SECOND EDITION

Mental Wellness in Adults with Down Syndrome

A Guide to Emotional and Behavioral Strengths and Challenges

Dennis McGuire, PhD & Brian Chicoine, MD

Mental Wellness
in Adults with Down Syndrome

A Guide to Emotional and Behavioral Strengths and Challenges

Second Edition

Dennis McGuire, Ph.D. & Brian Chicoine, M.D.

© 2006, 2021 by Dennis McGuire and Brian Chicoine
Second edition

The information contained in this book is not intended as a substitute for consultation with your healthcare providers. Although the authors, editor, and publisher made every attempt to ensure that the information in the book was up to date and accurate at the time of publication, recommended treatments and drug therapies may change as new medical or scientific information becomes available. Additionally, the authors, editor, and publisher are not responsible for errors or omissions or for consequences from application of this book. Any practice described in this book should be applied by the reader in close consultation with a qualified physician.

This is a copy of the second edition originally published by Woodbine House Publishing. After Woodbine House Publishing closed in 2022, the publishing rights reverted to the authors. The authors chose to make the book available to readers in this format at no cost.

Dedication

We dedicate this book to all who have taught us so much and supported our work, including our families, the team at the Advocate Medical Group Adult Down Syndrome Center, the team at the Denver Adult Down Syndrome Center, Advocate Aurora Health, the Global Down Syndrome Foundation, Advocate Lutheran General Hospital Family Medicine Faculty and Residents, the Woodbine House Publishing team, the generous philanthropic donors to the centers, our colleagues in the Down Syndrome Medical Interest Group, the National Association for Down Syndrome and other Down syndrome organizations, the staff of the agencies that serve people with Down syndrome, and the families of people with Down syndrome.

Most especially, we dedicate this book to people with Down syndrome, who have been our true guides and teachers for thirty years.

Table of Contents

	Introduction	6
CHAPTER 1	Mental Health Assessment	13
CHAPTER 2	Assessing the Physical Health/Mental Health Connection	22
CHAPTER 3	Family and Community Support	48
CHAPTER 4	What Is Normal?	66
CHAPTER 5	Concrete Thinking and Behaving	86
CHAPTER 6	Visual Memory and Cues	97
CHAPTER 7	Communication	111
CHAPTER 8	Self-Esteem and Self-Image	136
CHAPTER 9	Self-Talk Revisited	174
CHAPTER 10	The Groove and Flexibility	193
CHAPTER 11	Life-Span Issues	218
CHAPTER 12	Sensory Processing Differences and Disorders	242
CHAPTER 13	Social Skills	264
CHAPTER 14	Mental Illness and Its Precipitants	280
CHAPTER 15	Assessment of Mental Illness	304
CHAPTER 16	Treatment Approaches for Mental Illness	311
CHAPTER 17	Mood Disorders	367
CHAPTER 18	Anxiety Disorders	395

CHAPTER 19 Obsessive-Compulsive Disorders 414
CHAPTER 20 Psychotic Disorders 437
CHAPTER 21 Eating Refusal 443
CHAPTER 22 Challenging Behavior 454
CHAPTER 23 Self-Injurious Behavior 487
CHAPTER 24 Tics, Tourette Syndrome, and Stereotypies 497
CHAPTER 25 Autism 518
CHAPTER 26 Alzheimer's Disease and Decline in Skills 535
CHAPTER 27 Regression 561
References 578
About the Authors 588

Introduction

In the first edition of this book, the first few sentences of the introduction read: "As you hold this rather large volume in your hands, you probably have a number of questions. Perhaps the most natural one is, why is there so much to say about the subject? Is the mental health of adolescents and adults with Down syndrome really such a complex matter that we need an encyclopedic guide? You might also wonder: Are mental health problems inevitable in all people with Down syndrome? Will it be worth my while to read this book, even if the individual(s) with Down syndrome I know seem to be perfectly healthy?"

Some of the responses to those questions remain the same in the second edition. We would like to reassure you at the outset that mental health problems are not inevitable in people with Down syndrome. That is one reason we wrote this book—to point out ways that parents, adult siblings, teachers, paid caregivers, and others can effectively promote and maintain mental wellness in adolescents and adults with Down syndrome. But we also wrote this book because there are some biological differences as well as common environmental stresses that can make people with Down syndrome more susceptible to mood, emotional, and other mental health problems. We hoped to make it easier for other practitioners to recognize these problems and pursue treatments that will help the person regain his normal abilities and outlook on life. We also hoped to clarify that there are some common characteristics of Down syndrome that can be mistaken for evidence of mental illness but are nothing more than harmless quirks or useful coping strategies.

You may note that this edition is even longer than the first and there are many more references. Since the first edition was published in 2006, there has been a great deal more research regarding the mental health of people with Down syndrome. We have included much of that information. However, as in the first edition, we have also relied heavily on our experience of caring for adolescents and adults with Down syndrome . . . and we each have fourteen more years of experience to bring to this edition.

We have updated the entire book, added some new chapters, including one on regression, and invited Dr. Katie Frank to write a chapter on sensory issues. The importance of communication, concrete thinking, and visual memory is highlighted in this edition so that each of these topics now has its own chapter. The chapter on Alzheimer's disease has also undergone extensive revision in light of a plethora of

research studies and our vast experience treating individuals with Down syndrome and Alzheimer's disease since the first edition was published.

We do want to make it clear, however, that this is educational material. This book is not meant to take the place of the relationship between a person with Down syndrome and his health care provider. We do hope you can use it to improve your understanding of how you can enhance health promotion and address conditions of health impairment.

Our goal is not just to educate parents and support people about these issues. We would also like to help adults with Down syndrome actively participate in the process of achieving good health. Health is more than the absence of disease. It is a sense of physical, mental, and spiritual well-being. It is a process that involves health promotion, health screening, and early intervention for health problems. It is reasonable for adults with Down syndrome and their families to assume that they can (and should) be part of this process that leads to improved health. While there are clearly physical and psychological problems that affect the health of some individuals with Down syndrome, good health for most and improved health for all are reasonable expectations.

Many families have described how health problems of people with Down syndrome were ignored because the problem was assumed to be (or "written off" as being) "just part of the Down syndrome." In fact, people with Down syndrome are susceptible to many of the usual health problems that occur in people without Down syndrome. In addition, there are illnesses and conditions that are more common in people with Down syndrome, and some health conditions present differently in people with Down syndrome. These problems may alter health and, therefore, cause a change in the status of the person. These changes are often not directly linked to the syndrome but to one of these conditions. While there is no proven treatment for Down syndrome itself, most often the additional health issues are diagnosable and treatable. Therefore, changes that are just assumed to be a manifestation of Down syndrome can lead to unnecessary impairment because of underdiagnosis or undertreatment of other conditions.

These ideas are true for both physical and mental health issues. The focus of this book is on mental health issues. Physical health issues are addressed as they relate to or affect mental health.

What Is Mental Health?

By “mental health,” we mean having the emotional well-being that allows us to cope with the activities and stresses of everyday life. Mental health is more than just diagnosing and treating mental illness. It is part of each person’s life: seeking to optimize our enjoyment, our sense of purpose, and our ability to participate in the activities of daily life. Mental health is a process. It can be optimized through mental health promotion strategies. These strategies are ideally part of daily living, as well as part of regular health care promotion with your health care provider.

To promote mental health, it is imperative to understand the continuum of normal behavior and mental health issues. A behavior may be healthy and serve a useful purpose, but if it becomes excessive or impairs function, it may move along the continuum into the “abnormal” range. While there are clear diagnostic guidelines for mental illness, there is certainly some degree of subjective interpretation of the symptoms. In addition, the environment in which the person lives, if supportive enough, may help prevent the behavior from becoming—or from being labeled—maladaptive. Mental health promotion strategies can help emphasize the positive aspects of behavior and keep the behavior on the healthy side of the continuum.

As mentioned previously, it is important to understand that change in behavior is often not “just the Down syndrome.” Conversely, it is also critical to understand that there are typical strengths and weaknesses and common characteristics of people with Down syndrome. To optimally promote mental health in a person with Down syndrome, it is necessary to understand and appreciate both of these opposing concepts.

Who We Are

We, the authors, are the cofounders of the Advocate Medical Group Adult Down Syndrome Center in Park Ridge, Illinois. Dr. Chicoine continues as the Medical Director of the center, and Dr. McGuire has left the center and is now a consultant for the Global Down Syndrome Foundation. Understanding the history of how we came to cofound the center should help you understand our perspective on mental health issues in adolescents and adults with Down syndrome.

In the late 1980s, parents in the Chicago metropolitan area were becoming increasingly frustrated by the lack of good quality medical and psychosocial care for their adult children with Down syndrome. Too often, these parents found that when

their children with Down syndrome developed changes in behavior, health care professionals would tell them it was "just the Down syndrome" and there was no treatment. Another common concern was that anytime a person with Down syndrome experienced a decline in cognitive function, a limited evaluation always seemed to lead to a diagnosis of Alzheimer's disease. Families often felt that their son or daughter was not receiving a thorough evaluation. They wanted to be able to bring the person with Down syndrome to a place where he or she would get a thorough evaluation from practitioners who understood the medical and psychosocial issues of people with Down syndrome.

Many of these concerned parents belonged to the National Association for Down Syndrome (NADS), the oldest parent group serving people with Down syndrome in the United States. In 1991, the staff and parents of the group went to the administration of Lutheran General Hospital (on whose campus the center sits). They requested that a clinic for adults with Down syndrome be developed. This clinic opened in 1992. Initially, patients were served two mornings a month. The original staff included a physician (the second author), PhD-trained social worker (the first author), and a certified medical assistant. Dr. McGuire had been working with NADS through a fellowship the group provided to the University of Illinois–Chicago, where Dr. McGuire was employed. The clinic was a natural extension of his work because he realized that medical conditions often were part of the cause of the psychosocial problems he was addressing. Dr. Chicoine had recently joined the faculty in the Lutheran General Department of Family Medicine residency program. He had previous experience in working with adults with intellectual disabilities and eagerly helped develop the clinic. A nutritionist and an audiologist were also available to see the patients.

Today the Adult Down Syndrome Center has grown and is now open full-time. It includes a staff of fourteen and has its own building of about 3,400 square feet. In addition to receiving care at the center, individuals with Down syndrome are served when they are inpatients at Advocate Lutheran General Hospital, in a local nursing home, at a variety of residential facilities, and in their homes, when needed.

To date, the Adult Down Syndrome Center has served over six thousand patients, ages twelve to eighty-three. The patients use the center in one of three ways: as their primary health care center; for an annual complete evaluation and regular follow-up for specific problems (most commonly for psychosocial issues); or for annual evaluations only. The patients who live farthest away are most commonly in the last group. Each year approximately 2,000 unique adolescents and adults with Down syndrome are served through approximately 7,500 patient encounters. We work

as a team in treating patients, particularly when there are mental health concerns. This approach allows us to address all the issues that promote health (mental and physical) as well as contribute to mental or physical health problems.

This book is a compilation of the information we have gathered through serving adolescents and adults with Down syndrome and their families and care providers. We view the Adult Down Syndrome Center as a repository of knowledge. Through listening to people with Down syndrome and their families and care providers, we have learned much of what is written on these pages. When we heard something from one person, we asked others whether that was true for them as well. Through this process, we have developed a greater understanding of mental health and mental illness in people with Down syndrome.

"Two Syndromes"

In the first edition, we shared the following thoughts on "Two Syndromes." The distinctions among the syndrome are even truer today, as the benefits of improved health care, education, recreational opportunities, physical activity, etc. have resulted in still greater improvements in cognition, skills, and participation in society:

> *A word on the concept of "Two Syndromes" is important. Some families that include children or young adults with Down syndrome have heard us speak or have read a piece of material from us and have commented that it doesn't seem to fit with their life experience. Certainly, some of this interpretation may be due to the variance in people with Down syndrome and the variance in families. Some of it, however, may be because it sometimes seems as if there are "Two Syndromes." The childhood experience of families with older sons and daughters was often very different from that experienced by families now. In the past, based on the information provided by health care and education professionals, families often had very low expectations for their son or daughter. Good health care was often unavailable to children with Down syndrome. School, social, recreational, and work opportunities were often limited or nonexistent.*
>
> *We now know that early intervention is very beneficial for children with Down syndrome (Anderson et al., 2003; Guralnick, 1998). The benefits of early intervention, as well as more inclusive and academically*

challenging school experiences, are being appreciated and enjoyed in childhood and early adulthood. As this generation of young people ages, it will be very interesting to see what the long-term benefits will be. Studies in the general population suggest that improved learning and educational opportunities may reduce cognitive decline and reduce the risk of Alzheimer's disease (Snowdon, 2001; Levenson, 1978). What will be the effect on people with Down syndrome? Historically, impaired verbal and communication skills have had a great impact on both the physical and mental health of people with Down syndrome. Presumably, the improved skills being seen in many young people who have received speech therapy from a young age will affect their health in adulthood. The improved ability to report and discuss their concerns and to participate in treatment should reduce illness as well as lessen the severity of problems.

The concept of "Two Syndromes" is theoretical at this point. However, it will be very interesting to watch the differences in adults with Down syndrome who have had very different life experiences. It is also important to understand that we are also seeing the beneficial effect of these positive experiences for our older patients. In other words, it is not too late even if these opportunities were not available at a younger age. Good health care, work opportunities, and social opportunities have been very positive experiences for our older patients as well.

A Word about the Case Stories

Throughout this book you will find many case stories about adolescents or adults with Down syndrome we have seen. In general, the case studies are a compilation of multiple people. We have changed names and other identifying information to protect identities, but the problems described and solutions are real.

In addition, note that we sometimes refer to adolescents and adults with Down syndrome simply as "people" or "adults" for simplicity's sake, and we make it clear when we are using the word "people" to refer to individuals who do not have Down syndrome.

In Conclusion

As you proceed through this book, we recommend keeping "Joe" in mind. Joe is both an example and a compilation of our healthy patients. Joe is twenty-nine years old and is healthy, physically and mentally. What has helped Joe be healthy? What are the experiences that Joe has had that contributed to health?

- Joe is accepted as an individual.
- He is given choices.
- Expectations are placed on him that are neither too low nor too high.
- Joe regularly exercises.
- His need for routine is supported, but flexibility is encouraged.
- Joe gets annual health evaluations and good health care at other times when needed.
- Communication skills were emphasized from a young age.
- Vocational training was part of his schooling.
- He has an enjoyable, stimulating job that allows him to use his strengths.
- He is part of a supportive community.
- He has opportunities to help others.
- He participates in his religious community.
- He has opportunities to be integrated in society (with people without disabilities) but also has opportunities to congregate with other people with disabilities.
- And Joe is heard. When he expresses concerns, people listen.

Joe has taken these opportunities, works very well in his job, and has an enjoyable social and family life. In short, Joe is healthy. Our hope is that by sharing the information in this book, we can help all adolescents and adults with Down syndrome be healthy like Joe.

Return to Table of Contents

Chapter 1

Mental Health Assessment

Joseph, age twenty-three, was excited to get a job in a grocery store in his community. He thought he would enjoy doing the assigned tasks and interacting with the customers. But after an observation experience at the store, he had some worries about how he should respond to some situations he noted. A former teacher helped him and his family use role play to practice the potentially challenging interactions he had witnessed. A few days into the job, one of those situations arose, and Joseph remembered what they had practiced and handled it very well.

The evaluation of a person with Down syndrome for mental health issues should not begin when there is a problem. Where physical health is concerned, it is well recognized that health promotion, disease prevention, and early assessment of disease are essential to optimize health and limit illness. Similarly, mental health promotion, prevention of mental illness, and early assessment of mental illness are essential parts of mental health care. Educating people with Down syndrome, their families, and care providers about ways to optimize mental health is an important part of the process.

Even if an adolescent or adult with Down syndrome does not seem to have any mental health problems, it is very useful to assess the aspects of his life that can promote mental health, or, conversely, increase the likelihood of mental illness. Understanding a person's support system, his connection to friends, the activities available to him, his community involvement, and other aspects of his life help develop a picture of the person and can help provide guidance in ways to optimize mental health.

We recommend that patients seen annually for a comprehensive mental health assessment starting in adolescence and throughout the adult years. In addition, as with Joseph, we encourage anticipating challenging social situations to help prepare the individual with Down syndrome to manage potentially difficult situations. Experience and research remind us that repeated stressful situations—or in some cases, even one situation that the person is not prepared for—can affect mental health.

Of course, if the adult with Down syndrome experiences changes in mental health or behavior, he should have a thorough assessment as soon as possible. A complete evaluation of his physical health status will also often provide a great deal of insight into the cause of the problem.

In *To Kill a Mockingbird*, Atticus Finch stated, "You never really understand a person until you climb into his skin and walk around in it." We recommend an assessment that is designed to do this as best as possible. While we will never be able to understand all that it means to be a person with Down syndrome, our assessment is designed to maximize our understanding. The goals of the assessment include:

- understanding a person's strengths and weaknesses,
- appreciating the positive and negative aspects of the environment where he lives,
- assessing the contribution of physical health challenges, and
- discerning how the person deals with stresses in life.

If an adult with Down syndrome has experienced a change in his mental health status or there is a behavioral concern, the assessment is similar. It is important to understand his strengths and the supportive aspects of the environment. Those will be important issues to emphasize in developing a health-promoting treatment plan. In addition, we focus particularly on the areas that may be contributing to the problem so that these can be understood and addressed.

While assessing these issues, we typically evaluate the following:

- the relationship between social health and mental health
- expressive language skills
- self-esteem
- self-talk
- cognitive issues, including processing speed, understanding of time, abstract thinking abilities, and the need for repetition
- range of emotions,
- the presence of factors that could precipitate mental illness,
- symptoms of mental illness (if any)
- physical health.

These areas are discussed in detail below.

Where to Get an Evaluation

Of course, it is not practical or possible for all adolescents and adults with Down syndrome to be evaluated at a center specifically for adults with Down syndrome. If you are seeking a mental health assessment elsewhere, it may be up to you to ensure that all the above areas are evaluated as described in this chapter. Ideally, you will be able to take the person with Down syndrome to a comprehensive Down syndrome clinic where the staff understand these issues and where professionals with different specialty areas work together to evaluate individuals with Down syndrome. Global Down Syndrome Foundation has a web page that lists clinics for people with Down syndrome: https://www.globaldownsyndrome.org/research-medical-care/medical-care-providers/.

If you are not able to visit a Down syndrome clinic, you may need to seek a series of assessments from a variety of professionals. You can then ask one of them to be the case manager and help you interpret what all the separate assessments mean. We have found that the combined efforts of the following professionals work very well:

- a medical doctor (family physician or internist), to rule out medical problems and to prescribe medications, if needed;
- a psychologist, social worker, or other professional qualified to assess social skill and support issues, as well as emotional/behavioral disturbances;
- a teacher, job coach, or counselor at a job site, who can provide valuable information about the work or school environment, the person's behavior before the problem arose, and the problems the person may be experiencing outside the home.

In addition, a psychiatrist or a neurologist may be consulted, depending on the need.

Areas Assessed

The Relationship between Social Health and Mental Health

> *For anyone to receive unbidden words of encouragement, this is the material a true gift is made of.*
>
> —Christopher deVinck, The Power of the Powerless

Generally, no one is able to achieve mental health completely independently. To have a sense of well-being, it usually requires a sense of connection to others. For instance, most of us need to feel that we are loved and accepted by others to feel good about ourselves. This is just as true for people with Down syndrome. Because adolescents and adults with Down syndrome are more dependent on others in most areas of life, their reliance on others to achieve mental health is not surprising. However, the important contribution that friends and family provide is often overlooked as an important aspect of health for people with Down syndrome.

Several years ago, a large state facility closed in Illinois. The adults with intellectual disabilities were moved to new, smaller residences. Some of the residents had very significant communication challenges. There was little obvious social interaction between these residents. To the observer's eye, they did not seem to rely on each other for social stimulation. When the residents were tracked over time, however, it was found that those who moved to a residence together with other people from the old facility did better than those who moved to separate facilities. This was true even for those who had very significant communication challenges. Even the survival rate was better for those who moved with their peers. Social interaction is part of our life-sustaining activity, even when it is not obvious to others (Heller, 1982).

People with Down syndrome need interaction with family, friends, peers, and others, just as other people do. The absence of any of these groups can be a significant problem. This can be true even if there never was any perceived interaction with the group, as discussed in the preceding paragraph.

Also important for mental health is meaningful participation in community life. Participating in activities, hobbies, and community events promotes a sense of well-being, boosts self-esteem, and helps a person develop and improve social skills. Physical activity, social events, travel, and opportunities to learn new ideas and interact with other people while doing these activities can all be beneficial. So too is employment in a job that the person with Down syndrome finds interesting and fulfilling. Chapter 3 describes in more detail why social connections are essential to the mental well-being of adults with Down syndrome.

If possible, we recommend that a mental health worker (e.g., social worker or psychologist) assess the individual's social health. We find it helpful when information is gathered from the person with Down syndrome and his family via a checklist of questions about family support, friendships, recreational opportunities, other support systems, the work or school environment, and other issues of daily life. If

appropriate, staff of a residential facility or day program, teachers, and other important people in the person's life can also be questioned.

Ideally, each staff person who interacts with the person with Down syndrome should be aware of his or her role in assessing the patient's social health. Often the patient or a family member reveals an important piece of information during a less formal part of the assessment. For example, as part of the "warming up" part of the interview, the physician asks, "What do you do for fun?" This part of the interview is less threatening, gives the person with Down syndrome an opportunity to "warm up" to the interview, and frequently uncovers important information. The receptionist, nurse, or others may also gather clues related to the person's social health and share the information with other staff to optimize data gathering.

Expressive Language Skills

Another area to assess is the person's expressive language skills—that is, his or her ability to communicate a message to others using spoken language, gestures, or an augmentative or alternative method. The ability or inability to express oneself has a significant impact on promotion of mental health and prevention of mental illness.

We do a basic, informal assessment of communication skills. We focus on articulation, intelligibility, and overall communication abilities. If additional assessment or treatment is needed, we refer the adult to a speech-language pathologist. We have found that 75 percent of our patients are understood by familiar others most of the time. But only 28 percent are understood most of the time by people who are unfamiliar with their speech. Clearly, intelligibility is important to assess.

In addition to understanding verbal skills, it is essential to do a basic assessment of the adult's ability to express feelings. We also question the family about the adult's ability to express feelings. In our experience, most people with Down syndrome are open and honest in expressing feelings nonverbally, even though many have difficulty expressing feelings verbally. Unfortunately, we find that many caregivers have difficulty interpreting the cause or source of the person's nonverbal expressions.

We have found that when the family or caregivers of a person with Down syndrome have difficulty understanding his nonverbal expressions, the person is more likely to have a mental illness diagnosis. In contrast, 78 percent of our patients without mental illness diagnoses had caregivers who reported that they could

understand the nonverbal expressions of the person with Down syndrome most of the time. For those with a mental illness diagnosis, only 26 percent had caregivers who stated they could understand the person's nonverbal expression of feelings most of the time.

The cause and effect are not clear from these data. In other words, the data do not prove that the caregiver's inability to understand nonverbal expressions leads to a higher incidence of mental illness. However, we believe that this is the case. Therefore, knowing how well a person with Down syndrome can express his feelings is essential to understanding mental health and assessing for mental illness risk. Chapter 7 provides further discussion of how this information is used to promote mental health.

Diagnosis of Mental Health Disorders in Adults with Down Syndrome

For the assessment of mental health symptoms and disorders in adolescents and adults with Down syndrome, we use The Diagnostic and Statistical Manual of Mental Disorders, fifth edition (DSM-5), and the Diagnostic Manual-Intellectual Disability (DM-ID). Modification of the criteria in the DSM-5 is usually necessary, however, because of the verbal expressive language and conceptual difficulties that may limit the person's ability to report his symptoms. We should note, though, that over time and for a variety of reasons (early intervention, speech therapy, school programs, improved health, and others) the authors have noted that many individuals with Down syndrome are better able and interested in participating in the health care interview.

Emphasis in our adapted criteria is on the observed changes in behavior rather than self-report. For example, the DSM-5 criteria for major depression include such observable behavioral changes as withdrawal, a loss of interest in things previously enjoyed, change in sleeping and eating habits, loss of energy and fatigue, etc. Parents or other caregivers usually have no difficulty observing and reporting on these symptoms.

However, DSM-5 criteria also include such self-reported symptoms as verbal expressions of sadness and feelings of guilt and worthlessness, which are less commonly expressed by people with Down syndrome. Although feelings of guilt and worthlessness have no behavioral corollary, we have found that most caregivers readily observe evidence of sadness in facial expressions and body language (slumping shoulders, etc.), as well as in our criteria of a "loss of spark, life, and vitality." These

observable features are described in DM-ID as well. What helps in this process is that family members are often very astute observers of the person with Down syndrome in their care. We feel confident that even without self-report, the observed changes in behavior do allow an accurate diagnosis of mental health disorders.

See chapter 15 for further discussion of assessment of mental illness.

Additional Areas of Assessment

The mental health assessment also includes evaluations of several other important areas related to social and emotional functioning. These areas are assessed through questions in the medical exam and the semi-structured psychosocial evaluation, and more informally by talking with the adult with Down syndrome and his parents or caregivers. These areas include

- memory (discussed in chapters 6),
- issues of self-esteem (chapter 8),
- self-talk (chapter 9),
- the tendency toward repetition (chapter 10),
- processing speed (chapter 4),
- the understanding of time (chapter 4),
- the ability to use abstract thinking (chapters 4 and 5),
- the difference or similarity between chronological and developmental age (chapter 4),
- the range of emotions (chapter 4),
- lifespan concerns, such as issues of adolescence and geriatrics (chapter 11),
- factors that may precipitate mental illness (chapter 14), and
- the assessment of mental health disorders (Section 3).

Physical Health

A mental health assessment is incomplete if physical health is not also assessed. For all people, there is a great deal of interaction between physical and mental health. For people with Down syndrome, there appears to be more interaction and thus a greater need to assess and understand physical health problems.

Physical health problems can directly affect mental health. Some physical problems can include mental illness symptoms as a part of the disease. For example,

depression can be a manifestation of hypothyroidism. Physical problems can also indirectly contribute to mental illness. For example, prolonged pain or chronic illness can lead to depression. These are aspects of physical health that are well recognized in people with or without Down syndrome.

Another reason it is important to understand the interaction between physical health and mental health is that people with Down syndrome often have expressive language difficulties. People who have difficulty verbally expressing their physical discomfort tend to be more likely to express it with their behavior or through or a mental health problem. Limited expressive language ability seems to play a significant role in the interaction between physical and mental health issues for people with Down syndrome.

A thorough history and physical exam is part of the mental health assessment. In addition, if there are mental health or behavioral problems, additional testing is often necessary. This may include blood tests, x-rays, electroencephalograms (EEGs), and others. Often more testing is required for a person with Down syndrome than for someone without Down syndrome. If the person is unable to provide an accurate history or tell us if he is having symptoms of physical disease, additional testing is needed to assess for a physical cause for the behavioral change. Further information on the interaction between physical and mental health is provided in chapter 2.

Assessment Results

For many adults with Down syndrome, this will be the end of the assessment. They are doing well. We recommend that the person with Down syndrome, the parents, or whoever requested the assessment receive a written report of the results and an opportunity to discuss them. Areas of strength should be emphasized and other ways that mental health can be optimized discussed. Many times, parents or other caregivers are relieved to learn that the behavior being discussed is common in people with Down syndrome and should be viewed as a characteristic rather than evidence of a mental illness. Just knowing that the behavior is not a sign of mental illness can be therapeutic in and of itself because it can calm family or caregiver anxiety, reduce unnecessary redirection of the behavior, and prevent efforts to change the behavior from damaging the individual's self-esteem. Treatment recommendations, if any, should be thoroughly explained and implemented, or appropriate referral(s) should be made. If it appears that the adult with Down

syndrome may have a mental illness, further assessment may be recommended, as discussed in Section 3.

Conclusion

The assessment of the mental health of a person with Down syndrome ideally begins before there is a problem. It is important to understand the individual's strengths and weaknesses; to assess the environment, including the social contacts and support of family and friends; and to evaluate the link between physical and mental health. This information can be used to promote mental health as well as to better understand mental illness and the appropriate treatment. Additional issues to consider in evaluating a person with Down syndrome are addressed throughout the rest of the book.

Return to Table of Contents

Chapter 2

Assessing the Physical Health/Mental Health Connection

George's parents brought him to the adult clinic because they were concerned about his depressed mood, loss of skills, and hallucinatory behavior. During the assessment, we found that George, age thirty-six, snored and the family was noting pauses in his breathing. Before trying any psychotropic medications (medications to treat psychiatric conditions), we referred George to the sleep lab at his local hospital. He was diagnosed with sleep apnea and was started on CPAP (continuous positive airway pressure with a mask and CPAP machine). With this treatment, George had a remarkable recovery of his previous functional level as well as his mood. His hallucinations also stopped. He did not require any psychotropic medications.

As discussed in the previous chapter, every thorough mental health assessment should include an assessment of physical health to ensure that physical health problems are not affecting mental well-being in any way. This is true whether or not the adult with Down syndrome is suspected of having any kind of mental illness. Discovering physical health problems early in their course may prevent them from causing mental health problems.

When evaluating an adult with Down syndrome for changes in mental health or behavioral problems, it is important to do more than put on your "psychiatry hat." We find that there is often an underlying physical condition that is causing or contributing to the mental illness or the behavioral change. Furthermore, treating the mental illness or the behavioral symptoms without attention to the physical health issues will generally lead to at least partial failure of the treatment of the mental illness or the behavioral symptoms.

It may be advisable to have the physical exam before the mental health assessment. This may be especially true if there are no experts in mental health of adults with Down syndrome in your community.

We sometimes find that a physical problem is the direct cause of a mental health problem and sometimes it is a contributing factor. However, in either case, as

time goes by, the problem often develops other "layers," and treatment of the physical problem alone is often not adequate. It is necessary to address the problem from all aspects: physical, psychological, and social. For example, consider an adult with Down syndrome who develops a medical problem that includes the symptom of depressed mood. Often the person withdraws and becomes less interested in participating in activities, work, etc. In addition, her mood change may lead to interpersonal conflicts. Therefore, when treating the problem, it is not enough to focus on any one of these areas and neglect the others. It is important to evaluate and treat for the underlying physical problem but also generally necessary to address the psychological and social aspects of the problem as well. (This is addressed in detail in the chapters in Section 3.)

Sandy is a young woman with Down syndrome who had a birth trauma to her left shoulder, resulting in reduced use of her arm. Through therapy, however, the function of her arm had greatly improved. On her first visit to our center, Sandy had a depressed mood, and her parents reported she did not want to go to work and was avoiding social interaction with people other than her immediate family. In reviewing her history, we found that shortly before these symptoms began, she had slipped on the ice and fallen on her left shoulder, resulting in further impairment of her arm's function. In Sandy's case, physical therapy for her shoulder turned out to be a major part of the treatment for her depressed mood. She was able to recover with physical therapy, emotional support from her family, and a gentle reintroduction into her previously active social life.

Some patients will require greater intervention for the psychological and social issues. Some will require medications for the psychological problem, in addition to therapy and other interventions. However, we have repeatedly found that providing that care without addressing the underlying or contributing medical problems will limit the overall success of treatment.

Areas to Be Assessed

Table 2-1 shows the tests and procedures that should be considered to identify the physical problems that most often contribute to mental health problems in adolescents and adults with Down syndrome. The next sections explain in more detail why these problems are important to rule out. While the table suggests guidelines for assessing physical conditions, it is important to understand that any medical problem can contribute to making any psychological problem more significant. Any physical problem that makes a person feel physically worse will likely increase the symptoms of the psychological condition.

Table 2-1. Important physical conditions to assess

Condition/Problem	**Possible Impact on Mental Health**	**Test or Procedure***
Pain	Depression, behavior change, aggression, anxiety	Interview adult w/DS and family/caregiver; thorough physical exam; additional procedures as indicated based on history and physical exam.
Hearing impairment	Anxiety, apparent loss of cognitive skills, depression, agitation, aggression	Hearing test from audiologist at least every 2 years, or more frequently if indicated by possible change in hearing
Vision impairment	Anxiety, depression, apparent loss of cognitive skills, depression, agitation	Complete vision exam at least every 2 years or more frequently if indicated by possible change in vision
Seizures	Aggression, depression, apparent loss of cognitive skills	EEG and imaging of brain (CT scan or MRI)
Cervical subluxation	Loss of skills (particularly decreased ambulation	Thorough neurological exam (as part of physical exam);

	skills; loss of muscle function; incontinence), anxiety, agitation, depression, loss of interest in activities	lateral cervical spine x-rays in flexion, extension, and neutral; CT scan and/or MRI of cervical spine
Urinary tract problems (includes urinary tract infections and difficulty/inability to empty the urinary bladder)	Development of incontinence, urinary frequency, agitation, anxiety	Urinalysis (urine test) and possibly urine culture; ultrasound of the bladder and kidneys (a pre- and post-void ultrasound or a bladder scan is helpful to assess for problems with emptying the bladder)
Arthritis	Agitation; depression; apparent loss of skills	Physical exam; x-rays
Diabetes	Apparent loss of skills; urinary incontinence, increased urinary frequency and/or drinking of fluids; agitation; depression	Blood sugar; consider Hemoglobin A1C (further testing indicated if blood sugar suggests diabetes)
Dental concerns	Agitation, poor eating, depression, aggressive behavior	Thorough dental exam; dental x-rays as indicated
Hypothyroidism (underactive thyroid)	Depression; loss of cognitive skills; appetite change	Blood test for TSH and free T4
Hyperthyroidism	Anxiety; hyperactivity; depression; loss of cognitive skills	Blood test for TSH and freeT4 (and possibly free T3)

Sleep apnea and other sleep difficulties	Depression, loss of cognitive skills, agitation, psychoses	Observe sleep and keep a sleep log (although easy to miss sleep apnea with observation alone); formal sleep study in a sleep lab or at home
Gastrointestinal problems	Loss of appetite, depression, agitation, anxiety	Stool test for blood; blood tests for anemia (CBC), celiac disease, liver disease, gall bladder disease; x-rays, ultrasounds, CT scans, and endoscopy as indicated by history, physical, and other tests
Medication side effects	Can contribute to essentially any behavioral or psychological change	Careful history to look for potential link to medication; possible trial off the medication

* It is assumed that a complete history and thorough physical exam is performed for each of the presenting problems. Although areas of focus for the history and physical are specified here, additional testing may be indicated.

Pain

Pain is one aspect of physical conditions that can affect mental health. Many illnesses and injuries can cause pain. Common causes of pain in people with Down syndrome include the following: dental problems, gastroesophageal reflux, bladder problems (especially the inability to empty the bladder), ear infections, gastrointestinal discomfort secondary to celiac disease or constipation, and arthritis or subluxation of the joints (especially in the cervical spine). Pain can also be caused by emotional distress, but this section focuses on pain caused by physical health problems or injuries.

We are often asked, “Do people with Down syndrome have a reduced ability to perceive pain (an increased pain tolerance; delayed or less precise pain perception)

or do they have a reduced ability to communicate the pain so we only think they aren't experiencing the pain?" The answer appears to be "both and also delayed pain responses." Families often tell us that their son or daughter (sister, brother) has an increased pain tolerance. "He never complains. Even when he had the broken arm, he complained of very little pain."

Many individuals with Down syndrome seem to have reduced and delayed pain responses. They are not insensitive to pain, but their expression is delayed and less precise. It is hard to tell if they have a decreased ability to sense or localize the pain or a higher pain tolerance, or if the reaction to the pain is delayed. Impaired communication can also make it difficult to assess. It is important to understand differences in pain responses as a healthcare provider and family members caring for an individual with Down syndrome.

A study of specially bred mice provides supporting evidence for these observations. There are "mouse models" for Down syndrome. Mice with trisomy 16 (an extra sixteenth chromosome) have many similar health characteristics to people with trisomy 21 (Down syndrome). One study compared the response to pain of mice with trisomy 16 to mice with the normal number of chromosomes (for a mouse). The mice with trisomy 16 showed a significantly reduced response to painful stimuli. The authors concluded that the mice with trisomy 16 had a reduced ability to perceive pain or process pain (Martinez-Cue et al., 1999). While you might think that a reduced ability to perceive pain has some good points, there can also be important negative aspects. Having an increased pain tolerance or impaired processing of pain reduces a person's drive to escape a painful situation and avoid further contact with the painful stimuli. It also reduces the drive to seek help or treatment for the pain. We have had patients whose sole complaint was that they were "passing out" and found them to have a hemoglobin (blood count) of 4 or 5 (one-third of normal) due to a bleeding ulcer of the stomach. The situation became life threatening because their altered pain tolerance allowed them to avoid seeking help for the condition earlier.

Sometimes it is clear that people with Down syndrome are perceiving pain but have a reduced ability or desire to communicate the pain. Some patients avoid communicating painful conditions because they know from previous experience that if they tell someone, they may have to have uncomfortable or unpleasant tests or evaluations. Other patients try to communicate their discomfort but in ways that others have difficulty understanding. Particularly when someone has limited communication skills, she may communicate pain through behavior, as in the example below:

When we first began evaluating Patrick for depression, one of his symptoms was striking his head repeatedly. A CT scan of his head showed a chronic sinus infection in the area where he was striking his head. The depressive symptoms were improving with antidepressant medications, but the head-striking symptoms did not improve until the sinus infection was treated. The pain he was experiencing was contributing to his symptoms, and both the mental health and physical health issues needed to be treated to improve his condition.

Persistent pain can also have a negative effect if the person is unable to communicate the problem. Chronic pain can lead to depression and agitated behavior and exacerbate other mental health problems. There can be a great deal of interplay between chronic pain and depression, with each one contributing to or worsening the other. Therefore, addressing both is an important part of treating either.

Keys to Recognizing Painful Episodes

Watch for subtle signs. Grimacing, pointing to a body part, using a spoken phrase that is not typically uttered, sweating for no apparent reason, changes in appetite, and holding a limb differently can all be signs to note. There are probably many more that you have noticed as well.

Watch for behavioral changes. Pain can often be expressed by a behavioral change, especially if a person with Down syndrome has limited verbal or nonverbal communication skills. Behavioral changes could include less activity, more activity, seeking more attention, seeking less attention, a sad appearance, anger, emotional lability (sudden and frequent changes of emotions), reduced emotion, and many others. It is important to consider any behavioral change as a means of communication for a potential underlying physical problem.

Consider altered pain perception, reduced expression of pain, or less precise reporting. Remember that people with Down syndrome may have a reduced ability to perceive pain. Keep an eye on someone who has what appears to be a minor complaint. If the pain persists longer than expected or there are other symptoms that suggest something more serious (despite the person having little complaint), it could be time to for further evaluation.

Vision

Impaired vision also affects mental health. Declining vision can be a scary problem for anyone. Having a reduced cognitive ability to understand the decline can make it even more frightening. In addition, most people with worsening vision try to compensate by using their intellect and by increasing the use of their other senses. However, with an intellectual disability, there is "less to fall back on." Also, as noted below, hearing problems are more common in people with Down syndrome, limiting the ability to compensate. Therefore, vision loss can be even more traumatic for a person with Down syndrome.

Adults with Down syndrome have the usual vision problems, including nearsightedness, farsightedness, astigmatism, and glaucoma. There are also some problems that are more common. For example, cataracts occur at a younger age in adults with Down syndrome. In addition, many adults with Down syndrome have had strabismus (crossing of the eyes) from a young age. Strabismus can significantly contribute to impaired depth perception. Furthermore, many people with Down syndrome appear to have depth perception problems even if they don't have strabismus (perhaps due to a reduced ability of the vision centers in the brain to process the information). Depth perception impairment plays a role in mental health issues. People with poor depth perception may have more concerns about crossing from one surface to another or walking in places where there are different levels. For example, we have frequently heard that when people with Down syndrome are depressed and feeling more fearful, they are afraid to go on escalators or to walk on the second level of a mall where there is a glass wall that overlooks the opening to the lower level.

Clearly, possible changes in vision are important to consider when assessing mood changes, anxiety, an apparent loss of cognitive skills, and other changes. The loss of vision can be frightening and confusing.

Sara developed a progressive, uncorrectable loss of vision in her midthirties. She became aggressive and engaged in repetitive behavior consistent with obsessive-compulsive disorder (OCD). She also developed an increased tactile defensiveness (she was very frightened when touched). Treatment included prescribing medications to help with her

aggressiveness and compulsive symptoms and enlisting the help of experts on blindness to help her negotiate her environment and reduce the fear she was experiencing. Sara also received "desensitization therapy" from an occupational therapist to decrease her tactile defensiveness. In addition, the staff at her group home learned to give her advance warning before touching her or before asking her to move on to another task or activity. Reducing sudden, unexpected transitions to new activities made life less frightening for her.

Hearing

A decline in hearing can also be an emotionally difficult situation. Impaired hearing has a profound impact on our ability to communicate with others. In addition, hearing loss deprives us of a wonderful warning system that lets us know that someone or something is approaching or is nearby. If we don't hear people coming, their sudden, unexpected arrival can be anxiety provoking. Interaction with the world can be more frightening under these circumstances. Furthermore, daily joyful occurrences can be missed because good hearing helps let us know when opportunities are available and participate more completely. In addition, as with impaired vision, the person with Down syndrome probably has a reduced ability to compensate for this loss.

High frequency hearing loss (hearing of high-pitched sounds) is more common in people with Down syndrome. It can occur even in individuals who previously had normal hearing. One aspect of this loss is the reduced ability to distinguish different consonant sounds, making it harder to discriminate what is being said. Sometimes the person may seem to hear speech but not be able to discriminate the words. She may then do something different from what is asked, thinking that she is following the directions. In other words, what may appear to be defiance, oppositional behavior, or a decline in intellectual skills may actually be impaired hearing. Hearing assessment and hearing aids may be a significant part of the treatment of some mental health and behavioral problems.

Many children with Down syndrome experience recurrent middle ear infections and temporary loss of hearing related to fluid buildup in the middle ear. This can also occur in adults but is much less common than in children. However, when it does occur, it can cause temporary hearing loss and significant problems.

Cerumen impaction (earwax blocking the ear canal) is more common in people with Down syndrome. Removing wax from someone's ears may not typically be a part of treating psychological or behavior problems, but if earwax is contributing to reduced communication, it may be an important part of the treatment. Sometimes this relatively simple cause for hearing loss can lead to misunderstandings and frustration on the part of the person with Down syndrome. We have seen this situation leave adults with Down syndrome confused or escalate into more significant behavioral problems. It is gratifying when a person is brought in for evaluation for possible Alzheimer's disease and the problem is diagnosed as cerumen impaction and the person is "cured of Alzheimer's disease" by removing the earwax.

Seizures

Seizure disorders are more common in people with Down syndrome. The new onset of seizures has two peaks in people with Down syndrome. The first peak occurs in the first two years of life, and the second in adulthood. As discussed in chapter 26, sometimes seizures that start in adulthood are associated with Alzheimer's disease.

Particularly if seizures are poorly controlled, they can lead to episodes of confusion, recurrent injuries, and a sense of poor health and/or fear and frustration. Sometimes the fear becomes emotionally paralyzing and prompts the person to limit her activities. For some people, the unpredictability and randomness of the occurrences of seizures can cause significant psychological stress.

In addition, undiagnosed seizures can sometimes be mistaken for behavioral problems. Looking for other symptoms or clues during such a "behavioral episode" can lead to a more accurate diagnosis. Abnormal movements of the extremities or eyes, temporary loss of body control or consciousness, a period of fatigue or confusion after the episode, and other symptoms may suggest seizures. Some families have provided very helpful information by videotaping an episode. A careful neurological exam during the physical exam is recommended if seizures are suspected. An EEG, imaging studies (CT scan or MRI of the brain), and a referral to a neurologist may also be indicated.

Cervical Subluxation

Atlantoaxial instability, the slipping of the first vertebrae in the neck away from the second vertebrae, is more common in people with Down syndrome. There

are seven vertebrae in the neck, and slippage (cervical subluxation) can occur at any of these other vertebrae as well. It can also occur at other locations in the spine, but the cervical spine, or neck, is the most common area. Slippage can cause discomfort. When the slippage is great enough, the vertebrae can put pressure on the spinal cord and cause neurological changes, weakness of the arms and/or legs, bowel and/or bladder incontinence, and impaired gait (walking).The physician will also find increased reflexes (a more brisk movement) in response to the reflex hammer. In addition, there is often a significant emotional component to this problem since the fear of the discomfort and of the changes occurring in the person's nervous system can be quite disturbing.

A person with Down syndrome can develop cervical subluxation at any time. We have seen a number of patients who did not have the problem earlier in life develop it as they reached adolescence or adulthood. Significant trauma to the neck could cause this problem suddenly, or it may occur more gradually. Most of the time it seems to be related to aging and the degeneration of joints and tends to be more gradual. Osteoarthritis does seem to occur at a younger age in people with Down syndrome, and this can be a particular problem in the cervical spine. This can also cause pressure on the spinal cord by narrowing the spinal canal even if atlantoaxial instability is not present. It is important for the practitioner to regularly ask about symptoms that could be related to cervical subluxation and spinal cord compression and also perform a neurological exam for muscle strength and reflexes.

A young man with Down syndrome was evaluated for a global decline in his cognitive skills, as well as bowel and bladder incontinence and unsteady gait. As discussed in chapter 26, these are some of the symptoms seen in Alzheimer's disease, so his family was quite concerned. Our evaluation revealed that he was depressed and that he had increased reflexes. X-rays showed subluxation (slippage) of the third cervical vertebrae away from the fourth. Antidepressants markedly improved his mood and enabled him to regain his former cognitive abilities. He also underwent surgery to stabilize his neck and to eliminate the pressure of the vertebrae on his spinal cord. With the assistance of physical therapy, supportive counseling, and the antidepressant, he resumed his previous level of function and daily activities.

Arthritis

One of the medical conditions seen more frequently and at a younger age in people with Down syndrome is osteoarthritis (the arthritis generally associated with aging.) The joints degenerate, cause discomfort, and eventually become less mobile and less functional. This can cause "slowing down physically." Pain-related issues are similar to those discussed above. However, many of our patients with arthritis don't complain about arthritic pain; instead, they experience a change in function or lack of interest in activities due to dysfunction of the arthritis joint. Asking questions about joint discomfort or changes in mobility is part of the assessment for arthritis. The physical exam should include assessment of joint structure, signs of inflammation, and joint mobility. X-rays of the joints may also be needed.

Jean's story provides a good illustration of the issues of aging and osteoarthritis. When she was forty-six, she was brought to us for an evaluation because of "behavior problems." The problem started when she would walk into the center of her workplace, stop, and urinate on the floor. The staff claimed, "She is being defiant because she doesn't like to work." We discovered that Jean was actually doing a good job at work most of the time. She sat at her station and had a good production level. However, she was "frequently absent from her station for prolonged periods of time."

Our assessment found that, with age, Jean had developed a reduced bladder capacity and was also walking more slowly and cautiously. Unfortunately, her jobsite was in a building about the size of half a football field and her workstation was on the opposite end of the building from the bathrooms. Her frequent absences from her workstation were due to urinary frequency, and her "defiant" behavior was caused by an inability to get to the toilet in time. Her reduced walking speed secondary to her arthritis, the distance to the bathroom, and her reduced bladder capacity all made it impossible for her to make it to the bathroom in time. Jean's treatment included moving her workstation closer to the bathroom, using medications and physical therapy to reduce the discomfort and immobility caused by her arthritis, assessing and treating her bladder condition, and providing emotional support for the physical

changes she was experiencing. No psychiatric medications or other behavioral treatment were necessary.

Urinary Tract and Bladder Problems

Bladder problems can cause emotional issues, as well. Decreased muscle tone in the bladder, which appears to be more common in people with Down syndrome, can cause retention of urine and difficulty voiding. A large, distended bladder can, in turn, cause discomfort and overflow, leading to incontinence. A number of our patients have developed agitated behavior in response to this discomfort. Often, agitated behavior is treated with antidepressant or antipsychotic medications, many of which have the unintentional side effect of relaxing the bladder muscle even more and even further reducing the ability to empty the bladder. This can lead to even more agitation. Incomplete emptying of the bladder can also lead to increased urinary tract infections, which can be uncomfortable and result in behavioral changes.

If an adult with Down syndrome has a change in behavior, particularly if it is accompanied by any change in urination, we recommend performing a urinalysis. An ultrasound of the bladder pre- and post-void or a bladder scan may also be necessary to assess whether the person is emptying her bladder normally.

Diabetes Mellitus

Type 1 diabetes mellitus (insulin-requiring, previously called juvenile diabetes) is more common in people with Down syndrome. The incidence of Type 2 diabetes mellitus (previously called adult-onset DM and often treated with pills) is less clear but may be more common in people with Down syndrome. The symptoms of both types include polydipsia (drinking more fluids), polyuria (urinating more frequently), polyphagia (eating increased amounts of food), weight loss, and fatigue. People with new onset Type 1 also often have other symptoms and get sicker more rapidly.

The onset of diabetes can be subtle, particularly with type 2 and especially in individual who have difficulty perceiving or reporting the changes. The onset of Type 1 diabetes tends to be more rapid and cause a more acute and serious physical illness, but sometimes it may be somewhat slow and subtle initially. The sense of feeling unwell associated with diabetes mellitus can contribute to behavioral changes or a depressed mood. In addition, urinary incontinence may occur because of the need to

urinate so often, and this can be misinterpreted as a behavioral issue. Needing to drink or eat larger quantities can also be mistaken for a behavioral issue in someone with untreated diabetes mellitus

In addition, even after the diagnosis has been made, if the person's blood sugar goes too high or too low, behavioral changes may be seen. Particular attention must be given to preventing very low blood sugars. Very low blood sugars can result in significant behavioral changes, but, more importantly, can be a life-threatening situation.

It is anticipated that new health care guidelines will recommend screening all adults with Down syndrome periodically for diabetes mellitus, and more frequently for anyone who is obese, has a family history, or a previous borderline blood sugar. In addition, if, an adult with Down syndrome has a behavioral change or the new onset of psychological symptoms, a blood sugar test is usually indicated. Even if it that level is normal, some of the medications used for psychiatric problems can cause an increased blood sugar; therefore, the blood sugar should be known before prescribing these medications (see chapter 16 on medications).

Dental Concerns

Many adults with Down syndrome have significant tooth decay. Some of this is related to inadequate tooth brushing and flossing and some to misalignment of teeth (which is more common in people with Down syndrome). There may be genetic factors as well. In addition, gum disease is more common in people with Down syndrome. Pain, loose teeth, difficulty chewing, and other problems can occur because of the dental disease. Agitated behavior in response to dental pain is a common problem. Regular preventative dental care is very important. A good dental exam is imperative when there are concerning changes in behavior or emotional distress.

Underactive or Overactive Thyroid

When the thyroid gland is not producing adequate thyroid hormone, the condition is called hypothyroidism (underactive thyroid). Nearly 40 percent of people with Down syndrome have hypothyroidism. Some developed hypothyroidism when they are children, but many don't develop the condition until adolescence or adulthood. Hypothyroidism can cause numerous physical symptoms including constipation, dry skin, and weakness. When discussing mental health issues, it is important to note that

hypothyroidism can also cause lethargy, depression, and even a decline in skills or dementia. The symptoms may be subtle. Therefore, because of the frequency of the problem and the subtlety of the symptoms, an annual blood test is recommended for people with Down syndrome (Cohen, 1999).

Often, treating the hypothyroidism is not the whole treatment for the problem but is a necessary part of the treatment. Without treating the hypothyroidism, only limited improvement of mental health problems is generally possible. Treating the psychological problem directly may also be necessary. For example, sometimes a depressed person who is found to have hypothyroidism will respond to hypothyroidism treatment and not require further medication. However, sometimes the person will need also an antidepressant. Furthermore, even when someone responds to treatment with thyroid medication alone, if the dose needs adjusting, the symptoms may recur. It is important to do regular blood testing to confirm that the dose of thyroid medication is appropriate.

Hyperthyroidism (overactive thyroid), is also more common in people with Down syndrome, although nowhere near as common as hypothyroidism. Hyperthyroidism can cause weight loss, hyperactivity, anxiety, fatigue, loss of skills, and personality changes. Blood tests for thyroid function are an important part of any assessment for emotional or behavioral changes.

Gastrointestinal Problems

Gastrointestinal problems can be easily overlooked because many symptoms are not visible to an observer, and adults with Down syndrome may not report experiencing them. These problems can cause significant distress without any external signs or findings. Peptic ulcer disease, gastroesophageal reflux disease or GERD (heartburn), constipation, and other problems can cause discomfort that an adult with Down syndrome may express in a behavioral fashion if she cannot verbalize her discomfort.

During the physical exam, the practitioner should inquire about gastrointestinal symptoms such as diarrhea, constipation, and heartburn (making sure the adult with Down syndrome understands the terminology). If the patient can't clearly describe symptoms, it is sometimes necessary to treat her for a condition and see if it helps. One reason is that doing diagnostic testing to search for potential underlying gastrointestinal problems may be riskier than treating for the condition for a short time. For example, there may be signs that physical discomfort precedes a person's

behavioral change. If there is any history that suggests the possibility of GERD, treatment with medications to reduce acid in the stomach may be an appropriate adjunct therapy to treating the behavioral changes. This treatment might be tried initially instead of making a definite diagnosis by doing an upper endoscopy to look into the stomach. This test may be particularly difficult for some people with Down syndrome to comply with.

When assessing one young man with limited verbal skills for agitated behavior, one of the pieces of the history we obtained was that he was "fanning his chest" as if he were trying to cool off. Part of his successful treatment was using famotidine, a medication to reduce the acid in his stomach to treat his apparent heartburn.

Celiac Disease

Celiac disease is a gastrointestinal condition that is more common in people with Down syndrome. People with this condition are overly sensitivity to gluten—a protein in wheat, barley, and rye. These sensitivity leads to inflammation in the small intestine, which causes destruction of the villi, the small projections in the intestine that help us absorb food. This damage can cause a reduced ability to absorb food, vitamins, and minerals. Celiac disease can be associated with diarrhea, poor weight gain, weight loss, overeating, and fatigue. Some people may have constipation instead of diarrhea, probably because of large bowel movements secondary to undigested food. Many people with untreated celiac disease do not feel well. This can lead to irritability and contribute to a variety of behavioral, emotional, and psychological problems.

Celiac disease can begin at any age, so even if the person with Down syndrome has already been tested for celiac disease, it may be worth repeating the test. Blood testing is used as the initial assessment for celiac disease. The blood tests include the following: anti-tissue transglutaminase antibody, IgA and IgG (or anti-endomysial antibody), total IgA, anti-gliadin IgG, and anti-gliadin IgA. If the blood tests suggest celiac disease, the diagnosis is made by doing a small bowel biopsy, which is collected by doing an endoscopy. Some families choose to eliminate gluten-containing foods from the diet based on the blood tests alone to avoid the endoscopy. However, the diagnosis cannot be definitively made without the biopsy. Furthermore, in a small study we did, we found that the blood test is not as accurate in people with Down syndrome (Chicoine et al., 2014), so the biopsy is the best way to make the definitive diagnosis.

Another test that can be done to assist in diagnosis is HLA testing. For celiac disease, this genetic testing is used to check for the presence of HLA DQ2 and DQ8. People with these genes are more at risk of developing celiac disease. It appears that one must have one or both of these genes to develop celiac disease. Therefore, if someone does not have either gene, she does not appear to have the genetic predisposition to develop celiac disease. At this time, this genetic test is generally not part of testing for celiac; it is expensive and often not covered by insurance.

Treatment of celiac disease requires eliminating all foods that contain gluten from the diet. In people known to have celiac disease, changes in mood or increased irritation can be signs that they have strayed from the gluten-free diet.

Vitamin B-12 Deficiency

Vitamin B-12 deficiency appears to be more common in people with Down syndrome. Sometimes this deficiency may be a result of celiac disease, and it may have an autoimmune cause in some people. Vitamin B12 deficiency can cause a variety of psychological and neurological symptoms, including poor appetite, numbness, difficulty with balance, confusion, memory loss, and dementia. As has been described with several other physical health problems, correcting the vitamin B12 deficiency may not resolve the whole psychological problem. However, without optimizing physical health and treating the vitamin B12 deficiency, complete recovery is less likely. At present there is no recommendation to routinely screen all people with Down syndrome for vitamin B12 deficiency. However, we recommend testing each person who has neurological or psychological changes.

Menstrual Problems

Menstrual difficulties can cause significant emotional changes as well. Most of our female patients manage their monthly cycle very appropriately. With the right training in hygiene, explanations that it is part of a normal bodily function, and support as needed, it becomes part of their routine. For some, however, menstruation continues to be a challenge and it can contribute to behavioral problems. When a woman with Down syndrome is experiencing menstrual problems, it is important to first consider whether there may be a significant problem such as premenstrual syndrome (PMS) or dysmenorrhea (painful periods).

Premenstrual syndrome or premenstrual dysphoric disorder (when the dominant symptoms are emotional) can be quite disturbing. Symptoms include depressed mood, mood swings, irritability, difficulty concentrating, fatigue, swelling, breast tenderness, headaches, and sleep disturbance. We recommend keeping a log of symptoms to see if the symptoms are present in the premenstrual phase (approximately seven to ten days before menstruation) and absent during the rest of the cycle. Treatment includes getting daily exercise; eating regular, balanced meals; avoiding smoking; getting regular sleep; using stress reduction techniques, and counseling. A low-salt, low-caffeine, and low-fat diet with frequent small meals of complex carbohydrates may also be beneficial. Vitamin B6, calcium, and vitamin E have also been used. Antidepressants and/or oral contraceptives can be helpful when the symptoms are more severe.

If the periods themselves cause discomfort (dysmenorrhea), anti-inflammatory drugs can be of particular benefit. Ibuprofen (Advil, Nupren) and other anti-inflammatory medications block a chemical pathway that leads to increased discomfort. Acetaminophen (Tylenol) can help with discomfort but doesn't block the pathway and for many women does not work as well. Since painful periods may contribute to behavioral changes during that time of the month, treatment of dysmenorrhea may be a significant piece of the treatment for behavioral changes. Some women also get very significant improvement with the use of birth control pills or other hormonal treatments.

Medications

Medications can also be a source of health problems. Side effects from medications can contribute directly to psychological issues, create a sense of ill health that leads to behavioral challenges, or cause pain or other health problems that lead to psychological or behavioral problems. A medication that was tolerated previously may be less tolerated with age. Also, sometimes when a new medication is added, it may interact with a medication previously tolerated and cause intolerability of the first medication. To determine whether medications are contributing to mental health problems, it's important to do a thorough assessment of all medications—reviewing when the symptoms began relative to starting on the medication, changing doses, or adding other medications or natural products.

Sleep Disorders

Inadequate sleep, particularly if it is a chronic problem, has a huge effect on a person's ability to function in her daily activities. It can lead to irritation, problems controlling emotions, loss of concentration, attention problems, and apparent loss of cognitive skills. These difficulties are clearly apparent in many of our patients with sleep difficulties.

Sleep difficulties are very common in people with Down syndrome of all ages and can include the following:

- sleep apnea,
- hypopnea,
- restless, fragmented sleep, and
- difficulties related to routines or sleep environment.

Sleep Apnea

Sleep apnea is a serious health problem that is much more common in people with Down syndrome of all ages. If not treated, it can lead to heart and lung damage and can affect multiple organ systems. It also contributes to behavioral or psychological problems.

In order to understand what sleep apnea is, you need to know what normal sleep is. Normal, uninterrupted sleep consists of a cyclic pattern alternating between rapid eye movement (REM) sleep and non-rapid eye movement (non-REM) sleep. REM sleep is also called dream sleep. During REM sleep, many physiological changes are observed. For instance, there is generalized muscle relaxation except in the diaphragm, irregular breathing, and less chin muscle activity. Consequently, with the onset of normal sleep, the pharynx (airway) narrows because of muscle relaxation, leading to added resistance to air movement through the airway. During normal sleep, this causes reduced air movement and a slight increase in carbon dioxide in the body.

Sleep apnea is defined as a complete cessation of breathing from any cause during sleep, resulting in decreased oxygen in the blood or increased carbon dioxide (a greater increase than would be seen in normal sleep). The pauses in breathing usually last ten to twenty seconds but can last as long as two minutes. In severe cases, more than 500 attacks of sleep apnea may occur during a night. In people with Down syndrome, apnea is most commonly caused by obstruction of the airway. The

person keeps trying to breathe, but the obstruction prevents movement of air into and out of the lungs. In children with Down syndrome, obstruction is frequently caused by large tonsils, adenoids, or tongue; small respiratory passages; or low muscle tone in the mouth and pharynx. The tonsils seem to play less of a role with advancing age, but smaller airways and low tone continue to be a problem. Obesity can increase the risk of sleep apnea, but even trim people with Down syndrome can have sleep apnea. If recent weight gain is a problem and there is a behavioral change, sleep apnea should be considered.

Common signs of sleep apnea include a long history of loud snoring combined with restless sleep, excessive daytime drowsiness, and early morning headaches (although headaches are not a common complaint in people with Down syndrome). The person may also have difficulty concentrating, depression, irritability, and personality changes. We have even seen evidence of psychotic behavior that improved after treating sleep apnea (as per the case vignette at the beginning of the chapter). During obstruction, the person may aspirate secretions into the lungs, causing a cough or aggravating asthma symptoms. Shortness of breath and fatigue increase as the disease progresses. Sleep apnea also contributes to increased gastroesophageal reflux (GERD/heartburn).

Due to the high incidence of sleep apnea in people with Down syndrome, a history and physical exam should always include questions designed to determine whether apnea might be present. The doctor should inquire about how much the person with Down syndrome snores, restless sleep, daytime drowsiness, awakening during the night, whether the person's lips ever turn blue during sleep, and other symptoms of sleep apnea. Parents or other caregivers might want to videotape the person sleeping and bring it to the exam if they suspect apnea. Unfortunately, as has been shown in children with Down syndrome, observation alone is not a real accurate piece of the diagnosis (Shott et al., 2006). If there is a possibility of apnea, the doctor should make a referral for a sleep study at a sleep disorders clinic or hospital. A sleep study involves attaching electrodes and other sensors to the body to measure breathing effort, the passage of air through the airways, brain waves, oxygen content of the blood, and the restfulness of sleep. The test generally requires staying overnight in a sleep lab but sometimes can be done in one's own home.

If sleep apnea is diagnosed, treatment depends on the severity of the disease. Here are some options:

- A sock with a tennis ball inside can be attached to the back of the pajama top to prevent the person from sleeping on her back (if apnea only occurs when the person sleeps on her back).
- The person can use CPAP or BIPAP, which are means of delivering pressure to the airway to keep it open throughout the breathing cycle (and requires the person to wear a mask over the mouth and/or nose when sleeping).
- Surgery such as widening of the airway or a tracheotomy may help. However, we have had very limited success and significant complications with this as a treatment and generally no longer recommend it.
- A newer treatment that is beginning to be used and studied more in people with Down syndrome is a hypoglossal nerve stimulator—a surgically implanted device that moves the tongue forward and acts on other airway muscles to open the airway with inspiration (when the person breathes in).

Hypopnea

Hypopnea is similar to sleep apnea. Instead of the airflow stopping during sleep, however, it is reduced due to narrowing or obstruction of the airway. In people with Down syndrome, sources of this obstruction are similar to those in sleep apnea. If the airflow is reduced enough, oxygen levels drop and carbon dioxide levels rise, resulting in many of the same symptoms that sleep apnea causes. Signs of hypopnea can include snoring, labored breathing, odd sleeping positions such as sitting up during sleep, etc. Again, diagnosis is made with a sleep study.

Sleep Disturbances Related to Routines and Environment

Abnormal sleep can be a problem even if there isn't apnea. If a person does not get into a good sleep routine, poor sleep may result. As discussed in the section on "The Groove" (chapter 10), routine is important to many people with Down syndrome. For sleep, the downside to the groove is that without a set pattern of getting ready for bed, someone with Down syndrome may have a difficult time settling down and falling asleep. The upside is that once the person has established a bedtime routine, she can generally follow it successfully night after night.

Sometimes sleep disturbance is more of a social problem. Noise from roommates or other activity in the house may keep a person awake. This is generally

more of a problem in group residential facilities where staff may have nighttime duties that can make noise and be disturbing. However, a number of our patients who live in their family homes also have this problem. Many people with Down syndrome seem to be "light sleepers," and when other members of the family have different schedules, their activity may wake the person with Down syndrome. Others just like to make sure that everyone else is in bed before settling down or like to be up when others get up in the morning.

Sometimes inadequate supervision and too much independence at night may contribute to inadequate sleep. Too much independence can be a challenge for some of our patients who either live at home or in supported residential settings. Some of our patients need assistance making a scheduled routine for sleep and may need "refresher" courses periodically to stay on the schedule. Use of picture reminders or schedules can be very effective (see chapter 6 on visual memory and chapter 16 on treatment of mental health issues).

Restless, Fragmented Sleep

Even when all the above causes of sleep difficulties are eliminated, many people with Down syndrome have restless, fragmented sleep. Studies have shown that for unknown reasons, they may move all around the bed, fall out, sit up, lean their heads against the wall, or wake up frequently during the course of the night. These types of chronic sleep difficulties can lead to irritation and problems with attention, and emotional control. Parents and caregivers should be aware of these sleep difficulties as a possible source of behavioral problems, although there is no specific treatment for them. Sometimes a trial of medication that produces sedation might be beneficial.

Allergies

Allergies can be a significant contributor to behavioral challenges or psychological changes. While further research is needed to determine whether there is a direct link between allergies and behavior, it is very clear that there is an indirect link between allergies and behavioral changes. The sense of feeling poorly can lead to irritability and mood changes. We have seen adults with Down syndrome who have more behavioral challenges during the time that their allergies are symptomatic. An

assessment for allergy symptoms and a possible link to behavioral changes is, therefore, an important part of the history and physical exam.

Sensory Issues

Some people with Down syndrome seem to be more sensitive to stimuli around them. They may be sensitive to noise, touch, temperature changes, and other sensory input. And as discussed above, many seem to have an altered perception of pain.

Some people with hearing loss experience a phenomenon called recruitment. They generally hear poorly at softer volumes. As the sound gets louder, at some point enough hearing cells have been "recruited" that suddenly the person can hear at that loud level. This can be very startling and frightening.

Many of our patients also tend to have an increased sensitivity to touch. This is common in people with autism and is often a problem in our patients who have both autism and Down syndrome. However, it is also an issue for many people with Down syndrome who do not have autism. In particular, we have noticed that many of our patients are reluctant to use moisturizing cream for their dry skin. They don't like the sensation of the cream.

For some people with Down syndrome, it seems that the issue may not be so much a heightened sensitivity as an inability to filter out sensory input. Some people with Down syndrome seem to be sensitive to activity going on around them that others might just "tune out." This can lead to sensory overload. As noted in chapter 4, many of our patients are very sensitive to activities that are going on in the distance that do not involve them.

This awareness of activity is present even when the person doesn't appear to be paying attention. For example, in the exam room when the conversation moves away temporarily from the person with Down syndrome and we are speaking with the family, the person with Down syndrome will often appear not to be even paying attention, perhaps reading a magazine or playing a video game, when suddenly she makes a comment that clearly indicates she is hearing and understanding the conversation very well.

Loud noises, overly stimulating environments, and other contributors to "sensory overload" can cause the person to become upset, agitated, anxious, or depressed. In addition, she may be aware of events that others may not realize she is aware of. We often hear, "She doesn't even seem to be paying attention to the noise around her" or "She was not aware of the event because we never told her."

However, the stimulation or awareness is usually occurring even if the person makes no sign of being aware. These sensory inputs can be a source of behavioral and emotional problems and should be considered in the evaluation. See chapter 12 for more information on sensory issues.

Alzheimer's Disease

Alzheimer's disease is a progressively degenerative neurological condition that affects the brain. Alzheimer's disease is a form of dementia (a persistent impairment of a prior level of intellectual functioning). There is progressive destruction of brain cells, especially in certain parts of the brain. People with Alzheimer's disease experience gradually worsening impairment of memory, cognitive skills, and skills of daily living, as well as psychological changes. There is presently no cure for Alzheimer's disease, but there are some treatments that can, at least temporarily, reduce its effect. Unfortunately, studies have shown that these treatments are not beneficial for people with Down syndrome.

Research indicates that Alzheimer's disease is more common in adults with Down syndrome and that the symptoms can occur earlier in people with Down syndrome than in other people. It often begins in the fifties (and sometimes in the forties or rarely, even in the thirties) instead of in the sixties or seventies, which is more typical in people without Down syndrome. As we mentioned before, many cases of suspected Alzheimer's disease turn out to be something else; still, doctors should assess for it in adults with Down syndrome. It is a diagnosis we particularly consider if the person is over the age of thirty-five. The youngest person we diagnosed with Alzheimer's disease was in his late thirties. Alzheimer's disease is addressed in detail in chapter 26.

Attitude toward Health Problems

If an adolescent or adult with Down syndrome is aware that she has a significant health problem, it is important to assess her attitude toward her condition. This is because one of the possible secondary psychological impacts of illness relates to how we explain what is happening to us and what will potentially happen to us. If we understand the implications of the illness and the treatment, we can be empowered to participate in the treatment process. If someone has limited intellectual skills, difficulties understanding the process can limit her opportunity to

participate in it. For example, we often encounter this problem when people need physical therapy but cannot comprehend why the mental and physical challenges of therapy are needed (e.g., to regain mobility in an arm or leg after a fracture and casting). These individuals are often less able to participate adequately in the therapy.

We have often seen people with Down syndrome who understand that something is happening to them but either cannot understand what it is or are not able to discuss it. Fears about the possible implication of the illness can be an issue for people with Down syndrome, even if they do not express this fear verbally. For example, if an adult with Down syndrome saw someone in the hospital who required oxygen and subsequently died, this could lead to a markedly exaggerated fear of the need for oxygen in the event of her own illness. Her inability to verbalize this fear or to understand how her own illness differs from the deceased person's can have a negative impact on the treatment and lead to problems such as anxiety or depression.

Some of our patients are very capable of expressing their feelings about their conditions. They may share these feelings in the doctor's office or may give a clue about their feelings to someone else through the course of their day. This is important information for the practitioner to have, whether he obtains it directly from the patient or the family. Sometimes, however, it is necessary to try to put yourself in the person's shoes and ask, "Would I be frightened or anxious in this situation?" If so, then it is usually safe to assume that the person with Down syndrome may also have those feelings. Avoidance and denial are mechanisms that many of us use on some level in dealing with health problems. These should be considered when deciding on the treatment approach for people with Down syndrome as well.

Conclusion

Medical or physical conditions often play a role in psychological or behavioral problems. Treating the psychological or behavioral issue without addressing the medical problem will generally result in a less-than-satisfactory outcome. We have seen many people with Down syndrome who had psychological symptoms that were at least partly due to a physical problem. For example, a high school student who was crying and laying his head on the table, especially at school, was found to have atlantoaxial instability. A thorough medical evaluation of an agitated adult led to the diagnosis of an atonic (poorly contracting) urinary bladder. He was experiencing pain from an overfilled bladder that he couldn't empty. Another adult with Down

syndrome who was having difficulty maintaining his weight and had lost interest in activities was found to have celiac disease. These, and many other people with Down syndrome, had significant psychological issues for which at least part of the treatment was addressing an underlying physical problem.

It is essential to pay careful attention to potential physical or medical conditions when diagnosing and treating behavioral and psychological conditions. Addressing physical and medical conditions can reduce a person's discomfort and improve diagnosis and treatment of behavioral and psychological conditions.

Return to Table of Contents

Chapter 3

Family and Community Support

Jonathan lives in a three-bedroom apartment with two other men with intellectual disabilities. Once a month he visits his parents at their home. Often some of his siblings are there as well. One month, however, Jonathan's parents went out of town to spend time with a sick relative, and the usual visit to his parents' home was canceled. Soon afterward, Jonathan grew less interested in his activities, quicker to anger, and less friendly with his housemates and staff. Recognizing the likely reason for these changes in Jonathan, the staff of the agency that supported him reviewed pictures of his parents with him as well as a picture his parents had sent of his aunt who was ill. They helped him mail a get-well card for his aunt and arranged for him to use FaceTime with his parents and his aunt. He reported that he understood the schedule change and felt included in the family events that were occurring. His mood brightened, and he resumed his usual activities.

Family

Family is generally very important to people with Down syndrome. Just as for other people, it is a place of grounding—a group that loves and is loved, that supports and is supported. There is a commonality. There is a bond. Without this bond, people generally have a sense of something missing. As one mother said, "When I die, the best thing I can do for my son is arrange for someone who will advocate for him without being paid to do it." This was not a slight to the staff who cared for her son, but recognition of the importance of family. That certain bond of love that comes from family is still important, no matter how many other people are there for the person.

The definition of family has changed a great deal over time. It is not in the scope of this book to define family or to comment on the different types of families. However, it is critically important for the person with Down syndrome to be connected to people who mutually view one other as family. Maintaining a constant presence of some people in the life of a person with Down syndrome is the key. It is

particularly important to anticipate these needs as parents age and die or staff at a group facility turn over. Identifying others who have a committed, ongoing relationship with the adult with Down syndrome helps broaden the sense of connection as well as soften the impact of possible future loss.

Family is important whether the person lives at home or not. In group homes, this is particularly evident at holiday time. When the other residents go home to their families, the adults who have no family to visit feel a sense of absence in their life. They may show this through behavioral changes, sadness, irritability, or other changes. Family is not just a group that supports the person with Down syndrome. It is also the main group that the person with Down syndrome can support. We repeatedly see dramatic mental health benefits for people with Down syndrome who have opportunities to help others. The family home is the first place a person with Down syndrome can learn this behavior as a young person (and be encouraged to do it). It is also where he can continue to use and improve these skills. To care for others is a tremendous motivator and contributes to the development of a sense of self-esteem. Unfortunately, some people with Down syndrome spend their entire lives "being done for" and don't get an opportunity "to do for."

To Live at Home or Not?

Many adults with Down syndrome live at home with family throughout their lives. We are often asked, "Is this the best choice?" There is not a single answer to that question. For some of individuals with Down syndrome, living at home with their parents and/or siblings is the best choice. For others, living in a residential setting or in their own home or apartment is best. Some who live at home would be better off living in a residential setting or their own place and vice versa. It depends on the person, the setting, the needs of the family, and many other factors. Sometimes families only determine the best choice by trying something else and deciding that the original choice was the best fit.

Advantages of Living at Home

Elizabeth, age thirty-one, was evaluated for a depressed mood and a desire to move back home. She had begun living at a residential facility four years prior. Although it was a wonderful place where several other adults with Down syndrome lived, Elizabeth expressed concern that she

was missing out on family activities. Elizabeth comes from a large extended family whose members live close to each other, see each other frequently, and are very involved in each other's lives. She was less able to participate in these activities, and it made her unhappy.

Due to a shortage of residential placement openings, Elizabeth had waited a long time for the opportunity to move to her residential facility. Her family was reluctant to "give up her spot" unless it was the right thing to do. We discussed the issues and the options with Elizabeth and her family. Although we determined that Elizabeth was not depressed but "just" unhappy, in the end, Elizabeth and her family decided that she would return to live with her family, and that was the best decision for them.

Living at home has many advantages. There is a familiarity to the routine, the expectations, and the opportunities. At the same time, increased or different expectations and opportunities can be developed as the person ages and his skills and interests change. He can participate in housekeeping chores, have an outside job, and have his own friends, just as other family members do. As when any adult lives at home, both the adult and the family must work to expand the role from child to adult, work on independence and self-supporting issues, and ensure the adult's identity is separate from his parents'.

There is a sense of safety and support at home as well. Safety can be a concern for people with Down syndrome, and families have realistic concerns about how others may treat their family member with Down syndrome. As addressed later in this chapter, a balance should be found between providing a safe living environment and helping the person with Down syndrome to optimally develop his skills. With attention to these two needs, many families have ensured that their home situation offers safety, support, and opportunity for the adult with Down syndrome.

Living at home will also likely give the person with Down syndrome more opportunities to support his family. As indicated above, this can be very beneficial for him and the other family members. In fact, we have seen a role reversal in many families. As the parents age, their son or daughter with Down syndrome takes on greater caregiver functions. This can be of tremendous value to the parents and the person with Down syndrome. On the other side of the coin, we have seen many families who feel a sense of purpose because of their need to support their adult son

or daughter with Down syndrome. As described by many parents, their continued caregiver role for the person with Down syndrome keeps them "young and active."

One practical concern families have is what happens if and when the parents of the person with Down syndrome die or become unable to care for him. Clearly, families need to make a contingency plan ahead of time. These plans might include any of the following: the person with Down syndrome moving in with siblings; siblings moving into the family home; the adult moving to a residential facility; donating/selling the family home to a residential facility with the contingency that their son or daughter will continue to live in the home supported by the residential staff; and others. The option of being able to continue to live in the family home has been a very good choice for several of our patients. The continued familiarity and the sense of continuity and constancy can be very steadying in the face of the other changes.

One of the key planning pieces is making sure the person with Down syndrome has a connection to others in addition to those living in his home. For example, if he is living with his parents, we encourage participation in activities with other people outside the home and aside from being with his parents. This might include opportunities to spend time, including overnight, independently in the homes of other family members or friends. This not only provides more life opportunities and enables the person to experience more independent growth but also prepares the individual for a potential time in the future when living with his parents is no longer an available option.

Visual Supports

One of the advantages of living at home is that the same people support the adult with Down syndrome on an ongoing basis. With this constancy, family can learn about the person's needs, means of communication, desires, schedules, etc. With an active effort to use previous knowledge of the adult and build on it, this can be a dynamic learning process for both the person with Down syndrome and his family.

One way for families to build on this information is to use visual supports. For example, as discussed later in this chapter, picture schedules can be a very effective way to help the person with Down syndrome develop a sense of order to his schedule and take a more independent, self-directed role in managing his schedule. The visual schedule also lets others know what activities the person expects to occur and when.

If a change is anticipated, discussing and negotiating this with the person with Down syndrome beforehand can reduce the stress of the change.

Another very useful tool is a personalized book about the person with Down syndrome. The book can include a variety of sections, including "Who I am," health history, likes and dislikes, schedules, means of communication, etc. Creating this book can help each family member recognize the needs and wants of the person with Down syndrome and support him. These books are even more helpful in communicating essential information to nonfamily members such as employers, recreation directors, and other individuals. If the adult moves to a group home, we encourage continued use of these books or the development of some other tool to communicate this information to staff. We also encourage the person with Down syndrome to participate in deciding what is included. Those who support or meet the adult with Down syndrome can then use the information to optimize communication.

Advantages of Living Away from Home

For an adult with Down syndrome, there can also be some disadvantages to living at home and some advantages to living in a residential facility or his own apartment or home. Chief among the disadvantages to living at home are the risks of isolation and overdependence on the family.

Sometimes an adult with Down syndrome who lives at home may become more isolated. Perhaps the only nonfamily members he encounters are his parents' friends. As his parents age, they may leave the home less often or have fewer people to the house, and this may also create isolation.

A continued dependence on family may also occur. The familiarity with routine that is mentioned as a positive in the previous section may also be a negative if family members continue to do tasks for the person with Down syndrome that he could do or learn to do for himself. He may not develop skills or may lose skills if family members do too much for him or maintain a parent-child relationship without giving him the opportunity to grow into an adult. Families often must make a concerted, conscious effort to help the adult with Down syndrome continue to develop his skills. If they don't, the person with Down syndrome may not get enough mental stimulation or opportunity for growth. This can be particularly harmful if his home life is isolating as well.

Roger, age forty-three, lived with his mother, who was in her late eighties. She had many health problems that limited her ability to leave the house or to participate in activities. Roger was helpful to his mother and felt good about the care he could provide. However, he did not have opportunities to work, socialize with others, or participate in community or recreational activities. When his mother died, Roger went to live with his sister, who lived close to the Advocate Medical Group Adult Down Syndrome Center (1,500 miles from Roger's former home).

Roger's mother and his family had not made any real plans for him in anticipation of her death, and unfortunately, his sister, who had had a limited role in Roger's life for a number of years, struggled to care for him. One problem was that Roger had developed a very set routine with his mother, and he was resistant to change. For example, he had an extremely rigid and limited diet that was not healthy. Eventually, Roger moved to a residential facility. He gradually adjusted over several months but developed depression in the interim and required antidepressant medications.

Roger's situation illustrates several potential problems that deserve consideration. Roger and his mother had developed a very set routine that was functional in their home. However, nobody considered the possibility that Roger would move out in the future, and therefore no efforts were made to help Roger build in some flexibility. When his mother died, Roger had to deal with her death, a long-distance move, and a trial stay with his sister, who could not care for him. All these changes occurred simultaneously. Shortly thereafter, he also had to deal with moving to the residential home. It was a nice home with a few other residents and a very supportive staff, but it was another change.

We have often found that it is advantageous for the person with Down syndrome (as one mom put it) "To make the move before he has to." If the ultimate plan is for the person with Down syndrome to move to a residential facility when his parents become unable to care for him or die, there are advantages to making this move prior to the time it becomes necessary. One advantage is that he will not have to deal with the stress of the move at the same time he is dealing with the stress of his parents' incapacity or death. In addition, if the move occurs when the parents are still able to help, they can provide support during the time of transition. Another

advantage might be that an urgent situation could be avoided. At the time when the person's parents die or become unable to care for him, the choice of living arrangement (residential facility, apartment, home, etc.) may be unavailable. This can lead to rushed, anxiety-provoking efforts to find a place and may necessitate multiple moves that can add to the stress.

It might be easier and more reassuring if we could give a quick, one-word answer to the question, "Is this the best place for him to live?" However, to no one's surprise, this is not the case. There are many issues to address that are unique to each person with Down syndrome and to each family. To add to the complications, sometimes families conclude that a setting other than the family home is best, but the setting or the appropriate support system is not available. The availability of a particular type of housing and sufficient support obviously is one of the factors in the decision.

Each person with Down syndrome and his family assesses the issues they value and their goals along with the available resources. Safety, independence, continued learning and growth, accessibility to family, available residential settings, ability to live independently in an apartment or own home, available support systems, and many other factors are all weighed. The setting that is chosen is then optimized to support the values and achieve the goals. For many reasons, there are different right answers for different people with Down syndrome and their families.

Staff Turnover

The issue of staff turnover requires further comment. For people living in a group home, the staff often becomes like family. This is especially true if the person does not have other family involved in his life but is also true when the person's family is still very involved. When a staff person leaves employment at the facility, it can be very much like a member of the family leaving. It can be quite traumatic for the adult with Down syndrome. We have often wondered what it would be like to have our families turn over every six months, which, in effect, is the situation in residential facilities with frequent staff turnover. It seems that we ask people with less intellectual ability to deal with greater change than most of the rest of us could cope with. The challenge is to create as much constancy in the "family" as possible, to appreciate the stress that occurs because of these changes, and to be supportive.

Peers

In addition to family, people with Down syndrome need friends and peers. Having a variety of friends with varying interests is a reasonable goal. However, it is important to remember that most of us have friends who have similar interests and intellectual abilities. Like the rest of us, people with Down syndrome have "the right to congregation as well as the right to integration," as a member of the audience at one of our conferences suggested.

Particularly with the improved opportunities for people with Down syndrome to be more fully integrated into society, it is important not to discount the value of having peers with a similar level of intellect. Often as children with Down syndrome get older, both they and their classmates become more aware of the differences between them. While we are not advocating limiting these friendships, we are advocating not fostering them exclusively at the expense of friendships with other people with intellectual disabilities. Without these friendships, we have seen many people with Down syndrome find themselves "between two worlds" and feeling as if they do not fit in either one.

In addition, steering an adult with Down syndrome away from others with disabilities or only toward people without disabilities has posed unanticipated challenges for some people with Down syndrome. The message that may be heard is that a person with Down syndrome shouldn't associate with people with intellectual disabilities. Since he has an intellectual disability, he struggles with his own identity. If he shouldn't "like" others with disabilities, he cannot like himself, since he has a disability. For some of our patients, this has become a true existential crisis. This is further discussed in chapter 8.

Having friends with and without disabilities adds richness to life. We were so impressed by a young woman with Down syndrome who presented at a conference where we were also presenting. She told us of her "Rolodex." She had a card for each friend and acquaintance. When she wanted to plan an upcoming activity, she went through the cards in the Rolodex one by one until she found the person or people that she wanted for the activity. Her family promoted wonderful social interaction skills from an early age. Contact lists in phones can be used similarly to make it easier to reach out to friends and family.

Recreation

Recreational activities are an important part of the lives of people with Down syndrome. In addition to being part of the enjoyment of life, they are important for the promotion of both physical and mental health and play an important role in restoring health when there is an illness.

Participating in activities, hobbies, travel, and community events leads to a sense of well-being, helps develop self-esteem, and helps a person develop and improve social skills. We often recommend interesting events that are both physically and mentally stimulating, such as walking inside a museum. People with Down syndrome often learn by the example set by others, and some are great "imitators." Therefore, participation of family, friends, and care providers in these events is a wonderful motivator. For example, we have run a group at our center in which one of our staff members was "part of the group." She did the exercises, learned about and practiced good nutritional habits, and participated in the discussion right along with people with Down syndrome. Rather than just being a leader, she was a participant and an example, and the group flourished.

Good imitation skills can also, unfortunately, contribute to a reduction in the activity level of an adult with Down syndrome, as demonstrated in the following example:

> *Luke was a forty-seven-year-old man with Down syndrome whose father developed severe lung disease and required constant oxygen therapy. The father stopped going out of the house except on rare occasions, and he sat in the living room most of the day in his pajamas. Luke's activities decreased as his father's activities decreased. Luke's father had been a major source of activities outside the home for Luke. Luke reached a point where he refused to leave the house. He began sitting in the living room most of the day in his pajamas. He even went a step further than his father did because he would only wear blue pajamas.*

The reduction of activities is not always related to an illness in the family. As parents naturally slow down with age, they may become less active outside the home, and the person with Down syndrome often becomes less active as well. This can lead to less involvement in enjoyable activities, as well as isolation from friends and peers.

We have seen this contribute to the onset of depression in several of our patients. The problem can be compounded if the parent develops dementia. When a parent develops dementia, it is not just physical activity that decreases but also mental stimulation.

Wes was forty-five when his mother developed Alzheimer's disease. Although her mental capabilities were declining, she and Wes still lived by themselves. Wes stopped going to work as well as to many activities outside the home. Initially, Wes took on the caregiver role. However, as the situation became overwhelming for him, he too began to decline. He made comments such as "She doesn't like me anymore." He initially appeared to have Alzheimer's disease himself, but it later became clear that he was depressed. Wes responded to counseling, assistance in the home for both him and his mother, and antidepressant medication. Eventually, his mother moved to a skilled care facility, and Wes moved into a group residential facility.

For younger adults with Down syndrome, the possible impediments to recreational activities may appear to be much less dramatic, but the consequences may be just as damaging. As the person with Down syndrome ages out of the school system, often fewer activities are available. Therefore, when he is still a child, it is critical to help the person develop skills that will serve him well in adulthood. A planned transition from school to adult life is important. It is imperative that social opportunities be included in the planning. Each person's needs are unique, and a careful assessment of skills and personal preferences is important to determine what activities will best suit the person. In addition, as the person with Down syndrome ages, his interests and physical capabilities may change. Regular reassessment of the activities, his likes and dislikes, and how his activities match with his health status is important to help "get him off on the right foot" after leaving school and assist him in confirming ongoing availability of appropriate opportunities.

It is beyond the scope of this book to go into detail about locating and taking advantage of recreational opportunities for adults with Down syndrome. However, if you are concerned that an adult with Down syndrome is getting too little mental and physical stimulation, here are some places to begin gathering information about opportunities in your community:

- the local ARC or Down syndrome support group,
- a local chapter of Easter Seals;
- Special Olympics,
- your local college (many have programs or courses aimed at students with developmental disabilities),
- a community recreation center, or
- special recreation programs.

Employment

Employment is an important part of daily life for many people. In addition to providing one's livelihood, it can be important for self-esteem and can help provide a sense of purpose and direction.

Just as it is important to help adults with Down syndrome to select recreational activities, it is also important to consider personal preferences when looking for employment. The ability to choose and the ability to have an impact on choices being made are significant.

Cyrus had recently graduated from high school and was working in a job at which he seemed to be doing well. However, he began to have significant behavioral challenges there. These challenges had never been a problem before. Cyrus became depressed. Although he had good language skills, he had difficulty verbally expressing his emotions.

Through counseling and talking with his family, Cyrus was eventually able to verbalize that he was unhappy with his job. He seemed to be asking himself, "Is that all there is?" His mother had spent a great deal of effort helping get the work program up and running, which contributed to even more communication problems with regards to Cyrus's unhappiness. He seemed to have a sense that he had not had an impact on the decision to work there. As the problem became clear, discussions were held with Cyrus regarding his job duties, and he was given different ones. The new duties ended up not being significantly different from the old ones, except that he had had an impact on the decision. Cyrus was much happier in his job.

Like other people, adults with Down syndrome have a varied range of interests. Some people with Down syndrome enjoy repetitive jobs that fulfill their need for order and regimen. Some want to feel needed and achieve this by doing things for others. Still other people with Down syndrome have the desire and skills to handle a job in the community. There are often fewer other people with intellectual disabilities working at jobs in the community, but the job may be more attractive and rewarding than the opportunity to socialize with peers at work. For others, a setting where there are more people with intellectual disabilities is a better fit. Sometimes these jobs might appear to be less interesting to someone who doesn't have disabilities. However, to the person with Down syndrome, the job may be more fulfilling than a job in the community. Also, the opportunity to work with peers might be the most attractive aspect of the job. The key is personal preference.

Employment, like other activities, is ideally more than simply just "something to do." It is an opportunity for learning and for developing a sense of accomplishment and worth. An assessment of skills and capabilities must be part of the job selection process to ensure that the person ends up in a job where he can learn and become more accomplished. The job then has to be taught and organized in such a way that the adult with Down syndrome can be successful.

Barb, a young woman with Down syndrome, was a fantastic bagger at a grocery store. She learned the rules ("bread on the top," "be careful with the eggs," etc.) and could do the job very well. However, another part of her job was "facing the shelves." This task involved pulling the items on the shelves forward to make them more accessible and visible. In a store that covered thousands of square feet, this could be an overwhelming task. The manager was aware of this and helped Barb break the task down into pieces. Barb did a magnificent job. While many of the other people in the store found the task repetitive and boring, Barb relished the order and preciseness.

Unfortunately, when a new store manager started, he did not appreciate Barb's need to have the task broken down into pieces. When he only directed her by saying "Face the shelves," Barb became overwhelmed by the enormity of her responsibilities. She became immobilized and could

not even perform the tasks she previously did well. The manager believed she was being insubordinate and fired her.

A rewarding job that pays well is a wonderful, achievable goal for many people with Down syndrome. Depending on an adult's skills and the availability of jobs, however, this goal might not be realistic or may take some additional assistance that may or may not be available. If obtaining the right job is not possible or resources for assistance are not available, we recommend reassessing priorities. This might include eliminating the moneymaking aspect of the goal or at least reducing it on the priority list. One of the ways that some of our patients and their families have put this reassessment into action is to do volunteer work instead of, or in addition to, a paying job. Many people have developed an improved sense of accomplishment and self-esteem through volunteer work. In addition, often the person can learn new skills that can later be used in a paying job. Also, volunteer opportunities often bring the person out into the community and may lead to other opportunities for employment. Further information on employment can be found in the chapter on self-esteem, chapter 8.

Selecting Appropriate Activities

Decisions about jobs, recreational activities, and living arrangements can be difficult to make. Is the choice appropriate? Will it help foster independence and personal growth? Is it safe? Does the adult with Down syndrome have the skills to be successful? If not, what can be done to help him develop the skills? These are all challenging questions to be considered.

Helping a child develop independence is an important part of parenthood. While it can be argued that most of us are not truly completely independent, most people with Down syndrome will generally have a greater degree of dependence throughout life. The ongoing challenge for families and care providers is to help the person with Down syndrome achieve maximal independence. On a day-by-day basis, providing opportunities to develop skills and having appropriate and realistic expectations are key aspects to assisting a person with Down syndrome increase independence.

It is truly a challenge to help someone become more independent. Skills need to be learned and practiced; these skills must be regularly assessed so that reasonable

expectations can be made and an appropriate level of independence can be provided; safety issues must be addressed and monitored.

Teaching and practicing skills or activities of daily living usually is, and should be, part of training, both at home and at school, during childhood. This training should continue into adulthood. Generally, adults with Down syndrome can continue to learn throughout life. The training should not only include how-to-do skills (e.g., brushing teeth, washing dishes, traveling on public transportation). To be truly independent, a person needs to learn how to schedule and organize his time and activities. This is often a bigger challenge for people with Down syndrome. For many of our patients, schedules and calendars are helpful. Many do well when pictures of the activities are used rather than written words. Even some people with Down syndrome who read well find schedules or calendars with pictures easier to use. Actual photographs often are better than schematic drawings. Calendars and schedules can usually be pretty easily made at home. In addition, there are some commercially available products. We use the Boardmaker software program (www.goboardmaker.com). Many of our patients are now using schedules or calendars on iPads, smartphones, and other electronic devices.

At school and work, there is generally some sort of ongoing evaluation of skills. Families usually evaluate skills on a less formal basis. Regular assessment helps determine when the person is ready for greater independence. But it is also important to bear in mind that for many people with Down syndrome, "once learned is not always remembered." Therefore, in addition to ongoing assessment, ongoing practice of social skills for work, home, and other settings may be necessary.

Use Visual Supports

As discussed in chapter 6, visual learning and visual memory are commonly stronger than auditory learning and memory for people with Down syndrome. Picture schedules and reminders made symbols and pictures of the individual doing the activity can be strong visual reminders and learning tools. Videos also seem to be a particularly beneficial learning tool for adults with Down syndrome. We find that videos depicting the individual himself are particularly strong tools. We both develop such videos and search for useful videos made by others and share the information on the Resource page of the center's website (https://adscresources.advocatehealth.com/).

Clearly, we don't have the resources to develop a video of each person with Down syndrome depicting the desired behavior. However, watching another person with Down syndrome do the desired behavior is a good alternative to a video depicting the person himself. These videos include many "healthy behaviors," including good handwashing technique, eating at a slower rate to prevent choking, improving hydration, and many others.

Safety concerns are clearly a major hurdle for a person with Down syndrome who is developing independence. Safety issues can not only make it logistically more difficult to help a person develop greater independence but can also make families and care providers more hesitant to allow it to happen. Clearly, safety concerns prevent some families and care providers from encouraging adults with Down syndrome to gain more independence. This problem should be openly discussed so that family, teachers, care providers, and other professionals can develop strategies to address the concerns. Occupational therapists can help evaluate skills and develop ways to increase independence.

Too little independence is clearly a problem. It stifles growth and development of skills and can lead to a sense of frustration. Too much independence is also problematic. The individual can become overwhelmed and actually perform at a level lower than might be expected given his skills. This problem is further discussed below in the section on the "The Dennis Principle."

Expectations can have a major effect on an individual's ability to become as independent as possible. When expectations are too low, they can limit growth; when they are too high, they can be confusing and cause the person to "shut down" or give up trying. This can lead to depression (discussed in chapter 17), obsessional slowness (discussed in chapter 19), or other mental illness.

Once again, regular evaluation is needed to determine appropriate expectations. Adjustment of the expectations is the natural next step as the person's skill level changes. The key is to find the appropriate level of expectation and adjust it upward as the person develops. Again, the hurdle to overcome is finding the balance between appropriate expectations and safety. Some "falling down and skinning one's knees" is necessary for achieving the desired growth and optimal skill level.

The Dennis Principle

In business, the Peter Principle describes the phenomenon in which a person is recurrently promoted until he reaches a position for which he is not qualified. We have seen a similar phenomenon occur with several of our patients. We have named it the Dennis Principle (not because Dr. Dennis McGuire has reached a position for which he is not qualified, but rather because he was the first to describe it).

Many of our patients with Down syndrome have been recurrently "promoted" to less restrictive residential or work environments until they have reached a level that they cannot manage. Often, we find that they can manage the actual tasks. For example, they can cook for themselves at home or can do the actual task at work. However, they may not be able to develop a plan to use these skills without assistance or direction. They often need assistance with what we call "executive function," the planning and organization needed to achieve success in using the skills one possesses. In addition, when these adults reach the level they cannot manage, the emotional challenge can overwhelm their coping skills.

Sometimes the issue is that the adult lacks the self-initiation skills to use the appropriate behavior. Other times, difficulties revolve around dealing with roommate issues or interpersonal problems. Often the issue is not knowing how to use "downtime" or relaxation time. The person may be unable to decide on and initiate a recreational activity when there isn't a structured event. This can lead to isolation, frustration, or unhappiness if he spends too much time without anything to do. He may not have the skills to use his time in a fashion that allows for healthy relaxation.

If these issues are not addressed, the situation can become progressively stressful, and the adult may become depressed. Some people have become overwhelmed and have lost skills in several areas, even to the point of not being able to do the tasks they could previously do.

In chapter 8 we describe three women who were staying up late to watch movies and eat large amounts of food. They were depressed, fatigued, and experiencing a deterioration in their job performance. One woman had declined so greatly that the staff of her group home thought she was developing Alzheimer's disease. The women were making poor choices with their high level of freedom. After this was addressed with the women, their families, and the staff of the group home, increased assistance for the women succeeded in getting them back on track. Sometimes it is a matter of helping the person write down a schedule:

Brad, age twenty-five, had previously been very active in sports and was quite skillful in several. However, when he was brought in for evaluation, he was no longer involved in sports, was gaining weight, and was falling asleep at work. He was also having some interpersonal issues with his roommate.

Brad had recently moved from his family home to an apartment with a roommate. He had previously cooked, cleaned, and attended several activities after work and in the evening. He was doing very few of these activities in his apartment, and his new sloppiness was causing conflicts with his roommate. After a written schedule was developed for Brad, the problem resolved over several weeks. He was capable of all the tasks on the schedule and enjoyed doing them. However, he was not able to initiate the schedule without some assistance and needed a written plan to follow. (When he had lived with his family, the structure of the family schedule and subtle cues from his family had enabled him to do what he needed and wanted to do). Brad, like many people with Down syndrome, needed consistency and repetition (see chapter 10). He felt supported and much more at ease with the new schedule, which he helped devise with the help of the staff.

The Dennis Principle points out the need to try to assess not only "task abilities" but also the ability to self-initiate activity (including recreational or relaxation activities) and the need for some reminders. The reminders do not necessarily have to come from another person. In fact, a system that does not rely on another person for reminders promotes greater independence. Printed or picture schedules or calendars—on paper or in electronic format—are quite helpful for many people with Down syndrome.

Adults with Down syndrome become more personally responsible for emotional and interpersonal issues when they move into a more independent setting. The Dennis Principle stresses the importance of assessing and, if necessary, assisting the person with Down syndrome with these responsibilities. Assessing the needs before the change and reassessing the situation after the move can help reduce the stress and improve the likelihood of a successful adjustment to the new situation.

Conclusion

Family and friends are commonly recognized as essential people in our lives. Interactions with family and friends are just as important for people with Down syndrome. The key is to encourage and assist in the development of these relationships from a young age. Similarly, decisions about where the person with Down syndrome will live, what recreational activities he will participate in, and where he will work or volunteer must all be made carefully, with an eye toward mental health promotion.

Return to Table of Contents

Chapter 4

What Is Normal? Understanding "Normal," "Usual," and "Common" Behavior in People with Down Syndrome

Maya, age sixteen, came to an appointment with us with her mother. They were seeking our opinion on a recommendation they had received from another provider. Maya's mother was concerned about her daughter talking to herself. The other doctor had prescribed risperidone, an antipsychotic medication, but Maya's mother questioned whether psychosis was the correct diagnosis. Maya's father had died about four months previously, and since that time Maya had talked to herself a great deal. Her mother noted that Maya had talked to herself in the past, but now it was markedly increased. Maya had limited expressive language skills, and even family members often had difficulties understanding her speech. However, her mother reported that at times Maya would go into her room, close the door, say, more clearly than when talking to her mother, "It's going to be OK, Maya"; "Daddy loves you"; "But I do miss him"; and other similar phrases. Maya continued to attend school and her activities but sometimes cried at home, at school, and at her activities.

At our assessment, we discussed that self-talk is common in people with Down syndrome. We also discussed grieving. Our assessment of Maya was that she was going through normal grieving and her increase in self-talk was one of the ways she was coping. We did not prescribe any medications but provided her with counseling, using a variety of techniques, to help her through the grieving process. Over the course of the next few months, Maya's self-talk returned to its previous level.

"Is this behavior 'normal'?" "Why does my son do that?" "Do other people with Down syndrome also do that?" These are questions families and caregivers of people with Down syndrome often ask us.

Although the assessment process described in chapter 1 can ideally provide answers to questions like these, we realize that (1) getting a thorough mental health

assessment might not always be feasible; (2) you may not have access to healthcare professionals who know the answers to these questions; and (3) you just may want to be reassured, without getting an assessment, that the behavior your son or daughter is displaying is not unusual.

This chapter will help you understand the continuum of behavior that ranges from normal to abnormal, the strengths and weaknesses frequently seen, and the common characteristics of people with Down syndrome. These factors all need to be considered when deciding whether behavior is "normal" for someone with Down syndrome.

Normal versus Abnormal Behavior

There are clear definitions for abnormal behavior and psychological problems. *The Diagnostic and Statistical Manual of Mental Disorders*, 5th edition (DSM-5), published by the American Psychiatric Association, and the *Diagnostic Manual—Intellectual Disability 2 (DM-ID 2): A Textbook of Diagnosis of Mental Disorders in Persons with Intellectual Disability* published by National Association for the Dually Diagnosed describe the diagnostic criteria for mental illness used in the United States. (Other countries use a similar manual called the *International Classification of Diseases* published by the World Health Organization.) To be diagnosed with a particular mental disorder, someone must have a certain number of the symptoms listed for that disorder, and the symptoms must persist for the specified amount of time. However, even in people without an intellectual disability, there is room for interpretation, and this is certainly true for people with Down syndrome. Clinical assessment is important, and clinicians use their judgment to determine how behavior meets (or doesn't meet) the criteria.

The DSM-5 criteria are less clear when describing a person with an intellectual disability, since these guidelines were written for people without intellectual disabilities. The DM-ID 2 includes adapted criteria. The typical (or normal) behavior, the developmental stage, communication skills, and other aspects of a person with Down syndrome are different from those of a person without an intellectual disability and may alter the diagnosis. Therefore, there is often more room and need for interpretation of the criteria when applying them to a person with Down syndrome.

Particularly in light of the need for interpretation of behavior when it comes to applying the criteria for mental disorders, behavior can be looked at as occurring on a continuum from normal to abnormal. At one end of the spectrum is behavior that is

clearly abnormal, and at the other end is behavior that is clearly normal, but there is a vast middle ground in between. The same behavior may in one context be normal while in another be abnormal. For example, it is normal for an adult to cry and feel very sad after a loved one dies, but it is not normal for an adult to cry throughout the day merely because little things are going wrong.

Developmental Age

When trying to interpret behavior on this continuum, the first task is to define normal (or typical). When defining normal, a number of issues must be considered. It is particularly important to look at the developmental level of the person.

Psychological testing (including IQ testing) is often done as part of an assessment of a person with Down syndrome. Often at the end of the written report, a developmental age is written (for example, 6 years 7 months). This developmental age means that the person's skills, taken as a whole, are about what you would expect for a typically developing person of that chronological age. This is a reasonable place to start when assessing what might be normal for the person. As will be outlined in more detail in other chapters, there are behaviors that are normal for a person at each developmental stage. These characteristics would not be normal for a person without an intellectual disability at the same chronological age (who is at a developmental age that is the same or closer to her chronological age). For example, it is very normal for a four-year-old to have imaginary friends. When one of our daughters was four, she regularly invited Barney the Dinosaur to our dinner table and insisted we set out a plate for him. A four-year-old would not be treated for a psychotic disorder for this behavior. On the other hand, if her father had insisted on inviting Barney to dinner, one would assess his behavior differently. The importance of understanding who the person is and where she is developmentally is of critical importance.

Most adults with Down syndrome have developmental ages that are lower than their chronological ages. You must always have an idea of an adult's developmental level and behaviors that are appropriate for that developmental age before determining whether her behavior is normal or not.

An important caveat when looking at developmental ages for people with Down syndrome: do not forget that this score is, in a sense, an average of the different aspects of the person's personality.

If it is 100 degrees outside today and 0 degrees tomorrow, the average is a pleasant 50 degrees, but knowing the average doesn't make today any less hot and tomorrow any less cold. Similarly, an average developmental age score can be misleading. While the person may have a developmental age of five years six months, some aspects of her personality may be closer to someone who is four years old, and others may be on a much older level, even consistent with her chronological age. The key is to not put all the focus on the developmental age without considering the whole person. While her social skills may be closer to a four-year-old's, her social aspirations may be similar to those of a person who is twenty-two years old. And while she has many skills at a level comparable to a thirteen-year-old's, her judgment may be closer to a seven-year-old's. Without acknowledging these possibilities, expectations can be too high or too low. For a teen or adult with Down syndrome, it can be a challenge to develop an understanding of the multiple aspects of her personality and to help her optimally develop in each of them. However, you are much less likely to succeed if you only look at the person as having skills and abilities at the level of the "average" developmental age.

Understanding developmental age is an important part of helping a person with Down syndrome optimally develop her skills. Psychological testing provides insight into developmental age. However, care must be taken to appreciate the whole person as well as to understand the behaviors and characteristics that are commonly seen at each developmental age.

Common Characteristics

Robert, a thirty-six-year-old man with Down syndrome, came into the office for the first time. When discussing his family history, he suddenly started crying after mentioning that his father had died. After further discussion, we learned that his father had died fifteen years prior. Robert had an excellent memory; however, his sense of time was limited, as it is for many people with Down syndrome. To him, there was little if any difference between several weeks ago and several years ago. Since the staff understood these concepts, Robert was comforted and reassured, and the interview continued. There was no need to consider the diagnosis of prolonged grieving or even depression.

Besides considering developmental age when determining whether behavior is normal for a person with Down syndrome, many aspects of the person's personality must be taken into consideration. A first consideration is to acknowledge that the person has Down syndrome and that there are common or typical behaviors for a person with Down syndrome. However, care must be taken not to make this consideration the only one taken into account. We call this "two opposite issues."

The first "opposite issue" to understand is that there are some behaviors and characteristics that, while not universal, are common in people with Down syndrome. It is problematic to ignore that possibility because understanding these issues (as will be discussed further in this and other chapters) can assist in diagnosis, treatment, and health promotion. The second issue is that "All is not Down syndrome," meaning that there are many conditions that can coexist with Down syndrome—for example, depression, thyroid disease, and sleep apnea. Unlike Down syndrome—for which there is currently no specific treatment—each of those coexisting conditions is potentially treatable. Many families have shared that after presenting their concerns to a healthcare professional, they were told, "It's just the Down syndrome" as they were politely dismissed from the office. In other words, the provider did not and would not look for potentially treatable coexisting conditions.

While considering the first issue, it is important to understand that there are many behaviors that are commonly seen in people with Down syndrome. They are considered normal within the context of the person. In this section we will discuss a number of characteristics that are common in people with Down syndrome that should not be taken as evidence of mental health problems. These characteristics include the following:

- differences in emotional response and development
- language delays
- self-talk (talking to oneself, also called private speech by some researchers)
- tendency toward sameness or repetition
- lack of flexibility
- concrete thinking
- difficulties understanding time concepts
- slower processing speed
- memory strengths and weaknesses

Differences in Emotional Response and Development

The Myth of Perpetual Happiness

"Individuals with Down syndrome are always happy." Although this is a commonly believed stereotype, it is a myth. The corollary to that myth, which is equally incorrect, is that people with Down syndrome have no stress in their lives (thus, the reason they can be happy all the time). In reality, people with Down syndrome have a wide range of emotions. Their emotions can reflect their inner feelings, as well as the mood of the surrounding environment. Sometimes the emotion is a result of the stress the person is feeling.

The notion that all people with Down syndrome are happy all the time evokes a positive image of people with Down syndrome. While it may be beneficial in light of some of the negative stereotypes held by some, it sets up unrealistic expectations for behavior. This can lead to misinterpretation of behavior, since, as chapter 7 discusses, people with Down syndrome often have difficulty verbally expressing their feelings. We have heard many people express concern when a person with Down syndrome is not happy. Because of the misconception that all people with Down syndrome are happy all the time, it is assumed that something is "wrong" with the person with Down syndrome when she is not happy.

The range of emotions for people with Down syndrome is typically wide—sometimes even wider than in those without Down syndrome. People with Down syndrome certainly express sadness, happiness, anger, indifference, and other normal emotions. Generally, we have found that people with Down syndrome have a high degree of honesty when it comes to their emotions. They may show or even exaggerate the emotion they are feeling. This can be a very positive trait when it comes to optimizing communication. Unfortunately, it can also lead to tactless remarks or socially inappropriate or unacceptable behavior.

> *Joe, a twenty-seven-year-old man with Down syndrome, had a job bagging groceries at a local grocery store. When customers would rush him or upset him, he would voice the anxiety and agitation he was feeling. This behavior offended some customers, who complained to the manager. Joe was fired.*

The problem was not that Joe was unhappy, but that he inappropriately expressed his unhappiness. Negative emotions are just as "normal" in people with Down syndrome as they are in other people. However, the person with Down syndrome may be unable to hide her negative emotions in situations where neutral or positive emotions are expected.

Sensitivity and Empathy (Empathy Radar)

Mark, age fifteen, was with his parents at a school conference. The focus of the conference was expectedly on Mark. Suddenly, he changed the flow of the meeting by asking his teacher, "How are you doing? You seem upset." His parents, who had not noticed a problem, were somewhat startled by his interruption and confused by his apparent lack of understanding of the purpose of the conference. The teacher paused, became teary-eyed, and then told Mark and his parents that a close relative had recently died. She thanked Mark and spent much of the rest of the conference discussing Mark's empathy and compassion for others.

There can be some positive aspects to the honesty of expression of emotions. This is particularly true when this expression is accompanied by the real sense of empathy many adults with Down syndrome possess. Often adults with Down syndrome excel at sensing the emotions of other people. We call this "empathy radar."

At times, this strong sense of empathy combined with the honest expression of emotions is like a mirror. The emotions of a person with Down syndrome can be a reflection of what is going on around her. In particular, the emotion expressed can reflect the emotion of the person she is with. In a setting with people who are treating her kindly, this characteristic can be very positive. However, when those around her are expressing negative emotions, she may express similar negative emotions.

This is important for family and caregivers to be aware of and to acknowledge. The question "Why has Mary become so angry?" cannot be answered in a vacuum. In other words, an assessment of changes in the environment is essential. A child counselor and colleague used to describe it like this: "When families bring a child to

the office and drop him at the curb and say, 'Fix him,' you know you are going to have a real challenge." If families and care providers are unwilling to evaluate (and, if appropriate, acknowledge) the role the environment may play in a behavior change, the treatment will be more challenging and less likely to succeed.

The person with Down syndrome may reflect the emotions of a variety of settings. The emotions she expresses in one setting may actually be in response to something that occurred elsewhere. For instance, anger you see at home may actually be in response to something that occurred at school or work. In addition, physical health or biological issues may be contributing to her behavior or emotions. Therefore, any given environment may contribute little or nothing to a behavioral or emotional change. On the other hand, the environment may play a large role. Therefore, assessing each environment and reviewing the importance of this issue with people in each environment is an important part of the healing process, as illustrated by this example:

> *Jeff came home from work very upset and agitated. When this continued for over a week, his family contacted us. We called his worksite, but his supervisor could not explain the change in Jeff's behavior. There were no problems among the individuals in Jeff's work group, and the staff had not observed anything unusual or problematic for Jeff. After some investigation, we found out that Jeff was upset about a coworker who was having frequent outbursts and crying spells. Interestingly, this man was in a separate room some distance from Jeff (over two hundred feet away). Jeff actually had no contact with him during the course of his day. Still, he seemed to have picked up or absorbed the other man's tension and was greatly affected by it.*

We have found that many people, like Jeff, are not always able to filter out the emotions, stressors, tensions, and conflicts of others. In short, if someone with Down syndrome is showing emotions that do not seem to fit the situation, failure to assess for other possible external factors may lead to misdiagnosis and inadequate treatment. Consider these questions first: Is she mirroring the emotions of those around her? Is she displaying extreme sensitivity to the events around her? Do her emotional reactions seem exaggerated? These are all "normal" for people with Down syndrome. That is not to say that she may not need some help handling her emotions.

For instance, people around her may need to work harder to display positive emotions around her, or she may need to learn when it is tactful to be honest about negative emotions. Sometimes long-term exposure to stressful external factors can lead to mental illness and require treatment (see the chapters in Section 3).

Sensitivity to Conflicts between Others

We have found that people with Down syndrome may be very sensitive to conflicts or tensions between significant others in their lives. Depending on the type and degree of conflicts, they may be severely affected, as the example below illustrates.

> *Mary, a resident of a small group home, was brought for evaluation after a mild habit of scratching her skin became a more serious problem of digging deep cuts in her neck and arms. Even though she had been very social and capable, she developed symptoms of a major depression, including a loss of appetite, restless sleep, sad mood, loss of energy, fatigue, and a loss of interest in activities that she had formerly enjoyed. In her own words, she was "really down." When we questioned her group home manager, who brought her to the appointment, she complained that Mary's mother had caused her problems by being "overly protective." An example she gave was that Mary's mother would not allow her to go on outings that staff felt were beneficial for her. When we called her mother, she in turn complained that the house manager was trying to turn Mary against her, and this was the reason that Mary was under so much stress.*
>
> *After further exploring the situation, we found that the conflict between Mary's mother and staff had existed for some time. Neither the mother nor the house manager was necessarily wrong in what they wanted for Mary—they just held contrary opinions about what was best for her. For her part, Mary was extremely torn and stressed by this conflict because she loved her mother but also felt very close to her house manager. As the situation escalated and became increasingly intolerable for Mary, her agitation and depression increased.*

We have seen similar problems with people caught between two parents in conflict, such as when parents were having serious marital problems or having a contentious divorce. In fact, asking people with Down syndrome, or anyone, to take sides against those they love or who are important to their well-being is extremely dangerous. The stress that this creates invariably causes changes in mood and behavior in the person.

In theory, the solution to these problems is fairly simple. The person with Down syndrome must be removed from her position in the middle. This is possible if she is not asked to side with one parent or caregiver over the other. For example, in Mary's case, we relieved Mary of her go-between position by becoming the intermediary in the conflict and working out a solution that was agreeable to both parties. Her mother and house manager agreed to a compromise that allowed Mary to go out in the community, which is what her house manager wanted, but with a staff person accompanying her, in deference to the mother's safety concerns. In time, many other issues were also resolved in this way with the help of mediation.

Although Mary's problem was solved fairly readily, resolving conflicts may not always be simple, such as when people are in the middle of a contested divorce. In these instances, it is still critical to get the person with Down syndrome out of the middle. What is most successful is to set up firm ground rules to free the person with Down syndrome from taking sides. One absolutely essential rule is for each parent to refrain from commenting about the other parent in front of the person with Down syndrome. Even when the parties try to "hide" their emotions, the person's empathy radar may detect conflict and emotional upheaval. Working on the expression of one's own emotional state may be required to optimize the emotional state of the individual with Down syndrome.

Rules about how and when the person with Down syndrome makes transitions between her mother's and father's households are also of critical importance. Even when there are court-ordered visitation schedules, the rules surrounding this process must be carefully and meticulously reviewed with both parents. The reason is that whatever anger remains between parents will often be expressed in this process. Examples include late pickups or drop-offs, subsequent phone calls, and, of course, angry comments about the other parent directly to or within earshot of the son or daughter with Down syndrome. The reason for these rules must be stated very clearly to each parent: either you do this, or you could contribute to extreme stress and mental or behavioral changes that will be expressed by your son or daughter.

In some cases, the anger between parents is too strong, and the only solution is for the person with Down syndrome to move to a neutral environment, such as a group home. This does not solve the problems completely, but it does limit exposure of the person with Down syndrome to the tension. Once the pattern of transitions is established, the person with Down syndrome is free to respond to each parent without fear or concern of hurting the other. In time, people are able to go back to their normal lives unencumbered by the intense burden one often feels when experiencing this process.

Delayed Maturation

Throughout the lifespan, there are periods when certain emotions tend to be more prominent. This is as true for people with Down syndrome as it is for others. One aspect that is different for people with Down syndrome, however, is the timing. For example, many families report that when their sons or daughters reach their early twenties, they want to be left alone and assert themselves more. This can be seen negatively as depressed or agitated behavior. However, frequently it is all put in perspective when one question is asked: "Do you recall what your other children were like when they went through their teenage years?" This is typical adolescent or teenage behavior that is often, but not always, seen at a later age in people with Down syndrome. See chapter 11 for more information about the differences in maturation.

Delayed Grief Response

People with Down syndrome often have a delayed grief response. For example, when a family member dies, the person with Down syndrome may initially seem to be unaffected. Often, we have seen that she will begin to grieve approximately three to six months later. It is not completely clear why this delay occurs. However, it most likely has to do with slower cognitive processing (see below). The expectation that people with an intellectual disability don't grieve coupled with their impaired communication skills (Brickell & Munir, 2008) could also lead to a later presentation or appreciation of the grief. It may simply take people with Down syndrome longer to recognize and understand that a loss has occurred or that the loss is permanent (that the loved one is truly gone, etc.). Understanding and anticipating this response can

help prevent problems and prepare family and care providers to help with the grieving process when the time comes.

Language Delays

Language limitations in adults with Down syndrome can also lead to misinterpretation of their behavior. Many people with Down syndrome have language deficits. Often their expressive language skills are lower than their receptive language skills. That is, many people with Down syndrome understand what is going on around them but are unable to articulately express their concerns. Even some individuals who can express themselves well in other aspects of daily life may have difficulty expressing their emotions. This can be a real source of stress. It can lead to frustration, irritation, anger, and other emotional changes. Interpreting behavior change in light of this challenge can greatly improve the understanding of the behavior. Because language/communication skills can be a significant challenge for some people with Down syndrome, we devote chapter 7 to exploring them in depth.

Processing Speed

The ability to process data rapidly is an increasing demand of a world whose pace of activity is accelerating. Many people with Down syndrome have a limited ability to quickly process information. In addition, they have trouble shifting processing speed in different situations, which can be even more problematic. Many people with Down syndrome struggle when a situation demands a sudden acceleration in the pace of activity or a demand for rapid adaptation of a different processing mode. Responding to an urgent situation can be very distressing to them. This limits their ability to adapt to different settings, as the example below illustrates.

> *Neal, age seventeen, was having difficulty in school due to his problem switching classes. When the bell rang and the other students walked to their next class, Neal would not move. After discussing the situation with Neal and his family, it became clear to us that Neal required a short period of time to process the need to adjust from the quiet classroom where he was seated to the active, noisy hallway. A*

verbal warning about five minutes before the bell gave Neal time to prepare for the change in activity.

The fact that people with Down syndrome have a slower cognitive processing speed may seem evident based on their intellectual disability. However, in practice, others who are interacting with people with Down syndrome often don't appreciate this. This is a particular problem in fast-paced places of business and can lead to difficulties in the workplace or in the classroom, particularly when employees or students don't all have the same processing speed.

In our interviews of patients, we often see this slower processing speed manifested when we ask questions about a person's health. Over the course of the interview, we ask multiple questions. Not only may it take a moment or two for the person with Down syndrome to answer, but some of our patients are quite fatigued by the end of the appointment. They have spent a great deal of mental energy thinking about and answering the questions.

When asked a question, people with Down syndrome often pause before answering. This can lead to misinterpretation of their behavior and problems in interacting with others. Often others interpret the pause as meaning that the person with Down syndrome is ignoring what they said, is insolent, or has an attention problem. This has led to problems for many people with Down syndrome, especially at work or school. It can be a source of friction between the person with Down syndrome and her boss, teacher, or fellow employees.

In addition, if multiple directions are given before the person is able to process them, then she can become frustrated. We have seen or heard about many people with Down syndrome who stop attempting to process in that situation because they have become overwhelmed. The employer may also become frustrated and lose patience. This can lead to agitation and strained interaction between the two people.

These types of misunderstandings are a common source of conflicts at work or school and can lead to loss of employment or disciplinary issues. Interestingly, this type of issue has been shown to be a more common cause of job loss than lack of skills to do the job (Greenspan & Shoultz, 1981). Understanding and appreciating the challenge of slower processing and providing information at a rate that the person with Down syndrome can process will lead to a much healthier situation and less frustration and conflict.

Making Accommodations for Processing Speed

In light of these issues with slower processing speed, how can others optimize interaction with a person with Down syndrome?

- Understand that this is a potential issue. Being prepared to adjust one's approach is the first step.
- Be careful not to view this as a "behavioral" issue. It may be that the person is slower at processing the information rather than insolent or lazy.
- Anticipate that she may need a period of time to process the information. Start the request soon enough so that she will have that time.
- Get her attention. Wait for a response from her such as "What?" or "Yes" to indicate she is acknowledging you have her attention.
- Make the request or give the directive in an understandable fashion and confirm that she understood. Having her repeat the instructions, if able, may be beneficial.
- Give her the time she needs to process the request.
- After an appropriate period of time (this varies from person to person but may be several minutes, depending on the request), check with her to make sure she understood or that there is no impediment to her proceeding (rather than making the request repeatedly or louder).
- Bear in mind that many people with Down syndrome will just stop trying to comply if there is an impediment rather than try an alternative approach or ask for assistance.
- Try to find alternative ways of communicating (to speak to the person's strengths). For example, many people with Down syndrome benefit greatly from visual images that may augment or accompany a verbal communication or instruction. After all, visual supports are useful in any teaching situation. This is why presenters or teachers use blackboards, slides, or other visual aids when teaching. For people with Down syndrome, this may be especially useful because so many are visual learners (see chapter 6). For example, we consistently hear from job supervisors that people with Down syndrome can learn and reliably repeat even complex, multistep tasks if the task is broken down into smaller steps and shown to them.

Time Reference

Understanding the concepts of past, present, and future is something most people take for granted. Since these concepts are abstract, however, they are difficult for many people with Down syndrome to understand. This can lead to confusion for both the person who doesn't understand and for the people around her. When taken in context of the very strong memory (see chapter 6) that many people with Down syndrome have, this can result in even further confusion. Recall the example of Robert earlier in the chapter. He reacted as if his father had just died when questioned about him, when in reality, he had died fifteen years ago. The understanding of past and present seemed to be different for Robert than for people without Down syndrome. For him, a very strong memory and limited comprehension of time made at least some past events seem as real as present events.

Often, we find a less clear line of distinction between past and present than we would expect in someone who doesn't have Down syndrome. For the person with Down syndrome, the understanding of many concepts is much more concrete, and the concept of time may be too abstract.

This decreased sense of the difference between past and present can lead to much confusion in conversation with other people. Some people with Down syndrome have even been diagnosed as psychotic by other practitioners because the person with Down syndrome appeared to be disconnected to the reality of the present. Most often we have found this type of misdiagnosis to be due to miscommunication and to the practitioner's unfamiliarity with the way people with Down syndrome sense time. As noted before, when this characteristic is taken in the context of a very strong memory, the person with Down syndrome can recall far-off events and may seem to have a disconnection with the present.

If an adult has limited communication skills, this problem is exacerbated. It can be very difficult to ascertain if she is talking about an event that occurred recently or in the distant past. This can, for example, make it very difficult to get an accurate history regarding symptoms:

> *Carol, age twenty-five, has very limited verbal skills and usually speaks in one- or two-word phrases. She was complaining of ear pain. A thorough evaluation revealed no underlying problem. After further discussion, it seemed that this was more of a complaint of her past medical history of frequent ear infections.*

Sometimes the misunderstanding may be due more to difficulties using past and present verb tenses than to a true lack of understanding of time. Dr. Libby Kumin, a speech-language pathologist with a special interest in Down syndrome, has theorized that some people with Down syndrome never learn to use verb endings correctly because hearing problems in their formative years prevent them from hearing the final –s or –ed on verbs. Others do not master irregular verbs due to language learning difficulties and may answer questions such as "What did you do this weekend?" along the lines of "Saturday I eat dinner with my mom." In context, the listener can figure out that the person is speaking of a past event, but a less careful listener could be confused (Kumin, 2012).

As indicated, this different time reference can affect interactions with other people. The biggest problem for people with Down syndrome occurs when others assume they understand time and time references in a "typical" or "usual" fashion. This leads to misinterpretation of what they say and sometimes to disagreements or misunderstandings. It can also result in practitioners making an inaccurate diagnosis on the basis of an apparently altered thought process. Additionally, others may sometimes perceive the person with Down syndrome as untruthful when she is describing an event that occurred in the past but sounds like she is describing a current event.

Based on these findings, we have some recommendations for optimizing communication:

- Appreciate that someone with Down syndrome may have a different understanding of time. If you know that she may be speaking in the present tense about a past event, ask further questions to prevent confusion.
- What are the person's overall language skills? Have you previously heard her use past tense or words like "yesterday"? If not, when she is speaking in present tense, she may actually be speaking about the past.
- If possible, help the person put the event in the time context of another event. For example, ask, "Did it happen when you were in school? When you were working at the grocery store?" Particularly if the person says she doesn't know, look for other clues about when the event occurred. Parents are often very helpful in helping answer this question. For example, "I know that is a past event because she referred to Sally, who was a high school classmate."

Awareness of "Clock Time"

An interesting paradox is the incredible ability of many adults with Down syndrome to "know the time." This is often demonstrated in grooves (see chapter 10). Sometimes people with Down syndrome are very inflexible about times in their routines, insisting that meals, breaks at work, televisions shows, etc. occur at set times. Many people who follow set time schedules cannot "tell time," yet have an internal clock that is often extremely accurate. We have learned to pay close attention to time because people can be less tolerant of our questions and procedures when we intrude on their lunchtime.

Inflexibility about time can also cause problems in employment settings. For example, in the early days of the clinic, we employed a young woman with Down syndrome, Jean, who did an excellent job of data entry. At the time, we had limited space, and Jean had to share a small office with two other employees. One afternoon, Jean suddenly got up and literally climbed over both staff members to leave the room. She then left the building and caught her bus home. The staff members were surprised to find that she had not saved her work or turned off her computer before leaving. When questioned about this later, Jean could not understand their surprise. She explained that it was 2:30, which was "time to go home." Fortunately, Jean was able to learn a different routine, which included wrapping up her work activities appropriately in anticipation of quitting time.

Concrete Thinking

Most of us who are past the age of twelve or so can think both concretely and abstractly. Using our five senses gives us a concrete understanding of the world. However, it can be more challenging to think beyond what we can perceive with our five senses to think abstractly or theoretically. People with Down syndrome often think in a very concrete manner and frequently cannot think well abstractly. The concrete nature of the thought processes of most people with Down syndrome is very functional and can be very precise if allowed to flourish in an appropriate setting. Often people with Down syndrome do wonderfully in jobs that have concrete tasks. In fact, we find that most often when a person with Down syndrome has problems at work, it is not because she can't do the tasks. Often, she does the task extremely well and is a model employee because her concrete nature helps her to do it well

repeatedly. Since this is such an important topic, we have devoted an entire topic to it. Please refer to chapter 5.

Self-Talk

Another behavior that we frequently encounter is self-talk. As described in detail in chapter 9, self-talk is very common in people with Down syndrome. We became particularly interested in this topic when we found a great number of our patients being treated by other practitioners for psychoses, as in the case example of Sara at the beginning of the chapter. Talking to themselves seemed to be the major reason these misdiagnoses were made. While self-talk can be part of the diagnostic criteria for psychoses, these major psychiatric disorders are characterized by delusions, hallucinations, withdrawal from reality, paranoia, unusual mood, and an altered thought process. Self-talk without these other symptoms is not a psychotic disorder. When we assessed all our patients for self-talk, we found that approximately 83 percent of them talked to themselves and that many of the other 17 percent did not speak at all. When medical professionals do not understand or appreciate this finding, it can lead to overdiagnosis of "abnormal."

As mentioned above, self-talk is developmentally appropriate for many adults with Down syndrome, since many typically developing children under the age of six or so talk to themselves.

A similar behavior that is often appropriate for an adult's developmental stage is the use of imaginary friends. This too can lead to inaccurate diagnoses if the developmental stage of the individual is not considered. Self-talk, imaginary friends, and fantasy lives are addressed further in chapter 9.

Tendency toward Sameness and Repetition

Another truly fascinating aspect of the personality of many people with Down syndrome is the tendency to prefer sameness or repetition. We call this "the groove." The groove has many advantages, such as helping a person maintain order in her life and optimizing use of her skills. However, the lack of flexibility can make it difficult to deal with the realities of the changes and inconsistencies of life. In addition, if others do not understand this tendency, it is easy for conflicts to develop, because dealing with apparent inflexibility can be disruptive for people who have less "groove."

Many adolescents and adults with Down syndrome have “grooves” that can be misinterpreted as behavior problems, and we discuss the issue in detail in chapter 10.

Keeping Sight of the Continuum

Understanding what is normal or typical for people with Down syndrome helps define the continuum of normal to abnormal. This understanding gives us a reference point for comprehending behavior in adolescents and adults with Down syndrome. Revisiting some of the issues discussed above will illustrate this principle.

Looking at the tendency toward sameness or repetition, the normal (or typical) behavior for a person with Down syndrome is “the groove.” Abnormal is taking the groove to such an extreme that it interferes with the ability to function efficiently in daily life. Grooves can be quite useful if others in the environment recognize this tendency and are willing to work with the behavior. However, if the tendency prevents functioning in daily life—either because of the degree of compulsion or the inability of others to work with the tendency—obsessive- compulsive disorder may be diagnosed (see chapter 19).

Similarly, with grief, depending on the degree of the problem and the environment in which the person is grieving, the reaction may fit somewhere on the continuum from normal grieving to depression.

Self-talk is another behavioral aspect that can fit on a continuum. As indicated, self-talk is a common behavior in adults with Down syndrome, but self-talk can also be a feature of psychoses. A careful assessment of the nature of the self-talk, associated symptoms, environmental circumstances, and the person’s functioning, as well as the presence or absence of self-talk prior to the present concern, is necessary to understand where the self-talk fits on the continuum.

Another aspect of the continuum that is important to understand is that complete absence of the particular behavior is not necessarily healthier than the presence of the behavior. In other words, because “too much” of a particular behavior meets the criteria for a particular psychological problem (e.g., psychotic disorder), the complete absence of the behavior is not necessarily the goal. For example, families and caregivers often ask whether self-talk should be suppressed. As is discussed in more detail in chapter 9, people with Down syndrome often use self-talk as a means of talking through problems. Therefore, to suppress the self-talk may actually hamper the healing process. In this situation, eliminating self-talk is neither healthy nor the goal of therapy.

While it is important to consider these “typical” behaviors in light of the continuum, it is equally important to avoid the trap of “blaming everything on the Down syndrome.” A typical behavior that has become problematic may no longer just be “typical behavior of Down syndrome.” It is necessary to assess whether it has become a psychological problem. This is optimally achieved by understanding the individual’s behavior throughout her life, particularly if she has undergone a change in behavior. Developing an understanding of the person before the change, assessing the period of her life, evaluating the environment, and using other assessments as outlined in chapter 1 will help delineate the causes of a change in behavior. This evaluation will help clarify whether this is common or typical behavior for a person with Down syndrome, where it fits on the continuum, and whether further assessment and treatment are indicated.

Evaluating behavior with an understanding of the continuum has a several important advantages. It provides a framework for appreciating and acknowledging the unique qualities and common behavioral characteristics of people with Down syndrome. It also provides a structure to assess when behavior is abnormal and further intervention and treatment is necessary.

Return to Table of Contents

Chapter 5

Concrete Thinking and Behaving

Luis works at a job about ten miles from his house. He takes a bus about four miles, transfers to another bus and rides it about six miles, and then walks the last three blocks. He does very well in getting back and forth to work on time. One day, however, there was construction on one of the streets that his bus usually traveled on. The bus had to take a detour a block east, go north two blocks, and then return west to the street the bus usually travels on. Luis recognized as he traveled north on the new street that he was not on the usual route. He got off the bus and became lost. While his concrete thinking required for the daily trip was excellent and he was able to manage public transportation very well, the abstract thinking required by the change in route was difficult for Luis.

One major challenge to adaptive functioning for people with Down syndrome is a reliance on concrete forms of thought and behavior. This can create confusion and a lack of flexibility and adaptiveness in a number of key areas. In this new edition we have added an entire chapter to the concept of concrete thinking because we found it to be so important to our understanding of people with Down syndrome. Still, it is important to state here that like all the common behavioral characteristics addressed in this book, concrete thinking and behaving can be both a strength and a weakness.

Before proceeding, let us first explain what differentiates concrete and abstract thought. Most of us who are past the age of twelve or so can think both concretely and abstractly. Using our five senses gives us a concrete understanding of the world. That is, it enables us to see things as they actually exist. However, it can be more challenging to think beyond what we can perceive with our five senses to think abstractly or theoretically—to imagine things as they might be or to manipulate ideas in our minds without looking at visual representations. People with Down syndrome often think in a very concrete manner and frequently cannot think well abstractly. Sometimes this can lead to frustration and misunderstanding.

Eugene was in the office for a complete evaluation, which lasts approximately three hours. After seeing the social worker, the audiologist, and the nutritionist, he was now seeing the physician. One of the standard questions asked is about appetite. As the time was approaching noon, Eugene's response was, "I am hungry." The question was rephrased in several different ways in an effort to understand Eugene's appetite in a general sense, but the answers only became more emphatic as to his present state of hunger. His mother explained his eating habits, and we moved on to the next question (and as quickly as possible, to lunch).

The concrete nature of the thought processes of most people with Down syndrome is very functional and can be very precise if allowed to flourish in an appropriate setting. Often people with Down syndrome do wonderfully when completing concrete tasks at work, when taking care of daily living needs such as meal preparation, and in self-care. In fact, we find that most often when a person with Down syndrome has problems at work, it is not because he can't do the tasks. Often, he does the task extremely well and is a model employee because his concrete nature helps him to do it well repeatedly. Eugene is the best employee in his mail service job. He is responsible for delivering mail in a six-story office building. He goes from the mailing center in the basement to the sixth floor and precisely moves from floor to floor delivering the mail. In a concrete world, he flourishes. His inability to answer the physician's question about his appetite—to think abstractly—has no real impact on his usual work or daily life routine.

The challenge for many people with Down syndrome comes when a task changes, and they must take what they have learned and apply it to a new situation, as in the example of Luis at the beginning of the chapter. This problem with generalization is one of many that can result from difficulties with concrete thinking and is discussed below.

Benefits of Concrete Thinking

Aside from the benefits of being able to function in a concrete world, there are can be huge benefits of thinking concretely. Perhaps the best way to describe these benefits is to look at how abstract thinkers may think and behave that get them in

trouble. We have all likely heard about people who are so driven to succeed that they sacrifice time with family and friends. What happens to these people? They may work on earnings, possessions, and accomplishments, and then realize only too late that other things in life are also important. These individuals pursue an abstract idea or concept of succeeding (what is often called "keeping up with the Joneses") that may limit their ability to appreciate the people and things in their day-to-day lives.

While we as abstract thinkers are off working and thinking of this and that concept, concrete thinkers such as people with Down syndrome are far more likely to appreciate the richness of the world right here and now. They may see and experience things that abstract thinkers may be too busy or too preoccupied to see and experience. As Martha Beck stated in her book Expecting Adam, one of the joys of Down syndrome is the ability to literally and figuratively stop to smell the roses (Beck, 1999). In fact, being present in one's life is the focus of almost every religious and spiritual tradition. Meditation, prayer, and yoga are all meant to bring people into the present moment and out of thoughts of the past or future. People with Down syndrome often have no problem being in the present moment; the rest of us have to practice over and over. (It should be clarified, however, that if a person is getting "stuck" on a bad thing that happened to him somewhere and is anxious about it happening again, he is not in the present moment.)

Taking a concrete approach to life may also make it far easier for people with Down syndrome to do certain repetitive activities (e.g., reorganizing their bedroom) that may help to take their minds off their troubles and concerns. Concrete thinkers may also enjoy activities that require sustained time and energy (e.g., painting) and can have profound benefits. We discuss this further in chapter 10 on "grooves."

Practical Effects of Difficulties with Abstract Thinking

Flexibility

Abstract thought allows us to understand the relationship between things and events and not just the individual (concrete) case. This allows us to problem solve—to view a situation in many different ways and imagine how changing specific variables could lead to different outcomes.

For people who rely primarily on concrete thinking, problems can occur when there is some need for flexibility to adapt to changes or disruptions to their normal plan or schedule. For example, as in the case of Luis at the beginning of this chapter,

many individuals with Down syndrome have difficulty adapting to unplanned disruptions to their daily bus ride (on regular public transportation) when there is a change in the bus route due to construction or a winter storm. They may react to the change by simply getting off the bus, not knowing where they are or what to do, much to the horror of family and other caregivers. Concrete thought ties you too closely to one thing—for example, "the route." When that one route is no longer available, it is difficult to imagine there being any other route. On the other hand, abstract thought allows you to understand the concept of a route, consisting of many or all routes, which allows you to consider other options when the one route is not available.

Lou Brown, a well-known teacher and advocate, gave another good example of concrete thinking. He described a training exercise in which people with Down syndrome and other intellectual disabilities were trained to get on busses using huge cardboard models of busses. Unfortunately, when they sent this group out into the community to get on real busses, they did not. What happened? Obviously, they were waiting for the cardboard busses to arrive.

Concrete thinking may limit flexibility and adaptability in many other areas besides thinking through and adapting to differences in routines. For example, if an activity is written on a calendar, it can be difficult for people who think concretely to imagine that this activity may not be possible (due to changes in weather, a medical emergency, etc.) and to then consider other possibilities. Consequently, the best way to deal with this difficulty is to return to the calendar to erase the activity from one day and rewrite it on another day. This concrete change to the calendar is easily understood and seen by the person, and life can go on.

Generalization Skills

Another major limitation to a concrete style of thought is that it makes it harder for someone to generalize behavior learned in one setting to other settings. For example, learning an important social skill, such as greeting customers appropriately in a supermarket, does not guarantee that the person will transfer this skill to a job at another supermarket or a different type of work setting. In these situations, the person may need to relearn the skill in the new environment.

Difficulties with generalization can also limit the transfer of essential life skills from the classroom to the community. For example, safety training is of great importance for people with Down syndrome who are venturing forth into community

settings. Often an individual might attend group sessions to learn safety strategies, such as not responding to strangers' advances, and may appear to master those skills in class. But parents and teachers are often dismayed to discover that the skill does not really transfer out of the classroom to a real-life setting. Consequently, it is essential for the lion's share of practice runs to occur in malls or other community settings. Parents and others should be included as important adjuncts to the training to practice what is called "in the moment training," which is simply working on the new skill in their son's or daughter's day-to-day life as learning opportunities present themselves.

In addition to having trouble with generalizing life skills from one setting to another, people with Down syndrome may struggle to apply academic skills learned in the classroom to real-life settings. In particular, the tendency toward weakness in abstract thinking can frequently result in problems where money or other math skills are involved. Here are two examples of common problems:

Joshua was learning about money skills in school. In the classroom, he could identify currency and its value and add bills and coins to get a total value. However, when shopping, he was not able to calculate the amount of money he had to know if he had enough to buy the item he wanted to buy. With additional training, including some done in stores, Joshua was able to look at the price of an item, look at how much money he had with him, and calculate whether he had enough.

Valerie, who is employed as an office assistant, is able to count quite competently. However, if she is photocopying a long document for a coworker and the pages get out of order, she is unable to correct the pagination herself. She usually ends up in tears if she struggles with the jumbled-up pages too long.

Valerie also has trouble generalizing her excellent reading and writing skills to help her perform her job. Often when she is having problems remembering how to do something, a coworker will prompt her to write down the steps involved. Afterward, Valerie can usually follow the written instructions and complete the task independently. However, it usually does not occur to her to write needed information down for

herself, even when it is something (such as state abbreviations) that she repeatedly asks her coworkers for.

Problems with generalization can lead to difficulties at work and school. Others may assume the person with Down syndrome is deliberately pretending he doesn't remember how to do something in one setting that he could do in another.

Time and Abstract Thought

Time is another concept that can be too abstract for many people with Down syndrome to fully understand. For example, most adults with Down syndrome can tell time, using either an analog or digital clock, and therefore know exactly when it is time for meals, favorite activities, TV shows, etc. However, they often don't seem to have a good sense of the passage of time. That is, they may not be able to internalize how long different lengths of time are.

The idea of "five minutes" or "five years" may be just too abstract, and as such, these time concepts may be confusing to some people with Down syndrome. For example, a new employee with Down syndrome might be told by the boss to "take a twenty-minute break." The employee with Down syndrome may come back after five or even thirty minutes because he does not fully understand what "twenty minutes" means. The way around this is to work with a more concrete concept of time. You can do this by pointing to a clock and saying, "When the big hand is on 12, go to the break room . . . when it is on the 3, begin to come back, and when it is on 4, start working again." Or if the person understands precise time on a digital or analog clock, tell him, "At 3:00 go; at 3:20 be back." In our experience, analog clocks and watches are preferable because people can actually see the hands of the clock progress. Most do very well with these adaptations and in fact are often better at returning on time than other employees because they are often more conscientious than other workers.

Time-related problems and misunderstandings may also occur if the person with Down syndrome has a tendency to use the present tense even when talking about things in the past or future. Here is an example:

Henry was a resident at a group home that held yearly progress reviews for clients with all relevant staff and family members in attendance. In one such meeting, Henry stated that he had been beaten by

another resident in the group home. Staff were incensed and defensive. They said that Henry was lying and making up stories to make them look bad.

Fortunately, Henry's mother was present and was able to resolve the issue. Three years previously (and before the current staff started working with him), Henry was the victim of a bully in his residence, who was subsequently moved. This bullying incident had been discussed and dealt with at just such a review meeting, but again, several years in the past. The present meeting seemed to have jogged his memory of the past staffing incident. Henry's mistake was to recount this event in present terms, as he did frequently, and as is the case for many people with Down syndrome.

Caregivers or professionals who do not understand this difficulty with using the past tense may believe the person is showing some serious pathology, such as a dementia-like memory lapse, a psychosis, or even (as in the example) telling a story or lie with some harmful intent. People who think concretely may also have trouble with future time, such as gauging when a desired activity will actually occur in the future. This may lead to the repetition of such questions as "We are going to (whatever the desired activity), aren't we?" Parents or other caregivers may answer the question, only to find that the same question is asked again and again. Caregivers frequently comment on the futility and frustration of these repetitive questions. Why does this occur? We think that the usual answers given may satisfy a person who thinks abstractly but not necessarily a concrete thinker, because it does not solve the vagueness of time for these individuals. For example, the typical answer will be "Yes, we will go to _____ later, tomorrow, next week," etc., which, to people with Down syndrome, may not sound like an answer at all. Just as the concept of time segments of five minutes or five years may be too abstract and meaningless, the concepts of later, tomorrow, or next week may also not make sense.

The person may then continue to ask the question because he hasn't heard a solid answer to his question. (It is interesting to note here that typically developing children often respond in the same way to these types of answers, because they too are concrete thinkers and also may believe (correctly or incorrectly) that this is a brush-off. For the person with Down syndrome, these types of nonspecific answers may actually activate another strong characteristic—a groove-like response, which

may continue until the question is resolved. That is, the person with Down syndrome may get compulsively stuck on the question until he hears a satisfying answer. One way to get out of this is to capitalize on the individual's strengths in visual learning and use visual cues to show the individual on a calendar when the event will actually happen. This often allows people to gauge time in a way that is more satisfactory. Calendars and other visual cues are discussed in more detail later in the chapter, as well as in chapters 6 and 7.

Misunderstandings Involving Abstract Language

Messages are often communicated in a concrete way, when, in fact, they are meant to be interpreted using abstract reasoning skills. We often encounter situations where people with Down syndrome are just "too literal." For example, Eugene, above, persisted in telling us he was not working because he did not understand that we meant in general, not at that specific moment. In another instance, a young woman with Down syndrome was working in an office and was encouraged to call "anytime" if she had questions. After she made a few 3:00 a.m. phone calls to the home of a coworker, the true meaning of "anytime" had to be clarified.

Another office worker had trouble with tasks like these, which require abstract language skills:

- answering the phone and differentiating telemarketing calls—which should be screened out—from legitimate calls—which should be transferred to the appropriate coworker;
- understanding that when colleagues asked her what she did on the weekend, they did not want to hear a lengthy hour-by-hour report but a quick summary;
- recognizing and weeding out duplicate names when entering customer names into a computer program used for generating mailing lists (e.g., realizing that Thomas Dooley, T. Dooley, and Tom Dooley are probably one and the same person, especially if they live at the same address).

These kinds of misunderstandings are often the source of significant problems in the workplace. See chapter 7 for more information on communication issues.

Misunderstandings Involving Humor

Many people with Down syndrome have a wonderful sense of humor, particularly around good-natured teasing of and by favorite family members or friends. Still, we have also found that some people have difficulty with some types of humor due in part to the reliance on concrete forms of thought. It may be difficult to "get" the punch line of certain jokes that are too abstract or that have too many words or concepts that they do not understand. In these situations, people may feel they are being laughed at or the butt of the joke. Teens and adults with Down syndrome may also feel as if they are being laughed at if they inadvertently say or do something that is funny to abstract thinkers. For example, we were careful not to laugh at Eugene when he stated that he was not working at that very moment. These sorts of comments may be genuinely funny and even charming to family and friends but may not be funny to the person with Down syndrome.

There are some types of jokes that people with Down syndrome do "get" extremely well. In fact, many enjoy humor that they can see. Examples of visual humor include comedy such as *High School Musical*, *Glee*, *Mama Mia*, and *Grease* and TV shows such the *Three Stooges*, *Scooby-Doo*, and professional wrestling. We include professional wrestling in this category not because it is meant to be comedic but because most people recognize it as campy and "larger than life" theater that is just pure fun. It is interesting to note that despite the aggressive nature of the Stooges and wrestling (which often includes slapping, hitting, punching, insults, and verbal taunting), we have found an almost total absence of mimicking physical aggression by the people with Down syndrome who watch these shows. This is true, even though people with Down syndrome are superb imitators and if they wanted to copy the aggression seen in these shows, they could certainly do so. We think that people with Down syndrome get the joke—that they know it is drama and theater and not reality.

What Helps

What may seem to be an obvious logical next step when thinking in abstract terms may not be obvious to someone with Down syndrome. This can lead to misunderstandings, communication difficulties, and even "behavioral issues." (Either the inappropriate response is interpreted as a "behavior problem," or the person with Down syndrome may become frustrated and his behavior may change.) If the task is

broken down into concrete steps, however, the person is often quite capable of managing the tasks with little, if any, confusion or "behavioral issues."

Visual Supports

We have found that one particularly beneficial way to assist many people with Down syndrome make sense of abstractions is to develop visual supports for them. We discuss this in much greater detail in the chapters on visual memory and communication. Interestingly, even some of our patients who generally function independently and would seem to not need this assistance may benefit from visual supports. For example, coming to the doctor's office frightens some of our patients. We have made a book with pictures and a video of a visit to the office. As the person looks through the book or views the video, it makes it less abstract and more concrete, and generally, less frightening. Likewise, to assist people with Down syndrome in taking more responsibility for their own care, we have found it helpful to provide illustrated patient education materials that guide the person through self-care.

As discussed in chapter 16, we have also successfully used modeling to teach adaptive behavior. Videos of the person himself doing the desired behavior (Buggey, 2009) can be a particularly powerful tool. When a person with Down syndrome watches a video of himself, it reinforces and promotes the desired behavior. Viewing videos of individuals with similar characteristics—for example, other people with Down syndrome (peer-modeling)—is also beneficial.

While it may be initially more time consuming to make visual supports, it does give the person with Down syndrome more opportunity to direct his own care. It also reduces the confusion that can occur when directions are only provided verbally. In the long run, it will probably also save time in completing daily tasks.

Helping the person with Down syndrome work through issues related to concrete and abstract thinking (and, thus, help him be more independent) is extremely important in fostering independent-living skills. Being successful in self-care can lead to a great sense of accomplishment, improved self-esteem, and increased interest in caring for oneself. A personal sense of good health is an important part of mental health. In contrast, confusion about what is being asked or is expected can lead to an escalating set of problems. The loss of a job, the loss of self-esteem, and other negative effects can lead to more severe mental health issues.

In health promotion, the ability to understand (health literacy) and participate in one's own health also improves health outcomes (Parker, 2003). Clear communication, plain language, and visual communication are essential aspects of improving health literacy (National Institutes of Health, 2017) to promote personal health. For people with Down syndrome, pictures and videos are particularly helpful ways to convey the information.

We can use the person's visual strengths to teach a wide range of adaptive skills despite a deficit with abstract thinking. But we usually need to manipulate the images for people with Down syndrome and not depend on them to do so on their own. Chapter 6 on visual memory offers additional strategies beyond those discussed above that can be useful in helping people with Down syndrome understand abstract concepts.

Conclusion

Problematic issues related to concrete thinking are very important to recognize, particularly when it comes to promoting health and well-being. In a world that demands both concrete and abstract thinking, it is very important for those assisting a person with Down syndrome to consider these issues. The goal is to present the tasks of daily life in a way that capitalizes on strengths in concrete thinking but does not penalize the person for limitations in abstract thinking.

Return to Table of Contents

Chapter 6

Visual Memory and Cues

Samantha, age nineteen, came to us for an evaluation. Her mother explained that they were "butting heads" on a regular basis. She said Samantha would refuse to do as she was asked despite repeated requests. There were a variety of tasks, including taking care of her own hygiene, that Samantha was not doing on regular basis. Samantha's mother wanted her daughter to do the tasks without having to ask repeatedly, and Samantha wanted her mom to stop repeatedly asking.

We discussed adolescence issues (see chapter 11) and also that people with Down syndrome tend to be visual learners and to have stronger visual memories than auditory. We recommended a picture schedule and calendar to help Samantha remember and independently perform the desired household chores and self-care tasks. Her mom said they had used visual cues with Samantha when she was a young child but didn't think they would be necessary as she moved through adolescence to adulthood. However, she agreed to give it a try. Upon returning for a reevaluation a few months later, both Samantha and her mother expressed satisfaction with the progress they were making toward achieving their desires for the tasks to get done without constant verbal reminders.

There is a large body of research suggesting that people with Down syndrome have "auditory deficits." What this means is that they often have difficulty remembering and then acting on what they hear or are told to do. On the other hand, people with Down syndrome very often have exceptional visual memories. As a result, they tend to remember many things they see and are very good visual learners. Therefore, although people with Down syndrome may easily forget what they hear, they are less likely to forget what they see, which can be extremely useful in promoting adaptive skills and problem-solving strategies.

Because of their relative strengths in visual processing, people with Down syndrome can benefit greatly from visual supports. A visual support uses pictures, written words, or both to help prompt or remind an individual to do something. Most

of us actually depend on visual supports in our daily lives. For example, we use calendars, written shopping lists or to-do lists, schedules, diagrams, and other visual supports, whether on smartphones, computers, or paper. All too often visual supports are used for children with Down syndrome when they are in school, but then they are phased out in adulthood. Adults with Down syndrome can benefit from such supports just as much, if not more, than other adults, though, as discussed in this chapter.

Visual Dimensions: Visual Memory and Visual Cues

We find there are two important dimensions to capitalizing on the visual strengths of people with Down syndrome: (1) what visual images people remember (their visual memory) and (2) what people see and respond to (visual cues). We came to realize how important visual memory and visual cues are to people with Down syndrome through a number of discoveries. First, regarding visual memory, we heard from many families that their sons or daughters had exceptional memories, but the descriptions of what was remembered were almost always something visual. When we looked more closely, we were astounded to find this visual memory was often photographic-like. Many people with Down syndrome seemed to remember people, places, and events in vivid detail, particularly events that were of importance to them. Equally important, time seemed to have little effect on these memories. Events from the distant past (ten-plus years) could be recalled in the same detail as events from early in the same day. Related to this, we found that people with Down syndrome often seemed to be able to take in movies “whole” and then mentally replay them at will, to entertain themselves or to fill free time.

We also noticed something about this visual memory that was even more striking: people with Down syndrome would often remember past events as if they were happening all over again, with the same feelings and emotions of the original event (especially events involving strong feelings or emotions). We learned that what triggers a relived memory may be something simple or seemingly small in the immediate environment, such as an object, room, or piece of clothing worn during the original event.

We have discovered that both visual and auditory cues can encourage the recall of a past event. Pictures of the original event obviously have great power, but a story-like description by another person can also trigger or set off the memory. Time and time again families have reported experiencing this with family members with Down syndrome. For example, the brother of one of our patients reported that when he

displayed a picture from a vacation that occurred over fifteen years ago, his brother with Down syndrome not only remembered the vacation but also seemed to recall the whole experience in vivid detail. Sometimes just hearing someone else reminisce about an experience can lead to the person with Down syndrome replaying the memory in her mind, complete with the original emotions she felt at the time.

Five older teens who were quite well adjusted were participating in a series of classes meant to teach them adaptive skills in the community. In the middle of one evening class, a participant stated that his dog died, and this threw the group into absolute pandemonium as all group members expressed strong feelings of grief over some experience of loss that had occurred at some point in their own lives. This horrified and mystified the leaders of the group, who could offer no explanation to the parents who were waiting for their sons at the end of the class. Fortunately, we were able to clarify to family and teachers that it was not harmful or unusual for these students to relive old memories. We call this "in the moment" memory, and it involves the intense replay of an original memory, but this memory is usually over quite quickly if the original event was not so severe. This proved to be the case with these young men, who were back to normal within moments.

On the other hand, replayed memories can be quite a problem if the original memory is of a strong negative or traumatic experience. Given the ability of people with Down syndrome to relive past events, we were not surprised to find that they are likely to be more susceptible to post-traumatic stress. Additionally, we believe there is a higher rate of phobia in people with Down syndrome. This may be because when a person has a frightening or terrifying experience, such as with a storm or an animal, she tends to relive the original fear when she encounters the same stimuli again. Interestingly, we found that even if (or perhaps because) this frightening experience occurred when the person was young, the phobia often persists throughout the individual's life. We discuss strategies for dealing with this problem in chapter 18.

These findings led us to look very closely at the effect of visual images on people with Down syndrome. For example, we thought one way to deal with harmful effects of negative images would be to substitute positive images, something analogous to "fighting fire with fire." We assumed that the one thing strong enough to combat the effect of a negative memory was another memory that elicited strong positive feelings and emotions. At first our interest was pragmatic—to identify and neutralize the effect of negative images and memories—but this led us to look very closely at visual images and stimuli in general.

We learned that people with Down syndrome tend to be attracted to anything visual. This may include movies, DVDs, photographs, and visual images seen in their immediate lives (such as scenery viewed on a vacation). In our presentations, we tell families that grooves (see chapter 10) may be "onboard equipment," but "visual is king." Despite having auditory deficits, when someone with Down syndrome remembers a scene (for example, a family gathering, or a favorite movie), she can often remember what people said. Additionally, the love people with Down syndrome have for visual stimuli often extends to live theater and role-play. Not surprisingly many enjoy acting. In other words, teens and adults with Down syndrome are very receptive to visual images or cues, and we can use this to promote adaptive functioning and artistic expression and to solve problems that occur.

A second reason for our interest in visual cues developed from our work with individuals with Down syndrome who also were diagnosed with an autism spectrum disorder (ASD) (see chapter 25). The benefits of using visual supports with people with autism spectrum disorders are well known. We have incorporated many of the visual strategies used for people with ASD with people with Down syndrome and found them to be effective for most people with Down syndrome, regardless of whether or not they had an autism spectrum diagnosis. In short, visual cues can be an extremely effective technique for assisting people with Down syndrome. We discuss our experience with these strategies throughout this book and also look at how these strategies can be used in conjunction with other behavioral characteristics to create even more effective and lasting methods of helping people with Down syndrome.

Dependence on Concrete Thought

Despite strengths in visual memory and a great capacity to learn from visual cues and supports, at times the benefits may be relatively limited. One reason is because people with Down syndrome depend on concrete rather than abstract forms

of thought (see chapter 5). This may prevent them from learning from past experiences stored in long-term memory, since you need abstract reasoning to see the relationship between things or events. In the absence of abstract reasoning, the person is often not able to use a past visual memory to help deal with her current situation.

If you went to a restaurant intending to order the special chocolate cake for dessert but discovered later that the restaurant was out of chocolate cake, you might be disappointed, but you'd be able to solve the problem. You might picture yourself losing weight (or at least not gaining weight) if you just virtuously did without dessert. Or you might think about the remaining dessert choices and choose one that you remember enjoying in the past. Someone with Down syndrome, though, might just get upset—unable to let go of the idea of eating her favorite chocolate cake and to imagine herself eating any other dessert. Or in a more serious situation, you might lose your wallet while out shopping. In this case, you would probably mentally retrace your footsteps and then either call the stores that you visited and ask if a wallet was turned in or physically revisit the stores. Someone with Down syndrome, however, might not be able to independently figure out these steps in tracking down her wallet, even though she could likely visualize each place that she went if asked to.

Now, we can still use the person's visual skills to teach a host of adaptive skills, but we often need to manipulate the images for people with Down syndrome and not depend on them to do so on their own. This may include using concrete visual cues such as checklists, calendars, role-play, etc.

What may seem to be an obvious logical next step (if one has abstract thought) may not be obvious to someone with Down syndrome. This can lead to misunderstandings, communication difficulties, and even "behavioral issues." (Either the inappropriate response is interpreted as a "behavior problem," or the person with Down syndrome may become frustrated and her behavior may change.) If, however the task is broken down into concrete pieces that can be seen, however, the person is often quite capable of managing the tasks with little, if any, confusion and "behavioral issues."

Visual Supports to Solve Routine Problems

We have found that one particularly beneficial way to assist people with Down syndrome make sense of abstractions is to help them visualize the tasks or upcoming activities by providing pictures. We also find that schedules (typically using words and

pictures) are quite helpful, both in helping the person cope with routine situations and in dealing with less common problems that may occur.

Supports to Help the Person Remember and Follow Routines

When your child was growing up, you (or his teachers) may have used a variety of visual supports to help him stay "on task," follow a routine, or keep track of upcoming events or important dates. Most people with Down syndrome (like other adults) do not outgrow the need for visual supports or reminders as they grow older. Especially as they enter the workforce and/or leave the family home to live on their own or semi-independently, they may need new types of supports or more "adult" versions of supports they used when younger. Below are examples of types of supports that may help teens and adults with Down syndrome handle routine situations:

Visual Schedules and To-Do Lists

Visual schedules and to-do lists can help people follow the steps in a routine, learn a new skill, or complete a series of tasks. They can be designed using sketches or photos from the internet or magazines, or they can be illustrated and personalized with photos of the person herself. For people who feel stigmatized by using something that makes them look "different," schedules can be small and unobtrusive (contained on a smartphone or tablet).

Although many people with Down syndrome love routines and easily learn their own daily routines, visual schedules may be especially helpful when the person is first learning a routine or needs to follow a known routine in a new setting. For example, an adult who knows how to get ready for bed might struggle with following through on actually going to bed when her parents are not around to enforce bedtime. Breaking the nighttime routine into a schedule of concrete tasks can be very beneficial. We encourage the person with Down syndrome (with the help of his family or care provider) to select approximately five or six tasks that need to be done before bedtime. These tasks are selected from a series of pictures. The picture corresponding to each task is then pasted on a small piece of cardboard in sequential order. For other people, a system that allows them to check off boxes as tasks are completed is a better way to manage the bedtime routine. Interestingly, even some of our patients who generally function independently and would seem not to need this assistance may benefit from this type of system.

Checklists

The written word is visual and can be concrete, and thus it can be understood and accessible to many people with Down syndrome. Therefore, written words can be used in many proactive ways to help people learn new skills or adapt to new settings or new tasks. For example, checklists or a list of a series of tasks can be used to increase independence when doing self-care, choosing what to eat or drink, or interacting with others. These checklists can be changed in response to the person's age, maturity, and the developmental task at hand, as well as the person's current wants and needs, and pictures can be added for clarification.

Calendars

Calendars are highly valued and closely monitored by most people with Down syndrome because they are concrete and visual. They allow the person to see what her life and upcoming schedule look like, which is of great importance to most people with Down syndrome. We have found that once the activities are written on the calendar the person with Down syndrome will remind the caregiver, and not the other way around. Using a calendar to communicate schedule changes can be very helpful in avoiding power struggles and in helping the person accept the change. Again, processing auditory input is not a strength. Going back to the person's preferred calendar (on the wall, tablet, or smartphone) and noting the change is far more likely to be understood and accepted.

It is interesting, too, that pictures and written words on calendars can resolve problems related to adults exercising their right to say no to activities they might actually enjoy and benefit from. For instance, problems developed in one group home after Illinois state administrators mandated that adults with Down syndrome (and other disabilities) who lived in group settings had to be asked before engaging in any outside social activities. To administrators, this undoubtedly sounded like a good way to support independence, but what happened was that many people were enticed by the power of saying no. Consequently, the staff would ask and would often hear "no" even in response to requests to do activities that the person actually loved and highly valued. This resulted in many people staying home rather than going out to have fun and get needed exercise. We suggested that residents use calendars to write down the weekly and monthly social activities. Afterward, in most cases, there was no need

for staff to ask if there was interest in doing an activity. If the activity was written on the calendar, the person with Down syndrome would usually remind staff.

Videos and Video Modeling

Video can be a powerful medium for visual learning. As mentioned previously, most people with Down syndrome are quite enamored by all manner of visual images, including videos and movies. Video can be used as a particularly effective means to teach tasks. Appropriate videos may be found on YouTube, as well as on sites specifically for people with intellectual disabilities and autism spectrum issues. Examples include Watch Me Learn and the Adult Down Syndrome Center (https://adscresources.advocatehealth.com/video). Personalized videos can also be made to teach a skill. For example, it might be helpful to make a video highlighting particular job activities or showing how to do tasks such as loading a dishwasher or using a sleep apnea device.

Video learning may be especially useful in helping a teenager or adult with Down syndrome manage social skill issues such as fraternizing with or hugging customers when on the job. If possible, it is best to reinforce the video lesson with role-play practice. In some cases, it may be more effective to use "self-modeling"—making a video of the person herself engaged in some task or activity (Dowrick, 1991).

Time Management Supports

Visual cues can be used to assist people with a variety of time management issues. For example, there are apps that can be set up to show how much time is left on an analog clockface before the person has to move on to another task. For example, the clockface (sometimes coupled with a beeping mechanism) can show how much time remains to play a video game or dress before it's time to move on to breakfast. Incentives can be added to reinforce the person for heeding the reminder (e.g., earning a simple reward for arriving at breakfast on time). Reminder signs or graphics, perhaps with incentives attached, can also be used. Such reminders may work better in a work setting, for example to help with the transition between tasks. When verbal reminders about time are a source of conflict between the person with Down syndrome and others—such as when the person needs to stop an enjoyable activity and move on to something less preferred—having an app or other visual aid provide the reminder is often a good way to defuse the problem.

Visual Supports for School Settings

It is beyond the scope of this book to describe supports at school in any detail since our main focus is on adult issues. However, in school settings visual supports that build on strengths are essential for people to survive and thrive. Teaching materials that are adapted with visuals are far more likely to be effective. On the other hand, when teachers complain of "inattentiveness" or "distractibility," the problems may simply be because instructions are primarily spoken, and auditory processing in not a strength for people with Down syndrome. Behavior problems are also not uncommon when students are left out and left behind. They may try to let people know that they are not doing well through a change in their behavior. Of course, the longer this goes on, the more demoralized and behind the student gets.

Fortunately, most teachers of students in grades kindergarten through high school are aware of visual supports, but they may need to be reminded that students with Down syndrome can benefit from them just as much as students with autism spectrum do. In postsecondary programs parents may need to educate teachers about visual supports that worked for their son or daughter as a younger student.

Supports to Prepare the Person for New Experiences or Potential Problems

Visual supports can also be very helpful in preparing a teen or adult with Down syndrome for a new experience or to help her problem solve in a situation where abstract reasoning skills are called for. These types of visual supports are a bit trickier to develop, though, since the parent or other caregiver needs to anticipate a problem before it occurs. Here are some types of visual supports that can help in these types of situations.

Illustrated Books or Stories

When a person with Down syndrome is facing a new experience that is worrisome, it can be very helpful to preview what will happen using visual supports. You may be able to find publications or information on websites to show the person what will happen in a general way beforehand.

Coming to the doctor's office frightens many of our patients. We have made a book with pictures of a visit to the office and a video (ADSC, 2016). As the person

looks through the book or watches the video, it makes the upcoming visit less abstract and more concrete, and usually, far less frightening. Likewise, to assist individuals with Down syndrome in taking more responsibility for their own care, we have found it helpful to develop patient education materials that have pictures that guide the person through self-care.

Whatever the new experience the person might be facing—starting a new job, going on a trip, attending a new school, or having a medical procedure done—you can help prepare the person for the experience by looking at illustrated brochures or perhaps finding a library book on the topic to review with her.

Personalized Books/Stories and Social Stories

If you have the time to create a story showing the person with Down syndrome the exact surroundings, people, and activities she will encounter during a new experience, that will be even more helpful in preparing her. While it is initially more time consuming to make personalized books or stories, it does give the person with Down syndrome more opportunity to visualize herself in the new situation and understand what will happen there. Including information in this book to help the person with Down syndrome work through issues related to concrete and abstract thinking (and, thus, help her be more independent) is extremely important.

Social Stories are personalized supports that are often used to prepare people with autism for new experiences or to help them understand the expectations for behavior in a particular situation. Their use was pioneered by Carol Gray, and you can learn about making and using them in Gray's books and on her website. (https://carolgraysocialstories.com). You can also loosely adapt the technique by taking pictures of the individual who needs guidance and writing your own story. Here is an example of how personalized books can help:

> *Colton, a young man with autism and Down syndrome, was to accompany his mother on a vacation trip to visit his aunt thousands of miles away. This seemed like a daunting task at first, but then his mother thought that if she could get him to the dentist with the use of pictures, she could get him to go anywhere. She painstakingly put together a book with pictures for each step of the trip (from leaving from the airport and everything in between). After the vacation, Colton's mom shared pictures*

of him sitting on the beach with a big smile and demeanor that demonstrated this strategy paid off. Colton knew every detail of the trip ahead of time because he was able to follow the steps in his picture book as it happened.

Photographs

Sometimes you may not have the time to create a story about a new event but may still hope to prevent a foreseeable problem. For example, you might want to prepare an individual with Down syndrome for the possibility that her favorite food won't be available when she dines out. If so, you might visit the website together and look at the menu and photos of entrées and desserts available, talking about which ones you both like and dislike. Or you might want to preview the type of hotel room your family will be staying in on a trip. For example, a family from another state shared that they wanted their son with Down syndrome to accompany them to Disney World on New Year's Eve. In daily life, even going to the grocery store could be overwhelming for their son, so it seemed impossible to think he could manage the crowds and all the sound and fury of the night at Disney. His parents previewed the experience for him by outlining the entire trip via pictures of the anticipated events (getting on the plane, going to the hotel, the parks at Disney, fireworks, etc.) on his iPad, and he adapted splendidly.

If the person with Down syndrome is making a major transition such as moving out of the family home, photographs can play a role too. For example, you can gradually expose her to the house, the roommates, and staff "virtually" through pictures at first and then actual visits for increasingly longer time periods. Likewise, if a sibling will be going away, you might prepare the person with Down syndrome by showing her photos of the place her brother or sister will be going to and then perhaps visit the sibling there. For example, if the sibling goes off to college, look at photos of the campus and then perhaps visit the dorm or apartment where the sibling lives and the campus where he or she walks to class. Otherwise, the person with Down syndrome has no images to see and understand where her sibling is. It is as if the sibling disappears into a vacuum. Having an image allows the person with Down syndrome to maintain some type of object permanence, which makes all the difference in the world. You can follow up with Zoom, Skype, or FaceTime sessions to

allow the adult with Down syndrome to see her sibling and support a sense of continued contact.

Role Play

We have found that there is an effective way to teach more subtle rules and social roles to teens and adults with Down syndrome, and that is through role play.

How do you do role play? The parent or teacher plays the role of one type of person (e.g., authority figure, customer, friend), and someone else (e.g., the person or people with Down syndrome or someone who knows how to behave "correctly") plays the other role (or roles) in the desired situation. You set up a situation, such as interacting at a dinner party, and then demonstrate the right way to interact, wait until everyone is served, etc. Afterward, you have the person with Down syndrome try to do it correctly as well. Gentle guidance away from incorrect behavior and encouragement and praise for the parts the person does correctly can lead them to the desired behavior. We generally recommend only showing the correct way because if you also demonstrate the wrong way, the person may remember both ways but have a difficult time remembering which is wrong and which is right.

Role play capitalizes on the often-exceptional ability of people with Down syndrome to visually take in their environment in order to learn from it. We found that this happens even when people are engaged in role play and not just watching it. People with Down syndrome may be able to take in the scenes they enact in a role play "whole," just as many of them can remember verbatim favorite scenes from a movie and from their daily lives. They learn to be discerning about the rules of the setting by repeatedly acting them out.

Role Play at School or in the Community. We have seen role play successfully used to teach people with Down syndrome to not hug or fraternize with the other employees, customers, or even the boss. Just as importantly, they can learn that when the boss says, "Come see me" or "Call me any time," it's an expression and not a real request like it might be when with friends.

In explaining this need to practice interacting with others, we use the analogy of jet pilots who repeatedly practice what to do in the event of a catastrophic engine failure. Jets are heavy pieces of metal that fly at great velocity, and pilots have precious little time to do what they need to do to save their own and others' lives when a crisis occurs. Practicing survival strategies over and over allows them to override instinctive panic and impulsive actions, which could easily end their lives.

Similarly, for people with Down syndrome, survival in a work or community setting depends on overriding impulses that are inappropriate to the setting. The research is clear about this: people with Down syndrome and other intellectual disabilities don't usually lose their jobs because they don't do their job duties. Instead, when they lose their jobs, it is often because they break the rules of social engagement.

Now, one major difference between people with Down syndrome and jet pilots is that the former often have problems with generalization (see chapter 5). It's not enough for them to practice job-related behaviors outside of the workplace (such as in a school or agency setting). Practice must eventually be moved to the actual setting. Role play in the actual setting is no different than in a separate setting. The person with Down syndrome needs to go through the steps of responding to bosses and other employees in ways appropriate to the work setting. Ideally the actual boss and other employees would be involved and prepped to assist. If that is not possible, it can be helpful to do the role play in the setting with a job coach or other assistant. Once adults learn have to act in the workplace, they are quite conscientious about abiding by the rules, in part because they want to please, but also because that is the set formula or routine that they then feel comfortable following.

Safety Training and Role Play. Role play can also be useful in helping the person learn how to appropriately define her boundaries and protect herself from possible predators or others who might want to take advantage of her. Learning how to use verbal and nonverbal communication to assertively indicate "no," "stop," "go away," and the like is incredibly important for survival in the community. People with Down syndrome usually also need to be explicitly taught how to request or call for help. So, for example, it can be helpful to role-play scenarios in which they need to approach another person or a police officer to ask for help or dial 911 and report a problem. Practice makes perfect, but again, the practice should ideally occur in the actual setting (community, school, or work) in order to make certain it sticks.

Backup Plans

If you anticipate that an adult with Down syndrome may have a particular problem in a certain situation or if she has already experienced a confusing situation, you can create a written or visual backup plan to help her with problem solving. For example, many people with Down syndrome who take public transportation have backup plans consisting of pictures and words to guide them through different types of changes or disruptions in service that might occur. Smartphones in particular can

be very helpful in the event of an emergency or change of plans, since they enable phone communication to be made visual. Adults with Down syndrome can use FaceTime or similar apps to make video calls when confronted with a problem they can't solve on their own and—particularly if speech or reading is an issue—they can take photos of relevant buildings or signs nearby and text them to others if they get lost or do not understand written notices they encounter.

Return to Table of Contents

Chapter 7

Communication

Hunter, age twenty-three, was about to lose his job when he came to us for an evaluation. He sometimes was not completing the tasks he was asked to do, and this was considered a behavioral problem. When asked to do the task, he would eagerly set off to do it but sometimes would stop mid-task and not complete it. His supervisor did not understand that Hunter had wonderful receptive language skills (he understood what he had been asked) but limited expressive language skills (he had difficulty expressing himself). It was discovered that when Hunter stopped midtask, he typically had come across a problem he had not previously encountered and didn't know how to handle. He was unable to tell anyone about the problem or seek assistance, so he just stopped, confused as to his next step.

Once it became clear why Hunter was not completing assigned tasks, a system was set up that included his supervisor watching Hunter to see when he was confused and a method for Hunter to communicate that he had stopped working. Then, appropriate assistance was provided to help Hunter understand what to do. Over time, this successful strategy helped him gain confidence and actually reduced the times he had to stop in the middle of a task.

There is perhaps nothing so vital to us as human beings as our ability to communicate our thoughts and feelings effectively to others. Communication involves two key dimensions: (1) our ability to express our thoughts and feelings to others (expressive language), and (2) our ability to understand what is being communicated to us (receptive language). Expressive and receptive language go hand in hand. Our ability to respond or to communicate effectively with others requires that we understand first what they are communicating to us. We may take our communication abilities for granted, but communication skills cannot be taken for granted for adults with Down syndrome. We have found that many people with Down syndrome have significant problems with both expressive and receptive language, and this may greatly affect their understanding and ability to function in the world.

For people with Down syndrome, there are three major difficulties that may help to explain problems with receptive and expressive language:

- **An auditory deficit**: The person has difficulty with taking in, understanding, and remembering information he hears (or auditory input).
- **Concrete thinking**: This means that hearing and understanding can be even more difficult if others talk faster than the person can comprehend or in terms that are difficult to grasp because the concepts are too abstract (see chapter 5).
- **Slower processing speed**: Compounding the other two issues is a slower processing speed, which means the person often needs more time to understand what others say, even when the concepts are not too abstract.

Receptive Language

As mentioned above, our ability to respond or to communicate effectively with others requires that we first understand what they are communicating to us. At each step of development, and for many activities in our lives, our ability to benefit from the instruction, guidance, and wisdom of our teachers, caregivers, bosses, and coworkers depends on whether we truly understand what others are communicating to us.

Teenagers and adults with Down syndrome usually have better receptive than expressive language. As a result, they often understand more than they can express. This can lead to a variety of problems, especially if the person's wishes are not taken into consideration or if he is patronized or talked down to by people who are not aware of the gap in receptive versus expressive language skills. Sometimes what is perceived as a "behavior problem" is actually an attempt by the person to get his message across or an expression of his frustration at not being heard.

Strengths and Weaknesses in Receptive Skills

Despite the language limitations that people with Down syndrome often have, their receptive skills can be exceptional. This may be due to their strengths in visual memory. We have heard time and time again from families that their son, daughter, or other relative with Down syndrome seems to take in and remember many important things they see in their immediate environment. One problem with this

ability to take in and remember things they witness is that they do not always have the ability to understand or interpret what they are seeing. For example, they might witness a heated debate between family members about a political or economic issue. Even though they understand most of the words spoken, they don't understand the crux of the disagreement because they don't comprehend how the issues might affect them and other people they care about. Likewise, people with Down syndrome may be able to memorize advanced math formulas or the words describing a science concept but be unable to understand what they mean—no matter what visual supports are used—because they do not have the intellectual ability to do so. That is, there are some very real limits to language comprehension in most people with Down syndrome that can't be surmounted with visual supports.

Pragmatics

In contrast to the areas of receptive language in which people with Down syndrome struggle due to cognitive challenges, there is one particular area in which many teens and adults excel, as long as they are given sufficient support. This area is what speech-language pathologists call "pragmatics." Pragmatics refers to the social uses of communication and involves understanding and using gestures and facial expressions as well as spoken language.

Initially people with Down syndrome may have difficulty differentiating the rules of how to behave in different environments due to problems understanding the more subtle rules of social engagement. In particular, they may not comprehend how appropriate behavior differs in school, work, or community settings than in more informal contexts such as a bowling outing or a family gathering. For example, in a school, work, or community setting, should you greet others with a hug as you might in a social gathering? The truth is, the wrong behavior in the wrong setting can get you fired or create other problems.

Sometimes teens and adults exhibit problematic behavior at school, work, or in the community because others have inappropriately reacted to it as "cute" in home or social settings, inadvertently reinforcing the behavior. We have seen a number of people laid off from good jobs for behavior that was tolerated in their homes but not in a work or community setting. Examples of inappropriate behavior resulting in dismissal include "stealing" food for snacks in a grocery store, taking others' food from a refrigerator in the break room, flirting with others, and even slapping the backside of a fellow employee. As discussed in chapter 6, visual supports can be very

useful in helping people with Down syndrome learn the specific behavioral expectations in any given setting, including the subtle rules of socializing with coworkers or customers at work and handling interactions with strangers in the community.

Expressive Language Limitations

Expressive language is the communication of one's thoughts and feelings, through verbal and nonverbal language as well as the written word. Although many people with Down syndrome are quite adept at communicating with gestures and through other nonverbal means, speech is often a stumbling block. In particular, teens and adults often have trouble with intelligibility, or producing speech that is understood by others. Adults and adolescents with Down syndrome have a wide range of intelligibility, from those who are highly verbal and intelligible, to those who have no speech at all, but the vast majority of people with Down syndrome fall in the middle: they use speech as a primary means of communication but have some difficulty being understood by others, particularly unfamiliar others.

In the first edition of this book, we discussed a survey we conducted of 579 people in the Adult Down Syndrome Center. We found that 75 percent of caregivers reported that they were able to understand the person with Down syndrome in their care "most of the time." Only 28 percent of the adults with Down syndrome were reported to have speech that was understood by strangers most of the time, while 40 percent of them could be understood by strangers some of the time, and 32 percent very little of the time.

Table 7-1. How understandable the person with Down syndrome is to familiar and nonfamiliar others, as estimated by caregivers

	Most of the time	Sometimes	Very little
Understood by familiar others	432/75%	74/12.7%	73/12.3%
Understood by strangers or unfamiliar others	161/28%	233/40%	185/32%

Overstating Intelligibility

Since the first edition was published in 2006, we have had many discussions with families about our findings on intelligibility. Many family members have told us that regardless of how intelligibly the person with Down syndrome is capable of speaking, he is often reluctant to speak, even to close family or significant others, and even if he is quite intelligible to these others. Many caregivers have told us what happens when they ask their family member with Down syndrome routine questions such as what he did at work, school, or on an outing. The usual answer they get is only a shrug of the shoulders, a blank stare, or at most a curt "I don't know" or "stuff." This happens not just with general questions but even more specific questions such as "Were you able to take out the garbage today?" or "Did you finish your collage?"

What is going on here? We believe that regardless of how intelligible or how close they are to caregivers, many people with Down syndrome are simply not comfortable with spoken exchanges. We call this "taking the Fifth" (a reference to the Fifth Amendment of the US Constitution, which allows one to protect oneself from self-incrimination). The best explanation for this appears to be that they feel vulnerable when speaking. They are simply not on a level playing field and the risk of saying something wrong or that is misinterpreted or judged in some negative way appears to be very high, even by the most patient of parents or caregivers.

Please recall that the three issues discussed previously—auditory deficit, reliance on concrete thinking, and a slower processing speed—have a major effect on receptive skills, but they also have a profound effect on an individual's motivation and ability to speak. Many simply do not feel confident or safe when speaking to others.

In the first edition we suggested that the person with Down syndrome is not unlike a visitor to a foreign country who cannot speak the native language. The analogy was drawn from the first author's experience with speaking Spanish while visiting his late wife's native country of Argentina. Self-consciousness is a common problem both for people with Down syndrome and people speaking a foreign language. Both groups may be concerned about how they sound or how long it takes for them to formulate their ideas. This may make them hurry or force their speech, which may then interfere with the successful articulation of their thoughts. In turn, this may make them less intelligible to the listener, making them feel even more apprehensive. Over time, an unproductive pattern may develop in which the person

with Down syndrome, or the person trying to speak a foreign language, becomes more and more reluctant to speak, even to close family and friends.

In addition, teens and adults with Down syndrome also have other speech issues or deficits that create hurdles to successfully conversing, and these difficulties can easily aggravate their self-consciousness. For example, many individuals stutter or get stuck repeating the first word or phrase. Many also tend to speak at a low volume or speak too quickly or too slowly.

Finally, we believe that some people with Down syndrome tend to think in pictures (as explained in chapter 6). Therefore, in order to communicate a thought or an idea, they may need to translate back and forth between a visual image and a verbal statement. This may actually be very close to what it takes to translate back and forth from one's native language to a foreign tongue. Now, this is both good and bad news for our discussion. The bad news is that many people with Down syndrome have difficulty verbalizing, but the good news is that they may be able to use their visual minds to find alternative means to communicate, and this can be a highly creative medium to use to communicate.

Overreliance on Interpreters for the Spoken Word

When a teen or adult with Down syndrome has difficulties expressing himself through speech, parents or siblings often act as interpreters. In this role they help the person with Down syndrome get his message across to listeners who cannot easily interpret his communication. The interpreter enables the person with Down syndrome to identify and meet basic wants and needs and may also advocate for his rights and needs in the community, such as by helping to locate the most advantageous programs and services.

Often there may be a type of shared agreement between caregivers and the person with Down syndrome for the caregiver to take over much of the act of communicating. Parents or other caregivers appreciate how difficult it is for the person to speak, and it may also be easier for the person with Down syndrome to let someone else speak for him, even when, with time and effort, he could communicate for herself.

Despite the important role caregivers can play as interpreters, a number of problems may develop from this role. First, many people with Down syndrome may become too dependent on one person as an interpreter. If this person leaves or is no longer present, the person with Down syndrome may feel lost and disoriented. For

example, we have seen people who lose a special boss who had been taking the time to understand and communicate with them. In the absence of this person, they may flounder. This may also happen when siblings leave for college, or in residential settings when close staff or other residents leave. Of course, the death of a family member such as a parent may be extremely difficult.

Another drawback is that over time interpreters tend to develop a type of "shorthand" understanding of the individual's needs and wants. Having seen the person with Down syndrome respond to different situations over many years, they are often able to anticipate how he will respond and what he may want and need in a given situation. This is just part of being a good interpreter and advocate. Still, sometimes the interpreter may make assumptions for the person that are not correct. Additionally, as adults with Down syndrome age and mature, they often want to have more of a say in decisions affecting their lives. Even if the interpreter's assumptions are correct, the person with Down syndrome may still want to communicate for himself, just as most other adults do.

Promoting Strengths to Facilitate Communication

The good news is that people with Down syndrome can communicate far more effectively if they are allowed to use mediums of expression that are strengths for them. As we discussed in chapters 5 and 6, people with Down syndrome tend to think concretely and are also highly responsive to visual cues. Consequently, they often rely on words or images that are concrete and visual to express themselves, such as things they see and touch in their immediate world.

Someone may be very reluctant to express a thought with speech alone but may be more willing to express it if he can pair a picture with the spoken word. For example, a teen or adult might be unwilling or unable to state what happened on an evening out. What if instead of not talking, he takes out his cell phone?

- The person might display pictures of himself or others dancing (perhaps augmented with the word "dance").
- Or he may include pictures of friends who were with him (and may simply say the name of a friend, e.g., "Megan").
- Or he may show you a picture of the restaurant (accompanied by carefully chosen words such as "hamburger" or the name of the restaurant).

Through the visual medium of pictures, the person's world can open up to you. You may just have to encourage him to use easily accessible images. As the expression goes, "A picture is worth a thousand words," and not surprisingly, you may also be thrilled with the few words that accompany the picture.

Imagine that a family or group is sitting around a dinner table, and everyone except the person with Down syndrome is talking about their day. Again, pictures can open the person's experience up to others. Even if he does not have a phone or pictures, he may be able to visually share information about his activities, such as by drawing or acting out some aspect of his day. The fact that so many people with Down syndrome love to act and are often described as "hams" can, if others are receptive and patient, turn any gathering into a shared exchange of images, actions, and words that convey rich meaning and purpose.

Visual images used for communication need not be a picture, drawing, or a representation of an object but can in fact be anything that can be pointed to in the immediate environment. This can go a long way to solve problems that occur when someone cannot verbalize what has happened to him. However, parents or caregivers may need to be flexible and go to the environment in question if they suspect that there is something important to be communicated or that something is amiss that cannot be communicated through the spoken or even the written word. An example of this is discussed in the section on expressing more complex thoughts or feelings. All told, if you can encourage the person with Down syndrome to express himself using whatever medium he is comfortable with, you both may be greatly rewarded.

Using Written Words to Facilitate Communication

As mentioned previously in the discussion of receptive language, the written word is still visual and can be concrete. Therefore, it is *easily understood and accessible* to people with Down syndrome who can read. Written words can be used to help people communicate, as well as to adapt and solve problems in their worlds. Here is an example of using the written word to help an adult with Down syndrome communicate his thoughts or ideas to his mother:

> *The first author was asked to meet with Cody's family because of what his mother described as a "difficulty with communicating with her." When Cody came to the meeting with his mother, we were surprised to*

find what appeared to be a very friendly, engaging, and seemingly confident young man. He shook my hand firmly and talked about his impressive job in the mail department of a large office and about his successful social life. We then asked Cody's mother if she could explain the problem, and she stated that her son had become more and more reticent to talk to her. We asked Cody if he would talk to his mother so we could observe their interaction. Despite having spoken with us in a strong and engaging style, he turned and began to speak to his mother in a surprisingly timid and tiny voice. His mother was a very confident and forthright person, and it appeared that Cody was intimidated when talking to her. Neither of them could tell us when the problem had begun or what had transpired to create it, but it was not something the mother or son wished to continue.

It appeared that Cody had developed some aversion or fear when speaking to his mother even about trivial matters. This issue had grown worse over the past several years. We learned, however, that he enjoyed writing and was comfortable with writing his thoughts and ideas in a journal. Thus, we recommended that he begin to journal ideas that he would like to communicate to his mother. We suggested he bring this to his mother as a prop to facilitate communication when they had free time in the evening.

When we introduced this idea, we were intrigued by his mother's laughter. She stated that for many years her son had left Post-it Notes all over the house to communicate his thoughts and issues. There were not sure why or when this had stopped, but it clearly was a form of communication that was similar to our suggestion to journal. We suggested that Cody resume writing Post-it Notes to communicate basic wants and needs in the house but then use the journal to discuss more serious or thoughtful ideas that occurred to him in the course of his day. When we followed up with Cody's family, we heard that the suggestion had worked to allow them to communicate successfully despite Cody's difficulty speaking assertively with his mother.

Word Lists

In our experience, many teens and adults enjoy writing lists: lists of their favorite movies, actors, foods; lists of things they own or would like to own (e.g., birthday wish lists); lists of sports figures or teams and their standings, etc. Making lists helps people with Down syndrome organize their thoughts and their days and can be a productive way of spending time, just as it is for any of us. Lists can also facilitate the use of more appropriate or complex verbal language. Here is how one young man was able to use lists to learn to express his frustration more appropriately:

Alex was quite good at texting, but he had a limited repertoire of words to communicate some of his thoughts and feelings. He had established a texting relationship with his boss at his job in a bookstore to receive his work schedule and other important work-related information. Like many people with Down syndrome, he was very conscientious about getting to work on time, and he became increasingly annoyed when the cab drivers arrived late, making him late for work. On one occasion when Alex was especially late, his anger rose, and he texted his boss that he was going to "kill that SOB."

Alex's boss was very fond of him, but he was also concerned by the severity of this threat. He reached out to Alex's parents, who in turn contacted Alex's behaviorist. She worked closely with Alex to put together the following list, which was meant to allow him a more appropriate repertoire of words than "kill that SOB." His "I am mad strategies" included this list of more appropriate phrases he could use:

I am so mad
I am super mad
I am upset with the 242 cab (the cab company)
I am frustrated
I am angry with the driver
I am pissed OFF
I am unhappy
I am uncomfortable
I am nervous
I need a minute to calm down

Over time, Alex learned that when he used these phrases rather than "kill the SOB," other people didn't get mad at him but instead helped him solve his problems. Alex was also given additional help in this situation. For example, his boss reassured him that it wasn't his fault if the cab was late. He was also given help ordering the cab earlier to give him more time to be on time.

List writing also facilitated a transition to new staff for Alex. In this situation Alex was complaining to his mother about his new job coach. Following from what they learned from the behaviorist, his mother helped him to list what bothered him about what his new job coach was doing or not doing. Instead of carrying a grudge or harboring ill will that might have ended the relationship, Alex was then able to effectively ask for what he wanted and needed, and they have worked very collaboratively since that time.

We have found that Alex and other teens and adults with Down syndrome can use similar word lists to facilitate social communication. For example, many people write lists of favorite music or artists, favorite activities such as dancing or movies, or favorite sports teams or stars. If they keep their lists on their smartphones or in a small notebook, they can bring the lists with them wherever they go. In social situations where small talk is expected, they can pull up the list from their phone or notebook and then name or show others the list. This greatly opens up communication and bridges the great divide in spoken language that often keeps people with Down syndrome from socializing effectively.

Communicating More Complex or Intense Thoughts and Feelings

We have found that people with Down syndrome usually cannot help but express their feelings and emotions nonverbally through their facial expressions and gestures. Family and friends generally have no difficulty knowing when the person is happy, sad, or angry. Many cannot hide their feelings, and this can actually be very beneficial to them. For example, people with Down syndrome have a reputation for being refreshingly honest, direct, and unpretentious. See "If People with Down Syndrome Ruled the World" (McGuire, 2015). They are also less likely to be sneaky or devious because they express their emotions so honestly.

Unfortunately, they are also far less likely to be able to conceptualize and verbalize the cause or source of their feelings and emotions. This may make it more difficult for the person and others to identify and resolve problems. As a result, teens and adults with Down syndrome are often more susceptible to stress that occurs in their lives. This may lead to a maladaptive pattern of problem solving called "learned helplessness." This occurs when a person does not have enough experience identifying and resolving personal problems and may therefore tend to give up when faced with a major difficulty. In other words, he has literally "learned to be helpless" in the face of a major challenge or difficulty. (Seligman et al., 1968).

A host of problems can arise if someone cannot communicate the cause of a problem. However, it may help to remember that the world is concrete and visual for people with Down syndrome:

Milo was an engaging young man who had worked successfully in an upscale grocery store. He and his family were very proud of his success. Much to the horror of his family, he came home from work one day in a foul mood, stating, "I want to quit" and "I hate that place." No matter what they tried, he could not tell them why. They called us for assistance, and we explained that this was not uncommon. We suspected that he had been traumatized by some experience, and even if he could explain what happened, the act of retelling it could retrigger the trauma). We suggested that they go back to the store with him to see if he could show them what happened. It took a great deal of encouragement and explaining, but he did finally agree to go to the store with his mother and his trusted job coach, Jonathan. We needed to find a concrete and visual way to tell us what happened, and this was the best chance for that.

When his mother and job coach accompanied him around the store, Milo was able to point to the back room and show a slight but obvious bruise on his head. It became obvious to his mother and Jonathan that he had been hurt and traumatized when using a big trash compacter because it had bounced up and hit him in the head. With the worksite supervisor's blessing, Jonathan created a detailed schedule for Milo's workday that kept him out of the back room. Over time, we found a way to help him go into the back room (months later). Jonathan also set up a concrete plan

that included asking some other employee he could trust to help him use the trash compacter.

There may be situations when people with Down syndrome have trouble putting their feelings into words and it is far more difficult to uncover the reason. Sometimes this occurs if changes or problems occurring in another setting are affecting a person's feelings. For example, parents may not know why their adult child is upset if something at work is affecting him. The intensity of the problem may also determine how difficult it is to verbalize feelings. Here is an example of how both intensity of the trauma and location can complicate the situation:

Staff in a small group home noted that thirty-one-year-old Bruce would not get out of bed to go to work in the morning. This was very unusual for him because he rarely missed work, even when ill. Staff became increasingly alarmed when he began to isolate himself in his room and when they observed a marked increase in agitation and self-talk, including frequent bouts of angry speech. Bruce was brought for an evaluation on an emergency basis when he began to stay up all night and became so absorbed in his self-talk that he barely touched his meals. A thorough physical and psychosocial evaluation revealed that Bruce had hypothyroidism (see chapter 2) but no other clear reasons for his change in behavior. While hypothyroidism may create some significant depression-like symptoms, it did not seem to explain the severity of his symptoms.

Fortunately, after we contacted staff at his worksite, they helped to solve the mystery. Bruce had been victimized by an aggressive bully who had recently begun working with him. Staff in his residence had begun to note a change in his behavior just after the bully stared work. Shortly after this, the bully left for a more appropriate setting. At this point, we worked with Bruce and the staff from his residence and worksite to help him deal with his anxiety. Following our recommendations, Bruce was able to go back to work after much time and effort by all involved. For example, over a period of a week, residential staff members were able to get him up and out the door in the morning, but then he refused to go inside the workshop building once he arrived. Despite much reassurance, he seemed to be afraid that the bully was still at the workshop. Staff

worked patiently with Bruce, and he began to enter the building very cautiously. In time, he went back to a normal schedule with no further problems or symptoms.

The Nonverbal Expression of Feelings

In the preceding example, Bruce could not tell even close family and staff what was happening to him. More than likely, this was because he feared that he might relive the experience of being bullied if he talked about it. In our experience, Bruce's response is fairly common in people with Down syndrome who have experienced intense trauma or other emotional issues. That is, even if they are able to communicate with others about day-to-day occurrences, they may not be able to conceptualize and communicate more sensitive problems and issues. As a result, they may communicate these issues nonverbally through a change in behavior.

The problem with this type of nonverbal communication is that there must be a receptive "listener" or receiver of the message at the other end. Unfortunately, uninformed professionals or staff may too easily label nonverbal expressions of behavior and emotion as a "behavior problem" or a "mental health disorder." Such general labels tell us nothing about the possible causes or solutions to a problem. Moreover, these labels may actually point to solutions that maintain or worsen a problem. For example, what if we had not learned about the bully and had viewed Bruce's problem as a "behavior problem"? There is a good chance that we may have developed a "behavior plan" that forced him back to the worksite without eliminating the bully's threat. This would not have solved the problem and would most likely have intensified his fear and anxiety. Similarly, treating the problem as a "mental health disorder" with the use of medications may have temporarily reduced some of his anxiety, but it also would not have eliminated the bully's threat. This too would have most likely resulted in a continuation and worsening of the problem.

When people with Down syndrome have to communicate their stress behaviorally, the causes are all too often misinterpreted. If professionals do not take the time and effort to look for all possible causes, the problem may continue. For example, we find many people who are treated for symptoms of depression (lethargy, loss of energy, etc.) with an antidepressant medication, when in fact the cause or a major source of the problem is hypothyroidism or sleep apnea. While antidepressant medications may temporarily reduce the symptoms, failure to treat the underlying

medical condition may result in a continuation of the problem and the depressive symptoms. In short, it is important to not assume that the behavior that is being used to communicate the presence of stress is the problem.

Even when parents or professionals are receptive listeners, the message may not always be received. Nonverbal messages are rarely clear. They often show the presence but not the source of the problem. To complicate matters, the problem may have more than one source. Thus, we have learned that we need to be very thorough when trying to track down all possible causes. For example, Bruce had a medical condition (hypothyroidism) that may have aggravated his symptoms and behavior, even if the primary cause of the problem, the bully, had been dealt with effectively. Failing to treat this or other conditions or sources of stress may have delayed a resolution of the problem.

Numerous health conditions, sensory deficits, and environmental stresses may be associated with any given problem. Additionally, different people show stress in different ways, depending on their own characteristics and vulnerabilities. For example, many adolescents and adults with Down syndrome have compulsive tendencies or "grooves" (see chapter 10) that are beneficial to them. They are able to follow through with routines and schedules that allow them to complete daily living tasks and job tasks very reliably. Unfortunately, under stress, they may become too rigid in completing tasks, and this may begin to interfere in essential or beneficial activities.

Here are some guidelines to reduce the effects of an individual's difficulties in verbalizing his feelings:

- Consider teaching the person some skills to communicate with concrete and visual materials. This may include having him work with a counselor to identify and label feelings with pictures or teaching him to look at pictures of expressions and indicate which one represents how he is feeling.
- Encourage him to draw or show pictures from books, magazines, or the internet to communicate about issues and concerns when verbal communication is difficult or limited.
- Sometimes people may be willing to act out a scene that is of concern to them. For example, one man who had limited verbal skills was able to demonstrate how his roommate was punching him when staff were not present.
- Family members and other caregivers can help the person with Down syndrome identify his feelings during day-to-day activities. For example, if you witness a

situation that obviously upsets the adult with Down syndrome, help him label his feelings, such as by saying something like this: “I would feel really angry if that happened to me. Is that how you’re feeling?” Then, when possible, connect the word to a picture or icon representing that specific feeling or emotion to allow better understanding.

- Family members or longtime caregivers may need to teach less experienced staff (at school, work, or residential settings) to interpret the individual’s expressive face and body language and to use visual images to support understanding.
- Make sure that there is more than one person who is willing and able to interpret for the adult, as discussed above. The more people in more settings who are able to understand and respond to the adult’s expressed needs, the more competent he will feel. Also, the more responsive caregivers there are, the less effect the loss of one will have on the person with Down syndrome.
- When possible, family members should be active participants in meetings with work or residential staff in order to ensure the most advantageous or accurate interpretation of the adult’s expressed needs.

Communicating in Group Situations

If someone is reluctant to talk to a close family member one-on-one, imagine how difficult it would be to talk to a group. We have found that the act of communicating in group situations tests the endurance and mental energy of many people with Down syndrome, much as it stresses anyone who might try to speak a foreign language in a group. For example, the first author found that it was extremely difficult to maintain focus and respond to the conversation when in a group speaking Spanish to others. He found that it was easy to drift off in his own thoughts and had difficulty not appearing rude and uninterested in the conversation.

With this in mind, it is interesting to hear that a family member with Down syndrome will often “make an appearance” at family gatherings but then retreat to the relative safety and quiet of his own room. Families want to know whether this is an indication of a problem, such as a symptom of a depressive withdrawal or antisocial personality. We often find that this behavior is normal. Like nonnative language speakers, many people with Down syndrome simply cannot keep up with conversations because they move too fast for them to process and to respond.

Conversations may also revolve around topics they have little or no personal interest in, such as how the stock market is doing or traffic problems on the local highway. It is understandable that the person with Down syndrome would want to leave the room. How long a teen or adult with Down syndrome participates in group conversation may depend on several key issues. If there are topics of interest to the person with Down syndrome, he will stay longer. Additionally, if the other people in the group have a genuine interest in the individual, he will often sense this and tend to stay longer.

What Helps

Here are some dos and don'ts for family caregivers to help encourage participation by people with Down syndrome in social conversations:

- Whenever possible use concrete and visual terms to enhance understanding. It may be possible to show them pictures related to the topic discussion, such as photos of the band when discussing a concert attended or pictures from a recent vacation. If visuals are not available, use language that is concrete and down to earth.
- Whenever possible, ensure the person has access to visual aids to supplement speech. He might present a few photos, quickly draw an illustration, or briefly act out something. As mentioned above, he could also carry lists of things of interest such as favorite musicians, movies, or sports teams, and these can be easily read or shown to the others in a social gathering or group meeting.
- Help to keep the person with Down syndrome in the conversation by sitting near him and quietly paraphrasing the topics of discussion in understandable terms. This is not necessary with all topics of conversation but more so when the topics are abstract.
- Encourage other people in social gatherings to be patient with the person with Down syndrome to allow him enough time to think, talk, ask questions, and articulate answers to questions and if possible, to use visual supports to communicate.
- Make comments to help include the person with Down syndrome in the conversation. For example, say something like, "You know, Sharon had a similar experience once . . . Sharon, do you remember when you . . .?" Attempts to include the person with Down syndrome in talking about past events may also play to his memory strength.

- Encourage others to respond to the person with Down syndrome in a rewarding way and not just by saying "uh huh." The most effective way to do this is to simply repeat a part or all of what the person with Down syndrome has said, without making any interpretations or additional comments. This is often said with a slight inflection as if asking a question. "So, you went to the party?" This is called reflective listening and it is well known and widely practiced in the counseling profession (Rogers, 1951). It stimulates conversation by letting the person with Down syndrome know that you are listening and what he says is valued. This also allows you to facilitate and clarify the person's conversation without actually speaking for him or taking his voice.

If the adult's speech is difficult for others to understand, the above reflective listening technique may be very helpful. We also recommend these additional strategies:

- Help translate the person's remarks, but only when needed. It is best to translate the person's actual statements without adding comments or interpretations, which may distort or change what he is actually trying to say.
- Consider briefly stating or describing the context, topic, or background of what is said by the person with Down syndrome to improve others' understanding. We find this can help to orient the listener to the context of the conversation and make interpretation of the specifics much easier. For example, it may be helpful to say, "He is talking about this party . . . his job . . . this movie," or "He is talking about the time when . . ."
- Get the permission of the person with Down syndrome before either translating or giving the context or background of his comments. This helps to maintain respect for the independence and integrity of his thoughts and opinions. For the same reason, it is also important to ask the person with Down syndrome if your translation is correct, such as by saying, "What I heard you say is that you had a good time at the party. Is that correct?"

This brings up another important issue about the participation of adolescents and adults with Down syndrome in other types of group meetings. We have attended many meetings where the conversation moved too fast for the person with Down

syndrome. How can he understand and respond to the proceedings, which may involve key decisions about his life, if it all sounds like a foreign language?

In these situations, it is imperative to translate the information into concrete and visual form prior to the meeting and have the person who will be acting as interpreter go through the key points of the agenda in written form with the adult with Down syndrome. He may still require a translator/interpreter during the actual meeting to ensure that he understands the ongoing proceedings. The interpreter and the others attending the meeting must also allow the person to communicate verbally or nonverbally about the proceedings. This requires patience from others, but above all else, a belief that the person with Down syndrome has the ability to understand, to communicate for himself, and to contribute important thoughts.

In our experience, if someone with Down syndrome is subjected to a meeting without these supports, he may feel disenfranchised and devalued and thus subject to frustration, despair, and depression. His situation is similar to that of someone who is being tried in a foreign court, where the language is not his own and there is no attempt to inform him about what is going on or to solicit his opinion or feelings. Viewed from this perspective, it is easy to see how people with Down syndrome may begin to act "inappropriately" during such meetings. How can people help but drift off into their own thoughts and even to talk to themselves when they are so divorced from the process they are forced to attend?

Special Issues of Highly Verbal and Nonverbal People

As explained earlier, most people with Down syndrome are in the middle of the intelligibility spectrum and have moderate speech limitations. Although there are fewer people at either end of the spectrum (those who are highly verbal or nonverbal), these individuals have their own issues that merit special attention.

Nonverbal Adults with Down Syndrome

People who have significant speech limitations have to use nonverbal actions and behavior to communicate their thoughts, feelings, needs, and desires. We have found that most individuals with verbal limitations are able to find incredibly varied and creative means for communicating that go beyond facial expression, gestures, body language. This may include signing, showing pictures or visual images, or the simple but effective tactic of pointing to get their point across.

Many people who are nonverbal can also use a variety of alternative and augmentative devices such as speaking devices, visual communication books, iPads or other tablets, computers, or smartphones, to improve their abilities to communicate. This may greatly expand their communication potential—provided someone is available and willing to take the time and effort to set up and teach the person with Down syndrome to use the devices. Finding the correct fit between device and the individual is important. We recommend consulting with a speech-language pathologist, talking to other parents, and reviewing devices and apps on the internet or where apps are available on a tablet or smartphone. We will not make a specific recommendation here for devices or apps because this is a changing field with new devices and apps regularly being introduced. Entering "Augmentative and Alternative Communication Apps" into your computer search engine leads to a variety of choices.

For this type of nonverbal communication to be received, the listener-interpreter has to be attuned to all the subtle and idiosyncratic nuances of the behavior and actions of the adult with Down syndrome and must attend to any augmentative or alternative communication devices. Understanding nonverbal communication requires learning a unique language for each person. This obviously requires a listener who is a sensitive and patient observer. Not surprisingly, we have seen some of the most understanding and sensitive caregivers working with people with verbal limitations. These interpreters become very important to the adult with Down syndrome, so their loss is often more devastating than it is for people with better expressive language skills or who are able to more readily use visual supports such as checklists. If you know a nonverbal adult with Down syndrome who has had a behavior change, consider whether he has lost an important interpreter lately. Additionally, whenever possible, make sure there are always several people in his life who can interpret for him.

Caregiver-interpreters must be careful not to assume they already know what the person with Down syndrome is trying to communicate, especially when it comes to wants and needs. There may be a strong incentive to just decide or choose for the person with Down syndrome, especially if getting his opinion takes a great deal of work. Still, there is nothing more important to one's self-esteem than having one's opinion and choices heard and acted upon. This may be even more important to people who do not have a history of responsiveness from others.

Finally, most successful interpreters continue to look for ways to expand the person's communication abilities. Thus, it is important to ensure that the person learns to use some more standardized form of nonverbal communication, so he can

voice her wants, needs, and choices to those who are less familiar with her communication methods.

When Communication Problems Become Behavior Problems

For nonverbal adults with Down syndrome, it may be extremely difficult to communicate more serious problems and issues. This may be especially true if the problem is something new or has not previously been communicated to others. For example, one twenty-nine-year-old man with verbal language limitations was brought for evaluation by his family when he began hitting himself very hard in the head. It turned out that he had a painful sinus infection. He had been very healthy for most of his adult years and had had little previous need to communicate physical pain to his family.

Sometimes people in the person's environment may fail to "hear" his communication. This may happen because no one is taking the time and effort to understand what he is trying to communicate—for example, when a special caregiver-interpreter is absent or distracted by someone or something else. It may also occur if the person's skills and intelligence are underestimated, especially by inexperienced staff or professionals. These individuals may tend to discount or downplay the person's ability to understand and to communicate her thoughts, feelings, and needs to others.

Whatever the cause, we have found that when people are frustrated in trying to communicate a problem or need, they usually do one of two things:

1. They withdraw into depression and despair.
2. They communicate their frustration and need through anger or aggressive behavior (toward property, self, or others).

In our experience, withdrawal into depression may be potentially more dangerous. This is because it may go undetected for some time and because figuring out the cause may be more difficult. This is particularly the case if the person seems to have given up and makes no effort to try to communicate the source of the problem (see chapter 17).

The other way to communicate a problem, through anger and aggressive behavior, is potentially more constructive, because the behavior often gives stronger clues as to the cause of the problem. For example, when the man mentioned above hit himself in the head, he communicated the source of the pain as his head. The other benefit is

that it is often a more successful way to get help. In an insensitive environment, depression may be ignored, whereas aggression, particularly when directed at staff, often gets quick attention.

On the other hand, there is a danger that uninformed staff or professionals may misdiagnose aggressive behavior as a "behavior problem." This often means that there is a lack of understanding or interest in seeing the person's behavior as his primary means for communicating. Viewed from the "behavior problem" perspective, the treatment is often to chemically manage (sedate) the person rather than to try to uncover the source of the problem. Behavioral management techniques are also commonly used. These may be helpful but also may be too restricting, especially if there is no attempt to uncover the cause of the person's angry behavior. Unfortunately, these techniques may end up suppressing the person's means for communicating and often lead to more anger and despair. On the other hand, attempts to understand the person's behavior as communication can be very fruitful.

Highly Articulate Adults with Down Syndrome

On the other end of the intelligibility spectrum, people with Down syndrome who have excellent speech and language skills sometimes can have far more problems than would be expected. Unlike people who are nonverbal and whose skills are often underestimated, these individuals are often believed to be more capable than they are because of their spoken language skills. There are a number of reasons this may happen. First, many people with Down syndrome are excellent observers and have exceptional memories. As a result, they may memorize phrases that allow them to appear as if they understand more than they do. Second, many of these individuals want to participate in conversations and social situations like everyone else and therefore may use certain remembered phrases or comments that help them appear to fully understand what others are saying. Third, they may be able to converse very fluently and capably about concrete situations and concepts, leading others to assume that they understand abstractions equally well.

Sometimes parents and other caregivers may also put intense pressure on the person with Down syndrome to be more capable. Caregivers may accentuate the person's expressive skills as evidence of her superior abilities or even of her "normalcy" compared to others with Down syndrome. But it may also occur because teachers and other professionals have told the family that the person with Down syndrome just needs to try harder or be more motivated to achieve more.

In fact, for many highly articulate people with Down syndrome, the real problem is that they are assumed to be more competent than they are because of their verbal skills. Some of their other skills may not be at the same level as their verbal skills. In contrast to many of the individuals described throughout this chapter, these people often have verbal skills (receptive and expressive) that exceed some of their skills in other areas. If others assume that their skills in other areas are high, too, they may be allowed to manage aspects of their lives that are beyond their capability. This fits in with the fact that many people with Down syndrome have uneven skills. They may have excellent speech and often can do many of their own self-care tasks reliably. This does not necessarily mean, however, that they are as capable in other important areas of their lives—for example, in knowing when to go to bed, what food to eat, or how to plan beneficial free-time activities (see "The Dennis Principle," chapter 3).

Too often, we have seen highly articulate people with Down syndrome blamed for their "failures" in jobs or living situations when in fact the failure is due to a misreading or misunderstanding of their skills by caregivers or staff.

Preventing Problems Related to Misinterpretation of Skills

If you know someone with Down syndrome who fits this description, there are a number of things you can do to prevent or at least moderate these problems. First, it may be helpful to get a more complete picture of the individual's strengths and weaknesses. There are many excellent assessment tools that look at a wide range of adaptive skills and not just verbal language. In fact, some of these tools were designed to measure the functional skills of people who may have limited verbal language. They emphasize the person's behavior as observed and reported by caregivers rather than the person himself. These measures include the *Vineland Adaptive Behavior Scales*, *The Scales of Independent Behavior-Revised* (SIB-R), the *AAMR Adaptive Behavior Scales* (ABS), and, to a lesser degree, the *Inventory for Client and Agency Planning* (ICAP). These assessments can help you understand the person's strengths and weaknesses and help you advocate effectively for his real needs in work, residential, and other community settings. A standardized test administered by a professional can give your family an effective bargaining tool to help counteract inappropriate placements by well-meaning but misinformed staff in agencies and community programs.

Families can also ask professionals who have experience working with people with Down syndrome to consult with the agencies serving the needs of their sons and daughters. The purpose of this is to educate staff about the real strengths and weaknesses of the person with Down syndrome in order to avoid expectations that are either too high or too low. Interestingly, we often say the same things the family has been saying to these agencies, but as professionals, our opinion may carry more weight.

It may help to discuss examples of situations when expectations were too high for the person's skills and suggest some strategies to avoid a recurrence. For example, we have seen many people fail at jobs as cashiers in grocery stores. In these situations, the job placement agency failed to note that that just because the adult with Down syndrome had good verbal skills did not mean he could manage money. This misplacement could have been avoided if the job placement agency had tested the person's ability to manage money before finding him a cashier's job. Had we been consulted, we would have also recommended a test of money management skills. Use of one of the adaptive skills scales (discussed above) would have also identified this limitation.

Some people have also been given jobs as office receptionists because of their verbal skills. In smaller offices, some adults with Down syndrome are able to manage the job. For many others, however, handling a high volume of calls, writing phone messages, and doing other complex office tasks are well beyond their abilities. Again, job failure could be avoided if the placement agency consulted with professionals or family caregivers who were aware of the person's limitations.

Having a good understanding of a person's skills will allow a match of the right job with the right person. In these situations, everyone benefits: the person with Down syndrome, the employer, and the community. For example, we have found that many people with Down syndrome can succeed as stock clerks, even at grocery stores with many items, because they have exceptional memories and good organizational skills. Similarly, we know many adults with Down syndrome who do exceptionally well in mail rooms or on loading docks because of these skills. Additionally, if not placed in jobs with time pressures, many people bring a degree of precision and reliability that is valued by employers.

Residential settings can also be scenes of great success or great failure if expectations are too high, as described in some detail in chapter 3. In these situations, people often fail when they are assumed to have more mature decision-making capabilities regarding sleeping and diet than they actually have.

One important way to keep overly high expectations from becoming a problem is for the person with Down syndrome to learn to advocate for his own needs. For example, the individual can learn to tell someone in authority when a job or job task is too difficult. We have actually found that most people with Down syndrome have a much better understanding and acceptance of their own limitations than they are often given credit for. For example, when we ask people if they are able to do certain tasks such as using the stove, most are quite honest and realistic in their answers. As discussed in chapter 8, we have found that families who have a more realistic view of their child's strengths and weaknesses often have children who are also more accepting of their own abilities. This, of course, may be challenged somewhat during the adolescent drive for independence, but in general this holds true.

Finally, how can you get the right balance between tasks that are challenging but doable and tasks that are either too difficult or too easy, and thus demoralizing? How do we avoid either extreme? In short, people need to experience both failure and success. Tasks should be neither so easy that the person feels he is being treated like a baby nor so hard that the person cannot learn to succeed, given some time, effort, and encouragement from others. Each task at each stage of development should be measured to meet and appropriately challenge the person's skills and abilities. This can usually be gauged from the tasks that he has already mastered.

Return to Table of Contents

Chapter 8

Self-Esteem and Self-Image

On a visit to the park when Chris was nine, two young girls curiously asked him whether he was "retarded." He quickly answered no. When they persisted, he was irritated but he continued to play. Later that day at home, Chris asked his mother if he was "retarded." His mother had already explained that he had Down syndrome and that he might be slower than others in doing some things. Now she told Chris that she and Chris's father did not use the word "retarded" to describe someone who learns slowly, but that some people did. She then sat down with Chris and together they made a list of what Chris could do and things that were hard for him to do. After making the list, Chris concluded that he could do far more than he couldn't do and that "having Down syndrome wasn't bad at all."

The *American Heritage Dictionary* definition of esteem is "to regard with respect." Following from this, self-esteem is to regard oneself with pride and self-respect. The importance of self-esteem to one's health and well-being is well established: People with self-esteem are happier and healthier, live longer, and have fewer mental health problems, to name some of the many benefits (Seligman, 1998).

The promotion of self-esteem is any action that helps people regard themselves with pride and self-respect. This sounds simple enough, and yet, how do you promote self-esteem in people with Down syndrome if others stare at them because they think they look different? How do you encourage respect and pride when the world values speed, self-sufficiency, communication skills, and productivity? It will come as no surprise to anyone that people with Down syndrome move at their own pace, have far less independence and control in their lives, often have difficulties communicating, and have far fewer job or career opportunities. Similar to other minorities, people with Down syndrome are often viewed and treated differently in our society.

Despite all this, the majority of people we have evaluated do have strong self-esteem and self-respect. How is this possible? Many seem to have an innate sense of self-respect, but much can also be attributed to families and other care providers who have found ways to encourage and promote self-esteem. This chapter will describe

some key issues related to promoting self-esteem, including the importance of accepting one's disability.

People with Down Syndrome as a Minority

For people with Down syndrome, being part of a minority has advantages and disadvantages. One of the most difficult challenges for people with Down syndrome and their families is the lack of understanding and acceptance by some others in the community. Despite efforts by parent groups and advocates to change stereotypes, people with Down syndrome are still viewed and treated differently in society—they continue to be stared at, teased, and at times taken advantage of. Also, people with Down syndrome may lack positive role models from within their group or may have limited contact with others who have Down syndrome.

On the other hand, people with Down syndrome are recognizable as belonging to a group. The same physical features that invite teasing from some people signal more empathetic people of the possible need to take the disability into consideration during interactions (such as by being extra patient). This applies to adults as well as children. For example, one study found that school-age children were more accepting of peers with Down syndrome than of children with "invisible disabilities" such as learning disabilities (Siperstein & Bak, 1985). The authors of the study speculated that children with invisible disabilities may have looked like other children but had something different about them that may have confused or put the other children off. On the other hand, the distinctive physical features of the children with Down syndrome clearly identified them as having a disability. Similarly, a study (Healey, 2017) found that in soldiers returning from war, those with blindness, which was more visible to others, received more sympathetic responses than those with hearing loss, which tended to be less visible.

In our own experience, we have found that people with Down syndrome are generally accepted in community settings. When staring, teasing, or discrimination occurs, it often involves a small number of people who are uninformed or bullies.

Belonging to a group can also be an advantage by enabling people with Down syndrome and their families to connect with other people in the same situation. Some families of people with intellectual disabilities that are not due to a recognizable syndrome have shared with us that they do not have the same connection that families of people with Down syndrome have.

Family Acceptance and Self-Esteem

Self-esteem starts with acceptance of who we are. For people with Down syndrome, that includes accepting that they have Down syndrome. They cannot be proud of themselves if they cannot accept this fact. Acceptance increases the development of their own skills and abilities and encourages advocacy for their own rights and needs.

The development of pride and acceptance in the individual with Down syndrome often begins with the family's acceptance and willingness to discuss Down syndrome. This is not necessarily an easy or simple process. Anything that marks family members as different (race, creed, disability, etc.) may make a family susceptible to isolation due to real or perceived discrimination from others. For example, parents of children with Down syndrome may find some relatives distant or unresponsive, which may make it difficult or uncomfortable to attend family gatherings. Families often report that these types of issues extend well into the adult years.

Many families gain support and acceptance by joining organizations where they meet others who have children or adults with Down syndrome. Finding families with similar experiences can be a powerful form of support and promote positive attitude and self-esteem in the family and the person with Down syndrome. Still, some parents continue to struggle with acceptance of their child's Down syndrome. There are also families who have few, if any, problems accepting Down syndrome while their child is young but question their acceptance when their child enters adolescence or adulthood and fails to achieve some of the typical milestones associated with increasing independence.

It is beyond the scope of this book to explain in detail what family members can do to accept Down syndrome. If your adult child with Down syndrome is struggling with self-esteem, however, it may be worthwhile to ask yourself whether you are having trouble with acceptance. Also be sure to read chapter 3 on the role that families play in supporting adults with Down syndrome.

Self-Esteem in Individuals with Down Syndrome

The development of pride and acceptance in individuals with Down syndrome is a complex and often creative process, involving the person's own attitude and

abilities and the environment she lives in. In our experience, acceptance appears to be a five-step process that involves the following:

1. awareness,
2. developing a sense of competence,
3. developing realistic dreams,
4. developing one's own unique talents or gifts, and
5. feeling loved and accepted by family and friends.

Step 1: Developing Awareness

Awareness of any type of disability or difference in oneself may result in feelings of anger, loss, and sadness. For children with Down syndrome, awareness of differences in skills and opportunities compared to their peers' and siblings' invariably increases when they are included in regular classrooms and community settings. In adolescence and adulthood, many become even more aware of discrepancies. For example, a young child with Down syndrome may play on the same block and go to the same school as all the other children from the neighborhood. Later, the type and frequency of activities shared with peers without disabilities often decreases. This may occur as early as grade school, but differences are particularly noticeable when others learn to drive, date, go to college, get married, and have careers. Some people with Down syndrome have some or all of these opportunities, but we have seen a number who feel left behind.

Even people who attend self-contained programs throughout their school years are still in the community enough to experience the stares and the different treatment given them by others. Remember, people with Down syndrome are often very sensitive to the reactions of others.

When they compare themselves to others, people with Down syndrome are often forced to consider their own identities (of who and what they are). This is not unique to people with Down syndrome. We all have to figure out who we are. Furthermore, comparisons between people with Down syndrome and the general population are probably no less disappointing or humbling than the comparisons the rest of us make. After all, we may all dream of being what we are not, such as larger or smaller, more attractive, or more successful. Many of us even dream of being rock stars, sports legends, etc.

The process of coming to terms with one's identity often starts early in childhood and continues into adulthood. We have heard of many creative ways that families of children and adults with Down syndrome have promoted a positive view of Down syndrome. This may include dealing with the inappropriate or unkind actions of others and the self-blame that sometimes occurs in response to these actions (a common experience of minority members). Over time, the result of this process of awareness and acceptance leads to the development of a more honest, realistic, and positive view of self.

The story at the beginning of the chapter was shared by Sheila Hebein, the former executive director of the National Association for Down Syndrome (NADS). In it, she described an important event in her son Chris's development of acceptance and self-esteem.

It may be instructive to look at some of the steps Sheila followed in reacting to the incident in the park. First, Sheila did not intervene in the incident. After careful observation, she determined that the girls' questions were based on normal curiosity and offered a wonderful learning opportunity for Chris. Second, Sheila respectfully waited for Chris to initiate a discussion of the incident. She knew that he needed time to think through the issues, and she trusted that Chris would come to discuss the issue when ready. Third, her explanation of "Down syndrome" was direct and honest and yet also extremely encouraging and respectful of his skills and abilities. On the one hand, she did not gloss over the fact that Down syndrome was a disability that might make him "slower than others." On the other hand, she conveyed great confidence in his ability to deal with this issue on his own terms and at his own speed by waiting for him to initiate the discussion and by asking him to talk about things he could do. Finally, the sense of pride and acceptance Chris experienced came from a natural event in the community. This incident, in turn, was the result of a family decision to let Chris explore and experience the world.

Clearly, for Chris, the message from this experience was that he had the right and capability to be in the community regardless of his limitations. No doubt, the confidence and pride he experienced from this incident has carried over to subsequent incidents in his life. Chris is a now an adult working successfully in a community setting, and despite the challenges he has faced, he has maintained a strong positive view of himself and of Down syndrome.

Breaking the News

At this point it may be helpful to discuss what to do if you have an adolescent or an adult with Down syndrome in the family who is not aware of her diagnosis or has not recently talked with you about what it means for her to have Down syndrome. The following are frequently asked questions regarding educating teens and adults with Down syndrome about their condition:

Why do adolescents and adults with Down syndrome need to know they have Down syndrome?

As discussed earlier, it is difficult to develop your own talents and skills and to advocate for yourself if you cannot accept Down syndrome. This is true whether the person is a younger child or an adult.

Is the person with Down syndrome ever too old to learn about Down syndrome?

We do not think so. If anything, there is a greater need with increasing age, because most people with Down syndrome already know they are different than others. By adolescence, and certainly by early adulthood, most people have been stared at or treated differently literally hundreds of times. Regardless of how sensitive or insensitive you think the person is to these issues, it would be very difficult for her to miss the fact that she is different unless she lives in a vacuum. At this later stage of life, the question then is not whether they are different but how and why?

If we discussed these issues when she was younger, is it important to discuss them again when she is older?

Yes, because the discrepancy in skills between people with Down syndrome and peers in the general population is more apparent in adolescence and adulthood, so people's awareness of this difference is also greater at this time. Your son or daughter needs to discuss his or her Down syndrome in order to develop a positive and realistic self-image as a teen or adult.

What response should we expect from our son or daughter to this discussion?

Some people have difficulty accepting the message, but most experience some sense of relief after learning that they have Down syndrome. The discussion will often confirm and validate feelings and observations they have about being different. There

is a name for what they have been experiencing (Down syndrome). They have something that helps to explain what has been happening to them all these years.

Should we wait for the person to initiate a conversation about Down syndrome?

Certainly, if she brings up the subject, it is very important to respond. On the other hand, if she does not bring it up, it is not always advantageous to wait. Many people are very aware and sensitive to the world around them, but they may have difficulty articulating their feelings and thoughts to others. If you raise these issues, it may allow them to voice questions or concerns they have and have been unable to express. Additionally, people with Down syndrome are often sensitive to cues from others. If this topic is broached, they may feel free to discuss it; otherwise, they may not be sure that this is what the parent wants.

How do you talk about Down syndrome?

We recommend simple and honest statements similar to what Sheila told Chris in the above example. People need to know the name (Down syndrome) of what they have. This makes it a concrete reality. They need to know they have some significant differences, and what they are. For example, they may need more time to do certain tasks and need more help from others with finances, travel, or whatever the case may be.

One way to discuss Down syndrome is to discuss the person's strengths and weaknesses. It is important to praise and encourage the person for what she can do. It is also important to mention her special talents and gifts. On the other hand, being forthright about the person's limitations is also essential. If you are not honest, you run the risk of undermining your credibility, and, more importantly, of denigrating and patronizing the person with Down syndrome. Most people with Down syndrome are very perceptive about the veracity and authenticity of another's comments. If the person with Down syndrome thinks you are less than honest about her weaknesses, she will probably not trust your comments about her strengths.

Are there ways of conveying a more positive image of Down syndrome?

It may be helpful to point to other people with Down syndrome of different ages and levels of skill in the community who are good role models. Additionally, celebrities like actress Lauren Potter and the actors on Born This Way offer excellent role models of people who are successful and have good self-esteem (see peers discussion below).

Are there ways of normalizing Down syndrome?

There are ways to help someone with Down syndrome understand that while her Down syndrome may be unique, many of her issues and concerns are not. For example, it may be helpful to discuss the fact that we all have strengths and weaknesses. Many people dream of being rock stars, sports figures, or more successful in work or in love, but everyone has to live with what they were born with.

Additionally, it may help to point out that not all the individual's differences are related to Down syndrome. For example, an older sister may be able to have a cell phone because she has reached a certain age and not because she does not have Down syndrome.

When do you broach the subject?

There are many different types of situations where the subject may be broached:

- You might bring up the subject when your son or daughter is stared at, teased, or the victim of an unintentionally hurtful comment. These situations often demand some type of response and discussion by family members to help the person with Down syndrome learn from and deal effectively with these issues.
- An opportunity might also arise when the person with Down syndrome compares herself unfavorably with someone without a disability. This may happen when others have major life experiences that the person with Down syndrome is less likely to experience, such as getting married or going to college. It may also include more day-to-day issues, such as if the person with Down syndrome needs an escort to go into the community when her younger sibling is free to go alone.
- Parents may also broach the subject at a time when there are no negative issues or concerns related to the person's Down syndrome. This may allow you to discuss the issue under less stressful circumstances. This has some advantages and disadvantages. The individual may be more intensely interested in the subject when she is in the midst of a problem situation. On the other hand, when there are no problems, you can gradually introduce the issues. You can then discuss them over an extended period of time, allowing the person to more easily process the information. There may be many ways to bring up the

subject of Down syndrome—for example, if you see someone in the community or on television who has Down syndrome.

Step 2: Developing a Sense of Competence

Following awareness, the essential next step for positive acceptance is to emphasize the person's own strengths and skills. Over time, and with encouragement from family and friends, the person needs to change her perspective from "what cannot be done" because of his disability to "what can be done" because of her own skills and resources. Psychologists call this process the development of competence. Competence is the term used to describe every human's need "to do for self" as a means to gain some sense of control and mastery over the world. In childhood the promotion of competence begins at home, and over time, competence with daily living tasks leads to a greater sense of independence and to enhanced pride and self-esteem.

During our evaluations of adults with Down syndrome, we have found that the families who are most successful at promoting competence follow a "good enough" formula for each task at all levels of development. That is, parents are aware of their adult child's limitations but are also aware of her potential for independence and give her the message that she is good enough to do the task. Parents are there to guide and aid her as needed while also encouraging her to do the nitty-gritty concrete elements of the task herself, whenever possible. This includes doing daily living tasks at home at each stage of life, as well as tasks at school, work, and other community settings. The process of trial and error and of learning from one's mistakes is the same for people with Down syndrome as for other children; just the starting point and the level of skill attained are different.

Families who have more difficulty promoting competence may expect either too much or too little from the person with Down syndrome. When parental expectations are too high, the person may give up in frustration and failure. We have seen this occur in adults who become depressed and despondent when family expectations were too high and they could not achieve the same goals in sports, school, job, career, marriage, etc. as "normal" brothers, sisters, or same-age peers.

On the other hand, frustration and underachievement may result if families expect too little from the person with Down syndrome and do not allow her to do self-care tasks that would increase her independence. As a result, when opportunities for

independence occur, she may simply not have the experience or confidence to know how to respond effectively. Equally important, when people do not get experience in dealing with day-to-day challenges, they may develop a sense of helplessness and despair in the face of more serious problems and issues. This has been aptly described in the literature as "learned helplessness" and puts people at far greater risk for depression and a host of other health and mental health problems (Seligman, 1968). This is because if they do not have experience with meeting daily challenges, they will easily give up too easily when faced with a major problem because they have literally learned to be helpless.

Whether people expect too much or too little of the person with Down syndrome, the pride that comes from the development of independence skills goes unrealized. This occurs if the person is pushed to be something she is not or cannot be or if her talents and skills are grossly underestimated. These topics are further addressed in chapter 3.

Dos and Don'ts in Promoting Competence at Home

- Encourage the person with Down syndrome to do new tasks that she is physically and developmentally capable of doing. If the task is too far beyond her, this may demoralize her rather than create competence.
- If the task is too difficult, it may be possible to break it into manageable and doable steps.
- Tasks that are most important to the person with Down syndrome are an added incentive.
- Don't be too quick to "take over" when the person is trying a new task.
- Above all else, encourage the person to accept mistakes and a lack of success as a necessary part of the learning process. How else can people learn if they cannot learn from their mistakes and failures?

Competence in School

Families who support competence and self-esteem at school are careful not to confuse their own wishes and ambitions with those of the person with Down syndrome. These families are most likely to honestly and realistically appraise the person's skills and interests in order to find the right environment for her. They also

encourage school staff to build on existing strengths and positive experiences to increase confidence and motivation.

Again, families who overestimate or underestimate their child's skills and abilities may fail to find school settings that promote pride and self-esteem. True, in the United States, the most inclusive school setting possible is supposed to be the first one considered. However, the student's needs and abilities should determine the curriculum. Academic programs that are over the student's head or that simply serve as a babysitting service do nothing to promote independence, pride, or competence in dealing with the world. Also, when students with Down syndrome are included in classes geared to college-bound students, they often are not taught the practical job skills, money handling, reading, food preparation, self-travel, and other tasks needed to live as independently as possible. Additionally, including a student in a setting where there are few if any other students with Down syndrome or other disabilities may have a negative effect on the student's social and emotional development.

In the upper grades, education should focus on the realities of life after school. It is fine to continue to take academic classes, especially in subjects such as reading, writing, and math that adults need for independent living. However, most older students with Down syndrome also need to learn social and work skills so they can succeed in jobs and live independently in the community. Moreover, job experience is essential to any successful school program. Research has clearly shown that the greater one's job experience while still in school, the better the chances of being successful at finding and keeping a job after leaving school (Wehman, 1981). In our experience, exposure to different types of jobs is also valuable so people can choose an area of interest to them. Parents who do not understand the importance of job experience in school often find out too late that these experiences are critical to success in the job sphere.

Equally important, success in job experiences often requires a competent job coordinator and job coach. Successful programs will emphasize the skills and talents needed to succeed on the job, such as patience and persistence on job tasks, a strong work ethic, and taking care of one's appearance. In addition, the job coach will help teach appropriate social skills at the worksite (see more on social skills in chapter 13.

A good outcome is usually the result of a program that is both appropriate for the person's skills and addresses the life skills necessary after graduation. A nationally recognized school district in the Chicago area discovered these challenges. The focus of this district is on college preparation, since a very high percentage of the graduates go on to college. The approach was not altered for students with

intellectual disabilities, many of whom became frustrated and graduated without appropriate job skills. The school district then modified its approach and developed a transition program. This program emphasizes skills for living and working in the community and provides ample opportunity for supervised job experiences in a host of settings. The students are now having a much more positive experience and are better prepared for occupations after graduation.

The Importance of Having Friends with Disabilities

Why is it important for people with Down syndrome to have friends who have Down syndrome and other intellectual disabilities? If you think about your friends, they probably tend to be people who are on about the same intellectual level as you. They are the ones who are most likely to understand what you are trying to say, to share your interests, and to experience the world the same as you do. They serve as a type of mirror and validation for who and what you are.

The same applies for people with Down syndrome. They also need at least one or two friends who are likely to experience the world as they do. They may not always be able to talk to these others because of expressive language limitations, but this may not be necessary. As mentioned in chapter 4, people with Down syndrome are very aware of and sensitive to others in their environment. In observing how others experience and deal with the world, they come to feel that they are not alone in their perceptions and experiences. They validate who and what they are, which offers a strong sense of support and identity.

Postsecondary Programs

We have heard from a number of young adults with Down syndrome who attend postsecondary programs at colleges. Most of these individuals commute to programs at local community colleges and take one or two courses outside of their normal work and social schedule. Typically, these local programs make courses available to people with intellectual disabilities but offer little else in the way of support or organized social activities. Still, people with Down syndrome attending these programs enjoy their courses and often describe their participation in them with pride. Many seem to feel they are experiencing something that is fairly rare for individuals with disabilities. As one student described his courses, “I can go to college just like my

brothers and sisters." It is not clear how much substantive learning occurs in these courses, but this may be beside the point. For participants, the experience seems to be invaluable in and of itself.

There are also specialized community college programs that do have supports and counselors available for students with developmental disabilities. Some even have social activities available, ranging from drop-in centers on campus to dances or other social outings and events. The most successful and popular of these programs have courses that emphasize independent functioning in the community, as well as job training and adaptive skills in the workplace. The network of support with other students in these programs is also of great importance. These types of programs take up where secondary schools leave off and may be very beneficial in offering support and training for people in the community. Unfortunately, there are not many of these programs because they do not appear to be a high priority for state educators.

There are a number of more intensive programs, usually located on college campuses, in which participating students often live in dorms. There are two different types of campus programs: (1) those that are primarily for students with learning disabilities but sometimes also admit students with Down syndrome, and (2) those that are specifically designed to serve the needs of people with Down syndrome and other disabilities. We have found that the former are not always optimal environments for people with Down syndrome. Many of the students who do not have Down syndrome have learning problems, but most have average or low average intelligence. Because of the emphasis on these students, the courses are often too difficult for most students with Down syndrome. Perhaps more importantly, there is also more of an emphasis on academic rather than on work and community skills, so the programs may not suit the needs of adults with Down syndrome. Additionally, supervision in the dorms may be inadequate, since most of the students have the cognitive ability to manage their own lives and schedules. As discussed in chapter 3, adults with Down syndrome may be technically proficient with their own self-care, grooming, etc., but still be immature with decisions such as when to go to bed, what to eat, when to attend recreation activities, etc. We have heard of a few students who have done well in this type of environment, but many have not.

Given the problems associated with these programs, why would people with Down syndrome attend them? Many parents may hope that their son or daughter with Down syndrome may learn from and even rise to the level of the others in the program. While this may be possible to a limited degree, the demands on the student

and the lack of adequate support and supervision make this a bad fit for many people with Down syndrome. Many students with Down syndrome are simply overwhelmed.

The second type of campus program is more specifically designed for people with Down syndrome and other intellectual disabilities. The stated goals of most of these programs are to help students develop self-esteem and the skills for living independently in the community. Most of these programs are relatively new and have had to learn through trial and error how to best meet these goals. The most successful programs focus on adaptive living skills in the dorm, worksite, and community more than they emphasize traditional lecture-style coursework. Additionally, more successful programs have an understanding of the needs and limits of people with Down syndrome. For example, they are more active in helping students to structure schedules and routines for them to follow. The most successful schools also continue providing supports to students who have graduated and are living in the community.

Left to their own, young adults with Down syndrome may flounder, but with the right amount of help and guidance, many have matured and prospered in these environments.

Competence at Work

There is perhaps nothing as important to an adult's sense of competence and self-esteem as her work. We have found that most people with Down syndrome are very motivated and conscientious about their jobs. We have heard high praise from employers regarding the work ethic and performance of people with Down syndrome. Employers report that people with Down syndrome are not necessarily fast but are quite often thorough, persistent, and reliable (see chapter 10). Many are reluctant to take time off and are rarely tardy. Some employers actively recruit people with Down syndrome because of the positive experience they have had with this group. For example, one camera company hired a large number of people with Down syndrome for assembly of cameras and related items. The precision and care people bring to the job is valued by this and other employers.

Success in community jobs depends on a number of factors, including

1. exposure to different worksites;
2. adequate training in work tasks, ability to communicate with supervisors and others, social skills; and

3. ongoing support.

There has been much progress in the area of community jobs, particularly in the past ten years. Some of the most common jobs held by adults with Down syndrome are in grocery stores, janitorial and cleaning positions in offices, or fast food restaurants. We also frequently hear about jobs in offices, mailrooms, nursing homes, day care centers, and factories.

Exposure to Different Jobs. Exposure to a wide variety of different types of work is a key component of any successful school or postschool job training or placement program. This allows people with Down syndrome to try out different types of work and find out what fits their wants, needs, skills, and resources. Job trainers are able to see them perform at different worksites to assess strengths and areas that need additional training. When people are not given this type of exposure, they may be placed in jobs that are not appropriate to their skills or interests, as illustrated by these examples:

One man's family found the "perfect job" for him as a greeter in a large discount department store. Unfortunately, no one checked to see if he thought the job was perfect. His family found out fairly quickly that this was not what he wanted when he refused to get up and greet people. Fortunately, his family worked together with the manager so that he was able to try different work tasks in the store. After several different job tasks did not work out, he found the right job in the stock room doing merchandise preparation.

Dominic, another adult with Down syndrome, had been assigned a "prized job" in a grocery store by a job placement agency. After approximately six months in the position, he began to send carts out into the middle of a busy street that ran along the side of the store. He told his family that he had tried to do the job because he knew he was lucky to have gotten it but that he just could not take it anymore. Dominic now works in a greenhouse, which seems to be much better suited to his needs and interests.

There are additional examples of adults with Down syndrome failing at poorly suited jobs described in other parts of the book. All these failures might have been

prevented if the individuals had been exposed to different job environments in order to determine what they were capable of and interested in doing.

Adequate Job Training. Training to do the tasks required in the job is also essential. Job training works best if it is done at the worksite and if tasks are broken down and taught in manageable units that emphasize visual and concrete learning—strengths for many people with Down syndrome—by a patient job coach or trainer. We have found that it is also helpful if people with Down syndrome are encouraged to capitalize on their excellent visual memories, especially through role play, to memorize and repeat the steps required to do the task.

Unfortunately, we have seen people have problems and failures in jobs because they did not have adequate job task training and supervision. Here are several examples:

When Marion, age twenty-four, started working at a large discount department store, the only preparation her supervisor gave her was to hand her a rag and tell her, "Dust the store." The assignment was overwhelming, and needless to say, did not work out. Marion seemed to be paralyzed in one spot.

A similar problem occurred with Meg, a thirty-two-year-old woman working on a janitorial crew. She was given the job of vacuuming a large ballroom. Although she knew how to vacuum and enjoyed her job, she was also felt paralyzed due to the enormity of the task. When the supervisor came back three hours later, Meg had managed to vacuum only a twenty-square-foot section. Fortunately, the solution to Meg's problem was rather simple. Her supervisor and job coach partitioned the ballroom into more manageable sections, which she could see in a picture of the room, and Meg was able to continue doing her job as effectively as ever

For Marion, whose job was to dust the entire department store, the outcome was not positive; she lost her job. This should not have happened. A job coach should have been available to break the job down into more manageable tasks that were visual and concrete. The store supervisor also showed a lack of tolerance or patience for adapting the job to Marion's needs. Despite this, Marion actually had the last

laugh in this situation. She was able to find a job in a nearby competitor's store, and, with a little bit of direction from a good job coach, she became a model employee who is highly valued by her employer.

Social Skills Training. Besides training people in job tasks, perhaps the most important role of job trainers is teaching appropriate social skills and "job etiquette" at the worksite. Due to the importance of social skills, we devote an entire chapter to this topic (chapter 13). Suffice it to say for now that researchers have consistently found that deficits in social skills, not job skills, create most problems on the job for adults with developmental disabilities (Greenspan & Shoultz, 1981; Hill & Wehman, 1978).

Ongoing Support. A different type of social issue we have found is that some people with Down syndrome who have jobs in the community appear to be alone and isolated:

> *Ellen, age thirty, was diagnosed with depression. She had had recent losses in her life, but we also found she had no friends or confidants at work. She did cleanup in a fast food restaurant where there were few repeat customers and there was much turnover in the employees and management. Many of the other employees spoke primarily Spanish, and they had difficulty understanding or talking to Ellen. We started Ellen on an antidepressant medication, which reduced her symptoms, but we also strongly recommended that she work in a more supportive work environment. (Gains made from medication tend to be limited or not sustained if the person's environment does not change.) Soon afterward, Ellen moved to a different restaurant employing a number of people with disabilities, including a close friend of hers. She continued to make steady progress after this job change, and now, three years later, she has had no new bouts of depression.*

We have seen many people with Down syndrome do especially well in what are called "enclaves," where they work at a jobsite with several other employees with disabilities as well as employees without disabilities. Enclaves offer an excellent means of reducing the problem of isolation and make the most productive use of a job coach's time.

Some people with Down syndrome may find the most supportive work environment to be a sheltered workshop. These workshops were first established in the 1950s and ’60s to give people with disabilities access to training and work in a segregated (sheltered) setting. These centers usually have different levels to meet the needs of workers with differing degrees of skills. The lower levels emphasize teaching work and daily living skills, whereas the higher levels have work consisting of basic factory-like assembly work. Although the “gold standard” is community employment, we have found that some people do benefit from sheltered workshops. This may be especially true for people in their forties or beyond who missed out on the job training that is more widely available today. Unfortunately, some workshops still consist of loud, cavernous rooms with repetitious busywork. On the other hand, we have also seen a growing number that offer meaningful work and other activities. In addition, people with Down syndrome are often able to connect to peers in these settings.

Traditionally, workshops have offered piecework and assembly tasks as the primary type of paid work. Better workshops often have a variety of different assembly jobs, as well as other types of work, such as janitorial and shipping jobs, work in cafés or horticulture and garden centers, and a host of other settings and ventures. These workshops usually try very hard to recruit more work and a variety of jobs to keep employees busy and interested. They try to pay based on piece rate, just like any factory, so people who are most productive earn more money. Although this may not be a livable wage, it should still be a fair rate of compensation for a day’s work. We have found that many people with Down syndrome are proud of their paychecks even if they do not completely understand the value of the money. Historically, the wages that many earned from piecework was subminimum. New laws outlawing subminimum wage jobs are causing some workshops to shift their focus or to close.

Many of the better workshops have always also made social, recreational, and exercise programs available to employees during and after work. These activities have also become a bigger focus in some workshops responding to the subminimum wage laws. Aerobics exercise programs are becoming more frequent and not just during downtime but a regular part of the daily work routine. Additionally, we have seen arts and crafts programs in workshops. Some are simply glorified busywork, but there are a growing number of exceptional programs taught by professionals. These programs are beneficial to all participants in building pride and the joy of self-expression.

Many people with Down syndrome may also benefit from a combination of community work and time in a workshop setting. This allows the pride and excitement of a community job while also allowing access to friends and supports at a workshop.

Of course, we do not mean to suggest that all adults with Down syndrome need to work with "their own kind." Some adults with Down syndrome do quite well as the only employee with a disability. However, if you know an adult with Down syndrome who seems unhappy and withdrawn while at work, it may be worth having her try another job where she would have more coworkers with disabilities.

Competence at Home

Residential environments should promote independence, pride, and self-esteem. Parents, caregivers in group homes, or other supervisors need to follow a "good enough" model of direction and supervision. Adults with Down syndrome need to be given autonomy to do what they are able to, yet also receive help and guidance when needed.

Problems develop when people with Down syndrome are given either too little or too much independence. In our experience, too much independence is far more common. This may result from a disturbing tendency to judge a person's need for supervision on her ability to do self-care tasks rather on her level of maturity around certain key issues. For example, many people can independently do routine grooming, hygiene, and housekeeping tasks. However, they may make decisions about nutrition, sleeping, and free-time activities that are harmful to their health, well-being, and self-esteem. Many individuals have the skills to do the tasks but have more difficulty with what we refer to as "executive decision-making." It is the extra steps of organizing the day, being flexible, and adapting to change that can be limited and lead to problems in independent living.

We have also found that many people with Down syndrome in less supervised community settings do not have the skills or initiative to attend beneficial social or recreational programs if they are responsible for organizing these activities. This may be true even for individuals who have the capability to manage all other self-care tasks successfully. As a result, they may become isolated and therefore at great risk for depression or other health or mental health problems (see "The Dennis Principle" in chapter 3):

Peter, age thirty-one, moved from a fifteen-bed group home to a three-person apartment in a more residential neighborhood. His level of skills warranted a move to a more independent living situation. After a year in the new residence, his sister and case manager were concerned because he was becoming increasingly withdrawn and lethargic. He had also gained a considerable amount of weight due to inactivity. Peter had been active in social and recreation programs while living in the larger group home, but in his new residence, he was responsible for scheduling and getting to recreation activities on his own. Although he had the training and the skill, he seemed to lack the motivation or initiative to go to activities. As a result, he spent most of his free time sitting on the couch watching television. Furthermore, Peter was alone most of the time in his new apartment because his two roommates were involved in their own activities. Peter's sister scheduled an appointment with his doctor and social worker when he refused to visit her at her house, which was something he had always treasured.

At the first meeting, it became clear that Peter was depressed because of his social situation. After this, a second appointment was scheduled to include administrative staff from his current group home. In this meeting, the administrative staff initially stated that Peter had a "right" to choose whether to attend social activities. In response, the medical staff reported that his health and well-being were greatly affected by his inability to organize social activities. In the ensuing discussion, agency administrators developed an understanding that not only Peter but a number of other individuals living in less supervised settings were at risk for depression.

By the conclusion of the meeting, a program was developed to provide Peter and others with more choices of social activities. Staff agreed to work with Peter and the others to discuss their choices and desires and then to schedule and transport them to a full calendar of social and recreational events. For Peter, the "social plan" included riding to events in vans with others who lived in nearby apartments. Peter made some new friends among these people and reestablished friendships with others he had met over the years at social activities. After he began his new social schedule, he started to lose weight and regained his positive

mood and spirit. Within nine months, his sister reported that Peter was back to his old self.

For Peter, the good news is that competence and self-esteem will return when appropriate help and guidance are provided. Interestingly, it is often not people with Down syndrome or their families who need to be convinced of the need for such help, but rather residential service providers, who are often strapped with limited budgets and inadequate staffing levels.

Challenging Agencies to Provide Adequate Support

We have discovered that the problem of inadequate supervision shown in the above examples affects many people in community group homes. Agencies have limited finances for staff, but it may also be a little too convenient to attribute inadequate supervision to a "rights issue." We have found that families are often reluctant to challenge agencies to provide more supervision for fear of losing the residential placement. However, an interested third party such as a case management, social service, or healthcare provider may be able to join with the family to advocate for the needs of the person with Down syndrome. With this strategy, the family often feels that there is less risk of jeopardizing the placement.

We believe it is important to challenge agencies when they misuse the "rights issue" and persuade them to provide more supervision and instruction, particularly when inadequate support is a threat to the person's health and well-being. If needed, doctor's orders can be written specifying what is required to assist the person or provide the necessary guidance to help the individual make healthy choices. These orders supersede (legally and medically) any agency dictums regarding rights when the person's health or mental health is at stake. Again, the areas of concern often include more serious problems with sleeping and eating, as well as a failure to participate in beneficial social and recreation activities, which are of critical importance to the person's well-being. By staying active and avoiding a sedentary lifestyle, the individual is more able to stay fit and healthy. This is particularly important for people with Down syndrome, who need regular exercise and activity because of slower metabolism. Regular attendance at social and recreation programs is also essential to avoid social isolation.

Inadequate social supports are not just a problem in residential settings; supports can also be less than optimal when an adult with Down syndrome is living with parents or other family members. In our experience, some teens and adults living at home have little opportunity for social or recreation activities. Sometimes this is due to a lack of transportation, such as when parents are working or unable to drive. There may be different ways to solve this problem. For example, most communities have some form of transportation available for people who are elderly or disabled, such as a cab, van, or bus service. It may also be possible to get rides from the families of other program participants. Case managers of agencies serving the needs of people with disabilities and staff in the recreational programs are often knowledgeable about such services.

Another problem that families sometimes face is lack of appropriate social activities. These families may do well to band together and organize activities. For example, parents in the Chicago area reported a successful "pizza and movie" activity (*NADS News*, January 2004). This began as a get-together for two or three women with Down syndrome at the home of one participant and expanded to include eight to ten people who met at different participants' houses at least every other week.

Now there are a number of these types of groups formally organized by parent groups, special recreation programs, etc., as well as informal groups often done on a rotating basis in family homes. The beauty of this activity is that there is no plan. No one has to conform to any rules or minimum requirements for participation other than to show up at a specified time and place. People simply get together for the pizza and the movie or comparable activity. For people with Down syndrome, who are often told what to do for most of their day, this is quite a welcome change. What is surprising to the families is how the groups have evolved and participants have grown together. Over time, participants have felt free to share their feelings with each other, and a closeness and genuine friendship has developed between members. Participation is voluntary, but most people rarely miss these gatherings. (In fact, we tend to be encouraged by our patients to move an appointment along if it is occurring in the late afternoon when the group is meeting that evening).

More informal types of group activity may be even more beneficial in communities where there are relatively few organized social programs available, such as more rural areas or less populated areas. Parents in these less densely populated locales may need to travel some distance between households for these activities, but again, it may be well worth the effort.

Sometimes other resources are available in communities for organizing these types of groups. For instance, social groups have been formed by graduate students from the special education departments of universities. We have also been impressed with programs that pair teens or adults with Down syndrome with peers in the general population. Many high schools have such programs, which are called different names by the different schools, such as Peer Buddies. These programs often pair one teen peer with a teen with Down syndrome, but they also have group activities with all participants in the program. These programs often help to include people with Down syndrome with the rest of the students, especially in the extracurricular programs, which is where so much of the socializing occurs among high school students.

For young adults, there are similar programs that connect students with teens and adults with Down syndrome and other disabilities. Unlike with the high school programs, the people with Down syndrome rarely attend the colleges from which the students come. These programs go by different names at different universities and colleges, such as Best Buddies and Natural Ties. We have heard many comments from families and people with Down syndrome themselves about the benefits of these programs. Interestingly, those who report the greatest benefit are often the college or the high school students. They often comment on the affection and sensitivity of people with Down syndrome as well as the life lessons learned from them, such as to slow down and appreciate things here and now. We have also heard that many people continue their friendships after they leave college, which is an indication of the strength of these relationships.

If you are creative and persistent, you can probably think of many other options for broadening social horizons. For example, perhaps you could join with other families to hire a special education teacher or professional to organize a social calendar for adults with Down syndrome.

As we are wrapping up the revision of this edition, significant changes in social interactions are occurring due to the COVID-19 pandemic. Social distancing, virtual meetings, and other safety measures to prevent the spread of the virus are being used and developed. Interactions with others via Zoom, Skype, and similar technologies are not the same as in-person gatherings, and some people with Down syndrome are not able to participate in virtual gatherings using these platforms. However, many people with Down syndrome are doing very well with them. They are learning a new set of social interaction guidelines and rules, virtually enjoying time with their friends and family, and creatively participating in a wide variety of activities. At the time of this writing, it is too soon to make too many observations or conclusions about the large-

scale, sudden changes in social interaction. Going forward, however, it will be essential to review successful strategies for in-person social interaction, adjust them to develop virtual strategies, monitor their effectiveness, and modify them as necessary to promote healthy social interactions when in-person meetings are not possible.

"The Right to Choose" versus "The Need to Make Healthy Choices"

Whenever possible, adults with Down syndrome should be allowed to make their own choices and learn from their mistakes. But what if they continually make choices that are harmful to themselves? When and how should a family step in to protect the adult from the consequences of bad choices? The answer to this question will often depend on three key areas of concern: (1) safety, (2) people with influence, and (3) legal issues.

First, we believe that safety is the paramount concern. We think that families should intervene when the person consistently makes choices that put her at risk for physical or emotional harm. We have discussed some of the most common reasons this can occur, such as more serious problems with sleeping, diet, and social isolation. More immediate dangers can involve risks from being unaccompanied in the community or left alone in a home, exposing the adult to such risks as fire, unsavory people, etc.

The second consideration is whether there are differences of opinion as to the amount of independence the person with Down syndrome can handle. Parents, grown siblings, teachers, staff at a group home or worksite, or other people with influence over the adult with Down syndrome may all have differing agendas or philosophies that may compete for influence on the person's choices. This puts the person with Down syndrome in the middle, which may be very stressful. It may also be counterproductive if one of the parties encourages more independence than the person can handle and thus puts her at risk for harm (as in the above examples of Peter and the women in the apartment). In these situations, families may be well advised to work with a third party who may be able to negotiate a more positive solution (see discussion above). Again, agencies that have experience and authority to work with people with Down syndrome may be good candidates for this role.

The third issue has to do with whether the adult with Down syndrome is her own guardian. Guardianship issues may affect how easily parents and service

providers resolve differences of opinion over levels of supervision. Parents who are guardians have the legal right to challenge a service provider to provide appropriate supervision. It they fear losing the placement, they may work in conjunction with a third party (see above). But if the adult with Down syndrome is her own guardian, an agency or service provider may correctly say that she has the right to make up her own mind, even if her decision is harmful. See the section on legal guardianship in chapter 13 for more information about the pros and cons of appointing a guardian.

Step 3: Developing Realistic Dreams

Families of people with Down syndrome often approach the teen and adult years with great concern and trepidation because they believe that the "normal" opportunities for postsecondary education, career, marriage, and establishing a home may be very limited for people with Down syndrome. For example, how do you respond to your son's or daughter's wish to be like siblings and get married, have a home or career, or even become an athlete or movie star? Do you try to squelch this wish because it may never happen?

We think we have a good way to begin the process of encouraging dreams without denying reality. It may first help to look at a huge longitudinal study focusing on typical adults. This study found that people were healthier and better able to cope with life's challenges if they had what researchers call a "dream." This is a general plan and direction for developing their life and career. An important finding from the study is that people did not actually need to meet the goals of this dream to benefit from it. The major benefit was from just having a dream, which served to motivate and encourage them to develop and implement a concrete plan as adults.

Like everyone else, people with Down syndrome have dreams, and not surprisingly these dreams are often to be just like their parents and siblings, to be independent, and to succeed in jobs, relationships, careers, etc. In other words, people with Down syndrome clearly have abstract notions ("dreams"), and these dreams may drive and motivate them to take steps to fulfill their goals. Whether or not you have Down syndrome, success in life comes from taking the dream seriously and then filling in and fleshing out the concrete elements of the dream.

A dream can concern a major issue, like pursuing a career or moving out on one's own, but it may also be less weighty yet still very meaningful to the dreamer. For example, many teens with Down syndrome, like their siblings, see driving as a

dream involving independence and freedom. Images of the excitement of driving abound on TV and in the movies. When you consider your son's or daughter's request to drive, do you just say no to this dream, to protect them or save them from failure or humiliation, or do you find some way to take this seriously and then let them pursue it if at all possible? If you let the person pursue her dream, this might mean letting her take a driver education course, and if she passes the class and then the official road test, she can drive. Many adults with Down syndrome who have taken the course have not been successful, but it is important to note that the vast majority of these individuals were nonetheless satisfied and not discouraged by the attempt because they were given a chance, just like others with their dreams.

As people enter the teen and adult years, there is a focus on "work," but for many people with Down syndrome, this too is a dream (an abstract concept) that may not have concrete form and structure to them. They know work is valued, and they want to do what is valued, but what does it mean to work? Where does this occur, with what people (bosses, other employees, and perhaps customers), and what do you actually do on the job? Fortunately, high school transition programs are allowing most people with Down syndrome opportunities to actually try out different work environments. This allows them to actually experience different job sites and see what "fits"—in other words, to fill in the concrete form and structure of what it means to work.

How do you help the person with Down syndrome leave the relative safety and comfort of school for "adult life"? This is a huge change and an area of great confusion, mystery, and anxiety for many people with Down syndrome as well as their caregivers. One good place to start is to try to determine what the person with Down syndrome already understands about "adult life," including postsecondary education, establishing one's own household, and pursuing a career.

Postsecondary Education and College

Some people with Down syndrome profess an interest in college and postsecondary programs because their siblings or peers are going off to college. As discussed earlier in the chapter, there are now far more postsecondary opportunities available. But is this the right fit or approach for a particular individual? Junior or community colleges may be one way to test this idea because most people can commute to the college and stay at home or nearby where caregivers are still readily available. Going off to college is a more challenging option. Many typically developing

young people have difficulty adjusting to the academic and social-emotional demands of college life apart from the family. Compounding this problem, many young adults do not tell parents they are struggling, and the school protects students' privacy and rights (because they are legally adults).

How, then, do you test out whether the abstract concept fits the concrete reality for your son or daughter, whether or not he or she has Down syndrome? It may be helpful to do trial runs to see how your son or daughter manages in an environment away from home such as a camp, Special Olympics outings, or summer weekend programs. Also see the earlier section that discusses what else can be done to explore support on campus.

The Dream to Live Independently

When people with Down syndrome have a dream to live independently like others their age, how do you help them fill in the concrete details to flesh out this dream? Here you can definitely rely on their strength for visual and concrete thinking to break down the dream into doable concrete steps. Living independently means being able to complete a host of tasks that can be managed through checklists and calendar notations and worked on while still at home. As with pursuing a driver's license, even if an adult does not end up achieving her independent living dream, she can learn and develop significantly while trying, and again, for many that may be enough.

They are many alternatives to living independently or semi-independently in the community. How do you help someone develop a realistic picture of the options without destroying her dreams? Again, the best way is by filling in the concrete details of this reality. Adults with Down syndrome need to visit the types of residences available to actually see and experience what they are like. For instance, most group homes let prospective residents visit for dinner with other residents and stay overnight. The closer someone with Down syndrome can get to the real experience, the better able she is to understand what the concept means and to then decide whether the option is the right one for her, based on the reality and not a vague concept.

What Is Their Dream?

We also need to listen carefully to what the dream actually means to teens and adults in their own (concrete) reality. It may help to remember that people with Down syndrome often find it difficult to use speech to communicate, and thus it may take some time and effort to really understand their dream. As an example of the process involved, our colleague Bryn Gelaro, an experienced social worker in Denver, tells an interesting story of a young man whose brother had recently moved into his own apartment. Jim, the young man with Down syndrome, told his parents that he too wanted to "live in an apartment" like his brother. His parents were dismayed because they did not believe he was capable of this and would not even like living alone. Still, they wanted to take Jim seriously, so they helped him begin to do more self-care tasks on his own, in preparation for an eventual move to an apartment. But at the urging of Bryn, they asked him for more concrete details about his wish and asked in more creative ways.

One day when Jim's parents were with him in his brother's apartment, they asked him to show them what he liked about the apartment. He pointed to the refrigerator numerous times. From this, his parents gradually realized that what he liked about the apartment was that his brother had his own fridge and could get drinks any time he wanted. This was quite a revelation to his parents. They learned that he wanted to have this freedom but not necessarily live alone. With this insight, they began to look at the basement of the family house as an alternative to an apartment. They spent time and effort cleaning it up and designing the space according to his interests and needs, which included having his own fridge full of drinks, which made him very happy. His dream and the reality were in sync because his parents were eventually able to let him fill in the concrete details of his dream.

The take-home message is that just because we think we understand what an individual's dream is doesn't mean that we actually do. In addition, the person with Down syndrome might not always mean what it sounds she is saying. For example, when an adult or teen says she wants to go to college, she might mean that she wants to spend additional years listening to classroom lectures about academic subjects, doing homework, and studying for exams. But she might also not understand what going to a traditional college truly entails. She might actually mean that she wants to move out of the family home like other young adults do and/or have more freedom to decide how to spend her free time.

Step 4: Developing Unique Talents and Characteristics

In addition to being allowed to pursue dreams and developing a sense of awareness and a sense of competence in essential tasks at home, school, and work, there is one more area that is essential to the self-esteem of adults with Down syndrome. That is the identification and development of the individual's unique talents and gifts.

Some people with Down syndrome have conventional talents that others readily recognize—whether it be writing poetry, public speaking, playing a musical instrument, creating art, acting, swimming, or something else. Others are truly gifted in people skills, such as being able to read emotions or bring out the best in others, although communication skills may make their talents difficult to appreciate at times. Still other people with Down syndrome may not have talents that are readily appreciated by outsiders, but instead have relative strengths that friends and family members are aware of.

In fact, we have found that there is often a great unevenness in the individual's skills. This is true for all of us, but the unevenness is often far more pronounced and has a more profound effect on people with Down syndrome.

So how can we help people uncover and appreciate their talents? Two things may help. First, a guiding principle in the field of social work is "to start where the client is." For people with Down syndrome, this means we look at the person's progress in developmental areas and not her chronological age. Parents have a great advantage in assisting in this process because in our experience, no one knows the person with Down syndrome like parents and caregivers. Parents simply need to trust their knowledge and intuition about their child's abilities and passions. Second, we recommend that parents practice a concept in developmental psychology called "good enough" parenting. This means that the parent is available for love, support, and guidance when needed, but the child is allowed to experience manageable amounts of frustration and failure as an incentive to learn and develop her abilities.

Whether or not an adult is a "superstar" in the Down syndrome universe, she needs to be encouraged to develop her talents and be proud of her skills. For people with Down syndrome, who are often judged more by what they do not have or are not able to do, this is a way for them to say, "I am more than that."

Like anyone who is part of a minority group, an adult with Down syndrome wants to be seen both as a member of the group and as someone who possesses her own unique talents. These talents define the person as much or more than the Down

syndrome does and, therefore, need to be identified and nurtured. Most families know how important this is. So often we hear family members make statements like this: “Sure, she has limitations, like others with Down syndrome, but did you know that she . . . is an artist . . . can do this job better than anyone . . . has changed our family . . . is especially sensitive to others’ feelings and needs . . . has an exceptional memory,” etc. The pride and respect expressed in these statements is so important to people with Down syndrome. It says to them that they have something special and unique to contribute to their families and the world. A person may have limitations in some areas but strengths and talents in other areas, and this is who and what she is.

Dos and Don’ts for Nurturing Gifts and Talents

Here are some suggestions for helping an adult with Down syndrome to identify and appreciate her own unique talents and characteristics (with examples from different talent areas):

- Assume the person with Down syndrome has talents and gifts of some kind.
- Expose her to a wide variety of different activities to help identify these talents and gifts.
- Don’t assume she doesn’t have skills in certain areas. Try everything.
- Encourage talents that the person with Down syndrome shows real interest in. If it comes from the person herself, there will be true interest and pride from the talent.
- Look for ways to nurture the talent she is interested in developing. For example, if she is interested in art or music, it may help to have instruction from an appropriate teacher. If she is sensitive to others, find an avenue to express this (such as volunteering in a nursing home or at a good childcare program). For athletes, find different sports and recreation venues.
- Find ways to encourage talents at home. For example, for artists or musicians, have a place to work or practice with the appropriate instrument or art materials. For those sensitive to others, encourage them to use this talent with family and friends. For athletes, take time to play together or organize neighborhood sports activities.
- Encourage, but be careful not to overpressure. Nothing dampens a person’s spirit and energy like too much pressure from others.

- Take time to observe or acknowledge the person's talent. For example, look at her art, listen to her music, observe her volunteer at a nursing home, attend her sports activities, etc.
- Offer sincere praise, but don't overdo it. People with Down syndrome usually know when praise is not genuine.
- Excessive praise may increase the person's interest in doing the talent to please others rather than enhance her own self-esteem.
- Praise often flows naturally from others in the community when the person with Down syndrome puts her heart into her talent. (See the example of Emily, immediately below.) Sports, artistic endeavors, and other types of activities also generate praise from others, including peers (which is a valued form of praise).
- Finally, when you offer praise, encourage self-pride rather than pleasing others. For example, say, "You should be very proud of yourself" rather than "I'm so proud of you!"

One mother was concerned about how her twenty-nine-year-old daughter, Emily, would respond to her grandmother's deterioration from dementia and her move to a nursing home. After delaying a visit to the nursing home for some time, Emily's mother finally took her to visit her grandmother. Her mother was amazed and immensely proud to find that Emily was not only unusually sensitive and caring toward her grandmother, but she also responded sensitively to other nursing home residents, particularly those who were lonely and in greatest need of care. Emily's mother let her know how proud she was, but the residents who benefited from her caring interest also expressed their gratitude through their words and facial expressions. Emily returned to the nursing home many times before and after her grandmother's death. Eventually, the administrator of the nursing home asked her to continue as a volunteer, which she has done with much benefit to all, including herself.

A Word of Advice for Would-be Actors

Many teens and adults with Down syndrome dream of having a career in acting. Gail Williamson has been a talent agent in Los Angeles for over twenty-five years and has an adult son with Down syndrome who works as an actor. Gail has worked with many actors and actresses with Down syndrome, helping them pursue their dream of acting. She offers the following advice:

- Don't quit your day job. Rarely does acting lead to a real career, even for those who do find roles.
- Take many acting classes. Like every other dream, acting is a concept that has to be grounded in reality. Acting may look easy when done by professionals, but it requires a great deal of time and practice, like any skill.
- Location is everything. People living in Los Angeles or New York have the best chance of getting to auditions. Otherwise, Williamson recommends joining local theater programs, which may help to satisfy a person's desire to be onstage.

Love, Friendship, and Self-Esteem

As discussed earlier in the chapter, there are three key factors in the development of self-esteem: (1) accepting one's identity, (2) developing competence, which includes both nurturing a dream and understanding concrete reality, and (3) understanding one's own talents and gifts. A fourth, equally essential key to self-esteem is feeling that one is loved and lovable.

So much of counseling for all people revolves around their perceptions of being unloved and unlovable and helping them find ways and means for obtaining love in their life. While some people with Down syndrome clearly have difficulty finding the love they need, many are quite adept in this area. In any event, most people with Down syndrome are very sensitive to, and aware of, expressions of love. Most are also keenly aware when these expressions are absent in their lives. Often, they are so good at eliciting love that they are able to change the amount and intensity of love expressed in the family. Like all skills, this may have enormous benefits, but it may also have some negative consequences, as discussed below.

Peer Relations

Peer friendships are critical to everyone's health and well-being. Peer friendships are different from parent or teacher relationships, but they serve an

equally important role in the development of self and self-esteem. Like a parent-child relationship, a peer relationship involves the expression of positive feelings and support, but it also provides the all-important feeling of fitting in with one's peer group. Peers share common interests, struggle with similar developmental tasks and issues, and serve an important mirroring role in the formation of one's identity. Peers with similar disabilities play an even greater role in showing the way to pride and self-respect despite whatever limitations the disability causes. Peers with a disability who are in the public eye, such as actors, artists, musicians, and presenters at Down syndrome conventions, play an equally important role. They project a positive image of Down syndrome that people can take pride in and aspire to.

And yet, some families and researchers call into question the quality of peer friendships between people with Down syndrome and other disabilities. They believe that friendships between two people with Down syndrome may be less rewarding because one or both individuals may have trouble initiating and maintaining conversations and difficulties in showing interest in and taking another's perspective. On the other hand, many families report that even when there is an apparent lack of interactional skills, peer relationships are usually strong, longstanding, and critically important. Typically, these peer relationships develop over much time and familiarity, such as when people are in the same job or school program over many years. Although these friendships may take more time to develop, once established, they are an essential source of support and self-esteem.

Families often report that their family member with Down syndrome prefers to talk to the staff at social events where others with Down syndrome are present. But this does not mean they are not interested in socializing with or connecting to others with intellectual disabilities. We believe it is simply easier for them to talk to staff because these people are generally skilled at initiating and maintaining a conversation and comfortable with and knowledgeable about people with Down syndrome. It is interesting to note that events in which people can do things such as dance or play a game or sport are often easier to participate in than a cocktail party–like event where participants are expected to socialize by talking.

A very interesting study looked at what happed to individuals with disabilities when the large institutions in Illinois were closed. These institutions had housed people who had been institutionalized from birth and who had been described by staff as having few, if any, social skills. Yet when these adults were moved to smaller facilities, they tended to do better if they moved with other individuals from their wing. We cannot underestimate the connection people with Down syndrome have with

each other even if the verbal or social interaction appears to be limited (Heller, 1982).

Alienation

Families who have some difficulty accepting Down syndrome sometimes discourage the development of relationships with peers who have disabilities. Other families may inadvertently discourage these relationships if they are so intent on "inclusion" of their child with Down syndrome at school and in community activities that she rarely if ever encounters someone else with Down syndrome. This is not to say that friendships with peers without disabilities are not possible or that they are not extremely beneficial. However, these relationships are not as common as parents may hope, and when they do develop, they may be difficult to maintain over the long term.

Discouraging friendships with peers with disabilities is unwise. Some of the saddest people we have encountered are people who do not want to associate with peers who have Down syndrome (or other disabilities). These individuals can be caught between two worlds and have difficulty maintaining a positive self-image. On the one hand, they are not always readily accepted by typically developing peers, or they lose touch with them over time. On the other hand, they voluntarily cut themselves off from peers with disabilities who could be their friends and who often remain in the area when nondisabled peers leave for college or other typical adult pursuits.

Some of the individuals we have served live in what we describe as an "existential hell." Their high school friends without disabilities have moved on to other pursuits. And yet, they do not want to associate with people with disabilities. Furthermore, they struggle with their own identities because of their inability to accept or deal with having Down syndrome. They find themselves isolated between two worlds and aren't even comfortable with themselves. The approaches discussed in this chapter are much better used as prevention strategies to avoid this occurrence. However, if it does occur, we recommend starting with the concepts discussed at the beginning of this chapter and working on establishing or reestablishing self-esteem.

Dos and Don'ts in Encouraging Peer Friendships

In the meantime, there are some things families can do to encourage the person with Down syndrome to interact with, rather than to avoid, others with disabilities:

- Encourage participation in Special Olympics and other recreation activities for people with disabilities. This is important even if the person resists joining these activities. We find that teens and adults with Down syndrome often get caught up in the team effort, which helps to create a rapport and positive experience with teammates.
- Try to find situations where the avoider has to help another person with a disability. For example, have her teach a coworker how to do a job task or a housemate how to do a chore. This strategy does three things:
    - The incentive to show that she can do a good job at the task will often get the avoider past her initial reluctance to interact with others who have a disability. In setting this up, it may be helpful to appeal to the helper's skill in doing the job.
    - Taking on the role of helper will often change the avoider's attitude and demeanor from negative or apathetic to positive and helpful.
    - This in turn will usually change the attitude and response of the person who is being helped. This is important because many people with Down syndrome feel they are not liked or appreciated by others and usually avoid those who respond negatively to them.

  If the avoider has a positive experience as a helper, this may change her attitude and subsequent behavior toward others with disabilities. After a number of these positive experiences, she may develop a more permanent positive attitude toward others with disabilities.

- Find or create opportunities to interact with people with disabilities other than Down syndrome. We have found that some people who avoid others with Down syndrome are more open to interacting with those with different types of disabilities, such as a physical handicap. For example, some adults with Down syndrome enjoy pushing people in wheelchairs. You can then capitalize on this to praise the person for her sensitivity. Later you may be able to point out that Down syndrome is simply a different type of disability than a physical disability, which may help her be more accepting of her own disability.

- See if the adult is less resistant to assisting younger people, such as children in daycare settings. Many times the children they help look up to them as heroes and role models. This may not only help them view themselves and their Down syndrome more positively but also benefit the children.
- Arrange for the adult to attend Down syndrome conferences and conventions. At these conferences, she will be surrounded by people and programs that are supportive and positive about Down syndrome. Perhaps more importantly, she may be influenced positively by leaders and self-advocates with Down syndrome who have found acceptance and self-pride despite their own disability. There may also be leadership opportunities or other responsibilities in the self-advocate group that promote self-esteem.
- Keep a careful eye on your own and other caregivers' attitudes and behaviors toward individuals with disabilities. Your attitudes will strongly influence the person with Down syndrome. Even if you feel you are hiding your attitude, any negativity will often be detected by your family member with Down syndrome. If you do not have a positive attitude toward people with disabilities, try talking to someone who can help, or join a support group for parents.

Encouraging Friendships for Shy or Inexperienced Adults

Sometimes adolescents and adults with Down syndrome avoid others because they are shy or have little experience socializing—not necessarily because they are resistant to interacting with people with disabilities. These are some successful strategies used by families of teens and adults with Down syndrome:

- Encourage participation in Special Olympics and similar recreation activities. Participating in structured activities is far easier than participating in more unstructured social events. Over time, people often become more comfortable interacting with others, especially when caught up in enjoyable team activities.
- Dances may be a surprisingly good way for shy adults to interact with others. Many people with Down syndrome love to dance, alone or with others. At dances attended primarily by people with disabilities, there often does not seem to be the same type of social pressure that accompanies dances for teens or young adults in the general population.

- Participating in programs for teens and adults at Down syndrome conferences and conventions can be a good way to connect with peers. These programs often include activities that allow people to comfortably interact and may include activities specifically designed for building confidence in social situations. Additionally, experienced self-advocates in attendance are often good role models. This can translate into more confidence in social situations with peers.
- Volunteering or working at programs serving young people or children with Down syndrome may help to build more confidence in social situations. Again, if the younger people look up to the older person who is helping, it helps to build pride and self-confidence.
- Participation in Buddy Walks or other fundraisers may help instill confidence, particularly when participates offer strong support for people with Down syndrome. There is also exposure to self-advocates who have strong pride and self-esteem.
- Try to develop casual get-togethers for peers with Down syndrome such as the pizza and movie night described above. People often feel more comfortable attending these sorts of planned activities, especially in a comfortable home environment. Also, having different participants host the get-together allows them to develop confidence in taking care of others in a social situation. As an added incentive, people are often able to show off their music and other hobbies and interests, which can build interest and rapport with others.

Summary

Families who are most successful at promoting self-esteem in teens and adults with Down syndrome do the following:

- develop an understanding and acceptance of Down syndrome and encourage the same in their child with Down syndrome;
- develop an awareness of their family member's limitations as well as her potential for developing independent living skills and more unique talents and abilities;

- promote self-esteem and independence through competence—taking their child's dream seriously and teaching her the skills needed for self-care tasks and for meeting day-to-day challenges of adulthood;
- encourage expressive language skills and social relationships with peers;
- encourage helpful "grooves"—especially those that assist the person in reliably completing self-care and job tasks (see chapter 10);
- encourage participation in social and recreational activities;
- try to find the right school, work, or residential program to meet their son's or daughter's needs and abilities;
- encourage staff to build on their child's existing strengths and positive experiences to increase confidence and motivation in these environments.

Finally, families who do the best job of nurturing self-esteem do not just focus on their son's and daughter's deficits and on ways to overcome them. Instead, they develop an appreciative understanding of the areas in which their children excel. For example, they celebrate their son's or daughter's empathetic understanding of others' nonverbal language or their ability to teach valuable lessons on slowing down and on experiencing the here and now. In short, they allow themselves to see their child's strengths and then use that appreciation to fuel their efforts to develop their child's self-esteem.

Return to Table of Contents

Chapter 9

Self-Talk Revisited: A Tool for Learning and Responding to Challenging Tasks

Jason, age thirty-one, was seen for his annual evaluation. His mother asked about Jason talking to himself. She reported that if he got upset, he might talk loudly to himself in the family room, but most of the time, he did it quietly in his bedroom. His parents could often hear him through the bedroom door, and the subject matter was usually the events of his day. Neither Jason nor his parents were concerned, but at the urging of a family friend, Jason had been evaluated by a psychiatrist, who had recommended an antipsychotic medication. They had not filled the prescription and were seeking a second opinion.

Most people with Down syndrome use self-talk in their childhood, but as many as 81 percent continue to talk out loud to themselves throughout their teen and adult years (McGuire, Chicoine, & Greenbaum, 1997). When we first started evaluating adults with Down syndrome (in the early 1990s), we found that families were reluctant to tell us that their teens and adults had this behavior. Parents were justifiably concerned that others might view talking to oneself as indication of a mental health disorder such as psychosis or schizophrenia. In those early years, we did in fact see many adults with Down syndrome who were being treated with antipsychotic medications simply for talking out loud to themselves, and unfortunately this still happens today.

For over twenty-five years now, in our articles, books, and many hundreds of presentations, we have discussed the fact that self-talk is common in the teen and adult years and not usually a symptom of mental illness. We see self-talk as a social skill issue much like learning about public displays of affection, such as when, where, and who to hug in public.

Still, concerns about self-talk persist among many parents and professionals for several reasons. The first is that self-talk can be a major concern when conducted in

a public setting. In this chapter we will discuss why this may happen and what to do about it.

Self-talk can also be disturbing for two additional reasons:

1. It can appear dramatic to the observer, even when conducted in a private space and even when the content is positive. We will discuss why and how this happens and why concerns about these types of private displays are unwarranted.
2. Self-talk may also consist of negative or self-critical content, which is disturbing both because of the intensity of the presentation but also because it may mirror something that is actually going on in the person's life. We will explore why and how this happens as well as what to do when this occurs.

What Is Self-Talk?

We need to be crystal clear about what self-talk is to put concerns to rest. As mentioned above, we have shown self-talk to be a social (and not pathological) behavior. But we have also found both support and validation of our clinical experience, as well as findings in the research, some of which were quite surprising and far exceeded our expectations and understanding of self-talk.

For example, we were astonished to find that self-talk is one of the most revered concepts in the field of human development. This is because it plays a key role in the acquisition of a new developmental skill and simultaneously in the development of mental thought processes.

Perhaps the best way to explain how self-talk plays a role in mental development is to describe the steps in this process.

1. First the child observes or is shown a new task.
2. The child then borrows the parent's or teacher's words to literally *talk himself* through the new task in a process called guided action. This happens over and over with each new task in the child's development.
3. As children get older, they may use fewer words when talking to themselves, using only enough words to get the task done.
4. As children get older still, the self-talk will often evolve into *inaudible muttering* which is soft but still audible to a careful listener.

5. In time, the self-talk, is internalized into the child's own thoughts or his *inner speech*. As we mature into our teen and adult years, this inner speech then becomes the basis of our inner thoughts and what researcher Laura Berk calls "the conscious dialogue we hold with ourselves while thinking and acting."

Why then do so many teens and adults with Down syndrome continue to talk out loud to themselves? There are four compelling reasons for this:

First, although most typical children internalize their self-talk by six or seven years of age, children with a variety of challenges tend to internalize their self-talk as late as age ten or eleven. This includes children who live in impoverished environments as well as children with visual, hearing, or attention deficits. This is also clearly the case for teens and adults with Down syndrome, who have far greater challenges throughout their lives in expressive language and adaptive functioning than more typically challenged children.

Second, although most "typical" adults generally internalize their self-talk, research still shows frequent use of self-talk by adults, especially when doing newer or harder tasks. Again, this self-talk may be muttered and barely audible, but it remains a tool that helps us overcome obstacles and acquire new skills.

Third, like other teens and adults, people with Down syndrome may use self-talk to solve problems or challenges. But they may not be as sensitive or aware of the need to hide self-talk when near other people (Patti, Andiloro, & Gavin, 2009). Families often discuss this fact with some degree of amazement. For example when their son or daughter is criticized or told something that is not to their liking by the parent, the person with Down syndrome will often go off (although still close enough to be overheard) and talk out loud about his or her parent, such as to say "darn (or damn) her… she should mind her own business." This may even include such statements as "what a #+&$! miserable b— or SOB of a parent." If the parent approaches the person with Down syndrome with a comment like "what are you saying about me?" the person with Down syndrome will often be incensed because he assumed this conversation was private and that his private thoughts have been violated. Clearly, people with Down syndrome don't always understand that they are overheard.

Fourth, many people with Down syndrome may also have difficulty discerning what is and isn't private. For example, we have been called quite often to schools or worksites by concerned staff because a person with Down syndrome is talking out loud

in one of the public bathroom stalls. For the person with Down syndrome, the space may appear private (just like a bathroom at home), but it is clearly not.

Typical Examples of Self-Talk

We occasionally have a need to call computer tech staff because of problems with computer hardware or software. The tech staff will often stay on the phone while remotely logging onto our computers to solve the problem. Often the tech person is heard talking out loud to himself when walking through the steps to try to solve the computer problems.

In other settings and situations researchers have reported similar findings. For example, teachers and lecturers were found to use self-talk frequently to help organize their presentations, such as by stating something like "I talked about that already . . . let's see . . . maybe I can talk about this now." Similarly, adults learning a second language often use self-talk to help facilitate conversation in the second language. Audible self-talk has also been found to be quite common among adult athletes to help motivate themselves in competitive situations or when challenging themselves to do their personal best. In other words, the more researchers look for self-talk, the more they find regardless of age, stage, or presence of a disability.

Managing Self-Talk in Public

Given the research findings on the universal nature of self-talk, it seems reasonable to assume that we may all use self-talk, but we may simply be better at hiding this fact than most people with Down syndrome. Still, self-talk serves the same adaptive functions for all of us.

When teens or adults use self-talk in public, there are obviously some important social considerations. When viewed as a social skill, we can say that learning when and where one can talk to oneself is not that different from any other social skill, such as learning when, where, and who to hug, or even when and where to express one's sexuality through masturbation. Everyone, whether they have disabilities or not, needs to learn to curb these behaviors in public settings. (For more on other social skills such as displays of affection, see chapter 13.) This may be even more important for people with Down syndrome because they have distinctive physical features that may already draw attention from others.

Below are some strategies for managing self-talk in public.

Develop "Inaudible Muttering." As discussed previously, most people without an intellectual disability use "inaudible muttering" when talking to themselves, and we have found that people with Down syndrome can learn to do this too, especially when they encounter a challenging situation in a public setting. Like other social skills, this skill should be introduced when the person is young, as it may require time and practice. Use the person's visual strengths to help him learn. As discussed below in "Self-Talk on Steroids and in chapter 6 on visual memory, many people with Down syndrome are highly responsive to visual cues and the visual arts (movies, pictures, and theater) and acting, role play, or theater-like practice sessions can be a wonderful way for them to learn. They may practice keeping their self-talk under their breath with the assistance of parent, teacher, or some other coach.

Use Visual Reminders. Remember that the written word is a visual medium. Thus, a reminder to the teen or adult to talk under his breath, written on a notepad, electronic tablet, or smartphone can be very effective if he is a visual learner. It may be particularly helpful to write out and carry a reminder when entering a situation that may be challenging for any reason.

Find Teachable Moments. A "teachable moment" simply means that the teacher takes advantage of any opportunity at any time of the day to practice the skill, especially when the person with Down syndrome is in a natural or public setting. This is also useful in helping the person generalize a skill learned in one setting to another. (See chapter 5 for more on the issue of generalizing.) As with all teaching experiences, the teacher should be respectful to the learner with Down syndrome. For example, this strategy should be employed by someone who is a clear authority, such as a parent or teacher, and not necessarily by a younger sibling, unless this is respectful and accepted by the person with Down syndrome. Additionally, giving the information in a positive way can reduce teen-like resistance to learning.

Decide on a Subtle Reminder. We have found that it may be best to remind people to keep their comments under their breath by using an unobtrusive, prearranged gesture. Some have called this their "secret sign." This is particularly important as the person enters the teen and adult years and is more sensitive to his social environment.

Disguise Self-Talk with a Cell Phone. People can temporarily use cell phones or even Bluetooth devices (which are usually stuck in their ears) to talk into while they are learning to manage their self-talk. Talking on a cell phone may be considered rude by some, but it is not seen as a sign of pathology in the general public. This is

not an optimal long-term solution, however, because ultimately this habit needs to be unlearned if the person is going to truly manage this behavior. Also, using a cell phone is not usually tolerated at school or work, so it may be of limited use in these settings. Still, it may be a stopgap measure used as a temporary strategy when the person is in a new setting and self-talk makes the person too susceptible to a label of mental illness by uninformed others.

Self-Talk on Steroids: Dramatic Sessions with Imaginary Friends

If we only had to be concerned with simple conversations to oneself, then the self-talk would probably be more easily explained, justified, or managed. But the concern and controversy about self-talk continues in good part because of how dramatic it can look at times. Self-talk often includes the full range of facial expressions, hand gestures, and body movements, often appearing as if the person is on a stage and fully enacting a scene with at least one and quite often a host of imagined others. In these instances, self-talk becomes a three-dimensional representation of the person's thoughts, memory, feelings, emotions, and behaviors. Self-talk sessions that include pretend play and imaginary others often occur in private spaces such as a bedroom but can still be more than a little disturbing to observers, even close family and friends.

Is this "self-talk session" something to be truly concerned about, or is it actually creative and beneficial? Once again it may be helpful to go back to the literature for guidance about pretend play and imaginary others.

Pretend Play

As with self-talk, child development researchers have found that pretend play has a critically important role in a child's development. Pretend play gives typical children a wonderful stage or laboratory to try out and practice what they see and experience out in the world. Despite the use of the terms "pretend" or "fantasy play," this is rarely fantasy and most often it is a means to process and understand the real world. In most cases even the lives of fantasy characters, such as from favorite TV and movies, also mirror the real world or they may offer a way to manage real-world fears, challenges, and situations. For example, the child may try out different means for managing fears and anxieties by acting like a policeman, superhero, etc.

We have found that these same issues apply to the bedroom "self-talk scenes" enacted by people with Down syndrome (of any age). Families report that the scenes enacted may center on the person's experiences in the real world. As discussed in chapter 6, people with Down syndrome often have exceptional visual memory for scenes and situations and can accurately recall and then act them out as they experienced them. For example, many parents hear their son or daughter taking on the role of the boss at their job and then give orders to (imagined) others in the room. Sometimes dolls, stuffed animals, or action figures are assigned roles in the play. Other people with Down syndrome may act out scenes from other parts of their lives, such as by pretending to be the activity director at the fitness center or a priest saying Mass. Anything that is out there in the world— especially scenes that are important, puzzling, or memorable to the person with Down syndrome—can show up in self-talk sessions.

Imaginary Others

Just like younger children, some teens and adults with Down syndrome may have play sessions that include fantasy characters or characters from TV or movies. Witnessing the sound and fury of a stage-like session may be disturbing enough to observers, but noting the presence of one or more imaginary others within a self-talk session may be even more disturbing. Sometimes listeners worry about mental health concerns such as hearing voices, hallucinations, etc. Rest assured, however, that the clear majority of people with Down syndrome who talk to imaginary characters are quite sane.

It is important to remember that these imagined others are simply players in the person's self-talk session (in his private laboratory). Additionally, imagined characters often mirror people in real life and offer ways to manage challenges, feelings, and anxieties. It may also help to know that when typical children play alone or even with other playmates, they frequently talk out loud to themselves and to imaginary others. Again, the reason children or adults with Down syndrome talk to imaginary others during play or self-talk sessions is to replicate interactions and conversations with others at work, in social situations, and at other interesting or meaningful events.

Special Imaginary Companions

Many people with Down syndrome have one or more special "imaginary companions" who often appear in their self-talk sessions. For typical children and for teens and adults with Down syndrome, imaginary companions often have a special role, such as to offer support, comfort, and companionship. Some adults with Down syndrome may maintain these imaginary relationships for many years or even decades.

For people with Down syndrome, an imaginary companion may be one of three types:

1. Although imaginary means "in the imagination and not real," we have found the most common imaginary companion to be a real person who played a significant role in the person's life, either past or present. What makes the person "imaginary" is simply that he or she is not present at the time of the self-talk session.
2. This special companion could also be some type of celebrity character from a TV show or movie, a recording artist (live or deceased, such as Michael Jackson or even Elvis), or a pro wrestler. We have found some blurring of fact and fantasy about these characters. Many people with Down syndrome may have some difficulty understanding that this is an actor and not a real person. Still, in all but a very few cases we're aware of, these fantasy characters remained as players in the self-talk scenes and were not expected to be a part of the person's real life. Even if the adult with Down syndrome met the actor or musician in real life, he expected the actor or musician to continue to perform as usual and not become a real part of the person's life. As a variant of this, some people with Down syndrome have a special relationship with "inanimate" characters, including stuffed animals, dolls, or action figures representative of characters from movies or TV (e.g., *Grease* or *Glee*) or animated movies such as *Frozen*.
3. Finally, this special companion could also be a made-up fantasy character, which is what has been typically thought of as an imaginary companion. We have found these types of made-up imaginary friends to be less common among the teens and adults we have evaluated. However, we have found that inanimate objects such as dolls, action figures, and stuffed animals are often assigned unique and creative roles.

Not surprisingly, there is some controversy about the "normalcy" of having imaginary companions. In earlier times, there were concerns that children with these companions were "possessed" or, more recently, that they had a mental illness. Fortunately, the child development literature has challenged these and other misconceptions about imaginary companions, and the findings can be readily applied to teens and adults with Down syndrome who also have these special companions. For example, researchers in this area have learning the following:

- Up to 65 percent of all children have imaginary companions at one point in their childhood; some have companions through the school years and even into adulthood. Although we can't cite the percentage of the many thousands that we have evaluated, a significant number have been observed to have special companions in their self-talk sessions.
- Most children with imaginary companions are social and capable children whose primary reason for having an imaginary companion was just more companionship. Similarly, we have found that teens and adults with Down syndrome who tend to be social with peers also tend to be social with imaginary companions in their self-talk sessions.
- Children who don't have siblings tend to have imaginary friends for companionship, but researchers have found that children with plenty of siblings or available play partners also have imaginary friends. "Only children" may spend more time with imaginary companions simply because they tend to have more alone time. We believe that teens and adults with Down syndrome are even more likely to have special companions than only (typical) children because they are often less likely to have real companions and visit them at their houses and therefore may have more free time than most to play with a special companion.
- Time with an imaginary companion is generally viewed by a child as quality time. This is also the case for teens and adults with Down syndrome, as repeatedly confirmed by their families and caregivers.
- Finally, and contrary to widespread beliefs, typical children and adults with Down syndrome generally understand that their imaginary companions are imaginary or at the very least that they are not actually present during self-talk sessions. For example, one researcher reported numerous instances in

interviews with children who took her aside to say, “You know these are just imaginary, don’t you?” (Taylor & Mottweiler, 2008).

We have stories from many families of teens and adults with Down syndrome corroborating that these individuals usually understand that their imaginary friends are imaginary. One mother related a story about her twenty-two-year-old son, Anthony, that occurred before they came for an evaluation. Tony’s mom had heard him night after night in his room “talking up a storm to a host of people.” She finally had enough and one night banged on the door and yelled, “Who are you talking to in there?” Several minutes later the door flew open, and he yelled back with conviction, “Myself—who do you think?”

Alvin’s family reported a similar scene. In this case Alvin’s mom had also had enough, and she went to her son’s room and said with some edge to her voice, “Who are you talking to in here?” Her son’s response was to stand up and look slowly from left to right while saying, “Do you see anyone in here, Mom?” From this and countless other conversations with families in similar situations we can surmise that usually the person with Down syndrome realizes that these imagined others are not really there. Even when there is some blurring of lines of identity such as the true identity of a character in a TV show, the person with Down syndrome usually knows the person is not really present in the room.

What We Know about Self-Talk in People with Down Syndrome

From the preceding discussion, we know that self-talk is so much more than just talking out loud to oneself; it is one of the most important tools we as humans possess to learn, adapt, and manage our thoughts and behaviors in the world. We will now describe more fully what self-talk is and can be for people with Down syndrome, taking into consideration their unique characteristics, which can have a big influence on the form and expression of self-talk. To this end, we will discuss two different dimensions of self-talk. We will first look at some basic issues that may greatly affect how people with Down syndrome use and respond to the self-talk process. We will then look at the purpose or function of self-talk for people with Down syndrome. Finally, we will discuss when self-talk in an indication of a problem that needs attention.

Self-Talk: When and Where Displayed

As mentioned, learning when and where one can talk to oneself is not that different from learning any other social skill such as when, where, and who to hug. Learning when and where self-talk is appropriate is very important because mental illness may be suspected if the person talks outside of his private space. The lesson from all of this is to heed the importance of social skill training to avoid this possibility.

We hear from many families that a regular part of the teen's or adult's schedule is to spend alone time in his room reviewing the day's events, through what is often an extended period of self-talk. In this case, it's usually easy for families to tell what the person is talking about. But if the person is reviewing something that occurred in the past, it can be very confusing to caregivers, especially caregivers who do not have shared history with the person with Down syndrome. The past may be something as recent as the early part of one's day or as old as an event from early childhood (sometimes up to thirty years or more in the past). For example, people may recall a past holiday or birthday, in great detail, going back to almost any time in the past. This may be analogous to a video replay, but in this case the person is replaying events from his own (longer-term) memory bank.

Self-Talk with and without Props

As mentioned previously, it is not uncommon for people to use objects during a self-talk session. These objects will often have a specific purpose in the session such as to represent objects in a scene but may also represent a specific person such as a parent, friend, boss, special companion, or even the person themselves. In most cases, the use of objects should not be of concern unless the person is doing something negative or aggressive with the object—e.g., kicking it, punching it, or talking about injuring it.

Emotional Tenor of the Self-Talk Scene

An important dimension of a self-talk session is its emotional tenor. Self-talk can mirror the full range of human emotions. A self-talk session may reflect a calm and serene state of mind or include such positive feelings and emotions as

exhilaration, happiness, and triumph. But a session may also express such negative feelings as anger, rage, sadness, self-criticism, anxiety, and frustration.

Families and other caregivers often report that there may be a few sessions that include a mix of emotions such as sad and happy feelings, but in general, most self-talk sessions reflect only one primary type of emotion, either positive or negative. For example, some teens or adults replay a scene that was frustrating to them in the course of their day, such as someone being rude to them or witnessing someone getting angry. This in turn may trigger a memory of a past experience at the hands of others in other situations. Other replay scenes may include something positive such as praise from a well-liked supervisor. This too can set off similar memories of past positive comments from important others. Many families try to tune into this self-talk in order to get a reading on how the person's day went.

See "When Self-Talk Turns Negative" later in the chapter for more information on self-talk that may reveal a problem.

Purpose and Function of Self-Talk

In most cases the purpose is to enable the person with Down syndrome to learn and adapt to the world (as discussed immediately below). In other cases, a self-talk scene may be a way to help the person understand and attempt to cope with a minor or major stress or trauma. We discuss how to recognize and respond to signs of this function in the later part of this section.

Directed Action: Learning a New or Challenging Task

For people with Down syndrome, just as for typical children, one of the more important functions of self-talk is to walk oneself through a new or challenging task. This is known as *directed self-talk*. For example, this may occur when someone in trying to learn something new, such as knitting, making a new recipe, or to working out a math problem. Such directed self-talk is a normal part of childhood development as an aid to the acquisition of new tasks.

Review of the Day

Self-talk often involves a review of the day's events. The importance of this function cannot be underestimated for people with Down syndrome. Because of

intellectual and expressive language limitations, teens and adults with Down syndrome may have some difficulty understanding events and processes that occur in the course of their day. For instance, other people may move and talk too fast for people with Down syndrome to adequately understand them. Fortunately, many have the ability to visually record their day in memory. When they are home, their rooms serve as private labs and sanctuaries of learning. They can slowly take apart and replay each step and event until they comprehend it to their satisfaction.

Problem Solving and Anxiety Management

Some people may be able to use self-talk to solve problems. For example, they may think through different ways to help a parent or friend who is sad or to figure out different ways to reorganize their room or favorite tapes and CD's.

Self-talk may also help in the management of situations that might be challenging, frightening, or potentially anxiety provoking. For example, a self-talk session can allow people to walk through or prepare for a new job or new job task or even to come up with new tasks and activities to entertain themselves during free time. Similarly, someone may use self-talk to prepare for an upcoming social event such as a party or a big dance or to decide how to handle difficult situations such as a peer who frequently expresses negative feelings at school or work. One of the best ways for parents, teachers, or other helpers to take advantage of this skill is to use it as a part of a role-play session. This allows the person to see and work out different strategies for managing challenging situations and then take them into the environment where the problem occurs. See chapter 6 for information on conducting a role-play session.

Entertainment and More

Self-talk sessions may give us a glimpse of what we believe to be an extremely rich visual world of images and remembrances for people with Down syndrome going as far back as early childhood. As discussed in detail in chapter 6, many people with Down syndrome seem to have an exceptional photographic-like memory for events that are meaningful to them. As a result, their memory banks are filled with visual records of events lived in their daily lives. These scenes are often "taken in whole" and then replayed at will, sometimes in minute detail. The analogy we use for this is

that it is similar to replaying a video, but the screen is in the mind's eye and these video-like images can be enacted in a self-talk session.

Not surprisingly, people with these sorts of vivid memories may enjoy using self-talk for pure entertainment, replaying a favorite movie or a positive event in their life. Many families tell us that the person with Down syndrome enjoys replaying family movies (with an emphasis on special events such as vacations, weddings, and favorite holidays). Similarly, many people with Down syndrome can replay favorite scenes and events from memory. As with child's play sessions, these types of replay scenes will often be more than just entertainment. For example, people may replay positive memories to entertain but also as motivation—for instance, replaying an event in which they won medals in Special Olympics or even a movie with an uplifting theme. Other times, a person's self-talk may include scenes from favorite movies, but he may also insert himself into roles as if to try them out. For example, this may include being a firefighter or superhero who saves others from catastrophe.

Tools for Learning and Understanding the World

Self-talk sessions can be beneficial even when there is no anxiety or significant challenge associated with the experience. As mentioned previously, many families report self-talk scenes related to events at work, at school, or during religious services or social or recreation activities. For example, many people with Down syndrome act out the role of the teacher or boss and instruct or guide the other students or employees to do tasks. In other words, these individuals use self-talk and imaginative play to role-play scenes and situations that can be both entertaining and incredibly helpful and instructive.

As a Means to Escape

Sometimes the problem with self-talk is not what is replayed but when and where it is played. For example, if the person replays a favorite movie or scene from his life in his head or under his breath during school or work, it may interfere with the person's ability to do school or work activities. In most instances, self-talk at school or work is related to a lack of stimulating activities. Often a boring and understimulating environment simply cannot compete with the rich imaginative world of images that many people with Down syndrome seem to possess. The cure for this problem is to ensure the person has stimulating activities to engage in.

As a Break from Tensions and Pressures of the World

Like everyone, people with Down syndrome can find the privacy of their own room to be a wonderful and needed respite from the stresses of the outside world for people. Still, families often have questions about how much time their son or daughter should spend alone in their rooms, whether just relaxing or enacting scenes on their private stage. To address this issue, let's look to the research on typical children and teens.

Typical children make use of play to both process the world and to get away from it, which is also clearly the case for some teens and adults with Down syndrome. There is also ample evidence that typical teens need a great deal of privacy and alone time. The same goes for teens and adults with Down syndrome. They have to inhabit a world where others tend to talk and move way too fast for them. They need time in their rooms to reenact experiences and make sense out of them. Just as importantly, they need time to recover and decompress from the unique demands on them that result from adaptive and expressive limitations.

As discussed in chapter 7, people with Down syndrome can also be easily fatigued by trying to communicate verbally with others, so they need breaks when there are no demands for talking or interacting with others. Similarly, some are being told by others what to do during their free time and have relatively little control over their lives, especially compared to other "adults" in the world. Having a place to go where there are no demands or expectations is not just a good thing but an absolute necessity for people to maintain a sense of integrity and peace of mind.

So yes, alone time is necessary, but can there too be much alone time? The answer is that all of us need a balance between time spent alone and with others. As a rule of thumb, if there is a significant change in the amount of alone time and this change interferes with normal or essential activities of life, this could be a red flag that there is a problem. We all have different needs, but withdrawing into a world without family or friends, without social or recreational activities, is generally not a healthy behavior. If there is not enough contact with others (family and socially), then something may be wrong. It's best to try to set up a schedule that includes nightly dinners with family, friends, or people who live with the teen or adult and activities outside with peers. If the person with Down syndrome balks at these interactions, this may mean there is a problem. If the problem persists it may be helpful to consult a

mental health professional. See chapters 17 and 18 (on depression and anxiety) for more on this.

When Self-Talk Is Negative: Cause for Concern?

Self-talk scenes usually mirror real life—again, because the self-talk is a lab to process the outside world. As a result, expressions of anger and negative feelings may periodically be directed at even good and loving family and friends in the person's self-talk. This is a reflection of real life and should not raise concern, as long as the majority of self-talk scenes involve positive and constructive interactions with others.

There is more call for concern when the intensity and duration of negative feelings expressed is unusual or excessive. For example, self-talk may include derogatory comments such as "You are stupid [or bad]" or include swear words (especially those that are not commonly used by the person). It may also include self-critical comments such as "I'm stupid," etc. These types of remarks should raise red flags because they may actually be the expression of a real-life encounter with people who are abusive or harmful to the person in some way. This sort of self-talk should be looked at carefully to see if such a situation exists in real life. The self-talk may turn out to be the expression of a scene from a movie or TV show, but it is better to be safe than sorry.

How Self-Talk Can Express What May Be Too Difficult to Verbalize

As discussed in chapter 7, difficulty understanding and communicating with others, especially about complex or difficult issues, is a challenge for many people with Down syndrome. They may also have trouble communicating anything that is traumatic because they may reexperience the trauma by retelling it (see chapter 15). Even when traumatized, however, they may be able to express some key components of the event in question through self-talk. For example, as discussed above, they may enact critical statements expressed by people who are abusive to them. Family members are often able to tell from the tone and nature of self-talk whether it has been a good or not-so-good day, and they can pick up clues about specific problems from reenacted scenes. In fact, if a teen or adult uses self-talk, evidence of any mistreatment will usually surface in some form or fashion through self-talk. Like typical children, the person with Down syndrome may be compelled to replay negative experiences as a conscious or unconscious means to try to cope or manage

them. Below are a few key issues parents and professionals need to understand to interpret whether a self-talk scene truly is a cause for concern.

Under Conscious Control?

It is important to understand that the content of a self-talk scene is not always under conscious control, or fully understood, by the person with Down syndrome. As discussed above, people with Down syndrome sometimes replay certain memories at will, such as when walking themselves through a new task or replaying a movie to entertain themselves. There are other memories, however, that they do not seem to control. These may indicate something painful, harmful, frustrating, or anxiety provoking in their lives. What's interesting is that people with Down syndrome may engage in self-talk about situations even if they don't understand or cannot make sense out of what has happened. For example, self-talk may occur after someone is exposed to verbal and physical aggression for the first time. New or difficult-to-understand events are precisely the type of events that will show up in the person's private self-talk theater as he goes over and over the event to try to process and understand.

When a Good Thing Becomes an Impediment

It bears repeating here that people with Down syndrome often have excellent visual memories (chapter 6). They may be able to replay visual memories as if an event is happening all over again with all the original feelings and emotions. They may experience a trauma and then replay it, particularly when there is a reminder or trigger. This is why people with Down syndrome seem to be more susceptible to phobias (chapter 18). For example, once they experience a bad storm, they may become afraid even when the clouds just begin to darken.

To complicate things, people with Down syndrome can also replay positive memories such as of a favorite TV show or family event. They may use these positive memories to block traumatic memories. Unfortunately, they may get so caught up in trying to protect themselves (from traumatic memories) with positive self-talk scenes that they have difficulty focusing on the world outside and around them. See the example below of how this may happen.

Marie, age twenty-six, had recently shown a dramatic change in mood and behavior. Although she had previously been very social, articulate, and capable, she had withdrawn into herself and into a world consisting of a cast of characters from a favorite TV show. Despite being quite present and responsive to others previously, she talked and interacted with this cast of characters, and it was very difficult to bring out of her inner world. When her family tried to engage her in conversation, she would struggle mightily to leave her imaginary world. Due to her incessant focus on her imaginary characters, she also had trouble doing routine daily living tasks that she had previously done automatically.

Her family noted an event at her job that preceded this change. She had worked in a fast-food restaurant doing a variety of tasks. She seemed to love the job and was well regarded by her supervisors, the other employees, and the customers in the store. One day, however, she was found crying on the floor in the women's bathroom. She could not say what had happened to her. After this she refused to go to the restaurant and became noticeably afraid and agitated when they even went by the restaurant. This often led to agitated self-talk for the remainder of the day and even into the night, causing sleep disturbance.

In chapter 17, we discuss at length how people with Down syndrome react to stress and their difficulties dealing with negative emotions. In short, when a person is having trouble handling negative emotions, this can show up in the person's self-talk. In this case, when Marie was not discussing characters from her favorite show, her family began to hear her say "go away" over and over in her agitated self-talk.

When seen for a medical evaluation, she showed no evidence of physical or sexual trauma. What we believe happened was that someone said things to her in that bathroom that hurt her and were quite traumatic for her. This was enough to set off a protective mechanism of a replay of her favorite TV show to keep her memory of the events in the bathroom at bay.

We discuss treatment strategies for these types of traumas in detail in chapter 17. Suffice it to say for now that we recommended to her family that she not return to the restaurant either to visit or to work because of the high risk of a replay of the trauma. At first, they were reluctant to follow our recommendation because they had

fought so hard to get her into this job. In time, though, they recognized her negative reaction to being exposed to the restaurant. The minute they told her she no longer had to return to the restaurant, she began to heal and over time returned to her previous level of functioning.

Summary

To return to Jason, whom we introduced at the beginning of the chapter, we reassured both Jason and his mother of the normalcy of his self-talk. We expressed concern that suppressing his self-talk would be similar to telling someone else not to think about or contemplate the events of the day. Not allowing him to think through his day could have negative consequences. In addition, making him feel bad about self-talk could affect self-esteem. We also discussed Jason's awareness of the social skills component of self-talk: do it only in a private space. In addition, we encouraged Jason and his mother to return for further discussion if there was a change in quality or quantity of self-talk in the future. In general, this is the approach we recommend taking when a teen or adult with Down syndrome engages in mainly positive or neutral self-talk in the privacy of his own room. Only when self-talk turns negative or prevents the person from engaging in real-life activities, do we suggest intervening, as explained above.

Return to Table of Contents

Chapter 10

The Groove and Flexibility

Don would wake each workday at the same time and invariably follow the same routine. First, he would have his toast and juice, then shave, shower, and dress in the same meticulous way. He was always clean and well groomed for work, and his boss could count on him to be punctual and reliable. After work Don would have a snack, do his chores (take out the garbage, set the table), and make sure everything in his room was put away. Each Tuesday he would wash his clothes; Wednesdays he would pick up and vacuum the house. After dinner Don would relax in his room with his favorite movie or music while writing in his notebook or doing word search puzzles. Saturdays he would get up at the same time to eat breakfast; shave, shower, and dress; then head off to bowling and later to social club. Don's family was accustomed to his regularity; he was very reliable with his routines.

During the course of interviewing thousands of individuals with Down syndrome and their families, we discovered that an unusually high number of people with Down syndrome, like Don, need sameness, repetition, and order in their lives. We call this tendency "The Groove" because people's behavior tends to follow fairly well-worn paths or grooves. We have found grooves to be so common among people with Down syndrome that the absence of groove-like tendencies is notable for its rarity. In this chapter, we will describe different types of grooves and their advantages and disadvantages as well as ways to identify and resolve problems arising from "stuck grooves."

What Is a Groove?

A simple definition of groove is a set pattern or routine in one's actions or thoughts. We all need grooves in our daily lives, or absolutely nothing would get done. For example, if every morning we had to rethink when or how to take a shower, brush our teeth, tie our shoes, and make toast, we would never leave home in the morning. Consider all the other automatic, routine tasks we do every day at home, at work,

and in the community and it is easy to see that a world without grooves would grind to a halt.

People with Down syndrome are particularly good at developing and following grooves in their daily lives. Many follow grooves with a degree of precision that would impress a fussy accountant. Examples of grooves we have seen include the following:

- Having a set order and timing to daily routines, to include set morning, evening, and work routines, as well as routines for activities that are relaxing. For example, many people with Down syndrome draw, write, or copy words or letters during their time at home.
- Being quite meticulous in the care of their appearance, grooming, rooms, and possessions. People with Down syndrome often have set places for furniture and personal items in their rooms or living spaces and will return items that are moved or disturbed by others to the original location in short order.
- Developing grooves around less frequent activities, such as packing clothes, ordering in restaurants, or celebrating birthdays, holidays, etc.
- Having grooves that center on personal preferences for such things as music, sports teams, social and recreation activities, or celebrities, as well as for more personal issues such as a favorite relative or a love interest. These preferences help to define who the person is by what she likes to do and who she likes to do it with.

Advantages of Grooves

There are numerous advantages to these grooves. Grooves give an important sense of order and structure to people's daily lives. They are also of great benefit in increasing independence. Once an activity is learned and becomes a part of a daily routine, these tasks will be completed faithfully. Independence and performance may also be enhanced in the work environment. Employers are often impressed with how reliable workers with Down syndrome are in completing routine work tasks and in following time schedules.

Using Grooves to Relax

Groove activities may offer a refuge from the stresses and strains of daily life in the home or on the job. Typically, relaxing grooves involve repeating a specific

enjoyable activity in a quiet or private space such as a bedroom or bathroom, sometimes as part of the daily routine. Some of the most common activities people with Down syndrome enjoy repeating include reading, writing or drawing, listening to music, watching TV or videos, looking at family pictures, cleaning or organizing a room, or doing such crafts and hobbies as needlework or arranging collected items. In the bathroom, relaxing activities can include cleaning or grooming tasks, as well as just sitting and relaxing.

On the job, repeating a relaxing activity may give adults with Down syndrome a brief but valuable respite from interacting with others, from the noise and hassle of the workplace, or from the tedium of work. At work, the chosen space is quite often the bathroom because this is frequently the only place where there is some quiet or privacy. As at home, relaxing grooves may include doing some grooming tasks or just sitting and relaxing in one of the stalls. For instance, Debra, a thirty-two-year-old office assistant, relaxes during breaks by listening to music on her headphones or by doing "word searches" at her desk.

Grooves Related to Appearance and Possessions

Also of great benefit to teens and adults with Down syndrome are grooves involving the meticulous care of their own appearance, room, and personal items. Careful grooming and dressing convey a sense of pride, self-respect, and dignity to others and may increase the individual's own feelings of pride and self-respect. This may be especially important for people with Down syndrome, who have distinctive physical features that clearly mark them as different. These differences make them susceptible to discrimination, which may occur with any minority group. The self-pride that comes from attention to dress and grooming may go a long way to reduce the stigmatizing effect of being different.

Ordering Grooves

Ordering grooves are also very important to many people with Down syndrome. Ordering involves being neat and tidy with one's room, furniture, clothing, and other personal items (videos, pictures, books, etc.). Adults with Down syndrome are often very careful with folding and putting away clothing in drawers and on hangers in their closets. When ordering and arranging, they often feel a need to close doors and cabinets others have opened and turn off lights others have turned on. If taken to an

extreme, these ordering tendencies can be difficult for families to live with, as detailed in the section on "Disadvantages" below.

Grooves Related to Personal Preferences

Importantly, grooves can be a powerful means of expression and communication. This is especially true for people with Down syndrome who have a limited ability to express themselves verbally. Each groove is a clear and unambiguous statement of a personal choice or preference. For example, daily grooves and routines express how someone chooses to organize and manage such things as grooming, appearance, and personal items; participation in social, recreational, and work activities; and preferences in music, hobbies, and artistic endeavors. Each person's choices will, in turn, help to shape and define his own unique style and personality.

For some, grooves may even take on a life-saving role:

> *Cassie, twenty-eight, has Down syndrome as well as a genetic disease that causes progressive and irreversible muscle deterioration that will eventually lead to an untimely death. This horrible disease has already taken the life of all but one sibling. Cassie is well aware of her condition but maintains a strong positive attitude about life and about the people in her life. In fact, when we first met Cassie, one of the most difficult aspects of her disease was not the pain and discomfort it caused but the fact that she frequently missed seeing her friends at work or at social activities because she was too tired or ill to go out.*
>
> *We discovered that the primary reason Cassie is able to maintain her positive attitude is that she has developed a number of routines that are extremely relaxing and allow her to connect to others. Like many grooves, these routines often occur at a set time and in a set order. For example, Cassie often starts with a favorite activity: making one of her endless lists of things on her computer. She frequently follows this with a period of thoughtful letter writing to friends or extended family members. Finally, she carefully and meticulously writes personal thoughts in her diary.*
>
> *Despite Cassie's illness and the losses in her life, she has developed routines that serve at least three important purposes. First, she is able to*

avoid despair because she is simply too busy for self-pity. Second, her letters allow her to connect with family and friends, even if not in person. Thanks to her exceptional visual memory, she feels almost as if she is conversing with them while writing. She is even able to write to her deceased family members "in heaven," which gives her a great deal of comfort. Third, through her letters and diary, she can express both her positive feelings and her fears and concerns about her illness and her great sense of loss for her family. She reliably repeats her grooves every day, ensuring that she continues to get the benefits of these activities.

Disadvantages of Grooves

Although there are many benefits and advantages to grooves, there may also be problems and disadvantages. Some of the problems are not serious or need not be serious if handled appropriately by others. For example, an adult with Down syndrome may be interested in a particular issue, such as a favorite sports team, and bring it up repeatedly with family and friends. This may be a minor irritant to his friends is not necessarily a problem that interferes with important spheres of the adult's life.

There are also grooves that may be useful if done at the appropriate time or place but a problem if done at the wrong time or place. For example, a groove related to cleaning the bathroom may be greatly appreciated by family members unless it is done in the morning when everyone needs to get ready for work. A better plan would be to schedule this task into an afternoon routine. Similarly, a restaurant manager may be pleased with how clean an employee with Down syndrome keeps the bathrooms unless patrons have to wait for long periods while he does a meticulous job. A better plan may be for the employee to do this job when there are fewer patrons or before the restaurant opens.

It is not uncommon for ordering grooves to become problematic to a greater or lesser degree. For example, some people are described as "having their own sense of order." They arrange things in their room "just so" but not necessarily in a way that others would consider tidy or even practical. Books, clothing, videos, paper, and so forth may be arranged in piles in different spots on the floor, which may be inconvenient and make it difficult to walk through the room or clean it.

Some adults with Down syndrome have a habit of folding and putting away even clothing that has been worn. Some spend an excessive amount of time ordering or

rearranging clothing or other belongings, which may delay an outing or interfere with other activities. These adults may "over-arrange" everything, including clothing, furniture, magazines, and unusual items such as cut-up paper or bottle caps. Saving unusual items, as well more common things such as pictures, magazines, memorabilia, pens, and CDs is another pretty common groove. Many people with DS take saved or special items "on the road," carefully packing them in bags or backpacks and bringing them everywhere they go. The behavior becomes problematic and may be considered hoarding if the person resists giving up items, backpacks become excessively heavy, or hoarded items include garbage or refuse items. (See hoarding in chapter 19.)

Ordering can also be problematic if it involves inflexible insistence on always doing things the same way, such as sitting in the same fixed spot for meals, using the same cup, having food arranged "just so" on one's plate, etc.

The pace of completing routines is also an area of inflexibility for some people with Down syndrome. For example, attempts to move people along at a faster pace may only result in them slowing down further. Sometimes when adults with Down syndrome are pushed too far, they respond by shutting down or having a meltdown. They may even start the sequence of a routine all over again. We often hear of this happening during morning routines when there is a limited amount of time to get out the door. This may also happen when there are unexpected last-minute changes to the daily schedule. Additionally, shutting down may occur in response to a more significant change in an adult's life, as this story illustrates:

> *Susan, age thirty-nine, often refused to go out for recreational activities with the five other residents in her group home. This refusal had created growing resentment among the other residents, who enjoyed regular outings. It turned out that Susan had recently moved from a residence with fifteen older women who had a more sedentary lifestyle and far fewer outings.*
>
> *Staff became increasing more forceful with Susan because they believed her reason for staying home—so she wouldn't miss her nightly seven o'clock bath—was an absurd excuse. Predictably, she met their attempts to push her with even more resistance. At first, she slowed her pace when getting ready, which then delayed the outing. With increasing pressure, she not only slowed her pace to a crawl but also became more*

meticulous about her dressing and grooming and would start the routine all over again if not "just so."

A meeting between Susan's healthcare team and residential staff parties resulted in a recommendation that Susan move to a home with older residents, which was believed to be better suited to her needs. Before this plan was carried out, a second option became available. It was proposed that residents from several nearby group homes could go on joint outings, freeing up one staff person who could stay home with Susan. This latter plan was adopted as the best option for Susan. Interestingly, as Susan became more comfortable with her group home, she began to go out more often on planned outings. This significantly improved her relationship with other residents and also helped her with weight management.

In addition to hearing about routines in our clinical work, we have developed a healthy respect for the routines of the interns and employees with Down syndrome who have worked with us. Generally, these individuals are able to do assigned office tasks reliably. However, sometimes routines create minor problems. For example, one man had a set lunchtime at 12:00 and could not adjust his work routine to eat lunch at 12:30 with others in the office. He was allowed to have his own lunch schedule, but this meant missing out on a beneficial social period with other employees. Fortunately, after several weeks of gentle encouragement, he agreed to lunch with the other employees. In some cases, the wisest solution is to stay out of people's way when they do their routines. For example, it is best not to interrupt someone with Down syndrome who is doing a copying job in order to quickly make "just one copy." This is just too disruptive.

More Serious Problems

Sometimes a groove may become a more serious problem, and occasionally it may even meet the criteria of an obsessive-compulsive disorder (OCD). The groove may become OCD when it includes repetitive thoughts (obsessions) or repetitive behaviors (compulsions) that interfere significantly in normal and essential activities of life. For example, a groove is significantly problematic if morning or evening routines become so elaborate that someone consistently misses work or social

activities or feels compelled to do a particular activity even when they would prefer not to. Obsessive-compulsive disorder is addressed in chapter 19.

Risk of Maladaptive Grooves

People with Down syndrome are more susceptible to maladaptive grooves than other people are. After depression, maladaptive grooves are the second-most common mental health problem that we have diagnosed. On the positive side, maladaptive grooves, especially in the form of a more severe OCD, are not inevitable in adults with Down syndrome. Grooves clearly seem to be built into the chemistry of most people with Down syndrome (as described below), but we have found that these grooves have different degrees of strength.

One way to look at grooves is to compare them to the concept of temperament (Carey & McDevitt, 1995). Temperament is an accepted psychological term for our own innate personality traits or characteristics, which govern our moods and emotional nature. Any parent with more than one child will testify to the differences in temperament that each child displays from birth. This is not to say that families or outside forces do not have a strong influence on temperament—just that a strong biological component plays a major role in any aspect of one's emotions and moods. Grooves also appear to have an inherent biological basis, but, as with temperament, there appears to be a wide variation in the strength of this tendency. In other words, some people have an inherent tendency to be more intense or rigid in their grooves than others. This may affect how susceptible people are to the development of maladaptive grooves. In addition, in our experience, people with better adaptive skills overall are often more flexible and less susceptible to developing severe grooves or OCD.

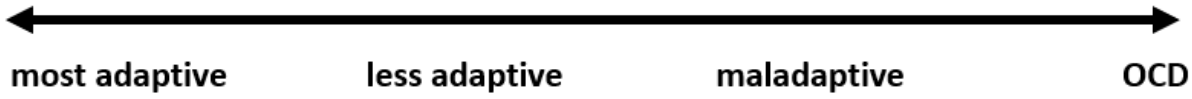
The Biochemical Basis of Grooves

To understand the tendency toward maladaptive grooves in some people with Down syndrome, you need some familiarity with the underlying chemical process in the brain. Researchers have known for some time that human behavior is the result of nerve activity in the brain. The brain's system of nerve pathways functions much like an electrical system, but with gaps at junction points. These junction points are bridged by chemical substances (neurotransmitters) that allow the nerve system to work properly.

Researchers have used sophisticated brain scans to locate specific nerve pathways and regions of the brain that are associated with particular types of human behavior (Saxena et al., 1998; Schwartz, Stoessel, Baxter, et al., 1996; Breiter, Rauch, Kwong, et al., 1996). For example, grooves (obsessions and compulsions) are associated with the nerve systems located in and between the frontal lobe and the basal ganglia of the brain. Researchers also found that deficiencies in the chemical substance serotonin (which bridges specific nerve synapses) may result in maladaptive behavior. In other words, specific nerve activity is associated with adaptive grooves and routines but may also result in less functional or maladaptive grooves when serotonin deficiencies exist at the nerve synapses. See chapter 16 for more information about brain chemicals.

The Groove Continuum

The fact that all grooves, whether adaptive or maladaptive, are connected to the same biochemical process may help to explain how maladaptive grooves develop in people with Down syndrome. In order to understand this, it may be helpful to conceptualize grooves on a continuum from most to least adaptive:



The left end of the continuum represents adaptive grooves, which we believe are essential to the day-to-day survival and functioning of all human beings. Grooves allow people to reliably complete daily self-care and job tasks. We could take this one step further and state that grooves are the basis of our own survival and serve as a foundation and structure of civilized society.

Closer to the center of the continuum, a groove may become less adaptive and less functional. This occurs when people adhere too rigidly to the groove, or when the groove has no useful purpose. Families usually preface their descriptions of these types of grooves with “I don’t know why he/she does this, but . . .” For example, some people with Down syndrome repeat the same question over and over when they already know the answer. Other common examples include arranging furniture or personal items in a room “just so,” turning lights on and off, or closing doors repeatedly before leaving a room or house. Grooves may also include repetitious

thoughts or activities that have no real functional purpose even when part of the activity is functional. For example, eating is necessary, but there is no need to arrange one's chair "just so" or sit in the exact same spot every time to eat dinner. Also, there is no need to arrange food on plates so as to avoid contact between different food items.

Even in the case of people whose grooves are overwhelmingly adaptive, there are invariably a few areas where there are nonsensical or unexplainable repetitious thoughts or behaviors. The truth is, most of us have some nonsensical repetitious thoughts or behaviors such as counting to ourselves, checking the stove numerous times, cleaning meticulously, or arranging things "just so." What is going on here? We have to consider again that the chemical process resulting in grooves may fluctuate or sometimes result in chemical deficiencies for most of us, resulting in some "nonsensical" behaviors.

When do we say that a groove is pathological? Actually, no matter how illogical, odd, or nonsensical the repetitious behavior, it may not be maladaptive or meet the criteria for an obsessive-compulsive disorder unless it interferes with functioning in the key social, home, or work spheres of life. For example, arranging your chair or food "just so" at meals is odd but not a problem unless this activity keeps you from eating the meal in a timely fashion. Similarly, repeating questions or comments about a favorite star, sports team, or holiday may drive other people a little crazy, but it is not a real problem unless the preoccupation interferes with work or social activities.

Moving farther to the right on the continuum, there is a point at which the groove begins to interfere with life activities and becomes increasingly maladaptive. Once the repetitious thought or behavior interferes significantly in normal life activities, then the criteria of an obsessive-compulsive disorder are met. It is important to note that we have found a difference in how OCD is presented in the general population and in adolescents and adults with Down syndrome. In the general population, the hallmark of this disorder is odd or disturbing thoughts that intrude on people's thinking; the persistent thoughts are unwanted and intrusive. This in turn results in debilitating rituals and repetitious behaviors that people do to try to stave off or manage these disturbing thoughts. For example, illogical fears of dirt or infection can result in repeated washing rituals. What makes this a problem in need of treatment is not necessarily the odd thoughts but the resulting changes in behavior that interfere with the person's life.

In contrast, people with Down syndrome are far less likely to report disturbing thoughts associated with compulsions. This may be due in part to expressive language limitations that may make it difficult to conceptualize and communicate such thoughts, or it may be that most people with Down syndrome do not have these disturbing thoughts—at least not consciously. However, sometimes we have inferred that disturbing thoughts are present. For instance, we have treated a few people who spent an excessive amount of time showering or doing cleaning tasks, which seemed to indicate fears about contamination from dirt, germs, or other harmful substances. Irrational fears may also be inferred if an individual has a ritual that consistently results in avoidance of a specific activity such as a community outing or a ride in a car. Even in instances where such contamination or safety fears seem obvious, people with Down syndrome are rarely able to verbalize fears and anxieties, even if asked by a trusted confidant. Regardless, the presence or absence of a disturbing thought may not matter so much as the presence of a repetitious behavior that interferes with their lives.

The intensity of maladaptive grooves may be reduced if stress is reduced and if the groove can be diverted to more productive ends, such as completing self-care tasks or work activities. However, when grooves continue to interfere in essential life functions, the person may benefit from the use of medication to normalize the biochemical imbalance. This is discussed in more detail in chapter 19.

How Stress Affects Grooves

Up to this point, we have described the chemical process underlying grooves and discussed what constitutes a truly maladaptive groove versus just odd or nonsensical behavior. We now need to describe some of the other causes of maladaptive grooves. To this end, we need to consider one simple but important notion, and that is that people with Down syndrome have a strong propensity to repeat thoughts or behaviors once begun. This may include thoughts or behaviors that are not always appropriate or may even be a serious threat to their health and well-being, and thus maladaptive. For example, many adults with Down syndrome may acquire “bad habits” by first mimicking and then repeating others’ behavior, such as hand flapping, hitting, picking at skin, and even faking seizures. People with Down syndrome may also develop maladaptive routines when they make poor decisions about behavior and it becomes a habitual pattern. For example, they may “get in the

habit" of staying up too late to watch movies or TV while also consuming unhealthy foods or soft drinks.

This propensity for grooves may lead to the development of maladaptive grooves when someone experiences too much stress. This happens because grooves represent preexisting tendencies or pathways, which then become natural conduits for expressing stress. In other words, a groove is similar to any type of physical predisposition. For example, people may have predispositions to have headaches or stomach or bowel problems, which become activated during periods of stress. These areas seem to be these individuals' "weaknesses" and are the location of physical symptoms secondary to stress. Similarly, the tendency of people with Down syndrome to develop routines and grooves may result in grooves that become "stuck" and unproductive with stress.

The specific process for developing a problem groove may be explained if we consider again the association of grooves to the chemical process in an obsessive-compulsive disorder. Under stress, the chemical needed to bridge the gap between nerve endings may become deficient and thus more like the chemical deficiencies seen in OCD. As a result, the groove may become more rigid and maladaptive. For example, when under stress, someone who is ordinarily somewhat flexible may rigidly adhere to a routine such as taking a bath at 7:00 in the evening, even if it prevents her from doing a previously enjoyable activity such as going to the movies. Another common example is to become so preoccupied by a favorite star or a love interest (real or imagined) that it begins to interfere with home or work activities.

In short, people with Down syndrome often have a tendency for repeating thoughts and behaviors that may be extremely beneficial but may also be a formula for major problems. Ironically, maladaptive grooves may be very useful as a communication method. Just as adaptive grooves are a powerful means for expressing people's choices and preferences, so too a stuck groove may be an effective way to express the presence of health, sensory, social, or emotional stress. When someone has a stuck groove, it may communicate stress related to any area of the person's functioning: health problems, a sensory impairment, or an environmental stress at work or at home.

Interpreting "Stuck" Grooves

We have found that stuck grooves may convey a general warning that an individual with Down syndrome is experiencing stress. In addition, they may communicate a more specific message about a problem.

Indications of Pain or Physical Problems

Returning to the analogy of a headache, a headache may be a general warning of a health problem but may also point more specifically to a problem in the region of the head, such as a sinus infection. Similarly, a stuck groove, such as repeated trips to the bathroom, may sound a general warning of a problem but may also point to a specific health problem, such as a bladder or kidney infection. In some instances, people with Down syndrome who have had ritualistic-like actions involving touching or poking their face or ears have been found to have serious sinus or ear infections. Similar actions may also be an indication of a hearing or vision impairment.

We believe that physical problems result in an increased sensitivity to the affected area of the body, which then triggers an accompanying repetitious pattern or behavior. When an adult with Down syndrome has a stuck groove, it is always important for her to see a medical doctor to check for an underlying medical problem, particularly if the groove has some association to the body or a bodily function.

> *Oscar repeatedly pushed his hand up his own rectum. He was diagnosed with a painful, uncomfortable bladder problem, which did not allow him to urinate. His actions brought some relief by stimulating his bladder to urinate. Although unorthodox and alarming, his actions also successfully alerted his caregivers of a serious health problem and pointed to the specific area of the problem. In fact, staff quickly responded to Oscar's problem, especially since Oscar was not careful to wash his hands and was smearing feces on the walls in his group home and bathroom. Treatment of the bladder condition did eventually eliminate the problem, but it took several months after the physical problem had resolved for him to unlearn the groove.*

Sensory Issues and Grooves

Sensory sensitivities or difficulties may also lead to stuck grooves. These types of grooves often sound a general warning but may also point to a specific sensory problem. For example, touch or tactile sensitivity is probably one of the major reasons underlying the unusual preferences some adults with Down syndrome have related to their clothing. For example, many people will only wear soft, loose clothing such as sweatpants due to an apparent sensitivity to certain types of fabrics. Quite a few people also refuse to wear new jeans, and some will not wear any new clothing. Some people even try to wear the same clothing every day or the same "uniform" consisting of the same broken-in shirts and pants.

Family members often come up with creative solutions and adaptations to deal with these idiosyncrasies. For example, if an adult will only wear one outfit no matter what the occasion (wedding, meeting the president, etc.), there is the nightly washing routine while the person sleeps. Other families wash new clothing twenty times before it is worn or purchase pre-worn jeans or carefully selected secondhand clothing.

These issues often wax and wane based on the degree of stress an adult is experiencing, maturity, or other factors. As a result, people are at times more able to try different solutions to these problems. For example, an adult may be more willing to try something other than sweats, such as soft cotton shorts in warmer weather, or soft cotton pants, which may look better at more formal occasions. Also, many people who tend to wear one outfit, or only one uniform, periodically change to something different. Still, if clothing-related grooves begin to interfere in essential home or work activities, we have been able to help people become a little more flexible by using behavior strategies, and, if needed, medications (serotonin reuptake inhibitors). See chapter 19 for more on this.

Many people with Down syndrome also have food-related rituals that may turn into maladaptive grooves. One of the many possible causes is an aversion to certain textures and tastes in the mouth. People with this problem are often described as picky eaters because they often avoid certain foods. Sometimes people may also refuse certain foods based on past experience with the food prepared in a way that is aversive for them (crispy, soft, etc.) Over time and through trial and error, most families find enough foods and enough ways to prepare the foods so that they are acceptable to the person. A few adults with Down syndrome develop more extreme aversions to food and may refuse to eat. Fortunately, this problem is not common and

not usually long lasting and generally responds to medication and behavioral treatments. See more on treatment in chapter 21.

Another common sensory issue in adults with Down syndrome is a type of depth perception or related visual problem that makes it difficult for them to climb up stairs or to cross uneven surfaces. This problem often seems to worsen with age, although some people have this problem even as children. Usually, this is not an unmanageable problem. Most people continue to walk across uneven surfaces (even wet or snowy surfaces) or to go up and down stairs, just very slowly and carefully. However, many adults with Down syndrome develop grooves and rituals around this difficulty that may cause problems in certain situations. For example, people with Down syndrome often have difficulty negotiating stairs in an auditorium, movie theater, or sports stadium, particularly if the facility is dark or very crowded.

Many people try to deal with the problem by moving very slowly and methodically, which may not always be practical at large venue events. The problem may be circumvented by arriving early for the event and then waiting until most people have left before leaving. Still, some people with Down syndrome flatly refuse to go to these types of settings, probably because of a previous negative experience. Difficulties with stairs may become a more severe problem if it affects the person's ability to negotiate stairs that are present in unavoidable settings such as shopping centers, school, or worksites. More severe stair avoidance may involve an anxiety disorder as well as a more severe maladaptive groove (OCD). See the chapters on anxiety and OCD for more on these problems.

Sometimes stuck grooves take the form of repetitious hand or body movements, including mild self-injurious behavior such as picking at sores, chewing fingers, etc. The cause of these behaviors is varied and may include anxiety or even tics, which may be outside of the individual's conscious control. However, these types of repetitious behaviors may also be linked to sensory issues, including with touch, taste, vision, hearing, and smell, as well as with lesser-known sensations such as proprioception and the vestibular system (which has to do with movement and orientation of the body in space). Such problems may be best evaluated by an occupational therapist (OT) who specializes in sensory integration issues. These OTs have expertise in identifying and resolving problems that arise from a malfunction of the complex sensory system. Sensory issues are discussed in detail in chapter 12.

Repetitive Behavior: Groove, Stereotypic Behavior, or Movement Disorder?

Repetitious behaviors are often called stereotypies or "stimming" (self-stimulation), and they are frequently observed in people with developmental disabilities. The most common of these behaviors include rocking or hand flapping, but they may also include utterances, particular postures, or any ritualistic or repetitious movement of any part of the body. Sometimes stereotypic behaviors involve objects. For example, we have seen people with Down syndrome wave socks, paper items, flags, pencils or pens, sticks or pencils with a string tied to it, and pom poms.

Although stereotypic motor behaviors often occur in people with autism (including those with Down syndrome and autism), they are also quite common in people who just have Down syndrome. Sometimes these behaviors are described as "autistic features," but we have not found that their presence is an indication of autism. These are common behaviors for a wide variety of individuals with intellectual disabilities and actually for all of us.

Why do these motor behaviors occur? There appear to be a number of reasons. Stereotypies may express some positive or negative emotional state. For example, these behaviors may occur when something is experienced as aversive or anxiety provoking or when the person is excited, happy, or exhilarated. For some, the behavior may meet a sensory need (see below). On the other hand, these types of behaviors frequently occur during quiet times when there is no stress or demand on the person, such as when sitting quietly watching a movie or TV show. In other words, this seems to be a more general discharge of motor activity that is set off by different conditions or stimuli. One thing to note about repetitive behavior is that it is far less likely to occur when the person is engaged in mental or physical activity. The more engaging the activity is for the person, the less likely she is to show stereotypic behavior. We discuss stereotypic behavior in greater detail in chapter 24.

Indications of Environmental Stress

Sometimes a stuck groove indicates the presence of some kind of environmental stress. One of the most common messages expressed by a stuck groove is one of avoidance. Often, a stuck groove that is used to avoid something develops from a groove originally used to relax. For example, as described previously, many people with Down syndrome relax by repeating an enjoyable activity in a quiet or

private space. If tensions or conflicts in an environment increase, they may spend more and more time in their private space doing their own relaxing or self-absorbing activity. Some teens and adults with Down syndrome move more slowly and become even more meticulous in completing their morning routines as a way to avoid a problem at school or work. If they are slow enough, they may miss the bus and avoid the conflict altogether.

People may try to avoid many different situations or conflicts, including physical or verbal aggression or an overprotective or intrusive care provider. On the job, adults may try to avoid serious conflicts or tensions with others as well as the noise and tedium of work, perhaps by staying in a quiet place such as a bathroom stall.

Other Reasons for Stuck Grooves

Sometimes stuck grooves communicate that there is a relatively harmless or benign issue. For example, an individual may get stuck on grooming tasks upon entering the self-conscious teen stage of development. (Remember, this developmental stage often begins later for people with Down syndrome; see chapter 11.) For instance, teens or adults may brush or comb their hair excessively or put on and take off many different outfits in the morning in an attempt to find the most becoming one.

Odd or out-of-character behavior, such as repeatedly talking about or drawing scenes with sexual or violent content, may alert others to the possibility of sexual or physical abuse. For instance, after Gary started repeatedly drawing sexually explicit sketches, careful investigation led to the discovery that he was being sexually abused. After he received appropriate treatment and was removed from the abusive situation, his repetitive behavior diminished.

Encouraging the Development of Healthy Grooves

Parents, siblings, coworkers, friends, roommates, and support people can all have an important effect on an adult's grooves. Indeed, how well the person understands, accepts, and responds to grooves can determine whether her grooves become adaptive and useful or maladaptive and problematic. Families and others in the adult's environment can encourage the development of healthy grooves by

interpreting behavior related to grooves in positive ways (for instance, viewing it as something the person needs to do rather than as deliberate oppositional behavior);

1. setting and enforcing rules that do not interfere with healthy grooves;
2. providing the right level of supervision to prevent bad habits that can turn into maladaptive grooves; and
3. encouraging flexibility.

Interpreting Behavior

It is relatively easy to misinterpret someone's need to complete routines or grooves as oppositional behavior. For example, most people with Down syndrome will try to finish a routine before starting a newly assigned task. Unfortunately, if the person who has assigned the task believes the motivation for delaying the assigned task is to resist authority, then an escalating conflict may ensue. Further pressure by the parent, teacher, or supervisor may cause further entrenchment by the person with Down syndrome. Is this oppositional behavior, or is this a behavior that is not completely under the person's control, involving a type of biological imperative? If you believe it is willful and oppositional behavior, then you will probably continue to force the issue, which will no doubt lead to continued resistance from the teen or adult with Down syndrome.

What may confuse the issue is that most people feel a push for independence and a normal urge to rebel in any situation where they are told what to do. However, biology is probably a stronger force here, and biology (like Mother Nature) is not to be trifled with or ignored or you pay a price. An analogy can be found in the tumultuous behavior and moods of teenagers who are in the midst of hormonal changes. Parents who successfully handle their teenage sons and daughters have a healthy respect for the way hormonal changes affect mood and temperament. They learn to respond very carefully and patiently to the teenager's moods since they know that reacting too strongly only makes things worse. An analogy may also be seen in people who have problems with low blood sugar and become moody and unreasonable when the blood sugar is low. Family members often learn to encourage the person to eat before discussing any important issues. Similarly, pushing hard to force someone with Down syndrome to do something will often backfire, as the person becomes more mired in the "problem behavior."

In our experience it is always important to look for reasons behind a person's behavior, including annoying or seemingly nonsensical grooves, before concluding that the person is just being stubborn and oppositional. As discussed throughout this book, people with Down syndrome are not always capable of articulating or communicating issues or concerns verbally and therefore may need to express them behaviorally. Since grooves are a natural part of these individuals' lives, they may be a logical means of communicating stress. Whenever possible, identifying and reducing stress in people's lives may also reduce problematic or annoying grooves.

That said, there are situations in which people with Down are not just compulsive but also oppositional. We are usually able to determine whether this is the case by noting the number and intensity of situations where the behavior occurs. For example, if someone's intention is to be oppositional, he may act this way whenever he is asked to do things by authority figures. If he is just trying to complete a groove in a given situation, then this should not affect other tasks when she has adequate time to complete them. Another clue is how the person responds to attempts to solve the problem. If she is given more time but her negative attitude persists, then the purpose of her behavior may be to oppose the person in authority rather than to gain more time. This person may very well need more freedom and independence, but this is a different type of problem than merely needing extra time to accommodate a groove.

Enforcing Sensible Rules

Problems may also occur if adults in charge set rules that interfere with the completion of grooves or that show a lack of understanding or allowance for grooves, as in this example:

> *Lynne, age forty-two, was assessed for behavior challenges that included yelling at the staff and other residents in her group home, as well as occasionally striking the person who was sweeping the floor. The rules of her house included sharing the chores. On Monday, Lynne swept the floor; Tuesday, she cleared the table; Wednesday, she took out the trash, etc. Each day she did a different chore, as did the other residents. However, Lynne really liked to sweep, and she was very good at it. She became upset when she had to do a different chore. Upon further*

questioning, we discovered that the other women did not care whether they swept the floor. The staff person who accompanied Lynne asked if this was an example of obsessive-compulsive disorder. It is an example of OCD, but not in Lynne. The rules were too compulsive for the situation and were creating unnecessary conflict. We encouraged a change in house policy, and when Lynne was able to sweep daily, the floor sparkled and peace reigned.

We most often see these types of problems with rules in work or residential settings when the staff or administrators have little experience with people with Down syndrome. Problems may also occur in the person's home if his family does not fully understand groove-like issues:

The Baker family continually had difficulty with their teenage son, Greg, who was chronically late getting ready for school in the morning. With the best of intentions, Greg's parents had made the same rules for him and his two brothers because they wanted Greg to be like everyone else. In many areas, he was just like others, but he definitely did not move as fast in the morning. Like many people with Down syndrome, he was slow, precise, and methodical in bathing, grooming, and dressing. As a result, he looked very neat and handsome, but still he was late.

Eventually the conflict and tensions in the morning reached a boiling point. In the parents' words, they had begun to "encourage" Greg to move faster. In his words, they were rushing and babying him. The harder they tried, the more he resisted. Greg even began to refuse to go to school, which was very unusual for him. During a consultation with Greg's medical team, everyone agreed that what they were doing was not working and that a new strategy was in order. Greg's parents patiently listened to an explanation about grooves. They recognized that their son had many groove-like tendencies and that they were generally beneficial for him. They had not considered that this was behind his morning slowness. All agreed that Greg would be better off with more time to get ready. He took the initiative to set his own alarm clock, and from then on there were no further problems around this issue.

We have also encountered situations similar to Greg's in school or work settings. For example, staff at one worksite complained that employees with Down syndrome were chronically late getting back to work after lunch. As it turned out, the break was only half an hour, which did not allow enough time for the employees to eat and return to work, given their slower pace. In negotiating a solution to these kinds of problems, it is often possible to discuss the benefits of the groove. Many employers will readily admit that the person's reliability and attention to detail makes her an excellent employee. Once they understand that the adult's pace contributes to her precision with her work, they will often agree to whatever extra time is needed (usually just five to ten minutes).

Similarly, we have heard that many students with Down syndrome are late to their next class after physical education class because they need extra time to shower and dress. Frequently parents try to communicate the need for more time to no avail. School staff often dismiss the parents' request on the grounds that parents are biased or overprotective of their children. We, however, are usually viewed as less biased and more professional, and thus staff member are often more willing to listen to us, even if we repeat the same things the family already stated. We have found that the students with Down syndrome usually just need a few extra minutes. Once this is understood, the problems are solved fairly easily.

Sometimes it may not be possible or in the person's best interest for her to take as much time as she would really like. For instance, extended time bathing or showering may aggravate dry skin problems. We have had some success using timers in these instances. Whenever possible, however, we put the person with Down syndrome in charge of the timer.

It is important to balance the person's needs and abilities against the needs of the family, school, etc. If the individual is physically incapable of moving fast enough to follow the rules, then it unfair to expect her to do so. If, on the other hand, the person is taking a long time as a delaying tactic, it often indicates a more serious problem. For instance, the person with Down syndrome does not want to be there for whatever reason.

We believe that one reason people with Down syndrome have a reputation for being stubborn is due to misunderstandings about their need to complete groove-like activities. Understanding this tendency and modifying your approach to setting and enforcing rules based on this inclination can avoid many problems.

Providing the Right Level of Supervision

Adults in authority also play a role in allowing or preventing the development of "bad habits" that could become maladaptive grooves. For instance, previously we presented the story of several roommates with Down syndrome who got in the habit of staying up late to watch movies or TV. This obviously involved a poor decision on the part of the adults with Down syndrome, but there was also a lack of supervision appropriate to the developmental age and maturity of these adults.

As discussed in chapter 4, developmental age can be different from the individual's chronological age. For example, a man with Down syndrome who is thirty years old may have many strong daily living skills, but his ability to make judgments may lag far behind. If others assume that his problem-solving skills are in line with his cleaning, cooking, and grooming skills, he may not receive the supervision or support he needs in other areas. In group living situations, inadequate support may be due to funding and staffing restrictions that may be rationalized with misguided or disingenuous notions of treating people in an "age appropriate manner."

A lack of understanding or recognition of the power and persistence of grooves often compounds these situations. Caregivers may not understand that there is a natural tendency to repeat a behavior once started. Unfortunately, once someone gets a taste of late shows and midnight snacks, for example, she may soon develop a bad habit of staying up too late. Not surprisingly, this can create major problems with sleep deprivation, daytime fatigue and lethargy, truancy, tardiness, and unproductiveness at work, as well as an increased risk for depression, weight gain, and a long list of associated health problems.

Discouraging Maladaptive Grooves

Remember that people with Down syndrome tend to do what people do and not what they say. Many people with Down syndrome are visual learners (see chapter 6). They learn from observing those around them. If you do not want the person to develop bad habits, then limit exposure to people who have them. Teens and adults with Down syndrome who spend time with people who eat, sleep, and exercise right generally follow these same practices.

Provide "good enough" parenting and supervision. Described in more detail in chapter 8, this is the practice of allowing people to have as much freedom as they are capable of handling while still maintaining their health and well-being. Too much

supervision may be stifling, but too little may lead to the development of unhealthy habits. In addition, when guiding, we generally find directing the individual toward a healthy groove is more successful than directing him away from an unhealthy one.

Limit exposure to situations with a higher degree of risk. Many different situations can put the person with Down syndrome at risk of developing maladaptive grooves. Exposing him to these situations would be like putting several boxes of chocolate in front of a "chocoholic." Chances are good that the chocolate will be eaten in short order. Likewise, some people with Down syndrome may be easily addicted to watching TV or movies or playing video games. Not providing them with guidance when deciding how long to engage in these activities will likely result in them spending too much time watching TV or playing video games. Fortunately, grooves may work both ways. Once the person with Down syndrome has a schedule that includes a more reasonable amount of screen time, she will generally follow it.

Take time to talk about it. Our habits and patterns of behavior are not just based on what we want or desire. We can reason and respond to others' influence. People with Down syndrome may have some difficulty with abstract reasoning, but they are still very sensitive to others' feelings and opinions. Taking the time to talk to them about why they should do certain activities may be very helpful and respectful. Even if they do not completely appreciate the benefits of a reasonable diet or social and recreational activities, they will care that this matters to those who are close to them. It may also help to explain issues in more concrete terms. For example, try explaining that a reasonable diet and exercise may help them to fit into their pants, have more energy, feel better, etc.

Encouraging Flexibility

While we certainly recommend respecting a person's groove, we also acknowledge that too much groove can be a problem. We therefore recommend encouraging the individual to develop some flexibility. This is an ongoing process done on a day-to-day basis. It involves respecting the groove while at the same time gently encouraging and directing the person to see other options.

Dos and don'ts of encouraging flexibility with groove-like behavior:

- Pick a behavior that is possible to change. Asking someone to make a change that is too difficult will lead to demoralization and more rigidity.

- Pick a time to encourage flexibility when you have the time to be patient.
- Explain clearly and patiently what behaviors are flexible alternatives to the current behavior.
- Break the activity down into manageable steps to facilitate learning.
- Use visual cues: pictures, a calendar, a demonstration of an alternative behavior, or the like to make learning and comprehension easier.
- Don't try to change a groove when the person with Down syndrome is under extra stress.
- Don't be judgmental or critical. (Nothing encourages rigidity quite like saying something along these lines: "It drives me crazy when you ____.")
- Give the person enough advance notice to prepare for the change but not so long to encourage obsessing about the change.
- It may be helpful to deliberately teach the word *flexible* by pointing out and praising instances when the individual is being flexible (see example below).

William, age thirty-four, came home from work and found his mother and his aunt, who was visiting from Europe, talking at the kitchen table. His mother invited William to go to a movie with them that evening. William indicated that it was Tuesday night and that on Tuesday nights he always did one hour of exercise to his favorite exercise video. His mother suggested that he could change his usual schedule and go with them to the movie, and perhaps make up the session later in the week.

William went to his room and his mother overheard him talking to himself regarding this issue. He returned to the kitchen and said to his mother, "I want to talk about the F word." His mother braced for the embarrassment of this (never-before-had) conversation in front of his aunt. William went on, "I want to talk about flexibility. I will go to the movie tonight with you and Aunt Jenny." Years of gently encouraging William to look at alternatives to his grooves when appropriate ended in a successful and enjoyable evening.

Conclusion

We have reviewed many ways that grooves are displayed. Each of them can have a beneficial function. Unfortunately, we have also seen patients with each of these grooves that have shifted to the right on the continuum to the point where they become significant challenges. These behaviors may become maladaptive grooves or may even become problematic enough to meet the criteria for obsessive-compulsive disorder. If the person is simply unable to be more flexible after a great deal of gentle prodding from others and his grooves are causing lots of conflicts or problems, then it may be time to consider an assessment for OCD or a trial of medication. Assessment and treatment options are explained in more detail in chapter 19.

Although many adults with Down syndrome have difficulty being flexible with routines, these routines usually do not interfere significantly in their lives. Most people with Down syndrome are able to adapt to change, given some time and encouragement from others. Even in situations such as Susan's, above, when rigid routines create problems, these problems may be resolved if others help the person to develop new, more productive routines or find environments that are more accepting and accommodating of her routines.

Grooves are clearly a common characteristic of people with Down syndrome. Attempts to completely eliminate grooves are not only likely to fail but can be detrimental. Using grooves in a healthy way can often be very advantageous for teens and adults with Down syndrome. We recommend continued efforts to respect a person's grooves, while at the same time striking a balance between the groove and flexibility.

Return to Table of Contents

Chapter 11

Life-Span Issues: "Teenage Behavior," Isolation, Withdrawal, Retirement

"Jimmy has changed over the last several months" his mom explained. Jimmy was a seventeen-year-old young man with Down syndrome when he came to see us. His mom went on to explain that Jimmy was spending more time in his room by himself, playing more video games, talking to his friends on the phone, and was "less respectful" of his parents. After several other questions that didn't reveal concerning mental illness, we asked the question, "Does Jimmy have older siblings who previously were teenagers?" Jimmy's mom paused, tilted her head, briefly smiled, and just said, "Oh."

None of us are the exact same person throughout our lives; the same at fifty years of age as we are at twenty or forty or the same as a teenager as at age ten. Growth, development, and the effects of our life's experiences affect each of us over time. Change is a normal and healthy part of our lives as human beings. Behaviors change, attitudes change, and personality can change. The degree of change varies from person to person, but some change is inevitable. People with Down syndrome, too, change throughout their lifetimes. Also, just as for people without Down syndrome, there are particular times of life when change is typically most pronounced. This chapter focuses on several stages of life that are likely to result in changes in adults with Down syndrome that may be misinterpreted by others as behavior problems.

Adolescent Behavior

Adolescence can be a challenging time, both for the teen and for the people around him. It is a time to develop one's sense of self and one's independence. A major task of adolescence is defining oneself separately from one's parents. The process of finding oneself while at the same time "fitting into the crowd" is a difficult

balancing act. An adolescent often struggles with wanting to be independent while simultaneously wanting to hold on to the safety of his parents. Mood changes, isolation, experimental behavior, and intermittent assertiveness of personal choice often mark this struggle. For the people around the teen, particularly the parents, understanding and accepting the process are first steps toward surviving this transition. But for parents, acceptance does not always come easily, particularly in the midst of the intense conflicts that often envelop the parent and teen in this period.

For parents trying to survive this developmental stage, one of the most useful books on adolescents is by Dr. Anthony Wolf, a father and seasoned psychologist who has treated thousands of teenagers and their families in his clinical practice. The title of his book, *Get Out of My Life, But First Could You Drive Me & Cheryl to the Mall: A Parent's Guide to the New Teenager, Revised and Updated* is a testament to his understanding of these issues and his sense of humor. His message to parents is one of hope and surprising simplicity in a period that can be extremely confusing and trying. He says that teens often act the way they do because they are so close to their parents. The drive to establish their identity and independence requires distance from their parents. However, the dependency on parents and desire for assistance can conflict with the desire to be independent and act without parental involvement. Carl Pickhardt, author of *Surviving Your Child's Adolescence*, quotes an adolescent: "If you want me to be more independent, just give me more support." The potentially competing goals can be confusing for both parent and adolescent. Of course, teenagers' hormonal surges and body changes, which make them irritable, moody, and unpredictable, don't help.

Given the difficulty of the teenagers' tasks and the physical changes they endure, parents may be getting off easy if all they receive is three or four years of the silent treatment mixed in with intense periodic expressions of anger, hostility, and defiance. Adolescents may challenge the rules, but they crave them, nonetheless. Wolf assures us that with time, teens' hormonal surges level off and the drive to establish their own identity and independence may become more reasonable and the teen more responsible.

A parent's continued support and maintenance of rules helps to guide a teen in the appropriate tasks of this life stage. In the early stage of adolescence (onset of puberty) this involves teaching about basic but essential grooming and hygiene tasks (deodorants, hair combing, and menstruation management for females). In the later stage, this includes working on social, academic, and job-related tasks that are

necessary for the teen to successfully transition to adult life and responsibilities. A teen's pride, self-esteem, and identity are built by his increasing ability to "do for self" (what psychologists call competence), at each stage of this process. What helps with this process is the development of cognitive abilities that increase teenagers' capacity for reasoning and abstract thinking. This helps them to better manage their emotions and to see the need to be responsible for, and not just in charge of, their actions.

Similarities between Teens with and without Down Syndrome

Physical and Hormonal Changes

What happens with teenagers with Down syndrome and their families is analogous to the changes that occur in teens without Down syndrome. Most teens with Down syndrome undergo the same physical and hormonal changes of puberty around the same time as teens and preteens without Down syndrome. Although some older studies found that girls with Down syndrome begin menstruating earlier (Evans, 1988), more recent studies have found that the age of onset is similar for girls with and without Down syndrome. Due to the changes of puberty, many teens with Down syndrome experience bouts of moodiness and irritability like their counterparts without Down syndrome do. They, like other teenagers, may also

- dress and groom more carefully,
- take forever in the bathroom, combing their hair, etc.,
- overuse cologne, deodorants, and hair gels,
- have problems with pimples and acne,
- become more interested in members of the opposite sex (or same sex),
- and, perhaps, begin to masturbate.

Parents also note gender-specific changes. Boys try to shave (like their fathers, whether or not they have facial hair) and to wear deodorant for the first time. Girls (like their mothers) may try using makeup and have to adapt to their menstrual cycles, and for some, the associated discomfort and emotional effects of premenstrual syndrome. In other words, teens with Down syndrome appear to respond to puberty and other physical and emotional changes of early teenage development in a similar way as other teens do.

Conflicts with Parents

Complaints about behavior and emotional issues of adolescents with Down syndrome are often similar to complaints from parents of teens in general. At times during conflict, some adolescents, particularly younger teens, may display behavior and emotions that are more childlike or regressive in nature. For example, they may sometimes use tantrum-like behavior they haven't used since early childhood. Parents also report that whatever issues had occurred prior to the onset of puberty seem to intensify or worsen with these changes, at least temporarily. Like many teenagers, they are generally less patient and tolerant of minor irritations and inconveniences. Of course, they may also resist direction from parents or others in authority.

The effects of these teen-era changes may have more of an impact on some people than on others. For some, this process may be somewhat delayed but show up in full force at a later date. Many parents report little or no emotional upheaval from their teens and preteens with Down syndrome. However, the same may be said for teens in general, who experience these years at varying degrees of intensity, presenting varying degrees of difficulty for their parents.

Desire to Do Things Themselves

One area of similarity is a need for the teen "to do for self." The starting point for the tasks that teens with Down syndrome wants to do for themselves may be different because of delays in development. For example, they may want to do grooming or hygiene tasks without parental assistance, such as showering by themselves. Most teens without Down syndrome would have mastered such skills at an earlier age but may want "to do for self" in another important area, such as going off by themselves in the community.

Because of the wide range of skills and development in children with Down syndrome, some will try to do some tasks for themselves at ages similar to other teens. However, the majority of teens with Down syndrome will be at a different level of skill than teens without Down syndrome are. What is similar between teens with and without Down syndrome is that parents often complain that they want to do things for themselves even when they are really not ready for or capable of doing the tasks. Particularly for teens without Down syndrome, this often involves testing the limits of freedom, such as how far from home they are allowed to go. Teens often try

to go farther and farther from home, regardless of the possible risks (and the gray hairs they are giving their parents).

Similarly, parents of teens with Down syndrome complain that their children insist on doing things that they are not quite able to do. This may be aggravated by the fact that many teens with Down syndrome are visual learners and they see what other teens have attained. They may want a degree of independence just like everyone else their age, at least in terms of dressing and grooming. This may not always be reasonable or in their best interest. For example, some individuals wash or dry their bodies inadequately, don't get soap out of their hair, or don't brush their teeth appropriately.

Dealing with these problems may require that parents get very creative. Being too direct or "parental" may cause the teen to shut the parent out. But allowing the teen to do the task inadequately may leave him open to teasing and criticism from peers. There may also be a detrimental effect on their bodies, such as the possibility of gum disease from poor dental hygiene or the development of rashes and painful boils from inadequate bathing. A creative solution for dental care might be to buy electric toothbrushes that are fun and effective. (For example, a timer on the toothbrush may improve dental hygiene.) A time-tested strategy may be to ask others who are more acceptable to the teen to teach him certain key tasks. These others may include older siblings, cousins, grandparents, etc. The best teachers are often mature individuals whom the teen looks up to. Life skills classes in schools may also be wonderful places to learn.

Regardless of how many general education classes the teen with Down syndrome is included in, he may still benefit from a separate time with students who have Down syndrome or other intellectual disabilities. Although some parents are reluctant to have peers with disabilities congregate together in regular school settings, this is often essential to the teen's own sense of acceptance and self-esteem. (See chapter 8 for more on this.) Often, the other teens with disabilities are struggling with the same developmental tasks and issues. In these situations, both the teacher and the other peers can be instrumental in the learning process. The teen learns from observing others, but there is also no better way to learn than to help teach others. (See more on peer modeling in chapter 13.) What may also help in this process is the tendency of teens with Down syndrome to follow grooves and routines. Once they learn how to do the tasks correctly, they may continue to follow through with them very reliably (see chapter 10).

Differences between Teens with and without Down syndrome

Abstract Reasoning

As mentioned above, it is normal for teens to challenge the rules and for their parents to become concerned as the teens' challenges seem to get bolder and riskier the older they get. For example, younger teens may want to pick out their own clothes or hairstyle, but older teens may want to stay out later and later with friends. Parents of teens without Down syndrome are relieved to find that as their children mature, they often become more reasonable. This is due to an increase in cognitive skills, resulting in the development of abstract reasoning. With these increased reasoning skills, teens begin to understand the reasons parents are setting rules and do not just see rules as something to challenge. For example, they may see why it is helpful to get in early on a school night in order to be alert the next day or why it is important to avoid certain people.

In contrast, most teens with Down syndrome continue to be fairly concrete in their thinking. This may adversely affect their ability to understand and resolve issues in this life stage. Yet we have found that these teens often have other strengths and attributes that may allow them to compensate for this deficit. For example, many people with Down syndrome tend to be very aware and sensitive to others' feelings and emotions (see chapter 4). Contrast this with teens without Down syndrome who are often described by parents as self-absorbed, self-conscious, self-centered, etc. In fairness, this focus on self and others like themselves is to be expected in this stage, given hormonal surges and the need to define one's own identity. This is not to say that teens with Down syndrome are not also self-conscious or self-centered—often just less so, when compared to other teens.

Compulsive Tendencies

A second key difference in teens with and without Down syndrome is that that the former often have neat, organized rooms, with clothing and personal items placed "just so." Compare this to other teens who are sometimes described as "slobs" by their parents and their bedrooms as "disasters." Even when teens with Down syndrome do have chaotic-appearing rooms, there is some order to the chaos--often they have specific piles for specific things. This is consistent with our observations that obsessive-compulsive behaviors or grooves are more common in teens with Down syndrome, just as they are for adults with Down syndrome (see chapter 10).

The problem with grooves is that they may become more rigid and inflexible, particularly with the type of stress that teens with Down syndrome experience during this stage of physical, emotional, and social change. For example, at the start of every school year, and until her anxiety about her new classes and classmates abates, Beth becomes more rigid and compulsive at home. She insists on making her bed "just so" before she eats breakfast, even if she is in danger of missing the bus, refuses to eat her breakfast if her toast is cut "wrong" or her juice is in the "wrong" glass, and repacks her whole backpack if her mother tries to put her lunchbox in it in an attempt to hurry her out the door.

On the other hand, even with the stress of adolescence, we have found that grooves may be very productive for teens with Down syndrome. They may serve as an effective means for the teen to express his independence and autonomy. As discussed in chapter 10, grooves and routines are a clear and unambiguous statement of a personal choice or preferences in such key areas as clothing and appearance, social and recreational activities, and music, hobbies, and artistic endeavors. Each person's choices will, in turn, help to shape and define his own unique style and identity, which is of critical importance to the developing teen. Grooves may also provide a less antagonistic, clever means for expressing one's independence to parents. This is because grooves do not necessarily require the teen with Down syndrome to express anger or a rebellious attitude to maintain.

Regarding this tendency for grooves, we have seen an interesting trend in some teens with Down syndrome in recent years. These teens seem to have had more experience doing things for themselves and thus have more confidence and assertiveness when dealing with parents and authority figures. As a result, these teens are often more like teens without Down syndrome in expressing their feelings and independence. Again, this may be expressed without the type of impulsiveness and angry outbursts more characteristic of other teens (although it may be expressed with impulsiveness and anger). Interestingly, however, the demands this group makes on parents is often in the form of groove-like preferences and routines, which is often not the case for teens without Down syndrome, who may vacillate more about their likes and dislikes, adherence to schedules, etc. Parents of teens without Down syndrome are often confused and befuddled by the constant changes, while parents of teens with Down syndrome may be mildly irritated by the demands for sticking to set patterns and behaviors. Fortunately, most parents learn to appreciate their child's unique ways and means of "getting there" (even if this means they are slobs, neatniks, save-a-holics, etc.).

Delays in Teenage Behaviors

A third major difference between teens with and without Down syndrome is that the parents of the former may end up dealing with adolescence issues at two separate periods of time compared to only one (albeit possibly long) period for teens without Down syndrome. There may be certain advantages to having a second period of adolescence, but it may also create a great deal of confusion if not understood by parents and other caregivers.

Let us explain how this may happen. For teens with Down syndrome, the first period of adolescent issues may occur when they undergo the physical and hormonal changes of puberty, which often occur at a similar time as for those without Down syndrome. However, most teens with Down syndrome do not have the level of skill and maturity at that time needed to do the adolescent tasks that help them make the transition toward adulthood. Consequently, this transition may occur at a much later age, compared to teens without Down syndrome.

We know that the individual's drive for independence is not unique to adolescents. Children strive for independence at all stages of development. For example, a toddler's drive for independence may be as strong as an adolescent's. The difference between the teen and infant is that the infant struggles to become independent within the family, and the teen struggles to be independent from the family. What then defines the stage of adolescence is not the teenager's physical changes, or even his rebellious behavior, so much as his progress toward completing the tasks for this stage of development. Some people with Down syndrome may move toward independence while they are still in their teens. This often occurs in just one or several specific areas, such as wanting to get a job or to have more independence in their living situation. However, many people with Down syndrome experience the first wave of puberty and the accompanying changes in mood and behavior, but then are not necessarily ready to deal with the tasks of separating from the family until they are older (in their twenties, thirties, or even forties). Even then, they may continue to be dependent on parents in certain areas.

This is called an "out of sync" pattern of development, because for many people with Down syndrome, the maturity of the physical body is not in line (in sync) with the maturity of the mind or of adaptive skills. It's not that the person's physical and mental maturity don't ever get in sync; it's just that the process may be delayed or modified for years. The fact that this process is out of sync and may occur when

adolescent issues appear to many parents to be long gone may result in confusion and misinterpretation.

We have seen many people with Down syndrome in their twenties to early thirties and occasionally late teens whose parents were concerned about these changes in behavior. “He doesn’t participate in family activities like he used to”; “she spends more time in her room.” These and other comments by parents reflect a change in their son or daughter. These changes are a challenge for all families. They are neither unique to having a son or daughter with Down syndrome nor to be unexpected for a person with Down syndrome. However, there are a few issues that are more likely to limit a successful transition:

1. Families, support people, etc. may not realize or accept that the person is going through a normal developmental process (because it is delayed) and may misinterpret behavior.
2. Families and others may have trouble giving the adult with Down syndrome the proper amount of independence.
3. It may be difficult to tell the difference between normal adolescent behavior and behavior that calls for professional intervention.

The Present and Future of Down Syndrome

We have seen an interesting phenomenon develop over the many years we have been caring for adolescents with Down syndrome. Many teenagers with Down syndrome from recent generations are becoming more independent than those from past generations—which is a desired goal. Sometimes families, as one mother put it, “are not sure what we have done.” That is, sometimes when encouraged to develop independence, individuals will make choices that are not the ones parents, families, or teachers would desire for them. When one promotes independent thought and behavior in another, a piece of that is accepting that the individual will sometimes make different decisions than the parent, sibling, or teacher was hoping for. Allowing and supporting that decision to be made or action to be taken can be a challenging step toward independence (both for the adolescent and the parent).

The challenge for the parent of any adolescent, with or without Down syndrome is to balance other factors important to the parental response. Safety is a significant issue for any teen and is usually a larger consideration for a person with

Down syndrome. Does the individual have the cognitive capability to understand the implications of his decision?

Ability is another consideration. Is the desired activity realistic? This requires careful thought as people with Down syndrome are involved in more activities and demonstrate new skills that people with Down syndrome could not do or were not given the opportunity to do in the past. The easy answer of the past that "A person with Down syndrome can't do that" has to be carefully evaluated. In addition, parents have to be cautious about shattering dreams. Providing support while allowing the natural course of events to occur is one approach. For example, one of the authors of this book had great aspirations of being a major league baseball player. While supporting that aspiration (despite clear evidence early on that it was a dream unlikely to be fulfilled), his parents wisely encouraged other interests as well. As the natural course of events played out and demonstrated that the dream was not supported by sufficient talent, the pursuit of the dream waned and the other interests that had been fostered developed further.

Unfortunately, individuals with Down syndrome aren't always given the opportunity to develop their abilities. Providing appropriate resources (such as early intervention) can have a significant impact on achievement of goals by expanding abilities. When needed, supports at school, in the workplace, and elsewhere are helping many people with Down syndrome participate in activities previously thought to be impossible and to more fully participate in society in general.

As many people with Down syndrome are making more independent decisions, conflicts can develop for many reasons. Perhaps the person overestimates his abilities. Or perhaps there are inadequate supports. It could be that the family, school, or parents are afraid for the person's safety or they feel a likelihood of success is low and could lead to heartbreak, agitation, or depression.

These issues must all be addressed when considering "the present and future of Down syndrome." There are both opportunities and challenges. People with Down syndrome are expressing their opinions, seeking new activities, and participating in society in ways not seen to this degree in the past. How will people with and without Down syndrome respond to these changes?

Recognizing and Accepting Normal Developmental Changes

In the past, before the life expectancy of people with Down syndrome increased dramatically, parents heard, often at birth, that their child would not live

into adulthood. They were often also told that their child would not walk, speak, or read and would have other severe disabilities. Even in more recent times, some of our younger families have been given very pessimistic views of their child's eventual development. Not surprisingly, parents who were led to believe that their child would not survive into adulthood, much less develop skills necessary to participate in the world, gave little thought to going through stages of life. The professional prediction was of a static human being, one who would not develop over time.

Many families rejected the concept of a static individual when it came to developing skills. Against medical advice, they took their children home and helped them develop skills that they were told were not achievable. However, seeing the person with Down syndrome as a continuously developing person who experiences the changes associated with going through the stages of life may still have been a challenge for them. Even younger families who were given better and more optimistic information at the birth of their child often have a difficult time understanding the normalcy of the development of the person with Down syndrome: the pattern is similar but delayed.

Understanding that the changes are likely to occur is the first hurdle to supporting the person with Down syndrome through these changes. When a family asks about the changes that they are seeing, the first question we usually ask is, "Do you remember the teenage years of your other children?" This is usually followed by a look of insight, then a smile and a quizzical frown, as with Jimmy's mom at the beginning of the chapter. Parents understand the challenge of helping a teen develop a sense of self, to "fit in," and to develop independence. It may not always be easy to accept or deal with the process, but parents understand that the process must occur.

We have found that when an adult has limited verbal skills, parents may have even more difficulty understanding that the person's behavior is a part of her drive for independence. Sometimes the person's behavior may be her only reliable means for communicating that this is occurring. For example, a mother called us to report that her thirty-three-year-old son, Richard, refused to get out of bed. When we investigated, we found that this man had no contact outside the family and had little or no independence because his mother did everything for him. His refusal to do anything was his only remaining strategy, but it was very effective. When Richard refused to do anything, his behavior sent a message

- to his **mother**,
- who called his **sister** (the only relative who had stayed involved),

- who looked up our website and called us,
- who investigated the problem and made recommendations for him to "have a life."

Although there is obviously much more to this story, the key problem and solution are both here. Richard's mother did agree to let him venture out to a day program and to different programs and activities that did in fact give him a life. She took some convincing, however, because some students had teased Richard when he was in school fifteen years ago, and she was afraid for him to be in the community. Still, she heard his message, and she was eventually pleased when he was happy with his newfound independence. We have found similar solutions to other situations that were less dramatic but that involved a concern with a "change in behavior." This may be something out of character or even an increase in a preexisting behavior, but the message is clear: "I need more independence!"

When this is the cause and the family sees the solution, they are relieved and pleased by the progress the person has made to communicate and gain some control and independence in her life.

Providing the Right Level of Independence

A challenge for all parents is finding the balance between directing, supporting, and letting go. Parents may be scared to let their child find her way, because when she spreads her wings, she may make mistakes, stumble, and even get hurt. For children without Down syndrome, the expectation is that they will eventually be "independent." In contrast, most people with Down syndrome have a greater degree of lifelong dependence, although some are more independent than others. In addition, the independence they achieve will take longer to attain. This makes the already difficult task of "letting go" that much more difficult. For most families, there is never the same sense of "letting go" as there is for children without Down syndrome.

Letting go may also be more difficult for parents of a son or daughter with Down syndrome because the parents may be older than they were when they let go of their other children. Their child with Down syndrome is usually the last to leave the nest. Sometimes parents are at a point in life where they don't have the same energy to assist their son or daughter with Down syndrome through the process as they did their other children.

The “rules,” however, are similar:

1. Understand that the process toward independence will occur.
2. Accept the process.
3. Be supportive. Help your child to develop as much independence as she is capable of.
4. Allow her to grow toward greater personal decision-making. She can only develop the ability to make decisions by making decisions and experiencing the outcome.
5. Pick your battles. The person has to make decisions as part of the process. Some choices the person makes will absolutely not be acceptable (see #6) and cannot be allowed. However, learning to pick battles takes time and practice. A child needs to make many decisions and experience the outcome over time to learn and grow. A person with Down syndrome will probably need more time for this process. If you repeatedly intervene, it will slow and possibly even stop the process.
6. Keep the person safe. Obviously, allowing a young child to play in the street and get struck by a car is an unreasonable way to teach her that playing in the street is not safe. There are many choices that will similarly not be safe as the person with Down syndrome progresses through adolescence and young adulthood. However, many decisions are obviously not “safety threatening.” Offering choices that are all safe can help shape decision-making. Allowing the person with Down syndrome to walk home in the dark by herself may not be safe. However, other choices can be offered, including arranging a carpool with her friends, taking a cab or public transportation, or other options available in your community.
7. When it comes to decisions that are not dangerous but may subject the person to ridicule, offer encouragement and choices, and discuss your concerns. However, in the end, her choice may be part of the learning process. Peers (and others) can also teach (sometimes in a less kind fashion). Be prepared to support the teen or adult later without any “I told you so.” In addition, she may hear the ridicule and decide she still likes her choice, feel a stronger sense of her independence, and be proud of herself for “not following the crowd.”
8. Remember that people with Down syndrome often imitate appropriate behavior much better than they follow verbal guidance. For example, if you want an adolescent to answer questions politely and not ignore you, then you should

answer her questions politely, rather than grunting if you are busy or distracted.

9. Recognize that some behavior that starts during adolescence may never be "fixed." For instance, the adult may henceforth always prefer to do things with friends rather than parents. Or she may be more prone to argue for doing things her way. Encouraging independence sometimes includes accepting choices you might not have made. (But then, if she made every choice you would have made, she wouldn't be truly independent). For example:

Kevin, age twenty-three, was isolating himself in his room and interacting less with his parents. His older brother, Steve, had recently moved out of the family home but made a concerted effort to go out regularly with Kevin to play video games, shoot baskets, or other similar activities. Kevin also became involved in a mentor program and met regularly with the parent of a young child with Down syndrome for a social event. Kevin continued to have less verbal interaction with his parents and to spend more time alone in his room than he had as a child, but he was getting out and enjoying himself. As for many teens, better interaction was seen with people other than his parents.

We would describe Kevin's behavior as "normal." Even when his behavior improved, he still was different than he had been as a child. That is not a failure of the intervention. That is the expected outcome. People develop as they go through their lives, and there are particular times when change tends to be greater and more rapid. The "adolescent period" is clearly one of those times.

Comparisons with Peers

The desire to be like peers is often part of adolescence. This can be a particular challenge for some adolescents with Down syndrome if there are things that they cannot do. They see their peers driving, dating without chaperones, going to college, moving away from home, and getting married. Teens and adults with Down syndrome may feel left out, sad, or frustrated if they can't do the same things. While this struggle has always been present in families as siblings of people with Down syndrome have matured and done these things, it is now more frequently encountered

with regards to peers. When people with Down syndrome are included in local schools and social settings, they have more peers who do not have disabilities and who are participating in these activities. Although interacting with peers who don't have disabilities has many positive aspects, it can be painful to see these peers participate in activities the adult with Down syndrome cannot participate in (see more on this in chapter 8).

Recognizing the Need for Professional Help

During times when people are going through rapid changes, there is greater stress and a greater likelihood of developing a mental illness. Adolescence is clearly one of those times. Depression is the most common mental illness we see, although anxiety may also occur. Sometimes it can be hard to distinguish adolescent behavior from symptoms of depression, as in these situations:

- While a loss of interest in activities with parents is not unusual, loss of interest in all activities is concerning.
- Sleep patterns often change in adolescence. Needs for sleep increase and adolescents want to stay up later and sleep later in the morning. However, sleeping all the time or not sleeping at all are concerning.
- Mood swings are to be expected, but uncontrollable anger and aggression can be signs of a more significant problem. Periodic mood swings may also be a sign of premenstrual syndrome (PMS).

Depression is discussed below, and more information is available in chapter 17 and should be considered if the intensity or duration of the behavior changes or seems unusual or excessive.

When More Serious Problems Occur in Adolescence

Depression. We have found that there are some teens with Down syndrome who respond to the stress of adolescence with depression that includes a more severe form of withdrawal and isolation. When this happens, the anger that is usually expressed between parent and teenager may only occur when the parent tries to get the teenager to move out of his isolation. This tendency to withdraw and seek isolation may be due to the concept of "learned helplessness" (Seligman, 1975),

which also results in a high frequency of withdrawal and depression in adults with Down syndrome.

Learned helplessness can occur when the person has limited experience solving his own problems or standing up for himself. As a result, when faced with a major challenge, such as the physical and emotional turmoil of the teen years, he tends to shut down and withdraw into a state of helplessness rather than meet or deal with the challenge. This, in turn, may lead to an internal focus on fantasies, movies, or past events because people with Down syndrome often have good visual memory skills (see chapter 6). Withdrawing in this way may help to remove them from the conflicts and tensions in their world, but it only delays the resolution of the tasks of this life stage.

Teens without Down syndrome may also withdraw and isolate themselves to a certain extent, but they are also more likely to fight back. The reader may say here that teens without Down syndrome are notorious for shutting out their parents. While this is a form of withdrawal, there is nothing passive about this behavior, as anyone who has tried to talk to a sullen teenager will agree. On the other hand, teens with Down syndrome are more likely to hide and isolate themselves, perhaps giving others the "silent treatment," than to express anger directly or indirectly to others.

Suicidal Gestures

The "good news" about teens with Down syndrome who are depressed is that they are far less likely to make suicidal gestures compared to depressed teens without Down syndrome. This may be in part because suicidal gestures often involve a high degree of anger/rage at oneself and others, which is less likely to be expressed in teens with Down syndrome. Lack of understanding of methods of suicide or the link between self-harm and death may also contribute.

Attention-Deficit/Hyperactivity Disorder (AD/HD). We have seen teens with Down syndrome who have more serious behavioral issues. When we look closely, we often find one or more health or neurological problems, which, added to the intense stress experienced in the teen years, make it very difficult for these individuals to control their behavior. Some of these teens have an attention-deficit/hyperactivity disorder, which is characterized by impulsiveness, distractibility, and attention problems. As for children and teens without Down syndrome who have AD/HD, the

right medication and an academic program adapted to their needs may greatly help to reduce the stress experienced by these individuals. Still, the stresses of the teen years added to the AD/HD issues may present a major challenge for these teens and their families.

Tourette Syndrome. We have also seen that adolescent hormonal changes may trigger the onset of neurological disorders such as Tourette syndrome and bipolar disorder. As discussed in chapter 24, Tourette syndrome (TS) often involves a constellation of three sets of symptoms: (1) attention deficit and distractibility, (2) motor tics, and less frequently, vocal tics, and (3) obsessive-compulsive behavior. People with TS may be misdiagnosed as having only one of the above disorders or as having another disorder such as oppositional-defiant disorder (if the person's tics are viewed as willful rather than biologically based). Misdiagnosis may limit treatment success. This combination of these symptoms and behaviors is debilitating for adults but for a teen may be especially devastating, particularly when the teen is trying to establish himself in school and among peers.

Bipolar Disorder. Bipolar disorder is a mood disorder that includes extreme fluctuations in mood and behavior. This disorder often begins in adolescence and has a devastating effect on the child and her family. Bipolar disorder is more difficult to diagnose in those with Down syndrome, especially in adolescence, because the changes in mood may be confused with adolescent behavior. Additionally, teens and adults with Down syndrome may be more likely to have a pattern of rapid cycling from up and down states of mood. It is not uncommon for the cycle to occur in as short a period as one day as opposed to weeks or months in the people who do not have Down syndrome. See chapter 17 for more information.

Autism Spectrum Disorder. Autism spectrum disorder (ASD) is characterized by difficulties in social and communication skills and behavioral management problems. It may become more intense and unmanageable with the onset of puberty. However, many teens with ASD and Down syndrome have been diagnosed before adolescence, so although symptoms may be more intense, they are not necessarily new to the families and the professionals working with the teen. Often, the same strategies for dealing with teens with Down syndrome alone are useful for teens with Down syndrome and ASD. These teens want independence just like any others but may need more adaptations to achieve it.

Health or Sensory Problems. Finally, we would be remiss if we did not discuss teens with Down syndrome who have health or sensory problems. When we look into reasons that teenagers with Down syndrome have behavioral issues, we frequently

find health problems, just as we have reported for adults with Down syndrome (see chapter 2 for a detailed discussion of this).

Additionally, sensory integration (SI) issues may have a profound effect on the teen. SI is the ability of the person to receive and effectively manage and integrate all sensory stimuli, including hearing, vision, touch, proprioception, smell, and taste. Although SI is a relatively new area of study, there is evidence that problems with SI may affect a higher percentage of teens and adults with Down syndrome compared to those without Down syndrome. SI problems may be misdiagnosed as AD/HD and a host of other disorders and may also add to existing health or mental health problems. For the teen, health and sensory issues may make it even more difficult to manage the stresses of adolescence and thus may result in the types of behavior issues that are noted in this and other sections of this book. To resolve the behavior problems and give the teen a fighting chance to deal with the intense pressures of the teen years, we need to first diagnose and treat the health or sensory issues. See chapter 12 for more information on sensory issues.

Behavior Changes in Adulthood

Although many people with Down syndrome begin acting like adolescents later than usual, the reverse is true when it comes to acting like an older adult. People with Down syndrome seem to age more rapidly than their peers without Down syndrome. Actually, in childhood and early adulthood, many people with Down syndrome seem younger than their age. However, as they progress through their thirties, they often begin to look and act older than their age. In our experience, we tend to think of people with Down syndrome beyond thirty-five or forty as being five, ten, fifteen, and even twenty years older than their chronological age. They begin to have the health problems associated with being older, and they tend to "slow down" sooner than others. Therefore, the issues of older adulthood may arise at a younger age.

For older adults *without* Down syndrome, changes or challenges include children leaving home, becoming grandparents, retiring, and dealing with changes in their own or their parents' health. A person with Down syndrome may experience many similar challenges: becoming an aunt or uncle, retiring, and dealing with changes in their own or their parents' health.

As people with Down syndrome age, they can continue to learn. Whatever developmental age or level of skills they have attained, they still have the benefit of

living and learning. Like other older people, as people with Down syndrome age, they often become calmer, more patient and accepting of themselves and others, and better able to make thoughtful and wise decisions. However, like some older people without Down syndrome, people with Down syndrome may become reluctant to learn new things or to accept any change and become "more set in their ways." They may also prefer quieter and more sedentary activities. They will often mimic older parents who prefer these activities. If they do not develop Alzheimer's disease, people with Down syndrome experience memory and cognitive declines similar to those of other older adults without Down syndrome.

Since many adults with Down syndrome were born to older parents, they are often relatively young when their parents encounter their own aging issues. Interestingly, some people with Down syndrome are still processing the issues of adolescence (e.g., siblings leaving home) when they began dealing with issues of older adulthood (e.g., parents declining).

Lawrence worked in a rather fast-paced job. As he got older, he found it more difficult to keep up with the pace. Both physically and mentally, the job was a greater challenge. When Lawrence was forty-eight, his father retired, and Lawrence saw that as a more attractive alternative to going to work each day. He began to refuse to go to work and eventually lost his job. He became very sedentary and now spends most of his time sitting in front of the television controlling the remote.

Melissa, age forty-four, was brought for evaluation by her sisters. They were concerned about depression because she was not interested in doing activities that she had enjoyed for so many years. These activities included going to dances and sports activities such as softball, volleyball, and swimming sponsored by the local park district, as well as track and field events run by Special Olympics. She continued to work at her assembly job, but she was not as productive as she had been in the past. They also found that she did not have as much energy or enthusiasm for longer shopping trips or family gatherings, particularly when a number of younger children were present.

Melissa's sisters were relieved when we found no evidence of depression. We explained that people with Down syndrome have a premature aging process, which may account for an approximately twenty-year difference compared to people without Down syndrome. Like others who were aging, Melissa was moving slower and was interested in activities that were less taxing and yet still stimulating to her. She liked arts and craft activities and bingo, as well as shorter shopping trips that put less strain on her feet. She also continued to participate in Special Olympics, but switched to bowling, a less strenuous activity. In other words, her patterns of behavior were not unusual for a person who was developmentally at an age closer to retirement.

Why did Melissa's aging process seem to be more positive than Lawrence's? We think it is because her family understood that not only do stamina, interests, and motivation change as people age, but people may also become a little more set in their ways. They learned that these changes tend to occur earlier in adults with Down syndrome and helped Melissa choose new activities that were stimulating for her but less taxing. They made these changes while being careful not to disrupt Melissa's grooves too much. As previously mentioned, it is common for older people to get set in their ways. However, because so many people with Down syndrome have a preexisting tendency for grooves, the tendency may become more pronounced with age. This is not to say that people have to be stuck in groove-like patterns, particularly if these patterns are no longer adaptive for them. We do find, however, that caregivers need to be a little more patient when trying to change grooves. Older adults with Down syndrome may simply need more time. Additionally, the longer a pattern continues, the more difficult a change may be. But this is not true for all changes. We have seen many people respond very quickly to changes that they themselves view as positive. For example:

Juan showed great enthusiasm after he moved out of his aging parents' home and into his new residential facility at the age of thirty-nine. His sister, who helped facilitate this move, could not bring herself to tell her parents how happy Juan was because they had been convinced that he would fall apart if he left their home. She dealt with this very

tactfully by telling her parents that he missed them but that he was making progress (because they had prepared him so well for this move).

On the other hand, we have seen people have enormous difficulty with abrupt changes such as an unplanned move to a group home when a parent suddenly died. This is not true for all adults with Down syndrome--some do very well with these types of changes. However, a significant number do have adjustment difficulties. We also see that people do adapt, given time and a sensitive caring environment, but this process may take years and much pain.

The message we hope parents will take from this is to prepare for your son and daughter to leave your home well in advance. This will allow the person with Down syndrome and his parents—who often have more difficulty with this transition than their son or daughter does—to deal with this productively. More importantly, parents who wait may have no say in where and how their son or daughter will be moved. Our advice and the advice of anyone who has had to assist a person with Down syndrome adapting to an abrupt change is that advance planning and placement is far preferable to the alternative.

Not Eternal Children

People with Down syndrome are often referred to as "kids" or "children," even when they are fifty years old. The person with Down syndrome is viewed as if he is frozen in childhood. There are many possible reasons for this. Certainly, the presence of an intellectual disability leads many people to mistakenly assume that the adult with Down syndrome is still a child. While the person may be "childlike" in some respects, there are also generally some very adult qualities, dreams, and hopes.

Another possible reason for seeing people with Down syndrome as perpetual children is related to the kind of world our present-day adults with Down syndrome were born into. Those who were born in the 1960s or earlier had little access to basic health care, much less school, social, and other opportunities, and there was little expectation that they would live beyond childhood. Those who were born a bit later were legally entitled to a public-school education (starting in 1975) but still may have faced a great deal of ignorance and resistance to inclusion in everyday life. Families of these older people with Down syndrome sometimes have difficulty seeing the person as an adult because the professionals and community members surrounding

them may have had low expectations and because there were more limited opportunities to develop skills.

Today, however, we have a new generation that was born into a world where early intervention and inclusion in school and community are rights. As we watch the new generation of children with Down syndrome grow, we often wonder if we will see a different syndrome with improved health as well as improved cognitive, social, and occupational skills. As we see people with Down syndrome living longer and developing greater skills, it is imperative that we understand and support their development into adolescents and adults and throughout their lives.

Retirement

In the case of Lawrence, above, deciding to quit work (retiring) was something he desired that others did not appreciate. He made his wishes known in the only successful way he found, declining to go to work.

The age at which people with Down syndrome are ready to retire is as variable as it is for people without Down syndrome. Some people with Down syndrome are ready to retire in their forties. (Considering the premature aging, this might be comparable to someone else retiring in their sixties.) Some people with Down syndrome are never ready to retire because work is very important to them. There isn't a set age; each person's needs must be individually taken into account.

If the workplace has become too great a challenge physically, mentally, or both, it is time to explore different options for the person. It isn't a time for the person to do nothing. Although retirement is often seen as a time to do less, it is actually a natural part of the development process. If families and staff do not see this change as a natural part of development, they can have difficulty accepting it. In addition, state regulations often require some form of day programming for people with Down syndrome who live in a group facility. Therefore, accepting the change is not only an issue of changing the mind-set but may also involve dealing with, and adjusting to, regulations.

If this developmental stage is viewed as "retiring *to* something" instead of "retiring *from* something," the chance of success is greater. A successful retirement program for an adult with Down syndrome should have the following characteristics: a slower pace, less "work," more recreation, and provisions for changing health needs. It may need to include a place where the person can rest during the day if he needs to.

Retirement programs should not be boring or less than stimulating. We have seen some programs that are concerning—where the adults' only activity is to watch TV or to find their own means for entertaining themselves. Fortunately, as the population of people with Down syndrome ages, we have seen a growing number of programs that meet adults' needs while also stimulating and enriching them. The better programs have excellent arts and crafts activities, including some that are taught by professionals (often producing gallery-quality work). These programs also provide regular outings to restaurants and shopping centers and include healthy doses of cultural, sports, aerobics, and other recreational activities. Many of these programs could complete with any senior program anywhere. The people in these programs may be retired, but they are anything but inactive and they love their new lives. Many group facilities have developed programs with these characteristics that also satisfy the requirement that each person living there must attend a day program.

Volunteer work can often meet these requirements as well. We have heard of a number of innovative programs involving volunteer or civic-minded projects for elderly people with Down syndrome. For example, one program brings a group of "retirees" composed primarily of people with Down syndrome to a nursing home. The purpose is to help residents in the nursing homes who are lonely, aged, and sick by talking and socializing with them. Staff from these nursing homes report that something magical happens when the group of volunteers with Down syndrome arrives. This should not be surprising, given the sensitivity of people with Down syndrome to the needs of others. For the people with Down syndrome, there is perhaps nothing so enriching and invigorating as having the opportunity to do for others and especially others who are so appreciative.

Even when excellent seniors' programs such as those described above are available, they may not be the best fit for some people. The key is finding the program that is right for the individual. Does the person want to retire? Is his job too stressful? Would modifying the job meet his needs? Would a different program be better suited? Retirement is not right for everyone, and there is no one retirement program that is right for everyone who is ready to retire.

Victor, age fifty-two, was getting less and less enjoyment out of work. He found work increasingly challenging and frustrating, and his production continued to drop. He still liked to keep busy, however. He enjoyed the slower pace of cleaning the house where he lived. He could

rest when he was tired and then start working again. His verbal skills are quite limited, so he made his desires known by staying up late at night to clean and then refusing to get out of bed in the morning to go to work. Victor was transferred to the "retirement program" that met in the next building. There he was able to spend part of the day cleaning like he wanted, and part of the day socializing with the retirement group. His sleep improved because he was able to do what he wanted to do during the day and didn't feel he needed to be up at night to do it.

Conclusion

People with Down syndrome are not static. This is true at retirement age, during adolescence, and throughout their lives. Their needs, desires, and wants change over time, just as everyone else's do. Many approaches that are successful for people without Down syndrome can be used to provide beneficial and caring support. In addition, however, it is important to remember the differences in people with Down syndrome—such as slower development and early aging—that may require some modification to the support.

Return to Table of Contents

Chapter 12

Sensory Processing Differences and Disorders

Katie Frank, PhD, OTR/L

We received a phone call from a parent who was concerned because of behavior changes over the past month in her nineteen-year-old son with Down syndrome. Tyler seemed to be angry and would leave the classroom at school without warning and yell at his teacher. There was also an increase in his self-talk. These outbursts of anger were occurring both at home and school, especially during transition times.

During an occupational therapy (OT) evaluation, a detailed history was taken including sensory processing challenges. It was noted that Tyler was taking medication for ADHD. Tyler's parents reported that the school had been attempting to incorporate sensory strategies throughout the day, but the assessment found that the strategies were not being used properly. During the evaluation a variety of proprioceptive sensory strategies were tried so see which ones Tyler preferred. Through trial and error, it was recommended that he receive sensory input breaks every two hours throughout the school day and at home. During these breaks, choices would be offered between two or three options, including joint compression, wall push-ups, weighted blanket/vest, Theraputty, handheld massager, etc. It was also recommended that the sensory breaks be kept short (no longer than twenty minutes, especially with weighted objects).

The evaluation occurred over a holiday break, but once school started again, the new strategies were incorporated and there was an immediate change in behavior. Transitions became easier and there were no anger outbursts. After a few months, the family followed up and continued to report positive behavior changes with the increase in sensory breaks throughout the day in addition to his medication. Helping keep Tyler's body regulated had a positive impact on his behavior at both school and home.

No one has a perfectly functioning sensory system. We all have sensory preferences and even sensory needs, each unique to how our central nervous system perceives and processes sensory information. Think about your sensory system being like electrical wiring. When there is a kink in a wire, it may cause lights to flicker. Sometimes we get a kink in our central nervous system, and that leads to mixed messages in our sensory system. Many times, we may not know what is causing that kink or how to stop our own internal lights from flickering. When sensory processing is disorderly, the brain cannot do its most important job of organizing sensory messages. When this interferes with someone's ability to function in daily life and becomes a disruption, the person may be diagnosed with sensory processing disorder (SPD).

Sensory processing challenges, which are relatively common among people with Down syndrome, do not just affect how a person moves and learns. These challenges also have an effect on how the person behaves; completes tasks at home, school, or work; and interacts with others. Individuals with SPD may experience touch, taste, sound, smell, movement, and other sensations differently from those without SPD. Some may feel sensations more intensely, others feel them less so, and some just do not get sensory information "right": up may feel like down or a penny feels the same as a button. These sensory responses can lead to challenging behaviors (Will et al., 2019). When adults with Down syndrome have sensory difficulties, even without a diagnosis of SPD, those problems should be addressed in order to help them live each day feeling regulated.

We Have Eight Senses

Most people are familiar with the senses of touch, sight, sound, taste, and smell. However, there are three additional senses that are not as well known: vestibular, proprioception, and interoception. We can also think of the five familiar senses as the *far* sensory system and the three additional senses as the *near* sensory system. The *far* sensory systems tell us what is going on in the world around us, while the *near* sensory systems tell us what is going on in our own bodies (Wild, 2015). We will discuss the importance of the near and far sensory systems below. Ideally, these sensory systems should work together like a team.

Far Sensory System

Tactile/Touch. The tactile sense provides our nervous system with information about touching and being touched. More specifically, it tells us not just that we have been touched or touched something, but how we interpret that touch or how that touch feels. For example, if we touch something that is small, circular, and smooth, we have to determine whether what we feel is a button or a penny (Kranowitz, 2016).

Visual/Sight. The visual sense provides information about what we see in the environment. Not only does it include visual acuity (i.e., what our eye sees and how clearly), but also visual processing (i.e., interpreting what our eyes see). For example, we see rounded lines on a piece of paper and have to decide whether it is the letter *C* or *O*. The visual sense also helps us filter out things we do not need to see; this can keep us from getting distracted by external stimuli. For instance, if we are looking for a pen in our junk drawer, we can filter out all the items in the drawer that are not shaped like a pen (Kranowitz, 2016).

Auditory/Hearing. The auditory sense provides information about what we hear. The auditory system includes not only what we hear, but also how we interpret (process) what we hear and filter out what we do not need to focus on (Kranowitz, 2016). This allows us to focus on our conversation with someone else in a crowd or hear someone call our name when we are watching TV or listening to music.

Gustatory/Taste. The gustatory sense provides information about what and how we taste. For example, it enables us to discriminate certain flavors like sweet, sour, and salty.

Olfactory/Smell. The olfactory sense provides information about what we smell. It also can influence our sense of taste. Our sense of smell can keep us safe because we may not eat food that has gone bad based on smell and we may exit a building because we smell smoke or gas.

Near Sensory System

Vestibular System. The vestibular sense helps our body move through space. Our vestibular system resides within our inner ear and helps us keep our balance, stand upright, and change our head position without getting dizzy. The vestibular sense is a master sense. It helps organize all the other senses because input to this system can be beneficial for up to eight hours. This input helps to keep our entire body regulated (Kranowitz, 2016).

Proprioceptive System. The proprioceptive system provides information about where our body is in space and how much pressure or force we use when we pick things up, walk, sit down, or otherwise use our body to interact with the environment. We receive proprioceptive sensations through our muscles and joints, and the system activates anytime we push or pull objects or our joints are compressed or stretched.

Interoceptive System. Interoception is a sensation related to the physiological condition of the body. There are internal receptors that help us process the sensations coming from within our body such as heart rate, thirst, hunger, elimination, digestion, and sweating (Kranowitz, 2016).

Importance of Near and Far Sensory Systems

It is believed that the *near* sensory systems need to be regulated first in order for the *far* sensory systems to be regulated (Wild, 2015). When a sensory system is regulated, it means optimal arousal is achieved; we are neither understimulated nor overstimulated. Difficulties with the *near* systems require activities (often involving therapy) in order to treat, but the *far* sensory systems can be addressed by providing accommodations and adaptations (Wild, 2015).

At the end of the chapter, the section on sensory diets discusses in detail how sensory difficulties are addressed; however, consider the following as an example in the meantime. If a person has deficits with proprioception (i.e., difficulties regulating her force, often doing things too hard or rough), heavy work activities that encourage more use of the muscles and joints can increase body awareness. If that same person does not like to be in gymnasiums during basketball games because of the loud noises (whistles, buzzers, fans, etc.), we can make an accommodation by encouraging the use of sound-reducing headphones. Oftentimes, because of lack of resources or time, school-based occupational therapists (OTs) are able to better address the far sensory systems with accommodations and not the near sensory systems with activities. These students, plus adults with Down syndrome who are out of school, may need to seek assistance from outpatient or private occupational therapy (OT) to determine strategies to regulate the near sensory systems.

What Sensory Processing Is *Not*

SPD is frequently misdiagnosed and treated with medication or therapies that do not address the underlying sensory issues. One reason may be that the American Academy of Pediatrics (AAP) issued a position statement recommending that pediatricians not use SPD as a diagnosis but instead evaluate for other developmental disabilities when a child has sensory symptoms (Zimmer & Desch, 2012). The AAP came to this conclusion because research has not definitively proven that treating sensory processing disorder as a stand-alone diagnosis is effective. Unfortunately, treating sensory symptoms is highly individualized, and creating a research study to evaluate effectiveness can be challenging. However, a few studies have demonstrated positive results. For instance, OTs have shown that using sensory strategies in acute psychiatric settings has been effective in creating nurturing treatment environments and allowing patients to self-organize (Champagne, 2005). Also, when researchers conducted a systematic review of the effectiveness of sensory integration (SI) interventions based on twenty-seven studies, they concluded that an SI approach tends to produce positive results, especially when compared to no treatment (May-Benson & Koomar, 2010).

Instead of diagnosing SPD, pediatricians are instructed to look into diagnoses such as autism spectrum disorder (ASD) and attention-deficit/hyperactivity disorder (ADHD) (Zimmer & Desch, 2012). Later on in the chapter, you will see that the red flags and behaviors of some sensory systems can appear similar to ADHD. We also know that sensory processing difficulties can be part of ASD, but the mere presence of sensory processing deficits does not mean a person has ASD, which is often misunderstood. In addition, SPD can look like an eating disorder, anxiety, bipolar disorder, or obsessive-compulsive disorder (Kranowitz, 2016). While SPD can co-occur with these diagnoses, it is important to evaluate the person thoroughly to determine the root of the issue; oftentimes an interdisciplinary evaluation can help with this.

Types of Sensory Processing Disorders

There are three different types of sensory processing disorders: Sensory Modulation Disorder (SMD), Sensory Discrimination Disorder (SDD), and Sensory-Based Motor Disorder (SBMD). Unfortunately, little is known about the occurrence of these different disorders in individuals with Down syndrome. However, recent studies have found that children with Down syndrome tend to have low sensory responsivity and

sensory-seeking behaviors, both of which can occur in SMD, and that they often have difficulty filtering auditory stimuli, which can be present in SDD (Bruni, Cameron, Dua, & Noy, 2010; Will, Daunhauer, Fidler, Lee, Rosenberg, & Hepburn, 2019).

Sensory Modulation Disorder

SMD has to do with problems related to a person regulating responses to sensations. That is, she has trouble focusing on only relevant stimuli and filtering out unimportant stimuli (Wild, 2015). Reponses may be more intense than the situation demands (over-responsive) or less than the situation demands (under-responsive), or the person may be motivated to obtain sensory stimulation, but this input causes disorganization and the need for even more input (sensory craver). When someone has a modulation disorder, it is because the brain has difficulty determining what is important (Wild, 2015). This is the most commonly seen type of SPD in general.

Sensory Over-Responsive (SOR). Individuals who are over-responsive respond to sensory messages more intensely, more quickly, and/or for longer periods of time than individuals with normal responsivity (Johnson, 2016). This is also known as being a sensory avoider or sensory defensive. Often these individuals require calming activities, and the behaviors we see as problematic are just how the person attempts to calm herself down (Wild, 2015). People can be over-responsive in any sensory system or combination of sensory systems, although the tactile and auditory systems are most commonly affected (Johnson, 2016). SOR can occur in combination with other types of sensory processing difficulties. It is also possible to be over-responsive in one sensory system and under-responsive or a sensory craver in another. For example, it is common to be over-responsive to far sensory systems and under-responsive to near sensory systems (Wild, 2015).

There are some common patterns of SOR and as a result, associated behaviors. For instance, people who are SOR (both with and without Down syndrome) may frequently be bothered by some of the following: textures, sounds (loud or unexpected or background noise), movement, bright lights, smells, haircuts, or being dirty. Because of this, the person may respond aggressively when overwhelmed by sensory information or may come across as fussy or moody. This is because the fight or flight response kicks in. Group activities may be avoided, and the individual may be cautious about trying new things for fear of encountering overwhelming sensory information. Often individuals with SOR are upset by transitions and changes to routines (Johnson, 2016).

Loud sounds bother Nicole. She has an especially difficult time in gymnasiums because the sounds of the crowd, buzzer, ball, and whistles echo. Her younger brother's team is playing in the state basketball finals, and she really wants to go cheer him on. Luckily, Nicole tolerates wearing sound-reducing headphones, so she is able to use those while watching the game. However, she still gets anxious knowing she is going to a gym. In the car on the way to the game, she listens to music from her favorite band and sits under a weighted lap pad, which helps relax her. She asks to bring the weighted lap pad into the gym in case she needs it during the game. These are examples of strategies that can be used to help someone who is over-responsive.

Sensory Under-Responsive (SUR). These individuals exhibit less of a response to sensory information than the situation demands. They may take longer to react or require relatively intense or long-lasting sensory messages before moving into action (Johnson, 2016). People who are SUR may be more socially withdrawn and are often thought of as "well-behaved." These individuals benefit from alerting or stimulating activities.

There are some common patterns of SUR and as a result, associated behaviors. These individuals do not cry when they get hurt. They do not seem to notice when someone has touched them or they are touching others. They prefer sedentary activities and are often unaware of what is going on around them. They do not seem to notice when they are dirty. They may not feel hungry or be aware of the need to use the restroom. These people often seem more passive and withdrawn. They are difficult to engage in conversation and slow to respond to directions, either because the world is moving too fast around them or once they are alert enough to respond, the conversation has continued without them. These difficulties are often seen in people with Down syndrome, especially when the individual has slower auditory processing. Sensory under-responsive people lack inner drive or motivation and may be uninterested in exploring the world around them (Johnson, 2016).

Anna has choking incidences. This is because her mouth is under-responsive to tactile and proprioceptive input. She cannot feel the food in her mouth when the bites are small, so she overstuffs her mouth, but then she has trouble chewing and swallowing.

Sensory strategies that are helpful for Anna include gentle vibration/massage to the inside and outside of her cheeks before mealtime. If she is out in public, she makes sure to drink out of a straw to "wake-up" her mouth. She also has a visual support reminding her to take small bites and chew five to ten times before swallowing. These are examples of strategies that can be used to help someone who is under-responsive.

Sensory Craver. Sensory cravers actively seek sensation in order to meet their sensory needs. However, the stimulation results in disorganization and does not satisfy the sensory need (Johnson, 2016). These individuals are often in constant motion and have no regard for safety. This behavior is often confused with ADHD. While it may seem like these individuals would benefit from calming activities, they actually require alerting or stimulating activities provided in an organized and structured manner (Wild, 2015).

There are some common patterns and behaviors observed with sensory cravers. For instance, they are constantly on the move and often engage in activities with no regard for safety. They have trouble taking turns, talk excessively, and interrupt others. They have little regard for personal space and frequently touch other people and things. They prefer their music and TV at loud volumes and may lick or chew nonfood items. Overall, these individuals are intense and may be hard to calm. They may even be angry when asked to sit still (Johnson, 2016).

> *Sierra frequently bites her fingers and licks the palms of her hand. When seated, she likes to rock back and forth in her chair. She craves sensory input to her vestibular, tactile, and gustatory systems. She could do this for hours. Unfortunately, this sensory input does not satisfy Susan's needs and leads her to search for even more stimulation. However, when proprioceptive and vestibular strategies are integrated into Sierra's day, she rarely bites on her fingers or licks her palms. She also rocks less. She prefers swimming and swinging on the porch swing. She has also started power lifting with Special Olympics and walks on the treadmill at home. She has started eating more raw vegetables as well. These are examples of strategies that can be used to help someone who is a sensory craver.*

Sensory Discrimination Disorder

Someone with SDD may have difficulty distinguishing and interpreting sensory input. More time may be needed to process information because of trouble figuring out what is being perceived as quickly and naturally as others do. Difficulties can occur in any of the sensory systems.

Depending on which sensory systems they have trouble with, people with SDD may have a variety of difficulties. For example, those who have trouble with visual discrimination may have trouble finding something in a crowded space. Those with auditory discrimination difficulties may have trouble recognizing differences between sounds and may be easily distracted by particular sounds. Some people with SDD have trouble distinguishing pleasant from noxious smells or the difference between sweet or spicy foods. People with difficulty with vestibular discrimination may not realize they are falling until they have fallen, and those with problems with proprioceptive discrimination may not know how hard to squeeze something without breaking it (Johnson, 2016). Due to sensory discrimination problems of this nature, individuals with SDD may have trouble following directions, get lost easily, have difficulty completing homework, or may need instructions repeated multiple times. They often need more time to transition as well.

> *Kelly has trouble regulating her force. She often pets the dog too hard and breaks pencil tips when writing in her journal because she presses down too hard. When she was younger, she loved to wrestle with her dad. Kelly has trouble with discrimination of the input into her proprioceptive system and therefore has trouble realizing when she is being too hard or rough. She has trouble determining the right amount of pressure. She needs a sensory-rich environment full of proprioceptive input to help her body learn the just right amount. In occupational therapy, Kelly was helped to first regulate her proprioceptive system by completing a chore like vacuuming the carpet or sweeping the floor. Her proprioceptive system was then challenged by having Kelly use tweezers or beginner chopsticks to eating a snack like popcorn or grapes. She had to use just the right amount of pressure to pick up the snack without dropping it or smashing it. After her snack, she would get more proprioceptive input like sitting under a weighted blanket or in the massage chair.*

Sensory-Based Motor Disorders

SBMD describes dysfunction that occurs when the "hidden" vestibular and proprioceptive senses that allow our bodies to move and sense our position in space are impaired (Johnson, 2016). These individuals have a regulated sensory system, but have difficulty with sensory motor activities including balance, coordination, motor planning, and sequencing. There are two types of motor disorders, which are discussed below.

Postural Disorder. Postural disorder is the result of having difficulty maintaining control over the body in order to meet the demands of a motor task (Johnson, 2016). For instance, when working at a desk, the individual often will be slumped over in order to try to support her body when writing or when watching a movie on her tablet. People with postural disorder often have low muscle tone, weak grasp on objects, and difficulty using both hands together. They may appear lazy or unmotivated and have decreased endurance. They have poor balance and tend to fall easily. Individuals with postural disorder may sit in a "w-sit" position or walk with a wide base of support, both of which are often seen in people with Down syndrome. While it is unknown if every person with low muscle tone also has postural disorder, the treatment strategies for postural disorder are often effective for individuals with Down syndrome because of their postural instability.

> *Ashley slumps at the dinner table. Her weak muscles, low tone, and poor core strength make it hard to sit and stay upright. She is clumsy stepping up onto a curb, reaching for her water bottle, kicking a ball, and other tasks that require her to position her body accurately. She would rather stay seated because moving takes so much energy. This affects her self-esteem because she is embarrassed when she is out in the community and has difficulty with tasks that others find easy to complete. Ashley would benefit from activities to help strengthen her core and postural muscles. An OT, physical therapist, or personal trainer could help her with this. Special Olympics and other recreation activities and sports are great ways to naturally work on increasing strength and endurance.*

Dyspraxia. Many people are aware of apraxia of speech. This occurs when the brain wants to say something, but the words or sounds that come out of the mouth are not the same as the message sent from the brain. Similarly, dyspraxia, which contains the same root word, refers to problems getting the body to move as desired due to difficulty translating sensory information into physical movement. People with dyspraxia have difficulty thinking, planning, and/or executing movements. These individuals have difficulty with multistep instructions or learning skills that require coordination, including handwriting and self-care tasks. They may appear clumsy and disorganized and become frustrated when unable to complete tasks (Johnson, 2016). Many of the tasks we do each day require a plan. We often hear that teens and adults with Down syndrome have difficulty remembering the steps to tasks like dressing or bathing. This often frustrates caregivers; they do not understand why the task is not more automatic for their loved one. Instead of thinking about it as a memory issue or fine motor issue, it is likely to be a motor planning issue. In order to help establish a plan, a visual support can be effective. A visual support can provide the prompts needed to complete a task, and with practice, help the activity become more intuitive.

> *After Georgia gets into the car, her mother waits a few minutes and then asks Georgia to put on her seat belt so they can leave. Georgia has to think about all the steps that she needs to take in order to buckle her seat belt. She becomes frustrated and finally begins to put on her seat belt. Three minutes later she manages to fasten her seat belt. Sensory strategies like proprioceptive input (which are discussed later in the chapter) could help regulate Georgia's body so she would be less frustrated and better able to focus on fastening her seat belt. A visual support could provide step-by-step prompts to remind Georgia of the steps and avoid the frustration of coming up with the steps on her own. An OT could also suggest a modification to the seat belt so fewer steps are needed to complete the task.*

Sensory Processing and Down Syndrome

Sensory processing problems are relatively common among adolescents and adults with Down syndrome, and certain difficulties are reported more often than

others. However, it is important to remember that each person is unique, and it should never be assumed that one person with Down syndrome will have the same sensory problems as another.

Sensory processing deficits may make daily tasks like bathing, dressing, and eating a challenge. We often see and hear that patients do not tolerate lotion on their skin, despite it being dry. Some individuals do not like water touching their face, which affects bathing and hygiene. People with Down syndrome may not like to wear socks, shoes, or other pieces of clothing because of the way they feel. Transitioning from one season's clothing to the next can also be a challenge.

Many teens and adults with Down syndrome have sensory-related eating problems. For example, they are often reported to be picky eaters. Others may stuff their mouth full when eating. This may be due to proprioceptive deficits. In order for a person to chew, she must feel the food in her mouth, and she might only have this sensation if she stuffs her mouth full to her cheeks. Unfortunately, this is not a safe eating habit. Other feeding issues that are reported include difficulties feeling thirst or satiation (full). This is actually related to the interoceptive system, one of the eight sensory systems!

Many people with Down syndrome have hearing loss or wax buildup in their ear canals, which can muffle sounds. Compounding these hearing problems, they often have difficulties with auditory processing. They often can be overwhelmed by loud or unexpected sounds yet prefer their music at a loud volume. In addition, because of slow auditory processing, they often perform best with visual cues or instructions verbalized in simple ways and repeated often. Depth perception is also a challenge for many individuals with Down syndrome and can make it difficult for them to go up and down stairs or maneuver on uneven surfaces. People with Down syndrome also tend to have low muscle tone, which can affect how they interpret sensory input coming in through their muscles and joints (i.e., proprioceptive input) (Bruni, 2016). Frequently they require more proprioceptive input into their muscles and joints in order to help regulate their bodies. This also makes it challenging for people with Down syndrome to regulate the amount of force they generate with their muscles; they often do something too hard (apply too much pressure to an object so it breaks) or not hard enough (apply insufficient pressure and have a hard time holding on to an object).

Evaluating and Treating Sensory Processing Disorders

When it comes to diagnosing SPD, there is currently no gold-standard assessment. However, occupational therapists (OTs) can help assess and treat people whose ability to function throughout the day is negatively affected by sensory processing disorders. OT is a health profession concerned with how people function in their respective roles and how they perform meaningful activities. OT views "occupation" as anything a person needs to do from the moment she wakes up to the moment she falls asleep. Unfortunately, occupational therapy is not a profession that is able to provide medical diagnoses; that is outside the scope of practice. Therefore, OTs cannot diagnose SPD. However, OTs can assess what is interfering with a person's ability to engage in activities, including an impaired sensory system. OTs use formal and informal assessments as well as clinical observation and caregiver interviews to evaluate and assess for sensory processing deficits and then use the results to create a treatment plan.

While all OT students receive education in sensory processing, not all practicing OTs are comfortable working with individuals with sensory dysfunction or have the equipment or support to provide these services. The other challenge is that pediatric and school-based therapists may address sensory needs, but who can an adult with sensory needs see for therapy? Many OT practitioners who do see adult clients work in rehab or other settings that are not well-equipped to address the needs of an adult with sensory processing deficits. It can be particularly difficult for adults with Down syndrome and sensory needs to find an OT who can help. These individuals may be permitted to be seen in a pediatric setting with clinicians who are skilled to address these needs, but many times insurance does not cover these services in that setting. Locating skilled therapists in your area to address sensory needs is unfortunately a huge challenge for many families.

Evaluation

When it comes to testing teens or adults with Down syndrome for sensory processing difficulties, there are just a few options. The only formal assessment for SPD that currently exists is the Sensory Integration and Praxis Test (SIPT), which is only standardized for children ages four to eight (Ayres, 1989). Lucy Jane Miller and her team are currently working on the development and standardization of a new formal assessment for individuals ages three and older. It is called The Sensory

Processing Three Dimension Scale (SP3D). There is, however, a questionnaire called the Sensory Profile 2 (Dunn, 2014), which covers individuals from birth to age fourteen and is available in Spanish. In addition, the original version of the Sensory Profile has an adolescent/adult version for ages eleven and older (Brown & Dunn, 2002). While parents/caregivers often complete this questionnaire, it is standardized only when completed by the individual herself.

An informal, free, and easy-to-complete checklist for families is the Sensory Systems Checklist (https://sensationalbrain.com/pdf/SB-sensory-checklist.pdf). This is not used to diagnose but can help create a nice picture of how the person engages with her environment. There is also a version of the checklist available in Spanish (Wild, 2015).

A second part of an evaluation process is to further assess motor skills. The Bruininks-Oseretsky Test of Motor Proficiency, second edition (BOT-2), is often used, but it is only standardized up to age 21 years, 11 months (Bruininks & Bruininks, 2005). The final aspect of the evaluation process is for the therapist to complete a detailed observation in a clinical setting and identify the person's strengths and weaknesses.

Treatment

There are many reasons to seek treatment for sensory processing deficits. First and foremost, sensory processing deficits are not something that are outgrown; they grow and change with us. Treatment will help a person function more smoothly and can also help to develop social skills. As stated earlier, individuals with sensory modulation deficits often have difficulty forming relationships with others. This is because they either respond to sensory input with a fight or flight response—perhaps yelling, hitting out, or running away when they are overstimulated—or they seek out more input—perhaps by chewing on their fingers or flapping their hands when they are understimulated. These reactions can be hard for others to understand and may make it hard for someone to want to be friends with this person because of their sensory responses. Once their bodies feel regulated, it can help them learn and be more emotionally stable. Finally, treating sensory processing deficits can improve family relationships (Kranowitz, 2016).

Think about your body/sensory system as a teeter-totter. There are times when your sensory system is over-responsive (you have received too much sensory input), perhaps because you are stuck in a loud or chaotic setting and just want to get out of

there. In these cases, you are up high on the teeter-totter. There are other times when you are under-responsive (you do not have enough sensory input), perhaps because you have been sitting listening to a lecture for an hour and feel as if you are going to fall asleep. In these cases, you are low on the teeter-totter. When you are regulated, the teeter-totter is even. Our goal is to ensure the teeter-totter stays even!

Sensory Diet

Treatment for SPD or any sensory processing deficit is highly individualized. An OT can provide direct therapy in an outpatient setting or direct or consultative therapy in a school setting. A therapist will create a sensory diet, which is a planned and scheduled activity program designed to meet a person's specific sensory needs. It can include a combination of alerting, calming, and organizing activities based on the person's needs in order to keep an optimal level of arousal and performance (i.e. an even teeter-totter). Some trial and error may be needed to determine the "best" sensory activities for each person.

Building and implementing a sensory diet should be more like choosing from a menu rather than following a recipe (Johnson, 2016). One main aim of a sensory diet is to prevent sensory and emotional overload by satisfying the nervous system's sensory needs (remember the teeter-totter image). For instance, if a sensory diet activity is done before another activity, it can help a person move through the transition or prepare for a change in routine more easily (Johnson, 2016). However, it can also be used as a recovery technique if the person does become overwhelmed. A sensory diet can be implemented at home and in the community (e.g. school, day program, or work). One key feature to remember, especially when considering behavior plans, is that sensory strategies should never be removed as punishment or given as a reward. If a person has a sensory diet, it is because it is needed for optimal performance, and her body begins to rely on it.

Activities in a sensory diet can be alerting, organizing, or calming. ***Alerting activities*** benefit people who are under-responsive and need a boost (i.e., the teeter-totter is low). Examples of alerting activities include

- eating crunchy food,
- taking a shower,
- bouncing on a trampoline or therapy ball,

- swinging in a circle, or
- swinging fast.

Organizing activities help regulate a person's responses so she can be more attentive. Sensory cravers often benefit from these activities. Organizing activities include

- eating chewy foods,
- heavy work activities (e.g. proprioceptive input and other activities that include pushing and pulling like vacuuming or pushing a grocery cart), or
- vibration.

Calming activities help decrease sensory over-responsiveness (i.e., the teeter-totter is high). These activities include

- sucking on hard candy,
- heavy work (exercising, household chores, joint compression),
- swinging back and forth or forward and backward,
- rocking in a rocking chair,
- deep pressure/massage/compression,
- weighted products (blanket, lap pad, pillow, or neck wrap),
- playing with a fidget, or
- listening to a sound machine.

Activities as part of a sensory diet should be offered periodically throughout the day (in order to keep the teeter-totter level). Sensory breaks can be short in length, often only for a few minutes. Sensory input should not be provided all day long. For instance, a weighted vest will often lose its impact after approximately ten to fifteen minutes. Think about what happens when you put on jewelry: you may notice the necklace or ring initially, but at some point, you forget you are wearing it. It is the same with sensory input. There is a point when the body gets used to the sensation and tunes it out, making it no longer effective.

Sensory Strategies and Mental Wellness

In addition to helping with sensory processing disorders, sensory diets and other sensory strategies are sometimes used to treat conditions like anxiety, self-talk, and compulsion. There have not been any formal studies proving the benefits, but we

have found sensory strategies helpful for some individuals with Down syndrome. In particular, proprioceptive input (e.g., deep pressure and other input into muscles and joints) appears to help calm anxiety in some of the individuals. Deep pressure input is alleged to have a calming and organizing effect on the central nervous system by lowering states of arousal, resulting in positive behavioral and emotional outcomes (VandenBerg, 2001). Many of these strategies can be alerting, organizing, or calming depending on the person's needs. Joint compression is like a mini massage, and no equipment is needed except two hands. If the person with Down syndrome likes joint compression, she can perform wall push-ups independently, which is similar to joint compression, but without the need for someone else to be present to assist. Other ways we have successfully provided proprioceptive input are with weighted objects (e.g., snake, lap pad, blanket) and vibration. Vibrating options are varied, but we often try handheld massagers, vibrating pillows, or vibrating mats.

We successfully used proprioceptive strategies with one patient who did not like to get his blood drawn. (This is actually something that is common among people with Down syndrome.) This person tried some sensory strategies prior to getting his blood drawn. He liked the joint compression, so vibration was offered next. He seemed to like that as well. Finally, he lay down on the exam table under a nine-pound weighted blanket for about fifteen minutes. After the blood draw, his mother returned and reported excitedly that her son had walked in, stuck out his arm, allowed the blood draw without being restrained, and even held the cotton ball on his arm until the band aid could be placed! Remember, however, that sensory input is highly individualized. Another person received similar sensory input prior to her blood draw, and while she enjoyed the sensory input, she still did not tolerate the blood draw very well. Many people with Down syndrome also do not tolerate going to the dentist or getting their hair or nails cut and have trouble with transitions and changes to routine. Incorporating sensory input prior to these anxiety-producing activities may improve their tolerance.

It is important to note something here about weighted products, especially weighted blankets. First off, it is recommended that a weighted product should weigh no more than 7 to10 percent of a person's body weight. However, it can be less if that works best! Many commercial weighted blankets advertise weights that are more than the recommended 7 to10 percent. Please do not refer to the manufacturer's weight suggestions when purchasing a weighted blanket. It is also highly recommended not to sleep under a weighted blanket, especially if there are concerns with seizures, asthma, sleep apnea, or cardiac issues. However, a weighted blanket

could be used as part of the bedtime routine in order to help an individual calm herself before falling asleep. It is best if weighted products are used with supervision to ensure safety.

Sensory Processing and Alzheimer's Disease

As people age, their senses tend to become less acute. For example, older people generally cannot see things up close as well as they could when they were younger and they often lose the ability to hear higher-pitched sounds. We have observed a variety of sensory-related changes as people with Down syndrome age. For instance, the vestibular system is affected, and individuals tend to lean to one side or the other in sitting, standing, and even walking. The visual system is also affected, and people with Down syndrome have difficulty with depth perception in addition to narrowing of their visual field. Their proprioceptive system is already affected because of the low muscle tone; then, as they age, their muscles tend to atrophy as they become more sedentary. Because of these physical changes, adults with Down syndrome often experience psychosocial changes like anxiety and agitation during activities that involve impaired sensory systems. For example, someone who has trouble with depth perception and her sense of balance may feel quite anxious when walking up or down stairs. If an older adult with Down syndrome develops dementia, it affects the brain, which further affects how she perceives the sensory input she receives (Champagne, 2005).

Caregivers need to use strategies to help individuals with Down syndrome and Alzheimer's feel safer. Sensory strategies that are calming or alerting can help the person, especially with vestibular and proprioceptive changes. Remember the teeter-totter image from earlier in the chapter? When the teeter-totter is high, we are overstimulated and may need calming strategies, and when the teeter-totter is low, we are understimulated and may need altering strategies. See the chart below for calming and alerting strategies for various sensory systems.

Calming and Alerting Sensory Strategies for People with Down Syndrome and Alzheimer's Disease

Calming	**Alerting**
Warm shower of bath	Cool shower or bath
Holding or stroking a pet	Holding ice or a cool washcloth in hand or to face

Sitting in front of a fireplace	Being in a cool room
Wrapping in a heavy blanket	Wrapping in cool bed sheets
Massage/deep pressure touch	Fast-paced, upbeat music
Isometric exercises/yoga	Alerting nature sounds (birds chirping)
Leisurely walks	Strong scents (peppermint)
Slow/rhythmic music	Light touch
Calming nature sounds (waterfalls, oceans)	Aerobic exercise
Humming/singing	Power walks
Soothing scents (lavender)	Rough or prickly materials or textures
Soft materials or textures	Fast or bumpy car ride
Rocking in a rocking chair or glider	Spinning on a swing
Swinging on a swing	Fast and/or jerky movements
Slow rhythmic motions (swaying to music)	Bright or flashing lights
Soft/low lighting	Drinking tea or coffee
Decaffeinated and herbal teas	Biting into a popsicle
Chewing gum or sucking on candy	Sour or spicy foods/candy

Adapted from Champagne (2011).

Individuals with visual deficits may benefit from environmental modifications. We find that they often have difficulty going through doorways or walking on floor surfaces that change (wood to carpet or cement to grass). Add reflective tape on stairs or walls to help provide a color contrast. Remove doorsills to prevent tripping through doorways.

- Using contrasting colors can make everyday activities like using the toilet and eating easier. For instance, installing a toilet seat that is a contrasting color to the toilet as well as the floor can help someone remain independent with toileting. This same principle applies to eating. Use plates that are a contrasting color to the placemat or table. It is also important to make sure the food contrasts with the plate color. Additionally, the person will be safer moving around their home if doors and walls are contrasting colors.
- Because of the smaller visual field, we often hear that bathing is a challenge and individuals become anxious when it is time for a bath or shower. It may be helpful to have a shower chair or transfer tub bench for physical reasons, but a handheld showerhead can help when the visual field decreases. Seeing the water come from the handheld showerhead can lessen anxiety.

- Various lighting options can also be difficult. Make sure to reduce reflective glares during the day and bright lights at night. You also want to make sure the lighting is even throughout the house.
- Removing mirrors in bedrooms and bathrooms is recommended, as individuals with Alzheimer's disease may not recognize themselves and may perceive the person in the mirror as an intruder.

Is It a Sensory Processing Issue or Challenging Behavior?

This really is the $64,000 question. The answer often depends on who you ask. A psychologist or behaviorist will often label an action as a problematic behavior unless behavior management strategies are not effective to decrease or stop that behavior or area of concern. If behavior management strategies are not effective, the professional may make a referral to a physician, to rule out a medical problem, or to an OT, to check for sensory issues. An OT will most likely consider both sensory and behavior issues and provide recommendations based on both. The thing is, everything we do is behavior! It only becomes a concern when the behavior is truly challenging or problematic. These are behaviors like aggression toward self or others, noncompliance, tantrums, and disrespecting authority.

It is important to analyze why problematic behavior is occurring. Challenging behavior is often the result of a lack of skills (Johnson, 2016). Teaching the person what to do can therefore decrease the behavior. For instance, an adult with Down syndrome might be constantly interrupting you and others. You cannot teach her how not to interrupt, but you can teach the concept of waiting. Sometimes, however, problems with interrupting may be related to a sensory-based motor disorder (i.e., dyspraxia or postural disorders). These sensory problems may result in the person being noncompliant or easily frustrated and thus more prone to interrupting others.

So how can you tell if a challenging behavior is related to a sensory issue? Sometimes the only way to tell is to see which interventions are working. You can also ask yourself a few questions, but remember, it cannot always be that easy: right? Do the person's actions disrupt her own and others' lives? That is, does she dislike certain places because of noises, crowds, or smells? And, as a consequence, do family members or others who plan community outings avoid those places? If so, it could be a sensory problem.

- Do the actions occur with everyone? If so, the behavior is likely sensory. If it only happens at home, but not at school or work (or vice versa), it is likely to be behavioral. It is also important to remember that it can be a behavioral response to sensory input. For instance, you know your daughter, Maria, is over-responsive to auditory input and often gets overwhelmed in restaurants. You are at a restaurant for a family member's birthday and all of a sudden, Maria throws her drink on the floor. This behavior does not usually happen during dinner at home, but frequently happens when out in restaurants. Therefore, this is behavioral, but because the sensory environment overwhelms Maria.
- Finally, does the individual stop the action when given a reward? If so, it is likely behavioral. In addition, if someone is having a meltdown because of a sensory issue, then time on an iPad (or any other reward) typically will not fix the situation or stop the behavior.

When a challenging behavior is purely behavioral, it is designed to achieve a specific result: avoid/escape a task or situation, get attention, receive a pleasant feeling, obtain something material). If so, the individual can often turn the action on and off like a switch. The behavior may include crying or tantrums, but often without tears. The person will likely respond well to structure, boundaries, and clearly defined rules and expectations. Consistency is key, and visual supports are often helpful. On the other hand, if the challenging behavior has a sensory cause, the individual will have trouble calming down even after getting what she wants, and the response is the same with everyone. This is when a sensory diet should be put into place. Perhaps offer a positive time-out in a calm space. It is also important to determine the sensory triggers and find ways to avoid, modify, or adapt to them (Johnson, 2016).

Often an approach that utilizes both sensory input and behavior management techniques will work best. Remember, though, sensory input should never be removed as part of a behavior plan or provided as a reward. Sensory input should not be considered a treat and should not only be offered when a person is acting appropriately. In actuality, sensory input is necessary to help a person regulate her body and should be offered even in times when she is acting out. When a sensory diet is implemented as designed by an OT, input should not be withheld because of challenging behavior. Sometimes, you may not be able to differentiate between

sensory and behavior, and that is OK. It really should not matter. The important thing is to find the best solution!

In summary, everyone has sensory processing differences. When these differences impair a person's ability to function day to day, we may want to consider therapy options to help. When people have a regulated sensory system, they are better able to pay attention, follow directions, and perform at an optimal level. This is true for people both with and without Down syndrome.

Return to Table of Contents

Chapter 13

Social Skills

Jasmine, age twenty-two, had recently finished the transition program at her high school. She had learned many skills in her transition program, including social skills. However, there had been limited opportunities to practice those skills in a wide variety of community settings. One skill she had learned was expressing her concerns and needs. She used this skill very capably at home and at her physician's office. Unfortunately, however, Jasmine still had some trouble understanding when and where it was appropriate to express her needs. While bagging groceries at her new workplace, she made several comments about her personal health and hygiene needs. These were accurate expressions of her needs but were not appropriate to make while on the job. With some additional social skills training, Jasmine learned not to share information about her personal health when she was at work.

We are social beings and depend on others for our health, our well-being, and our very survival. Social skills is the term we use to describe our ability to effectively engage with and respond to others in our world. There are a number of key areas of social skills in both private and public settings. They include the ability

- to express and receive love and affection;
- to be socially sensitive to positive and negative feelings and emotions;
- to develop and maintain positive social roles in work and community settings;
- to use social skills in positive ways, not manipulatively to our detriment;
- to develop and maintain friendships in work, social, and recreation settings (addressed in chapter 8).

Expressing and Receiving Love and Affection

One of the most prevalent stereotypes about people with Down syndrome is that they are unusually loving and affectionate. For the most part, we find that this stereotype is true, but of course, not all people with Down syndrome are naturally

outgoing and affectionate, and some actively resist being touched by others, especially strangers.

Whether or not a particular individual is naturally inclined to be demonstrative or is merely assumed to enjoy expressing and receiving affection because of his diagnosis, he will face some unique issues. Most importantly, he will need to learn when, where, how, and with whom it is appropriate to express affection, as well as when, where, how, and with whom to turn away physical affection.

When a teen or adult with Down syndrome is physically demonstrative, the biggest concern is with expression of affection toward strangers or people who are not family or close friends. This makes sense. There are unscrupulous people who may take advantage of people with Down syndrome, just as there are people who take advantage of children. And even well-meaning strangers may make an individual with Down syndrome very uncomfortable if they mistakenly assume he loves hugs and intrude on his personal space. Families' concern for the safety and well-being of their sons and daughters with Down syndrome is justified, and the openness and affection shown by many people with Down syndrome no doubt intensify it. Conversely, the person with Down syndrome may hug an unfamiliar child or adults who does not welcome the hug and is offended or worse, interprets the action as sexually aggressive.

One of the most effective ways to help people with Down syndrome with safety issues is to capitalize on their strengths with concrete and visual images, for instance, by conducting safety training in role-play groups and then practicing skills in the settings where they will be used. There is a large and growing literature by knowledgeable authors like sexuality educator Terri Couwenhoven, who use visual and concrete learning resources to help people with Down syndrome to learn.

Lack of Love and Affection

Recently more attention has been paid to safety issues in both the general population and the field of disabilities. We believe this may be contributing to a different problem that we have encountered. The problem seems to occur most often when adults with Down syndrome move from their family home to a group home. At home, people with Down syndrome are usually free to give and receive love and affection. In group homes, however, staff are strongly discouraged from showing their clients physical affection due to growing concerns about sexual abuse.

We think that many adults feel a great sense of loss when they move to a more sterile or affectionless environment. An adult with Down syndrome may or may not understand why staff members are not allowed to show him physical affection. He may think, "This is my house and this is a person I care about and who cares for me. But he (she) never hugs me." The problem may be exacerbated if the adult takes longer to connect to people because of expressive language limitations. The lack of physical contact and speech difficulties may be major reasons why people have difficulties adapting to a group home as well as problems coping with turnover of certain staff members.

Dos and Don'ts for Safe Expression of Affection

The needs for individuals to express and receive physical affection, to be protected from sexual abuse and sexual predators, and to refrain from giving unwanted affection are very important. There is not an easy solution to address all these needs.

Some guidelines to help ensure safety for adults with Down syndrome include the following:

- Seek out programs designed to teach people with developmental disabilities to be street-savvy and security-conscious. Examples include Terri Couwenhoven's workshops, and programs such as "Circles," which may be offered by local ARCs, parent groups, or agencies serving the needs of people with intellectual disabilities. These programs can be quite helpful, particularly if what is learned in the program becomes a part of the adult's regular routine. For example, one man who was sometimes left alone in his house learned never to open the door to anyone, even when it was just a neighbor borrowing sugar.
- Teach the person which physical demonstrations of affection are appropriate. Remember that most people with Down syndrome are visual learners and often learn best from seeing the appropriate behavior. Role-play is a very effective way to teach these skills but is most effective if it occurs in the actual setting where they are needed rather than in a classroom setting.
- Bear in mind that although adults with Down syndrome often have a general understanding of appropriate physical touch, many situations may be confusing and seemingly contradictory. For example: Why is it OK to hug people at a wedding reception but not at a mall? Why is it OK to show affection to parents

and not to staff in a group residence, especially if these people behave much like parents? Because of these types of contradictory expectations, people with Down syndrome may need more specific guidelines about circumstances where it is appropriate to show affection. The general (abstract) concept needs to be filled in with more specific concrete examples. The individual may need to specifically identify each person he can show physical affection to (concrete) rather than just identifying a class of all people (abstract), such as all relatives.

Here are some guidelines to help adults with Down syndrome safely give and receive physical affection:

- If they move out of the family home, try to ensure that they still see family members regularly and have ample, appropriate opportunities to be physically affectionate with those family members.
- If the adult with Down syndrome enjoys dancing, encourage him to attend dances with friends where physical contact is part of the activity.
- Ensure that the adult has the support, opportunities, and privacy needed to show affection with a date or at get-togethers with a boy- or girlfriend. We find that such get-togethers infrequently involve sexual intimacy and the handholding and other forms of affection are very enriching.
- Caring for a pet can also provide a safe way for people with Down syndrome to give and receive affection and experience the rewards of taking responsibility for the well-being of another living thing.
- Taking care of others in need may be a very beneficial way for people with Down syndrome to share their caring and affectionate nature. It is important, however, to ensure that the person receiving care is not taking advantage of the individual with Down syndrome. By giving of themselves, people receive affection and thanks in return and develop a sense of accomplishment and pride from helping others.

While it is important to ensure the safety of teens and adults with Down syndrome, the solution cannot be the complete loss of physical contact. A hug, a pat on the shoulder, and other nonsexual acts of physical contact are very important for all

human beings. Continued efforts, monitoring, and working with compassionate, caring care providers are essential to maximizing emotional support and safety.

Social Sensitivity to Feelings and Emotions

People with Down syndrome often seem to be especially aware of and sensitive to their immediate social environment. Most are quite adept at reading certain social cues, especially picking up on the feelings and emotions of others around them. Because of this, people with Down syndrome have a reputation for having very good social skills. And in fact, many are friendly, social beings who relate well to others and are empathic and responsive to the feelings and emotions of others. Often family and caregivers describe them as wanting to please others. This may be due in part to a wish to avoid conflict and reduce any negative emotions, but it may also be due to a genuine sense of affection and caring for others.

Having good social skills and social sensitivity benefits people with Down syndrome in many ways. It can help them to make and maintain positive relationships at home with family, with friends or staff in social and recreation settings, with students and teachers in school and educational settings, and with bosses and fellow employees at work. This social sensitivity can also help them create goodwill with others, which can be useful if they make mistakes in relating to others, as discussed later in this chapter.

Unfortunately, there can be a major downside to this social sensitivity. We have found that people with Down syndrome may be too sensitive to negative feelings and emotions. Understandably, they may be quite sensitive to criticism directed at them, if only because this goes against their wish to please others. They may also be especially sensitive to feelings such as sadness, fear, or anxiety experienced or expressed by others. They may be more sensitive to these feelings in close family and friends but may also be affected by these emotions experienced by anyone in their environment, even a stranger. It appears that they may have difficulties blocking the effect of these feelings on themselves. In our experience, people with Down syndrome may be even more sensitive to anger than to other emotions. Again, this is true whether anger is directed at them or is directed at or between others. Many parents have reported that their sons and daughters get upset even when others just appear to be angry, such as when talking loudly with one other.

We believe their sensitivity is related to their superior intuitive and empathic ability to pick up on others' emotions. They have such good receptors that they may

even pick up on feelings that are not openly expressed by another person. Many caregivers have told us that their family member with Down syndrome knows what they are feeling before they do. Unfortunately, many people with Down syndrome seem unable to effectively manage negative feelings once expressed or experienced by others. They may be overwhelmed because they cannot block or fend off the feelings like others can. This may be due in part to expressive language limitations, which make it more difficult for them to get help from others to process or vent these feelings. Because of their reliance on concrete forms of thought, they may also simply not know the cause or source of the feeling or have difficulty understanding that others' emotions are separate from their own. Due to all these reasons, they may lack a good enough defensive structure to block negative feelings or emotions.

This inability to manage or fend off negative emotions in their environments may be one of the most underappreciated and yet significant causes of stress for people with Down syndrome. This source of stress may not always be considered by others. In fact, we have heard people state that individuals with Down syndrome don't have stress. (See "The Myth of Perpetual Happiness" in chapter 4.) While it is certainly true that they may not experience the type of pressure that many adults feel related to making a living, paying a mortgage or rent, etc., we have found that they may be at even greater risk for stress, particularly from this issue. There is some variability, and some may be more sensitive than others, but still, most people with Down syndrome have a heightened sensitivity to others' emotions. Sometimes the stress from exposure to strong negative emotions can be quite severe and debilitating, resulting in trauma, depression, anxiety, and an increase in obsessions or compulsions.

What to do about this sensitivity? First, it is important to recognize that this is part of the person's basic makeup. We cannot eliminate this sensitivity, and we don't want to. It does lead to many positive benefits in their relationships with others. Still, we must help people find ways to manage negative feelings because they will inevitably encounter these emotions throughout their lives.

We have had some success with providing people with Down syndrome several strategies to reduce the impact of negative emotions. These strategies can be boiled down to three approaches:

1. Help the person learn to recognize and label his emotions.
2. Role-play methods of dealing with negative emotions.
3. Teach the person to get away from the disturbing situation, if possible.

Recognize and Label Emotions. We believe that when people with Down syndrome experience a strong expression of negative emotions by another, this emotion in a sense "invades" their own body, senses, and feelings. They may experience this as physically and emotionally shocking, overwhelming, and even paralyzing. Often, they simply have no good way to understand or manage what is happening. Being able to put a name to the emotion, such as "anger" or "sadness," and, whenever possible, point to the source of this emotion may make it more manageable. This may sound too simple, but in effect what you are doing is turning the event into an abstract concept for the person with Down syndrome.

Once the person is able to name and categorize an overwhelming emotion, the event begins to have some meaning apart from his own feelings. The individual may then be able to see this as something that is both experienced in himself (as stress or tension from the emotion) but also separate from himself (emanating from another). This, of course, is not a simple process. It needs to be repeated over and over, but there are plenty of tools to help the person with Down syndrome with this. For more information on teaching about emotions, see the sections on "Communicating More Complex or Intense Thoughts and Feelings" and "Nonverbal Expression of Feelings" in chapter 7.

Use Role Play. Another particularly effective way to manage negative emotions is to role-play a situation involving those emotions. (See chapter 5 for more on role play.) This can be done either to prepare people for situations involving a particular emotion in the future or in response to a recent situation in which they encountered the emotion. The role play allows the participant to create a visual memory that he can easily access when a situation involving negative emotions arises, or even to replace the memory of a recent situation that was emotionally arousing. This strategy will probably be needed more often in settings that include others with intellectual disabilities because these people may have more trouble managing their own emotions.

Having a strategy to name, categorize, and practice ways to deal with an emotion in the environment does not mean that the person has to manage his emotions alone. In fact, once he can attach a name (e.g., "anger") and a source to the emotion (another person), he may be better able to ask for help with this problem. For example, a staff person, supervisor, or teacher may be able to help him manage the source or at least reduce the intensity of the negative emotions.

Get Away from the Stressful Setting. As discussed in chapter 4, teens and adults with Down syndrome may also be taught to leave or to distance themselves from a person or situation that is too emotionally intense or stressful for them. Sometimes this is impossible because it involves a work or school situation that the person cannot leave. In these cases, it may help to do an activity that allows him to focus on something other than the source of the stress. For example, he might be able to do a calming activity, such as copying letters or words or listening to music with earphones (if allowed in the setting).

Helping the Person Cope in the Moment

Parents are quite often superb observers of sons and daughters with Down syndrome, but any caregiver can be taught to recognize signs of emotional stress in a person in their care, even if he has difficulty verbalizing the cause or source of the emotions. Most people with Down syndrome cannot help showing their feelings through their facial expressions or gestures, which will alert caregivers who are present and attentive to the expression of emotions. How can you help when you notice that an adult with Down syndrome is having difficulty coping with a stressful emotion?

- Remember that the person may not know or cannot easily verbalize what is happening.
- Let him know that you notice he is having some type of emotional experience or response. For example, say, "I see that you are upset [feeling stressed]."
- Then, try to help him name the emotion. The best way may be to use picture images of emotions (see chapter 6).
- Try to identify the cause or source of the emotion in the individual's current environment. If the cause is not easily identifiable, there may be a number of other possible explanations:
    - The person may be too fearful to discuss the source, especially if the source is still in the environment. In this case, try to note behavior that might indicate the cause, such as avoiding certain people or places.
    - The source may have occurred in another environment earlier in the day, but the person is still experiencing the emotion. Naming the emotion will at least allow him to have some tools to deal with the emotion. He may also be able to better name and manage the emotion

in the future, especially if he is given a visual representation of the emotion to keep.
    - Something in the immediate environment may also have reminded the individual of a past event involving a negative emotion. (For more on this, see chapter 6.) In this case the best response is again to help him name the emotion and if possible, identify the trigger. The trigger may be very difficult to identify and discuss, though, because naming the cause may trigger the negative or traumatic memory again.
- Finally, whenever possible, have the individual role-play the situation and name the emotion. This will help him understand how he can identify and then respond to the emotional experience in the future.

Sensitivity to Effects of Behavior on Others

Although teens and adults with Down syndrome may be sensitive to others' emotions, they may not be so aware of their own impact on the feelings of others. People with Down syndrome are often refreshingly honest and direct, but this can sometimes lead to hurt feelings. For example, they may draw attention to a sensitive issue for another person, such as their height, weight, hairstyle, or glasses, or make other comments that are inadvertently hurtful. Now, we do have to state clearly here that the experience and familiarity of the person with Down syndrome with the person who is the object of the potentially hurtful comment will obviously affect whether it is seen as insensitive or hurtful.

Angela, a very social, friendly, and well-spoken woman with Down syndrome in her late forties, has been working in the same small office since age twenty-one. She is quite competent at completing her work responsibilities, including light word processing, filing, helping with mailings, and answering the phone. However, Angela has a long-standing habit of laughing at her coworkers' minor misfortunes. For instance, if someone trips, stubs their toe, spills coffee on their clothing, or drops an armload of papers, she almost always laughs. Over the years, her coworkers have often pointed out to Angela that these accidents are not funny. They have asked Angela if she likes it when people laugh at her when she falls or has another mishap, and Angela always admits that she

does not like it. Even though Angela's colleagues then point out that she should not laugh at other people because they do not like being laughed at either (and because it sometimes hurts), Angela still often laughs at her coworkers' misfortunes.

Unfortunately, people with Down syndrome may not learn to be accountable for their behavior, which can lead to difficulties in work or community settings. One way we recommend helping people learn appropriate skills is to practice "in the moment" learning, responding immediately in the environment in which the behavior occurs. This should be followed up with role play to help the person practice more appropriate or sensitive responses to a social situation. This is a powerful and effective way to help people see and understand what happened and help them find ways to correct their behavior.

For Angela, in-the-moment teaching might include, at the time of the incident, asking Angela to step out of the office. Then her coworker or supervisor could take these steps:

- Discuss whether she would like to have someone laugh at her in a similar situation.
- Find a picture of a simple injury or mishap (similar to the event of concern), show it to her, and discuss not laughing.
- Discuss an appropriate consequence for laughing inappropriately at coworkers.
- If these steps do not work, try using role play to practice a more appropriate reaction, such as saying, "Are you all right?"

Social Skills in Work and Community Settings

As discussed previously, many people with Down syndrome have a positive attitude and social demeanor that may shield them from some repercussions when they make social blunders. However, a positive attitude and demeanor may only go so far on the job. In fact, research studies have consistently found that what creates problems on the job for adults with developmental disabilities is a lack of social skills, not job skills (Greenspan & Shouts, 1981; Hill & Wehman, 1981). As noted by one group of researchers (Stainton et al., 2006), "Skills needed for positive non-task related social interactions with coworkers as well as skills needed to complete work

tasks are essential conditions of meeting the basic requirements for success in entry-level jobs." To succeed on the job, employees need to use appropriate social skills with the boss, other employees, and customers or the public.

Dealing with Supervisors and Coworkers

It is especially important for employees to use appropriate social skills with the boss. Having difficulties in this area is one of the primary reasons for job dismissal. For example, when dealing with an authority figure, such as a boss, one common problem is to misinterpret or take literally a "figure of speech" (an abstract notion) for a statement of fact. For example, the statement "my door is always open" is often used by bosses or managers to convey that they are accessible if there is a problem. But the person with Down syndrome may take such statements literally. (This is discussed in more detail in chapter 7 in the section on "Using Role Play to Teach Social-Language Skills.")

Additionally, some adults may not understand that coworkers besides their job coach or immediate supervisor may be in charge. The idea of multiple bosses may be difficult to grasp. For example, one young man working in a grocery store was almost fired when he told the store manager that he only took orders from his "boss" (his immediate supervisor). Still others may take orders from coworkers who are not in charge, as illustrated in this example:

> *In the midst of a bitter battle between union and management in a grocery store, Samantha was manipulated by angry employees to write down complaints she had about her boss. Some of her complaints were appropriate, such as about her boss's disorganization around weekly scheduling. Other complaints were true but should not have been expressed. For example, she described her boss as "grumpy," "yelling sometimes," etc. Unfortunately, Samantha showed a lack of common sense by delivering this list of complaints directly to her boss, much to the horror of her family and the employees who had put her up to this.*
>
> *Fortunately, Samantha's boss had a sense of humor and realized others had manipulated her. Additionally, the boss had enough knowledge of Samantha's needs that he called a meeting with the family, the job coach agency representative, and our staff. Interestingly, the job coach*

agency representative explained that Samantha did not have regular meetings with her job coach because she already knew her job. The agency representative steadfastly refused to accept responsibility for Samantha's social skills on the job. On hearing this, the family decided to contract with a new job coach agency. The new job coach met with Samantha regularly to help her work on social skills, and she has not had any additional problems since that time. Samantha continues to do her job as competently as ever.

Dealing with the Public or Customers

Like other employees, adults with Down syndrome are expected to interact politely and appropriately with the public or customers they encounter on the job. Sometimes, however, they lack the social skills to handle a disgruntled customer properly or do not know when it is best not to engage with someone. For example, Dr. McGuire's late wife, Elina, came home from work one day with an interesting look on her face. On the morning in question she had been late for a meeting at work and was in a grocery store getting a few food items for the meeting. She happened to have a young woman with Down syndrome bagging her food, and without thinking, because she was late, Elina grabbed an item and put it in the bag. The response by the woman was immediate. She looked up with a look of pain and disgust on her face and said, "You . . . people . . . are all the same. You are always . . . in a hurry." Elina stood there for a minute temporarily stunned. She truly understood where this woman was coming from and appreciated the sentiment, but the truth is that others might not. Although she was a psychologist, she did not feel it appropriate to intervene. In this case she was simple a customer.

It is hard to imagine that this woman's behavior, especially if repeated over time and with several other customers, would not have gotten the bagger dismissed from the job. As mentioned previously, the research is clear that social skill issues and not performance with job tasks is often when gets people fired.

Sometimes, too, people with Down syndrome can be less capable of defending themselves from the bad intentions and emotions of others, which can make them vulnerable on the job, as illustrated below:

Declan had a job busing tables in a restaurant. One day he just happened to meet "the couple from hell." The restaurant was very busy that night, and they came up to the table as Declan was finishing up cleaning their table. It seems he wasn't fast enough for them because they called him "slow" and made snide and insulting comments about his skill, speed, and intelligence. He continued to finish the job as best as he could, but as he walked away, he had had enough, and under his breath he mumbled "mean people." Unfortunately, they heard this and went to the manager of the restaurant, demanding that he be fired because of his "rude" behavior. The restaurant was one of a chain, and the couple followed their complaint up with a nasty letter to the home base. Fortunately, Declan had worked in the restaurant long enough and was too loved and too appreciated, so he was not let go, but he may have been with a less understanding manager.

In a similar situation, a young woman with Down syndrome almost lost her job as a mail clerk because she "talked back" to a woman in the office who chided her for being too slow in delivering the mail. (She actually said, "I am going as fast as I can.") The woman complained to the administrative office of the business, which told her direct manager to put her on probation. Fortunately, this young woman had a good job coach and was also well liked by the manager and the other employees and was able to keep her job.

Building Social Skills for Real-World Conflicts

The moral of the last few case stories is that whenever possible, people with Down syndrome should be taught social skills that will prepare them to deal with all kinds of situations and dangers when in the community or at work. As discussed in previous chapters, an excellent way to do this is through role play in which people enact scenes that may occur. This may include encountering members of the public who are irate, "toxic," or insensitive, but can also include people who may simply apply the same standards to adults with Down syndrome as they apply to others. In addition to role play, there are other visual strategies that can help prepare teens and adults with Down syndrome for new experiences or potential problems, including

Social Stories, personalized books, and video modeling. See chapter 6 on visual supports for more information.

In particular, it is important to identify types of situations or interactions that may cause teens and adults with Down syndrome to lose their cool (whether it be others' insults or comments, facial expressions, or particular actions) and then help them learn how to control their comments and anger. Like the people profiled above, they must learn to identify what to do (concrete behavior) when in a particular situation (abstract concept) that would usually provoke their anger.

It is important to remember that the reliance of people with Down syndrome on concrete thinking may make it difficult to generalize a skill from one setting to another. Thus, if the place this skill is learned is not the actual workplace, then it needs to be transferred to that site. For example, if a skill is learned in a social skills or safety training class, it needs to be practiced in the work environment to take hold in the real world. Of course, not all situations can be prepared for through role play or visual supports, but if at least people are prepared for others to be negative and not all "nice," they will hopefully respond more appropriately when confronted with unfriendly behavior.

Using Social Skills in Positive Ways

Some of the preceding examples may give the impression that perhaps people with Down syndrome are a little too honest or too innocent for our world. But as many families have told us, "They may have Down syndrome, but they are not dumb." Many know how to "play" people or wrap them around their little fingers. For example, when told by one parent that something may be harmful or not in their best interest (such as eating a bowl of ice cream every night), they may then go to the more indulgent parent. This is the more manipulative side of social skills.

Manipulation can occur in any context. For example, residential staff often report that people in their care gravitate to the person who will do things for them. This is not a good skill, as it undermines their own independence. Interestingly, many caregivers have told us about discovering that the person with Down syndrome has "hidden skills." For example, they may hear from relatives that their son or daughter is able to do many tasks they did not know the person could do. This may be because the person with Down syndrome does not object when these tasks are done for him at home, but others expect him to do it on his own.

Over the long term, allowing the person with Down syndrome to manipulate others to do things for him can lead to a loss of skills and an unwillingness to become more independent. He can learn to be helpless. As discussed in chapter 11, learned helplessness can lead to depression. Manipulation of others can also lead to problems with interpersonal relationships, as the people being manipulated may come to resent being manipulated and the feeling of being "used." The best way to guard against this is to ask the person to be responsible for his own life and activities to the extent possible.

There are some strategies to address a long-standing pattern of allowing an adult with Down syndrome to manipulate others:

- As discussed in multiple places in this book, using lists, calendars, and schedules, particularly of a visual nature, is helpful. Their use takes it out of the hands of the parent (or other individual) and puts it on the schedule and the person with Down syndrome. Rather than having endless discussions, the parent or other can direct the individual with Down syndrome to the calendar. This is much more likely to be successful if the person with Down syndrome participates in the development of the calendar and there is some discussion and agreement to the process. Use of rewards or "if /then" statements that result in the individual getting something he desires if he does the tasks as outlined on the calendar or schedule can be beneficial.
- Demonstrating by example can also be very helpful. "This is something we all do" or "something adults need to do" can be motivating.
- Individuals in the adult's life have to agree to work together. If one person is giving in and the other is promoting more independent use of skills, it is unlikely to be successful and will likely lead to behavioral issues or conflicts.

If one person generally refuses to let the adult do something that is not in his best interest, but another person usually allows it, it may help for everyone involved to discuss what is going on together. For example, perhaps Connor's father often agrees to take his son to a fast food restaurant for lunch after Connor's mother has refused. Meanwhile, everyone is aware that Connor is overweight and needs to learn healthier eating habits. The next time Connor asks his father to take him out to eat, Dad could bring Mom into the discussion. Together, the three of them could talk nonjudgmentally about the need for the whole family to make fewer trips to fast food restaurants—e.g., because everyone needs to make healthier choices, eat more

vegetables and less salt, and perhaps save money. They might then agree on the number of times per month they will eat out and even schedule them on a calendar. After the discussion, Connor should theoretically know that it will no longer work to appeal to Dad when Mom says "no," but Connor will probably test his father to be sure.

For a plan such as the above to work, the obliging parent (Dad) will have to hold firm and put up with repeated efforts from the adult with Down syndrome to get him to give in like he used to.

Developing and Maintaining Friendships

One of the most common concerns we hear expressed by caregivers is that people with Down syndrome tend to seek out conversation with staff rather than their peers with Down syndrome. The fact that this is so common should be somewhat of a relief to families. We believe the reason for this is that it is simply easier to communicate and to be heard by these others. As we discussed at some length in the preceding chapter, the connection to others with disabilities including others with Down syndrome may be there, but just not evident from the conversation patterns that occur.

Conclusion

Social skills are an important part of participating in society and interacting with family, friends, people we regularly encounter, and even strangers. Lack of social skills or impaired social skills is often interpreted as a behavioral problem for people with Down syndrome. The lack of these skills and the challenges created in daily life by not having them can lead to impaired self-esteem and even mental illness. Learning, relearning, and practicing social skills are important components of mental health promotion.

Return to Table of Contents

Chapter 14

Mental Illness and Its Precipitants

Janine, age twenty-two, became depressed. Her parents had worked very hard to get her into a work setting, but Janine did not like the job and felt trapped. She had a limited ability to communicate these feelings, however. With time, the problem became clear and her parents were able to find her a different job. The new job was very similar to the previous one, but Janine was much happier. Much of her happiness seemed to be related to her sense that she could have an impact on her environment.

Up to this point in the book, we have addressed a number of ways that mental health may be promoted and optimized in people with Down syndrome. We have pointed out that good mental health in adults with Down syndrome does not necessarily look the same as good mental health in others, due to common behaviors such as self-talk or the "groove" and differences in language and memory abilities. These differences are sometimes mistaken for signs of mental illnesses but are generally nothing to worry about if parents and professionals react to them appropriately.

On the other hand, sometimes adolescents and adults with Down syndrome are affected by mental illness. A mental disorder (mental illness) is defined in the *Diagnostic and Statistical Manual of Mental Disorders*, 5th edition, as

- "…a syndrome characterized by clinically significant disturbance in an individual's cognition, emotion regulation, or behavior
- that reflects a dysfunction in the psychological, biological, or developmental processes underlying mental functioning."

According to the DSM, mental disorders "are usually associated with significant distress or disability in social, occupational, or other important activities." An expected or culturally approved response to a common stressor is not a mental disorder. For example, grieving is not a mental disorder.

Some types of mental illnesses include the following:

- mood disorders (disorders in which disturbance in mood is the predominate feature) such as depression, bipolar disorder, dysthymic disorder, and mood disorders due to a general medical condition;
- conduct disorder and oppositional defiant disorder;
- schizophrenia and other psychotic disorders (disorders in which the symptoms of delusions or hallucinations are the primary features);
- anxiety disorders, obsessive-compulsive disorder, and post-traumatic stress disorder;
- attention-deficit disorder;
- impulse control disorder (failure to resist an impulse, drive, or temptation to perform an act that is harmful to the person or others and is not caused by another mental illness).

Mental illnesses are fairly common in adults without Down syndrome. The National Alliance on Mental Illness (2018) reports the following statistics:

- Annually 43.8 million adults in the United States—18.5 percent (nearly one in five)—experiences mental illness. Each year, 9.8 million adults in America (about one in twenty-five, or 4 percent) experiences a serious mental illness—defined as one that "substantially interferes with or limits one or more major life activities."
- In the past year, sixteen million adults in the US (6.9 percent) experienced at least one major depressive episode.

According to the National Institute of Mental Health (2017), neuropsychiatric disorders are the leading cause of disability in the United States. However, most mental disorders go untreated. In 2016, only 43.1% of adults with any mental illness received mental health treatment.

In people with Down syndrome, one researcher (Mantry, 2008) reported a prevalence of mental illness of any type, excluding specific phobias, of 23.7%, and another researcher (Vicari, 2013) reported 18 to 23 percent. Twenty-three percent is higher than the number cited above by the National Alliance on Mental Illness for the US population as a whole. Although mental illness is higher in people with Down syndrome than the population as a whole, the first researcher reported that the prevalence is lower in people with Down syndrome than in those with other intellectual disabilities. In addition, the study reported that for people with Down

syndrome, the highest prevalence was for depressive episode (5.2 percent) and dementia/delirium (5.2 percent).

Precipitants to Mental Illness

In general, there are two ways that mental illness may be precipitated in anyone: either through physical causes, such as biochemical or structural differences in the brain or an illness, or through what laymen usually refer to as "stress" ("stressors"), or a combination of these factors.

In chapter 16, we explain some of the structural and chemical differences in the brains of people with Down syndrome that can make them more susceptible to some mental illnesses. For example, differences in the transport of serotonin may make people with Down syndrome more likely to have symptoms of depression. In chapter 2, we discuss many of the physical problems that can lead to mental health issues in adults with Down syndrome. In this chapter, we will focus on the stresses that can sometimes lead to mental illness in people with Down syndrome.

Stress

It is interesting to note that hypertension (high blood pressure) is uncommon in people with Down syndrome. When we share this finding with others, frequently their response is "that's probably because people with Down syndrome have no stress in their lives." This is a myth. Not only do people with Down syndrome have stress in their lives, but they perceive it and it can affect them. Stress is in the eye of the beholder. In other words, because what you perceive as stress is not stressful to me, it doesn't negate that it is stressful for you.

In addition, individuals with Down syndrome have shared that they experience stress. Some are able to tell us with words, some with actions, and some with a change in behavior. Terms used to describe stress have included "feeling edgy" and "overwhelmed." Individuals have also described stress as "confusion," "frustration," "having too many pictures in the head," and "pulling your brains out," Stress is there and comes from many different sources.

There are clearly some challenges in life that people with Down syndrome don't usually have to deal with. For example, it is rare for an adult with Down syndrome to worry about how she is going to afford housing. However, this is not to say that stress from other sources is not in their lives. Sometimes adults with Down

syndrome are expected to deal with challenges that other adults are not. For example, many of our patients live in residences in which there may be several other people who have undesirable habits, who are awake at night, or whom they just don't like. However, they may have very little room to negotiate a change or to live with whom they would like. Furthermore, the intellectual disability secondary to Down syndrome may limit the person's ability to cope with stress. Therefore, at times an adult with Down syndrome may experience just as much, if not more, stress as anyone else, but may have less ability to cope with the stress.

The presence of stress in someone's life usually does not trigger a mental illness. Receiving appropriate support, removing oneself from the situation, participating in activities that reduce stress, and other strategies can all help prevent the development of mental illness. However, it is often necessary to first recognize the stress in order to actively develop strategies to deal with it. Adults with Down syndrome may not recognize their own stress or may not be able to verbalize their concerns. Care must be taken not to downplay stress in a person's life simply because she doesn't verbalize a concern. It is important to recognize that the person with Down syndrome may be experiencing stress and may need assistance to develop healthy strategies.

Common stresses that can precipitate mental illness in adults with Down syndrome include

- learned helplessness,
- lack of opportunity,
- lack of respect,
- expected stressful events,
- unexpected stressful events, and
- grief.

Common Sources of Stress

Learned Helplessness

One particular precipitant to mental illness occurs when a person senses that she has no ability to affect her environment. This is similar to the phenomenon seen in some infants called "learned helplessness." If an infant repeatedly cries and no one

responds, eventually she learns that it is hopeless and stops crying. Essentially, she gives up. She has learned to become helpless.

We have seen a number of adults with Down syndrome who were frustrated or unhappy with their situation. After repeated, unsuccessful attempts to communicate their concerns or have their concerns taken seriously, they developed a sense of despair and gave up. Often, they not only gave up trying to make that particular change but also became apathetic in general, as well as depressed.

Sometimes a person's limited language abilities keep her from expressing her concerns understandably. Other times, the people who hear the concern may not take her seriously or may see her concerns as unimportant. Limited resources may also be a challenge. For example, if the person does not like her group home, but there are no other choices, no change is likely to occur.

For an environment to be supportive, it must be responsive, as in Janine's case at the beginning of the chapter. Listening is the first step in providing support or care. Without understanding the problem, one cannot offer a truly beneficial solution. Clearly, it can be a major challenge to understand the needs of a person with Down syndrome who has minimal communication skills. While this book outlines a number of issues that are common to people with Down syndrome, it also points out that each person with Down syndrome is an individual and has unique needs, desires, and hopes. When someone expresses these needs but is not listened to, it can be very frustrating and eventually cause despair.

Trying to understand individuals from their point of view is a key element in diagnosing and assisting teens and adults with Down syndrome. One method that is helpful is to think of comparable situations in our own lives and how we might react. The frustration of not being heard is seen in this simple analogy. If you want to understand how frustrating it must be to have people not listen, imagine entering a department store with a nice shirt that you recently purchased. After waiting in a lengthy line at Customer Service, you reach the front and place the shirt on the counter. Before you have a chance to say anything, the clerk smiles, takes the shirt, and hands you your money back for the returned item. You are pleased with the clerk's politeness, cheerfulness, and eagerness to help. On the other hand, as you see your shirt being carried away to be returned to the shelf, you become frustrated. The purpose of your visit to the store (that you didn't get to voice to the clerk) was to get the security tag removed because it had not been removed when you purchased it. You really liked the shirt, and now it is being taken away. The service was great; it

just wasn't what you needed. This type of occurrence can be quite frustrating, especially if it happens day after day, multiple times a day.

Many individuals with Down syndrome have shared analogous events in their lives that led to frustration and could eventually cause them to stop trying. Anyone who has her attempts to change or affect her environment ignored or misinterpreted can become frustrated. If this happens recurrently, it can be a significant precipitant to mental illness.

Lack of Opportunity

Lack of opportunity can be very stressful, frustrating, and challenging to self-esteem (see chapter 8). Opportunity is not just something to do or something that fills our time. It is an interesting challenge that allows us to feel creative and excited. For some adults with Down syndrome, the tasks or jobs available do not begin to fill these criteria. For some, there may be no tasks or jobs available at all, due to lack of funding and other reasons.

The individual determines how fulfilling any given task or job is. Therefore, what may seem boring or unfulfilling to you may be quite fulfilling for someone with Down syndrome. The opposite is also true. Unfortunately, many adults with Down syndrome have little input into the work they do, and this can lead to frustration.

Living situations can also be a source of frustration or stress. Many people with Down syndrome have limited choices in residential options. Funding issues, the need to rapidly find a home after the death of parents, and other issues all contribute to lack of available and appropriate living situations. Certainly, it is ideal if families can investigate opportunities well in advance, but this does not always occur. In addition, there are often fluctuating funding issues. A well-made plan can become problematic at the time of need because of a change in funding by the state or other provider.

Many adults with Down syndrome continue to live in their family home with their parents and/or siblings and it works very well. Many individuals move to group living arrangements and that works very well for them. However, sometimes when someone wants to move out of her family home, no opportunities are available. Others have moved into group facilities and the fit is wrong. Once again, individual assessment and planning is important to optimize the situation and minimize stress.

Inclusion and Choice

For a number of years, the educational philosophy has been moving toward teaching students in the "least restrictive environment." The goal is to expand full inclusion in schools so that people with intellectual disabilities can attend classes with typically developing students and have as much access as possible to the regular curriculum. Inclusion is increasingly being recognized as a goal for adults with intellectual disabilities as well. To be included in our society, adults should be able to participate in activities that appeal to them. One of the highest hallmarks of participation in our society is having choice. Unfortunately, many adults with Down syndrome are not truly included because they do not have choices. Often, financial/funding issues or society (or professionals) limit them because only one way for individuals with Down syndrome to do something is recognized. Fortunately, as in schools, over time we have heard and seen a great number of individuals with Down syndrome have the opportunity and successfully participate in a more inclusive way in society in a variety of ways.

Even when someone is living in a fine agency-supported home, there can be many stresses. While many adults enjoyed living in a dorm in college, there are issues that would make living there forever a challenge. Noise issues, tolerating others' habits, and compromising to meet the needs of all individuals can all present problems. Although these are issues in any family, sometimes a group facility is more akin to a college dorm, with a larger number of individuals who make it more challenging. In addition, difficulties with language and the ability to express concerns (see chapter 7) can limit the ability to deal with these problems. In addition, some individuals with Down syndrome are quite sensitive to the behavior or actions of others and can compound the challenge.

To address these problems, the challenge for the family or staff supporting people with Down syndrome is to make the living situation the "home" of the person with Down syndrome. This is important for family members to remember and acknowledge. When a person with Down syndrome goes to live with a brother or sister, the family also faces the challenge of making it the home of the person with Down syndrome. Staff members at group homes have the same challenge.

For some adults with Down syndrome, lack of opportunity may also be a problem in the areas of education, recreation, and travel. As in the other areas, the first step is to recognize that these may be areas of stress for the person with Down

syndrome. Once the problem is recognized, the adult needs encouragement to continue to find and develop opportunities to optimize participation in these areas.

The Desire for More Inclusive Job Opportunities

"I don't want to be with people with developmental disabilities."

When an individual we are serving shares this sentiment, it can be a concerning statement. Typically, the person sharing it has good communication skills (and thus is able to verbalize this preference). The message behind the statement can have many meanings. As we have assessed the concern through further conversation, we have teased out some of these concerns:

- Some people have had a wonderful, inclusive school experience, and as they leave the school system and move into the working world, they assess (often correctly) that the opportunities available to them in the "developmental disability" world are not as attractive as those of their siblings and nondisabled peers. We anticipate as more individuals with Down syndrome have an improved school experience that this will be a bigger issue and the employment opportunities will need to "catch up" to the level of school opportunities to meet the needs and expectations of individuals with Down syndrome who are better educated and more experienced.
- Some of the individuals around them with intellectual or developmental disabilities are "less predictable." A number of adults with Down syndrome have expressed a desire to work with individuals without a "developmental disability" because of what they perceive to be unpredictable or erratic behavior of some people with intellectual disabilities.

One challenge to eliminating all others with an intellectual disability from someone's life is that friends and family without an intellectual disability often move on to other aspects of their lives (school, work, marriage, etc.) and the person with Down syndrome may feel left behind. We generally encourage a blend, if possible, of friends and coworkers with and without disabilities. We have often seen that having a variety of friends, with and without a disability, can lessen the impact of someone leaving. On the other hand, we do need to be cognizant of the stress that some may be sharing and help them understand and cope (manage their own stress) so that they have the opportunity to be more comfortable with a greater variety of individuals in the community.

Lack of Respect

Unfortunately, another source of stress for people with Down syndrome is dealing with people who do not respect them. It is stressful to deal with people (both children and adults) who make derogatory comments or call them hurtful names. Unkind or unscrupulous individuals can be very hurtful. Even kind or well-meaning people can cause discomfort by not recognizing or appreciating the skills of a person with Down syndrome.

> *Bill, age twenty-seven, loves his job at the grocery store. However, the customers sometimes upset him. It is easy to see how the rude customers who call him "retard" are a real stress in his life. The other customers who are a challenge for Bill are the ones who assist him with the bagging. Bill takes pride in his work and interprets this assistance as indicating that people do not feel he is capable of doing his job. Despite all the benefits Bill gets from his job, not the least of which is improved self-esteem, his daily interactions with customers could be precipitants to mental illness. Bill has opportunities to discuss his day with the staff of his home each evening after dinner. He receives support and then participates in activities that he finds relaxing. He continues to do very well despite the challenges at work.*

Here are some ways you can help an adolescent or adult with Down syndrome deal with perceived or real disrespect:

- Have her ask herself why the person might be acting that way. Help her determine whether the person means to be disrespectful or whether she is merely perceiving it as disrespect.
- Help her understand that the person who is purposefully being disrespectful is the one with the problem. Encourage her to ignore it as best as possible or to address (or have someone else address) the person who is being disrespectful if it is a situation that can be changed.
- Encourage her to talk about episodes in which she feels she has been treated disrespectfully. Acknowledge and affirm her feelings and support her in dealing

with them. Encourage and support her in the development of self-esteem in other ways (see chapter 8).

- Help her develop strategies to deal with how she feels. These may include relaxation techniques, physical exercise, and other strategies.
- As explained in other sections in the book, use of pictures or videos and art therapy may help the individual communicate concerns and learn ways to manage them.

Expected but Stressful Events

There is little doubt that the environment and the passage of time will present situations that can be stressful. Some of the events and the timing are certain or to be expected, such as graduating from school and entering the work environment. Other events are less predictable. An illness, the death of a family member, divorce, and other life events are all relatively unplanned. Preparation is helpful for the person with Down syndrome to deal with both expected and unexpected events.

Remember, change can be difficult for many people with Down syndrome. A particularly difficult event is when a sibling moves out of the house, leaves for college, or gets married. These changes tend to affect the person with Down syndrome in two ways. First, she experiences the sibling's move out of the family home as a loss and goes through a grieving process. Second, she often senses that these are normal events that she will never participate in. For instance, Joan, after becoming upset at her sister's wedding, said, "I'll never get married and have children." As a person with Down syndrome enters adolescence and adulthood, the differences between her life and the lives of siblings or nondisabled peers can become more evident.

The grief of the loss experienced when a sibling moves should not be minimized. It certainly is not minimized in the minds and hearts of many people with Down syndrome. As outlined below in the section on Grief, there are a number of ways to assist people with Down syndrome in this process. Obviously, a significant difference between a family member moving and a family member dying is that when the person has moved out of the home, physical contact with him or her is still possible. Involvement of the family member in the life of the person with Down syndrome helps with the loss. While in-person contact is usually ideal, many families have found that using Skype, FaceTime, or visual communication methods to stay in

touch is more beneficial just talking on the phone. In addition, people with Down syndrome can use many other ways to connect, such as texting, social media, and others.

Consistency and regularity are essential in this contact. There is no question that people with Down syndrome are generally hurt by unfulfilled promises (for example, when a family member misses a scheduled event). In addition, the tendency toward order and repetition (see chapter 10) make irregular visits particularly difficult.

Improving the availability of opportunities and inclusion in society are clearly part of the solution to concerns related to having fewer opportunities. However, many people with Down syndrome have challenges that prevent their life activities from being just like those of siblings or peers. Accentuating the positive aspects of the person's life, seeking opportunities that mirror the opportunities of siblings or peers, and strengthening self-esteem and social skills can all help to minimize these challenges.

Accentuate the Positive

We all have some strengths and some limitations, whether we have a diagnosed disability or not. With the encouragement of family and others, each of us can recognize our strengths and focus on them, and we can find meaningful activities we enjoy. The same is true for people with Down syndrome. With a little effort, many individuals with Down syndrome discover untapped abilities. Artistic talent, musical flair, incredible memory, organizational skills, and others are all gifts that can be real assets. These strengths that can be part of a very fulfilling life are discussed further in several chapters in Section 2 of the book. Assessing, guiding, teaching, and, particularly, modeling, the use of talents can lead to great satisfaction for people with Down syndrome.

Seek Similar Opportunities

Seeking opportunities that mirror the opportunities of others can be very beneficial. We know several adults with Down syndrome who have expressed interest in being able to drive a car, get married, and/or move out of the family home and then have accomplished those goals. As people with Down syndrome become further included in society, opportunities to achieve these sorts of goals are increasing.

However, not all people with Down syndrome can realistically be expected to participate in all opportunities. Again, assessing interests and capabilities, guiding the person to alternative opportunities, and then teaching and modeling them can lead to life satisfaction and self-esteem and acceptance of the changes occurring in life.

Darryl, age eighteen, was frustrated about not being able to drive. He had participated in driver's education but did not have the manual skills or judgment to drive. As his family tried to help him understand these challenges, they discovered that the lack of independent travel was a significant part of his frustration, so they worked on teaching him how to use public transportation. Darryl has found it particularly satisfying that his family no longer drives everywhere but sometimes uses public transportation instead. When his older brother comes home from college and they go to a movie, museum, or other outing, they usually take public transportation. Darryl takes the lead role in use of the public transportation because his older brother is not as familiar with the system.

Although a lack of skills or opportunities is a challenge to overcome, promoting healthy self-esteem is still ultimately the goal as changes in life occur. Further information on promoting self-esteem is discussed in chapter 8.

Preparing for Events

Calendars and schedules are very helpful in allowing the person with Down syndrome to prepare for events that are planned ahead of time. As discussed in chapter 6, teens and adults with Down syndrome often have strong visual memories, so visual cues are often more beneficial than verbal. Therefore, using a calendar or schedule with pictures or a video is very useful. We have often found that a photograph or video of the upcoming event works best, especially if it shows the person herself participating in the activity.

Vanessa, age twenty-eight, had severe atlantoaxial instability. She required traction for several weeks before the surgery to optimize the

alignment of her neck. This was really a challenge for her to bear. A picture that signified surgery was placed on her calendar on the date of her upcoming surgery. This helped her deal with the time in traction by improving her understanding that there was a concrete end date to the use of traction.

In addition, advance warning is often necessary. On a day-to-day basis, giving a warning before transitions is generally a good idea. For example, it can be helpful to tell the person five or ten minutes in advance that it is almost time to stop work and get ready to go home. Giving the warning too far ahead is often not helpful because the person forgets or becomes re-involved with the job. Too little or no warning does not allow her to prepare for the transition.

Warnings for larger transitions, such as a move or changes in family, are also appropriate. A picture that signifies the event can be placed on the day on the calendar. Perhaps a picture of the adult with Down syndrome standing in front of her new home would work well or a video depicting the new house. Pointing to the date and counting off the days as they go by may also be helpful. Again, giving a warning too soon can be problematic (although how soon is "too soon" varies from person to person).

Often, parents or others delay telling the person with Down syndrome about an upcoming change in hopes of preventing her from perseverating about it. Again, the timing depends on the individual. Frequently, however, she learns about the change anyway by overhearing conversations. In addition, many people with Down syndrome have a much greater sense of upcoming events or change than is assumed. They have a radar detection skill that helps them perceive changes in others' demeanors or in events or activities. In those situations, it is better to have an open discussion and to help the person prepare for the transition rather than to delay telling her. Otherwise, she may jump to the wrong conclusion about what is going to happen or why, which may cause more worry. She may also come to the correct conclusion, but, because she is "not supposed to know," she may not get an opportunity to address her feelings about the situation.

Many times we have heard that a patient's behavioral change or alteration in mental health began some time before an upcoming event. "She hadn't even been told about the change yet when her depression started." She was apparently already aware, however, and was struggling with the change. Always consider the incredible

ability of many people with Down syndrome to pick up on events they have not been told about.

Unexpected Stressful Events

If an event is unplanned or unexpected, clearly many of the recommendations outlined in the section on planned or expected events are not applicable. However, there are still a number of similarities.

Many unexpected events, such as a death, illness, or divorce involve a sense of loss. The death of a family member, friend, or care provider can be particularly challenging; the grieving process is therefore discussed in some depth in the next section. Many of the strategies used to support a person with Down syndrome through the grieving process can be used for other unexpected events.

A key principal is to assess where the person is and help her from there. Ask what she knows or understands about the loss first. This can eliminate a great deal of confusion as to how to help. If the person is nonverbal, strategies discussed in chapter 7 may be helpful.

Loss is a form of transition, and transition can be difficult for many people with Down syndrome. If possible, warn the person that the transition is coming and then let her proceed at her own pace. If you carefully observe her response—whether spoken or conveyed through her body language—you can avoid overwhelming her with information. Share what she is ready for and able to grasp and come back as the need and readiness present themselves.

When the loss does not involve death, there are often continued opportunities for the person with Down syndrome to interact with the individual(s) undergoing changes. It is once again essential to assess the person's ability to deal with the changes. A question often arises as to whether someone with Down syndrome should visit a sick family member. We generally encourage regular visits. Rarely, the changes in the ill person can become too disturbing for the adult or teen with Down syndrome. In these situations, we advise that the person with Down syndrome reduce or eliminate visits.

It can be helpful to provide pictures or videos of the person with Down syndrome together with the person (or people) that she appears to be losing due to illness or other events. The strong memory that so many people with Down syndrome possess can be a blessing here. The pictures help them remember happier times, which can be a real comfort.

A calendar or schedule, especially one with pictures, can help adults with Down syndrome deal with the aspects of a loss that can be anticipated. For example, if treatment start and stop days or a surgery date can be placed on the calendar, it can provide a time frame or some sense of order.

Similarly, if the person with Down syndrome becomes ill or needs surgery, as discussed previously, a calendar can be used. Adding structure or increasing the predictability as much as possible helps the person with Down syndrome deal with the illness. It is important to explain the treatment or procedures at a level that is understandable. For example, for patients who are anxious about coming to a medical office, a book with pictures and words or a video that walks a person through an office visit can be helpful. The aids could include depictions of history taking, examination, blood drawing, and other aspects of the appointment. This information would be helpful to review before the appointment.

There are many changes or losses in life that can challenge adults with Down syndrome. Helping them put these losses in a structure and optimizing predictability is usually helpful. Using strengths in memory and visual learning skills to help a person with Down syndrome deal with loss can improve the benefit of the intervention.

It is important to realize that even good change is still change. Good news such as a job promotion, an invitation to go on a trip with a friend, or the birth of a new niece or nephew can all be very positive events in the life of a person with Down syndrome. However, they can also cause stress. Using the strategies outlined above can help prevent these positive events from becoming negative events.

Dealing with Divorce

Divorce has aspects of both unexpected and expected events. Usually it is not something that is suddenly sprung on the person with Down syndrome. Often, even if she hasn't been told of problems, she is aware of them. Therefore, in preparation for divorce, here are some helpful tips:

- Reassure the person with Down syndrome that it is not her fault.
- Reassure her that her parents still love her.
- Reassure her that she will continue to have a home, will be taken care of, and will see both of her parents.

Don't put her in the middle of the two parents. Don't put her in a situation where she is "turned against" one parent or the other.

- Maintain her schedule as much as possible.
- Give her permission to discuss her concerns and opportunities to do so.
- Encourage activities that help to reduce stress.

Grief

Grieving is a very individual process. People grieve in a variety of ways. However, there are some common features that we have seen in teens and adults with Down syndrome. First, people with Down syndrome often have a delayed grief response. Second, they need to grieve in their own way and time. And third, their strong memories often complicate the grieving process. Difficult or prolonged grieving can sometimes lead to mental illnesses such as depression.

Delayed Grief Process

The grieving process may be delayed in people with Down syndrome. It is not uncommon for an individual with Down syndrome to do well for three to six months or more after the death of a loved one and then to start expressing her grief. Sometimes an adolescent or adult with Down syndrome will do fine for a long time, and then another loss will occur that triggers the grief response. For example, a very significant person (such as a parent) will die and our patient seems to do fine. Then, months or sometimes years later, what appears to be a less significant death occurs (for example, a distant relative or the turtle at the group home dies), and then the individual expresses grief about the previous, more significant death. We have seen several such instances in which an adolescent or adult with Down syndrome suddenly started talking about the loss of the significant person in her life after several years.

There are many reasons a person with Down syndrome may have delayed grieving. The person may not have been given the opportunity to grieve, may take longer to sort through her feelings and emotions, and may not have had the skills to understand the process and how to go through it. In addition, we have often wondered about the ability of many people with Down syndrome to comprehend the concept of time. Sometimes, in describing the death of a family member, an adolescent or adult with Down syndrome will talk as if it happened in the last few weeks when it actually happened many years before. We have seen this difficulty with time related to other issues as well. Therefore, what may appear to others as an

unusual time to be grieving may be connected to differences in how time is perceived (see chapter 4). In addition, some individuals seem to take more time to understand the permanence of death and initially don't seem to comprehend the difference between death and a long trip or other prolonged separation.

Joel, age twenty-six, was quite healthy when he came to see us. His parents had recently updated their will and had decided to purchase their cemetery plots. They were relatively young and healthy, and there was no immediate anticipated use of the plots. While they were buying their plots, they had also purchased a plot for Joel. He had become very upset and eventually required antidepressant medication because he was so frightened by the message he perceived in this purchase of a plot for him. He had great difficulty grasping that the purchase was for a later, undetermined time. He was full of fear that he was going to die in the near future despite being healthy. Planning for the future was a concept beyond his skill level.

If Joel had been given a clearer explanation before the plots were purchased, he might not have had this reaction. On the other hand, if his parents could have predicted how he would respond to the news, given his response to other, similar events, a better approach might have been not telling Joel at all.

Memories and Grief

People with Down syndrome often have incredible memory capability. This ability can serve them well but can also cause problems if they remember painful times. For many people without Down syndrome, the grief process involves the easing or forgetting of the pain while remembering the person. However, the intense memory of many people with Down syndrome can make this process difficult. Using strategies that help the person with Down syndrome remember happy times with the deceased can be helpful. This is discussed in more detail in chapters 5 and 6.

The Right Time to Grieve

In helping a person with Down syndrome through the grieving process, it is generally best to provide assistance at the time she is ready for the help. We have

often found that having a person attend a "Grief Group" at a designated time can be troubling for many individuals with Down syndrome. At 4:00 on Tuesday afternoons, the person may not be ready to discuss the death of her mother. In fact, attending the group at that time may actually bring back painful memories and cause her to perseverate on the pain. Scheduled grief groups may work for some, but for many people with Down syndrome, they just bring back the pain that they had been able to set aside for a time.

Often it is more helpful, although more challenging, to allow a person to work through her grief when she is ready and interested. When everyone is dressed and heading out the door for an event and she brings up the recent death of her mother, it may not seem like the most convenient time; however, it is likely to be the most successful time. We recommend letting the person with Down syndrome guide the discussion: in her time, in her way, in her place.

When the person is interested in talking about the deceased, it is important to allow that opportunity. Particularly if the individual's network of family and friends is small, talking about a departed loved one may help her feel connected to that person through memories.

Literal Thinking and Grief

As discussed in chapter 5, many people with Down syndrome seem to be concrete and literal in their thinking. This concrete thinking can help them to get through their day, to succeed at their jobs, and to make sense of their world. However, some degree of abstract thinking is also helpful in many situations, and difficulties with that ability can be problematic, especially if others do not recognize those difficulties.

> *Scott was thirty-four when his mother died. He was placed in a grief group at his group home. The group was called the "Mourning Group" and met every other Wednesday afternoon. We were never able to figure out if it would have been helpful to Scott. He got so locked into the fact that the group met in the afternoon despite being called the "morning" group that he would not participate.*

Helping the Grieving Person

Here are some additional methods to assist with grieving:

- Spend time with the person.
- If she is open to it, talk about the death and the person who died.
- Share feelings.
- Encourage the person to attend the visitation/wake and the funeral or memorial service.
- Prevent other losses or transitions, if possible, at this time.
- Allow the person to make choices (about how she needs to grieve).

Charlene Luchterhand (1998), the author of a pamphlet on helping people with intellectual disabilities cope with death, offered the following advice when teaching about death:

- Use simple words and avoid words that have more than one meaning (e.g., "fell asleep").
- Teach by using examples in everyday life (such as the death of an animal or a famous person).
- Use many examples over time.
- Allow the person to see how you deal with losses in your life.
- Allow her to show emotion.
- Encourage questions.
- Talk about stages of life: birth, childhood, teen years, adulthood, aging, death, etc.
- Identify someone who can lead a class or group to talk about death and grief and then ask the person if she wants to attend, assuming it's the right time for her.
- Describe what is good about death: the person will no longer suffer; he or she will move on to an afterlife (if that is part of her religious beliefs). Don't, however, introduce religious concepts that she's not familiar with.
- Help the person feel safe now. Reassure her that she and other family members are healthy (in a truthful way).

Based on our experience with grief groups, we do advise determining whether participating in a group would most likely be helpful or problematic for the adult with Down syndrome. Does the person tend to perseverate on other issues? If someone else mentions the death of her loved one, does it seem to help her or cause her more problems? Has she previously done well in structured groups? Trying to answer these questions may give some insight into whether a grief group would be beneficial. If you are uncertain, doing a trial in a grief group for people with Down syndrome or other intellectual disabilities may answer the question one way or the other.

Another strategy we have found very helpful is making a video or a book with pictures of the deceased. It is particularly helpful to include pictures of the person doing enjoyable activities that are good memories for the person with Down syndrome. It is also generally helpful to use pictures that show the deceased and the person with Down syndrome together. The goal is to help the person ease and forget her pain while remembering the person and focusing on the happy memories. As long as you are very careful not to trivialize the grief or to "sweep the pain under the rug," it can be very positive to redirect the grief toward happy memories. We call this strategy "acknowledge, empathize, and redirect." Redirecting can particularly be of benefit when the person is "stuck" or perseverating. However, redirecting without acknowledging and empathizing often becomes dismissive and not helpful to the grieving process.

Avoiding other losses is only partly under our control. Obviously, we do not have complete control over many losses in our lives. However, we do recommend avoiding or delaying changes or losses, if possible, such as moving, or job changes, during the time of the most intense grieving.

Even the fear of another loss can be very disturbing to someone with Down syndrome. This is particularly true of the fear that others close to her will die. We have often observed this concern in people with Down syndrome who feared losing another parent, a sibling, or other person who is close. Reassuring her and remaining in close contact can help diminish these concerns.

Sometimes grief goes on for long time, severely interrupts a person's life, and can lead to depression. It is imperative to try to help the person work through her grief as outlined here, but if the grief becomes prolonged and appears to be more than just a grief reaction, further assessment and treatment may be necessary. Assessment for mental illness is discussed in the next chapter.

Remember, grieving is a unique process for each person. What is beneficial for one person may not be for others. The aspects of grieving that are common in the

people we have seen are certainly not universal. Often someone who knew the person with Down syndrome before the loss can best determine how to help her grieve. In addition, careful observation with an open, compassionate heart and a willingness to respond when she asks for help are keys to assisting the person with Down syndrome.

While everyone may grieve differently, it can be useful to learn how other people have helped a person with Down syndrome deal with grief. In response to an article on grief in our newsletter several years ago, Sheila Hebein, the (now former) Executive Director of the National Association for Down Syndrome, sent us the following letter:

My son, Chris, has had many losses to deal with in his life. His first one came when my father died, but Chris was only 5 at the time. Then he lost his paternal grandmother (Nona) when he was 11. He had been extremely close to her, but we were open with him and answered his questions as honestly as we could. He was at the wake with the rest of the family. He also participated in the liturgy by taking up the gifts with two of his cousins. Since he has been an adult, his paternal grandfather died and again, he was very close to him because grandpa lived with us on and off for several years. Chris was 19 at the time and we involved him in every way we could. He was with the whole family throughout the wake—I believe he was the only grandchild who didn't leave to get something to eat. He would go back to the casket and touch his grandpa's hand and kneel and pray. He was a pallbearer along with his cousins and we talked frequently about grandpa at home.

Four years ago, my mom died in England and Chris wasn't able to attend the funeral, but we talked about "Nana" a lot. Then my sister's husband died and that also was very difficult. We went to the funeral in New Hampshire and Chris was with his cousins and participated in all aspects of the funeral. We have spent many family vacations in New Hampshire and I usually make videotapes because all our family members live out of state or out of the country and the tapes have helped Chris "stay in touch" with everyone. He played the tapes of his nana, his uncle, and his grandpa often after they died, and I think that helped him.

However, the past year brought several more significant losses. One of Chris's former teachers died—she belonged to our church where Chris is

an altar server. Arlene struggled with cancer and when we last visited her before she died, she told him that she was counting on him to serve at her special (funeral) Mass and he promised that he would. It was such a sad day. Chris managed to get through it, although he did shed tears during and after the funeral.

Another big loss for him (and for us) was when my husband's brother died. Uncle Jim was a priest in Upper Michigan and he was on a ventilator in intensive care during the New Year's holiday. We spent 4 days with him at that time and Chris would pray with him, moisten his mouth, and just sit quietly with him. At one point he was sitting with his Dad in the waiting room and he started to cry. He said, "I'm afraid Uncle Jim is going to die." At that time, we didn't think Jim was going to die, but 3 weeks later he did. Again, Chris was involved in all aspects of the funeral. Because Jim was a priest, the Bishop celebrated the Mass and there were over 50 priests, all in their white robes. It was truly a celebration of Jim's life. Chris played the piano during the Mass and during the visitation time in church. He was also a pallbearer with his cousins.

Chris was very close to his cousin Julie who lives in England and 3 years ago he was an usher in her wedding. In April, Julie had her first baby, who died when she was just 2 weeks old. That was very hard for Chris, because he had been looking forward to seeing the baby when we went to England in June. I was touched by his openness with Julie and her husband, Ciaran. He put his arms around her and said, "Julie, I'm so sorry about your baby, Sinead," and at another time I heard him talking to Ciaran and telling him how sad he was that he lost his daughter. Some people would tell him to not talk about this terrible loss because it would upset Julie and Ciaran, but I think it's healthy and helpful to express your feelings and I know his sensitivity and love moved them. I also know that they are thinking about Sinead all the time anyway and Chris was able to shed some tears with them.

In most instances we tried to prepare Chris for the death of our loved ones. Whenever possible, we took him to visit them in the hospital or at home and we told him how very sick they were. I think that helped prepare Chris to deal with the loss.

Chris is a very spiritual person and we have shared our faith with him. Therefore, I know he truly believes that when a loved one dies, they

go to heaven. Every day he prays for the people he loved and has lost. When we visit the graves of our loved ones, he prays with his arms outstretched and asks the Lord to take care of his nana and granddad and baby Sinead or his nona and grandpa and Uncle Jim. I think that by openly talking about them and praying for them, it helps him and us.

Chris has a pretty good sense of time, but I know that not all of our adults do. I'm not so sure that time matters so much when you are thinking of someone you loved and have lost. If the person is thinking about them then I think it's probably OK to talk about them. If someone seems to be "stuck" and only thinking about a person who died several years ago, it might be helpful to acknowledge that you miss the person too. You could perhaps say a prayer together and then try to focus on something else, but I do think it is important to validate the person's feelings and not dismiss them. My nephew was killed several years ago in a car accident when he was 25 years old. My sister can still barely talk about Neil without crying, and I wouldn't dream of telling her that she should move on. We are all different and deal with loss in our own way, whether we have Down syndrome or not.

Reaction to Stress

Whether the stress is grief, a loud roommate, a divorce, or any of a number of challenges that life can present, individuals with Down syndrome may react to it in a variety of ways. Those who have good verbal skills may be able to talk about the stress. However, those with more limited verbal skills may have difficulty, and even some individuals with good verbal skills may still find it challenging to verbalize their feelings or discuss stress in their life (see chapter 7). Stress can affect us in a wide variety of ways, and the way each individual is affected and expresses stress can be unique.

Some individuals may have a behavioral change related to stress, as discussed in chapters 1 and 2. In addition, people with Down syndrome may experience physical and emotional reactions. Here is how some of our patients have described how stress affects them:

- "muscles get sore,"
- "heart beats faster than expected,"

- “feels hot,”
- “having too many pictures in the head,”
- “pulling your brains out.”

Dealing with Stress

Throughout the book, we discuss strategies to promote mental health as well as ways to treat mental illness. Since there is a close interplay between stress, mental health, and mental illness, reviewing and using those strategies can be helpful in managing stress. In addition, here are some of the ways our patients have shared that they manage stress:

- Coloring. (Adolescents and adults with Down syndrome have used this technique for decades, and others often questioned if it was age appropriate. Now many adults without Down syndrome are coloring as a stress reducer. People with Down syndrome were just ahead of the times!)
- Exercising
- Listening to music
- Taking deep breaths
- Closing their eyes
- Talking to family and friends

There are many stresses in life that can precipitate mental illness. Some of them are relatively small issues that are part of daily life, some are larger but expected, and some are larger but unexpected. Many strategies can be used to help a person with Down syndrome manage her stress. The first step is to acknowledge the possibility (and probability) that a teen or adult with Down syndrome will have stresses in her life. The second step is to listen to her concerns and help her devise ways to manage the stress. Individualize methods based on the individual’s needs and personality.

Return to Table of Contents

Chapter 15

Assessment of Mental Illness

We were assessing Randy for aggressive behavior. No one had previously asked him why he thought his behavior had changed, but he told us that his housemate Alvin was the cause. Alvin was entering Randy's room in their group home each evening after the staff went down to the office. Alvin was irritating Randy and taking his belongings. Randy did not take any action when this occurred but would ruminate overnight or perhaps for several days. Then he would retaliate in an aggressive, physical manner in front of the staff. (Because he has limited verbal skills, he did not know how to report Alvin's activities to the staff, and the staff had not asked him if anything was wrong.) Without the history provided by Randy, the staff had labeled his behavior as an "unprovoked outburst." The history obtained from Randy was invaluable in addressing his behavioral challenges.

In chapter 1 we discussed in a general way how we assess the mental health of an adolescent or adult with Down syndrome, whether or not a mental illness is suspected. When it seems likely that a mental illness is present, we proceed further with the evaluation. We seek more details about the chronological order of events, possible precipitants, family history, previous treatments, and the effects the change has had on the person and the family, other questions about the illness and about physical health symptoms and conditions.

Assessing a person with Down syndrome who has a mental illness or a behavioral change can be quite challenging. A number of barriers can make it difficult to obtain an adequate history. The impairment of language skills, conceptual thinking, and overall cognitive functioning are all hurdles to be overcome. This is because a large component of the diagnostic criteria for mental illnesses devised by the American Psychiatric Association relies on the person's ability to self-report subjective feelings. These criteria are described in the *Diagnostic and Statistical Manual of Mental Disorders*, 5th edition, or DSM-5). (Modified criteria are available in the *Diagnostic Manual – Intellectual Disability 2*, or DM-ID, to account for some of the limitations of using the DSM-5 with people with intellectual disabilities). For example,

people without an intellectual disability who have depression often report sadness, a lack of energy, a loss of interest in things enjoyed previously, and feelings of guilt and worthlessness. People with anxiety often report feeling anxious and fearful in certain situations. These pieces of the history may be less available from individuals with Down syndrome and other intellectual disabilities.

For people without intellectual disabilities, standardized questionnaires are available to help diagnose mental illness. These questionnaires essentially help the practitioner assess the individual on the basis of the DSM-5 criteria. Generally, teens and adults with Down syndrome have difficulty answering these questionnaires.

Because it is often difficult to obtain a clear history of emotional or behavioral changes in people with Down syndrome using conventional methods, we try to gather the information through a multipronged approach:

1. We obtain as much history as possible from the adult himself.
2. We ask parents and other caregivers for information about the history of the emotional or behavioral change.
3. We observe the person's behavior.

Obtaining a History from the Adult with Down Syndrome

As mentioned above, most people with Down syndrome have trouble sharing, or answering questions about, subjective feelings. Even some people with Down syndrome who have good verbal skills have difficulty articulating their feelings. As discussed in chapter 7, this can be an even greater problem if the adult with Down syndrome is speaking to an unfamiliar person. Nevertheless, we recommend getting as much history from the adult as possible. Although he may not report subjective feelings, he often provides very useful information, as illustrated in the example of Randy at the beginning of the chapter.

Unprovoked Outbursts

The term "unprovoked outburst" often causes us to have a "provoked outburst." Unfortunately, the term is sometimes used to avoid the responsibility of thoroughly assessing the environment and aspects of the individual's life that might contribute to her behavior. True, sometimes a person with Down syndrome will behave in an inappropriate manner with no apparent cause. However, as we have

previously described, people with Down syndrome may see the world differently than others. For example, strong memory abilities, a tendency to ruminate about events, a different understanding of time, and sensory issues (see chapter 12) may all contribute to a response that seems unrelated to a given event. Therefore, the label "unprovoked" may be much more of an indication of the observer's lack of understanding than it is of the person with Down syndrome's random loss of control. The implication of the term "unprovoked outburst" is that the person with Down syndrome has to be "fixed" and the environment has no role in either the cause or treatment of the problem.

When obtaining a history from a person with Down syndrome regarding a change in behavior or possible mental illness, we recommend the following:

- Bring him out of the situation, if possible. For example, if there is an inappropriate interaction between two people, separate them to help diffuse the conflict and allow for privacy to ask him questions.
- Reassure him. You might say something like this: "Thomas, you know I love you, but this behavior is not like you, and I think you know it's not appropriate." Or this: "Julie, you seem so unhappy lately. I am concerned about you and want to help."
- Start with an open-ended question: "Can you tell me what happened?" Or, "Can you tell me what is bothering you?"
- If he is not able to answer open-ended questions, try to probe with more direct questions without giving her an answer (which may or may not be the real problem). Be careful not to inappropriately lead the person. For example, if you have observed that Rosie has difficulty in the class that follows physical education class, it would be appropriate to ask, "Rosie, has something been happening in physical education class?" A question that would probably be too leading is, "I bet George is bothering you in physical education class. Right?" As previously discussed, many people with Down syndrome do seek to please others and will often answer questions in the way they think you want them answered. They will therefore often answer leading questions in the way they are being led.

If a person with Down syndrome cannot provide a history verbally, attempts should be made to get it in other ways. Having the individual draw a picture of his concerns has helped us successfully determine contributing causes in some individuals. Similarly, an adult with Down syndrome who likes to write might provide valuable clues about the history of his problems in his writing. Therefore, it may be beneficial for some individuals to ask them to draw or write about what is bothering them.

Obtaining a History from Parents/Caregivers

Although teens and adults with Down syndrome are frequently able to provide only limited information about their subjective feelings, close family members are quite often able to observe key symptomatic changes in behavior. For example, we find that most people with Down syndrome do not report sadness, but their families notice a definite change in personality, which we call a "loss of spark, life, and vitality." Most people with Down syndrome also do not verbally report a loss of energy or a lack of interest in doing previously enjoyed activities, but close family members usually observe this behavior change. Additionally, we have rarely heard a person with Down syndrome say that he feels worthless, so we do not rely on this self-reported symptom when evaluating someone for depression. Likewise, many people with Down syndrome do not verbalize feelings of anxiety, but body tension and other indications of anxiety are still usually quite obvious to family observers. (For more on the criteria for depression and anxiety, please see chapters 17 and 18.)

Another way that families can learn important information is by listening to self-talk. We have evaluated a number of adolescents and adults with Down syndrome who were not able to provide clear answers to questions regarding the issues that were bothering them. However, their families often heard them talking about the issues through self-talk (see chapter 9). Interestingly, many families have found that the person with Down syndrome speaks more clearly when self-talking than when speaking to someone else.

An additional piece of history that family members can provide is the family medical history. Many mental illness conditions run in the family. The family history may, therefore, give further insight into causes for the change in the mental health of a person with Down syndrome. In addition, sometimes the response of a family member to a particular medication may guide the decision as to what medication should be used.

Another important piece of information parents and caregivers can provide is how the person with Down syndrome has responded to previous medications. When we learn that a particular medication did not work, we ask ourselves, "Is the diagnosis correct? Is the medication correct? Are there other factors we have not considered?" For example, imagine that someone has problems with depression, but we do not obtain the history of mania that he also has. In this situation, prescribing an antidepressant for depression may induce mania. If that occurs, it can be a valuable piece of history. In addition, because obtaining the history is challenging, ongoing reassessment and history-taking is essential (particularly if the treatment is not successful) to determine if there are pieces of the history that were either previously overlooked or were not recognized as significant.

Difficulties in Interpreting Second-Hand Information

Even though we try to obtain as much history as possible from the individual with Down syndrome herself, often much of the information must come from family members or care providers. Unfortunately, this adds a level of interpretation. Observers bring their own biases, interpretations, and the possibility for error in reporting. In addition, another individual may assign a different level of importance to a behavior than the person with Down syndrome would. This may lead to under- or over-reporting of behavioral changes. This can be particularly problematic when the observer is unfamiliar with people with intellectual disabilities. The observer may interpret the common, typical behaviors described in Section 2 of the book as abnormal. This may cloud the whole perception of the events or presentation. Furthermore, frequently we will get completely different histories from families, residential staff, day program staff, and other service providers. These challenges make it necessary to obtain as much history as possible from multiple sources and try to sort out the discrepancies.

Dos and Don'ts in Reporting the History of a Behavior

When observing the behavior of a person with Down syndrome, we recommend taking these steps:

- Write your observations down. It can be difficult to remember the specifics, and often a connection to a cause will only be seen after reviewing the notes.

- Record chronological events. When did the symptoms first start? What changes were seen over time? If it is episodic behavior, what is the order of events during any given episode?
- Determine what else was occurring at the time the behavior occurred. What else was occurring in the family, at work, at school, with friends? For episodic events, it is important to look at the surrounding events, people, etc. As previously discussed, because many people with Down syndrome have such strong memories, it is important to observe for clues that may seem unimportant at first glance but may indicate a cause. For example, could there be a smell, an object, or some other trigger that reminds the person of a negative event in the past?
- Be as objective as possible. Avoid subjective observations such as, "He acts like he hates me." When observing, just as when interviewing, take care not to lead the observation in an inappropriate direction before you understand the problem. The same is true for assuming cause and effect. Even though an event occurred, and the behavior followed, it is certainly possible that a preceding event, an internal trigger (e.g., pain), or some other unrecognized occurrence actually contributed to the symptom or behavior development. In our experience, teasing out the symptoms and cause(s) is best done through careful observation, caution in assigning cause and effect, and keeping an open mind to all possible causes rather than focusing quickly on one cause.

Making Observations

In addition to obtaining as much history as possible from the individual with Down syndrome, as well as from family and care providers, we often find it necessary to do some observation of our own. Sometimes we obtain the best information by watching or being with the person in his home or work setting. As noted in earlier chapters, language difficulties, memory, the concept of time, and other issues for people with Down syndrome may make the history unclear. Putting the verbal history in context by visiting the person in her usual environment can be invaluable.

Quentin, age thirty-four, was brought to see us for outbursts of aggression. A visit to his home at 7:30 a.m. quickly revealed the cause of the problem. Quentin's bus arrived that day on time at 7:50, and he

became agitated when we continued to talk and tried to briefly delay him from getting on the bus. There was a clear compulsive component to his behavior. After discussing my observation and questioning the staff again, they acknowledged something they had previously denied due to not recognizing the significance—namely, that Quentin had many compulsive behaviors that interfered with certain activities. For example, he needed to finish one task, such as putting away items in his room, before moving on to another task or activity. If a staff person tried to divert him to another task before he was finished, he could become aggressive. With this clearer understanding of Quentin's problem, we suggested that staff organize his activities to minimize times that he was kept from completing tasks, and Quentin's aggressive outbursts significantly improved.

When we feel the need to directly observe problematic behavior, we generally prearrange a visit. We recommend arranging this with the family or staff and informing the person with Down syndrome. Occasionally, if we have arranged to see one person at a group facility (residential or work program), we might be asked to observe someone else or might inadvertently see someone else. In any case, we recommend asking family, coworkers, or caregivers if the behavior observed is typical. Observation itself might alter behavior, and it is important to know whether the behavior is not what is typically seen.

Conclusion

The assessment of mental illness in an adolescent or adult with Down syndrome can be difficult. It can be challenging and time-consuming to try to obtain a clear history from the individual, to get a history from a variety of people, or to directly observe the person, if necessary. However, careful, ongoing assessment using this multipronged approach is essential for understanding the nature of the person's problem and for developing the therapeutic plan that will be outlined in the next chapter.

Return to Table of Contents

Chapter 16

Treatment Approaches for Mental Illness

Janet, age twenty-seven, was evaluated due to changes in her behavior that affected her performance at work. Janet had a fantastic job in the community that she enjoyed, where her work ethic, friendly personality, and attentiveness to customers were appreciated. Recently, however, Janet's boss had noted that she was no longer getting along well with others. Customers who had previously gone out of their way to interact with Janet were now more likely to avoid her; some had even asked the manager if Janet was unwell. Her family had also noticed that Janet was more reluctant to participate in family activities, had developed erratic sleep patterns, and was more irritable than usual.

The assessment did not reveal any particular stresses or contributing physical health conditions. Janet started counseling with a social worker, but her symptoms worsened and she became less involved in activities, even her own self-care. She continued counseling but was also started on an antidepressant/antianxiety medication. While most of Janet's symptoms improved, she was still reluctant to participate in activities and resisted her family's efforts to encourage self-care. Janet was then assessed by an occupational therapist, who helped develop picture schedules to guide Janet to good self-care. In addition, Janet joined a social skills group, where she practiced interacting with others; this helped her feel more comfortable with returning to her previous activities in the community. Her boss had been kind enough to give Janet time off to work through these issues, and several months later she was able to return to her job. Once again, the customers gravitated toward "the old Janet," whom they loved and respected.

Once a mental illness is diagnosed, the next step is to develop a treatment intervention. Chapters 17 to 27 discuss the specific treatments that are often effective for adolescents and adults with Down syndrome who have been diagnosed with particular mental illnesses or additional conditions. However, because many of these treatments involve either counseling or the prescription of certain classes of

medications, we will provide an overview of general issues related to these treatment approaches here, in order to avoid repetition from chapter to chapter.

PART 1: WHEN COUNSELING IS NECESSARY

Counseling may be extremely helpful to some adolescents and adults with Down syndrome. Under the right circumstances, counseling from a trained, sensitive, and knowledgeable therapist may

- give people support and encouragement,
- enhance pride and self-esteem,
- help people to identify and resolve life problems,
- be a useful part of the treatment for more serious problems such as depression, anxiety, obsessive-compulsive disorder, etc.

This sounds wonderful, and it can be, but a number of conditions need to be met for people with Down syndrome to benefit from counseling.

To make an informed judgment as to whether counseling would be helpful, an adult with Down syndrome and her caregivers need to know more about what counseling entails. If they decide to try counseling, the following conditions should be met:

1. The counselor has adequate education, training, experience, and sensitivity.
2. There is a good fit between the personality of the counselor and the person counseled.
3. Counseling is a safe and secure process.
4. The counseling process has meaningful goals, and a means to obtain these goals and assess an outcome.

Counselors

Counselor is a generic term we are using to refer to any professional who is trained to identify and help resolve emotional and behavioral problems that people have in their lives. A number of different professionals are trained to do counseling, including

- social workers (MSW, DSW, or PhD),
- psychologists (PhD or PsyD),

- counselors (MA, MS, or EdD),
- psychiatrists (MD or DO),
- pastoral counselors (MDiv, DDiv, or PhD), and
- marriage and family therapists (MFT).

Depending on the situation and the particular needs of the individual, any of those included in the list may be a good fit. Their ability to establish rapport as well as their training and experience are what matters most. The ability to use a specific approach to counseling also depends more on the counselor's training than on his or her profession. For example, behavioral counseling began over sixty years ago in the field of psychology, but this approach is now used by all the different counseling professions. As a second example, marriage and family counseling began approximately fifty years ago by counselors from all different professions and it continues to be practiced by all these fields as a specialized area of training and practice (see below for more on this approach and an example). There is also a relatively new field of counseling—marriage and family therapy—that specializes in this approach.

Like behavioral and marital/family counseling, most other counseling approaches and techniques may be used by all the different counseling professionals if they are trained in these approaches. There are, however, several professions that do specific tasks that are unique to these professions:

Psychologists are trained to do counseling, but they also have unique expertise in *psychometrics*. This is the use of standard tests and instruments to measure different areas of mental and behavioral functioning. There are many different types of standard tests (neuropsychological batteries, personality tests, adaptive skills and maladaptive functioning measures, AD/HD tests, etc.). However, the tests that are often referred to as "psychologicals" are the standard intellectual quotient (IQ) tests that measure a person's level of intellectual functioning. These types of IQ tests are often required by state governments for people with Down syndrome and other intellectual disabilities to establish the degree of the person's intellectual disability.

Psychiatrists also have somewhat of a unique role in the mental health field. Psychiatrists are physicians (MD) or Doctors of Osteopathy (DO) who are trained to treat people with mental health problems. They often have training in counseling, but many specialize in managing psychotropic medications. This is the class of medications for the treatment of mental health problems, such as antidepressants and antianxiety medications, and antipsychotic medications. Physicians who are not

psychiatrists, as well as nurse practitioners and physician assistants, are also able to prescribe psychotropic medication.1 This is particularly the case for the conditions with more commonly known symptoms, such as depression and anxiety. Physicians may also refer a patient for a consultation with a psychiatrist, just as they would to any other medical specialist. Additionally, many people may initially consult with a psychiatrist to help diagnose and recommend a medication for a mental health problem. Afterward they may have their own primary doctor refill their prescription and follow them medically for the condition.

Does the presence of a more serious mental health problem require the treatment of a psychiatrist or physician? The answer is both yes and no:

Counselors who diagnose mental health conditions in their practice and who are not able to prescribe medications will quite often refer patients to a physician for a consult on a psychotropic medication. In these cases, the counselor often continues to see the person in counseling while a physician manages the medication. Likewise, many physicians who prescribe medications for mental health problems will refer the patient to a counselor for ongoing counseling. In either case the patient benefits from the combination of both medication and ongoing counseling. Research has consistently shown that either medication or counseling may be beneficial, but the combination is far more effective in resolving these types of problems and symptoms (Frank et al., 1990; Cuijpers et al., 2014).

Payment for Counseling Services

We would be remiss if we discussed counseling without talking about how people pay for this service. The truth is that counseling and mental health services are often underfunded by insurance and government sources in the United States. There has been great progress in this area over the last forty years or so. The Affordable Care Act has provisions to improve reimbursement for mental health services. However, a lower reimbursement for some mental health services (compared to physical health) as well as a lack of coverage for some others remains. It is very important to reviewing the person's personal insurance coverage before embarking on counseling.

[1] In some states, certified nurse practitioners (CPN) and physician assistants (PA) are also able to prescribe medications.

Education and Training

Individuals with Down syndrome need fully qualified counselors. Just because their thoughts and feelings might seem less complicated than those of others their age does not mean that someone who is not a counseling professional would be a "good enough" counselor for them.

Like other adolescents or adults, people with Down syndrome should receive counseling from individuals who have, at a minimum, a master's degree from an accredited program in their chosen profession (four years of college and at least two additional years of a specialized master's program in a counseling field). They must also have a license in their chosen profession from the state in which they practice. In most cases, state licensure is dependent on passing an accrediting exam and working a required number of hours, supervised by a senior clinician, after the completion of a formal educational program. Training and subsequent work experiences should be documented in a résumé, and the counselor should be able to furnish this and a copy of his or her diploma to anyone seeking counseling.

In the disability field there are a number of different types of case managers, including Qualified Intellectual Disability Professionals (QIDPs). There are also managers and direct caregivers in residential living situations and at worksites. These individuals have critically important roles and are often very caring and sensitive people, but they have not had training or education to do counseling and therefore should not be considered appropriate for this role.

Personal Issues

Counselors should be sensitive, insightful, and caring human beings. Additionally, the counselor's life experience may be an important factor in the success of counseling. Counselors who have "been there" (having experienced different aspects of life: marriage, raising children, grief, etc.), may be more aware and less judgmental than those who are younger or less experienced. In fact, researchers have found that successful therapists tend to be more experienced and try a variety of approaches to suit the person and her problem. Younger therapists may need to be more rigid in their approach simply because they do not have the life or treatment experience to try different things that work. Fortunately, young counselors are often supervised by more experienced therapists. This type of

arrangement may offer the "best of both worlds"—the energy and enthusiasm of a youthful therapist coupled with the wisdom of a seasoned supervisor.

Another consideration is whether you should seek out at counselor who has previous experience working with people with Down syndrome. In our experience, a counselor who has this experience may help the adult with Down syndrome have a positive counseling experience—one in which she feels as if the counselor truly understands and appreciates her, both for her strengths and weaknesses. The counselor who "knows" asks the right questions and responds to the right issues; this says "I understand" to the person being counseled. If the counselor conveys a sense of interest and genuine "like" of the person with Down syndrome, it can have a dramatic benefit in the counseling relationship.

Sometimes it may be very difficult to locate a trained counselor who has experience with adults with Down syndrome. In this case, finding someone who is willing to learn might be the best option. In fact, when we first started the Adult Down Syndrome Center, we had very little experience or understanding of people with Down syndrome. We did, however, have a great deal of respect for the knowledge of family caregivers. We listened and respected the opinions and ideas of caregivers who had spent a great deal of time, energy, and effort trying to understand and advocate for the person in their care.

If a counselor is able to bring his or her own expertise and still be open to learning from the family, then this collaboration has an excellent chance of working. In contrast, if the counselor seems to look down on the person with Down syndrome or lack understanding or respect for her or the caregivers, then the counselor should be avoided.

Conflicts of Interest

A cardinal rule in the counseling field is that a counselor cannot have a "dual role" with the person counseled. This means the counselor can only play the role of counselor for the person being counseled. He or she cannot be a friend, parent, caregiver, manager, supervisor, salesperson, and certainly not a lover or sexual partner. Residential or worksite staff or managers, even those who have training and experience as counselors, cannot be counselors to the people they work with or supervise. This simply does not work.

It may get a little confusing when staff who are assigned to work as case managers or direct caregivers are called "advocates" or even "counselors." While

they may appropriately advocate for the person's needs and services and assist the person in their care, they cannot serve as her counselor. This must be left to counseling professionals in order to maintain the integrity and security of this process. (See the sections on safety and confidentiality below.)

Safety Issues

Counselors may be caring human beings, but they must also carefully maintain a professional role and observe boundaries to ensure the safety and security of the counseling experience. Public awareness of safety concerns has been growing in light of allegations of abuse leveled at a variety of professionals and caregivers who provide services for children. Families are also justifiably concerned with the safety of family members with Down syndrome who are dependent on the care of others.

There may be some relief to know that safety and confidentiality have been the focus of the counseling field for as long as there have been counseling professionals. Just as medical professionals are careful to have nursing staff present during a medical procedure, counselors should be careful when they do individual counseling. For example, one way to help ensure safety during individual counseling sessions is to leave the door open several inches. The person in counseling is not heard outside the door (to protect her confidentiality, see below) but other counselors or family members are able to see in.

Sometimes it is wise to have a second staff person present during counseling sessions, as illustrated in this story:

> *Teresa, twenty-eight, came for counseling about issues related to sexuality and dating. She had many questions about sex and her relationship with her boyfriend. Her questions showed a great deal of misinformation and confusion about what "sex" entailed. For example, she asked whether she was pregnant when she had her period and if she could get pregnant by kissing her boyfriend.*
>
> *Although Teresa was very verbal and capable of managing daily living skills and her job in the community, she was immature in certain respects and did not have a good sense about what was private. She would ask anyone she encountered questions about sex. This included customers at work and strangers at social events. When her mother learned this, she*

was appalled and tried to answer her daughter's questions. Unfortunately, she was not terribly comfortable with discussing these issues, and her daughter did not seem to understand her explanations.

Teresa had a complete physical exam with a nurse practitioner, and she and her mother met with counseling staff to discuss her inappropriate behavior. Although Teresa requested individual counseling sessions, she agreed to have a female staff member present with the male social worker. This staff person was helpful in ensuring safety (for the benefit of both the counselor and Theresa) and also helpful in bringing a female perspective to the discussion.

In order to manage her problem social behavior, Teresa agreed to write down any questions she had about sex and dating and to discuss them only when she met with the counseling staff (or her mother). A sex education meeting with the outreach staff person and the nurse practitioner was also set up for her. Interestingly, during counseling and sex education sessions, staff learned that Theresa did not actually want to have sex but just to be affectionate with her boyfriend. Over time, these and other issues were sorted out to the family's and Teresa's satisfaction.

As this example illustrates, careful attention to safety issues protects the integrity of the counseling process while still allowing the discussion and successful resolution of even the most sensitive personal problems and issues.

Confidentiality

Confidentiality is also a key issue in counseling. How can someone feel free to talk about sensitive personal issues ("to spill their guts"), if this information is not kept private and confidential? The counseling profession is clear about this issue for adults who do not have an intellectual disability. Counselors are allowed to discuss facts of the counseling with a supervisor (who must also maintain strict confidentiality). Otherwise, the only time confidentiality is breached is when the person being counseled makes a credible threat to hurt herself or someone else, or when the person allows limited information to be given, such as to her insurance company for payment purposes.

Confidentiality is more complicated for adolescents. In a strict legal sense, guardians are allowed access to records up until the adolescent is the age of majority, which is eighteen in most states. However, for a number of conditions, including mental health, adolescents as young as twelve may obtain some services without parental consent. The conditions covered, the age, and other particulars of the laws vary by state.

The issue becomes far more complicated for adults with Down syndrome. Chronologically, they are adults, but developmentally they are often more like children or young teenagers. The legal issues are also confusing for many families of people with Down syndrome. Generally, adults with Down syndrome who have legal guardians (established through the appropriate court) are subject to the same rules as children. Guardians have access to their records. On the other hand, adults with Down syndrome (over the age of majority) who have no legal guardians are considered their own guardians and therefore have all the legal rights of other adults. This sometimes presents problems, such as when people who are their own guardians do not agree to needed medical treatments. However, we have not found that this confidentiality issue is a major problem when counseling people with Down syndrome. Once their rights are carefully explained, most adults with Down syndrome agree to share relevant information with caregivers. This is fortunate, because it is rare for us to do individual counseling without the ongoing involvement of caregivers.

In addition to guardianship, there are alternatives that legally establish a relationship that allows family members (or others) to participate in the decision-making and consent processes. Through the process of obtaining guardianship, the individual who is assigned a guardian is declared incompetent and thus loses some rights. Alternatively, arrangements such as supported decision-making and power of attorney do not require that the person be declared incompetent and allow her to maintain rights of personal decision-making. Even without such an agreement, an individual with Down syndrome, just like anyone else, can agree or request to have others, such as family, assist in decision-making and health care assessment and treatment.

We like to involve caregivers because expressive language limitations make it difficult for many people with Down syndrome to communicate relevant issues and concerns to the counselor. Individual counseling is not useful if it is conducted in a proverbial vacuum. Therefore, gathering information from caregivers in different environments may help to keep the counseling focused on the adult's real needs and

issues. The more information available to the therapist, the more understanding and helpful the therapist may be.

When there is a need for more open communication between the counselor and caregivers, the issue of confidentiality must be explained very clearly to the person with Down syndrome and her caregivers before the start of counseling. Whenever possible, the counselor must respect the needs and wishes of the person with Down syndrome. For example, some individuals are dealing with adolescent-like issues of independence with parents. (For people with intellectual disabilities, these are often delayed until their twenties or thirties.) In these situations, we may try to negotiate more privacy for the person with Down syndrome from caregivers (just as counselors of adolescents without an intellectual disability do), even if the caregivers are the guardians. Additionally, if parents are given information or progress reports on the counseling, we try to get permission to do this from the person with Down syndrome. Although this is not legally necessary if someone has a guardian, it is necessary to maintain trust in the therapeutic relationship.

We are also careful to ask the person in counseling first before discussing any information with her parents or other caregivers. It may be helpful for the person with Down syndrome to be present when this information is discussed with the caregiver, either in person or by phone. Again, this helps to maintain trust and confidence in the integrity of the counseling process. We have not had any problems creating a collaborative relationship between all parties when we have dealt with these issues openly and sensitively from the start.

The Wrong Counselor?

What makes a good counselor for a person with Down syndrome? What makes a good counselor for this specific person with Down syndrome? What if there are concerns that this person is not the right counselor?

Some counselors have gone through clinical training and internships, have extensive life experiences and even experience with people with Down syndrome, but still may not be the right counselor for a particular person with Down syndrome. Perhaps they are not very insightful in general or in a specific problem area, despite their training or experience. Or they may lack the ability to understand and convey a sense of warmth and caring to the person they are counseling.

Even when a counselor is trained, understanding, and sensitive, he or she may still not be the right person for the job. The truth is, everyone has a unique style and

personality, and sometimes the personalities of the counselor and the person counseled may clash. Counseling is as much art as science, and much of the art involves the counselor's personal style.

Counselors are very aware that personality is a critical component in counseling and often recommend that the patient interview them before starting therapy. Many also recommend a trial period before a commitment is made for counseling. If you are seeking a counselor for a family member with Down syndrome, it is highly recommended that you sit in on at least one session to see if the counselor is the right person for the job. Issues of personality and style can only be determined through face-to-face contact. When you observe the counseling process, trust your intuition and your sense of whether or not the counseling feels right for you and especially for your family member with Down syndrome.

Most people with Down syndrome have definite feelings about the counselor they are seeing. In our experience, they usually have an innate sense of the practitioner's interest in and comfort level with them. The question is how to gather this information from the person being counseled. People with Down syndrome often try to avoid being negative or critical, but someone who knows them well can usually determine their true feelings from the intensity of their response (enthusiastic to less than enthusiastic). If the adult with Down syndrome gives a half-hearted endorsement or response to the counseling, this should be looked at very closely. Perhaps the person needs some time to get used to the process of counseling because it is new. However, if her response continues to be half-hearted or lukewarm after a number of sessions, this may mean that the counselor is not a good fit for the person's needs and personality. Any good counselor will not have a problem referring the individual to another counselor who is a better fit due to personality issues.

Types of Counseling Helpful for Adults with Down Syndrome

Supportive Counseling

As part of a multidisciplinary evaluation, a psychosocial assessment is completed by counseling staff to gather key information on the person's social and adaptive skills, as well as her network of family and peer support. In our experience, during this assessment, the person's caregivers often ask her if she would like to talk alone with the counselor. They explain that this may be a way for the person to express her feelings, "to get things off her chest," etc.

Although many people with Down syndrome do not feel a need to talk alone, some do. Interestingly, quite a few of the individuals who want to talk have no current or urgent problems or issues. Instead, they often want to discuss some meaningful experience or event from the past. Common themes include being hurt or treated unfairly by another, being jilted by a love interest, or losing someone close. The issue may also appear fairly insignificant to others, such as an accidental breaking of a dish. Frequently the person will share the same story at each session.

Regardless of how seemingly insignificant or repetitious the issues, we take the counseling process very seriously. We listen very patiently and respectfully, ask for clarification when needed, reflect on the feelings expressed, and give advice when appropriate. We have learned that being able to express themselves to someone who really listens to them is extremely important to adults with Down syndrome and enhances their self-esteem. For these individuals, the verbal message may be less important than the communication of some feeling that is heard and responded to with respect and understanding.

This type of counseling is called *supportive counseling*, and despite the apparent absence of an urgent problem or issue, the process can be very beneficial to the person counseled. This approach is used only if the person is offered and accepts the invitation to talk to an experienced counselor. In our experience, those who accept this invitation often feel they are able to say what they need to in one or two meetings, although some people continue for a more extended time. After the counseling is completed, we try to "check in" with the person with Down syndrome with brief individual meetings when she returns for routine medical follow-ups. This often helps to keep a therapeutic connection between the counselor and the person with Down syndrome long past the initial counseling sessions. Additionally, and perhaps most importantly, this gives many people enough of a positive experience with counseling that they ask to come back to talk when there are new issues and concerns that are bothering them.

Good counseling always involves elements of supportive counseling such as being sensitive to the person's thoughts and feelings, regardless of the verbal message. The counselor communicates a message of human kindness, understanding, and respect. This message may be communicated to someone who is nonverbal or who has limited verbal skills just as well as to someone who has good verbal skills. The process of respectful listening and responding is extremely beneficial to all, regardless of their expressive language skills. We know this is the case because people have communicated in clear nonverbal messages their wish to continue counseling.

For example, some people use the sign for talk or move their hand outward from their mouth. Still others put their hand over their chest, indicating a need to express feelings (from the heart).

Some individuals with Down syndrome can get "stuck" when talking about certain events. They repetitively talk about a topic or event and have difficulty moving on. In this case, we use a three-pronged approach similar to that noted in the preceding paragraphs: (1) acknowledge, (2) empathize, and (3) redirect. In addition to listening, part of the support comes from redirecting the individual to participate in other activities and limit the perseveration that may be occurring. This approach is primarily supportive but also contains a component of behavior-changing approaches addressed below.

Insight-Oriented versus Behavior-Changing Approaches

All good counseling includes some type of learning or insight about oneself or one's behavior. Even in supportive counseling, the person may learn that she is valued and that she has more skills and resources than she had assumed. Often, supportive counseling is sufficient. Other times, more extensive measures may be needed. In these cases, the goal of counseling is often identifying and changing maladaptive ways of thinking and behaving that are creating problems for the person.

Insight-Oriented Approaches

Some counseling approaches emphasize the need to change how people think. These are often called insight-oriented approaches. Proponents of this approach assume that if people understand the cause of a problem, they will be motivated to behave more appropriately. For example, for Teresa, one key insight was that it was inappropriate to address questions on sexual issues to anyone other than her mother or the counseling and medical staff.

Other approaches emphasize changing behavior as a way to gain insight. For example, many approaches use carefully designed tasks to stop maladaptive patterns of behavior. The assumption is that it is easier for someone to gain insight once she does a task and sees the positive result. In reality, insight-oriented and behavior-change approaches work hand in hand, components of both are often used in any counseling situation. To return to the example of Teresa, we can see that she gained insight into her problem behavior (discussing personal sexual issues inappropriately)

both before and after she did the assigned task (which was to discuss these issues with counseling staff).

Behavior-Changing Approaches

There are also a host of approaches that emphasize behavior change and not insight. These so-called behavioral approaches include the behavior modification and applied behavior analysis approaches. Behavioral counselors identify behaviors in people's lives that create problems and then reinforce more desirable alternative behavior. Behaviorists often break the problem down into manageable steps of change. Behaviorists are also known for systematically tracking the frequency of the targeted behavior to evaluate any changes.

Behaviorists have not generally been known for their supportive counseling techniques. But when they work with adults with Down syndrome, they need to be sensitive to the thoughts and feelings of the individual and her family in order to develop a good working relationship. For example, when designing a behavior plan, successful counselors listen carefully and respectfully to caregivers in order to identify problem behaviors and issues. Since caregivers are often the ones who reinforce the adult's behavior, the counselor needs their full cooperation and acceptance of the plan. He or she needs to give some insight and rationale for the use of the approach and the expected change in behavior.

Equally important, the successful behaviorist will work very hard to obtain the understanding, acceptance, and cooperation of the person with Down syndrome. At the very least, the behaviorist must carefully consult the adult regarding what reinforcers are most desirable. Helping the person become invested in the process is not only respectful but also greatly increases the chance of success. This is because the person feels she has a hand in managing her own behavior. Along these same lines, adults with Down syndrome may be able to use charts to monitor their own behavior rather than just rely on caregivers to do the charting. These and other strategies make the behavioral approach more palatable for caregivers and people with Down syndrome. There are a number of good examples of this approach throughout the book. See, for example, the story about Jamie and OCD in chapter 19.

Combined Approaches to Counseling

Several counseling approaches combine insight-oriented with behavior-changing strategies. Here we describe the two that we have found to be especially effective in working with people with DS: the social learning and the cognitive behavioral approaches.

Social Learning Approach

The social learning approach is one of the most popular approaches in the counseling field. This approach has been shown to be effective with a wide variety of problems, including depression and anxiety. Through this approach, people change problem behavior by first watching others do a task and then learning to do it themselves, through a technique called modeling.

We have found this to be a particularly effective strategy for people with Down syndrome because they tend to think in visual images and to have excellent visual memories. As discussed in chapter 6, this is why many people with Down syndrome can remember past events in great detail. The ability to learn through visual observation and recall is also often very impressive.

In chapter 19, we give an example of an adult with Down syndrome using the social learning approach to model a desired behavior changes first demonstrated by his sister. The adult, Charles, had developed a habit of buying the same toiletry items over and over, even though he already had more than enough of them at home (such as ten bottles of shampoo). After his parents died and he moved in with his sister, she consulted with us to learn how to break him of this habit. The first step was to have Charles look in his bathroom cabinet to see what items he truly needed. His sister then helped him find a picture or make a drawing of the item he needed. Afterward, he took this picture to the store to help him locate and buy the item.

The new routine gave Charles a sense of independence and purpose during shopping trips. His sister was also very pleased because it helped him develop independence while keeping him from buying and hoarding items unnecessarily. His sister followed up the shopping trips by watching him put the acquired items away in his cabinet. She would then heap praise on him for a job well done. After doing this routine together for several shopping trips, Charles began to do the task by himself.

Perhaps the most interesting and innovative use of the social learning approach is to use a photograph or a video playback of the person herself. This approach is called self-modeling. As discussed in chapter 4, people with Down syndrome are

drawn to photographs and videos of family and friends but are especially interested in images of themselves. For example, we find that people with Down syndrome are far more likely to follow an aerobics exercise routine if they watch a tape of themselves doing the routine. Another example of the social learning approach is the story of Brian, who refused to see his brother after his brother was unable to come to a holiday gathering (chapter 6). In order to get the two brothers together, their sister followed our advice and showed Brian pictures of positive past family events that included both brothers. Not surprisingly, the photos that had the most influence on Brian depicted him with his brother. This concept is called self-modeling.

Showing the person images of herself can also be a great way to motivate appropriate behavior. The beauty of videos in particular is that they can be edited to show desired behavior that the person is not engaging in at present:

Rosemary, age thirty-four, had been independent in completing basic self-care tasks, but she began to refuse to leave her house to go to work in the morning. After a thorough evaluation, we determined that this was due to a number of factors, including hypothyroidism, fatigue, and a conflict with a staff person where she worked. The hypothyroidism was treated, and her fatigue was resolved by moving Rosemary in with a new roommate who did not keep her up late at night. Conflicts with the staff person were avoided by having a different staff person interact with Rosemary in the morning.

After these changes were made, Rosemary's morning routine went more smoothly, but she still required constant prompting to complete her routine in a timely manner. We believe this was because the problem had gone on long enough to become a habitual pattern for her. We suggested that staff videotape her morning routine, to include the constant prompting from staff. Following our directions, the staff then edited the tape. The edited tape left out all prompting and approximated a normal speed for completing her morning routine. We then asked the staff to show Rosemary the edited tape in the morning after she woke up.

The first morning Rosemary was shown the video, staff were very surprised by the result. Not only was she mesmerized by it, but she then went on to follow her routine at a much faster pace and without needing much prompting. For the next few days, she continued to watch the tape

every morning and then to move at a faster pace. By the end of the week, she was completing her routine as quickly as she had prior to the onset of her problem behavior. The only change in her routine was that she looked at the video every morning. She continued to do this long after the problem was resolved.

Although self-modeling is likely to be most effective, peer modeling can also be helpful. In peer modeling the individual who demonstrates the desired behavior in the video is similar to the individual watching the video. We are developing a series of patient education videos (available on the Advocate Medical Group Adult Down Syndrome Center web page) in which the actors and actresses have Down syndrome. Peer modeling videos can result in improved attention to and learning of the subject matter. These videos may not be as specific to the desired behavior, but many behaviors can be successfully addressed this way.

Cognitive Behavioral Approach

The cognitive behavioral approach is also very effective for people with and without Down syndrome. This approach emphasizes changing thoughts that influence someone's mood and behavior and is especially effective for treating depression. Research has shown that people who are depressed have negative thoughts about themselves ("I am worthless"), about their ability to affect the world ("I cannot do anything"), and about the world (a cold and insensitive place). It is easy to see how this then affects their self-esteem and makes them highly susceptible to depression. In the cognitive behavioral model, the counselor helps people to identify negative thoughts and change them into positive thoughts and behavior.

Sometimes a person's negative thoughts have some basis in fact—for instance, if she cannot do a task due to limited skills. For a person with a strong bent toward negative thoughts, "failing" in a task often supports her own beliefs that she is bad or deficient in some way, which may then lead to depression. Sometimes, however, the failure may simply be due to lack of instruction in completing the task. Once the person learns to do the task, she is successful. Other times, the task may be beyond the person's capability. In these situations, the person is encouraged to do a task that is more appropriate for her skills. In either case, once she is taught to do a task successfully, or encouraged to do an appropriate task successfully, she is then encouraged to listen and accept positive feedback from others. In addition, she is

encouraged to practice making positive comments about her success to counteract any continuing negative thoughts and behaviors.

When using this approach with someone without an intellectual disability, a therapist will carefully examine the person's thought patterns to look for negative and self-defeating thoughts that result in a problematic response to a challenge. For example, the therapist will ask the person what she would say to herself when she tries to deal with a problem (e.g., "I cannot overcome this challenge"). The counselor will then help the person to substitute a more positive thought that is more likely to result in a positive solution (e.g., "I have the strength and skill to meet this challenge"). This approach can also be used for people with Down syndrome who have very good verbal skills.

For people with Down syndrome who are less verbal, self-talk (see chapter 9) can be an excellent vehicle for examining negative thoughts. Caregivers often know or are able to learn the content of the person's self-talk. When negative self-talk messages are identified, the counselor can help the person to substitute phrases that are more positive, just as counselors do with people without an intellectual disability. A good example of this is seen in the case of Ben, discussed in detail below. When people have more positive thoughts, they behave more positively. They then receive more praise and feel increasing pride and self-esteem, leading to more positive thoughts and behaviors (on and on in a more positive spiral).

The example below illustrates how a cognitive-behavioral approach might be used with an adult with Down syndrome as well as how we might use a combination of different counseling approaches to resolve a problem:

> *Ben, age eighteen, lived with his parents and older brother and attended a transition program that emphasized community work skills. He was able to communicate effectively to familiar others but sometimes had problems verbalizing his feelings. According to his family, everything was fine until he began his second year of the transition program. At that time, he showed symptoms of depression, anxiety, and an obsessive-compulsive disorder and was moody, tense, and irritable. He became withdrawn and started refusing to go to social and recreation activities he had previously enjoyed. He also stopped doing the free-time activities he loved such as listening to music, watching movies, and playing video games. In addition, his appetite had decreased and he had a great deal of*

difficulty falling and staying asleep at night. He was listless and appeared to have little energy during the daytime.

Ben's family also became concerned with his increasingly odd compulsive behavior. In the past, his compulsions ("grooves") had been generally beneficial for him and his family. He was neat and orderly and careful with his grooming and appearance. Ben was also able to reliably complete self-care and work/school tasks because they were part of his daily routine. This changed as his grooves became more rigid and began to interfere with his normal functioning. For example, he began to take the garbage out every hour and to hoard ever-larger amounts of food in his room. He also became more extreme about the need to keep items in precisely the same spot. He had previously insisted on exact placement of items only in his bedroom, but as this need expanded to rooms in the rest of the house, it became a safety issue. The objects he moved now included massive items such as a piano, TV, couches, large glass objects, etc.

Perhaps most disturbing to Ben's family were his nightly tirades. Although he was not aggressive toward family members, he became increasingly angry and agitated as the night went on. The content of his tirades involved any negative or teasing comments directed toward him from others, and he used his superb memory to draw on up to fifteen years of such comments. He tended also to repeat the same incidents over and over through self-talk every evening, with greater intensity as the night wore on. Sometimes Ben seemed to be carrying on conversations with imagined others (which is not uncommon; see chapter 9), but his family recognized most of these conversations as replays of past negative experiences. It appeared to Ben's family as if he was building a strong negative case against himself.

The diagnosis and treatment of Ben's problem began with a medical evaluation, which revealed hypothyroidism and impaired hearing. These issues no doubt had an effect on his current symptoms. However, his parents also reported that he had recently been a victim of abuse by a female student with a disability. This happened while Ben and the other student were participating in a special recreation group at a community center. The other student had serious problems of her own and had vented her anger on Ben, who was much smaller than she was. On at least one occasion, the female student had groped at Ben's genitals. His parents

believed that the sexual groping was far more disturbing than the physical aggression to Ben.

Once we learned of the abuse, Ben's behavior became more understandable. His agitated self-talk was a clear example of the type of self-blame and self-recrimination that is common among victims of abuse. His exceptional ability to recall past negative events only added to his sense of shame and self-blame. On the positive side, his anger was a far better response to the abuse than withdrawal into a severe state of depression, which happens to many individuals who experience this type of abuse. His increase in more rigid compulsiveness is also a common response to stress, particularly for people with Down syndrome who have strong preexisting compulsive or groove-like tendencies.

Our treatment strategy was multifaceted, including medical treatment for his hypothyroidism and a referral for a hearing evaluation. After careful consideration, Ben was also prescribed an antidepressant medication to help reduce the intensity of his compulsive behaviors and his upset mood. At the same time, we offered him supportive counseling to boost his pride and self-esteem, which had been badly damaged by the abuse. We also counseled Ben's parents, who were very upset and concerned for their son. In family meetings we praised Ben and his family for having great strength of character in responding to the crisis. This helped to reduce their sense of self-blame.

Additionally, we used several counseling strategies that were discussed above. For example, at our suggestion, Ben's parents located many photographs and home movies of Ben involved in positive experiences. This served as a potent form of self-modeling, showing Ben as a strong, proud, and capable young man who enjoyed life. It also served as a substitute for his memory of negative comments and experiences that plagued his evenings. His parents were able to get him to focus on these positive memories in the "quiet time" after work when he was most susceptible to his memories of negative past events. They also commented positively on the pictures: "Look how good you looked . . . how much fun you had . . . how well you did that," etc.

A modified version of a cognitive behavioral technique was also helpful to Ben. He agreed to use a simple but effective strategy of "changing the channel" whenever he had negative thoughts. One of Ben's

parents described this technique as a "multimedia production." Ben would raise both hands up as if to turn the channel on an imaginary TV, while saying loudly and clearly "turn the channel." Ben's parents helped by reminding him to "turn the channel" whenever they noticed the negative self-talk. They also helped him to substitute negative comments with positive statements such as "I am a good person, and my family and friends love me." Although these were simple statements, they were very effective in helping him counteract the negative comments. He would then repeat these comments over and over, particularly in the evening when he was most susceptible to negative self-talk.

After a number of practice runs and reminders, Ben was often able to use his positive self-talk and "turn the channel" technique fairly automatically. In time, he even was able to remind himself if he was aware of the negative self-talk. His parents also helped him to focus on something positive when he changed the channel. They would either remind him of a favorite memory or show him a photograph of a favorite past event.

Over time, Ben showed a positive response to both the medical and counseling approaches. His mood improved, his obsessions and compulsions became less rigid and more productive (or groove-like), his anger dissipated, and he showed a renewed interest in all the activities that he had enjoyed before. After several years, he continues to do well. He was even been able to work with the young woman who abused him. Fortunately, this woman had been treated and was watched closely by staff.

Counseling for People with Acceptance or Identity Issues

One important reason for counseling is to help people better understand and accept who they are. In our experience, this often involves helping people accept the fact that they have Down syndrome. As discussed in chapter 8, this is very important because acceptance increases the person's willingness to use and develop her skills and abilities as well as advocacy for her own rights and needs.

Although there are relatively few people with Down syndrome who have "acceptance of self" or identity issues, for those who do have them, these issues may

have a profound effect on their lives. We see this in a number of key areas of life, including in the social arena and on the job.

Individuals with identity issues tend to have an aversion to socializing or associating with peers who have Down syndrome or other intellectual disabilities. For some people, this is a minor problem. Some prefer to socialize with staff, while others are selective about whom they socialize with at social gatherings, preferring people who are more capable. Generally, these individuals do not have an aversion to participating in activities with others with disabilities, and more importantly, do not have a negative view of their own disability. Of course, we would not force these people to make friends they would not choose on their own any more than we would force someone without Down syndrome to interact with people they do not choose. As long as they have good self-esteem and a positive view of their Down syndrome, their socialization habits are not a problem.

On the other hand, we have seen people who clearly do not want to associate with people with disabilities and have a negative view of Down syndrome. Many of these individuals view themselves as different from others with disabilities, and they may actually say that they are "not like them" (referring to others with Down syndrome or other intellectual disabilities).

There is a wide range of skills in people with intellectual disabilities, and some higher functioning individuals have reported feeling lumped in with those with lower cognitive abilities and "judged" as having fewer skills. This can be a realistic concern if they do not have opportunities to use the skills they have or are only offered opportunities that all the individuals with intellectual disabilities are capable of. Some adults with Down syndrome express that they are not comfortable with the actions or noises made by some others with intellectual disabilities. One person described a fear or concern with the "unpredictability" of individuals with an intellectual disability. The result for these individuals can be a complicated mixture of frustration at having their skills prejudged based on the diagnosis of Down syndrome (rather than being assessed as individuals), discomfort at being around other people with intellectual disabilities, and a lack of acceptance or a type of rejection of self (some have even called this a self-hatred), which leads to poor self-esteem. For people with Down syndrome, this identity issue may have a significant effect on their lives.

Unfortunately, counseling is often difficult for individuals who have identity issues because they may be very reluctant to discuss or acknowledge the fact that they have Down syndrome. For example, Sam voiced his lack of acceptance by stating

that he wanted a cure for “it” (Down syndrome). This admission was actually a breakthrough in counseling because for months he wouldn’t even admit that “it” existed. What harm comes from these negative feelings about Down syndrome? These individuals choose not to associate with people with Down syndrome and other intellectual disabilities but often have trouble being accepted by people without intellectual disabilities. Consequently, they may exist in a limbo of loneliness and despair because they are often cut off from the people they want to be with and choose to cut themselves off from people with disabilities.

Additionally, if they do not understand or accept their own limitations, many of these individuals have problems in the workplace. For example, we have seen many people who lost good jobs (in offices, grocery stores, etc.) because they did not feel the job was up to par when compared to jobs siblings or others without disabilities had (as executives, lawyers, doctors, etc.). From this perspective, no job will be acceptable because no job will be good enough compared to what others have attained. In addition, the inclusive opportunities in school are often not matched by inclusive opportunities in the workforce. Opportunities postschool, which often don’t match the quality of opportunities in school, are a real problem and can contribute to the challenge these individuals face. The job failures that result only add to the person’s sense of despair and poor self-esteem.

Although acceptance or identity issues can be difficult to treat, there are a few factors that may increase or decrease the success of counseling. One of the most important issues is whether the person’s family accepts the Down syndrome. Lack of acceptance may be manifested in a myriad of ways, such as when the family avoids or makes negative comments about people with Down syndrome or when they keep the family member with Down syndrome from participating in social gatherings or community outings because of embarrassment, etc. If the family has these types of acceptance issues, then our job is far more difficult, and we are less optimistic about the outcome of treatment. Often these families ask us to treat the symptoms (depression, despair, etc.) without discussing the real cause of the problem: the lack of acceptance of the person’s Down syndrome.

Another challenge arises if the individual does not have an opportunity to use her skills. While being a lawyer or a physician like her sister may not be reasonable for a woman with Down syndrome, working to find or develop opportunities in which she can fully use her skills is a key element. In the past we looked at dissatisfaction with job opportunities only as an acceptance issue. It is now clear that there is often an identity issue in that the individual is realistically appraising her situation and

finding it does not meet her expectations. With the expansion of opportunities for people with Down syndrome comes the opportunity to explore how ideas such as expanding horizons, safety, personal realities, and the need to allocate resources all come together. Exploring these issues can be an important piece of counseling. What can be changed? What must be accepted?

When an adult with Down syndrome struggles with acceptance, we have had some success in turning the tide of acceptance. This happens for two reasons. First, through counseling and exposure to other individuals with Down syndrome while at the Center, we are able to promote a positive view of Down syndrome. This may occur even if the issue of Down syndrome is not discussed directly, at least not in the early stages of counseling. Second, we have found that people may become more receptive to the message that it is OK to have Down syndrome as they age and mature. In these situations, patience and persistence will pay off, as in this example:

Judd has been followed in the clinic for many years. Over this period, he has come in at various times with complaints of loneliness, depression, and problems keeping jobs, which were all related to his lack of acceptance of Down syndrome. Each time he came for counseling, we listened and supported him, while we also promoted a positive view of Down syndrome and of his own unique talents and resources (despite and because of his Down syndrome). We encouraged him to view his problems as solvable if he could accept himself, appreciate his talents, and figure out ways to use them.

After years of pain and hardships, Judd has finally begun to show signs of self-acceptance and pride. One thing that helped to turn the tide for him was a successful job experience, after a string of job failures. Interestingly, this job involved assisting people with physical disabilities in a rehabilitation hospital. We recommended Judd for the job because we had found this to be a good experience for a number of others who had acceptance issues. Like the others, Judd did quite well in this job, perhaps because the supervisors were familiar with people with physical and intellectual disabilities and were very patient and encouraging. In addition, the act of helping someone else was transformative for Judd, just as it is for all of us.

Helping others is not an experience many people with Down syndrome have. They are taken care of but are less commonly given the opportunity to care for others, even though people with Down syndrome are often very sensitive and responsive to others. Judd responded to this opportunity by working patiently and sensitively to help others in his job. For this, his supervisors gave him praise and acknowledgment, which he sorely needed. Equally important, he developed a greater understanding of, and level of comfort with, the idea of disabilities, which he could use to view his own disability more positively.

Patience and persistence may also pay off in our work with families who have acceptance issues. Families who are initially reluctant to accept Down syndrome may become more accepting over time with exposure to the Center and other families. In effect, the meetings at the Center become a type of family counseling related to acceptance issues even if not formally defined as such. This is very important because, if there is acceptance in the family, we are far more likely to resolve an acceptance issues in the family member with Down syndrome. One reason is that these accepting families often encourage the family member with Down syndrome to participate in social and recreation activities with people with disabilities, despite whatever reluctance the person with Down syndrome has to do so. These individuals often learn to "tolerate" social events with peers with disabilities, and, over time, may actually develop friendships.

Our experience is that people may initially choose to stay to themselves when they attend social and recreation activities. Usually this does not last, as other participants begin to talk to them and they cannot help but respond when this includes a team activity. For example, one young man was initially aloof with his softball team but when his group began to compete for Special Olympics medals, he was caught up in the team effort. Another adult became more social because he was courted by a very friendly and attractive young woman with Down syndrome. Similarly, Judd's boss encouraged him to attend Special Olympics, and he has become more and more involved in these types of activities as a result.

We have seen some adults with Down syndrome who have difficulty accepting that they cannot drive, go to college, get married, and have their own life like siblings or peers. Here is an example of a woman who struggled with these acceptance issues:

Bridget "made a mess" (in her family's words) during her younger sister's wedding. She was a bridesmaid but was in a foul mood and would not participate in the dancing or other festivities at the reception, even though she loved parties and dancing. Her siblings and parents tried to talk to her and bring her out of her bad mood, but she simply could not talk or change her mood for the wedding. Over six weeks later, Bridget was able to explain to her sister, Colleen, that she was upset that her sister would have a family, career, and independence, while she would not. Colleen was very sympathetic to her and brought Bridget to discuss this further in counseling.

Over a number of sessions, Bridget, her counselor, and Colleen discussed Bridget's dreams and her limitations. Eventually, she was able to understand that others also had dreams that were not going to be fulfilled. For example, her sister admitted that she had not married her childhood sweetheart and that she had not been able to go to medical school or be a singer in a rock band as she had always dreamed of doing. Still, she was able to find happiness in what she was able to do and become. Later, the counseling shifted from focusing on what Bridget was unable to do to what was possible.

Bridget decided that she had three key goals for her life: (1) to go to college, (2) to find a good job, and (3) to live independently. She, like her sister, found good enough solutions to these goals in time by (1) attending courses at a college, (2) finding an enjoyable and good-paying job in a grocery store, and (3) moving to an apartment where she was given as much independence and support as she needed (by a good agency serving the needs of persons with disabilities). Bridget was seen at a follow-up counseling appointment after several years' absence and admitted that she still sometimes wished she was able to have the life her sister had but that she was much more content now and prouder of what she was able to do.

In many respects the counseling that Bridget or others with Down syndrome need is not different from the counseling anyone else receives when trying to come to terms with their own dreams and the reality of their limitations. For people with

Down syndrome, the counseling may simply be more focused on a different standard of attainment, but a standard that still challenges them to make the best use of the skills and talents they have.

Family Counseling

Because of the critical role families play in the lives of people with Down syndrome, this section offers an extended discussion of family counseling approaches. As with individual counseling, family counseling approaches include elements of support and insight. Goals of providing a good counseling experience for the family include fostering the feeling that they and their family member with Down syndrome are welcome, valued, and understood. Furthermore, the family's feelings and opinions are heard and their expertise as caregivers and advocates is greatly respected.

The psychosocial interview with the caregivers and the person with Down syndrome may explore substantive issues and concerns, which may be very insightful to the family. This may include sensitive issues as well as family concerns that may result in the person with Down syndrome feeling criticized or scrutinized. Often, what happens in the ensuing assessment process is a type of family counseling that is educational in nature.

In interviews with thousands of families since 1992, we have identified many issues of key importance in understanding people with Down syndrome. During the course of the assessment we can use this knowledge to help explain what is normal or common (or not so normal or common) to the family and other caregivers. We look at this process as a way to bring to the meeting the shared wisdom of all the families and caregivers that we have seen. We have heard so many times from so many families about these issues that we can talk with authority about them. Frequently, this brings an enormous sense of relief and understanding to both the person with Down syndrome and her family. For example, if the person with Down syndrome talks out loud to herself, we are able to state that this is a common and normal behavior in most instances. Similarly, we often discuss that the need to follow set routines and patterns may have many benefits. In regard to these and other issues discussed in this book, we become a source of insight for the families and for the person with Down syndrome. Equally important, we can provide these families with insights and strategies shared by thousands of other families, which may solve problems resulting from such issues as self-talk, grooves, etc.

What transpires in these meetings, then, is a type of counseling that helps to normalize and educate people with Down syndrome and their families based on the wisdom and expertise of other families.

Not all families have access to a center serving the needs of adults with Down syndrome, but there are other ways of obtaining education and support. For example, you may attend conferences offered by local or national Down syndrome parent groups. You may benefit from the educational workshops as well as from the process of exchanging information and experiences with other parents and caregivers at the conference. We have found that when we offer group counseling sessions to people with Down syndrome, there are frequently spontaneous support group meetings between the parents of these participants. Often when we notice this is happening, parents agree to have their own counselor present to facilitate these meetings. We have also found that whenever parents get together to meet and talk, they often exchange useful information and support.

Supportive Family Counseling

When the person with Down syndrome has more serious problems, supportive family counseling may be essential to the success of the treatment process. For example, when an adult with Down syndrome has severe depression and is quite withdrawn, family members often curtail their own social and work activities to care for the adult. In effect, the whole family is at risk for depression because of the problem. In these and other related situations we have learned that supporting the family is critically important. After all, family caregivers usually play a vital role in the person's life even when there are no problems. When there is a mental health problem, they have an even more important role in supporting the treatment and recovery process. If the family is overwhelmed and stressed, then the person with Down syndrome will also be overwhelmed and stressed and the severe problems will continue.

In dealing with these issues, it is important to act as quickly as possible. The number one goal of treatment is to get the person with Down syndrome to return to her normal social and occupational schedule so that the family can do the same. Getting back to a more normal schedule allows the family to continue to be a strong base that supports the gains made by the depressed adult. Unfortunately, some employers are reluctant to allow employees with mental health issues to return to work, particularly if there are more severe-appearing symptoms, such as agitated

self-talk (yelling at people who are not there). We often advocate for the person to go back even if severe symptoms are still present. We try to convince staff that this behavior is not uncommon for people with Down syndrome who have depression and that it will go away with time. What we, and the agencies, have found is that once people are back in a regular routine, much of the "crazy behavior" dissipates as they become absorbed in their work and the social activities that surround them on the job. On the other hand, some individuals need to go back more slowly, step-by-step, feeling their way and feeling safe with each step. Those individuals may not be able to go back until the symptoms have sufficiently abated.

Other types of problems in a family member with Down syndrome may be as challenging for families as depression, but for different reasons. For example, people with Down syndrome with more severe obsessive-compulsive behaviors may have rituals that control and upset the whole family's routines. In this case, counseling helps the family to learn the best ways to understand and respond to compulsive behaviors. For example, they learn that anger or attempts to stop the behavior may actually make things worse, whereas gently diverting the behavior to something else may be far more productive. This process helps the family learn how to reduce the rigidity and intensity of the person's rituals, which helps them get back to normal.

The third reason for family counseling is to resolve conflicts between members of a family or group that affect the person with Down syndrome. The counseling approaches that deal with these problems look at a family as a system of relationships that have a mutual influence on each other. This influence is generally beneficial, such as in the case of parents who take care of the emotional and material needs of someone with Down syndrome. However, problems occur when there is conflict between parents or other caregivers. It seems that the greater the dependence of the child or adult on the caregivers, and the greater the intensity and duration of the conflict between caregivers, the greater the stress for the person with Down syndrome in their care, as shown in this example:

> *Andre, age twenty-eight, was brought for evaluation by his parents and several older siblings. He had become increasingly withdrawn and depressed and would often not get out of bed, even to go to work or to social activities he had previously enjoyed. It turned out that his parents had a bitter, long-standing conflict, but they decided not to divorce because of religious convictions. Andre's three older siblings had moved*

out of the family house to go to college and to establish their own homes, while Andre stayed at home with his parents.

After the last of Andre's siblings moved out, the conflict between his parents had worsened. His father dealt with the conflict by spending more time at work, and his mother by focusing more of her attention on Andre. During the day, Andre's mother doted on him, doing more and more of the tasks that he was able to do for himself. At night, his parents got into frequent fights that were ostensibly about him. They blamed each other for Andre's depressive symptoms: Father accusing Mother of spoiling Andre, and Mother accusing Father of abandoning Andre (and her).

As time went on, Andre had become increasingly depressed by the conflict. His refusal to get out of his bed was an indication of the depths of his despair and perhaps also his way to send a message that the situation needed to be fixed.

At the first appointment, Andre's siblings explained how the problem had come about and how they were trying to remedy the situation for Andre. Some time ago, they had put him on waiting lists for group homes in the hope that he could escape from the parental conflict (like they had). In fact, there was an opening available in a desirable group home, but Andre's parents were reluctant for him to proceed with the move. Andre's siblings were concerned that their parents opposed the move because they wanted to keep Andre home as a buffer for their own conflicts.

After a number of family meetings, Andre's parents agreed to let him move out. Once he was in his new residence, his depression gradually lifted. Within several months he had adapted to his new home, and in time he was back to his old self and was participating in important work and social activities.

The counseling freed Andre enough from the parental conflict to move to the group home. This occurred because the counselor was able to channel a considerable amount of the parental conflict off of Andre (as the third party in the marriage) and onto the counselor himself. To facilitate this, a number of sessions were held between the counselor and the parents without Andre being present. During these meetings, the couple agreed to follow through with a marriage therapist for intensive counseling of their own. Additionally, Andre's siblings kept him at their

own houses when their parents met for marital counseling. They were extremely helpful in setting up and following through with the residential placement. This gave Andre some needed space from his parents, which helped to prevent the parents from pulling him back into the marital conflict. They also brought Andre to many individual counseling sessions to support the process of separating from his parents' conflict.

As explained above, family and marital counseling is a specialized approach to counseling. You cannot assume that counselors who do individual treatment are trained or experienced in family or marital counseling. One way to locate a counselor with this training is through a local chapter of the American Association for Marriage and Family Therapy. This is a national group with affiliates in every state. They have a referral service representing most of the counselors with this training and may even have counselors in different locations who have experience working with the families of people with Down syndrome or other disabilities.

The Need for Goals and Outcome Measures

As we discussed in the introduction of this chapter, counseling should have meaningful goals and a means to determine whether these goals are reached. Behavioral approaches are very clear about defining and evaluating change. Insight and supportive counseling approaches will often emphasize more subjective goals, such as "enhanced self-esteem" or "a better attitude," etc. Often, goals for these approaches may also include more objective behavioral measures, such as increased participation in beneficial activities, more smiling, etc. Outcome may also be measured as a decrease in negative emotions, such as angry outbursts.

Close family members or caregivers can often assess whether counseling goals are achieved, since they are often excellent observers of the person with Down syndrome in their care. If the counseling is helpful, caregivers will notice a definite change in some key areas of the person's mood, temperament, or behavior. The counselor should discuss outcome goals very clearly at the start of the counseling, and progress toward goals should be monitored throughout the course of the counseling.

Some counselors may also use standard assessment tools, which are normed for people with Down syndrome and other intellectual disabilities, to help them assess mental health symptoms and maladaptive behaviors. These tools may also be used at

the end of treatment to assess improvement. The most widely used assessment tool for this purpose is the *Reiss Screen for Maladaptive Behavior*. Some counselors may also use maladaptive scales from the following assessment tools: *The Inventory of Client and Agency Planning* (ICAP); the *American Association of Mental Retardation Adaptive Behavior Scales* (AAMR ABS); and the *Scales for Independent Behavior* (SIB-R). These measures may be especially helpful to clinicians who have less experience with diagnosing and treating people with Down syndrome.

Counseling Benefits for Less Verbal People

Some counselors question the benefits of counseling for people with Down syndrome who use little to no speech to communicate. In our experience, counseling may be conducted very successfully with this group through the creative use of nonverbal mediums such as signing, pointing, and pantomime. This may also include the use of augmentative communication devices such as talking computers and lower tech devices such as picture books, written notes, etc. In other words, a counselor may use whatever medium someone customarily uses to communicate with others.

Parents and other caregivers often play an important role as intermediaries in this process. They may help to interpret nonverbal communication, particularly more idiosyncratic gestures and communications that the person uses. They may also give a description of relevant history and of the day-to-day events that transpire between sessions. Here is an example:

> *Molly, a young woman with Down syndrome, had been hurt when her boyfriend chose another woman over her. Molly had relatively limited verbal skills, but her adoptive mother, Joan, told us Molly's story. As she explained, Molly was very sensitive to this type of loss. She had been put up for adoption at birth because her parents could not accept her Down syndrome. Molly was aware of this fact because she continued to see a grandmother, several siblings, and an aunt who were accepting and supportive of her. While her relationships with these family members and her adoptive family were very loving, her feelings of loss from her parents were still strong. This was particularly the case when she experienced a new loss, such as of her boyfriend.*

Despite her limited speech skills, in counseling Molly was very expressive with facial expressions, gestures, and written notes. Additionally, her adoptive mother helped explain the historical issues and the events that occurred during the two to three weeks between sessions. Molly's adoptive mother often sat in with her and the counselor for approximately ten or fifteen minutes prior to their individual meetings. This gave the counselor reference points that Molly and her counselor could discuss during their sessions. For example, the counselor could ask Molly how she experienced an event, and Molly would communicate her response through facial expressions or in writing.

According to Joan, the sessions were very helpful for Molly. Molly had her own unique sign to indicate she wanted to continue the counseling. She expressed this by moving her hands from her heart outward. She continued to use this sign for approximately three months until she felt she no longer needed the sessions.

Although parental input is usually helpful for people with limited speech, there are several circumstances in which parents should not be used as interpreters. First, parental input may be counterproductive if the person with Down syndrome wants to vent about her parents or if they are a part of the person's problem. Usually, this problem is very evident to the counselor when the person with Down syndrome responds to her parent's comments and interpretations with negative facial expressions and body language. If this happens, the counselor should look for alternative sources of information from other caregivers who are part of the person's daily life and activities. For example, there may be siblings living in the family home or nearby, teachers in school, or staff or supervisors in residential, work, or recreation settings. If these individuals cannot come to the appointment, we try to go to them in order to get the information we need to better understand what the person with Down syndrome needs to communicate.

There is also another type of problem that can arise when parents act as interpreters. Some people with Down syndrome have no major problem or complaints about their parents, but they do want to speak for themselves. They often try to block their parents from speaking and try to communicate on their own. While this is laudable, this creates a problem for the counselor if the person's speech is incomprehensible. In these instances, the counselor will often find some excuse to

talk to the parent or parents before and during the individual meeting to get relevant history and help with interpreting the person's speech. In most cases, the parents are very helpful in clarifying the person's comments because they usually involve some key issues or events that occurred in the recent past.

Photographs and Memory in Counseling

We have found that photographs may be an especially rich medium to help people express thoughts and feelings in counseling. This is particularly the case for people with Down syndrome because they tend to have exceptional visual memories, whether or not they have good verbal skills. They may use pictures of past events to express a significant theme or issue. In addition, if the adults who accompany the person with Down syndrome to counseling participated in the event shown in the photograph, or are aware of the particulars of the event, they can help by describing more of the feelings and actions captured in the picture. In this way, as the expression goes, a picture truly is worth a thousand words.

For instance, in the above example of Molly, pictures from two different family gatherings helped her to talk about her feelings for her family. The first set of pictures was from her sister's wedding. Molly was not invited to the wedding because her parents were present. The wedding pictures were a poignant example of her parents' lack of acceptance of her and of the many family gatherings she could not attend. Up until she looked at the photographs, she had avoided talking about her feelings about her parents' lack of acceptance. It was much easier for her to talk about missing her boyfriend. The pictures helped her to see the real reason why she was so sad about her boyfriend.

Molly and the counselor also looked at a second set of pictures showing the baptism of her sister's baby. Molly was able to attend this event because her parents were out of the country. This set of pictures helped her appreciate the love she had from accepting family members and from her adopted family (who also attended this ceremony). This not only took the sting out of her parents' rejection of her but also allowed her to accept her situation and to get on with her life.

As this example shows, pictures may be an excellent therapeutic tool for people with Down syndrome, with or without verbal limitations. They may help people to work through losses and problems, as well as to accept the positive resources and supports they have in their lives.

Art and Music Therapy

Art and music therapy are types of counseling that use other rich mediums to help people express themselves, whether or not they have verbal skills. We have seen great benefit to people from having individual and group sessions with trained and sensitive art and music therapists. People who participate in these types of counseling do not have to be accomplished in either art or music to benefit.

Art therapists teach people how to paint, create sculptures, or use other forms of art to express themselves in a nonjudgmental and supportive environment. They then help the person to identify the themes and feelings expressed through her art. Similarly, music therapists help people, in a supportive and nonjudgmental setting, to produce music using a wide variety of percussion and musical instruments. The therapist is often able to encourage the person to express some emotion through the music, from exhilaration and gladness to fear and sadness, and then to help the person to interpret what she is expressing through the music.

These therapies may be used effectively in individual or group therapy. In group meetings, the therapist helps people to communicate with each other as well as to the therapist.

Occupational Therapy

Another therapy that we have found to be beneficial for both mental health promotion and mental illness treatment is occupational therapy. A good occupational therapist is able to assess life activities and sensory issues that may be contributing to a change in mental health, recommend activities that promote both mental and physical health, develop and promote strategies to help the individual return to her previous level of function, and develop stress-reduction or anxiety-ameliorating measures and then practice them with the individual.

As described in the case of Janet at the beginning of this chapter, many people with Down syndrome become less involved in activities or more reluctant to leave the house during a mental illness, and this reduced activity may persist even after the other symptoms have improved. Use of pictures, social stories, videos, and schedules may help them return to typical function. In addition, social skills groups run by an occupational therapist, social worker, psychologist, or others can give the individual in-person demonstrations of and practice with social skills that may help the person regain her previous level of function.

Signs That a Counselor May Be Biased

In our experience, there is some bias in the counseling field that may make it difficult for people with Down syndrome to find knowledgeable counselors, committed to working respectfully with them to resolve their issues. When interviewing a counselor to determine whether he or she may be an appropriate counselor for a family member with Down syndrome, look for someone who does not express the following biased beliefs:

People with Down Syndrome Are Incapable of Insight

Some counselors mistakenly believe that people with Down syndrome are not capable of using insight to change their behavior. Practitioners with this bias assume that reliance on concrete forms of thought may prevent people from understanding how and why they behave certain ways.

In fact, extensive research has shown that there are many different types of intelligence and understanding that may lead to many different forms of learning. For example, one of the ways people with Down syndrome may compensate for difficulties with abstract thinking is by being very sensitive to the feelings and emotions of others (what we call "emotional radar"). They often interpret and respond to other people's behavior through this lens of understanding.

Even when people with Down syndrome show a lack of understanding of the effects of their behavior, this does not mean that they are insensitive to others or that they cannot learn. For example, it may have appeared that Teresa (above) was unaware of her behavior because of her inappropriate questions to strangers about private sexual matters. However, she readily stopped her questions when she learned that they were offensive to others and harmful to herself. What may have prevented her from changing her behavior earlier was that the strangers may not have felt comfortable telling her that her behavior was inappropriate. Her mother may also have been unable to teach her because of an existing adolescent-like conflict with her daughter. Like many adolescents, Teresa tended to rebel against anything communicated by her mother.

Modifying Someone's Behavior without Involving Her in the Process Is OK

Some behavioral counselors underestimate the ability of people with Down syndrome to understand and participate in the counseling process. This may lead to an emphasis on modifying the person's behavior without her personal involvement in the process. The person with Down syndrome may then feel that she is "done to" rather than a participant in the counseling. The counselor may not ask questions about what the person wants or needs, which may limit her personal interest and cooperation with the process. These types of problems can often be avoided if the counselor consults parents, adult siblings, or other caregivers. Even if the counselor has a bias, caregivers may be able to bring the person's feelings, preferences, strengths, and human qualities to life in the process.

Understanding the Message behind the Behavior Is Not Essential

Another more serious problem may result if an inexperienced counselor looks only at a person's behavior and not at the reasons for the behavior. As we have stressed throughout this book, many people with Down syndrome cannot easily verbalize thoughts and feelings, so they may sometimes rely on behavior as their primary means for communication. Unfortunately, inexperienced or uninformed counselors may not look for the message in the behavior. They may downplay the notion that the person with Down syndrome is aware of and responsive to her environment. Consequently, they may believe that all that is necessary is to identify a problem behavior and eliminate it, as in the example below:

Gina was refusing to go to work in the morning. Staff in her group home had attempted a variety of strategies, ranging from patient encouragement to forceful insistence, without success. An agency behaviorist then proposed that Gina be rewarded for returning to work with a Coke and a token. If she earned five tokens, she was to be rewarded with an outing with her favorite staff person. Unfortunately, this, too, did not seem to make any difference to her.

The real reason for Gina's refusal was discovered when her sister accompanied her to an appointment with her therapist. Unbeknownst to group home staff, Gina had had a bowel accident in her sister's car during a recent weekend visit. (Gina had lactose intolerance, which resulted in occasional diarrhea.) The accident in her sister's car was very upsetting to

Gina because she was very meticulous with her hygiene. She began to fear that car rides would put her at risk for more embarrassing accidents, especially on longer rides, such as between the group home and her workplace.

Once we understood the reason for Gina's behavior, we developed a plan that included adjusting her diet to better control her lactose intolerance and making regular stops at bathrooms on the route to work. Gina was shown pictures of the route with the bathrooms at different sites highlighted (bathrooms were located at several fast food restaurants, a doughnut shop, and several other agency sites). The plan included gradually increasing the distance between stops, with each bathroom on the route representing a checkpoint. Interestingly, Gina never needed to use the bathrooms during the process. She merely wanted some assurance that she could if she needed too.

Staff continued to stop at each bathroom on the route for many months to appease her sense of anxiety. Additionally, for any other outings into the community, staff made sure that Gina had a plan for bathrooms on the way to allow her to feel comfortable with the outing. This overall plan has now worked well for Gina and the staff at her group home for many years.

Clearly, behavior is frequently a nonverbal message that must be understood before any intervention is attempted. Otherwise, the results may be less than satisfactory.

PART 2: WHEN MEDICATION IS NECESSARY

Medications can also be a very important part of treatment for mental health conditions. Sometimes, however, parents or other caregivers are reluctant to consent to the use of "brain altering" medications or think of medications as a treatment of last resort. In reality, however, certain mental health conditions are related to altered brain chemistry and using medications to restore brain chemistry to normal may be an essential part of treatment.

This part of the chapter will help you understand why medications may be medically necessary for people with mental health conditions in general, as well as

why they may be helpful for people with Down syndrome in specific. In addition, this section covers several issues that must also be addressed before prescribing a medication:

- obtaining consent,
- discussing the philosophy of the use of medication, particularly in a person who may have a limited ability to understand the implications of the treatment, and
- developing a plan that includes medications as one part of an overall treatment strategy.

The medications used to treat specific mental health conditions are discussed in the chapters on those disorders (chapters 17–27).

Brain Chemistry and Mental Health

Most medications that are prescribed for mental health conditions have an effect on the chemicals in the brain. For you to understand how these medications work, you need a basic understanding of how brain chemicals or neurotransmitters communicate with one another and the specific functions of some of the brain chemicals that play a role in certain mental health disorders.

Neurotransmitters

Our central nervous system (brain and spinal cord) and peripheral nervous system (nerves outside the brain and spinal cord) contain millions of microscopic nerve cells called *neurons*. When a nerve cell is stimulated, a small electrical current is generated at one end. This current travels along the nerve cell. This current must be transferred to the next nerve cell (and then the next and the next) in order to send a signal from one part of the nervous system to another. The cells are separated by small gaps called synapses. The first cell must send the message to the next cell, which is done through chemicals. The electrical signal causes the release of a chemical that crosses the synapse and causes an electrical change in the next cell. The signal is carried along the next cell and then crosses the next synapse until it reaches its destination. For example, if you want to move your left arm, an electrical change occurs in the nerve cells in the part of the brain that controls left arm movement. The signal passes along cells and across synapses until it reaches the left

arm, where the electrical signal has an effect on the muscles in the left arm, causing the appropriate, desired movement.

Neurotransmitters are the chemicals that the brain and peripheral nervous system use to communicate from one nerve cell to another. A neurotransmitter is released from the sending (*efferent*) end of the first cell, crosses the microscopic space between the cells (the *synapse*), and then attaches or "binds" to the receiving (*afferent*) end of the next nerve cell. The neurotransmitter binds to a receptor on the second cell. The act of binding causes the small electrical change to occur. Depending on the cell, the receptor, and the neurotransmitter, the effect of the electrical charge may be either (1) *excitatory*—it excites the cell (increases activity in the cell), or (2) *inhibitory*—it inhibits the cell (decreases activity in the cell).

Again, depending on the cell, the receptor, and the neurotransmitter, the effect may be rapid or slow. After the neurotransmitter binds to the cell and causes the change in the cell's electrical activity, the neurotransmitter is broken down by other chemicals or reabsorbed. This stops the electrical change, and the cell can prepare for another message from the adjoining cell(s).

After the neurotransmitter has been bound to the next cell for a short time, often the effect of the binding stops. Therefore, if the neurotransmitter is not broken down, the receiving cell cannot "reload" electrically to get ready to fire again when another molecule of the neurotransmitter binds to its receptor. If the neurotransmitter is not removed or broken down, additional neurotransmitter does not have any additional effect on the nerve cell.

Medications can be used to change this process at any of the steps or sites. Medications can affect the neurotransmitter level between the cells by increasing or decreasing the release of the neurotransmitter or the breakdown of the neurotransmitter. The sensitivity of the receptor of the cell receiving the message can also be affected. If the receptor is made more sensitive, then either less neurotransmitter is required to obtain the same effect in the cell or the same amount of neurotransmitter has greater effect. In addition, if the receptor binds the neurotransmitter more tightly, the neurotransmitter cannot be released. If it is not released, more neurotransmitter cannot bind or cause the electrical change. This reduces the passage of the next signal from the first cell.

Types of Neurotransmitters

There are several types of neurotransmitters in the brain. Each has one or more functions. The function is determined by the part of the brain the cell is in, the amount of the neurotransmitter in that area of the brain, the relationship to the other neurotransmitters in the brain, the type of receptors, and other factors. All these issues complicate the use of medications.

Below are the neurotransmitters that play a role in some of the most common mental health disorders. Although it is difficult to directly measure the level and activity of neurotransmitters in the brain, researchers have deduced that many mental health disorders are caused either by deficiencies or excesses of one or more of these brain chemicals.

Glutamic Acid. Glutamic acid (glutamate) is the most common excitatory neurotransmitter in the brain. It appears to play a role in learning and memory. One glutamic receptor is called the NMDA (N-Methyl-D-Aspartate) receptor. Excess glutamic acid binding to the NMDA receptor can cause toxicity and has been implicated in destruction of neurons (nerve cells) in such disorders as Alzheimer's disease.

Gamma Amino Butyric Acid. Gamma amino butyric acid (GABA) is an inhibitory neurotransmitter. It is the most common inhibitory transmitter in the brain. It reduces signals in the brain and thus helps prevent overstimulation of the brain. It appears to have both an antiseizure and calming effect on the brain.

Acetylcholine. Acetylcholine is the main neurotransmitter in the peripheral nervous system, but there are relatively few acetylcholine (cholinergic) receptors in the brain. It is usually an excitatory neurotransmitter. It is the main neurotransmitter controlling skeletal muscles, so it is needed for making most voluntary movements. It is also used by part of the nervous system (parasympathetic nervous system) that controls involuntary functions such as bowel and bladder function, digestion, etc. (Norepinephrine is the other neurotransmitter used in the peripheral nervous system and is described below.) An enzyme called acetylcholinesterase breaks down acetylcholine. Blocking this enzyme with medications results in a greater effect of acetylcholine. Although this neurotransmitter is only present in small amounts in the brain, the neurons with acetylcholine play an important role in Alzheimer's disease. The destruction of these nerve cells in certain parts of the brain is involved in the development of some of the symptoms seen in AD.

Dopamine. Dopamine can act as either an inhibitory or excitatory neurotransmitter, depending on the location of the cell in the brain. Depending on

the location of the cells, dopamine facilitates the body's posture, the speed that muscles move, attention, and feelings of pleasure.

Norepinephrine. Norepinephrine is also known as noradrenaline. Like acetylcholine, it is found in both the peripheral nervous system and the brain. It is synthesized from dopamine, so levels of norepinephrine in the brain are directly linked to levels of dopamine. If dopamine is increased, norepinephrine will increase; if dopamine is decreased, norepinephrine will decrease. So, it's important to keep this in mind when prescribing a drug that increases dopamine. In the part of the brain where over 40 percent of the neurons that contain norepinephrine are found, electrical stimulation causes increased arousal and attention. This area of the brain has been identified as the pleasure center. Both norepinephrine and dopamine play a role in attention, arousal, and feelings of pleasure. If norepinephrine levels are too high, anxiety can result. Conversely, norepinephrine is lower during sleep.

Serotonin. Most of the serotonin in the body is found outside the brain, primarily in the blood. However, it is hypothesized that the small amount in the brain plays a large role in mental health. Serotonin is synthesized from the amino acid tryptophan and is used to synthesize melatonin in the part of the brain called the pineal gland. Melatonin, in turn, plays an important role in regulating daily sleep patterns. Too much melatonin can lead to drowsiness; too little, to problems falling and staying asleep.

Serotonin plays a role in pain perception and in mood. When serotonin levels are too low, it can contribute to anxiety, impulsive (and violent) behavior, and depression. Aggressive behavior has also been linked to low serotonin levels.

Endorphins. Endorphins are inhibitory neurotransmitters that reduce pain and can give a sense of euphoria. This sense of euphoria may be what leads some people to seek out self-injury. Because when you hurt yourself, endorphins increase to reduce the pain, the increased endorphins can be pleasurable to some people. Opiate drugs such as morphine, codeine, and other narcotics work by binding to these receptor sites.

Folic Acid/Folate. While not a neurotransmitter itself, folic acid/folate is an important building block of neurotransmitters. Folate and folic acid are consumed in the diet or folic acid is taken as a supplement and converted to the active form, methylfolate, in the body. Unfortunately, some medications and some genetic conditions may block the conversion to methylfolate. For example, the MTHFR gene provides instructions for making an enzyme called methylenetetrahydrofolate reductase. This enzyme is part of the multistep process that converts the amino acid

homocysteine to another amino acid, methionine. The body uses methionine to make proteins and other important compounds. The inability to complete these conversions has implications for building neurotransmitters and the treatment with medications that affect neurotransmitters. Individuals with the enzyme deficiency may require supplements with methylfolate (rather than folic acid found in most vitamin supplements).

Brain Chemistry in People with Down Syndrome

There are differences in neurotransmitters and brain structure in people with Down syndrome. Some of these differences mean that people with Down syndrome may be at increased risk for mental health conditions linked to particular neurotransmitter abnormalities. That is, there are biochemical reasons for some of the cognitive, emotional, and behavioral difficulties that people with Down syndrome may experience. In addition, people with Down syndrome have fewer nerve cells in their brains than is typical, as well as fewer connections between the nerve cells. This means there are fewer nerve cells to produce neurotransmitters, and fewer places for the brain chemicals that are produced to bind. This is more evident in some parts of the brain than others; therefore, the amounts of some neurotransmitters are affected more than others. For example, one group of researchers found that the brains of fetuses with Down syndrome had reduced levels of serotonin, gamma-aminobutyric acid, taurine, and dopamine in the frontal cortex (Whittle et al., 2007).

Imbalances of neurotransmitters are believed to be involved in a number of mental health conditions. We say “involved” rather than “the cause” because more than one factor is thought to be involved in causing most of these disorders. For example, depression is believed to be connected with deficiencies in serotonin and the sensitivity of serotonin receptors. But we also know that depression can be related to life events. So, if a person with Down syndrome has reduced levels of certain neurotransmitters as a result of having Down syndrome, she may be more susceptible to disorders that are linked to deficiencies of those neurotransmitters or imbalances between those and other neurotransmitters.

In addition, people with Down syndrome have differences in the neurotransmitter receptor proteins. The receptor proteins are the parts of the receptor where the neurotransmitter attaches. The different receptors may be more or less able to bind to the different neurotransmitters in people with Down syndrome.

It is important to point out that the differences in neurotransmitters may not be the only reason that some mental health conditions are more common in people with Down syndrome. Due to differences in brain chemistry, some have accepted that more frequent mental health problems are inevitable. While differences in brain chemistry probably do play a role, we can also take the same steps to promote mental health as we do for people without Down syndrome—namely, addressing educational, recreational, social, occupational, and other needs and opportunities. Historically, inadequate attention to these needs has very likely contributed to increased incidence of some mental health conditions. Attending to those needs is not only appropriate from an ethical standpoint but also likely to improve the mental health of people with Down syndrome.

The next section briefly reviews how neurotransmitter abnormalities play a role in several common mental health disorders, with special reference to the neurotransmitters that are known or believed to be affected by Down syndrome.

Neurotransmitters in Diseases and Conditions

Depression

The role of neurotransmitters in depression has not been completely delineated. It has been hypothesized that an imbalance of serotonin, norepinephrine, and dopamine plays a role. This is based largely on the fact that changing the level of these neurotransmitters with medications can successfully treat depression. Another clue is that contraceptives with high levels of estrogen can cause depression, apparently by lowering serotonin levels in the brain.

As discussed in chapter 17, depression is one of the most common mental health concerns among adolescents and adults with Down syndrome. The reasons are complex, as explained in that chapter, but reduced levels of these neurotransmitters in the brain probably play a role for at least some individuals with Down syndrome who develop depression.

One class of medications that can effectively treat depression is the tricyclic antidepressants. Some medications in this class increase norepinephrine by inhibiting (blocking or slowing) its reuptake. Others affect both norepinephrine and serotonin. It is interesting to note that while the medications immediately increase the neurotransmitters, it may take several weeks for the person to become less depressed. The antidepressant effect may actually come from a modification of the

receptors that occurs over time, rather than from the more immediate effect on the level of the neurotransmitter.

Another class of medications, the serotonin reuptake inhibitors (SSRIs), can also be part of a successful treatment plan for depression. The SSRIs reduce the reuptake (recycling) of serotonin back into the first (efferent) cell. The net effect is to increase the serotonin in the synapse. Similar to the tricyclic antidepressants, the effect on the level of serotonin is rather immediate, but the effect on depression can be delayed. This again suggests a possible role of the medications in modifying the receptors. We sometimes see a brief (and unexpected) improvement in the first few days, followed by a return of the symptoms. Then a few weeks later, the expected improvement occurs. This may reflect the initial effect on the level of serotonin and later the effect on the receptors.

A third class of medications used to treat depression—the serotonin-norepinephrine reuptake inhibitors (SNRI) (e.g., duloxetine/Cymbalta and venlafaxine/Effexor)—inhibit the reuptake of serotonin as well as norepinephrine and dopamine. That is, they block or slow down the recycling of these neurotransmitters in the brain, increasing their availability. Another class of medications is the serotonin modulators, which are thought to inhibit serotonin reuptake and turn on certain serotonin receptors. Medications in this class include vilazodone (Viibryd), trazodone, and vortioxetine (Trintellix). Finally, depression may also be treated with norepinephrine-dopamine reuptake inhibitors (NDRI) such as bupropion (Wellbutrin). These medications block reuptake of norepinephrine and dopamine, increasing the availability of those neurotransmitters in the brain.

See chapter 17 for more information about medical treatments for depression.

Anxiety

People with anxiety disorders are thought to have imbalances of the neurotransmitters norepinephrine, serotonin, and GABA. Cholecystokinin may also play a role. Since people with Down syndrome tend to have abnormal levels of norepinephrine and serotonin in their brains, it makes sense that anxiety disorders are fairly common.

As discussed in chapter 18, there are several types/classes of medications that can help normalize the levels of these neurotransmitters, often reducing anxiety. The benzodiazepines, such as diazepam, (Valium), work to increase the inhibitory effect of GABA. This decreases neuron activity (reducing anxiety) and also reduces the

release of norepinephrine. The selective serotonin reuptake inhibitors (SSRIs) such as paroxetine (Paxil) increase serotonin, which can also reduce anxiety.

Alzheimer's Disease

Alzheimer's disease, one of the most common causes of memory loss and dementia in older adults, occurs more often in people with Down syndrome. Furthermore, adults with Down syndrome tend to develop symptoms of the disease at an earlier age than others, sometimes as early as their forties.

Alzheimer's disease results in changes in a number of neurotransmitters, as well as other changes. Although we still do not understand completely what causes the disease, some research indicates that excess glutamic acid overexcites the NMDA receptors and can lead to toxicity and neuron destruction. There is also a reduction in neurons that use norepinephrine and serotonin. In addition, there is a reduction in cells that use the neurotransmitter acetylcholine. We know that when the effects of acetylcholine in the brain are blocked in otherwise normal individuals, it results in memory loss. The reduction of these cells may, therefore, play a role in memory loss in Alzheimer's disease. Alzheimer's disease has a complicated effect on the brain, and much study is ongoing to understand its causes and effect.

The medications donepezil (Aricept), galantamine (Razadyne), and rivastigmine (Exelon) are cholinesterase inhibitors. They block the enzyme that breaks down acetylcholine. By blocking this enzyme, the acetylcholine remains active longer, which improves the function of the cells that use acetylcholine. This does not cure or change the course of Alzheimer's disease, but it can temporarily improve the cognitive function of some people with Alzheimer's disease. Studies in people with Down syndrome with Alzheimer's disease and our clinical experiences have not shown significant benefit to using these medications.

Memantine (Namenda) blocks NMDA receptors. This reduces the overexcitation of the cells and may also slow cell destruction. Unfortunately, studies (and much experience) have not demonstrated much, if any, benefit of this medication in people with Down syndrome who have Alzheimer's disease.

Research into Alzheimer's Medications and Down Syndrome

Donepezil (Aricept) has been assessed in a few small studies in people with Down syndrome who don't have Alzheimer's disease. Some of these people showed

improvements in speech skills. In our small study, donepezil seemed to increase the amount of speech that the person was capable of but didn't help articulation. In other words, a few people who ordinarily did not speak much talked more when taking donepezil. Although their articulation did not improve, their increased speech helped them interact with others, make their interests and wants known, etc.

After our study was completed, some of the subjects chose to continue the donepezil. However, we did not notice any clinical benefit for those who had not experienced benefits during the study. For those who had benefited during the study, the benefit faded over several weeks to months, and their speech returned to the level it had been before starting the medication. Other, larger studies have also found that using donepezil did not significantly improve cognitive or speech function in people with Down syndrome (who don't have Alzheimer's disease).

Medications for People with Down Syndrome: The Art and Science

The practice of medicine has long been described as both an art and a science. Anyone who prescribes medication for someone with Down syndrome who has mental health or behavior issues must, of course, understand the science of medications, neurotransmitters, brain chemistry, and the particular issues in people with Down syndrome. But the art of medicine also comes into play for a number of reasons: each person is different; medications work differently in different people; different situations demand alternative approaches; and sometimes the expected result is not what occurs.

Some of the questions that need to be answered have more to do with clinical nuances or idiosyncrasies of the patient. Issues we consider include the following:

1. Is it a good idea to try medication?
2. How can the person with Down syndrome be involved in the decision as to whether to use medication?
3. Which medication should be used and at what dose (which may depend on administration issues, side effects, need for trial and error)?
4. Should genetics testing be used to evaluate the individual's metabolism of psychotropic medication?
5. What is the best way to monitor medication use and make decisions based on the patient's response (watching for paradoxical effects, deciding when to change or discontinue the medication)?

Deciding Whether to Try Medication

Once a mental health condition is diagnosed and we recommend considering medication, there are still additional issues to be addressed. In fact, there is a step even before the medication is recommended. The first question is, "Is a medication the correct approach?" That is, is the behavior part of "normal" or "characteristic" behavior for a person with Down syndrome who is not in need of any treatment but rather just education and/or reassurance? If treatment is needed, is a medication needed at all? Therapies and behavioral approaches are addressed above. Often these approaches can be used in place of medications.

However, sometimes therapy and/or behavior approaches are not enough by themselves. This may be because the condition is beyond the person's control or beyond the control of therapy. For example, the drive to perform an obsessional behavior can be so strong that no amount of counseling or behavioral therapy may be able to overcome it. Counseling may help the person cope with the problem, avoid situations that trigger the obsession, use an alternative behavior to avoid doing the behavior, etc. However, the drive may still be there, and without reducing the drive, the person may not be able to avoid the obsessional behavior. Other times, an adult has a condition that will require medications because of the nature of the problem or the severity of the symptoms. For example, when someone is severely depressed, various therapies and behavioral approaches may help (and may actually be enough treatment). However, sometimes these approaches are not sufficient, and medication is required. Also, the severity of the symptoms often necessitates use of medications to get to a healthier place more rapidly and to ease the suffering more quickly.

We generally look at medications as therapy used in addition to counseling and/or behavioral approaches. Many times, we view the medication as helping the person reach a point where she will be able to respond to therapy or behavioral approaches. There are a number of issues that must be addressed once the practitioner reaches the decision to recommend a medication.

Involving the Patient in the Decision to Use Medication

The first issue to consider before starting an adult with Down syndrome on medication is whether she will consent to treatment with the medication. Our

approach is to recommend therapy, explain the risks and benefits, and help the person, family, or guardian make a choice that is best for them.

Sometimes people with intellectual disabilities are not able to fully participate in the decision. We make a concerted effort to make the person part of the process, though. If a patient is her own guardian, we obtain the consent from her. In addition, we ask her permission to discuss it with her family. If the patient is not her own guardian, we obtain consent from the family/guardian. We also obtain "assent" when we can from the patient who is not her own guardian. Assent is agreement on the part of the person (with Down syndrome) who is not her own guardian and therefore cannot legally consent to the medication. While not a legal agreement to take the medication, assent includes the person in the decision and makes her part of the process. She agrees that she understands why we are recommending the medication and she agrees to take it.

Having the person with Down syndrome be part of the process is not only (in our opinion) "the right thing to do" but is also critical in achieving clinical success. Although observers can give some indication of benefit and side effects, no one else can give feedback about how the adult feels on the medication. Not all adults with Down syndrome can give this feedback, but it is important to discuss the use of the medication and to obtain personal history of benefits and side effects whenever possible.

Another consideration is whether the person can manage her own medications. There are a number of questions to consider:

1. Has she been able to learn other important tasks that involve regimens that must be followed for potentially dangerous activities (if done incorrectly)?
2. Has she demonstrated that she can do these activities repeatedly?
3. Is she interested in managing her own medications and willing to do so?
4. If she lives in a residential facility, are there policies that would allow or prevent her from managing her own medications?
5. Does she understand why she is taking the medications, the potential for side effects and the need to report them, and the importance of taking them on the prescribed schedule?

If the answer to these questions is yes, the adult may be successful in managing her own medications.

If the person with Down syndrome is resistant to swallowing any medication, we try to avoid "hiding" the medication as much as possible. Again, we want to make

the person a partner in the treatment to the extent she can participate. Hiding medication may hinder the relationship between the patient and physician or between the patient and the family (or care provider) who is giving the medication. This may have a deleterious effect on treatment. In addition, we have seen adolescents and adults with Down syndrome become reluctant to eat certain foods (that the medication was put in) or even to eat at all. However, if the problem requires a medication and the guardian consents but the patient doesn't assent, it may be necessary to hide the medication or use other methods to get the person to take the medication.

Legal Guardianship

In most states in the US, a person is considered legally competent to make her own decisions once she reaches the age of eighteen. While a family may still provide support in a variety of ways, the person is legally able to make independent decisions for herself, including regarding medications. Adults with Down syndrome are considered legally competent, just like anyone else, unless a guardian is appointed for them.

Parents frequently ask us whether they should get legal guardianship for their son or daughter. We are not attorneys and don't provide legal advice, but we do discuss some of the issues that families can consider and then direct them to an attorney. The decision to obtain guardianship is technically a legal one that a judge makes (using information provided by medical professionals) after determining that a person is not legally competent.

We have patients who live very independent lives, have their own bank accounts, pay their bills, and have jobs that provide a livable income. We also have patients who are very dependent on others for much of their care. Many of our patients fit somewhere in between.

In the ideal world, none of our patients would need legal guardianship. Society would recognize that some people need more assistance and would provide it. Unfortunately, there are unscrupulous people who, instead, try to take advantage of adults with Down syndrome—sometimes in financial matters and sometimes in medical situations. We have seen some adults without a guardian who were put on a medication for a behavioral or mental health condition without their family's knowledge. This may be appropriate if the person with Down syndrome has the ability

to understand the issues of concern and the potential benefits and side effects. However, often the person with Down syndrome really had very little, if any, input into the decision.

Having a guardian provides a legal safety net to help prevent the person from being treated inappropriately. However, it may also mean that the adult loses the right to vote or do other things. There are alternative approaches to guardianship, including power of attorney for health, financial, or other legal decisions. POA grants another person the power to make decisions about specified issues (e.g., medical care) for the person with a disability. Another more recent option is a supported health care decision-making agreement. A person can select her own supporter and can change supporters at any time. The agreement is an authorization to provide support but not to take over all decision-making. Consenting to this agreement generally requires less mental capacity than understanding a contract. It does not take court involvement. The United Nations Convention on the Rights of Persons with Disabilities advocates for supported decision-making to replace power of attorney or guardianship.

We recommend exploring issues of independence, safety concerns, and the family situation before seeking guardianship, power of attorney, or another arrangement to assist an adult with Down syndrome with decision-making. There is no one answer that fits all, and an attorney can help the family make the appropriate decision for their situation.

Choosing a Medication and Dosage

As discussed in the chapters on specific mental or behavioral disorders, there are often many choices of medications for a given disorder. Sometimes, there are considerations that can help us narrow down which one might be best to try first; other times, there is no way to know for sure which medication will be most effective for this person with this condition. In a sense, we must use a trial-and-error approach to find the best choice(s) for this individual.

Genetic testing may contribute in the selection process by identifying which medications have metabolism patterns that can reduce efficacy and also assess folic acid metabolism (and the potential benefit of supplements). Genetic testing is still undergoing evaluation and is not a widely accepted standard of care. Therefore, it may not be covered by insurance. Based on observations of use in our patients, we

have found some benefit, but we have not done any rigorous research on genetic testing. As the results on ongoing research become available, we anticipate a clearer picture of the role of genetic testing in prescribing medications for mental illness.

For adults with Down syndrome, one important consideration is often whether they can swallow pills and if not, which of the available medications can be used. For example, sometimes we must use pills that can be crushed, capsules that can be opened and sprinkled on food, or medications that come in liquid forms. This may limit or alter the choice.

Another consideration is whether any of the possible medications have side effects that may be especially advantageous for a given patient. For example, sedation can be an unwanted side effect of some medications. However, for people who are having sleep disturbance as part of their symptoms, this side effect can be an advantage:

> *Thanh, age twenty-six, lived at home with her father. Her mother had died a year earlier, and for the last several months Thanh had been displaying symptoms of depression. In addition to waking during the night, she had trouble falling asleep. When we prescribed an antidepressant for her, we chose mirtazapine (Remeron), which has caused sedation in some of our patients, and had her take it in the evening. Within a few days, Thanh began to fall asleep more readily. As the full antidepressant effect was realized over the next few weeks, her other symptoms improved, including her waking during the night.*

Further discussion regarding using side effects to advantage is included in the chapters on specific conditions.

Finally, as unscientific and as bothersome as it may seem at times, there may be a certain amount of trial and error in prescribing medications for any given individual. Although there are helpful trends for treating adolescents and adults with Down syndrome and genetic testing may aid in selection of medications, individuals still may have different responses. What works for the majority may not work well for the particular individual. Likewise, although a solitary medication may work for most, a combination may be necessary for the individual who is being treated.

Wendy, a forty-two-year-old with bipolar illness, responded very well to a combination of ziprasidone (Geodon) and carbamazepine (Tegretol). This combination was reached only after a number of unsuccessful treatments. She had a reaction of tardive dyskinesia (abnormal facial movement) to multiple other similar antipsychotic medications. Once on ziprasidone (Geodon), she had an increase (although not to the previous level) of agitation. The addition of carbamazepine (Tegretol) helped tremendously.

Finding the right dose of the medication is also critical. If there is a general rule we follow, it is "start low and go slow." By that we mean start at a low dose and work slowly to higher doses. A number of other factors, however, also guide dosing the medication, including severity of symptoms, age of the patient, weight of the patient, other health problems, previous reaction to medications, family history of medication effects, and others. Some medications have standards for appropriate blood levels and, therefore, blood tests can guide dosing.

Another rule is to increase a medication to maximum dose (if additional benefit is being gained with increasing the dose) or until intolerable side effects occur before adding additional medications. We generally try to avoid using multiple medications at lower doses because this often increases side effects without getting clear therapeutic benefit from any of the medications. Occasionally, however, we find that a person with Down syndrome cannot tolerate a larger dose of any medication but will respond to a certain combination of medications at lower doses. Dosing specifics will be addressed as medications are discussed in the following chapters about specific conditions.

It is important to assess the use and the dose of the medication on an ongoing basis. Is the medication still needed? Is it a condition that, once adequately treated, is unlikely to recur when the medication is stopped? For example, someone who develops depression in response to a traumatic event and who has never had depression before might be successfully weaned off the medication after a period of time.

In addition, as people get older, go through puberty or menopause, lose or gain weight, take other medications, and experience other changes, the needed dose may change. Sometimes symptoms increase. Other times an increase in side effects will be the indicator that the dose needs to be changed.

Another reason to assess the dosage on an ongoing basis is to ensure that we are maximizing benefit and minimizing side effects. Decisions to change the dosage in this case are made in discussion with the person with Down syndrome and her family. For example, if the patient is 95 percent improved, we will consider increasing the medication but only after we discuss the potential for greater side effects that may impede the patent's recovery (and potentially reduce the net effectiveness of the medication) versus the potential for continued improvement in the condition.

Monitoring Medication Effects and Usage

Because some people with Down syndrome have difficulty accurately reporting how a medication makes them feel, it is important to plan at the outset how to monitor the medication's effects. For example, monitoring might be done by tracking behaviors before and after the medication is started by using a checklist or writing symptoms down, or by observing the patient for ourselves, in the office or at home, work, or school.

When first beginning a medication, it is important to watch for "paradoxical reactions"—or reactions that are the opposite of what is expected. For example, sometimes people become more depressed when they take an antidepressant. Some people become more agitated and anxious when taking one of the antianxiety benzodiazepine medications. It can be very difficult at times to determine whether the problem is that the condition or behavior is worsening or that it is a side effect of the medication. At times, the only choice is to discontinue the medication. We will sometimes later reintroduce the medication to see if it appears more likely that the change is a side effect of the medication.

Monitoring is also vital if any changes are made in dose, time of administration, etc. In a sense, we have found that "any change is a change." Another way to say this is that "even a good change is a change." We often see patients who have an initial negative reaction to changing the dose, time of day taken, or adding or subtracting another medication. For these patients, the behavior or condition may temporarily worsen for a few days, a week, or sometimes more. After this initial phase passes, however, the change may be positive. Whether we have the time to "wait out" the change depends on the medication, the severity of the symptoms, and the effect it is having on others.

For some patients, it is necessary to confirm that the medication has been swallowed:

Kyle, age twenty-four, was doing very well with his treatment regimen for several months. Then his original symptoms unexpectedly returned. After a few medication changes failed to improve his symptoms, his mother solved the mystery one day when she was vacuuming. She glanced into the heating vent in the floor of their living room and discovered a pile of Kyle's medication. The medication showed signs of being slightly dissolved (from a brief stay in Kyle's mouth) but was essentially intact. The previously successful treatment regimen was restarted, with Kyle's mother now confirming the pills were swallowed, and Kyle returned to good health.

Finally, it is important to monitor for potential long-term side effects. It is especially important to monitor for these effects:

- *Downregulation*, in which the neurotransmitter receptors adapt to the medication so that their sensitivity or number is reduced. This results in "tolerance" and the need for a larger dose to produce the same clinical effect.
- *Upregulation*, in which the receptors become more sensitive to the medication so that a smaller dose is needed to produce the same effect.

When upregulation or downregulation has occurred, there may also be effects when the medication is discontinued. This is because when the medication is removed (particularly suddenly), the receptors don't immediately return to their previous number or sensitivity. So, for example, if a benzodiazepine has caused downregulation, the brain has fewer or less sensitive receptors for GABA, which ordinarily inhibits anxiety. If the medication is stopped, increased anxiety can occur until the receptors regain their usual sensitivity to GABA. The increased anxiety and other symptoms that occur are withdrawal. The effects of tolerance and withdrawal demonstrate dependence on the medication.

Some people who stop taking SSRIs also have a withdrawal-like reaction. These medications should be weaned instead of stopped "cold turkey." With many medications, even if an individual doesn't have a true withdrawal on a chemical basis, reduction in the medication may make her feel different for a time and cause a sense of ill health. We have often found it necessary to wean a medication even when there isn't a recommendation on a "chemical basis."

Questions to Ask the Doctor

If you are a parent or caregiver of an adolescent or adult with Down syndrome, make sure you understand what to watch for before a medication is started. Ask the doctor the following questions:

1. What are the common side effects?
2. Should you call him or her immediately if any particular effect occurs?
3. Is it OK to discontinue the medication if an adverse reaction occurs?
4. Should the medication be taken at a certain time of day, spaced apart from other doses in a specific time frame, taken with or apart from other medications, or taken with or apart from food or drink?

Conclusion

In subsequent chapters, we discuss how the principles outlined in this chapter are applied to specific mental health problems. For each problem, assessment (and often reassessment), the use of specific medications, counseling, and other therapies, and an assessment for contributing (or causative) physical problems are described. Wherever possible, we provide examples of ways in which medications benefited patients with Down syndrome we have treated.

Return to Table of Contents

Chapter 17

Mood Disorders

Jim, age nineteen, attended his local high school. He had been included in regular education classes, participated in activities with students with and without disabilities, and enjoyed being with his family and friends. However, in the last several months, he had begun to isolate himself more, refusing to participate in activities he'd previously enjoyed, sleeping much more than previously, and always seeming hungry. He had graduated from high school in the spring several months before we met him, and his brother, Calvin, had gone off to college several weeks after his parents began to note the changes in Jim.

Our evaluation did not reveal an underlying physical health problem. We diagnosed Jim with depression. In discussions with Jim and his family, it was clear that the changes in his life were contributing factors in his depression. He was now in a transition program school that was very different from his high school program. Although initially his brother's departure was discounted as a contributing cause because Jim's symptoms started before Calvin left, it was clear that all the college preparations had begun to affect Jim even before Calvin left home.

We discussed treatment options and started counseling. The discussions in counseling prompted Jim and his parents to work with his school to tailor the transition program to his particular desires, goals, and needs. In addition, Jim spoke with Calvin about missing him and wanting to move away from home at some point as well. Jim and his brother started to communicate regularly via FaceTime, and Jim visited Calvin at college. During counseling sessions, Jim and his family also talked about his future plans.

Jim's symptoms improved but did not resolve, so in discussion with Jim and his parents, we decided to add an antidepressant. We selected bupropion (Wellbutrin) for its antidepressant properties but also because we've often found that people are less likely to gain weight on it and it may actually help reduce appetite and/or contribute to weight loss. With Jim's increased appetite, the potential effect was an anticipated benefit

of bupropion. Jim's symptoms continued to improve, his appetite was modestly reduced, and he became more involved in activities.

Mood disorders are very common in our society in general. Depression is the most frequently diagnosed mood disorder. Not surprisingly, mood disorders are also common in adults with Down syndrome. In fact, depression is the mental illness we have most commonly diagnosed. Because of the frequency of this condition, this chapter will discuss in some detail the various causes and manifestations of depression in people with Down syndrome.

Bipolar disorder and mania are two other types of mood disorders that may occur in people with Down syndrome. Bipolar disorder is characterized by periods of depression alternating with periods of mania. This chapter discusses bipolar disorder at some length because the symptoms can be very severe and debilitating. It also takes a look at mania, which may also be severe, but is less common in adults with Down syndrome.

Depression

Depression is a primary mood disorder that is characterized by a sad mood and/or a decreased interest in things the person previously enjoyed. As described below, there may be several accompanying symptoms. These symptoms, as well as the persistent nature of the problem, differentiate depression from merely feeling blue or sad. A major depressive episode lasts at least two weeks. A related diagnosis is dysthymic disorder, which is characterized by at least two years of depressed mood for more days than not, but there is less of a change in mood and fewer effects on the individual than with depression.

What Are the Symptoms of Depression?

There are a variety of symptoms that define depression. In the United States, mental illness, including depression, is usually diagnosed by comparing a person's symptoms against the diagnostic criteria that are listed in the *Diagnostic and Statistical Manual of Mental Disorders*, 5th edition (DSM-5), published by the American Psychiatric Association. For individuals with Down syndrome, it may be more beneficial to use The *Diagnostic Manual-Intellectual Disability* (DM-ID 2), which

modifies the criteria in the DSM and was created to aid in diagnosing and treating mental disorders in people with intellectual disabilities.

The DM-ID criteria for depression require that four or more of the following symptoms be present in the same two-week period:

- depressed or irritable mood,
- loss of interest or pleasure in activities,
- weight loss or gain,
- change in sleep patterns,
- slowing down of physical movement or restlessness,
- fatigue,
- feelings of worthlessness (sometimes evident by negative self-statements),
- decreased concentration or ability to think, and/or
- recurrent thoughts of death (sometimes evident by preoccupation with thoughts of death or individuals who have died).

The symptoms must cause distress or impairment in social, occupational, or other areas of functioning.

In a review of the literature on depression in Down syndrome (Walker, 2011), one researcher included differences from criteria in people without an intellectual disability. Self-report of guilt and low self-esteem is less likely in adults with Down syndrome, while observable behavioral symptoms such as a lack of interest or participation in previously enjoyed activities, social withdraw, and psychomotor slowing or agitation may be more reliable symptoms of depression. Since some diagnostic criteria rely on the individual to self-report particular feelings—which is not always possible for people with Down syndrome—it is clear why some observable criteria are beneficial in the diagnosis.

In our experience, other symptoms of depression may include the following:

- psychotic features (extreme withdrawal, hallucinatory self-talk, etc.),
- inappropriate fears or avoidances of people/things, and
- a strong reluctance to leave the home.

In addition, other disorders may occur along with depression. These are called “co-occurring” disorders. For example, an anxiety and obsessive-compulsive disorder may occur along with depression. Many medical conditions may also be considered co-

occurring (see chapter 2 and below for more on this). Behavior disorders (chapter 22) and psychotic features (chapter 20) can also accompany depression.

Psychotic features are fairly common in people with Down syndrome who have depression; they involve symptoms that may seem very odd or make the person appear to be out of touch with reality. These symptoms may include agitated or hallucinatory-like self-talk, extreme withdrawal, and self-absorption. These symptoms are rarely an indication of a true psychosis, which is why they are called psychotic "features." These types of symptoms are also more common in children without Down syndrome who have depression. For both groups, the line between fact and fantasy is often blurred, which may result in more odd-appearing symptoms. Additionally, depression may be more debilitating for people with Down syndrome and for children. This explains why they are more likely to demonstrate severe withdrawal and self-absorption.

Diagnosis of Depression

In people without cognitive disabilities, depression is primarily diagnosed through a face-to-face interview with a mental health professional. A physical exam, laboratory tests, and history from others may provide additional information.

The diagnosis of depression may be more complicated for people with Down syndrome. As described in chapter 15, the impairment of verbal skills, conceptual thinking, and overall cognitive functioning makes it more challenging to obtain the appropriate history from the patient. Thus, there is a greater reliance on information offered by family or care providers. This adds a level of interpretation of symptoms and can lead to under- and over-reporting of symptoms. Added to this, people with Down syndrome may have some symptoms that appear more severe than they are. They may have some difficulty distinguishing between fact and fantasy, particularly when they are experiencing other symptoms of depression. For example, as mentioned above, hallucinatory-like self-talk, skill loss, or extreme withdrawal can be symptoms of depression for people with Down syndrome. If these symptoms are overemphasized, while other symptoms are missed or underemphasized, the person may be misdiagnosed with a primary psychotic disorder. This may result in the inappropriate use of a medication for psychosis.

As discussed in chapter 15, these limitations often make it necessary for the person making the diagnosis to do some observation of his or her own. Sometimes the best information can be obtained by observing or spending time with the person at

home or at work. Having the person draw how he is feeling may also reveal important information. In addition, the individual may learn how to articulate his feelings better through a trial of counseling.

Causes of Depression

In people with Down syndrome, as in everyone else, three general factors can contribute to the development of depression:

1. social and environmental stress,
2. physical differences or changes within the brain, and
3. medical problems.

Stress

Examples of stress that can lead to depression include the following:

- personal losses (such as the death of a parent, or the loss of a sibling's company when he moves out of state);
- environmental stress (such as a problem at work that the person finds stressful);
- changes in care providers.

Previous chapters address many issues that are important for mental health promotion. A change in, or loss of, any of these positive, healthy aspects of a person's life may contribute to depression.

Understanding how social and environmental stress can lead to depression can be useful in reaching a diagnosis. Looking for potential contributing social factors not only helps put the problem in context but can also be very beneficial in developing a treatment plan.

Some people respond to stress or loss with anger and aggressive behavior. Although some teens and adults with Down syndrome react this way, the more common response seems to be to become more passive when depressed. The person may withdraw from family and friends and from participation in activities he formerly enjoyed and may even refuse to leave the house. We believe that this more passive response is due to a sense of helplessness that comes when individuals have a sense of having little control of their lives and few opportunities for solving problems for

themselves. Depression may also be a protective mechanism to conserve life and energy, especially if the person feels overwhelmed.

Differences within the Brain

Biochemistry also plays a role in depression, at least some of the time. The brain functions much like an electrical system, with the gaps between brain cells bridged by the release of chemicals called neurotransmitters. Depression is thought to be related to a decrease in the neurotransmitters serotonin, norepinephrine, and/or dopamine. In addition, the sensitivity of the receptors of the cells to which the neurotransmitters bind plays a role in depression. What causes the change in the amount of the neurotransmitters or sensitivity of the receptors is still being investigated. Genetics, environmental stress, social factors, and other issues may all contribute to these changes.

Studies have suggested that there may differences in the amount of these neurotransmitters and/or the nature of the receptors in the brains of people with Down syndrome. The role of serotonin and other neurotransmitters, the receptors, and other factors related to neurotransmitters in people with and without Down syndrome are all areas that are still being studied.

Medical Problems

Many medical conditions, particularly those that last a long time or are severe, can contribute to depression. The frustration of not feeling well, the changes in routine, the inability to participate in activities, and the discomfort and inconvenience of procedures and tests can all contribute to depression. In addition, several medication conditions can directly affect the brain and contribute to depression.

Persistent or chronic pain can also contribute significantly to depression in people with Down syndrome. First, being in pain can be depressing. Second, the frustration of not being able to communicate one's pain can also contribute to depression. This cycle is further discussed in chapter 2. However, an issue that bears repeating here is that treating the painful condition and the pain is an important aspect of treating the depression. If pain persists and depression increases, it can become a chicken-and-egg situation. Which came first, or which is causing or

exacerbating the other, can be difficult to determine. In truth, deciding on cause and effect is much less important than evaluating and treating both conditions.

In chapter 2 we discuss in detail the medical problems that most commonly contribute to mental health problems in adults with Down syndrome. Of those disorders, the conditions most like to cause depression include the five described below.

Hypothyroidism (Underactive Thyroid). About 40 percent of the individuals with Down syndrome we have evaluated have hypothyroidism. It is often associated with lethargy, decreased interest in activities, and depressed mood. At times, it can cause a full-blown depression. Hypothyroidism is diagnosed through a blood test.

Treating hypothyroidism often improves an individual's depression and sometimes is the only treatment needed for the depression. If a person is depressed and his hypothyroidism is not treated, treatment for the depression often will not be completely successful. Conversely, sometimes treatment with thyroid medication is not all that is necessary. In that situation, it may be that the hypothyroidism is not the cause of the depression but only contributing to it. It is also possible that the hypothyroidism was the initial cause, but now other factors have developed. In either case, additional treatment is needed. This may include counseling, antidepressant medications, and other interventions.

Lyle, age thirty-five, was seen in his home because he refused to go outside. For several months, he had refused to get out of bed, even to go to the kitchen to eat or to the bathroom to use the toilet. He was found to have hypothyroidism and was started on the medication levothyroxine. Afterward, he became more animated and began spending time out of bed. A number of other issues needed to be addressed before further improvement was achieved, but treating his hypothyroidism was a successful first step.

Sleep Apnea. In people with sleep apnea, breathing either stops for a period of time or there is blockage of the airway during sleep. This results in decreased oxygen or abnormally increased carbon dioxide in the blood and an abnormal sleep cycle. Sleep apnea can cause depression. It has an indirect effect on the brain due to persistent fatigue and a direct effect on the brain due to inadequate sleep cycles and oxygen deprivation. The depression may even have psychotic features such as

hallucinations and delusions. Diagnosis and treatment of apnea are discussed in chapter 2. Treatment may include weight loss, changing the position of sleep, a mouth appliance, a CPAP or BIPAP machine, or surgery.

Celiac Disease. Celiac disease is caused by sensitivity to gluten, a protein found in wheat, barley, and rye. When foods containing these grains are eaten, the person with celiac disease develops an inflammation in the small intestine, leading to poor absorption of food, vitamins, and minerals. Symptoms may include weight loss, diarrhea, fatigue, and a sense of ill health. Particularly when experienced on a chronic basis, these symptoms can contribute to depression. In addition, the vitamin and mineral deficiencies caused by celiac disease can contribute to depression.

Celiac disease has been found to be more common in people with Down syndrome. Particularly since the symptoms can be subtle and people with Down syndrome may have difficulty describing their feelings of poor health, celiac disease should be considered when a person is depressed.

> *Alberto, age twenty-seven, was evaluated for chronic dysthymia (sadness). While he did not have full-blown depression, he seemed unhappy, fatigued, and reluctant to participate in activities. He was also known to need iron supplementation to treat and prevent iron deficiency anemia and he frequently experienced crampy abdominal pain. After the diagnosis of celiac disease was made and Alberto began a gluten-free diet, his abdominal pain decreased, he no longer needed iron supplementation, and his mood remarkably improved. His fatigue and reluctance to participate in activities gradually improved as well. He required no further treatment and "returned to his previous self."*

Vitamin B12 Deficiency. Vitamin B12 deficiency can contribute to depression. It may be more common in people with Down syndrome, perhaps due in part to celiac disease. A blood test for Vitamin B12 deficiency is therefore recommended when a person with Down syndrome has depression. Treatment may consist of increasing B12 in the diet (through vitamins or dietary changes), treatment of the cause of poor absorption of B12 (such as celiac disease), or regular B12 injections.

Vision or Hearing Impairment. Impaired hearing and/or sight can be significant challenges for anyone. If someone has a reduced intellectual ability to compensate for the loss, difficulties hearing or seeing can be particularly

problematic. The challenge may be more than some people with Down syndrome can manage, and this can lead to depression. If the problem is correctable, direct treatment of the impaired sense is clearly also helpful in improving the depression. When not correctable, treatment consists of optimizing the function of the sense as well as teaching the person to use compensatory mechanisms. Often antidepressant medications are also necessary.

Treatment of Depression

In addition to diagnosing possible medical problems that cause depression and providing medical treatment for them, a variety of other strategies are also used to treat depression. They include

- counseling (see chapter 16),
- identifying and reducing stress (discussed throughout the book, and especially in chapter 14),
- medication (see below), and
- encouraging participation in affirming activities and exercise.

Medications

We have found that medications can be useful in treating depression in adolescents and adults with Down syndrome. The goal of prescribing medications is not only to improve the depression but also to help the person be more responsive to the other treatments. The other treatment methods listed above are also beneficial.

When choosing an antidepressant medication, both the effects and potential side effects must be considered. If several medications are equally effective, it is usually best to select the medication with the lowest potential for side effects. However, in some situations, a potential side effect may be useful, and therefore the medication with that side effect might be chosen. For example, a medication that is sometimes sedating might be helpful for someone who is having difficulty getting to sleep.

The antidepressant medications can be quite effective. It often takes several weeks, though, to see the effect of starting the medication or increasing the dose.

There are five types of antidepressants that may be prescribed:

1. tricyclic antidepressants (TCAs),

2. selective serotonin reuptake inhibitors (SSRIs),
3. serotonin-norepinephrine reuptake inhibitors (SNRIs),
4. atypical antidepressants, and
5. monoamine oxidase inhibitors (MAO) (not discussed further here because these tend to be older medications with greater side effect potential and we never prescribe them).

Tricyclic Antidepressants. Tricyclic antidepressants were the first medications specifically developed to treat depression. Examples include amitriptyline and nortriptyline.

These medications are often quite effective. However, their side effects can be problematic. Particularly of note are the anticholinergic side effects. These side effects are caused when the effects of choline, a neurotransmitter, are blocked in the peripheral nervous system or the brain. Choline plays a large role in the autonomic nervous system (control of body functions that do not require conscious thought, such as heart rate) and skeletal muscles. Anticholinergic side effects include dry mouth, constipation, urinary problems, dizziness, and low blood pressure, among others. People with Down syndrome tend to be particularly sensitive to these side effects. Therefore, we use these medications infrequently.

Some tricyclic antidepressants tend to be sedating. This side effect can be used to advantage if difficulty sleeping is one of the symptoms of an individual's depression. Using these medications in the evening or at bedtime can lead to improved sleep. We have found doxepin (Sinequan) and amitriptyline (Elavil) to be particularly sedating and potentially helpful. However, the anticholinergic side effects usually outweigh the benefit, so we use them only infrequently.

Selective Serotonin Reuptake Inhibitors (SSRIs). As discussed in chapter 16, selective serotonin reuptake inhibitors (SSRIs) work by slowing down or blocking the reuptake (recycling) of the neurotransmitter serotonin in the brain. Since depressed people are thought to have insufficient serotonin, using these medications to increase the availability of serotonin in the brain can be effective in reducing symptoms of depression.

In our experience, the SSRIs citalopram (Celexa), escitalopram (Lexapro), paroxetine (Paxil), fluoxetine (Prozac), and sertraline (Zoloft) are all effective in improving the symptoms of depression. Fluvoxamine (Luvox), another SSRI, does not have an FDA indication for depression (it is indicated for obsessive-compulsive

disorder), and we have not found it to be particularly helpful for depression. As explained in chapter 16, the choice of medication is determined by the expected benefit, the side effect profile (using side effects to the patient's benefit or avoiding medications that tend to have certain side effects), and by individual issues (for example, a liquid is preferable if the person cannot swallow pills).

We have found that SSRIs tend to have fewer side effects for our patients than the tricyclic antidepressants do. They are not without side effects, however. Some of our patients with Down syndrome have developed agitation as a side effect when taking an SSRI. In our experience, this is most common with fluoxetine (Prozac), and therefore we tend to use it less frequently. With fluoxetine, the agitation is often delayed several weeks or even a few months. We have also seen agitation with paroxetine (Paxil), but it tends to occur earlier, within a few weeks of starting the medication or of increasing the dose. In our experience, agitation occurs less frequently with citalopram (Celexa), escitalopram (Lexapro), and sertraline (Zoloft). Agitation with an antidepressant may also be "unmasking" bipolar disorder (see below).

Sedation can be a side effect with these medications. Alternatively, these medications may make some people more awake. There doesn't seem to be any clear pattern as to who is affected in which way. We generally start by having the individual take medication in the morning and may switch to the evening if sedation is a side effect. A trial of the medication is the only way to determine whether someone will have either of these side effects (although genetic testing may give some insight; see box). Interestingly, despite the similarity of the medications, a person may not tolerate one of the medications because of these side effects but may tolerate another one.

Using Genetic Testing to Guide Medication Choice

There are now available some genetic tests to help in guiding medication selection. At this time the testing doesn't tell us which medication to choose based on symptoms or the patient's diagnosis, but it helps with selection based on factors such as metabolism of the medication by that individual. Testing can also give information about the individual's folic acid metabolism, which can affect effectiveness of the medication selected. The testing is typically done on a sample collected by swabbing inside the person's mouth (cheek).

At present, no solid data indicate that these tests improve treatment of mental illness. Therefore, they are usually not covered by insurance. We anticipate continued study over time to determine clinical usefulness of genetic testing.

Weight gain and increased appetite can also be side effects of SSRIs. Paroxetine (Paxil) is the SSRI that most often has this effect on our patients with Down syndrome. While this can be a detriment for some patients, for those who have a decreased appetite as part of their depression, the weight gain side effect can be a huge benefit.

In people without Down syndrome, increased suicide risk has been described as a potential side effect of these medications, particularly in children, adolescents, and young adults. We have not observed this side effect in people with Down syndrome. In fact, although patients have occasionally discussed thoughts of suicide, in our experience actual suicide attempts are extremely rare in people with Down syndrome.

Another aspect of treatment with SSRIs is the need to wean the medications when they are discontinued instead of abruptly stopping them. For some individuals, there appears to be a withdrawal-type phenomenon when discontinuing the medications. The weaning process can take many weeks or months.

Serotonin-Norepinephrine Reuptake Inhibitors (SNRIs). We have also found the serotonin-norepinephrine reuptake inhibitors beneficial in treating depression. These include duloxetine (Cymbalta), venlafaxine (Effexor), desvenlafaxine (Pristiq), and others. These medications inhibit (block or slow down) the uptake of both norepinephrine and serotonin in the brain. Our experience with SNRIs is similar to that with SSRIs. They are pretty well tolerated (limited side effects), have varying effectiveness from individual to individual, and should be withdrawn gradually. For individuals who have lethargy as part of their symptoms, we sometimes find that there is a “stimulant-like” effect, but this is certainly not universal and varies in each individual. We usually start the medication in the morning but will switch to the evening if the person has the opposite effect of sedation. We will often try an SSRI first because they have been available longer and we have had good success with them. However, choosing an SSRI vs an SNRI depends on the individual’s history including family history (e.g. if a family member has responded well or poorly to a particular medication), the symptoms, and if a potential side effect may have benefit.

Serotonin Modulators. These antidepressants appear to inhibit serotonin reuptake (block or slow down the recycling of serotonin in the brain) and also bind to and activate certain serotonin receptors. This results in an increased availability of serotonin in the brain. Some of the medications in this class—e.g., vortioxetine (Trintellix) and vilazodone (Viibryd)—are newer, and we have less experience with them. One older member of this class, trazodone (Desyrel) is an antidepressant that also has an FDA indication for depression but not insomnia. We have not found it to be particularly beneficial for treating depression in adults with Down syndrome. However, we have found it to be a useful additional medication when the individual has difficulty sleeping. Trazodone has a side effect of sedation that can be used to advantage by prescribing it at bedtime only. As the depression improves with other treatment, the trazodone can often be weaned.

Other Antidepressants. In addition to SSRIs, SNRIs, serotonin modulators, and tricyclic antidepressants, other medications are sometimes used to treat depression. They do not fit neatly into one category because they act on a variety of neurotransmitters. We have found medications in this fourth category of antidepressants to also be effective for adults with Down syndrome. Often, they contribute to improvement when there are additional issues besides depression to be addressed.

Bupropion (Wellbutrin) is an antidepressant that increases serotonin, norepinephrine, and dopamine. It can be a good antidepressant and seems to help some people lose weight. When increased appetite or weight gain is part of the depression, Wellbutrin can be of particular benefit. Seizures, however, are a known, but infrequent, side effect, so we avoid using bupropion with individuals who have a history of seizures.

Mirtazapine (Remeron) is in the subclass tricyclic/tetracyclic antidepressants. When it is prescribed to our patients with Down syndrome and depression, we have found that both sedation and weight gain are common side effects. Therefore, we reserve mirtazapine's use for those with sleep disturbance and decreased appetite. We start this medication in the evening and monitor for effect and side effects.

Treatment Duration

We usually treat depression with medications for six to twelve months beyond the time the symptoms resolve. If the symptoms were severe before treatment (for example, aggressive behavior accompanied the depressive symptoms) or there were

other difficult circumstances (such as the person refusing to take medication when he is depressed), we consider treating longer or indefinitely. If depression recurs after the medication is weaned, we also recommend considering treating subsequent episodes longer or indefinitely. Each time a person has a recurrence of symptoms, he is more likely to have another recurrence when the medications are discontinued.

When Depression Isn't the Only Problem

As mentioned above, other mental illnesses sometimes accompany depression (so-called "co-occurring conditions"). In these instances, it is often helpful to prescribe a medication to treat the co-occurring condition, in addition to an antidepressant.

Anxiety

Generalized anxiety often occurs with depression. Several of the antidepressants can also be helpful in treating anxiety (FDA-approved medications include paroxetine, escitalopram, duloxetine, and venlafaxine). Panic disorder may also co-occur and the antidepressants may be helpful in treating it as well (FDA-approved medications include fluoxetine, paroxetine, sertraline, and venlafaxine). In addition, social anxiety disorder can coexist with depression (FDA-approved medications include fluvoxamine, paroxetine, sertraline, and venlafaxine). However, it may take several weeks to see any anxiety-reducing benefits, just as the antidepressant effect of the medications may take time. Often an individual who has both depression and anxiety is most disturbed by the anxiety. Therefore, it can be beneficial to start an antianxiety medication along with the antidepressant. Then, once the person starts to feel the effect of the antidepressant medication, the antianxiety medication can be weaned.

In this situation, we use a short- or mid-acting benzodiazepine. A benzodiazepine is a medication that reduces anxiety, and the most recognized name in the category is Valium (diazepam). Alprazolam (Xanax) and lorazepam (Ativan) have both proven beneficial for our patients with Down syndrome. Short- and mid-acting benzodiazepines act quickly after they are taken, and their effect is short-lived (several hours). These medications reduce anxiety very quickly, and the dose can be adjusted every few days. Since these medications cause sedation, the goal is to find

the dose that gives the maximum benefit with the least sedation. See chapter 18 for more information on treating anxiety.

Drew, a thirty-four-year-old man with Down syndrome, had become withdrawn. He was declining to go to work, having difficulty sleeping, and would become agitated when his mother suggested leaving the house. He sometimes got aggressive with his mother. When we met him, these symptoms had been increasing for the previous four to five months. No underlying medical conditions were found.

We treated Drew with sertraline (Zoloft) and alprazolam (Xanax). His anxiety and agitation improved within a few days. Over the next several weeks, his mood also began to improve. The alprazolam was weaned so that he only needed it if he became agitated or was going someplace that tended to make him anxious. The dose of sertraline was gradually increased over several months until most of Drew's symptoms were gone and he had nearly returned to his previous level of function. Over the next year, he resumed more of his previous activities, and his symptoms continued to improve.

Sleep Disturbance

Sleep disturbance can also accompany depression. Antidepressant medications often benefit sleep disturbance, although once again, this benefit may not be seen immediately. As mentioned above, choosing an antidepressant that is more sedating can be helpful. Sometimes, however, it helps to start a medication that benefits sleep more directly and quickly.

A short-acting benzodiazepine such as alprazolam can be an effective sleep aid. Using a shorter-acting benzodiazepine makes it less likely that the person will be sedated or have difficulty arising the next day. As mentioned above, trazodone has been beneficial to a number of our patients as a sleep aid. Sleep agents that interact with the GABA-benzodiazepine system can also be helpful. GABA is an inhibitory neurotransmitter, and it tends to reduce excitation in the brain. In our experience, zolpidem (Ambien), zaleplon (Sonata), and eszopiclone (Lunesta) can be helpful. Caution is advised for people with sleep apnea (which is more common in people with

Down syndrome) when starting any medication that causes sedation. Sedating medications can worsen sleep apnea.

In addition, melatonin has benefitted many of our patients with sleep difficulties. Melatonin is a hormone that is used by travelers to help improve sleep problems associated with travel across time zones (jet lag). We have found melatonin to be a useful sleep aid both for people with and without depression, although we recommend caution when using it in a person with depression (it can contribute to worsening depression). We usually start with 2 mg at bedtime and increase to 4 mg a week or two later if 2 mg is not enough, and up to 6 mg in a few weeks if sleep continues to be a problem. There are also slow-release versions of melatonin. One formulation has both the immediate and slow-release types. This can be helpful for people who are having difficulty both falling asleep and staying asleep. Further research is needed regarding long-term use, however, before we would recommend using melatonin indefinitely.

Magnesium is an electrolyte that helps some people sleep. There are various formulations of magnesium: magnesium oxide, magnesium sulfate, and others. Some individuals find one works better than another. Some find that magnesium powder or pills that contain a few of the different formulations work better. We recommend checking a magnesium level and kidney function before starting magnesium to avoid toxicity. People with kidney dysfunction should consider an alternative form of sleep aid.

Psychotic Symptoms

Psychotic symptoms consist of hallucinations (an experience involving the apparent perception of something not present; usually visual or auditory) and delusions (false beliefs about yourself or others). Some people develop psychotic symptoms as part of their depression. In some individuals, it can be difficult to determine which are truly psychotic features and which are characteristics that sometimes occur in people with Down syndrome who have developed depression. For instance, self-talk often increases in people with Down syndrome who are depressed. So, too, can talking to imaginary friends. These can be strategies that help the person with Down syndrome cope or work through his problems. However, they can also be signs of a psychotic process. In addition, symptoms of extreme withdrawal and abnormal thought processes can occur.

Often psychotic symptoms will improve with the treatments for depression discussed above. Sometimes, however, an antipsychotic medication can be helpful. We have found the newer "atypical" antipsychotics to work particularly well. In addition to treating the psychotic symptoms, they also can improve the depressive symptoms. Risperidone (Risperdal), olanzapine (Zyprexa), quetiapine (Seroquel), aripiprazole (Abilify), ziprasidone (Geodon), and others have all proved beneficial for adults with Down syndrome. Unfortunately, though, side effects of these medications such as weight gain, sedation, and elevated blood sugar have been particularly problematic for many people we have treated. When side effects occur, careful assessment of the side effects and the benefits for the individual is essential to determine the best course of action.

The older antipsychotics such as thioridazine and haloperidol can also be helpful, but we have found that the side effects are even more problematic. Furthermore, these medications don't have the additional benefit of treating depressive symptoms. A potential side effect of particular concern is tardive dyskinesia (TD). (This can also occur with the newer antipsychotics but seems to be more common with the older ones.) Tardive dyskinesia is a neurological syndrome characterized by involuntary and abnormal movements. It can be permanent, even after discontinuing the medication.

In our experience, using an antipsychotic medication is frequently not necessary in treating depression, even if an adult with Down syndrome has apparent psychotic symptoms. Particularly when the symptoms are self-talk and use of imaginary friends, an antidepressant can often successfully treat the condition. Again, these symptoms may be more coping strategies used by the patient rather than psychotic symptoms.

Sally, age twenty-nine, had been talking to herself in an agitated manner for several months. Previously she had just talked to herself calmly in her own room, but now she was talking to herself in many locations. She was waking during the night, refused to participate in the activities at her group home, and had lost her sense of humor. We found no underlying medical causes for Sally's behavior changes. However, a staff member Sally had especially liked had left the group home a few months prior to the onset of her symptoms, and the company she was working for had gone out of business a month before that.

We discussed the treatment options with Sally and her mother. The psychotic-like symptoms of agitated self-talk were particularly disturbing to both of them. However, the possible side effect of weight gain with medication concerned them, since Sally already weighed 183 pounds at five foot one. We decided to try an SSRI, since the chance of significant weight gain is usually less than with antipsychotic medications. We also thought there was a good chance that Sally's symptoms would respond to antidepressants alone. Sally was started on sertraline (Zoloft), and her dose was adjusted over the next few months. With the help of Sally's family and supportive staff who encouraged her back into activities, Sally's mood, sleep patterns, and willingness to participate in activities all improved. As Sally began to feel better and was able to verbalize her concerns, her self-talk decreased, and she began to self-talk only in her bedroom again. Her mother excitedly said, "Sally is back."

On the other hand, Frank, age thirty-six, required antipsychotic medications. For several months, he had been severely withdrawn. He had lost interest in activities, was often agitated, awoke during the night, had a decreased appetite, and was doing more self-talk. Frank had limited verbal skills, so it was very difficult to delineate the content of his thought processes or his self-talk. Frank was found to have hypothyroidism, and appropriate treatment was provided. However, in light of the severity of his symptoms, we also prescribed an antidepressant. There was little improvement in Frank's symptoms, so risperidone (Risperdal) was added at bedtime, and his sleep and agitation improved fairly rapidly. With time and dose adjustments, his other symptoms also improved. In addition, Frank regained a sense of pleasure from activities that he had formerly enjoyed.

Mania

As discussed in the previous section, depression is one of the mood disorders in the DSM-5. Mood disorders may also include an opposite state of emotion called mania. Mania may include feelings of well-being or euphoria, and, at the extreme, a manic-like state that may include agitation, sleeplessness, hyperactivity, angry rages,

and even self-destructive behavior. Fortunately, mania by itself does not appear to be common in adults with Down syndrome. We have seen just a handful of patients with mania alone (not associated with depression).

In November, Penny, age twenty-four, was brought in for any evaluation by her concerned parents. Her mother and father had begun to notice changes in her in January, when she lost a valued job as a result of her company moving out of state. She became increasingly restless and preoccupied over the course of the year. She had not been able to find a new job and had therefore spent some time alone at home doing hobbies and things of interest to keep her busy. Despite losing her job, she continued to attend many social or recreational activities every week. However, as the year wore on, her parents began to notice that she seemed distracted and had difficulty focusing on the activities and socializing with her peers. They did not observe any evidence of a sad or depressed mood or any other evidence of depression. For example, Penny did not withdraw, lose interest in doing activities she enjoyed, or have any change in her appetite.

In September and October, Penny's parents noticed new changes in her behavior. First, she focused more intently on playing the piano, for which she had considerable talent. Her parents were initially pleased because they wanted to support her talent. Their pleasure quickly turned into alarm, however, as Penny's piano playing became obsessive. She played day and night as if she couldn't stop. Her playing became more frantic, until one day she suddenly stopped and did not play again.

Afterward, her parents noted other worrisome changes, and her father observed that Penny's anxiety seemed to go "through the roof." Although she'd had restless sleep earlier in the year, now she didn't appear to sleep at all. Even though Penny appeared tired in the day, she did not take any naps. She began to speak in an extremely fast and pressured manner that was garbled and very difficult to understand—unlike her usual excellent speech. When her parents could understand what she was saying, they found the content of her speech disturbing. She was repeating statements about petty concerns that she appeared to have blown out of proportion. For example, she was worried that she had

offended a neighbor by not waving to her. In the daytime, when not occupied or moving about, Penny would often pick at her skin or fingernails. At the appointment, she grimaced constantly and seemed irritated and uncomfortable with herself. She couldn't seem to relax or stop her constant movement.

At Penny's initial evaluation, we diagnosed her with a hyperthyroid condition that could result in manic-like symptoms. This mania continued after treatment for the hyperthyroidism. Penny then began taking psychotropic medications. After a trial of several different types and classes of medications, she responded well to a combination of an antipsychotic and an anticonvulsant medication. To support the gains made from the medications, we encouraged Penny's parents to get her back into beneficial social, recreational, and vocational activities whenever possible. As her mania began to recede, she became more social, attentive, and engaged in the activities. Equally important, Penny's parents and job coach agency were able to find a new job that was similar to her previous position. As her symptoms began to dissipate and she resumed her normal activities, she regained her sense of well-being and eventually her pride and self-esteem. In time, she also began to play the piano in a more normal and enjoyable way.

See the section on "Treatment of Bipolar Disorder" below for more on medications and other means of treating both mania and bipolar disorder.

Bipolar Disorder

As discussed in this chapter, depressive disorders are the most common type of mental health disorder found in people with Down syndrome, and mania is one of the least common. What we have seen slightly more often than mania is a condition called bipolar disorder in which people fluctuate between manic and depressive mood states. (This condition used to be called manic depression.)

To explain this condition in people with Down syndrome, it may be helpful to first look at bipolar disorder in people without Down syndrome (or another intellectual disability). People with bipolar disorder who are in an "up," or manic, state often have feelings of exhilaration and euphoria. They may have boundless

energy to work day and night on projects and activities of interest. Unfortunately, as the mania increases, people often "go over the top" and become more extreme and unreasonable in their grandiosity and their behavior. For example, they may buy extravagant items on a whim, gamble away life savings, or engage in risky sexual behavior. Mania may also lead to agitation, restlessness, sleeping problems, and anxiety.

What goes up invariably comes down in this disorder. At some point, the pendulum moves back toward the other pole, and the person begins a descent into depression. In this depressed state, people often shut themselves off from the world for weeks or even months at a time before the pendulum once again moves toward the other (manic) pole. Like all mental health disorders, bipolar disorder varies in intensity or severity, but the pattern of fluctuation in mood states remains a hallmark of the condition.

Although it has been known for many years that bipolar disorder occurs in adults, it is now also recognized in children. It may have taken some time to discover this because children often have a very different pattern of symptoms compared to adults. Similarly, it may have taken time to confirm that adults with Down syndrome may also have bipolar disorder because they have a symptom pattern that is often similar to that of children without Down syndrome who have bipolar disorder. In fact, because the pattern is different than it is in adults without Down syndrome, it was only recently realized that adults with Down syndrome do get bipolar disorder.

What are these differences compared to adults without Down syndrome? First, children and people with Down syndrome who have bipolar disorder often cycle more rapidly between up and down mood states. They may cycle back and forth in as short a period as a day or even over several hours, compared to adults without Down syndrome, who typically cycle between mood states over weeks or even months. Also, symptom presentation may be different. In the down or depressive state, children and people with Down syndrome are far less likely to verbalize feelings of sadness, worthlessness, or guilt. However, they do show such observable changes as an irritable mood, withdrawal, and loss of interest in activities formerly enjoyed. (See the above section on depression for more on this.) Similarly, mania in children and people with Down syndrome may not include the out-of-control spending or sexual behavior displayed by adults without Down syndrome. Nevertheless, the pattern of overstimulation, agitation, restlessness, angry rages, and hyperactive behavior occurs, and these changes are unmistakable symptoms of mania to parents or other caregivers, once they are educated on this condition.

Jacqui, a nineteen-year-old with Down syndrome, began showing symptoms of bipolar disorder six months before her parents brought her for evaluation. She exhibited a pattern of cycling back and forth from mania to depression and back to mania over the course of just one day. Before the onset of symptoms, Jacqui and her family had been proud of how capable and independent she was. She had also been meticulous about her grooming and her appearance. This changed once the symptoms started, particularly in the morning. Every task was a challenge and a struggle between her and her parents. Invariably as the morning routine progressed, things would become more and more tense as Jacqui resisted her parents' urgings to get ready for work. She would eventually "lose it" and unleash a barrage of screams, yells, and foul language, and she would often become physically aggressive with them.

This was all behavior that Jacqui's parents had never seen before, and they were horrified by it. Most upsetting to them was the cold look in her eyes when she raged at them. As her mother stated, this was "not her." She appeared as if "possessed." Although Jacqui weighed only a hundred pounds, her father, who was a big, muscular man, was intimidated by her during these rages.

By the time her parents finally got Jacqui on the van to work, they were physically and emotionally exhausted. Jacqui's supervisor reported that upon arriving at work and throughout the morning she was lethargic and listless and did little work, although she had previously been a good worker. She was also quite often in a foul mood. By midafternoon, she would begin to perk up and often cooperated with work tasks for several hours. Later, she became first silly and then anxious and agitated. In this state, she would fight any attempt by her supervisor to get her to do her normal tasks and would have angry outbursts, just as at home. Although the worksite staff had been tolerant or her behavior, as her angry outbursts became louder and more threatening, her parents worried that she would lose her job.

When Jacqui returned home from work at around 4:00, she often withdrew to her room until after 7:00 or 8:00 in the evening. She sometimes took short naps during this period, but much of the time she

just lay on her bed staring at the ceiling. When she finally came out of her room, she was "like a different person." She often began by laughing and joking with her parents, but this could turn into uncontrollable laughter. As her up mood escalated, she often became more irritable and unreasonable. Her parents tried to walk around her very gingerly, but she could respond with shouts of anger if she became upset about anything, including the simplest of requests.

Later, as her exhausted parents prepared for bed, they were dismayed that Jacqui actually seemed to become more alert and agitated. In this state she simply could not settle down. Finally, by 1:00 or 1:30 a.m., she would "crash in her bed." Afterward, she often woke up several times in the night and one of her parents would try to settle her down once again. In the morning, Jacqui's parents would get up and try to get her ready for her job all over again. Every day they became wearier, and Jacqui's symptoms seemed to get even worse.

At Jacqui's evaluation, her parents described her behavior and reported one additional issue that was disturbing to them: during her more active or manic periods, Jacqui displayed an odd, ritualistic behavior consisting of rubbing her hands together and making a loud, raspberry-like sound with her mouth. This was particularly upsetting to her parents, who were already on edge from her behavior and from sleep deprivation. They were also concerned about her doing this in public, believing that it would draw negative attention to her and the family. As a result, they had stopped sending her to social and recreation programs. This cut Jacqui off from her friends and from physical activities that had been positive influences on her life. Equally importantly, they had stopped going out as a family, which made them feel like they were prisoners in their own house and of the situation.

Treatment of Bipolar Disorder

The essential first step in successfully treating this condition is to make an accurate diagnosis. Health care providers must be careful to take a very complete and detailed history of the symptoms. If they are too hasty, they may only hear about one mood state, which may lead to inappropriate or ineffective treatments. For example,

if they only hear about the depressive symptoms, they may use an antidepressant medication that may actually worsen the problem by increasing agitation or other symptoms of mania (in a sense, “unmasking” the mania symptoms). Once bipolar disorder is identified, the goal of a treatment is stabilizing the extreme mood fluctuations.

Medications

There are a number of choices of medications to treat mania and bipolar illness. The medications include lithium, antipsychotics, and antiseizure medications. Some people benefit from one medication alone, while others need a combination of medications.

Lithium is a medication that has been used for a long time but is still a very effective choice. It affects the transport of sodium into nerve cells. How that action affects mania or bipolar illness is not clear, but lithium does stabilize mood. However, lithium has a number of side effects. Care must be taken to make sure the person is drinking enough fluids. Anything that reduces the output of urine, including dehydration or kidney problems, can raise the level of lithium in the bloodstream to dangerous levels. Lithium can affect kidney function, cause hypothyroidism (underactive thyroid), and cause disturbances in the heart rhythm. Drowsiness, tremor, and frequent urination are more common side effects.

Antipsychotic medications can also be an effective treatment for mania and bipolar illness. Ziprasidone (Geodon), risperidone (Risperdal), quetiapine (Seroquel), olanzapine (Zyprexa), lurasidone (Latuda), and aripiprazole (Abilify) are all approved for treatment of bipolar disorder and acute mania. They can very effectively improve the symptoms. We have found them to be particularly helpful in reducing agitation and aggressive behavior. Some of the possible side effects include weight gain, sedation, hyperglycemia (elevated blood sugar), diabetes mellitus, and swallowing dysfunction. We regularly monitor blood sugar tests for our patients on these medications. In addition, we monitor for tardive dyskinesia (TD). TD is a potentially permanent movement disorder that is less common with these newer medications than it was with the older antipsychotic medications.

Antiseizure medications can also stabilize mood for people with mania or bipolar disorder. Valproic acid (Depakote) is approved for mania. Carbamazepine (Tegretol) is approved for mania and mixed (bipolar). Lamotrigine (Lamictal) is approved for maintenance therapy once the person has been stabilized. There are a

number of potential side effects with these medications, too. Weight gain (particularly with valproic acid), effect on liver function, decrease in white and red blood cells and platelets, very serious rash (with lamotrigine), and other side effects can potentially be drawbacks to the use of these medications.

Antidepressants, as discussed in detail earlier in this chapter, can be very helpful for the depressive symptoms of bipolar disorder. However, care must be taken because they can also cause mania, particularly in people with mania or bipolar illness.

Counseling and Support

Although it is critically important to stabilize bipolar disorder and mania through treatment with medications, practitioners also need to be extremely sensitive to the emotional havoc the disorders wreak in people's lives. Bipolar disorder is one of the most trying and stressful disorders for affected individuals and, of course, for their families and caregivers. It is difficult to understand how stressful and demoralizing this disorder is unless one has had a similar experience.

One of the first steps in helping people cope is to educate them on the cause and nature of the condition. Like many medical conditions, such as asthma or type 2 diabetes, the condition may be aggravated by stress, but it is clearly not something that the person (or caregivers) has caused. Understanding this may help to reduce people's sense of self-blame. Additionally, many parents or caregivers often feel demoralized and like failures when, no matter what they do, the person's behavior seems to worsen. Educating people about the neurological basis of the condition helped Jacqui's parents to regain a sense of confidence. For her part, Jacqui was also very upset by her behavior. She did not feel she had control over herself, and yet she felt very bad about "hurting" her parents and her supervisor at her worksite. She needed many individual and family meetings to understand that this was not her fault. Her parents were especially helpful in explaining to her that they did not blame her or feel angry at her for something that was not her fault.

Getting on with Life

Once the treatment helps to even out the person's moods, it is important for others whose lives were disrupted to begin to resume their normal patterns of life. For Jacqui's parents, the first step was to finally get some rest at night. They also

needed to go back to work and to other important activities that they had suspended during the crisis. Jacqui's parents, who had both stopped going to work in the morning to cajole Jacqui to go to work, were able to return to their jobs once her behavior moderated. This allowed them to reconnect with friends and colleagues and to think about other things in addition to Jacqui.

For three to four weeks during the peak of her symptoms, Jacqui stayed home on leave from her job. As soon as her mood fluctuations began to diminish, we strongly encouraged her family and the restaurant management where she worked to let her go back to her work. To facilitate this, we scheduled a meeting with her parents, the manager, and our staff to work out the details of her return. This helped to appease the fears her coworkers had about her displaying more extreme behavior and gave her parents a plan for making this happen. In this meeting, her manager requested a simple behavior plan to help encourage Jacqui to return to a positive work routine. She was to earn a sticker for each day that she stayed on task and had no outbursts, and when she collected five stickers, she could "cash them in" to download two songs from iTunes. After three weeks the behavior plan was suspended. It was no longer necessary—partly because Jacqui was motivated to do her job on her own and partly because her paycheck became the reward and she could purchase music with her own money.

As Jacqui's symptoms began to dissipate, especially her raspberry-like noise and hand wringing, her parents felt comfortable letting her resume her social and recreational activities. This allowed her to reconnect with her many friends. She was also able to get some exercise, which was very helpful in reducing whatever manic-like symptoms remained.

As Jacqui's mood fluctuations were reduced, the entire family began to get back to normal. Once Jacqui's noises and hand wringing became less of a problem, they once again were able to go out and were freed from their self-imposed imprisonment in the house.

Bipolar Disorder as a Lifelong Condition

It is important to recognize that bipolar disorder is a chronic, lifelong medical condition that requires ongoing monitoring and attention throughout the person's lifetime. Finding the right medication to treat the condition at the onset of the disorder (often in the teen years) is essential, as in the case of Jacqui. In addition, symptoms need to be monitored closely by caregivers and treating professionals. After the person is stable, we see our patients every three to six months. Of course, if active symptoms reemerge, the person should return to the treating practitioner immediately.

During the course of treatment, there may be some periodic fluctuations in the disorder that adversely affect the effectiveness of the medications. This may be due to stress in the person's life or to physiological or neurological changes the individual experiences. When this happens, a change in medication or dosage may be needed to once again treat the condition effectively. For example, Jacqui has had a number of medication changes during the three years she has been treated.

It is also extremely important to ensure that the person complies with taking the prescribed medication. In people without Down syndrome who have bipolar disorder, one of the well-known problems is that they may go off their medications when they are feeling up because they like the feeling of euphoria and concentrated energy the disorder gives them when in the manic state. This may be less likely to occur with teens and adults with Down syndrome because family members or guardians usually have a say in their treatment. Still, this may be a problem if a caregiver or guardian believes the person is doing better and appears to no longer need the medication. This may occur if they are not aware that bipolar disorder is a lifelong condition usually requiring continuation of the medications. In these instances, we often receive crisis calls once the symptoms begin to get out of hand again. We try to prevent such crises by actively including caregivers in the treatment process and by educating them about the condition.

In summary, mania and bipolar illness are less common in people with Down syndrome than depression. However, bipolar disorder in particular appears to be more common than previously suspected. The reason, again, is that in people with Down syndrome, bipolar disorder often looks more like the condition seen in children than in adults without Down syndrome. With that appreciation and assessing for symptoms of mania in acute mania and for symptoms of depression and mania in bipolar illness,

the diagnosis can be effectively made. Effective treatment depends on the correct diagnosis.

Conclusion

In diagnosing and treating mood disorders in people with Down syndrome, it is important to pay attention to psychological, social, biological, and medical issues. Therapy needs to be tailored to meet the unique aspects of the person's personality and environment as well as the symptoms. If depression is suspected, it is imperative that the people diagnosing and treating the condition understand characteristics often seen in adults with Down syndrome, such as self-talk, strong visual memory, and others previously discussed.

Additionally, it is important to recognize that depressive symptoms may also be part of bipolar disorder. In adults with Down syndrome, this typically includes a rapid fluctuation between depressed and manic mood states. The manic state may be characterized by agitation, overactivity, and angry outbursts, which is similar to how mania appears in children rather than in adults in the general population. Moreover, mania, like depression, may be a separate mood disorder, or it may be part of a bipolar pattern. Understanding how mood disorders in adults with Down syndrome may differ from the textbook definitions may significantly improve the diagnosis and treatment of depression, mania, and bipolar disorder.

Return to Table of Contents

Chapter 18

Anxiety Disorders

Cole, age twenty-two, recently completed his high school transition program three years after graduation. He began a new job working in the grocery store. Cole, who is somewhat of a perfectionist, was happy to have the job but said he was "nervous" that he would make a mistake. Initially he did well at the store, but as time progressed, he began to struggle. He worried about the quality of his work, became irritable if anyone made suggestions about his work, developed sleep disturbance, was fatigued during the day, and had difficulty concentrating on the job and ultimately in other settings as well. The irritability eventually became more universal at work, at home, and at various activities.

When we evaluated Cole, we did not find any physical problems contributing to these symptoms, and he'd had no episodes of panic attacks. We gave Cole the diagnosis of generalized anxiety disorder. We referred him for counseling, but we all appreciated that Cole had a limited ability to discuss his feelings and that his problem at work was urgent (with a potential threat of losing his employment). Therefore, we decided to start a medication at the same time we referred him for counseling. He started taking duloxetine (Cymbalta) and attending counseling. In addition, after discussion with his employer, Cole began regularly receiving positive feedback when he performed well at his job. Within several weeks, he was back to his old self and was doing much better at his job and enjoying life outside work.

Nearly all people would admit to feeling anxious at one time or another. That is, they feel worried or apprehensive and may experience physical symptoms such as a pounding heart, shortness of breath, or "butterflies" in the stomach. Depending on the circumstances, anxiety is often a normal reaction and not an indication of mental illness at all. For example, it is normal for a student to feel some anxiety about taking an important test, or for an adult with Down syndrome to feel anxious about moving to a new residence. This sort of normal anxiety usually has a clearly

identifiable cause and is short lived. Once the test (or whatever) is over, the person no longer feels anxious.

When anxiety interferes with someone's day-to-day life on a long-term basis, however, an anxiety disorder may be diagnosed. The *Diagnostic Manual- Intellectual Disability (DM-ID 2)* defines a number of types of anxiety disorders. We will discuss the types that are the most common among adolescents and adults with Down syndrome:

1. generalized anxiety disorder,
2. agoraphobia, and
3. panic disorder.

We will also discuss selective mutism. Obsessive-compulsive disorder is related to anxiety disorders and is described in detail in chapter 19.

Generalized Anxiety Disorder

For a person with intellectual disabilities to be diagnosed with a generalized anxiety disorder, the symptoms need to occur on most days and last at least six months. The symptoms include excessive anxiety and worry and difficulty controlling the worry. For the diagnosis of anxiety, DM-ID 2 requires a number of symptoms. The number varies based on the level of intellectual disability. The symptoms include restlessness or feelings of being keyed up or on edge, being easily fatigued, difficulty concentrating, irritability, muscle tension, and/or sleep disturbance (difficulty falling or staying asleep, or restless, unsatisfying sleep. The symptoms cause distress or impairment in social, work, or other settings and are not related to substance abuse, a medical condition, or another mental disorder.

Like many people, teens and adults with Down syndrome have worries and concerns, but this does not mean they have a disorder. For some, these worries may be fairly strong and yet still not debilitating. However, anxiety and worry may become a disorder when they begin to interfere in essential activities. For example, it is not uncommon for people with Down syndrome to dislike thunderstorms or other types of weather conditions. If they are reluctant to go outdoors because the meteorologist on TV has predicted that particular weather condition will occur, it is not necessarily a problem, as long as their reluctance doesn't prevent them from doing essential activities such as going to work or school. Sometimes this sort of fear is a little more complicated, though. We have seen a number of people who are afraid

of certain weather conditions because they fell when there was rain, ice, or snow on the sidewalk. If a person's fear of these conditions is strong and the particular weather condition occurs often, a major problem may result, such as refusing to leave the house to work. The anxiety becomes generalized when it extends beyond the weather condition and other situations or events cause anxiety.

Symptoms of Generalized Anxiety Disorder (GAD)

As with other mental illnesses, the diagnosis of generalized anxiety disorder is usually made on the basis of subjective reports. That is, the individual complains of feeling anxious, or is able to articulate what happens to his body when he is worrying. People with Down syndrome may not verbalize subjective feelings of anxiety or anxiousness. Nevertheless, for experienced caregivers, the changes in behavior of a person with anxiety are unmistakable. Additionally, many people with Down syndrome are not good at hiding their feelings. Facial grimaces, body tension, and pacing can all be clear indications of anxiety and agitation. Some people show their anxiety through mild self-injurious or habitual behavior such as hand rubbing or wringing, picking at sores or at parts of the body, hand chewing, or excessive nail biting. These behaviors often seem to be out of the person's conscious awareness.

Many people with Down syndrome also develop a sleep disturbance when they are keyed up or worried. The sleeping problem may or may not be detected by caregivers, especially if the person has minimal supervision in the evening. However, whenever other symptoms of anxiety are present, sleeping habits should be looked at very closely, such as through nighttime observations (see the section on sleeping problems in chapter 2). Anxiety may also result in irritability and loss of concentration, but these symptoms may also be associated with depression or other types of disorders and thus are not as readily identifiable as signs of anxiety.

Excessive worry is also a common symptom of an anxiety disorder. People who are verbal may express worry directly, but for those who are less verbal, their behavior is often a clear indication of worry. For example, many "weather watchers" closely monitor TV and radio weather reports on storms. When a dreaded weather change is forecast, families or caregivers often observe the telltale signs of anxiety listed above. This pattern is often repeated over and over, so that the association of anxiety with the weather or some other concern becomes obvious in time. Anxiety about the weather alone can be classified as a phobia. However, we have also seen

this “triggering” event develop into a generalized anxiety disorder with other triggers also causing anxiety.

Linda, a twenty-eight-year-old woman with Down syndrome, was brought for evaluation because of suicidal statements. This was very unusual for Linda, who had been functioning well in all areas of her life up until this point. She was well liked and supported by her family and friends, active in sports and recreation activities, and was a talented artist. One evening, however, she was noticeably upset. When questioned by staff members at her group home, she reported wanting to hurt herself.

At first, Linda could not explain her suicidal thoughts. Over the course of the evaluation, the mystery began to unravel. The first clue came from the staff person who shared with us the agency’s concerns that Linda might be psychotic as well as suicidal. This concern was based on the observation that she made odd statements to staff (in the present tense) about people and events of which they had no knowledge. In the meeting, we were able to explain that many people with Down syndrome confuse past and present tense and have a tendency to relive past traumatic events.

Linda’s parents figured out that her comments reflected times in the past when she was very frightened by severe thunderstorms. Linda’s mother would often stay in her bed with her on evenings when there were storms. This led to a discussion of her worry about storms and her need to monitor the weather closely in order to avoid such storms. The staff person confirmed that Linda was often reluctant to leave the house on stormy days. This led to the discovery that her roommate had been away on vacation for several weeks. During this time, there had been frequent and severe thunderstorms. Typically Linda’s roommate gave her a sense of comfort in the same way her mother did at home.

Unfortunately, Linda could not easily discuss her worries or concerns with others. Thus, over the two weeks that her roommate was gone, her fears and worries had escalated until she could no longer tolerate the situation. This had resulted in her suicidal comments and the appointment.

For the remainder of the appointment, we determined that there was no real threat of suicide and developed a plan to deal with Linda's concerns. Her roommate was to return from vacation that day, which would help with her fears in the evening. Beyond this, staff in her group home would meet with her daily to assess her degree of weather fear. On days she was more worried and particularly during stormy weather, she would have special attention from staff to help her. Finally, staff would help her relax by having her look at pictures of favorite people and past events so that she could substitute comforting pictures for the mental images of frightening experiences related to past storms. One year later, Linda still had worries and concerns, but they were far more manageable because of the awareness and assistance of staff and her family.

Agoraphobia

We have found that worry among adults with Down syndrome is often also associated with another type of anxiety condition, agoraphobia. Agoraphobia is a fear of being in certain places or situations. We have seen many people with Down syndrome who are terrified of going to a medical clinic, no doubt because of some painful experience in the past. We have heard that for others a negative experience at a store, shopping mall, work, or recreation site have led to similar fears and a refusal to leave home. This is because the person worries that the feared place will be visited during the outing. Although the original experience may have occurred quite some time in the past, the fear may still be very intense.

Jane had been molested by another resident in a group home when she was twenty-one. Now at the age of thirty-eight, she had lived

successfully in a foster home arrangement for fifteen years. However, she still was very fearful of leaving her sister's house after visiting her sister for fear of being taken back to the group home she had left seventeen years ago.

Due to strong visual memories, people with Down syndrome may be more susceptible to having traumatic experiences live on in their memory to terrorize them over and over. For example, it is easy to imagine how weather-related fears may be based on a previous traumatic experience. The person may re-experience the fear of the original event every time he hears a weather forecast that predicts the feared weather condition is coming. He may become overwhelmed with worry and express his fear by refusing to leave the house—hence the agoraphobia.

Treatment of Agoraphobia

Treatment of agoraphobia and other anxiety conditions may take a great deal of time, patience, and detective work. People with an overwhelming fear need to regain some sense of control in these situations. Medications may be helpful to "take the edge off the fear." (See below on medications.)

The next step is to help the person identify situations that are legitimately frightening. It may take some careful assessment to understand why the person doesn't want to leave the house. Sometimes families or other caregivers may not know what specific place or event is feared. In these situations, someone in the person's environment often holds the key to the problem. For example, in chapter 7, Bruce refused to leave his house. When workshop staff were contacted, they supplied the critical piece of information: he was being bullied at work. This information marked the turning point in treatment. Often, the person's behavior gives clues about the source of his fear. For example, we assume that people who are terrified of coming in for a medical appointment had a negative experience in a previous hospital or medical clinic visit.

Once the source of anxiety has been identified or surmised, family members and other caregivers may need to spend a great deal of time explaining that the feared place will not be visited. This may allow the person to go about his normal business in all but the feared place. As discussed in chapter 6, pictures may help to reduce fear because so many people with Down syndrome have a strong bent toward

visual images. For example, the woman who was afraid to leave her sister's house was shown pictures of her real destination (her current foster home). This was a safe place for her and not the place where she encountered the previous trauma. With the continued use of pictures, a brief period of antianxiety medication, and the patience and encouragement of her sister and foster family, she regained the confidence to leave without experiencing debilitating fear. For her and for others, the ability to do what was feared provides an enormous sense of relief and confidence, which is therapeutic in and of itself.

If the source of anxiety cannot be pinpointed, sometimes the best solution is to change the person's environment or routine in hopes of distancing him from the source of anxiety. As in the example below, if you are pretty sure an individual's home life is OK, you change his work situation.

> *Anthony, a thirty-four-year-old man who was nonverbal, began refusing to go to his job at a community pool, even though he had worked there for five years and was typically very enthusiastic about this position. After repeated attempts to get him to go to the pool, he was transferred back to his previous workshop program, where he seemed to function without fear. Only later was it learned that a supervisor at the pool had been fondling another employee. Anthony's dramatic change in behavior coincided with the presence of this supervisor. Unfortunately, if this supervisor had victimized him, he could not communicate it to others.*

Desensitization

Desensitization can be an important part of the treatment strategy for any type of phobia or agoraphobia. Some of the steps in desensitization were touched on above but not formally explained. Desensitization is a means of gradually reducing a person's fear by incrementally helping him tolerate being around the thing he fears.

Ideally, a step-by-step plan for desensitizing the person should be written down. You start by determining the maximum exposure to the feared item that the person can tolerate (e.g., just a picture or a model of the thing that he fears viewed across the room). The next step might be actually holding the picture or the model.

Over time, he is increasingly exposed to the item until his fear and anxiety are decreased.

For example, we have learned to be very patient with people who are afraid to come in for a physical exam. We often recommend that the person go through a gradual process of exposure to the health facility. This may start with a drive by the building on one day, a drive into the parking lot on another, and then a walk up to the front door, etc.

Medication may help in this process by reducing the level of anxiety and may be particularly important at the place of greatest fear, such as the point where the person enters the building or the exam room, takes a needle, etc. Sometimes the desensitization process takes months, but the need to tolerate medical exams and procedures is so great that it is all worth it. For example, one man with Down syndrome finally allowed a blood draw after practicing with staff for nine months to have everything but the needle in his skin. This man turned out to have hypothyroidism that needed to be treated and was not diagnosed until the blood was successfully drawn.

One fairly common and particularly difficult version of agoraphobia is the refusal to leave the house and get into a vehicle. The same process of gradual exposure to the feared condition applies as in the above medical situations. Again, medication may help reduce the person's level of anxiety, allowing him to be more receptive to the desensitization process. Family members or other caregivers can then very gradually and patiently expose the person to the feared situation by a daily process of moving him farther and farther from home and into the car. In extreme cases, a beginning step might be for the person just to stick one arm outside the door or to stand on the porch for ten seconds before returning inside. Eventually, the person will work up to standing next to the car, then briefly touching the car, etc.

Once the individual has been desensitized the person to riding in the vehicle, his caregivers can often get him to ride to most daily events. It is important, however, to watch closely to see if any particular environment generates more anxiety. This may help in pinpointing the specific place that triggers anxiety, even if it does not provide insight into what occurred in this setting. This place may then be avoided. If this is impossible, then desensitizing the person to the place by gradual exposure is the best option.

Another treatment that helps some people is the use of sensory strategies to reduce anxiety. There are devices and techniques that may induce relaxation and

perhaps lessen the anxiety the person feels about going to a location or activity. See chapter 12 for more information.

Panic Disorder

Panic disorders are among the most common types of anxiety disorders and are also a cause of agoraphobia in the general population. The DM-ID 2 describes panic disorder as a condition that has discrete periods of intense fear and discomfort that reach a peak within minutes. Symptoms can include the following: palpitations, pounding heart, or accelerated heart rate; sweating, trembling, or shaking; shortness of breath, choking, or smothering sensations; chest pain or discomfort; nausea or abdominal distress; feeling dizzy, faint, chills, or a heat sensation; numbness or tingling; and fear of losing control.

These reactions can be spontaneous or can occur in response to a trigger—whether or not the person recognizes it as a trigger (Landon & Barlow, 2004).

We have seen panic disorder in patients we have evaluated, although in our experience, it does not occur as often in people with Down syndrome as some of the other anxiety disorders. It may, however, be more common than has been observed. The diagnosis may not be clear-cut because many people with Down syndrome have difficulty verbalizing subjective feelings due to verbal and articulation limitations. Nevertheless, some of the symptoms are definitely observable by sensitive caregivers, as illustrated in this example:

Sean, age twenty-four, was having repeated problems at work. He would become very agitated and sometimes strike others. His mother told us that on two recent family trips, he had become similarly agitated on the plane, sweating, breathing heavily, and having trouble sitting still. We asked the staff at his workshop to observe him more closely, and they discovered he was having similar symptoms before episodes of agitation.

We diagnosed Sean with a panic disorder. He responded very well to a combination of sertraline (Zoloft) and sustained-release alprazolam (Xanax), an antianxiety medication. We also gave his family short-acting alprazolam to use at the time of the symptoms. Sean has done very well with this combination. His ability to go to work regularly without

difficulties has been therapeutic in itself. Doing well at his job and not losing control of himself is a very positive experience for him and quite rewarding. The better he feels about himself, the better is his self-esteem and the less anxious he is in many situations.

Desensitization may also be beneficial for panic attacks. However, sometimes avoidance of the situation may also be necessary. Medications, including antidepressants and antianxiety medications, may be beneficial.

Selective Mutism

Selective mutism is characterized by consistent failure to speak in specific social situations. In our experience, it is not common in people with Down syndrome, but we have observed it. In addition, some individuals develop a more generalized mutism in which they don't speak in a variety (or all) environments. Other people won't talk in some settings and only whisper in others. We have evaluated individuals who have no (or limited) other symptoms but have also seen it as part of obsessional slowness (chapter 19) and regression (chapter 27).

Treatment may include counseling, antianxiety medications, and speech therapy but may only be partially effective. In several individuals with Down syndrome, only time was the healer as they slowly lost the fear or anxiety that had contributed to the start of the mutism and eventually started to talk again. Sometimes they would only talk to certain trusted individuals with whom they apparently felt comfortable.

Distinguishing Medical Conditions from Anxiety Disorders

When diagnosing an anxiety disorder or panic attack, it is important to rule out possible medical conditions that can contribute to or mimic these problems. Ideally, the doctor will evaluate the individual for these disorders when first considering an anxiety diagnosis, as treating these medical problems, if present, may make it unnecessary to treat for anxiety.

The first consideration is hyperthyroidism (overactive thyroid). In the general population, nervousness is a very common symptom of hyperthyroidism. Studies suggest that 85 percent of people with hyperthyroidism experience anxiety (Reid &

Wheeler, 2005; Katerndahl & Vande Creek, 1983). In people with Down syndrome, nervousness is also a common symptom. Often when hyperthyroidism is medically treated, the anxiety disappears as well.

Another medical problem that can contribute to anxiety is sleep apnea. Sometimes the chronic sleep deprivation associated with sleep apnea may be manifested as anxiety. Sleep apnea may also cause hypoxemia (low oxygen in the blood) while sleeping. Hypoxemia can cause a person to feel anxious or even have a sense of panic. At night, he may awake in a panic or feel anxious when he experiences the low oxygen. During the day, there may also be a persistent sense of anxiety.

Anything that causes hypoxemia can contribute to anxiety. For instance, adults with Down syndrome who have congenital heart disease that has resulted in chronic hypoxemia (low oxygen) may develop anxiety. Particularly as the hypoxemia progresses, the sense of difficulty breathing or "air hunger" may lead to anxiety. When the person is more active, his oxygen level may fall even further, causing increased anxiety. Some adults with Down syndrome with this problem have required antianxiety medications, but often oxygen therapy can be very beneficial physically and also reduce psychological stress.

Unfortunately, however, sometimes the recommendation of oxygen may be stress inducing. For example:

> *Leah, a thirty-seven-year-old woman with cyanotic congenital heart disease, was slowly but progressively declining. Her heart function was decreasing, and she was becoming short of breath with less and less activity. When we discussed the possibility of oxygen, she was very fearful. She adamantly refused to even consider oxygen therapy.*
>
> *Through discussions with Leah, her family discovered the source of her fears. Leah's grandfather had had severe emphysema. Shortly before he died, he had required oxygen therapy. Leah equated starting oxygen therapy with her grandfather's death. She thought that if she began oxygen therapy she would also die in the near future. Over time, we were able to reassure Leah, and she began the therapy. As she experienced the benefits, she became even less anxious and began to readily use oxygen.*

Another possible cause of the symptoms of anxiety is alcohol withdrawal. Alcohol withdrawal is experienced when a person's body becomes accustomed to drinking a lot over a long time and then the alcohol is stopped. While we have only seen this extremely rarely in our practice, it is something to consider. For the three people with alcoholism we have evaluated, making alcohol unavailable was enough to solve the problem. Interestingly, two of the individuals did not later express craving for alcohol and either "forgot" or denied they ever drank heavily. The third person described cravings until she began to develop memory impairment and then seemed to forget her previous cravings.

Illegal drug usage is even rarer among adults with Down syndrome (we have not seen it) but is theoretically a possibility. Much more likely is the consumption of too many caffeine-containing beverages. Soda (or pop) is an even bigger problem than coffee. Caffeine can cause anxiety symptoms. It can also disrupt sleep, leading to anxiety. We recommend slowly weaning the caffeine-containing beverages and replacing with noncaffeinated beverages the person likes.

Finally, it is important to rule out illness as a cause of anxiety. Just as illnesses can lead to depression, they can also be anxiety provoking. This is particularly true if there is uncertainty about the diagnosis or treatment. In addition, if an adult has a limited ability to communicate his concerns, pain, or other symptoms, that can be anxiety provoking. And, as with anyone else, the person may feel stressed if he knows from previous experience, or is told, that bad outcomes or suffering may be associated with his illness.

Treatment for Anxiety Disorders

In the case histories in this chapter, we have already alluded to some of the treatments for anxiety disorders, including treating underlying physical problems, counseling, desensitization, sensory approaches, and medications. Considerations in treating underlying physical problems are mentioned above. In this section we will focus on medications and counseling for anxiety.

Medications

The medications for anxiety fall into three categories:

1. benzodiazepines,

2. nonbenzodiazepine anxiolytics,
3. antidepressants.

Benzodiazepines are a class of medications commonly known as “tranquilizers.” They enhance the effects of the neurotransmitter GABA in the brain, leading to sedative, antianxiety, and muscle-relaxing effects, among others. The benzodiazepines such as diazepam (Valium) and lorazepam (Ativan) act relatively quickly compared to other types of antianxiety medications (such as buspirone, see below). There is usually some effect with the first dose, and with increasing regular doses, the effect often increases. These medications can also be used on a short-term basis while starting other medications that take longer to take effect. One downside to benzodiazepines is that they can be addictive, and tolerance may occur over time, requiring higher doses.

Technically, *anxiolytic* means “antianxiety,” but buspirone (Buspar) is the only one of the nonbenzodiazepine anxiolytics used for anxiety. Its exact mechanism of action (how it affects brain chemicals) is not known, but it does bind to serotonin and dopamine D2 receptors. It does not have an immediate impact on anxiety. It may take several days to weeks to see the effect. However, the advantage over the quicker-acting benzodiazepines is that it does not the same potential for addiction or tolerance. We have had some, but limited success in treating anxiety with buspirone.

Antidepressants can also be used to treat anxiety. We have found the selective serotonin reuptake inhibitors (SSRIs) and serotonin-norepinephrine reuptake inhibitors (SNRIs) to be beneficial for most adults with Down syndrome who are experiencing anxiety. Among the SSRIs, paroxetine and escitalopram have FDA approval for generalized anxiety disorder, and paroxetine, sertraline, and fluoxetine are approved for panic disorder. For the SNRIs, duloxetine and venlafaxine are approved for generalized anxiety disorder, and venlafaxine is approved for panic disorder. We have also used bupropion (Wellbutrin) with some success, although it does not have FDA-approval for this use.

It may take several weeks to see the full effect of these medications; therefore, we sometimes temporarily use a benzodiazepine while we are waiting for the SSRI, SNRI, or bupropion to take effect. Some people require long-term use of antidepressants because their symptoms recur if the medication is discontinued. Antidepressants are further discussed in chapters 16 and 17.

Steve, thirty-four, was heavily sedated when we first met him. He was on a high dose of thiothixene (brand name Navane, no longer available), an antipsychotic medication. He had become aggressive in his group home and struck one of the staff people. Steve was very sensitive to activity going on around him, and we learned that he would become agitated and tremble in certain situations, particularly when other people in the home were agitated. We diagnosed him with a generalized anxiety disorder. After starting him on buspirone (Buspar), we were able to wean him off thiothixene. Subsequently, he was much calmer and more tolerant when other residents became agitated.

Counseling

Many of our patients with anxiety have benefited not only from medication, when warranted, but also from counseling. Counseling provides the person with an opportunity to express his concerns and helps the practitioner determine if and how the environment needs to be adjusted to decrease anxiety.

Edward, age thirty, and Gabe, thirty-nine, were brought for evaluation by their respective mothers because of similar symptoms of a generalized anxiety disorder. Their symptoms consisted of agitation, body tension, pacing, and difficulty sleeping. Both also had mild self-injurious behavior. Gabe chewed on his hands, while Edward picked at sores on his arms and legs. Both had lost their fathers some time ago, and their mothers were struggling with some of their own issues of loss, having experienced deaths of many family members and friends as they aged. Both Edward and Gabe were sensitive to their mothers' feelings of loss, which definitely created some anxiety for them. But job-related stress seemed to have precipitated a dramatic increase in their anxiety.

Edward worked in a cavernous building, and his job had become more and more onerous to him as the noise level and conflicts between

others increased. Making matters worse, several men at his worksite teased him unmercifully once they learned that their teasing upset him.

Gabe had a different problem. The agency managing his employment changed from a workshop setting to a program that found and placed people in community jobs. Although this was a laudable goal, they closed the workshop before they had found community jobs for most of the program participants. As a result, most people reported to the inactive workshop and sat idle for hours on end. This was extremely difficult for Gabe, who needed to work and be productive. After Gabe had spent six months of sitting doing nothing, his level of anxiety and hand biting had increased dramatically.

Edward and Gabe were brought for evaluation within several months of each other. After we ruled out any significant physical health problems, a diagnosis of generalized anxiety disorder was given to each, and treatment plans were developed. Antianxiety medications were prescribed for both to help reduce the intensity of their anxiety and to aid them with sleep. Counseling was also recommended and started for both families.

Edward and his mother had both joint and individual counseling sessions to discuss their feelings and issues. Edward was quite verbal and was able to discuss many of his concerns openly and honestly. Sessions with Edward's mother focused on her own needs for more friends and fulfilling activities (which she found through greater participation in her church and a local social club).

Counseling for Gabe and his mother was different, in part because his mother's family viewed counseling as an admission of failure. As a result, she was less able to articulate her own issues and concerns for herself. Therefore, we did not offer, and she did not request, individual meetings for herself. Nevertheless, she was very concerned with helping us find a solution to Gabe's problem and was extremely helpful in teaching us what we needed to know about Gabe. For example, she told us how important Gabe's work was to his sense of well-being. Gabe met for

several short sessions (fifteen to twenty-five minutes) of individual counseling. These meetings were productive for him, even though he was not as verbal or articulate as Edward. (See chapter 16 for more on counseling adults who are less verbal.)

Despite some differences, there were several key similarities in our counseling strategies for both Edward and Gabe. First, we held meetings with the staff at their respective worksites regarding problems at these sites. Second, sessions were held with both men and their mothers to introduce relaxation techniques to help them deal with anxiety.

Both families went with us to meet with worksite staff and administrators to try to agree on a solution to the problems at these sites. We were successful in negotiating a positive change for Edward, who was allowed to move to a smaller and quieter workspace. We were less successful with Gabe, at least initially. His staff could not or would not find suitable work for him either at his current worksite or in the community. Fortunately, through the combined efforts of Gabe, his mother, counseling staff, and a local case management agency, Gabe was able to move to a new worksite that was near his home and had plenty of work for him.

As mentioned above, both Gabe and Edward were taught to use relaxation techniques during counseling. These were planned, practiced activities that gave them some sense of control over their anxiety by enabling them to relax when faced with anxiety-provoking situations. To develop the right relaxation activities for someone with Down syndrome, we try to capitalize on a person's interests and strengths. For example, for Linda (above), who was terrified of thunderstorms, we used her superb memory and interest in pictures of past experiences to help her focus her attention on these positive experiences. She learned to use this technique both when actually experiencing storms as well as when concerned about the possibility of a thunderstorm. (See more on the use of visual memory techniques in chapter 16.)

We found that Gabe, like many individuals with Down syndrome, had certain set routines or grooves that he did every day. One of these repetitious activities was

copying letters or words in notepads. He would do this for hours at a time, particularly in the evening. This was extremely relaxing for him. Unfortunately, he was so anxious as a result of his worksite problems that he stopped doing this activity. In individual and joint meetings with his mother, we encourage him to resume this activity. We then had him progress to copying words to relax when he was in a stressful situation. At first, his mother or another caregiver needed to remind him to do this activity when he was stressed, but in time he became skilled at identifying when to start it himself. For example, Gabe had some stress and anxiety at his new workshop program when there was some downtime. His new supervisor reminded him just once or twice, and then he began to do this task routinely whenever he had downtime or stress at work—or anywhere, for that matter.

For Edward, we used modifications of two well-known relaxation strategies used in the general counseling field. One strategy is called progressive muscle relaxation. This technique involves tightening and then relaxing different muscles of the body. The second strategy is to control one's breathing, such as through deep breathing and exhaling exercises, to induce relaxation.

For teens and adults with Down syndrome, we use versions of these techniques adapted to the ability of the person. We usually depend on a simple isometric exercise, which may be combined with a breathing exercise. Isometric exercise employs muscle resistance created by the person himself. For instance, the person pushes the palms of his hands together. Like progressive relaxation, this exercise may include different muscles and different parts of the body. People may push or pull their hands, push down or pull up their legs, etc. Typically, we teach the person to do each exercise for five seconds while he or another person says "go" and then counts "1,001, 1,002, 1,003 . . . stop." When someone else counts aloud, some people may also be able to add a breathing exercise. This involves taking in a deep breath when doing the exercise and letting go of the breath when the exercise is complete. Additional information on relaxation strategies can be found in chapter 16 and on sensory techniques in chapter 12.

Some people simply do not like or are not able to use these techniques, but many others, like Edward, can. We have had some success with other types of relaxation techniques, again modified to meet the needs of the person with Down syndrome. Some people have been able to use smooth stones, called rubbing stones or worry stones, which they rub with their thumb as a healthier and socially appropriate means to express some tension or anxiety. In public, people are often able to keep these stones in their pockets and rub them unbeknownst to others. A number of

different items may be used besides rubbing stones, such as stress balls or rabbits' feet, to allow people to keep their hands busy on more acceptable means for expressing tension.

To summarize the treatment and counseling strategies for Edward and Gabe who both were diagnosed with generalized anxiety disorder, the treatment consisted of a multipronged approach that included

- a complete physical to rule out health problems;
- medications to reduce the intensity of the anxiety symptoms and promote better sleep;
- counseling (which allowed both individuals and at least one family member to express their feelings and concerns and helped at least one parent to become involved in beneficial social activities);
- interventions at their worksites to promote healthy, less stressful work environments;
- relaxation techniques tailored to each individual's needs, skill level, and interests to help them relax in the face of anxiety-provoking situations.

Although both men in the example above happened to have a generalized anxiety disorder, we have found that counseling may be essential to the successful treatment of any type of anxiety disorder in people with Down syndrome. For example, people who require desensitization training may benefit from counseling to help them better tolerate the process of being gradually exposed to a feared thing or activity. Counseling helps the person to express his fears and concerns more fully. Counselors are then able to help develop a plan that is better attuned to the individual's issues and fears. Whether or not the training manuals for these types of treatments specify the need for counseling, in real life with real people, there is nothing more important to the success of these endeavors than to develop a therapeutic alliance through the counseling process. We find, too, that different types of counseling involving different numbers or groups of people, including the individual, family, and staff from worksites or residences, may all be necessary and helpful in resolving different types of anxiety disorders.

Conclusion

Anxiety can be a very debilitating illness. As with other mental illnesses, assessment and treatment must be directed toward underlying medical conditions, psychological issues, social issues, and the use of medications. After treatment is completed, anxiety may recur, so continued observation for symptoms is important.

Return to Table of Contents

Chapter 19

Obsessive-Compulsive Disorder

Jill, age twenty-five, enjoyed working with Carmen, a young staff person at her job. She became obsessed with Carmen and started drawing pictures of her, following her around at work, calling her repeatedly, and putting notes on her car. Jill even cut and saved magazine pictures of the model of Carmen's car. This obsession interfered significantly with Jill's daily life because she became so preoccupied with her obsession that she was unable to sleep, work, and participate in recreational activities. Although Jill received counseling and her symptoms improved, they did not stop and Jill's ability to function was still impaired by her obsession with Carmen. Jill was then prescribed sertraline (Zoloft), and her symptoms continued to improve but did not resolve. Ultimately, Carmen changed jobs, and Jill's obsessions diminished enough that she regained her usual level of function.

As discussed in the chapter on grooves, many people with Down syndrome have a tendency toward sameness and repetition. Grooves may be very beneficial—for example, if they enable a person to reliably complete routine self-care and work tasks. Grooves may become problematic, however, if thoughts or actions become stuck or rigid. Sometimes this tendency may lead to problems that are diagnosed as obsessive-compulsive disorder. Other times, however, the development of obsessive-compulsive disorder has no connection to previous grooves.

What Is Obsessive-Compulsive Disorder?

Obsessions are thoughts that preoccupy the mind. Compulsions are acts that one feels compelled to perform. In "classic" obsessive-compulsive disorder (OCD), these compulsions are linked to a desire to lessen the anxiety arising from the obsessions. For example, someone who is obsessed with the idea that he is going to accidentally burn down the house might constantly be compelled to check and

recheck that the stove is turned off. When he sees that the stove is off, his anxiety is temporarily reduced until he starts worrying about burning the house down again.

Ordinarily, people with OCD realize that their obsessions and compulsions are abnormal or excessive and would like to be rid of them so they will also be rid of the anxiety. In contrast, people with Down syndrome often don't have the desire to be rid of the obsessions and compulsions.

Compulsive behaviors that interfere in people's lives can be just as problematic as obsessions. For people with or without Down syndrome compulsions may be odd or nonsensical. For example, Sam, a thirty-six-year-old man with Down syndrome, had a compulsion to turn glass and other breakable objects around and hang them off the edge of shelves in his apartment. He seemed compelled to do this activity over and over. His compulsion resulted in many broken items, and it interfered in essential work and recreational activities of the people he lived with.

OCD occurs in about 1.5 to 2.3 percent of the general adult population in a given year and 2.5 percent of people over their lifetimes (Kessler et al., 2005). OCD seems to be more common in people with Down syndrome. Approximately 6 percent of the patients we have evaluated have been diagnosed with OCD.

Symptoms of OCD

Obsessions are characterized by recurrent and persistent thoughts that are more than just excessive worry. Compulsive behavior includes repetitive actions or speech that the person is driven to perform. The symptoms become problematic when they are disturbing to the individual and/or interfere with the activities of daily life. For example, if someone feels compelled to ensure that group home staff are doing their jobs and stays up all night to check on them, this would definitely interfere with his daily activities. Symptoms also need further evaluation if the person becomes very agitated, upset, or angry when someone interferes with his routine. For example, an adult with Down syndrome may be compelled to turn all the lights on and then become very angry when asked to stop because it is time to go to sleep (with the lights off).

Causes of OCD

People with OCD are thought to have an abnormality in the serotonin system (a reduction in serotonin or an abnormality in serotonin receptors). People with Down

syndrome are thought to have a greater incidence of abnormalities in serotonin and, therefore, seem to be more susceptible to OCD. In addition, stress, the amount of support from family and others, and other precipitants to mental illness (as discussed in chapter 14) can contribute to development or nondevelopment of OCD tendencies.

Diagnosis of OCD

When diagnosing OCD in adults with Down syndrome, it is often necessary to deviate somewhat from the diagnostic criteria for OCD listed in the *Diagnostic and Statistical Manual of Mental Disorders*, 5th edition (DSM-5). The *Diagnostic Manual-Intellectual Disability 2: A Textbook of Diagnosis of Mental Disorders in Persons with Intellectual Disability* (DM-ID 2) can be helpful because in includes modifications for unique features associated with people with intellectual disabilities.

In people without Down syndrome or an intellectual disability, OCD may be diagnosed if they have either obsessions or compulsions that are debilitating. In many cases, clinicians find that compulsive behaviors are linked to obsessions. This is because people will engage in compulsive behavior in an attempt to ward off a disturbing thought or fear that has become an obsession. However, given the expressive language limitations of many people with Down syndrome, it can be difficult to determine whether or not their compulsive behavior is linked to obsessive thoughts. Additionally, as mentioned above, OCD is not typically diagnosed unless the person recognizes his symptoms as being abnormal or undesirable. With people with Down syndrome, however, we may diagnose OCD even if the person seems to take pleasure in his obsessions or compulsions. A key component is the impact on daily function.

On the other hand, it is important not to attach too much importance to the presence of compulsive-appearing behavior alone. The presence of grooves in people with Down syndrome can be overdiagnosed as OCD. Careful assessment of the symptoms, bearing in mind that people with Down syndrome do tend to have grooves, is essential to making an accurate diagnosis. Attention must also be given to those who interact with the person with Down syndrome. If those around him do not appreciate the normalcy of grooves, they may contribute to the situation becoming problematic, as this example illustrates:

Lynn, age forty-three, really liked to sweep the floor in her residence. She was compelled to sweep and was very good at it. The staff had set up a system in which each of the four after-dinner jobs were divided among the four people who lived there. However, on the nights Lynn was assigned to do a job other than sweeping, she took the broom from the person assigned to sweep, which confused and frustrated her housemates. The staff also became frustrated with Lynn's behavior and concerned that she had OCD. But in fact, this was a case of a groove that was not being appreciated. Lynn's housemates did not care if they swept or not, and Lynn was a good sweeper. Letting her sweep every night did not eliminate her groove, but it did eliminate the problem. With a modification in the schedule that didn't negatively affect Lynn or her housemates, there was no longer a problem; there were, however, very well swept floors and peace in the house.

Keeping a record of the person's behaviors and reporting these to the professional who is assessing him or her for OCD can help prevent a misdiagnosis. A record of his typical groove, how it has been managed in the past, changes in the pattern of behavior, and stresses that may have contributed to a change can provide valuable information. In addition, remember that for a diagnosis of OCD to be made, the behavior must significantly interfere with the person's ability to complete day-to-day activities. It is therefore very useful in diagnosing OCD if parents or other caregivers can keep track of the amount of time the person spends engaging in his obsessions or compulsions and how that interrupts his life.

Treatment of OCD

Sometimes, no treatment of OCD is necessary. This might be the case, for instance, if a compulsion or obsession is somewhat annoying but does not interfere significantly with daily activities:

Daniel, age fifty-eight, had a compulsion to touch Kleenex boxes. Whenever he came into our office, he would stand up about every five minutes or so, walk over to the Kleenex box and touch it, and then sit back down. He did similar activities at his home. Although his compulsion was odd, it did not ordinarily interfere in essential activities of his life. However, after Daniel experience a period of stressful losses and developed hypothyroidism, his compulsion did become a problem. He was not sleeping at night because he was getting up repeatedly to touch a Kleenex box. He was treated for hypothyroidism and given a mild sleep aid. Once he was able to sleep, he continued to touch Kleenex boxes during the daytime, but, as before the onset of his sleeping difficulties, this did not interfere with his daily work and home activities.

To summarize, treatment of OCD in teens and adults with OCD is necessary under these three circumstances:

- if the OCD interferes with the person's life activities;
- if it is a major cause of strife within the family;
- and/or the obsessions or compulsions distress the person.

Treatment of OCD is multifaceted. As described in chapter 16, treatment includes attention to psychological, social, and biological issues. Treatment components may include redirection, medication, or a combination of strategies.

Redirection

For OCD, redirection is an important aspect of the psychological and social treatment approaches. By redirection, we mean trying to interest the person in another activity either just before or just after he begins a compulsive or obsessive activity. Keys to successfully redirecting someone include the following:

1. Select an interesting or preferred alternative activity in advance.
2. Do not get angry when trying to redirect the person.
3. Suggest rather than insist that the person try the other activity. Try this over the course of days; expect gradual, not instantaneous changes.

4. Offer rewards for doing an alternative activity; this may help the person get started on the activity.
5. Only select one obsession/compulsion at a time to reduce.
6. Remember that physical prompts may cause the person to become agitated.

Working with the person and others in his environment to redirect him away from the object, person, or activity involved in the obsession or compulsion can help reduce the obsession or compulsion and encourage him to participate in other activities. In the above examples, both Jill and Sam benefited from staff attempts to redirect them toward more beneficial thoughts and activities.

Medication

Medications can be of tremendous benefit in treating obsessive-compulsive disorder. Medications often reduce the strength and intensity of an obsession or compulsion enough to allow redirection to work more effectively. This was certainly the case for Jill, the women who was obsessed with her coworker. She was prescribed an antidepressant medication (selective serotonin reuptake inhibitor, SSRI) that is also used to treat OCD. As a result, Jill was more amenable to staff attempts to shift her obsession with Carmen to more normal work and recreation activities.

Note that to successfully treat OCD, it is often necessary to use higher doses of the medications than are used for other disorders and to use them for a prolonged period of time (and sometimes indefinitely).Therefore, it is particularly important to consider possible side effects of a medication before prescribing it and then monitor the person to ensure the benefits outweigh any downsides.

When prescribing medications for OCD, we have had the best success with the selective serotonin reuptake inhibitor (SSRI) medications. We most commonly use sertraline (Zoloft) because of its effectiveness and low incidence of side effects. Other SSRIs approved by the US Food and Drug Administration (FDA) that we have found beneficial include paroxetine (Paxil) and citalopram (Celexa). Paroxetine in particular can help stimulate the appetite of people experiencing a decrease in appetite as part of their OCD symptoms. Fluvoxamine (former brand name Luvox) and fluoxetine (Prozac) are also SSRIs approved for OCD, but we tend to use them less because we have found fluvoxamine to be less effective and fluoxetine to have more side effects. Escitalopram (Lexapro) has also been used effectively by some, but it does not have FDA approval for OCD.

We have also successfully treated OCD with the serotonin-norepinephrine reuptake inhibitors venlafaxine (Effexor), duloxetine (Cymbalta), or desvenlafaxine (Pristiq), but these are not FDA approved for OCD. We have not used the serotonin modulators for OCD, and they are not FDA approved for that indication. Bupropion (Wellbutrin), a norepinephrine-dopamine reuptake inhibitor, is not FDA approved but has been used by some with mixed results to reduce appetite when increased eating is part of the compulsive behavior.

For some adults with Down syndrome, obsessive-compulsive disorder seems to take on a psychotic nature. The obsessions or compulsions go beyond the usual level, with a greater degree of self-absorption, detachment, and inability to participate in activities of daily life. For these individuals, a small dose of an antipsychotic medication (in addition to or in place of the SSRI), may be very helpful. Risperidone (Risperdal), olanzapine (Zyprexa), quetiapine (Seroquel), ziprasidone (Geodon), lurasidone (Latuda), and aripiprazole (Abilify) have all been found to be beneficial. None of these is FDA-approved for OCD, but they are approved for schizophrenia, a psychotic illness. When difficulties sleeping are a problem, we select olanzapine because it is usually more sedating than the other choices, but it also seems to cause the most weight gain.

Unfortunately, weight gain is a potential problem with all these antipsychotic medications. This concern must be addressed through good nutrition and increased activity. As mentioned above, we have seen the greatest weight gain with olanzapine. Sometimes this side effect can be used in a positive way for patients who have weight loss or decreased appetite as part of their symptoms. Ziprasidone, lurasidone, and aripiprazole seem to cause the least weight gain in our patients, but for some individuals, all of them can cause weight gain. Ziprasidone can cause cardiac rhythm disturbances, so periodic EKG monitoring is recommended. In addition, tardive dyskinesia is a potential side effect of all these antipsychotic medications. Tardive dyskinesia most commonly occurs after long-term use of a medication and causes the person to have involuntary movements, grimacing, or similar problems. We have only seen tardive dyskinesia in a few adults with Down syndrome out of the several hundred who have been prescribed these drugs.

To limit weight gain, some providers add metformin (e.g., Glucophage) when a patient is taking one of the antipsychotic medications. We have had limited success adding metformin to reduce weight gain.

Combining Treatments

Sometimes when compulsions or obsessions are more difficult to resolve, treatment requires both creative approaches to redirection and the use of psychotropic medication. The first step in treating these more complicated obsessions and compulsions is always to look for causes or precipitants. We try to pinpoint possible health and environmental stresses through the evaluation process described in chapter 15. We also talk with the adult, family members, or others who know him well to get some history of his previous groove-like tendencies. This gives us a sense of the relative strength of this tendency for the individual.

In addition, we find out how family members and other caregivers have been responding to the person's grooves. How others respond to grooves may play a critical role in either the development or reduction of a problem, as in the following example:

Charles had lived with his parents all his life. When he was forty-three, his last parent died and he moved in with his married sister's family, which included young school-aged children. Although his sister, Zoe, had always been actively involved in Charles's and his parents' lives, the transition to her household was difficult. Zoe reported that Charles had always had a strong tendency for following set routines and that both he and his parents had become more set in their ways as they got older and assumed a more sedentary lifestyle.

When he moved to Zoe's house, Charles had habits and routines that did not always fit in with the family routines. For example, Charles demanded ice cream at 9:00 at night, which disrupted his sister's attempts to get the children to bed. After several weeks, Charles was able to adjust to an earlier time for ice cream and was even able to substitute healthier snacks. He also had a habit of bringing his dirty clothing to the basement every night after he changed into his pajamas. His mother had dutifully washed and returned the items to him the next day. This was impractical for his sister. After repeated practice and much

encouragement, Charles was able to put his clothing in a hamper, which was brought to the basement once a week to be washed.

There were other issues as well. For example, one of Charles's favorite activities was buying toiletry and other personal items at the store. While living with his parents, he had developed a habit of buying duplicates of items he already had at home. Zoe found out about this habit the first time they went shopping and Charles refused to leave the store unless he got the items he wanted. To prevent this problem from recurring, Zoe helped him develop a new routine before going to the store. The first step involved looking in his bathroom cabinet to see what items he truly needed. Zoe would then help him find a picture or make a drawing of the item, which he would take with him to the store to locate and buy. Charles was very pleased with this alternative because it gave him a routine and also a sense of independence and purpose. Equally important, it allowed him to cooperate with his sister's needs and wishes.

Later on, Zoe discovered a similar problem with movie rentals. Charles often wanted to rent movies that he already had at home. Following the tried and true parental strategy of picking one's battles carefully, Zoe wisely chose to let him do this activity. This practice did not cost much money, and, perhaps more importantly, it did not result in the accumulation of unnecessary items. Many situations like this occurred over the course of a year but were solved effectively (and often very creatively). In time, Charles developed routines that fit into those of his sister's family.

After two years with his sister, Charles moved into a nearby group home. Once again, he had difficulty adjusting to his new living situation over the course of the first year. Fortunately, the house manager and key staff members were experienced and understood Charles's need for routines. Staff wisely listened to suggestions from Zoe, whose knowledge and experience was an invaluable aid during this process of adjustment.

Unfortunately, toward the end of Charles's second year in the group home, there was a great deal of staff turnover. The new house manager ignored Zoe's offers of assistance because she viewed Charles's behavior as oppositional, rather than related to his need for routines. Over the next few months, Zoe watched with growing concern as problem rituals and routines that had been dealt with effectively in the past reemerged. For example, Charles once again began to take his laundry to the basement to be washed. He began to refuse to go to bed until he had his evening ice cream, even though previous staff had successfully substituted a healthier snack at an earlier time. Most problematic, however, was the new staff members' refusal to follow Zoe's proven strategy of providing Charles with a picture of needed items before going shopping. As a result, Charles once again refused to leave stores without buying his desired item.

Unfortunately, when the staff attempted to force Charles to comply, he had "aggressive outbursts," which he had never shown toward any previous caregiver (family or staff). One of these outbursts occurred on a Friday evening when Charles was with an inexperienced staff person. The police were called to the store, and Charles was transported to a local hospital emergency room for a psychiatric evaluation. There he was given an antianxiety medication and recommendations for follow-up for psychiatric treatment. This was the last straw for Zoe, who requested an emergency meeting with the staff and us to help resolve the problem. Unfortunately, the "aggressive incidents" only seemed to solidify the house manager's belief that this was oppositional behavior and that Charles was a danger to staff and other residents.

This situation was resolved only after Charles moved to a new group home. Predictably, his response to the stress of the move was to become even more rigid in his routines, and on several occasions, he was aggressive toward staff and other residents. During this transition period, he was started on an antidepressant medication that helped reduce his

anxiety and the rigidity of his compulsive behaviors. Fortunately, the new staff people were patient, took a positive view of routines, and collaborated with the family and us to ease his transition. In time, Charles became comfortable with his new home. He has now lived in this residence for several years and is doing extremely well.

Common Compulsions

Among the most common types of compulsions we have seen in people with Down syndrome are the following: (1) ordering, (2) hoarding, and (3) excessive rigidity in completing a routine.

Ordering

Putting objects in order, or "ordering," is a common and often beneficial groove among people with Down syndrome. Many people order pictures or collected items in their bedrooms. Similarly, some people have a need for a sense of order when it comes to lights. They are only in order when they are turned on (or off, depending on the individual). Closing or opening doors is another behavior that establishes a sense of order for certain individuals. However, ordering becomes maladaptive when the person's life increasingly revolves around the need to put things in order. This was clearly the case with Sam, who was described at the beginning of this chapter. He had a compulsion to put breakable objects in a certain set place. Ordering becomes a problem, as it did for Sam, when people begin to miss work, social gatherings, or previously desired activities to continue to arrange certain items "just so."

Ordering may be a form of compulsion that reduces the anxiety the person feels if things are not arranged just right. It may also reduce stress that is being experienced from other causes. For example, after a rough day at school, one preteen girl got extremely irritable and out of sorts if she was not allowed to put her things just how she wanted them. She was obsessed with keeping the top of her dresser tidy, for instance, and felt compelled to put things back right where they were if someone moved them. She would insist on having the blue pillowcase on her pillow because the yellow one did not look "right" with her bedspread (and she would hunt all around the

house for the missing pillowcase and hound her mom mercilessly until the “right” pillowcase was found).

In addition, ordering things can be related to the person wanting to assert control over her life. In this situation, ordering appears to give the person a sense of control.

Hoarding

Hoarding items is another compulsive behavior we frequently see. Hoarding may go hand-in-hand with ordering objects. Sometimes teens and adults with Down syndrome collect and store excessive quantities of specific items; unnecessary items (pens, soap); useless or other nonsensical items (trash, ripped paper); or food (including perishable items). As described above, Charles had a tendency to save soaps and other grooming supplies. Hoarding can be a problem if the individual cannot participate in his usual activities because he is so focused on hoarding items. It can also become a public health concern if the person hoards perishable food items, trash, or other things that make for unsafe conditions.

It can be difficult to redirect hoarding. The first objective is to help the person avoid hoarding items that create an unsafe situation. Replacing the item(s) with something that does not create an unsafe situation is recommended. The next step is to help the person put some limits on the hoarding. As with Charles, helping him developing an understanding of what is needed by using pictures and giving him control of buying needed items can be helpful. Making available other fun and interesting activities may also be beneficial. The staff of one agency took pictures of Don’s favorite collectible items (packs of gum, combs, pens, and more) and put a certain number of the pictures of each item on a large piece of cardboard (e.g., three pictures of packs of gum, two pictures of combs, etc.). They worked with Don so that he agreed that at the end of the day, he would empty his pockets and backpack and place the items on their corresponding pictures. The pictures were a good visual reminder that when he had more than the agreed-upon number, he would give away those extra items.

Rigid Routines

Perhaps the most common type of maladaptive compulsion we have seen among adults with Down syndrome is being overly rigid in completing some routine

activity. This was clearly a problem for Charles (above example), as well as for Susan, the woman described in chapter 10 who refused to change her bath time to go out with residents of her new group home. These types of rigid routines seem to occur most often when people are asked to make dramatic and difficult changes in their lives.

We frequently hear of maladaptive bathing routines. This occurs when people take baths or showers repeatedly and for extended periods of time. Such bathing routines are no longer productive when they interfere with work or social activities. They can also cause dry skin. Often, parents or other caregivers have no idea what led to the development of these maladaptive routines. However, they frequently report that there has been stress in the person's life, such as a move, and that the person had a preexisting tendency to take longer baths or showers. We have not heard reports that extended bathing or showering in people with Down syndrome who are under stress is linked to an obsession with germs (a common compulsion in those without Down syndrome), but it may be related to concerns about cleanliness.

The compulsion to do things in a certain order during the day or through the week is also fairly common and can be considered a form of ordering. For many people, this is part of a healthy groove, as discussed in chapter 10. However, this tendency can be problematic and consistent with OCD if it significantly disrupts daily life. Care must be taken, however, to avoid overdiagnosing OCD on the basis of ordering behavior, because this tendency is so common and part of the typical behavior for many people with Down syndrome.

In addition to stress, underlying health problems are a common precipitant to overly rigid routines. When an adult with Down syndrome is diagnosed with OCD, it is imperative to look for a physical problem that could have triggered the problem. Sometimes a health issue that creates sensitivity to a specific area of the body can lead to rigid routines, as in this example:

> *Jamie, thirty-two, had a surgery that required an incision across her lower abdomen. During her recovery, she had quite a lot of bleeding and needed a frequent change of bandages. After leaving the hospital, Jamie began to shower morning and night for excessive amounts of time. She also developed a habit of fixing her underwear until "just right." Her*

family believed that these rituals were an attempt to manage a feeling of uncleanness or contamination linked to the heavy bleeding in the hospital.

As a result of her extended showers and "fixing" behavior, Jamie was getting to work later and later in the morning. She also began to spend increasing amounts of time in the bathroom at work fixing her underwear, which began to interfere with her work. In the evening, her shower and fixing behavior interfered with social and recreational activities.

For Jamie, treatment included the use of an SSRI medication and behavior management to take the edge off her compulsive routines. Resolving her problem also took a great deal of patience and persistence from her family. To limit her time in the bathroom, she agreed to use a timer, which signaled an end of her shower and was then reset to signal the end of her fixing behavior. As an incentive, Jamie was given cards representing a specified amount of money she could us to buy her beloved music CDs. Gradually, the timer was set to shorter times. After approximately three weeks, Jamie was back to a more normal pattern of showering and fixing of underwear. This then became her regular routine and the incentive was no longer necessary.

Interestingly, Jamie's family noted that as she spent less time arranging her underwear, she began to substitute a new ritual of arranging certain personal items in her room "just so." Fortunately, she was far less rigid about the time spent on this activity and was able to continue her daily tasks without getting sidetracked on this activity.

At the same time that Jamie's family was giving her incentives to alter her bathroom routines, staff at her job provided her with incentives to return to work more quickly after visiting the bathroom to fix her underwear. These incentives included time and attention from her favorite staff person and also a prized job of delivering mail if she returned from the bathroom after lunch in a timely manner. Eventually, these incentives helped her regain a more normal work pattern, and this

new behavior then became her routine and she no longer required incentives.

Many other compulsions can result from health issues. For example, for some people, chronic sinusitis may lead to compulsive nose blowing or wiping, coughing, or throat clearing. For others, excessive handwashing may interfere with social and work activities and lead to serious problems with dry skin. For example, Allen began to wash his hands excessively after an extended period of persistent diarrhea. Even after he recovered from the diarrhea, he continued to wash his hands often, for prolonged periods of time. For many people with Down syndrome, OCD seems to be the outlet or the manifestation or expression of the stress associated with a variety of physical, psychological, or social stresses.

Dos and Don'ts in Dealing with Compulsive Routines

For people like Allen and others who develop compulsive routines, we recommend first determining whether the behavior really interferes in the person's life. If it doesn't, it may not be worth the effort of pushing to change the behavior. If it does interfere with some key area of functioning, or is a problem because it occurs in a public setting, we suggest these steps:

1. Try to keep the person busy with physical activities or activities that require some attention or concentration. At home, involving the individual in sports and recreation activities or even playing video games can reduce the time he spends on compulsive routines. On the job, ensuring that he has meaningful work to occupy his time can do the same. In our experience, people with Down syndrome are more likely to engage in compulsive activities when they have nothing else to do.
2. Limit sedentary activities such as watching TV.
3. Consider replacing the problem behavior with a more appropriate alternative behavior. This strategy often involves trial and error and creativity to find the right alternative behavior. Sometimes the alternative may be fairly simple. For example, some people with sinus problems may simply need to use a Kleenex (instead of their hand) when repeatedly blowing or wiping their nose. Sometimes the substitute behavior requires more thought. We have found that a worry stone, rabbit's foot, or foam- or sand-filled stress balls may keep

people's hands busy enough to begin to reduce the repetitious habit. With encouragement, the behavior may become a more appropriate regular habit. If encouragement alone doesn't work, a concrete reward may help some people.

It is usually best *not* to try to forcibly stop a compulsive behavior. Gently directing the individual to a new, healthier behavior seems to be a more successful strategy than just trying to stop the concerning compulsive behavior.

Obsessions

As previously defined, an obsession is an idea or issue that preoccupies the mind to the extent that it interferes with the ability to focus on other ideas or issues. It is more than just daydreaming. The person is limited in his ability to control the thought process.

In contrast to compulsions, obsessions can be harder to detect in a person with Down syndrome. That is, you can observe a compulsion, but an obsession takes place in the mind. Therefore, if the person is not very verbal or does not talk about his obsession, it may require some detective work to discover that he is obsessing about something. If the person is not verbal, he may repeatedly show a desire to see the source of his obsession (either the actual person or item or a picture of them).

Adolescents and adults with Down syndrome can develop obsessions about anything that particularly interests them. Some of the more common obsessions we've seen include the following:

- obsessions about fictional people (movie or TV characters) or people the person does not associate with in real life (celebrities),
- obsessions with people the person knows,
- obsessions with food.

People as Objects of Obsession

We have found that obsessions with people may be either negative or positive, and they are some of the most intense and persistent of obsessions. We believe this is due to the complex and provocative nature of human relationships, which may be fertile ground for the development and maintenance of intense obsessions. Sometimes the object of an obsession is an imaginary person or a celebrity. Sometimes the object

of an obsession may either look like or act like someone else in the person's life. This person may be gone (due to a move or death) or may be someone whom it would not be possible or advisable for the adult to talk to directly.

We are rarely able to understand the real cause of someone's obsession. Even people without Down syndrome cannot usually articulate why they have a particular obsession. It is even more likely for people with Down syndrome to have difficulty expressing the underlying cause of an obsession. We do know, however, that as with other types of grooves, stress may play a major role in the development of a problematic obsession.

Preoccupations with Imagined Others

We have seen a number of people who are intensely preoccupied with an imaginary person or a celebrity whose movie role or persona is the object of the obsession. Sometimes the adult has had fleeting contact, such as at a concert, but in general there is little contact with the person who is the object of the obsession.

Many parents of preteens and teenagers without Down syndrome have reported celebrity obsessions as a phase of adolescent development. (Remember: adolescent behavior often occurs at an older age for people with Down syndrome; see chapter 11.) This type of obsession usually subsides with age. However, in some instances the obsession continues and begins to interfere with normal functioning, as was the case with Sheri.

Sheri experienced a number of intense stresses, including the untimely loss of her beloved stepfather, who died of a massive heart attack. After this loss, Sheri developed an obsession with members of a rock band. (She had previously had a harmless interest in the band.) She created an imaginary family of these band members. Other individuals were also added to this imagined family, including a fireman who had been discussed in the news as a local hero. Sheri's mother believed this first responder took on a role of a protector in her imagined family.

Sheri became more and more obsessed with her imaginary family, to the detriment of her normal life activities. She would listen to the band's music or talk incessantly about her imagined "family members" to others

and through her self-talk. Her interest in the fireman developed into an obsession with anything in the news about the fire department. Over time, her obsessions with imagined friends interfered more and more in her life. Even though Sheri loved her job and was a conscientious worker, her work suffered as she spent most of her time in her fantasy world. A previously willing participant in social and recreation activities, she began to withdraw to her room and her imagined world.

Treatment for Sheri included an SSRI (sertraline), coupled with an intensive strategy to get her to return to her real-life work and social interests. After several months with no change, a small dose of an antipsychotic medication, risperidone, was added to the antidepressant medication, and she began to respond favorably to this treatment. The medications helped to reduce the intensity of her obsessions, and perhaps more importantly, helped to reduce her resistance to participating in real-world activities.

The first step in reawakening Sheri's interest in activities was to focus on her strong desire to manage her weight. With a little encouragement, she agreed to maintain an exercise chart of daily walking and stationary bike riding. This helped her live in the here and now and bring her out of her room. Over several months, she was bolstered by the successful loss of eleven pounds. She also agreed to keep a journal of her thoughts for discussion at ongoing counseling sessions. This served a number of important purposes. It saved her family from some of the frustration of hearing her repeated and relentless comments about her imagined family. If her comments went on for any length of time, they could direct her to her journal to write out her thoughts, which she did. Counseling gave her someone other than family members who would listen to her, although in a time-limited format (which is the nature of counseling).

Although the counseling may have seemed to encourage her attention to her imagined family, writing her thoughts actually reduced

the obsession. Putting her thoughts down on paper gave them concrete form and structure, which somehow reduced the need for her to verbalize the thoughts over and over. It may have also been enough of a real activity to help her come out of her fantasy world. The counselor also gave her homework assignments, such as listing some of her favorite activities to further encourage her attention on real-life events and activities. In time, Sheri also began again to attend social and recreational activities in the community. This allowed her to reestablish her friendships and to get additional exercise, as well as to move more into the real world and out of fantasy.

Preoccupations with Real People

We have also seen quite a few people with Down syndrome who have had positive or negative obsessions with real people in their lives. Positive obsessions mean having pleasant, enjoyable thoughts about the person; negative obsessions mean having bothersome, upsetting thoughts about the person. Here is an example of a negative obsession:

Jennifer, age thirty-two, lived in a cottage in a larger residential facility. She was accompanied to her appointment by her brother Keith, who lived nearby, and staff from her residence. Keith reported that she had a history of negative obsessions, often occurring at stressful points in her life. The object of her present obsession was another resident in her cottage. As in the past, this obsession took the form of constant complaints about this woman, expressed most often through bouts of angry self-talk. Jennifer also complained to staff or to her brother about the woman. Her brother noted that the obsession was typically accompanied by other compulsive behaviors such as a tendency to be more "stuck" or rigid with her routines. Additionally, she had been very meticulous with her self-care and reliable with work routines but now was

so absorbed with her obsession that she required more reminders to do these tasks. Despite repeated attempts by her brother and staff to distract her from her obsession, it persisted and worsened.

Aside from having obsessions and compulsions, Jennifer was also depressed, as evidenced by her irritable mood, restless sleep, poor appetite, and loss of interest in the things she had formerly enjoyed, such as music and dancing. She tended to stay by herself in her room, complaining about the object of her obsession.

For Jennifer, successful treatment was similar to the treatment offered to Sheri, discussed above, although she did not need an antipsychotic medication, just an SSRI. The staff made an intensive effort to get her out of her room and back to normal activities. In time, she became less obsessed and depressed and had resumed a normal life. In this case, there was no need to change her contacts with the woman she was obsessed with. Observation of their interactions had not revealed reasons for Jennifer to complain about her so much. However, if a problem had been found, addressing this with Jennifer and the other person would have been essential, and, if no resolution could be found, separation of the two may have been necessary.

Approximately three years later, Jennifer began complaining about a different person in her residence. Not surprisingly, Jennifer was under stress again: from a construction project in her home, from staff turnover, and from the loss of a close friend who had left the home. Additionally, Jennifer had been diagnosed and treated for hypothyroidism, which undoubtedly was another source of stress for her.

Jennifer was immediately scheduled for an evaluation. Treatment was started quickly, and within a short time she was back on track. For her family and staff, the message from all this was to look for early signs of an obsession and to seek treatment as soon as possible. Of equal importance, they learned to reduce stress whenever possible and to

predict the likelihood that an obsession would happen based on the presence of unpredictable or uncontrollable stress in her life.

As noted above, obsessions with people may also be "positive." For example, when Christina began a new job, she seemed to develop an obsession with the husband of one of her coworkers. She would flirt with him, send him love notes, talk about him as being her "honey," etc. For Christina, explaining to her that her behavior was problematic would be the first step in treatment. If the object of her obsession was willing to gently discuss with her why her obsession with him was not appropriate, it might also be helpful. Sometimes, however, the only solution is for the person who is the object of an obsession to cease all contact with the person with Down syndrome. Redirection, counseling, and medication may also be necessary.

"The Pace" and Obsessional Slowness

Obsessional slowness is an apparent form of obsessive-compulsive disorder that appears to be more common in people with Down syndrome. At this point, much more needs to be learned about obsessional slowness. Whether it is actually a diagnosis or perhaps part of some other diagnosis has been questioned. However, we have seen a clear obsessional slowing in several adolescents and adults with Down syndrome, and we will therefore describe this slowing with the understanding that there is uncertainty about the diagnosis.

The pace of life in our society always seems to be increasing. The persistent stress of the rapidly moving environment can lead to anxiety, depression, or other psychological problems in some people. People with Down syndrome can also perceive the stress of the fast-paced world. One particularly difficult response to this stress is obsessional slowness, which we have also labeled "The Pace."

For some people with Down syndrome, the world seems to move too fast. High expectations (whether real or just perceived by the person with Down syndrome) may play a role in the development of this problem. They may sense a need to perform at a level or pace that they can't keep up with. They may feel as if they don't have control of their lives.

When they cannot keep up with the pace of the world, some adolescents and adults with Down syndrome (consciously or unconsciously) slow down. We have seen people who eat slowly, walk slowly, and take an inordinate amount of time to do

daily tasks. They seem to slow down or even shut down when they can't live up to the expectations of the normal, fast-paced society. This slowing down may be a direct "benefit" to them—for example, if they move slowly enough, they miss the bus and don't get to their stressful job. For others, there doesn't seem to be anything in particular they are directly avoiding. These people have a more global avoidance of activity. They are not avoiding something specific but have slowed down in all (or nearly all) activities.

For a long time, we could only speculate that some of the individuals with obsessional slowness perceived that the world was going too fast. However, a few individuals who moved very slowly confirmed our suspicions by reporting that they feel that "the world is too fast."

Obsessional slowness often (but not always) begins fairly abruptly. We have found that usually there is not one triggering event. Instead, it seems more likely that a chronic buildup of frustration or desperation causes the problem.

Treatment must start with an acknowledgement that for these individuals, part of the acceptance of their disability (Down syndrome) must include an acceptance by those around them of their need to move at a slower pace. They will probably not move at the pace that society sets. They will probably not be able to function at their previous pace because that rate eventually led to their decline.

While acceptance of a slower pace is a large part of the therapy, there are some additional helpful approaches. Giving an allotted time for some activities and then moving on or discontinuing the activity at the end of the time can be helpful. For example, some people who eat very slowly will suddenly start eating more rapidly as they see the clock reach the final minutes of the allotted time for eating. Occasionally the person may become aggressive or very angry if you take away his food, turn off his shower, etc.

Telling the person to hurry up is usually not effective and instead may contribute to agitation. We don't recommend physically forcing the person to hurry up. However, it might be beneficial to help him do tasks in some situations. For example, if he is in danger of losing his job because of tardiness, you might consider helping him get his shoes on so he can get to work on time. However, it is also important to assess whether the job is appropriate for the person or is a cause of stress that could be contributing to the problem with obsessional slowness.

Counseling may provide some limited benefit in giving the person an opportunity to share his concerns. A change of environment—such as having the

person switch to a job where the pace is slower, or to a residence where the other people are more sedentary—may also be helpful.

We have not had tremendous success in treating obsessional slowness with medications. In some patients, however, we have seen some improvement with the use of medications for obsessive-compulsive disorder (see the section on SSRIs above).

A common response to the rapid pace of our society is to race to keep up. However, occasionally this can be overwhelming to the point where people with Down syndrome cannot or will not keep up and may actually move very slowly as a defense against this stress. “The Pace” at which they function can be described as obsessional slowness and requires a degree of acceptance as well as some intervention.

Some individuals who move slowly have been diagnosed with catatonia. This is further discussed in the chapter on regression (chapter 27). It is not clear if obsessional slowness and catatonia are variants of the same problem or two separate problems. We more frequently see that the person with obsessional slowness can actually move fast in certain situations. For example, one man with obsessional slowness who typically moved very slowly and spoke softly suddenly and rapidly jumped out of the chair while watching a sporting event on TV and yelled very loudly about his displeasure with a play on the field. This sort of occurrence is less likely with regression. Those with regression typically don’t have dramatic, episodic differences in behavior. Furthermore, individuals with obsessional slowness typically seem to retain the skills they have previously learned (although that is not always clear) but just use them at a slow (or incredibly slow) pace. As noted in the chapter on regression, our understanding of these conditions is incomplete and evolving.

Return to Table of Contents

Chapter 20

Psychotic Disorders

Elisa was twenty-four when she was brought for our assessment. Over the previous few months she had developed an intense "relationship" with a Hollywood actor that included a great deal of "talking out loud to him." She believed she was married to him, was eager to move to California, and was having difficulty participating in work, family activities, and other social activities. Elisa herself was able to provide little history or insight. Although self-talk and imaginary friends are common among people with Down syndrome, in Elisa's case, the level of dysfunction related to these behaviors was significantly impairing her life. The history, physical exam, and lab testing did not reveal contributing causes.

After weighing all the factors, including the potential for side effects, Elisa's family elected to proceed with treatment with aripiprazole (Abilify) for a diagnosis of a psychosis. The initial low dose resulted in only marginal improvement, but Elisa improved markedly with an increase to a low-moderate dose. She still had a fascination with the Hollywood actor, but she was able to participate in daily life, her self-talk became much more manageable, and her enjoyment of life seemed to significantly improve.

Many more adults with Down syndrome are initially suspected of having a psychotic disorder than actually turn out to have one. This is especially likely to occur when people with Down syndrome are seen by mental health professionals who have little experience with Down syndrome. In reality, adults with Down syndrome do not commonly have psychoses.

A psychosis is a psychiatric disorder in which the individual experiences delusions or hallucinations, and these symptoms interfere with the ability to function in daily life. In the general population, types of psychoses seen include schizophrenia,

schizoaffective disorder, brief psychotic disorder, substance-induced psychotic disorder, psychotic disorder secondary to a medical condition, and others. These specific diagnoses seem to be less common in people with Down syndrome. However, occasionally adults with Down syndrome have psychotic symptoms, as discussed below.

What Is Psychosis?

Psychosis is a disorder that includes at least some of these symptoms:

- delusions (a false or irrational belief);
- hallucinations (seeing, hearing, or sensing something that is not present);
- withdrawal from reality (for example, an intense preoccupation with hallucinations in place of reality);
- paranoia (an irrational fear or distrust; for example, being afraid that someone is out to get you when it is not true);
- flat affect (lack of emotional responsiveness);
- altered thought processes and disorganized thinking and speech (for example, stringing thoughts together that have no connection to each other).

The diagnosis of the different types of psychotic disorders is based on the symptoms and the duration. It is also important to evaluate for other causes of psychoses such as a substance (a drug or medication) or a medical condition (such as sleep apnea).

Diagnosis of Psychotic Disorders

Psychoses can be quite difficult to diagnose in people with Down syndrome. In order to determine that a thought process is abnormal or psychotic, it is necessary to understand what the normal thought process of the person was prior to the change. This can be a challenge in adults with Down syndrome, particularly those who have limited verbal skills. In addition, there are a number of issues to consider in people with Down syndrome that we have previously discussed. These include the occurrence of self-talk, imaginary friends, and other behaviors that have been described as "psychotoform" behavior (Sovner & Hurley, 1993). When taken out of context, these behaviors can be misinterpreted as psychotic. However, when taken in the context of a person with an intellectual disability, they are usually not psychotic but consistent

with the function of a person at that developmental level, as illustrated in this example:

Leonard, age twenty-nine, was brought to us because of concerns that he had developed a psychotic disorder. He had a long history of self-talk, but it had recently increased. He was also isolating himself more in his room. At work, he was refusing to go out to the parking lot to gather the grocery carts. A psychologist told his family that Leonard was psychotic, and they came to us for a second opinion.

We encouraged Leonard's mother to listen outside his door to hear what he was saying to himself. He kept repeating a phrase about no one helping him get up. Discussions with the family and with a counselor eventually led to a better understanding of the problem. They learned that Leonard had been struck by a car while working in the grocery store parking lot. He had been knocked down, the driver had not stopped, and no one had helped him up. He had not told his boss because after he had been struck by the car, it had taken him so long to get back into the store with the carts that the boss had become angry with Leonard.

Leonard's family discussed the situation with him and then arranged a meeting with his manager. The manager agreed to let Leonard just bag groceries without having to gather carts in the parking lot. In addition, Leonard underwent regular counseling. His self-talk returned to the previous level, he became more involved in activities again, and he did well at work. Leonard was not psychotic. However, he continues to have a fear of parking lots and prefers to walk close to someone when walking through one. He also still periodically expresses concern that no one helped him up. When he mentions this concern, he is reassured that people care for him and that the person who struck him did not act appropriately.

Further complicating the diagnosis of psychosis in people with Down syndrome, the emotional and psychological response to an underlying physical problem may sometimes result in symptoms that appear psychotic. One particular physical health problem to consider when assessing a person with psychotic symptoms is sleep apnea. Both the chronic sleep deprivation and the oxygen deprivation can cause significant psychological symptoms, including psychotic symptoms.

To diagnose an adult with Down syndrome with a psychotic disorder, it is generally necessary to carefully observe her in a variety of settings. Observation in the physician's and mental health professional's office is helpful, but direct observation or talking to others about their observations is often required to make the diagnosis. Sometimes the diagnosis is made by excluding other diagnoses that don't fit the symptoms and "reading into" the behavior that the person is displaying. An example of the value of observations in reaching a diagnosis follows:

Rodney, age forty-seven, had a long history of self-talk and interaction with imaginary friends. At first, his family was able to redirect him when necessary to help him function in his daily activities, he participated in his work program, and he was active with his family. However, his family noticed a change in his behavior over time. His self-talk became more intense, it was more difficult to redirect him when he talked to himself, and he was spending more and more time interacting with his imaginary friends to the exclusion of his family, coworkers, and friends. The diagnosis of psychosis became clear when he began reporting seeing monkeys swinging through his home. Rodney responded well to risperidone (Risperdal), an antipsychotic medication, as well as to his family's efforts to redirect him to his usual daily activities.

Treatment of Psychotic Disorders

Treatment of psychoses includes the following:

1. emotional support for the person and the family or care providers,

2. attention to medical issues that may be contributing to the psychotic symptoms or that may have been a result of the person being less able to care for herself, and
3. medications.

Chapter 16 discusses the importance of counseling and evaluation of, and intervention in, the environment. These are necessary to assess whether the situation is clearly one of psychosis and to give clues as to how best to intervene.

Medications seem to always be an essential piece of treating psychoses in people with Down syndrome. Doctors have two types of antipsychotic medications in their arsenals: the older antipsychotic medications, and newer, atypical antipsychotic medications.

The older antipsychotic medications include drugs such as haloperidol, thioridazine, and thiothixene. (The previous trade names for these were Haldol, Mellaril, and Navane, respectively, but these are now only available in generic form.) Although these medications work well, our experience has been that people with Down syndrome have more side effects from these medications. The anticholinergic side effects are particularly common and problematic. These include constipation, urinary retention, difficulty urinating, dizziness, and others.

The newer, atypical antipsychotic medications are often a better choice. (These are often called second-generation antipsychotics, as compared to the older, typical antipsychotic medications.) Some of the atypical antipsychotics are risperdone (Risperdal), olanzapine (Zyprexa), queitapine (Seroquel), ziprasidone (Geodon), lurasidone (Latuda), brexpiprazole (Rexulti), paliperidone (Invega), and aripiprazole (Abilify). Clozapine (Clozaril) also belongs to this class, but we don't use it because of the possible effect on the white blood cell count, although some psychiatrists use it very successfully. These medications work well and seem to have fewer side effects than the older medications. In addition, these newer medications can help with the depressed mood that often occurs with psychoses.

We have found that olanzapine (Zyprexa) generally causes greater sedation. This can be used as an advantage when sleep disturbance is part of the problem. It also tends to cause the most weight gain, which can be an advantage if poor appetite is a symptom but is often a troubling side effect.

Elevated blood sugar is a possible side effect of the atypical antipsychotics, and this can lead to diabetes mellitus. We have seen elevated blood sugar to a greater degree with risperdone (Risperdal) and olanzapine (Zyprexa) than with

queitapine (Seroquel), ziprasidone (Geodon), or aripiprazole (Abilify). Initially, lurasidone (Latuda) was touted as causing less elevated blood sugar, but our experience has been mixed. Some individuals have developed elevated blood sugars on lurasidone. We use blood draws to regularly monitor blood sugar in our patients who are taking these medications. In addition, monitoring for the development of cataracts is recommended when using quetiapine (Seroquel). Increased cholesterol can also be a side effect of these medications.

When using any antipsychotic, it is important to monitor for signs of tardive dyskinesia (TD). TD is a neurological syndrome characterized by involuntary and abnormal movements. It most commonly affects the muscles of the mouth or face but may affect any muscle. It most often occurs after long-term use of an antipsychotic medication, particularly when used in high doses. TD seems to be less common with the atypical antipsychotic medications but can occur with either the newer or older drugs. Discontinuing the medication is often enough to eliminate the symptoms, but sometimes they continue indefinitely even after the medication is stopped.

In older patients with dementia (Alzheimer's disease), it is recommended not to use these medications due to an increase in stroke risk.

Conclusion

Psychotic disorders seem to be relatively less common in people with Down syndrome compared to people without Down syndrome. However, some of that difference may be falsely low. Due to difficulties with communication skills, it is not always possible to understand what the individual is thinking, making it impossible to understand whether her thought processes are consistent with a psychosis or not. Psychotic disorders are also less common than other mental health problems in people with Down syndrome. Careful assessment is necessary to delineate true psychosis from "psychotoform" features or characteristics. Fortunately, psychosis in adults with Down syndrome does usually respond to counseling, environmental intervention, and medications.

Return to Table of Contents

Chapter 21

Eating Refusal

James, a twenty-five-year-old man with Down syndrome, was eating very little when he was brought to us for evaluation. Six months prior, his family had noted that his appetite seemed to be decreasing and he had lost a few pounds. While James has good verbal skills, he has never been able to communicate symptoms of discomfort or ill health, so his family took him to his primary physician. The history, physical, and lab evaluations did not reveal a cause for the changes in his appetite. Dietary supplements were recommended. However, his appetite continued to be limited, and eventually he lost 22 pounds, down from 147. Although James was not underweight, the unexplained loss of appetite and weight loss remained a concern.

When we evaluated James, he showed signs of dehydration and poor nutrition, and we recommended hospitalization. In the hospital, tests confirmed he was dehydrated and had mild iron deficiency anemia. We consulted a gastroenterologist, who performed an esophagogastroduodenoscopy (EGD or "upper scope") and found a stomach ulcer. The ulcer was treated with omeprazole, and James began iron therapy. James's appetite improved somewhat but not back to his baseline. It was clear that James had developed an obsessional fear of eating and compulsive avoidance of food, apparently in response to the discomfort he had experienced while the ulcer was active. We started James on paroxetine (Paxil), a selective serotonin reuptake inhibitor that has FDA approval for obsessive-compulsive disorder. With the medication and gentle encouragement, James's obsessive thinking and compulsive behavior decreased, and he was able to resume his normal eating.

Since the first edition of this book was published, the diagnosis Avoidant/Restrictive Food Intake Disorder (ARFID) was added to the DSM-5. (It replaced and extended the DSM-IV diagnosis of Feeding Disorder of Infancy or Early Childhood.) ARFID is marked by a feeding or eating disturbance that causes weight loss, nutritional deficiency, dependence on supplementary feeding (oral or by tube or intravenously), and interference in the person's psychosocial functioning. Physical illnesses can contribute to the change in eating seen in ARFID, but the decreased eating is beyond what can be directly accounted for by the physical symptoms of the medical condition. Phobias, anxiety, depression, obsessive-compulsive disorder, and other mental illnesses can co-occur with ARFID or may instead be the only mental health diagnosis (instead of ARFID). One mental illness, in our experience, that is rarely the cause of ARFID in people with Down syndrome is anorexia nervosa.

It can be challenging to make a definite diagnosis of ARFID in adolescents or adults with Down syndrome, particularly when the individual is not able to provide a medical history of symptoms and discuss his feelings. It is clear, however, that some people with Down syndrome do develop the symptom of "eating refusal." Since eating refusal can be a symptom of so many mental health and physical problems in people with Down syndrome, we have chosen to devote a separate chapter to the topic. ARFID is the likely diagnosis for some or perhaps many of these individuals. However, since ARFID can be difficult to differentiate and is an example of the interaction between mental and physical health problems, we will refer to it in this chapter as the symptom of eating refusal.

What Is Eating Refusal?

By *eating refusal*, we mean a significant change in an individual's eating or drinking pattern that produces significant weight loss or other health risks. Any of the following would be labeled eating refusal:

- stopping eating and drinking altogether,
- refusing to eat all but a small, select group of foods,
- refusing to eat certain textures (for example, the person will drink but not eat solid foods), or
- eating small, inadequate amounts of food.

We have seen a number of people with Down syndrome who have declined to eat and had significant weight loss. The eating refusal generally seemed to be a symptom of a physical problem or to have started as a complication of a physical health problem. Often the person stopped eating in response to pain related to the health problem. For example, James initially began eating less at least partly due to the ulcer. Like James, some individuals continue to refuse food even after their physical pain has gone away. In these cases, eating refusal often appears to have become a learned behavior, or the person has developed a mental illness that often consists of an obsession about the physical symptom of pain and a compulsion to avoid eating and the resulting pain. We have also evaluated people with depressive or anxiety symptoms that include eating refusal that were apparently triggered by the original physical problem.

Decreased appetite can also be a direct symptom of mental illness (other than ARFID). Loss of appetite is a common symptom of depression. When eating refusal accompanies the onset of symptoms of a mental health disorder such as depression, we consider the eating problem to be part of the symptoms of that disorder (or can be a dual diagnosis of depression and ARFID). There may not be a contributing physical problem.

However, in most of the individuals with Down syndrome we have seen with significant and problematic eating avoidance, a physical health issue contributed to the onset of symptoms. Frequently, the symptoms of a mental health disorder come later. In these individuals, the mental health condition seems to be a secondary problem that occurs in response to the stress of the initial medical condition and the eating refusal, as in the example of Nathan, below:

Nathan, age thirty-eight, had had a mild swallowing problem for many years. However, as long as he ate slowly and his food was well cut, he did well. Then, at a local carnival, he choked on a hot dog. He developed a fear of eating and would only drink fluids. He became distraught over his fear as well as others' attempts to try to get him to eat his normal diet. Over time, he became depressed. Treatment had to focus not only on the original medical issue (the swallowing problem) but also on his phobia and depression and on counseling his family and others as to how to address his swallowing and eating issues.

Diagnosis of Eating Refusal

When we see an adolescent or adult with Down syndrome who is eating less or refusing to eat, our first step is to assess for underlying physical health problems. We do this even though the only symptom of a disorder may be a refusal to eat. If we were to assume that the eating refusal was completely behavioral in origin, we could miss a significant underlying health problem. If there was an underlying health problem and we failed to treat it, the result would generally be incomplete resolution of the eating problem and continuing, unnecessary discomfort for the person.

Below are some of the medical conditions we consider:

- gastroesophageal reflux and esophagitis (with and without esophageal stricture),
- peptic ulcer disease,
- dental problems,
- swallowing dysfunction,
- celiac disease,
- hypothyroidism,
- multiple causes of nausea (e.g., kidney disease, diabetes, calcium abnormalities, pancreatitis, etc.),
- intracranial abnormalities (such as brain tumors or other causes of increased pressure within the brain),
- medication side effects,
- other possibilities that the history and physical might indicate, such as sore throat, masses in the mouth or throat, etc.

We often find that stomach ulcers, inflammation in the esophagus, or other conditions in the gastrointestinal tract are the root cause of the problem. As part of the workup for these problems, it is often necessary for the person to undergo an upper endoscopy (esophagogastroduodenoscopy or EGD) to evaluate the gastrointestinal tract. We make this decision only after thorough discussion with the patient and family. An upper endoscopy involves placing an endoscope (a tube that can be looked through) into the mouth and advancing it through the esophagus down to the stomach and the first part of the small intestine. This is generally done under

sedation, but many of our patients with Down syndrome need a greater degree of sedation and often require general anesthesia.

Whether further testing is needed depends on the findings of the history and physical. These include tests to assess for the problems noted above and others as guided by the findings. Careful observation of the individual's behavior and activity can sometimes result in finding clues suggesting discomfort or other symptoms that ultimately lead to the diagnosis.

Treatment of Eating Refusal

When the person has an underlying medical problem, treating the problem sometimes results in a return to normal eating. Other patients, however, like James, continue to refuse to eat even after their physical problem is successfully treated. In these instances, we have found or suspected that the symptoms began due to a physical condition, but the medical condition served as a stressor and contributed to the development of obsessive thinking and compulsive behavior (i.e., the refusal to eat), depression, ARFID, or other conditions. It is truly amazing how some people with Down syndrome in this situation seem to be able to ignore their body's hunger cues and continue the compulsive eating refusal.

A common question is whether this problem is comparable to anorexia nervosa. Although there are some similarities, there are also differences between anorexia nervosa in people without Down syndrome and the eating refusal we see in people with Down syndrome. While adolescents and adults with Down syndrome with this problem do avoid eating, we do not generally hear them express concerns that they are overweight, have body image concerns, or want to continue to lose weight—as is the case with people who have anorexia nervosa. Instead, eating refusal appears most often as a response to a medical condition, followed by the development of the psychological components as outlined above.

While we don't generally see anorexia nervosa in adolescents or adults with Down syndrome, we have seen some people who had atypical thinking and approaches to their dietary intake:

Jessica was placed on a strict low-fat diet because of elevated cholesterol. She was regularly reminded to avoid food that was higher in fat. She had always been one who developed "grooves" (see chapter 10) in

her approach to her daily routine, and she did this with her diet as well. Unfortunately, she developed such a groove that she became extremely selective in what she would eat. She became fixated on the dietary advice she had been given and followed it beyond reason. Her weight went down to her ideal body weight (120 pounds) and kept right on going, down to 101 pounds. Fortunately, we were able to redirect Jessica and her family to support a healthy diet without going overboard. Jessica was reassured that some dietary fat was not only acceptable but good. Over time, her diet became more reasonable, and her weight returned to a healthy level.

If someone continues to refuse to eat once a medical problem is treated (or if we fail to find a physical problem underlying the eating refusal), we often recommend treatment with medications for obsessive-compulsive disorder or depression. These treatments can include the following:

- counseling for the person with Down syndrome and for his family or caregivers,
- swallowing therapy to encourage the individual to try eating again,
- support until the person is eating adequately, and/or
- medications.

Support

Supportive therapy, reassurance, and redirection toward eating can all be helpful. When the physical problem has been treated, helping the person understand that the pain has resolved and eating will be pain-free can require ongoing support, "hand-holding," and encouragement. Often this must be done where the person lives (and eats). Therefore, family and caregivers need to be part of the team that encourages and supports the individual. Other people who interact with him at school, work, and recreational settings will also have to be aware of the situation and be part of the solution. In addition, it can be helpful for family and staff to consult a counselor or psychologist for guidance in supporting the person with Down syndrome.

The first goal is to make sure the person is eating and drinking enough to get the calories, vitamins, minerals, fluids, and so forth that he needs. As long as that is occurring, there is less pressure to push him to eat a "normal" diet. Therefore, if the

person refuses to eat solid foods, he may drink multiple cans of a supplemental drink (such as Ensure or Boost) and eat small amounts of food that add up to adequate calories, fluids, and other essential nutrients. Pressuring him to eat a "normal" diet may actually create anxiety and make it less likely he will consume the adequate amount he is presently. Support and gentle encouragement (and acceptance that this may be a slow process) are much more likely to be successful. This gives the adult with Down syndrome and the family time to work with a counselor, swallowing therapist, and/or medical practitioner to try additional treatments as outlined below.

Counseling

Caregivers and families usually need coaching and counseling as well, since trying to encourage someone to eat can be very frustrating. The family and caregivers may need support to stay away from confrontational situations, keep their cool, avoid taking the eating refusal personally, and reduce their frustrations. Confrontation often leads to the person with Down syndrome digging in his heels and can slow the healing process. A social worker, psychologist, or family therapist might provide beneficial counseling.

Swallowing Therapy

Swallowing therapy with a speech therapist can also be helpful. This therapy may sometimes be started as soon as the eating problem is recognized and while the underlying cause is being evaluated. Other times it is acceptable to wait until the underlying medical cause is found, because treatment of that problem may be all the treatment that is needed.

Swallowing therapy consists of reteaching the person to swallow and eat by progressively introducing foods. If some swallowing skills have been lost because the person has not swallowed appropriately in a long time, the therapy can help him regain these skills. Some adults with Down syndrome find swallowing therapy to be frightening or anxiety provoking (just as eating may be). If so, antianxiety medications may be necessary before proceeding with the therapy.

Medications

Medications are often part of the treatment for eating refusal. When depression, anxiety, or obsessive-compulsive disorder complicates the problem, antidepressant medications can be helpful. We have found paroxetine (Paxil) to be of particular benefit, because, like other antidepressants (SSRIs), it treats the depression, anxiety, or obsessive-compulsive symptoms. In addition, many people with Down syndrome develop an increased appetite and weight gain with paroxetine. Mirtazapine (Remeron) is another antidepressant that can cause significant weight gain. Since it can also cause sedation, it is best to take it in the evening. It does not have an FDA indication for obsessive-compulsive disorder or anxiety. Other antidepressants discussed in the chapters on depression, anxiety, and obsessive-compulsive disorder can also be helpful.

Sometimes a thorough evaluation of the situation may lead to a diagnosis of a psychotic disorder (see chapter 20). We have found that all antipsychotics cause weight gain in some individuals with Down syndrome. Olanzapine (Zyprexa) seems to be particularly associated with weight gain and can therefore be a good choice for those with eating refusal. More information about medications for psychotic disorders can be found in chapter 20.

Michael, age forty-three, was brought for evaluation because his sister was concerned that he was losing weight. His appetite had significantly declined; he had lost about 40 pounds and was down to 110 pounds. When we saw him, he would not eat solids, would only drink liquids, and occasionally would vomit. He had limited speech capabilities but could nod yes or no. He nodded that he didn't have any pain. His hemoglobin (red blood cells) was a little low, indicating he was slightly anemic, which could have been due to inadequate intake of nutrients or because of blood loss.

Michael was hospitalized to treat the dehydration that had occurred because of decreased fluid intake. While in the hospital, he underwent an endoscopy (EGD) and was found to have inflammation in his esophagus (esophagitis) and narrowing of the esophagus. His esophagus was dilated during the EGD. He began treatment with omeprazole (Prilosec), to reduce

the acid in his stomach, which was refluxing into his esophagus, causing inflammation and ultimately scarring and narrowing of the esophagus. He was also treated with iron for his anemia. In addition, Michael was found to have and was treated for hyperthyroidism (overactive thyroid). Although hyperthyroidism can cause weight loss, it is often accompanied by an increased appetite (which was not the case with Michael).

Treatment for these conditions was only mildly successful in improving Michael's appetite and weight. He was diagnosed with ARFID with depression based on his decreased appetite, mood, lack of interest in his usual activities, and increased sleepiness. He was started on paroxetine, and his mood improved, and, over time, his appetite did as well. As Michael gained weight and became more active, he regained strength and his overall demeanor markedly improved. Over the course of several months, he gained back his forty pounds. The paroxetine was weaned, and he continued to eat well. Treatment to address his reflux continued, as did his medication for hyperthyroidism.

Michael was a classic example of eating refusal. When he first stopped eating, he did not show symptoms of mental illness. These symptoms occurred later.

Paul, age thirty-three, required greater support. When we first saw him, he had lost 35 pounds and was down to 105 pounds. Other than not eating, the only other problem his caregivers reported was a lifelong pattern of compulsive behavior. He tended to do almost everything in a ritualistic, compulsive fashion. His physical exam was unremarkable except for his weight loss, and labs were all normal. Paul underwent an endoscopy, which showed just a little inflammation in his esophagus but no narrowing. Upon further questioning, he did seem to have had possible heartburn-type symptoms around the time he had begun eating less.

Paul was prescribed medications to reduce the acid in his stomach, but there was only minimal improvement in the amount he would eat. Despite counseling and paroxetine for presumed obsessive-compulsive disorder and ARFID, he continued to lose weight. Next, he was fed through a nasogastric tube (a tube through his nose into his stomach). However, he continued to refuse to eat, and eventually a gastrostomy tube (a tube through his abdominal wall into his stomach) was surgically placed to feed him. Supportive nutrition was given through the gastrostomy tube while swallowing therapy, further counseling, and other medications were tried. Paul did get to a point where he would eat limited amounts "for pleasure," but he continues with a gastrostomy tube for nutritional support.

Paul's problem with eating demonstrates a fairly common finding among people with Down syndrome with symptoms of eating refusal. By the time the problem is evaluated, the original medical condition may be resolving. Alternatively, the medical condition may not seem very serious. Paul, for example, had minor inflammation in his esophagus. However, this relatively minor problem seemed to have set off a progressively downhill course that resulted in ARFID with obsessive-compulsive disorder as well as other behavioral changes. While it can't be stated for sure, it would seem that early intervention prior to the compounding of problems would improve the treatment of eating refusal among teens and adults with Down syndrome,

Conclusion

Eating refusal can be a very difficult problem that can pose a significant threat to health. In people with Down syndrome, there can be an interaction between both physical and mental health issues. A thorough evaluation for underlying physical health problems is an essential part of the diagnosis and treatment of the problem. Counseling, environmental evaluation and support, and medications to treat associated psychological diagnoses may all be needed. Nutritional support may also be necessary. In adults with Down syndrome, significant weight loss is more the

exception than the rule, so parents and other caregivers need to be vigilant about bringing changes in weight or appetite to the attention of health care professionals.

Return to Table of Contents

Chapter 22

Challenging Behavior

Derek, age thirty-four, was brought to the emergency department after severely damaging the sink in the bathroom of his group home. For the few days prior to the event, Derek was "not himself," seemed upset, and was less active. Although he had reasonably good speech, he had difficulty verbally expressing his concerns about stressful situations and had a history of infrequent aggressive behavior in response to physical, emotional, and social stress. In the ER, Derek was found to have an ingrown toenail that was causing him discomfort and was apparently the source of his agitation. His ingrown toenail was addressed with appropriate medical treatment, and his behavior returned to normal.

Not all emotional and behavioral problems fit neatly into one of the diagnostic categories in the DSM-5. Yet, even though some behavior problems may not be formally labeled as disorders, they can still be challenging to deal with. In this chapter, we use the term challenging behavior to refer to behavior that is maladaptive or disruptive to an individual's own life or others' lives. Behavior problems may include

- verbal or physical aggression or damage to property;
- oppositional, defiant, or disobedient behavior to authority figures;
- antisocial or criminal behavior, including lying, stealing, or sexually inappropriate behavior, or behavior that is intentionally harmful to others;
- behavior problems associated with impulsiveness, including behavior that is "discharged" without thinking or prior planning, such as blurting out inappropriate comments about others, darting into the street, grabbing food from others' plates, etc.

Challenging behavior may also include behavior that is socially inappropriate or offensive but generally not harmful to others. This includes inappropriate behavior

that is displayed in social or public places (poor hygiene, touching the genital area, expelling gas, nose-picking, etc.).

People with Down syndrome are more likely to have behavior problems than are people without an intellectual disability (Kahn et al., 2002; Patel et al., 2018). However, studies have mixed results as to whether the frequency of behavioral problems are more, less, or the same for people with Down syndrome compared to people who have other causes of an intellectual disability (Ebsensen et al., 2008; Mantry et al., 2008; Dykens et al., 2015).

Reasons for Challenging Behavior

Why do people with Down syndrome have behavior problems, and why do they have more problems than those without an intellectual disability? There are a number of possible reasons. Perhaps most importantly, adaptive and expressive language limitations may make it difficult for someone with Down syndrome to conceptualize and communicate the presence of problems or issues. Although this may be more obvious for people with limited verbal skills, it may also be the case for people who have better verbal skills. This is because adults with better verbal language may still be unable to conceptualize and communicate thoughts or feelings related to certain problems or issues. For example, Derek, the man in the example at the beginning of the chapter, had good verbal skills in much of his life but had trouble sharing his thoughts when he became stressed and upset.

Likewise, a woman with Down syndrome had similar incidents that were very uncharacteristic for her. Her family was at a loss to explain these changes until they learned that she had been recently dumped by her boyfriend and had also lost a close friend who moved to another town.

Any type of behavior problem, including aggressive, oppositional, or inappropriate social behavior, may communicate the presence of a problem. These behaviors may serve both to vent the person's frustration and to obtain attention from others. As a form of nonverbal communication, the message requires an interpretation by others (see chapter 7). We have found that family members and other caregivers are usually able to help us interpret the message. Below are some of the most common causes of challenging behavior in adolescents and adults with Down syndrome.

Underlying Physical Causes

It is particularly important to look for possible physical causes when evaluating a person with Down syndrome for difficulty with controlling behavior or aggressive behavior. Discomfort may lead to a reduced ability to control emotions or an exaggerated or aggressive response to another stimuli or event.

Malcolm, a thirty-seven-year-old man with Down syndrome who had obsessive-compulsive disorder, was being treated with sertraline (Zoloft) and redirection by the staff of his group home. The treatment was generally successful. Periodically, however, he would become aggressive when staff attempted to redirect him. Malcolm had had mastoid (ear) surgery many years prior, and, periodically, the area would fill with debris and require suctioning. He was recurrently found to need this treatment during times of aggressiveness. He appeared to have discomfort associated with the physical problem but communicated this with his behavior, rather than words.

Attention-Deficit Disorders. We also see individuals with behavior problems that are secondary to physiological or neurological conditions. People with these conditions may have more difficulty with controlling their impulses and behavior. Among the most common of these conditions are attention-deficit disorders (AD/HD) (with or without hyperactivity). AD/HD has impulsive behavior as one of the three core symptoms, along with attention problems and distractibility. Because of the impulsive behavior, the person with AD/HD may have more trouble waiting or difficulty keeping unflattering comments about others to herself. Some individuals may also have difficulty controlling their emotions and behaviors, and they may be likely to be aggressive when frustrated or stressed. Correctly identifying the physiological cause of the condition will help to ensure a more successful course of treatment. See below for more information about attention-deficit disorders in adolescents and adults with Down syndrome.

Seizure Disorder. Occasionally we evaluate adults who have aggressive or impulsive behaviors associated with simple or complex partial seizures. While relatively uncommon, these conditions need to be identified in order to provide the

right pharmacological treatment. (See below and chapter 16 for more on the use of anticonvulsant medications.)

Impulse Control Problems. We have also seen some individuals with Down syndrome who have impulsive behavior but show no evidence of an attention deficit, a seizure disorder, or any other physical, emotional, or environmental precipitants. We continue to look for causes, but these people may simply have a greater disposition to impulsive behavior than others. As a result, like people with seizures and attention deficits, these individuals may have more difficulty controlling their impulses, and this may at times include antisocial or aggressive behavior.

Attention-Deficit Disorders

Attention-deficit disorders are neurological disorders that have the following symptoms: problems with attention, impulsive behavior, and distractibility. Some individuals have primarily an inattentive presentation. Others have either a primarily hyperactive/impulsive presentation or combined inattentive and hyperactive/impulsive symptoms. (Note that in the DSM-5, both types of attention-deficit disorders fall under the umbrella category of attention-deficit/hyperactivity disorder, or AD/HD. To aid in making distinctions between the two subtypes, we will use the abbreviations ADD-In for AD/HD without hyperactivity and ADHD for AD/HD with hyperactivity.)

With studies showing prevalence rates at about 11 percent in children without an intellectual disability (Visser et al., 2014), AD/HD is one of the most common neurological condition diagnosed in children. In children with Down syndrome, one study found a prevalence rate of 43.9 percent (Ekstein et al., 2011), although previous studies had found similar prevalence rates to children without Down syndrome (Cohen & Patterson,1998; Myers & Pueschel, 1991). Some clinicians have written that other conditions, including sleep problems, impaired vision and hearing, and thyroid disease, can look like AD/HD in children with Down syndrome (McBrien, 2012) and that AD/HD could therefore be misdiagnosed.

AD/HD often has a disastrous impact on a child's academic and work performance as well as her social and emotional functioning and development. Attention problems and distractibility may make it very difficult for people to focus and follow through in an organized fashion with essential tasks at home, school, or work. Additionally, because of impulsivity, relations with bosses and friends may suffer because the person may have trouble waiting or keeping her thoughtless or

unflattering comments to herself. She may also have difficulty concentrating on conversations, which may make it appear as if she is disinterested in other people. Some individuals with this condition also have difficulty controlling their emotions and behaviors because of the impulsivity and may become aggressive when frustrated or stressed.

Childhood AD/HD has been recognized for many years, but only recently has it been found to affect a significant number of adults. Adults may have the same problems with inattention, impulsivity, disorganization, and distractibility as children, and this may have the same effect on their social, emotional, academic, or occupational functioning as it does in children. Hyperactivity appears to be less common for adults with AD/HD, even in people who had hyperactivity as children. Apparently, people may grow out of the hyperactivity as they age into adulthood.

Although there is no available research on the rates of attention-deficit disorders in teens and adults with Down syndrome, we have evaluated a significant number of individuals who have this condition. This includes many individuals with a history of attention-deficit disorder with hyperactivity who seemed to grow out of the hyperactivity in adulthood. Like adults without an intellectual disability, they often continue to have problems with attention and impulsivity and therefore often benefit from medication to help them better manage these issues. We have also seen some adults with Down syndrome who continue to have hyperactivity, even though ADHD with hyperactivity is less common in adulthood in the general population.

Symptoms of ADHD

What does ADHD (the hyperactive form of AD/HD) look like in adults with Down syndrome? To answer this question, it may be helpful to first describe typical symptoms in children. Although many children have high activity levels, children diagnosed with this disorder are so active that parents often describe them as "bouncing off the walls." Adults with Down syndrome who have ADHD show some of the same overactive behavior that children show. Many have trouble sleeping, talk constantly and distractedly, and most cannot stay still or focus long enough to do sports activities, let alone essential school or work tasks. The level of activity in adults with Down syndrome may not be quite as intense as in children with ADHD (with or without Down syndrome), but in comparison to other adults, their level of activity is quite extreme. Obviously, this type of behavior may be very trying for caregivers of adults as well as children, as this example illustrates:

Marna, twenty-one, was brought for evaluation by her desperate and exhausted parents six months after she graduated from school. Marna was a friendly and good-natured young woman who was driving her parents and her boss crazy with her constant questions, distracted talk, and movement. At work, she could not stay in one place long enough to complete her job tasks. At home, she was only able to get ready for work or do other tasks if her parents were present to corral and prompt her at each step along the way. Marna was at her best when participating in sports, but even when engaged in these activities she had great difficulty staying focused. For example, Marna's coach described her as being "like a balloon with the air let out" when she was put into a game. She would fly around the field with great speed and intensity but was not necessarily focused on the actual events in the game.

Marna's parents reported that she was hyperactive as a child and had been diagnosed with ADHD by a school psychologist and an experienced pediatrician. She was very active throughout her school years, but a stimulant medication, methylphenidate (Concerta), greatly helped to keep her hyperactivity, inattention, and impulsivity in check.

During the time after Marna graduated from school up until her evaluation, her parents saw a marked increase in her ADHD, even though she was still on her medication. Marna's parents believed that the process of leaving school and starting her job may have been too stressful for her. Many of her friends were at the same worksite, but the setting was much noisier and distracting and less help was available from staff than at school.

At her evaluation, Marna showed no apparent health or sensory problems that could have aggravated her symptoms. The health care providers confirmed the diagnosis of ADHD from her history and her current behavior and began a course of treatment. Marna showed a positive response to bupropion (Wellbutrin), an atypical antidepressant

medication that is effective in treating adults without an intellectual disability who have ADD and ADHD. During this and subsequent appointments, Marna participated in counseling to discuss her feelings and expectations for herself. She was demoralized and her self-esteem was shaken, but she was encouraged to see herself in a more positive light, particularly as her hyperactive behavior and her inattention improved with the treatment.

For the first month, Marla continued to have some problems controlling her anger at work and settling down to sleep at night, but this improved with counseling and an increase in medication. During the second and third months, Marna's parents reported that her behavior was much improved at home and there were no negative reports from work. At a six-month follow-up Marna was still doing well at home, but her parents reported receiving notice from her worksite that she could be suspended because of angry outbursts.

Shortly after this meeting, outreach staff from her health care provider arranged to visit her worksite to get a better understanding of the situation. What they found was enlightening. A number of fellow workers were aggravating Marna with taunting and teasing remarks. Apparently, these individuals had seen her overreact to situations before starting her medications and they seemed to want to goad her into making angry outbursts as she had in the past. Because Marna had a history of past outbursts (prior to starting the new medication), she was threatened with suspension from the program. After being informed that other workers were provoking Marna, staff agreed to closely monitor the situation and to block the others from harassing Marna. After several weeks and several incident reports that named the provocateurs, the teasing and goading stopped.

It has now been three years since Marna's initial evaluation, and she continues to be monitored. She has had some minor problems and some medication adjustments, but overall, she continues to do very well.

Is It ADHD or Something Else?

The good news about ADHD is that people get help because of the intensity of the symptoms and the stress and strain on caregivers. The symptoms simply cannot be ignored. Additionally, ADHD is a widely researched and known condition affecting children and adults. Because of this, teachers, pediatricians, and other practitioners are likely to diagnosis this condition when a child or adult with Down syndrome is brought in with hyperactivity. The bad news about the widespread knowledge of ADHD is that hyperactive behavior may be diagnosed as ADHD when in fact the hyperactivity is caused by something else.

In reviewing referrals of people who have been previously diagnosed with ADHD, we have found that there is often an alternative explanation or diagnosis for the observed behavior. Accurate diagnosis may be even more of a problem for people with Down syndrome who have a limited ability to verbally report problems or symptoms. For example, in our experience people with bipolar disorder may be misdiagnosed as having ADHD because manic behavior may look like hyperactive behavior. However, manic behavior is only a part of the symptom picture and viewing it this way may lead to treatments that may actually worsen the problem. For instance, stimulant medications may increase manic behavior or increase the intensity of mood fluctuations. A careful history, which may be more likely to show the mood fluctuations (between mania and depression) that are characteristic of this disorder, can improve the accuracy of the diagnosis.

Similarly, mania may also be misdiagnosed as ADHD. As with bipolar disorder, this may lead to the use of stimulants, which may worsen and intensify the mania. Mania is often a condition that occurs cyclically, and it often ebbs and flows, whereas ADHD is usually more consistent in symptoms and intensity level.

We have also seen people with autism spectrum disorders who were diagnosed with ADHD. Autism and ADHD can coexist in the same individual and both can coexist in a person with Down syndrome. Careful assessment is needed to decipher whether the diagnosis is ADHD, autism, or both. For example, some individuals may have hyperactive-like behavior, particularly when anxious or overstimulated. If the focus is only on the hyperactivity and diagnosing ADHD, overlooking key aspects of the autism disorder such as the lack of relatedness to others and communication impairment may lead to an inaccurate diagnosis. Without the correct diagnosis, individuals may not receive essential behavioral and medical treatment for the autism component.

Anxiety may also be easily misdiagnosed as ADHD. This is particularly the case for people with Down syndrome who cannot verbalize their feelings but express anxiety through agitated and overactive behavior. How do you sort out anxiety from ADHD? We recommend being particularly sensitive to the history and longevity of the symptoms presented. ADHD is present in early childhood and occurs throughout the person's life. The intensity of the symptoms may change with age, but the disorder will still be present in some recognizable form in adulthood. On the other hand, if the person's "hyper" behavior seems to begin during a stressful time, then it is more likely that the behavior is actually anxiety. Additionally, if the person's ADHD occurs in only certain environments, such as a school classroom, then this may simply mean that the environment is stressful. Often, we find that the person is either over- or understimulated in the stressful environment.

Finally, and perhaps most importantly, symptoms of ADHD may simply be the person's preferred means of communicating behaviorally the presence of a stressful situation. Again, this is particularly likely for children and adults with Down syndrome who have limitations in verbalizing thoughts or feelings. Therefore, behavior that looks like ADHD may be communicating that there is a physical disorder, a sensory deficit (visual or auditory), a stressful change or loss, or an environmental stressor. As we have emphasized throughout this book, we can only get to the cause or source of a behavior if we, as practitioners and caregivers, become detectives and examine as many areas as possible, (e.g., physical, sensory, environmental, and life stage changes). This may be the only way to determine whether any other reasons or explanations for the person's behavior are possible.

Symptoms of ADD-In

If ADHD is overdiagnosed, ADD-In (inattentive, without hyperactivity) is underdiagnosed.

The good news for caregivers is that children and adults with ADD-In are far less disruptive and difficult to manage than those with ADHD. The bad news is that they be may far less likely to be diagnosed and treated for this condition. Studies have shown that a fairly large number of children and adults in the general population are not identified and treated for this condition because of the more subtle nature of the symptoms (especially when compared to people with hyperactivity) (Ginsberg et al., 2014; Jensen & Cooper, 2002; Murphy & Barkley, 1996). We have found that diagnosis may be even more of a problem for people with Down syndrome than for

other groups. Aside from the difficulty of identifying symptoms, ADD-In may not be considered because it may be too easily attributed to the Down syndrome, even when behaviors are not characteristic of Down syndrome (Reiss et al. 1982).

Children and adults with this condition may float along as if in a fog or dreamlike state. They have great difficulty concentrating on school or work tasks. They may have trouble in social situations because they have trouble listening to others or reading social cues. The distracted, dreamlike state may be even more of a problem for children with Down syndrome because they tend to have excellent visual memories, which they may draw on to "space out" (see chapter 6 for more on this).

Even if caregivers or teachers do not see the problem, these children may be teased by peers as being "space cadets" or "dreamers," but this is no laughing matter. ADD-In may have a profound negative effect on the individual's school, work, or social relations, and this in turn may have a disastrous impact on the person's self-esteem.

How, then, do we diagnose and treat ADD-In in people with Down syndrome, given the nature of the symptoms? We need to be very honest here with you, the reader. We have been treating adolescents and adults with Down syndrome for some time, and yet we find this a challenging diagnosis that has the potential to be overlooked in people with Down syndrome. Still, we have found that a number of clues may help us, as well as caregivers and parents, identify ADD-In in this group. These clues relate to different symptom presentations in people with Down syndrome who have ADD-In compared to people with the disorder in the general population. There are also important differences between people with Down syndrome who do and do not have ADD-In that may be instructive.

First, regarding the presentation of symptoms, a key characteristic of people with ADD-In in the general population is that they are often chaotic and disorganized. They often have great difficulty setting up and following consistent routines, making it difficult to do daily tasks reliably and efficiently. The resulting lack of a predictable order is often very frustrating for themselves and for family members.

We have seen a similar pattern for some people with Down syndrome who have ADHD (AD/HD with hyperactivity), but not for people who have ADD-In (without hyperactivity). These individuals have a sense of order despite having attention-deficit symptoms. They are often able to reliably complete daily living tasks, home chores, and work tasks, as long as these activities are part of their regular routine. Their routines and grooves then seem to carry them along despite their attention problems.

The key difficulty these individuals often seem to have is in dealing with free or unstructured time that is outside of their routine rather than an activity that is part of their routine. Although their routines usually carry them along, they have problems organizing activities when not doing their routines. This brings us to an important difference between people with Down syndrome who do and do not have ADD-In. In a nutshell, people with Down syndrome and ADD-In have great difficulty entertaining themselves. This is in contrast to the majority of people with Down syndrome, who are usually very good at entertaining and occupying themselves during free time.

Over thousands of interviews, we have repeatedly heard about adolescents and adults who have special activities they enjoy doing in their free time. Examples of these activities include drawing, copying words or letters, needlepoint, watching TV or movies, looking at family photographs, or even cleaning one's room. In fact, most people with Down syndrome are so good at entertaining themselves that parents and caregivers often complain that they may spend too much time doing these activities. Therefore, when someone cannot do this, it should be a red flag for caregivers and practitioners, even if the person is otherwise able to follow daily routine, as illustrated in this story:

Alida, age twenty-three, was initially brought for an evaluation because of several aggressive incidents that occurred at work. Her parents and case manager were surprised and concerned because Alida had been a model employee for the two years she had been at her job. Her boss had been very pleased with her because she loved to do some of the most challenging jobs and her production rate was very high. Her parents and case manager also noted that she was very friendly and personable and was not prone to acts of aggression.

We learned from the case manager that there had recently been an extended period of no work. This was the first time this had happened in the two years that Alida had worked at her job. She seemed to become more and more restless and bothersome to others during this period. She did not want to participate in some of the downtime activities, including

doing arts and crafts or watching movies, which her case manager admitted were not very stimulating.

Alida's parents described a similar problem at home. Alida seemed to be fine when she could do things that were part of her regular routine. She enjoyed doing her daily chores, such as dusting the house and cleaning her room. However, when she had free time, she could not seem to settle down to do anything to entertain herself. Although her parents tried to give her things to do, such as watching a movie, drawing, or doing word search puzzles, she simply could not do these things for any length of time. Her parents stated that she had always had some problems with entertaining herself in the past, but she had three very active brothers who seemed to supply her with an endless number of interesting activities. Although her brothers complained at times that Alida was a "pain," they loved her and were very tolerant of her participation in their activities. Unfortunately, in the last year, two of her brothers had moved out of the family home, and the other was rarely around.

Aida's parents tried to keep her busy outside the house participating in social and recreation activities. She did fine in more active sports or recreation activities but had a difficult time in more unstructured social gatherings. Unfortunately, her participation in outside activities did not seem to help her manage her free time at home.

Finally, Aida's school history was consistent with a diagnosis of ADD-In. Her family noted that she had had problems in grade school with staying on task. Her teachers had described her as having a short attention span. She also had some problems playing with others during recess activities, but they attributed this and her attention span to her Down syndrome. Alida had seemed to do better in high school, particularly in the last three years, when she participated in an excellent vocational training problem involving experiences in a wide variety of jobs. She was very busy and challenged in this program, and this seemed to be the formula she needed. Indeed, she had found her job very challenging and

satisfying until the sudden extended period of downtime that had preceded her aggressive outbursts and our initial assessment.

In order to verify ADD-In, we referred Alida to a psychologist who had expertise in evaluating people with Down syndrome with attention-deficit problems. She confirmed our suspicion that Alida had ADD-In. After a trial of several different stimulant medications, Alida showed a positive response to methylphenidate (Ritalin XR). On the medication, she was able to settle herself and developed a healthy interest in a host of activities including needlepoint and word search puzzles. Equally important, she was far more able to tolerate downtime at her worksite. We did, however, recommend to the workshop that they develop a more interesting program of free-time activities. Subsequently, a bank of computers was set up, which Alida loved to use during her free time.

Medications for Attention-Deficit Disorders

Medications are an important part of the treatment for ADHD and ADD-In. The medications can help improve attention, reduce impulsivity, and reduce hyperactivity. The medications fall into two general categories: stimulant and nonstimulant.

Stimulant medications stimulate the central nervous system. Interestingly, this reduces the symptoms, including hyperactivity. The approved medications include methylphenidate (Concerta, Metadate, and Ritalin; also available in a patch, Daytrana), dextroamphetamine (Dexedrine), dexmethylphenidate (Focalin), and amphetamine/dextroamphetamine (Adderall). Side effects of stimulant medications in adults include nervousness, difficulty sleeping, motor tics, palpitations, loss of appetite, and others. Finding the right medication may take some time. Even if someone doesn't respond well to one stimulant, she may respond to one of the other medications.

The other choice is a nonstimulant medication. FDA-approved choices include atomoxetine (Strattera) for children and adults and guanfacine (Intuniv) for children. Atomoxetine inhibits the reuptake of the brain chemical norepinephrine, which is believed to play a role in regulating attention. It may cause side effects including

sleep problems, fatigue, increased sweating, fatigue, palpitations, and others. Guanfacine stimulates alpha-2 adrenergic receptors, and side effects include low blood pressure, low heart rate, sleepiness, and others. Some people benefit from using a combination of a stimulant medication and guanfacine and/or atomoxetine. Bupropion (Wellbutrin) is an antidepressant that has also been used for ADHD and ADD-In. Clonidine (Catapres) is in the same class of medications as guanfacine and has been used for ADHD, but it does not have FDA approval.

Underlying Mental Health Disorders or Stress

A behavior problem may signal the presence of a mental health problem. For example, we have seen many people who become aggressive if prevented from doing their obsessive-compulsive behaviors and rituals. The presence of aggressive behavior often signals a marked increase in the severity of obsessive-compulsive symptoms. Caregivers who may be reluctant to seek treatment for obsessive-compulsive rituals may be more likely to seek help when symptoms are accompanied by physical aggression.

We have also seen similar patterns of behavior for some people with symptoms of depression (see chapter 17). This is particularly the case for individuals who withdraw and isolate themselves within their bedroom or another private space. Although these individuals are not characteristically aggressive, they may display aggressive behavior when caregivers try, out of desperation, to get them to leave their rooms to resume normal social or work activities.

Challenging behaviors may also communicate the presence of more extreme environmental stress, such as due to intolerable living or work situations. One of the most common causes of environmental stress is conflicts or tensions with or between others.

Other causes of stress that may result in a behavior change include noxious or overwhelming sensory stimuli in the home, on the job, or in the community. Loud cavernous worksites may be particularly to blame for this kind of stress, but other types of sensory input may be involved as well.

More Serious Behavior Problems

The Diagnostic and Statistical Manual of Mental Disorders (DSM-5) defines three major behavior disorders for the general population. Two of these, conduct

disorder and antisocial personality disorder, are very similar except that the latter is diagnosed in adults and the former is usually diagnosed in children. The third is oppositional defiant disorder. We will define these disorders and discuss whether these behavior problems may be diagnosed for people with Down syndrome.

Conduct Disorder/Antisocial Personality Disorder

The diagnosis of conduct disorder is usually reserved for children under eighteen. This disorder is defined as a repetitive and persistent pattern of behavior in which the basic rights of others or major age-appropriate societal norms or rules are violated. The person exhibits aggression, criminal behavior, and wanton disregard for the feelings and well-being of others. The disorder may range from mild forms of antisocial behavior to more severe criminal behavior. The DSM-5 also defines antisocial personality disorder as the continuation of a conduct disorder into adulthood. As with conduct disorder, there is a lack of empathy and concern for others, as well as antisocial or criminal behavior. People with this disorder have been called "sociopaths" or "psychopaths" to describe their lack of feeling and remorse for their harmful behavior.

In our experience, it is rare for people with Down syndrome to have conduct or antisocial personality disorders. We have only seen a handful of people who have at least a moderate degree of conduct problems.

Hue had a history of sexually inappropriate behavior throughout his childhood. We first evaluated him as a young adult when he was brought by a concerned older brother and staff from his group home and worksite. His brother and staff described a series of sexually inappropriate or sexually aggressive behaviors that had occurred in the bathroom at his worksite and at his group home. None of these had resulted in criminal prosecution, but Hue's brother believed that it was only a matter of time before this would happen. Hue's brother also revealed that Hue was the victim of sexual abuse by an uncle. This uncle was never prosecuted because the abuse was kept secret by the family. Unfortunately, like some victims of childhood abuse, Hue developed an insatiable desire for sexual

gratification. He also seemed to have little concern for how his sexual behavior affected those he targeted to meet his needs.

We developed a multimodal treatment approach to deal with Hue's difficult problem. It is important to understand that there is no definitive way to treat sexually deviant behavior except though strict monitoring and supervision. First, all his caregivers agreed to develop a highly structured environment for Hue. At no time would he be left alone with others without very close supervision. For example, he had his own bedroom and an alarm was rigged to go off if he left the room in the evening. He was also given a special changing room at the gymnasium where he exercised to keep him away from others in the locker room, especially children. Second, Hue was started on an antidepressant medication to reduce his sexual drive. (For Hue, the antidepressant effect of the medication was less important than the side effect of lowering sexual drive.) Because of the close supervision and the effectiveness of the medication, no further sexual incidents have been reported. Staff and family continue to meet at least every three months or more to ensure a continuation of the plan.

A second example of a person with a conduct disorder can be seen in a seventeen-year-old teen with Down syndrome, Beatrice:

Beatrice had grown up in a household with some family members who had an extensive history of criminal behavior. She had spent much of her early school years in classrooms for students with behavior disorders because of her aggressive and oppositional behavior. She also had some of the same sexually inappropriate behavior as Hue, although not to the same degree. She also had a history of stealing money, food, and valuables from others. Her behavior continues to be difficult to manage in her current high school BD program and at home with her mother.

Beatrice may continue to need a highly structured environment, similar to the program developed for Hue. However, Beatrice is much younger than Hue, and her school and the appropriate state case-management agency are looking for a therapeutic facility for her. There is hope that in the right program she may have a chance to overcome her behavior problems. There is evidence that some children do grow out of conduct or other behavior disorders if they are given appropriate guidance and treatment before they reach adulthood.

Less Severe Sexual Conduct Problems

We have assessed a number of men and women with Down syndrome who have made inappropriate sexual comments or who have touched people inappropriately. These adults have generally responded to parent or staff attempts to stop or redirect their behavior. In a few instances, parents have had trouble stopping sexual behavior due to normal teenager-like resistance by the person with Down syndrome to parental control. In these cases, we have collaborated successfully with parents to support their efforts to control the behavior through increased supervision. When we do encounter a problem of a sexual nature, we have the adult with Down syndrome return for regular scheduled appointments (weekly or every other week) until we are satisfied that the problem is completely resolved or managed.

Behaviors That May Look Like a Conduct Disorder

Stealing

Sometimes adults are brought for evaluation by concerned parents or other caregivers because of incidents of stealing from others. Occasionally, there are concerns that this behavior may lead to other types of antisocial behavior. We generally find that this behavior has more to do with an intellectual or conceptual limitation in the person with Down syndrome than a problem of criminality. We often call this behavior “creative borrowing,” rather than stealing, because the person may simply not understand the concept of stealing. Like many younger children without an intellection disability, some people with Down syndrome have difficulty understanding

that others have personal property, even when they are well aware of their own personal possessions.

For some people with Down syndrome, "stealing" may be related to a compulsion to save or hoard a special item (such as pens, paper, etc.). The person does not necessarily conceptualize this behavior as taking from others, but rather as simply adding to her collection. (See chapter 19 for more on hoarding.)

In evaluating instances of "stealing," we look for the presence or absence of other forms of antisocial behavior to determine the severity of the behavior. Beatrice, discussed above, is a good example of a person with a conduct disorder. She stole from others and had many other serious behaviors such as a lack of sensitivity toward others' feelings, aggressive behavior, etc. For adults who just have instances of stealing, without other antisocial behavior, we work with caregivers to try to reinforce simple behavioral strategies to limit this behavior. For example, the person may earn a reward, such as buying a desired pen from a store, if she does not take that item from others. This allows her to acquire desired items without "borrowing" from others.

Lying

Parents and other caregivers sometimes have similar concerns about lying. This behavior, too, may involve a conceptual limitation. Some people with Down syndrome simply may not understand the concept of lying, just as many younger children do not. Another reason that people with Down syndrome may tell untruths or "lie" may be that they tend to be very sensitive to others. As a result, they may try to protect others from feeling bad by not telling them something that will hurt their feelings. They may also try to please the listener or even to protect themselves from the other's anger. The fact that they may not truly understand what it means to lie may make the "lie" less of a problem for them in certain situations, such as when protecting themselves or others from real or perceived harm

Other times people with Down syndrome are thought to lie when in fact they are not lying. As discussed in chapter 6, many people with DS have exceptional memories but have difficulty understanding the concept of time. As a result, they may speak in the present tense about past events. If listeners are not aware of this lack of time orientation, they may think the person is lying rather than describing a past event. A good example of this was a young man with Down syndrome who complained that he was abused by someone at his worksite. His mother was called to

a meeting to discuss why he was lying about the abuse. Fortunately, she was able to explain that he was actually talking about a past event. However, it is easy to imagine that staff would interpret his statements as lies or false accusations if his mother was not present to explain the confusion. To avoid this type of problem, any comments need to be assessed as possibly having some basis in the past and not just the present.

Additionally, many people with Down syndrome may have difficulty with sorting out fact and fantasy and may discuss imagined events or events viewed on TV or in the movies as if they are factual. What appears to be a lie may actually be the result of a very active and vivid imagination and memory. Special care should be taken to understand the confusion of fact and fantasy when assessing statements made by people with Down syndrome.

Oppositional Defiant Disorder

The DSM-5 describes oppositional defiant disorder (ODD) as a recurrent "pattern of angry/irritable mood, argumentative/defiant behavior, or vindictiveness." Although this diagnosis is only used for children or teens in the general population, it may be applicable to adults with Down syndrome, because they often continue to have caregivers who are responsible for them.

We have seen a small number of people with Down syndrome who exhibit ODD behavior. Sometimes this seems to be something the person is born with (part of the person's temperament). Other times, the environment may play a role in the development of this problem, as in this example:

> *Robin, age thirty-six, was raised by parents who were very strict and controlling of her behavior. After her parents died, she moved to a group home that she shared with three other women. The stated philosophy of the house was choice, independence, and a respect for the rights of others. This seemed to work for the other women but not for Robin. She seemed to experience the house as a place where she could dominate and control others like her parents had controlled her.*
>
> *Over time, Robin became more demanding and less cooperative with staff. When staff tried to control her behavior, she rebelled and began to throw major tantrums, harass other residents, and behave aggressively.*

Eventually, Robin was transferred to a therapeutic group home, which was set up to better manage people with behavioral issues. The house followed a structured behavior plan where people earned rights and freedoms for cooperating with the rules. After several intense months, Robin learned that cooperating with the rules allowed her to have some control over her own situation. Although she continued to struggle, she learned that cooperating with the rules did not mean others controlled her. She also learned that if she respected others' rights, they would respect her rights too. After approximately three and a half years in the structured group home, she had shown enough maturity to move to a less structured house, and she has done very well ever since.

We have seen others like Robin who have problems with accepting caregiver authority. We should say, however, that we do not frequently diagnose them with ODD. As discussed earlier, we have found that the behavior may actually be an attempt to communicate the presence of a problem or an issue that people cannot easily verbalize. Again, this could be anything from physical pain and discomfort to environmental stress. Therefore, whenever this type of behavior occurs, we recommend that the caregivers and professionals involved try to identify any message communicated through the person's behavior.

In addition, sometimes what caregivers call "oppositional behavior" is actually a message about inappropriate constraints being imposed on the person with Down syndrome. For example, we have had referrals from families who have difficulty with a son or daughter who is legitimately struggling for their own independence. In these situations, it is very important to diplomatically but emphatically support the person's legitimate need for independence. A good example of this type of situation may be seen in Andre's story in the family counseling section in chapter 16.

Finally, another reason the diagnosis of ODD is not used as often as one might think is that oppositional behavior is often one symptom of a much larger problem or condition. For example, many people with bipolar disorder, a dual diagnosis of autism and Down syndrome, or even a conduct disorder, may express some oppositional and defiant behavior toward caregivers as part of the condition, along with other behaviors and symptoms. The treatment for these conditions may be very different

than for just ODD. For example, multimodal treatment approaches are often needed for bipolar disorder; autism may require very different behavioral strategies; and a more intensive behavior program may be required for someone with a conduct disorder, such as discussed for Hue above.

Misinterpreting Disorders as Behavior Problems

A number of disorders may be misinterpreted as behavior problems by professionals and caregivers. Most notably these may include obsessive- compulsive behaviors, tics associated with Tourette syndrome, and similar problems with stereotypic behaviors.

Obsessive-compulsive or groove-like behaviors may be misinterpreted as behavior problems. Uninformed or inexperienced authority figures may misinterpret the set routines and behaviors that people follow as oppositional behavior. For example, a teacher asked one of her teenage students to stop what he was doing to do another task. Like many people with Down syndrome, this student continued doing the first task and did not move on to the new task. He was not trying to have negative behavior or attitude but just to finish the first task out of a compulsive need.

If a teacher believed this kind of behavior was opposition or disobedience, she might intensify her efforts to force the student to stop. Force may work if the student is in fact oppositional, but if it is a compulsion, the teacher's behavior will usually only intensify the person's compulsive need to finish. See chapters 10 and 19 for information on recognizing grooves and compulsions, as well as guidance on helpful responses to compulsive behavior.

Motor and vocal tics that occur as a result of Tourette syndrome may also be misinterpreted as a behavior problem. For example, verbal tics involving grunts, mouth noises, or the expression of certain words may be viewed by a teacher or employer as oppositional behavior, particularly when the person has difficulty stopping the behavior. An adult with stereotypical behavior (repetitive movements) such as hand flapping may also be erroneously labeled as oppositional, since attempts to block such behavior are often counterproductive. See chapter 24 for information about appropriate ways of dealing with tics and repetitive movements

Treatment of Serious Behavior Problems

True oppositional behavior, aggression, difficulties with impulse control, and the other behaviors described in this chapter can be particularly problematic challenges. Whether it is an older person with Down syndrome who has begun to strike her elderly parents or a middle school student who is running out of the classroom and fleeing the school, these behaviors can be some of the most challenging. It is at least as important with these challenges as with any other behavior or mental health issue to assess for underlying physical causes, to assess the social context of the behavior, and to employ a multifaceted treatment approach with attention to psychological, social, and medicinal treatments.

The treatment of behavior problems can include (1) behavioral treatment and (2) medications.

Behavioral Treatment

Behavioral treatment can consist of reward systems, redirection attempts, and modeling of appropriate behavior and response to stressful situations. Families or other caregivers should look for a trained mental health counselor who specializes in behavioral therapy to assist them with behavioral treatment. (See the section on counseling in chapter 16 for more on this.)

Reward Systems

Reward systems can be used as part of the treatment plan for a variety of behavior issues in people with Down syndrome. This type of behavioral therapy is often directed only at responding to the occurrence (or absence) of an undesirable behavior. For example, if the person with Down syndrome has no aggressive episodes all week at work, she is allowed to purchase a soft drink on Friday afternoon. There are some additional features that should also be included in the behavioral approach.

First, make sure that the person is not being inadvertently rewarded for inappropriate behavior. If, in the example above, the woman has repeated episodes of aggressive, oppositional, impulsive, or antisocial behavior throughout the week, she will not receive her soft drink at the end of the week. However, if she does not really care for the soft drink and what she is really looking for is increased attention

from the staff, the attention she gets when she is aggressive may be much more rewarding than the soft drink. The behavioral program may actually be rewarding the behavior it seeks to extinguish.

The timing of rewards is another important aspect of the behavioral program. A reward that is three or four days away may have very little impact on some people with Down syndrome. Their attention span may be too short, or the reward may be too distant in the future for them to link their behavior with the reward. Rewards that are given more frequently based on a shorter period of appropriate behavior are more likely to be meaningful to some adults with Down syndrome.

Preventing Problems

Another critical aspect of behavioral programs is usually preventing problems. This involves first analyzing the events that typically lead up to the problem behavior and then figuring out what the person accomplishes with her behavior. (Does she receive attention? Does she get out of doing something she does not want to do?) Afterward, a variety of strategies can be tried to prevent the behavior from occurring. For example, if you learn that a behavior is apparently triggered by a certain event, you can try to prevent that event from occurring. Or, if you can anticipate the event that often provokes the behavior, you can redirect the person before she begins the behavior. Then again, if you learn that the behavior is related to difficulties the adult has in asking for a break, you can teach her a different way of communicating that message.

The systematic process of determining the function of a behavior by analyzing the antecedents (what happens before the behavior) and consequences (what happens after the behavior) is known as *functional behavior assessment* (FBA). It is beyond the scope of this chapter to go into detail about conducting an FBA. It is worth learning more about the process, however, if the adult with Down syndrome you care for has serious behavior problems. For an overview of the process, you may want to read the book *Functional Behavior Assessment for People with Autism* (Glasberg, 2006), or speak to a behavior analyst about conducting an FBA. However, in the meantime, it may be very useful for you to try the redirection strategies described in the next section.

It is important to note that prevention may be more difficult where impulse control problems are involved. For impulse control problems, there is often no clear antecedent event that triggers the behavior. Therefore, preventing the problem may

be difficult because it occurs “out of the blue.” Sometimes, though, there are antecedent events that we are not recognizing. It is often worthwhile to observe the situation several times to see if something might be triggering the situation.

It may also be possible to reduce impulsive behavior if it is associated with a treatable physiological condition, such as AD/HD or a seizure disorder. (See the section below on medication treatment.)

Redirection

Rebecca, a young woman with Down syndrome, would periodically become so frustrated with some of her colleagues at work that she would turn over a table when she got home. Usually if she had had a frustrating day at work, she would pace back and forth in the doorway before entering her home. When they saw this behavior, the staff knew that if they interceded and guided Rebecca to her room to sit down and listen to music, they could usually help her relax and prevent an escalation of her behavior.

Rebecca’s story shows how others can redirect a person with Down syndrome before she becomes aggressive. How and why does redirection work?

Redirection works when a person can successfully change a negative emotion or behavior to a positive emotion or behavior. A major principle of behavioral treatment is that people cannot have two contradictory emotions at the same time. If people feel calm and happy, they cannot experience the negative emotions of anger, sadness, etc., at the same time. Following from this, the goal of treatment is to identify the early stages of a negative emotion and behavior to help to redirect the person to a positive mood and behavior.

Keys to successful redirection:

- Redirection to positive emotions and behavior works best when started before the person is too far into a negative emotional state.
- In order to redirect someone before she becomes angry, try to identify early warning signs that she may display prior to expressing the anger. Rebecca’s

early warning sign was pacing in the doorway. Others may use a wide variety of different and idiosyncratic behaviors as early warning signs.
- Remember that warning signs may change, so continue to observe the person's behavior to identify new signs she may show before expressing anger.

Identifying positive alternatives:

- Regularly observe quiet-time activities to identify activities the person enjoys and that are relaxing to her.
- Try to have a number of relaxing activities from which the person may choose. This gives her more of a say in the process.
- Giving someone a choice of relaxing activities is also a way to direct her attention to something positive without eliciting anger.

Attitude and behavior of caregivers:

- Be very careful in how you approach the person in these situations. If you are too forceful or confrontational, you run the risk of actually provoking anger rather than redirecting the person away from anger. For example, you are much more likely to succeed if you say very calmly to someone who is getting angry, "Would you like to listen to music or draw?" than if you confront the person with this: "You look like you are getting angry. You need to listen to music or draw something. Now!"
- Maintaining a calm tone and attitude is particularly important for the parents of teens and young adults, who are far more likely to rebel against parental authority.

Self-Redirection

Sometimes adults with Down syndrome can be taught self-redirection. That is, the person can learn to identity her own early signs of anger and to redirect herself. This is not an easy task for people with average intelligence and it may be even more difficult for people with Down syndrome. Nevertheless, we have found that with time and practice many people with Down syndrome can learn to redirect themselves. It

may be possible to first teach the person with Down syndrome to respond to cues from others, particularly if a parent or caregiver cues her when early signs of anger are observed. Over time, she may be able to recognize her own patterns of behavior and to begin to cue herself when her mood and behavior changes.

It may also be possible to use self-talk to help someone redirect herself when she is feeling that she is about to act inappropriately. As described in the chapter on self-talk, people with Down syndrome will often say aloud what other adults would silently say to themselves. People can use their own self-talk to cue themselves to redirect an inappropriate behavior. Some will even speak to themselves in the third person when they are redirecting themselves. Annie, age thirty-seven, periodically strikes out at other people. Many times, others have witnessed her saying such phrases as "Annie, don't hit Tommy" when she is agitated. People who hear Annie say this take it as a cue that she needs to do an activity that is relaxing for her and assist her with this until she is relaxed.

We have been able to take this self-talk redirection strategy a step further with many adults with Down syndrome. Not only can some people learn to remind themselves not to express anger inappropriately, but they are also able to redirect themselves to do a positive or relaxing activity as in this example:

When Marvin began to feel himself getting angry and agitated, he would repeatedly tell himself, "Don't yell or throw things." Then he would instruct himself to breathe deeply. He would sit down and breathe in and out deeply for several minutes until he was calm and no longer agitated. At times Marvin had great difficulty practicing this strategy, especially if he was confronted too quickly or forcefully with a very stressful or frustrating situation. For example, once as he entered the dining room of his residence, he encountered a housemate who was having an angry, tantrum-like outburst. Marvin tried to start his redirection strategy, but he was too shaken by the other resident's anger. Fortunately, we had worked with staff to deal with just this type of situation. Several of them gave Marvin a prearranged sign (pointing their thumbs in the direction of the door). This was the signal for Marvin to

leave the immediate situation. He then was able to go to the quiet living room to talk himself through his redirection and deep breathing exercise.

Marvin has been very reliable with starting this strategy when he feels angry. In a few instances, he has needed another signal to remind him. This consists of staff members acting like they are taking a deep breath. This is usually enough for him to start his redirection and deep breathing exercise.

Steps to Take When Someone Is Already Expressing Anger

Sometimes, the person may already be angry and the opportunity to divert her anger to something positive is already past. In these situations, the following guidelines for moderate or more severe angry behavior may help to deal with the problem.

Assess the degree of anger. If the person is out of control and there is a risk that she could hurt herself or someone else, then follow the recommendations in the section below on "More Extreme Anger and Aggression." Otherwise, the following steps may be helpful for managing the anger:

1. Remain calm and in control of your own anger, if any. A caregiver's anger will only further anger or agitate the person with Down syndrome.
2. If possible, give the person room to express her anger. It is particularly important to get children or others who may not understand the danger out of the way. Remember, too, that in a fit of passion any behavior is possible. Therefore, clear away anything that can be thrown or used as a weapon.
3. Once the worst of the storm has blown over, approach the person in an unthreatening way.
4. In a calm voice, gently coax her to sit down to be in a more relaxed physical state. Once she is seated, repeat calming statements.
5. Family members may also gently hug or hold the person with Down syndrome, if she is comfortable with this, to help her relax.
6. Once she is calm, try to engage her in an enjoyable activity, such as listening to music, looking at pictures, etc.
7. When the situation is stable, examine the sequence of events that led to the anger. This may help you identify and resolve whatever problems led to the

outburst. It may also help to identify behavioral precursors to the anger that can cue you to redirect the anger.

Managing More Extreme Anger and Aggression

Dealing with the Immediate Crisis. Assess the aggressive behavior. If the person is out of control and there is a risk that she could hurt herself or someone else, she may require an immediate intervention. If she lives in a larger group home, there may be set procedures for dealing with more extreme aggressive behavior. Usually these procedures involve using some form of physical restraint that is maintained until the person calms down. For example, one larger residential agency has approximately eight staff members who are specially trained by a psychologist. When there is an incident, at least four of these trained personnel are paged to respond immediately to the situation. Following planned procedures, these individuals then carefully but firmly hold the person down so that she cannot hurt herself or others.

If the person lives at home, in a small residence, or anywhere where there is no formal procedure for managing aggressive behavior, then outside help may be needed. Sometimes experienced staff or family members are able to calm the person down when others cannot. For example, one staff person was called at home when Georgia had an outburst because she had a particularly good rapport with Georgia. She responded immediately by coming to the group home, and Georgia responded very positively to her presence. In another situation, an angry adult with Down syndrome calmed down after talking on the phone to his mother.

Sometimes the police may be called when interventions tried by family or staff have not been effective. The presence of police officers may help to calm down some people who have aggressive behavior. This is usually the case when the person does not have an extensive prior history of aggressive behavior. However, caregivers may need to be very active any time the police are called. Most police officers have very little experience or training in dealing with people with Down syndrome or other disabilities. They will often look to the caregiver for guidance. Caregivers may use the police presence to help stabilize the situation but not necessarily to make decisions about treatment options (discussed below). However, the police may be enlisted to transport to the person to a treatment facility if needed (discussed below).

Otherwise, if the person is calm, the staff or family members may be able to safely transport the person to an appropriate facility, if deemed necessary.

Seeking Treatment. Once the adult is calm and the crisis is over, there are two primary courses of action involving outpatient or inpatient treatment. Many times, staff will bring people with more extreme aggression to the attention of mental health professionals. This usually involves a consultation with a behaviorist and/or mental health professional in a medical or mental health outpatient setting. We have been asked to assess and treat many people on an emergency basis who display more extreme forms of aggression. In our experience there are many different causes, explanations, and possible treatments for these behaviors, which we have discussed in detail throughout this chapter and throughout the book.

Sometimes when adults with Down syndrome have extreme behavior, they are taken by staff, police, or their families to a hospital emergency room to be assessed for possible hospitalization in a mental health ward. In larger cities, there may be hospitals with wards specifically for people with intellectual disabilities. However, in most cases, the only option is a general psychiatric ward.

We have found that hospitalizations are not always as beneficial as people would hope. Ideally, hospital staff attempt to identify and treat mental health and health conditions and to work with family and staff to identify sources of stress. Too often, though, staff do not have enough experience or comfort with diagnosing and treating people with Down syndrome for mental health or behavioral problems. They often neglect to do other medical testing to identify health problems. Hospital staff may also not make an effort to contact the family or staff to help identify and resolve environmental conflicts or stress. As a result, the hospitalization may become little more than an extremely expensive respite, which buys time, and gives family and staff a break, but solves none of the problems or issues that may have caused the problem to begin with.

Sometimes hospitalization actually worsens the problem. This is because caregivers may rely heavily on hospitalizations for crisis management, but this does not necessarily help to resolve the problems leading to the behavioral outbursts. Additionally, some people with Down syndrome actually like the experience of being hospitalized. They may be doted over by staff and there may be little pressure on them to do anything constructive. Some adults with Down syndrome experience this as a type of vacation. Unfortunately, this is an extremely expensive “vacation,” and again, not necessarily productive if there is no attempt to identify the underlying problem or to teach the person how to better manage her behavior

We have also found that many hospitals will not admit people with intellectual disabilities to the psychiatric ward even if they are seen in their emergency departments. Sometimes hospital staff do not feel comfortable treating a population for whom they have little experience. Often the person who is taken to the emergency department will eventually calm down, even if she is still agitated upon arrival. Some may lose their anger and hostility when they see others in the emergency department who have more serious physical or emotional conditions. Waiting to be seen for hours in an ER may also deaden any anger that remains. Failing to be hospitalized is not necessarily a bad thing. This is especially the case if the hospitalization is not geared to identify and resolve the problems causing the behavior, as discussed immediately above.

Additionally, many times people who are turned away from the emergency department will then look for resources in the community. Once the immediate crisis is over, they may have more time to locate an outpatient facility with more experience serving people with disabilities. These facilities will have professionals who may have more success with resolving the causes of the behavior problem.

Evaluation of the Causes of an Extreme Behavior. Once the crisis of an extreme aggressive incident is over, a thorough evaluation of the possible causes of the problem needs to commence. Efforts to treat without this evaluation may lead to failure if the real reasons behind the angry outbursts are not identified and resolved. Often, once the cause of the problems is dealt with, the behavior will become more manageable. Once this occurs, the guidelines discussed above for more moderate forms of anger may be followed to help manage any remaining aggressive outbursts that occur.

Here are some of the most common causes of extreme behavior:

- The behavior may be due to extreme environmental stress. Consider this possibility particularly if there is no history of prior behavioral outbursts. For example, Bret had simply had it with being victimized by a bully in his residence. His behavior was a “wake-up call” to staff to move him or to better manage the behavior of the bully, who had been abusing a number of people in his residence. Once this problem was solved, there were no more outbursts from Bret or any of the other residents in the household.
- As mentioned previously, health problems may create extreme pain and discomfort and may contribute to or cause extreme behavior changes. A

thorough physical exam should be conducted whenever there are changes in behavior, especially more extreme changes.

- Extreme behavior may also be related to a mental health problem. As mentioned throughout this book, many people with Down syndrome have difficulties communicating problems verbally. A mental health problem may surface as an extreme change in behavior. Therefore, if the person's aggressive behavior is not explainable by environmental stress, by a medical condition, or by any other type of behavior disorder (conduct disorder, oppositional defiant disorder, etc.), then a consultation with a mental health professional may be advisable. For example, people with previously undiagnosed bipolar disorders have been brought to our office with a recent history of aggressive outbursts.

Medications

As discussed previously, behavior problems, and especially impulsive behavior, may be caused or aggravated by physiological or neurological conditions such as AD/HD, seizure disorders, Tourette syndrome, and tic disorders. The behavior problems that result may be far more manageable if medications are used to treat these conditions. For example, people with attention-deficit disorders may greatly benefit from a stimulant medication, while those with seizure disorders may respond positively to an anticonvulsant medication, and those with Tourette syndrome/tic disorders may respond to antipsychotic or other medications. Similarly, we have found that some individuals with more severe impulsive behavior may respond to anticonvulsant medications even when there is no apparent physiological cause. This may be due to the fact that some seizures are difficult to detect because they occur intermittently and have subtle symptoms.

Certainly, a host of other medical conditions may also cause or aggravate behavior problems, as discussed previously. These may include thyroid disorder, B12 deficiency, and other disorders discussed in detail in chapter 2. As discussed previously, appropriately treating physical problems that are contributing to behavioral problems is necessary to optimally reduce the abnormal behavior.

Additionally, medications may be helpful when mental health symptoms coexist with behavior problems. One of the most common of these symptoms is anxiety, which is often manifested as agitation and body tension. We also find mood disorders to be fairly common with behavior problems. A mood disorder may include the more

severe fluctuations in mood and behavior associated with bipolar disorder (see chapter 17), but far more often it includes less severe symptoms of moodiness and irritability.

The anticonvulsant (antiseizure) medications are an effective treatment approach for behavior problems, including agitated and aggressive behavior, associated with mental health symptoms. Valproic acid (Depakote, Depakene) and lamotrigine (Lamictal) are FDA approved for bipolar disorder. Carbamazepine (Tegretol) and oxcarbazepine (Trileptal), though not FDA approved, are also used for bipolar disorder. People on these medications need to have periodic blood tests to check medication levels, as well as tests such as CBC, liver function, and electrolytes to check for side effects. We have also had some success with gabapentin (Neurontin), which is not as widely recognized as a choice and is probably not as effective as the others and is not FDA approved for bipolar. The advantage of gabapentin is that there is less need to monitor blood work, and therefore it is better tolerated by our patients who dislike blood drawing. Although none of these medications are FDA approved for "behavior issues" when there is no diagnosis of bipolar disorder, we often find they are beneficial.

Antipsychotic medications can also be quite beneficial in treating these types of behavior problems with mental health symptoms. Risperdone (Risperdal), olanzapine (Zyprexa), queitapine (Seroquel), ziprasidone (Geodon), and aripiprazole (Abilify) have all been effective in some of our patients with Down syndrome and are FDA approved for bipolar disorder. As indicated in chapter 17, however, monitoring for sedation, weight gain, and elevated blood sugar is necessary.

Antidepressants are not generally beneficial when used on their own to treat these types of behavior problems but may be helpful in combination with other medications (such as antiseizure medications). The antidepressant trazodone (Desyrel) can be particularly helpful as an adjunct medication in treating behavior and mental health problems. It is especially helpful as a sleep aid. Therefore, it may be a good addition for patients who have sleep disturbance as part of their symptoms. Similarly, use of melatonin (a hormone used for sleep disturbance and jet lag) or other sleep aids such as zolpidem (Ambien), eszopiclone (Lunesta), or zaleplon (Sonata) may play a role in treating these conditions.

Finally, if the challenging behavior is associated with an obsessive-compulsive disorder or with major depression, an antidepressant medication that treats these conditions may help to reduce the behavior problem.

Conclusion

When an adolescent or adult with Down syndrome is aggressive, impulsive, or extremely oppositional, or is engaging in other challenging behaviors that interfere with daily life, it is extremely important to try to determine what is triggering this behavior. A careful assessment for underlying medical issues, as well as for possible environmental triggers, is essential. As with the treatment of other mental health problems, addressing psychological, social, and biological aspects increases the likelihood that treatment will be successful.

Return to Table of Contents

Chapter 23

Self-Injurious Behavior

When Joshua, age twenty-four, became frustrated or upset, he would kick his foot against a piece of furniture to the point of bruising. A careful assessment revealed no physical problems that could contribute to this behavior. Joshua had limited expressive language skills, but better language comprehension. Through counseling we were able to help Joshua and his family identify occurrences that would typically trigger self-injurious behavior. Limiting exposure to the events that he found frustrating or upsetting was part of the initial treatment. Strategies were also developed to help him express his frustration in alternative ways such as playing a brief stepping game. Over time, Joshua learned to do the stepping game earlier and earlier when frustrated so that he was calming himself long before he previously would have gotten to the point of hurting himself. He also learned to use the strategy more subtly so that it was barely noticeable to those around him.

On the surface, self-injurious behavior may seem one of the most difficult-to-comprehend mental health problems. Most of us do not expect people "in their right minds" to willingly hurt themselves. However, self-injurious behavior is not always a symptom of mental illness. When it occurs, though, it requires diagnosis and treatment, which is why it is included in the Mental Illness section of this book.

Self-injurious behavior can be a means to express discomfort, displeasure, or even pleasure. It can be seen in a variety of mental health problems. In addition, it may be a means of communicating physical pain.

What Is Self-Injurious Behavior?

By *self-injurious behavior*, we mean behavior that causes injury to oneself. This may include striking oneself, biting oneself, falling, running into walls, and other activities that cause injury.

Self-injurious behavior is not common in adults with Down syndrome but does seem to occur more often than in people who do not have developmental disabilities. One reason is that self-injurious behavior often appears to be a form of communication. Often the person who does self-injurious behavior has limited communication skills. This makes it that much more difficult to develop an understanding of the problem and to treat it.

Causes of Self-Injurious Behavior

In people with DS, self-injury may occur for a variety of reasons, including the following:

- The person finds self-injurious behavior pleasurable or rewarding.
- The person has autism in addition to Down syndrome.
- Self-injury helps relieve anxiety or stress.
- Self-injury is an effective means of communication.
- Self-injury is related to pain or a medical condition.

Finding Self-injury Pleasurable. Difficult as it may be to believe, self-injury seems to be rewarding for many people who engage in it. They don't seem to experience this injurious behavior as painful and may actually find it pleasurable. Difficulties in understanding how this injurious behavior is pleasurable or rewarding to the person can pose obstacles to understanding the behavior and developing an effective behavior program. However, endorphins may play a role. Endorphins are a natural substance produced by the body in response to pain (and certain other causes) that stimulate the opiate receptors, reduce pain, and can cause euphoric or positive feelings.

Self-Injury in Autism. Self-injurious behavior may be one of the symptoms seen in autism spectrum disorders. There are a number of theories as to why this behavior occurs in people with autism. It may be part of self-stimulation, may release endorphins and cause pleasure, may be a way to get attention, or may be a symptom of a subclinical seizure. Autism is discussed in chapter 25.

Relieving Anxiety. If someone is feeling anxiety or stress, self-injury might reduce that sensation. It may also distract the person from the anxiety. For example, if you are worrying about something and accidentally cut your finger or drop something on your foot, you immediately start thinking about your finger or your foot

instead of your worry. In addition, if the person is overwhelmed by the anxiety, he may respond irrationally when the anxiety "boils over." That is, he may respond to the sense of loss of control caused by the anxiety by striking or otherwise injuring himself. Some people with an intellectual disability have less ability to control their actions or to understand what an appropriate response is.

Self-Injury Related to Medical Problems. Self-injurious behavior can be a symptom of a variety of physical health problems. Its purpose may be to inform others of the discomfort or perhaps to eliminate the pain. For example, we had a patient who was depressed and had a chronic sinus infection. Whenever discomfort from the sinus infection occurred, he would repeatedly strike his head. Once the infection was diagnosed and treated, he stopped striking his forehead.

Often the person will strike a location on his body that is different from the location of the discomfort. This appears to be a general display of pain or frustration or a generalized call for help. We have treated a number of people who bit their hands, struck their chests, or hit their head against objects when uncomfortable.

Self-Injury as Communication. Self-injurious behavior can be a wonderfully effective communication strategy—particularly if the goal of the behavior is to gain attention. When someone observes a person who is performing self-injurious behavior, a very natural reaction is to attempt to stop him or her. This effort rewards the person with attention.

Self-injury can also be a very effective way to demonstrate displeasure with something that is going on in the environment:

> *Samir would slap himself in the face whenever he did not like what someone else was doing. His roommate, Oscar, had a tendency to quietly tease Samir in such a way that the staff members of their group home were unaware that it was happening. However, whenever Samir would slap his face in response to Oscar's teasing, the staff would intervene and tell Oscar to stop bothering Samir.*
>
> *Telling Oscar to stop teasing Samir whenever Samir slapped his face did stop the slapping immediately. However, it also encouraged Samir to continue to slap his face to get Oscar to stop his teasing. After assessing the situation, the staff observed Oscar more closely for teasing behavior. They worked on intervening before Samir started hurting himself. In*

addition, they watched for early signs that Samir was beginning to get agitated (if they had missed Oscar's teasing). When they saw these signs, they praised Samir for not injuring himself and redirected Oscar away from his teasing.

Samir learned that he did not need to strike himself to cause a change in his environment. Over time, the staff taught him to use a communication book that included a picture that indicated he was unhappy with something. He and the staff were able to learn a new way for him to communicate and he no longer felt the need to hit himself to let staff know he was unhappy.

When assessing self-injurious behavior, it is important to ascertain if it is a form of communication. Some steps to assess the behavior as communication include the following:

- Analyze what the person gets as a result of the injury. (Does he get attention, removal from a situation he finds troubling, or something he wanted?)
- Consider whether the behavior may be linked to a health issue (or is there some other indication that he may be in pain or have an illness)?

As discussed in the chapter on depression (chapter 17), suicide seems to be uncommon in people with Down syndrome. This generally does not seem to be the motivation for self-injurious behavior.

Treatment of Self-Injurious Behavior

Treatment depends on the underlying reason for the self-injury. If it's a form of communication, as described above, the solution is often to teach the person a different way of communicating the problem. A speech-language pathologist may need to be involved, especially if the person is not very verbal and may need an alternative communication system.

Redirection

When there is no apparent cause for the behavior or no apparent communicative intent for the behavior, just redirecting the person away from the behavior is sometimes the most effective intervention. For example:

> *Louise, a nonverbal woman in her forties, had a tendency to strike herself in the head. We found no evidence that she was trying to communicate something, that she was in pain, or that there was an underlying physical problem. We instructed staff to observe her and initiate a clapping game with her whenever she began to hit herself or looked like she was going to hit herself. Eventually, Louise was able to start the clapping game on her own.*

To redirect someone away from self-injury, follow these steps:

- Choose a substitute behavior that is incompatible with the self-injury. Louise struck herself with her hands, so staff used a clapping game to occupy her hands (as well as her attention) and reduce the self-injurious behavior.
- Watch for warning signs that the behavior is about to occur and intervene before the behavior starts.
- If the person is using something readily accessible (such as a sharp corner of a piece of furniture) to cause injury, safety-proof the environment.

Helping someone learn to redirect himself is a very effective method for reducing self-injurious behavior. One way is to have the person tell himself out loud not to hit (or otherwise hurt) himself. Start by asking him to repeat the phrase after he hits himself or engages in other self-injury. Next, work with him to say it during an episode of self-injurious behavior. Finally, watch for warning signs and work with him to say the phrase before he hits himself. Interestingly, many people with Down syndrome who use this technique address themselves in the third person when they do it. For example, when David redirects himself, he says, “David, don’t hit.”

Counseling

Counseling may also be effective for some adults with Down syndrome. Particularly if a stressful situation tends to trigger the self-injurious behavior, counseling may help the individual discover the event or events that lead to the behavior. Counseling may be done with the individual alone or with the family or caregiver participating in the session. It depends on the individual. However, it is often fruitful to give the person with Down syndrome an opportunity to participate in individual therapy with the counselor. If he is nonverbal, alternative means of communicating may be tried such as drawing pictures, using a communication app, etc.

Through counseling, adults with Down syndrome can be taught other ways to deal with stress. Use of devices to promote "self-redirection" may be effective, as in this example:

> *Indira had a tendency to hit herself when she became anxious or agitated. She was able to communicate what she was worried about, which allowed the counselor to help her see the connection. They were also able to develop a system together to allow her to redirect herself. Indira agreed that when she was feeling anxious, she would pull her "worry stone" out of her pocket and rub it instead of striking herself.*

Similarly, other people with Down syndrome have been able to learn to go to their rooms, sit down, listen to music, and relax when they are feeling anxious.

Medication

In addition to behavior techniques and counseling, medication can be beneficial for people with self-injurious behavior. There may be an underlying psychological problem that causes or contributes to self-injurious behavior. An assessment for an underlying psychological problem will help guide the medication selection. Further information on the medications discussed below can be found in chapter 16.

Helpful Medications When Anxiety and Depression Are Involved

When anxiety is associated with self-injurious behavior, antianxiety medications can be helpful in reducing self-injury as well as anxiety (see chapter 18). We have had limited success with buspirone (Buspar), but it often takes several weeks to see the benefits. The benzodiazepine medications (e.g., alprazolam, lorazepam, etc.) can be used while waiting for the effects of buspirone to be felt. In addition, the benzodiazepines can be used as the primary treatment. The longer-acting choices, in particular, can be used on a regular basis. However, the potential for developing tolerance, addiction, and withdrawal symptoms (when the medication is discontinued) may limit their benefits.

Antidepressants can also be used to treat anxiety. We have found the selective serotonin reuptake inhibitors (SSRIs) and serotonin-norepinephrine reuptake inhibitors (SNRIs) to be beneficial for most adults with Down syndrome who are experiencing anxiety. Among the SSRIs, paroxetine (Paxil) and escitalopram (Lexapro) have FDA approval for generalized anxiety disorder, and paroxetine, sertraline (Zoloft), and fluoxetine (Prozac) are approved for panic disorder. For the SNRIs, duloxetine (Cymbalta) and venlafaxine (Effexor) are approved for generalized anxiety disorder, and venlafaxine is approved for panic disorder. We have also used bupropion (Wellbutrin) with some success, although it does not have FDA approval for this use.

When self-injurious behavior is a manifestation of depression, antidepressants can again be quite beneficial. As indicated above for anxiety, the SSRIs, SNRIs, and bupropion can be helpful in some individuals.

Helpful Medications When Sleep Disorders Are Involved

As discussed in chapter 2, sleep disturbance is more common in people with Down syndrome. When sleep disturbance is chronic, it can lead to agitation. Self-injurious behavior may be the expression of the agitation. Restoring a more normal sleep pattern can be a very effective way to reduce agitation and any self-injurious behavior associated with it.

Sleep apnea is more common in people with Down syndrome and can cause agitation. If an adult with Down syndrome has sleep apnea, it should be brought under control as much as possible through CPAP or BIPAP, supplemental oxygen, position change, or surgery (see chapter 2). Untreated sleep apnea can affect both physical and mental health.

Sleep disturbances due to other causes, or unknown causes, might be treated with supplements or medications. We have found melatonin (a hormone used as a sleep aid and to treat jet lag) to be helpful in reducing self-injury associated with poor sleep. We have also used valerian root and magnesium supplements. The antidepressant trazodone (Desyrel) is also an effective treatment for self-injurious behavior, particularly when there is sleep disturbance. In our experience, trazodone seems to work better as a sleep aid than as an antidepressant in people with Down syndrome. We have also prescribed short courses of other sleep aids such as zolpidem (Ambien), eszopiclone (Lunesta), or zaleplon (Sonata).

Once the person gets back on a regular sleep pattern with the help of the medications, he may continue to sleep well (at least for a while) without the sleep aid. However, it is important to watch for recurrence of sleep disturbance and self-injury and to consider prescribing the medication again, if necessary. Some individuals require long-term use of a sleep aid to optimize their sleep (and behavior).

Other Medications

Antiseizure Medications. Antiseizure medications (anticonvulsants) can also be an adjunct to behavior techniques in treating people who have self-injurious behavior. For example, valproic acid (Depakote) has FDA approval for mania and has been found to be beneficial for agitated behavior, impulse control disorder, and self-injurious behavior. Carbamazepine (Tegretol) and oxcarbazepine (Trileptal) have also been used for these problems, although they are not FDA approved for this indication.

Gabapentin (Neurontin), another antiseizure medication, is less well known as a treatment for self-injurious behavior, but limited research data supports its use in mania. Although we knew there was neither FDA approval for this medication for self-injurious behavior nor a history of recognized benefit, we first tried it in several patients with self-injurious behavior who would not allow us to draw blood. (Drawing blood to monitor for possible side effects is usually recommended for other antiseizure medications such as valproic acid and carbamazepine.) We found that gabapentin is an effective treatment for some individuals for self-injurious behavior, aggressive behavior, and impulse control disorder. It can be used with less concern (although not no concern) about the need for blood test monitoring for people who are particularly resistant to blood drawing.

Atypical Antipsychotics. The atypical antipsychotics are quite beneficial in treating self-injurious behavior in people with Down syndrome. We have successfully

reduced self-injurious behavior with risperidone (Risperdal), olanzapine (Zyprexa), ziprasidone (Geodon), quetiapine (Seroquel), and aripiprazole (Abilify). Again, these are not specifically FDA approved for self-injurious behavior. In addition, we have unfortunately seen tremendous weight gain in some individuals with Down syndrome on these medications. We have also sometimes seen significant elevations of blood sugars (necessitating discontinuing the medication).

It is important to note that the people with Down syndrome and self-injurious behavior we have treated generally do not have clear psychotic symptoms. We are not using the medication to treat psychosis in this situation, nor using it as a sedative. In fact, if sedation occurs as a side effect, we usually reduce the dose or change the medication.

Naltrexone (ReVia). As mentioned above, some people may find self-injury pleasurable because pain can cause the body to release natural substances called endorphins. Endorphins stimulate the opiate receptors in the nervous system, which may cause the person to perceive the sensation as enjoyable. For this reason, medications developed to block the effect of opiates (narcotics) on the opiate receptors can be helpful in treating self-injurious behavior. Possibly, by blocking the opiate receptors from working as usual, any pleasurable effects of self-injury can be blocked as well. The opioid antagonist (blocker) we have had some success with is naltrexone. Since it can be difficult to tell whether someone with Down syndrome "gets pleasure" from self-injurious behavior, it might be reasonable to try naltrexone even in individuals who don't seem to find the self-injurious behavior pleasurable.

Eating Feces

Another problem that appears to be similar to self-injurious behavior is eating feces. This behavior has significant potential to hurt the individual. In people with Down syndrome, eating feces often seems to occur in association with one of a couple of issues. First, we have seen a few people whose vision was declining and may not have recognized that what was on their hands was not food. In these cases, treating the vision problem often helps reduce the eating of feces.

Second, some people with Down syndrome seem to find eating feces pleasurable. As with self-injurious behavior, it is difficult to understand how this can be so. Generally, we have found that individuals with Down syndrome who eat feces are more severely cognitively challenged and have limited or no verbal skills. Some

also have autism or pica (eating inedible substances). Counseling is often of little benefit in these situations.

Redirecting and closely monitoring the person may improve the situation. Giving him something else to eat, especially food that is a similar consistency and texture, may be helpful. Sometimes giving the person something else to hold in his hands helps, but some people might eat that instead.

Tailoring clothing to make it harder for the individual to reach stool may be helpful. Long-legged pants or one-piece garments that open in the back where the person can't reach have been successfully used. We generally only recommend this strategy for people who are not able to use the toilet independently because this clothing would prevent independent toileting.

Medications are usually the mainstay of treatment. The medications discussed above may also be helpful to reduce eating feces. Clearly, it is also important to monitor and treat the person for illnesses such as diarrhea or upset stomach resulting from eating feces.

Conclusion

Self-injury can be quite harmful to the person who is engaging in the behavior. Witnessing self-injury is usually quite disturbing to those around the person as well. Careful assessment is needed while keeping in mind that the adult with Down syndrome may be using self-injury to communicate that he is in pain or experiencing stress. Counseling, redirection, behavior programs, and medications may all be part of the treatment for individuals with this problem.

Return to Table of Contents

Chapter 24

Tics, Tourette Syndrome, and Stereotypies

Lily, a fifteen-year-old girl with Down syndrome, came for an evaluation due to changes in her behavior. Of particular concern was an increasing amount of compulsive behavior, which was impairing her ability to participate in daily activities. For example, on the way to the exam room, she stopped and closed every door she passed. She was unable to go directly from the waiting room to the exam room because of her compulsive need to close doors. Also during the exam, she had intermittent hand movements and eye blinking and periodically made incoherent vocalizations, although she had good verbal skills. Lily's family reported that the movements had been present for several years and that as a young child she had been diagnosed with attention-deficit hyperactivity disorder. There was no decline in Lily's skills, although she was having difficulty performing at her usual level due to her rituals and hand movements.

After a thorough evaluation, we diagnosed Tourette syndrome. We initially prescribed sertraline (Zoloft) for the obsessive-compulsive symptoms and later added clonidine (Catapres) for the tics. Lily's symptoms did not completely resolve, but they improved significantly and she was able to return to her previous activities and level of function. Lily and her family decided not to try higher doses or other treatments because she was doing quite well, and they had concern about possible side effects or a "backward slide" with any additional changes.

Many people with Down syndrome repeatedly make movements or sounds that others may consider odd or annoying. For example, they may grind their teeth, hum to themselves, wring their hands, or rock back and forth when listening to music or watching television.

Sometimes these movements and sounds are what are called stereotypies (also known as *self-stimulatory behavior* or *stereotypic behavior*). Stereotypy is defined as a motor behavior that is repetitive, often seemingly driven, and nonfunctional. It includes repetitive motor behaviors and the repetitive movement of objects. These motor behaviors may at times interfere with normal activities and may result in self-inflicted bodily injury.

Sometimes repetitive movements may be related to other things such as compulsions, although compulsions are usually more complex than stereotypies. Stereotypies involve the repetition of more simple behaviors, such as hand flapping, body rocking, or head banging, whereas compulsions often entail a more complex series of steps such as arranging personal items so that they are "just so." Repetitive movements may also be manifestations of stress, agitation, anxiety, or excitement rather than stereotypic behaviors. In addition, some types of motor movements are side effects of neuroleptic medications (see chapter 16). Some movements may also be related to medical conditions such as seizures or seizure-like conditions in Alzheimer's disease, or to less common health conditions such as Huntington's disease or strokes. Finally, some repetitive movements are actually tics, or involuntary sounds or actions that are due to biochemical differences in the brain. Tics may at times be difficult to differentiate from stereotypies.

Because repetitive movements and sounds are so common among people with Down syndrome, it is important to understand the different underlying reasons for them, as well as what, if anything, should be done to reduce their occurrence. With tic disorders, for instance, medical treatment may be extremely beneficial, but with stereotypies, medical treatment may not help and may sometimes actually do more harm than good.

Stereotypies

Jasmine's whole body gets involved when she is excited about something. For example, at home, when watching a favorite video, she frequently wrings her hands when she knows a "good part" is coming, or she may shoot her arms straight out in front of her and scrunch up her face in a happy grimace, her eyes wide with pleasure. She may wring her hands, stretch her fingers out, and grimace dozens of times in the course of a movie. At school or in other settings where she is less relaxed and

more aware of others' reactions, she rarely, if ever, wrings her hands or grimaces.

Like Jasmine, many people with Down syndrome have some seemingly odd, purposeless actions that they do over and over again. These stereotypies are presumed to occur more frequently in people with developmental disabilities. We have found that many types of stereotypies are fairly common and occur regardless of the person's level of skill or functioning. Common stereotypies include

- hand flapping or hand wringing,
- rocking back and forth or side to side,
- humming or making other types of mouth noises, and
- manipulating objects in some repetitious way (rubbing, twirling, etc.).

Of course, many people have their own idiosyncratic versions of these behaviors involving specific types of objects they manipulate or unique movements or sounds they may make.

Frequency-of-Occurrence Continuum

Among people with Down syndrome, there is a continuum from a very high rate of stereotypies to a very low rate of occurrence, with the vast majority of people with Down syndrome falling somewhere in the middle. At the low end, there are a relatively small number of people who rarely engage in these behaviors and at the higher end, a small number who have a high rate of behaviors.

People with a High Rate of Stereotypies

Many of the people at the high end of the continuum spend a considerable amount of their waking hours engaged in stereotypies. These individuals tend to have more severe impairments in intellectual and adaptive functioning. This group may also include people with autism spectrum disorders, and especially those with significant social and expressive language limitations.

These adolescents and adults often have the same types of stereotypies as those in the more moderate group (discussed below), but the frequency and duration of their behaviors greatly interfere with their functioning in other important life

spheres. Many of these individuals also have self-injurious behaviors, often more severe forms of self-injury. Examples of self-injurious behavior include rubbing, biting, or chewing on hands, knuckles, or other body parts, as well as picking at skin and at sores. Often there is also a history of hitting or slapping themselves in the face or body, head banging, or other forms of self-injury.

Some people in this group have more odd types of behaviors. For example, they manipulate objects in unusual ways, such as dangling or shaking action figures, or they manipulate unusual objects such as strings, paper items, pieces of clothing (socks, underwear, etc.), or shiny metal objects.

Some of these behaviors pose a significant challenge to caregivers. For example, some people have more onerous behaviors, such as licking or smelling objects or people, anal digging, fecal smearing, masturbation, or grabbing at genitals, etc. The treatment for these severe problems is an intensive and often lifelong process of diverting the person's attention to more productive social and adaptive behavior. (For more on the treatment of these behaviors, see chapter 16.)

People with More Moderate Rates of Stereotypies

As mentioned above, many people with Down syndrome do have noticeable stereotypies. These may include such behaviors as hand flapping or wringing, rocking or swaying, mouth noises, and manipulating certain objects. Some of these individuals may also have self-injurious stereotypic behaviors, particularly when anxious or stressed.

For people with more moderate rates of stereotypies, we have found that it is possible to predict when these behaviors are more or less likely to occur. These behaviors are less likely to occur when people are engaged in work, social, or recreation activities that require more of their attention and physical engagement. The behavior is more likely to occur when people have downtime, such as when they are idle at work or when relaxing at home in the evening listening to music or watching TV. Often the behavior also occurs more frequently and intensely when the person is having some type of emotional experience, such as the pleasure and excitement Jasmine felt in the above example. On the other hand, stereotypies may also increase when the person experiences negative emotions, such as stress or anxiety.

What Causes Stereotypies?

There are theories but no definitive answers as to what causes stereotypies or why this behavior occurs more frequently in persons with developmental disabilities. Understandably, much of the research has focused on people with more severe impairments who have a higher rate of stereotypies. Still, these theories may help those with more moderate stereotypies as well. Some researchers believe stereotypies may be caused by a deficiency in the central nervous system, resulting in a need or craving for intense stimulation. This could help explain the increase in the behaviors when someone is relaxing and is mildly understimulated. Other theorists believe the opposite, that people are overstimulated and use the behavior to try to block input from their environment. Still others believe that stereotypies may be self-soothing.

Each of these theories may have validity for some people. On the other hand, in many situations (such as in the example of Jasmine, above), stereotypies may simply indicate the presence of something that is stimulating and not necessarily something that is under- or overstimulating. Then again, it is possible that the behaviors may simply be a complement to whatever else an individual does to relax, such as play music, watch TV, etc.

Understanding stereotypies that result in self-injurious behavior may be more difficult. As discussed previously, we have seen a relatively small number of individuals who do more serious self-harm. What causes this? Ironically, this behavior may actually be self-soothing for some individuals. Researchers have found evidence that more severe self-injurious behavior may release pleasurable endorphins in the brain.

Although the release of endorphins may help to explain the contradictory behavior of people who seem to find pleasure in hitting or harming themselves, it may not help to explain the more moderate forms of self-injury we encounter more frequently in our clinical work. Examples include picking at skin or sores, scratching, and chewing or biting at fingers and knuckles. We have found that these more moderate self-injuries tend to occur when people experience, or are overstimulated by, some degree of stress in their lives.

When to Seek Help for Stereotypies

Families are often concerned that these behaviors are a sign of some health problem or an indication of an autism spectrum disorder. The key question here is whether the behavior interferes with normal life activities or results in harm to the person or to others. If it does neither of these, then we recommend that caregivers try to ignore the behavior, particularly if done in a private space.

There are some situations, however, when the behavior itself is not the problem but rather when and where it occurs. For example, hand flapping or rocking back and forth may be a problem if done in a shopping center, a workplace in the community, or in some other public setting. If this behavior occurs in public, it may draw attention and lead to possible ridicule from others. If so, the person may need to learn to limit the behavior to a private space. For instance, a capable young woman who worked in a bank was flapping her hands and drawing attention from others. After talking out possible solutions with her supervisor and parents, she decided on her own to curb this behavior because she wanted to fit in. In other situations, the solution may be for others to change, rather than the person with Down syndrome. For example, in the case of one fifteen-year-old boy who often flapped his hands, enlightened school staff used sensitivity training to educate other students as to the normalcy of this behavior, and the teasing stopped.

Of course, there may be other important considerations such as the type of behavior, the attitude of caregivers who are present, and when the behavior is displayed. For example, many people with Down syndrome (like Jasmine, above), briefly display hand flapping, a rocking behavior, or facial grimacing when they are happy or excited. After they express these behaviors, they are over with fairly quickly. Many family members simply tell others that this is the way the person shows excitement and enthusiasm.

Although some people may briefly rock when excited, more pronounced rocking may be a problem in public. The bad news about rocking is that compared to hand gestures or other forms of stereotypies, rocking is a bigger and bolder behavior and often more obvious to others. More importantly, others are more likely to associate this behavior with people with intellectual disabilities. As a result, it may serve to mark the person as disabled like a big neon sign and thus may make it more difficult for her to go about her normal business in public. This may be even more of a problem at school, where students with Down syndrome may be subject to ridicule from peers.

The good news about rocking behavior is that it is also more obvious to caregivers, and if they are present, they can often redirect the person to a different activity or behavior. We recommend finding less obvious ways of reminding the person that she is rocking, such as by touching her shoulder. You might also develop private signals to alert her when this is going on. It may also be possible to predict when this behavior is likely to occur and help her find alternative ways of expressing herself in these situations.

When Stereotypies May Need to Be Changed

In some situations, the stereotypic behavior itself is harmful or interferes more prominently in the person's life and therefore requires more of an intervention to correct or change it. Following from the research, it may be helpful to regard these more problematic stereotypies as possible indications that the person is under- or overstimulated.

In our experience, one of the major causes of unproductive stereotypies, including self-injurious behavior, is an unstimulating workplace. This is particularly the case for people with Down syndrome who are conscientious about their work. Being idle or having downtime at work can be "deadly" for them. Too often, supervisors respond with activities that are not interesting or stimulating. For example, many worksites that serve adults with developmental disabilities show movies or TV or fill the time with what one employee called the "same old tired busywork" (unpaid assembly tasks such as sorting different sizes of nuts or bolts solely to pass time).

Worst of all, some agencies do absolutely nothing. Employees are simply left to their own devices. We have heard from many families who have tried to find ways to help their sons and daughters deal with downtime. They often send the adult with Down syndrome to work with favorite free-time activities, such as paper to write or draw on, books and magazines, needlepoint, CD players, etc. These activities can occupy people for some time, but asking them to entertain themselves all day is not reasonable. Naturally, this kind of downtime results in an increase in stress and anxiety as well as unproductive stereotypies.

During downtime at work, we strongly recommend providing activities that are physically and mentally stimulating such as the following:

- beneficial recreation programs such as dancing, aerobics, or walking clubs

- higher caliber arts and craft programs that challenge and inspire people to produce high quality work
- outings into the community to mundane places such as shopping centers, but also to more interesting places such as museums and cultural events
- interesting volunteer work in the community (for example, one worksite has a very popular outing where people work doing a variety of clerical and janitorial task at local churches)

On the other hand, people with excessive stereotypies may also be communicating the presence of a stress or condition that is overwhelming or overstimulating to them. We recommend taking these steps to resolve the problem:

1. First, try to identify and reduce the cause or source of the stress. For example, people with Down syndrome are often very sensitive to the feelings and conflicts of others, especially of significant others (see chapter 14). Some people may be victimized by others (see chapter 6). Life changes or losses may also be especially stressful for people with Down syndrome, who crave consistency in their lives (see chapter 10). People may also be exposed to situations that overload their senses such as loud group homes or worksites.
2. After reducing stress, try to keep the person busy with interesting activities. This helps to distract her from preoccupations and stress.
3. Finally, when the stereotypy itself is a problem, remember that it is easier to divert the behavior to something more appropriate than to try to stop the behavior altogether. For example, an adult who picks at her skin may need activities that occupy her hands. Some people may find rubbing worry stones (small smooth stones) helpful. These stones may be rubbed while in the person's pocket or they may be small enough to not be noticed by others. Keeping a pad and pencil handy to write on may help some people keep from scratching or some other behavior. Still others may find that chewing on a toothpick, gum, etc. may help to keep them from biting nails or fingers.

Tic Disorders

People who have tic disorders, like people with stereotypies, also make seemingly odd or purposeless movements or sounds. A big difference, however, is that

tics may be suppressed for short periods of time, and tics are associated with a premonitory urge. That is, before the tic occurs, the person senses that the tic coming on, much as you might feel the urge to sneeze. The table below lists other differences between stereotypies and tics.

Stereotypies versus Tics

Stereotypies	**Tics**
Fixed, identical, foreseeable	Suggestible
Arm or hands, wavelike, posturing, jiggling	Blinking, grimacing, jerking
Rhythmic	Variable pattern, not rhythmic
Intermittent, repeated, prolonged	Quick, sudden, aimless
No pre-movement sensorimotor phenomena	Associated with a premonitory urge
Triggers: stress, excitement, boredom, fatigue	Increase with stress of excitement
Suppressible by external distraction	Suppressible—temporarily
Rarely responsive to medication	May respond to medications
Often enjoyable to the person	Often associated with distress or discomfort

(Mills & Hedderly, 2014)

There are a variety of types of tic disorders. Some of these involve just motor (movement) tics, some vocal tics, and some mixed. There are simple and complex tics. Simple tics, as the name suggests, are usually brief and more localized to specific areas such as the face, neck, or shoulder. Complex tics can involve elaborate sequences similar to compulsive behavior, persisting for varying lengths of time and with more purpose. In our experience, simple tics are not more common in people with Down syndrome than in other people. However, people with Down syndrome do seem to be at increased risk of having a more complicated type of tic disorder: Tourette syndrome.

Tourette Syndrome

Tourette syndrome (TS) is a hereditary, chronic neuromuscular condition consisting of motor and vocal tics. Tics are sudden, involuntary, brief, repetitive, stereotypic movements and vocalizations.

Examples of motor tics include jerking head movements, sudden movement of the extremities, facial twitches, and others. Examples of vocal tics include guttural sounds, yelling out, repeating a word or phrase, and others.

Tics begin in childhood and can change in location, number, frequency, and complexity over time. For a diagnosis of Tourette syndrome to be made, the tics need to begin before age eighteen, be present for at least one year, and not be due to a stimulant medication or a medical condition. In addition, people with classic TS always have at least two different motor tics as well as one vocal tic, but not necessarily concurrently. Attention-deficit disorders and/or obsessive-compulsive disorders often occur with Tourette syndrome.

We have seen a number of adults or adolescents with Down syndrome who have a Tourette-like disorder. We often label this as *Tourette-like* (or *atypical Tourette syndrome*) because the full complement of criteria is usually not present. (Most commonly there is an absence of vocal tics.)

Symptoms of Tourette Syndrome

Most of the people we have seen with both Down syndrome and Tourette syndrome have obsessive-compulsive symptoms that began in adolescence or adulthood. These patients have usually been diagnosed with attention-deficit disorder in childhood. Further questioning will lead to the discovery of the tics.

Motor Tics. Motor tics may include repetitious and at times sudden mouth, tongue, face, head, trunk, or limb movements. Tics appear fairly odd or bizarre— for instance, facial contortions, twirling, squatting, eyes deviated up or to the side, or sniffing at objects.

Vocal Tics. The people with Down syndrome plus TS that we have seen often do not have vocal tics in combination with motor tics. They sometimes have motor tics without vocal tics. However, we have seen a number of individuals with vocal tics, including the expression of words as well as such sounds as clicks, grunts, sniffs, hoots, snorts, coughs, throat clearing, and other types of mouth noises.

People without Down syndrome with Tourette syndrome occasionally have *coprolalia*, or the involuntary expression of obscenities or other inappropriate language. We have not observed coprolalia in people with Down syndrome. However, we have seen several people with Down syndrome who have vocal tics that include negative comments along with other vocalizations. These comments often resemble expressions of self-talk (see chapter 9), but they have a different feel. They appear to

be more sudden and spontaneous compared to regular self-talk, and it appears that these comments are not under the person's conscious control.

Waxing and Waning of Tics. In Tourette syndrome, tics change over time. A tic may persist for several months and then be replaced by a different tic. The intensity and frequency of tics change over time too. Sometimes, a tic may only occur several times an hour; at other times, it may occur dozens or even hundreds of times an hour. Often stress seems to increase the type, intensity, and frequency of tics, as in the example below.

> *We have treated Reggie, age thirty-three, for many years. Like many people with Tourette syndrome, he has an attention problem, obsessive-compulsive disorder, and both motor and vocal tics. Additionally, he is overweight and has an associated sleep apnea problem. He has lived in a number of residential settings that have been extremely stressful for him, primarily due to other residents' behavior problems.*
>
> *Reggie's Tourette symptoms have been greatly affected by the stress of his health and environmental difficulties. Under stress, his obsessive-compulsive behaviors are more incapacitating. He often gets "stuck" and refuses to move at transition points in his day. At these times, his motor and vocal tics also increase markedly. His motor tics include repetitious head and trunk movements. His vocal tics include frequent outbursts and expressions that often sound like self- criticism, as well as nonsensical comments. (Although these comments often resemble self-talk, they appear suddenly and are seemingly out of Reggie's control.)*
>
> *When his Tourette symptoms increase, Reggie moves less. As a result, his weight and sleeping problems also worsen. This in turn makes him more tired and sluggish in the daytime.*

Diagnosis of Tourette Syndrome

We have found that misdiagnosing the symptoms of Tourette syndrome may delay the effective treatment of the disorder in people with Down syndrome. This

often happens when motor and vocal tics are misdiagnosed as behavior problems, as in this example:

Manoj, age sixteen, was referred by his school. He attended a public school for children with disabilities who have behavioral and learning challenges because he had been diagnosed with an attention problem at a younger age.

The school had dealt with the attention problem through a structured curriculum, but they had recently noted a significant increase in compulsive behavior. Manoj had become more rigid in his routines and less capable of adapting to changes. His teacher and family also noted an increase in "odd" ritualistic behaviors such as bending his knees every five feet when he walked and repetitious movements of his arms and head. Most problematic to the school, he frequently uttered nonsensical words and other sounds that were becoming louder and more disturbing to his teacher and the other students.

We diagnosed Manoj with Tourette syndrome because of the presence of attention issues, obsessive-compulsive behaviors, and tics. For the school staff, it was most helpful to hear our opinion that both his "odd" behaviors tics and his expressions of words and other mouth sounds were tics. With this knowledge, staff and Manoj's parents began to see that these behaviors were not under his willful control. Consequently, the school reduced their insistence that he control his mouth noises. Interestingly, once teachers stopped pressuring Manoj to stop these sounds, the intensity and frequency of his vocalizations markedly decreased. Researchers on Tourette syndrome have reported similar results when caregivers understand the nature of tics as involuntary behaviors (Rosen, 2002).

The school followed up with an effective behavior plan to deal with Manoj's vocalizations when they became too disruptive. This included plenty of activities to divert his attention from the tics. Additionally,

when his vocalizations became a little too loud, he would be asked very quietly and diplomatically if he would prefer to leave the class to express himself more freely in a vacant room next door. Manoj sometimes took advantage of this option, but in general this was not necessary. He was also treated with an antidepressant to reduce the intensity of his compulsive behavior, which helped him to be more flexible with his daily schedule.

If you suspect that an adolescent or adult with Down syndrome may also have Tourette syndrome, the first step in diagnosis is to get an assessment of her history. Looking back to the person's childhood is important. As discussed above, it is common for individuals with Down syndrome and Tourette syndrome to have a previous diagnosis or symptoms of ADHD. Compulsive symptoms are also often seen, frequently beginning later than the attention-deficit hyperactivity symptoms (often starting in adolescence). Often the tics are the unrecognized symptom. They may have been overlooked in the past or thought to be side effects of medications. (Tics can increase with the stimulants frequently used for attention-deficit disorders.) The person's family doctor may be able to make the diagnosis and provide appropriate treatment, or a referral to a neurologist or psychiatrist may be necessary.

Treatment of Tourette Syndrome

Our treatment for Reggie, described above, has been multifaceted to meet his needs. He has responded positively to an antipsychotic medication that has helped to reduce his tics and the "stuck-ness" of his compulsive behaviors (see more on medication below). This, in turn, has helped him better manage his weight and to sleep better in the evening. We have also targeted his environmental stresses by advocating for him to move from several different problematic residences. These interventions have helped to get his symptoms under control and allowed him to get on with his life.

In general, the treatment for people with Down syndrome who have Tourette syndrome is the same as for people without Down syndrome who have Tourette syndrome. In our experience, people with a dual diagnosis of Down syndrome and Tourette syndrome respond best to a multifaceted treatment approach involving both behavioral interventions and medications.

Behavioral Strategies and Interventions for Tourette Syndrome

A number of chapters in the book deal with the obsessive-compulsive symptoms and the attention problems that often coexist with tics in people who have TS. This section will emphasize behavioral strategies for dealing with tics because they have not been discussed elsewhere.

In many respects, the behavioral strategies for dealing with tics and stereotypies are similar:

1. Stress may increase the occurrence of both conditions. Therefore, reducing the source of the stress may be helpful. (See above.)
2. Additionally, both conditions occur less often when people are engaged in more active pursuits such as sports or recreation activities.
3. We have also found it helpful to keep the body part that is most often engaged in the motor movement busy. For example, if the tic or stereotypic behavior involves the hands, it may help to engage the person in favorite hand activities, such as needlepoint, writing, drawing, video or computer games, etc. As an added benefit these activities are often stimulating yet relaxing and therefore may also help reduce stress.
4. Certain items may be used to keep hands busy when tics result in self-injurious behavior such as picking at skin or sores. These items may include the worry stones described above, or items with a sensory texture. These items may be chosen by the person herself. For example, we have seen small rubber hoses, paper bags, and small stuffed animals used. By helping to reduce the incidence of self-injury, these items may also help to reduce anxiety created by the response of others to the self-injury.
5. Finally, boredom and understimulation may increase the occurrence of tics. As suggested above in the section on stereotypies, it may be very important to find work that is interesting and mentally challenging. People who are productive and busy are generally happier and less stressed. As a result, they

may be less likely to display tics and stereotypic behaviors. At home, interesting mental activities such as educational computer games, word search puzzles, reading, or video games may help.

There is a major difference between tics and stereotypies, which may result in very different treatment and behavioral strategies. The primary difference is that stereotypies may be more under the person's voluntary control, while tics are not. Some people may be able to repress tics for short periods of time but not control them. For example, students may keep their tics suppressed or less obvious when in class but then may need to let loose with a barrage of tics after class. In contrast, stereotypies can often be managed by external distraction. This difference needs to be appreciated by family members and other caregivers.

Caregivers may use one of three strategies for dealing with tics based on how well informed and experienced they are regarding tics and Tourette disorder.

1. Those with less understanding may try to block or stop the person from engaging in the tic. This may work for a short period but will inevitably fail because of the involuntary nature of the tic. Not surprisingly, the anxiety this creates in the person with Down syndrome often results in more tics.
2. Those with more understanding may have some success by trying to redirect the tic, particularly if redirecting it to something that is of great interest to the person with Down syndrome. However, this strategy may also backfire, particularly if the person with Down syndrome experiences this as just another attempt to block the tic.
3. Those with the most understanding and experience often back off and let the tic run its course. This strategy often reduces stress on the person, which then helps reduce the incidence of the tics.

Managing the Effect of Tics and Tourette Syndrome on Others

Families may need to develop strategies for dealing with tics when they affect the relationships between people in the home and other key settings. This may occur when tics are annoying or disruptive to others, adversely affect the demands and expectations of the person with Down syndrome or are misinterpreted by others as

behavioral or attitude problems. Finally, we will discuss when to seek professional help for tics that interfere with daily life or become harmful.

Dealing with Disruptive or Annoying Tics

One issue that families need to sort out is how to deal with tics that are disruptive or annoying to family members. Often the best course for families is to educate their other children to be tolerant of the tics. It is also essential to be aware of the effect of tics on other family members, however, and to try to plan effective strategies to reduce the possibility of conflicts and tensions, as in the example below.

John, age twenty-two, has tics and Down syndrome. He had a job he loved, but he sometimes came home from work very stressed. As a result, he would have loud vocal tics that were disruptive to his two young sisters when they were trying to do their homework, read, or watch TV. His sisters responded to his tics by yelling at him to stop or complaining to their mother to make him stop. This only seemed to make the tics worse because John felt upset and persecuted by the complaints.

To deal with this problem, John's parents had a family meeting and proposed the following solutions. John's sisters would not yell at him or complain to his parents. Instead, they would ask him politely to go to his room, where he could relax and unwind by listening to his favorite music. This suited him fine. Additionally, John's parents helped to better insulate John's room. This allowed him to tic as loudly as he needed without disturbing others. In time, John was even able to remind himself to go to his room when he had a stressful day.

Not surprisingly, we have run across similar issues in residential settings. We often recommend similar meetings between residents of group homes to figure out how to deal with tics are irritating to others. In many cases, a resolution can be found that reduces annoyance to others as well as the stress experienced by the person with the tics.

Demands and Expectations

In our experience, families and other caregivers have to be careful not to let the person's tics affect the demands and expectations for her to behave normally in every other respect. For example, if a parent of a teen who has tics asks her to do her homework or to pick up her room, the presence of tics should not prevent this from happening. If it does, this may actually reinforce the expression of more tics or tic-like behavior. The teen may learn that expressing tics may let her off the hook for homework or other jobs she is expected to do. It may be a simple task to fake a tic "when needed." Excusing the person with tics from obligations may also create tension with siblings, who may feel their brother or sister is given special treatment. There may be other repercussions as well. For example, if the individual with tics is permitted to skip homework, she may experience anxiety at school when she has no homework to turn in.

In order to prevent tics from altering demands or expectations, we recommend the following strategies:

1. Discuss with the person with Down syndrome and TS that she has the same responsibility for completing chores or obligations as everyone else in the family, regardless of tics. Couch this in positive terms—for example, by stating that the tics should not affect her ability to develop her talents, skills, and independence.
2. It may help to explain that tasks may be temporarily delayed if a tic is disruptive, but only until the tic no longer interferes with task completion.
3. Be very careful to follow through with this expectation. If a tic is allowed to stop and not just delay the completion of a task, this will reinforce maladaptive patterns of responding to tics.
4. If a tic is not disruptive to the person or the task, then the person should be encouraged to complete a task whenever possible.
5. Bearing in mind that stress may increase the expression of tics, assess situations in which demands are made on the person with Down syndrome and TS. If the task is challenging but still in line with the person's ability, then you should encourage her to do the task, even if it results in a tic.
6. Provide assistance with a task only because it is too difficult and not because of the presence of a tic. As a test of this, ask yourself if you would be helping if the person did not have tics.

Similar problems with decreased demands or expectations may occur in settings other than the family home. Schools, worksites, and residential settings may also give the person with Down syndrome and Tourette syndrome special treatment or fewer responsibilities because of the tics. As in families, this may trigger resentment from others. It may also result in an underestimate and the possible underdevelopment of the person's skills and abilities based on a misperception or misunderstanding of tics and Tourette syndrome.

Misunderstanding of Tics and Tourette Syndrome

Sometimes families have to deal with a lack of understanding by people in different settings who may not be familiar with Tourette syndrome or tics. This can be very challenging, particularly if family members encounter service providers who assume the person's tics are actually the result of an attitude or an oppositional behavior problem. You can avoid or decrease this problem by educating caregivers on the nature of tics, as shown in the previous example of Stephanie (above).

In order to avoid this type of situation, we recommend the following:

1. Meet regularly with any staff or authority figures in the person's educational, work, or residential settings to educate them about the presence and nature of tics.
2. When needed, enlist the help of knowledgeable professionals to help educate staff who work with or serve as caregivers for your son or daughter.
3. Even if meetings are held with providers, problems may recur if there are changes in staff. Monitor these changes to avoid problems developing over an extended period of time.
4. If staff cannot or do not want to understand the nature of tics, then some changes should be made to avoid undue hardship and trauma for the person with Down syndrome. When Tourette syndrome is accurately diagnosed, it must be viewed like any other handicapping condition or disability. If caregivers do not accept this diagnosis and this has a negative effect on the person with Down syndrome, then the family has a right, and an obligation, to change either the offending person or the agency.

When to Seek Professional Help

Even the most experienced and effective families may need to seek professional help for their sons and daughters if tics and other symptoms result in physical harm or interfere in normal life activities. When this happens, the family may try to reduce stress, keep the person occupied with interesting activities, and try to patiently to wait for the tics to subside. Sometimes this may still not be enough to reduce a problem with tics. In these situations, medications may reduce the intensity and frequency of the tics to better allow the behavioral strategies to work.

Medications

Our behavioral interventions worked successfully for Manoj for close to one year. However, by the middle of his second year of high school, his tics were once again very disruptive in school and his compulsive behavior more and more debilitating. He was brought for our evaluation after he became physically aggressive when his teacher encouraged him to remove himself from the classroom. We discovered a new health problem that was then treated successfully. His vocal tics and his behavior continued to be problematic, however, so we prescribed a small dose of an antipsychotic medication. This helped to turn the corner on the problem.

At this point, Manoj is back in school. He still has some disruptive vocal tics and some inflexibility with his rituals and routines, but to a much lesser degree. We continue to follow his progress very closely to ensure that this positive progress is maintained.

We have found that the reason some individuals with Down syndrome do not respond well to medications for ADHD or OCD is that they actually have Tourette syndrome (or atypical Tourette syndrome). The usual treatment medications for the symptoms of ADHD and OCD may not work as well when those symptoms are part of Tourette syndrome.

Often, when these individuals were treated as children with the usual medications for attention-deficit disorders, their response was less than optimal. For example, Manoj responded to his structured school curriculum but did not show a

positive response to stimulant medication. His attention did not improve significantly, and his family also noted the presence of "odd" behaviors (most likely tics) with the use of the medication. Other parents have also reported that tics were first observed when their child took medications for attention-deficit disorder. And in fact, among people with Tourette syndrome who don't have Down syndrome, tics often seem to be triggered by stimulant medications used in the treatment of ADHD. Other caregivers have reported that tics were not observed until adolescence or adulthood, although it is quite possible that tic activity occurred at a younger age but was not observed and diagnosed as a tic.

We have also found that for many of the people we have seen with Tourette syndrome—including both Manoj and Reggie—obsessive-compulsive symptoms generally became more noticeable or problematic in adolescence or adulthood. Attempts to treat the obsessive-compulsive disorder have had mixed results for these individuals. Treatment with the usual antidepressant medications is often less than optimally successful. For example, an antidepressant medication temporarily helped to reduce the intensity of some of Manoj's compulsive behaviors, but it was not helpful for his tics and seemed to be less effective for his compulsions after approximately one year. Reggie also had a trial of an antidepressant medication for his compulsive behaviors, but it had no effect in reducing the problem symptoms.

Clonidine (Catapres) or guanfacine (Intuniv, Tenex) can help to reduce tics. These medications have an FDA indication for tics in children. Often, we have had the best success in treating Tourette syndrome in people with Down syndrome with an atypical antipsychotic medication. These medications are especially helpful in reducing the intensity of tics and of the more debilitating compulsive behaviors. We have used pimozide (Orap), risperdone (Risperdal), olanzapine (Zyprexa), quetiapine (Seroquel), and aripiprazole (Abilify) generally with good results. Only pimozide has FDA approval for treating Tourette syndrome in both adults and children and aripiprazole for children.

There are some newer atypical antipsychotics available, but we have not yet used them for Tourette syndrome. Side effects can be a limiting factor for the atypical antipsychotics. Weight gain and sedation are particular problems, and elevated blood sugar and type 2 diabetes mellitus can also occur. We recommend monitoring blood sugar on a periodic basis while taking these medications. Tardive dyskinesia, a movement disorder, is also a potential side effect. Chapters 16 and 20 have more information about the side effects of antipsychotic medications.

The Need for Long-Term Follow-Up

In people without Down syndrome, if the tics abate before adulthood, they generally do not recur. However, if they continue into adulthood, they tend to be persistent and recurrent. In our experience with adults with Down syndrome, symptoms tend to be persistent and recurrent. Often, some of the tics resolve, but they are generally replaced with others. We recommend ongoing monitoring of the symptoms. Most of our patients with Tourette syndrome have required ongoing treatment with medications.

Return to Table of Contents

Chapter 25

Autism

When Nicole was about four and a half, her parents increasingly noticed that she had developed a variety of differences compared to other children with Down syndrome. She had intense stereotypic and repetitive motor behaviors, including hand flapping, spinning, and rocking. Nicole also seemed uninterested in interacting with other children; instead, she was fascinated with inanimate objects such as strings and lights and preferred to play in rigid ways (e.g., lining up toys or other objects in fixed positions). In addition, Nicole seemed overly sensitive to hearing, touch, taste, and smell. Her parents also faced a host of behavioral challenges, including frequent tantrums and outbursts as well as some verbal and physical aggression.

Nicole's parents wondered if their daughter had autism and sought help from a local university medical center. However, the staff told them that autism did not coexist with Down syndrome. Nicole was instead diagnosed and started on medications for obsessive-compulsive disorder, impulse control disorder, and atypical psychosis. This treatment did little for her symptoms and behaviors and instead caused weight gain, agitation, and sedation. Fortunately, after several years, her parents found a center where Nicole was correctly diagnosed with autism spectrum disorder. This center connected the family with practitioners in the community who were experienced with autism. Nicole began receiving behavior therapy, which helped her family care for her more effectively and enabled her to benefit from a special program for autism in their school district.

When Nicole was fifteen, her parents brought her to us for an evaluation due to intense and often unmanageable outbursts and behavior at home and school. It appeared that the positive strategies they had used when she was younger were undermined by the developmental, physical,

and environmental changes and challenges in the teen years. These changes and challenges were creating a more extreme and unmanageable level of stress for Nicole and her family. We also diagnosed her with hypothyroidism and celiac disease, which were affecting both her physical and mental well-being. Additionally, she had sleeping difficulties, as well as bowel and bladder issues. She often exerted her need for control and independence by refusing to "go" as needed. This affected her ability to attend school and recreation programs that might have been a good fit for her needs.

After explaining to her family that there was no "magic bullet" to address all Nicole's difficulties, we recommended a multidisciplinary approach. We began by treating her coexisting health problems with medication and a gluten-free diet. In addition, we referred the family to an occupational therapist for help addressing Nicole's sensory sensitivities. We also recommended that her family work with the school to ensure that overstimulation was kept to a minimum (e.g., by placing her in a smaller class with less exposure to the noise and chaos of class changes) and to ensure that Nicole had a communication system that enabled her to better express her needs and wants. In particular, we suggested that the school and family teach Nicole visual strategies she could use to make sense of her world. (Research has shown that people on the autism spectrum respond extremely well to visual cues, such as the Picture Exchange Communication System [PECS], picture schedules, and video modeling.) Nicole's family found that she was very responsive to visual cues, and these cues were extremely helpful in teaching Nicole skills, enabling her to better manage day-to-day functions and frustrations and to communicate with others.

As Nicole grew into young adulthood, the pressures and stresses of adolescence subsided. She was then able to move into a good residential setting that met her needs and offered employment opportunities and an exceptional art program, for which she had great talent. Her family

continued to be very actively involved in supporting and advocating for her to get the best possible care and opportunities to show her artistic gifts.

What Is Autism?

Autism is a disorder that primarily causes problems in three domains:

1. significant difficulties in communication skills;
2. significant difficulties in social skills;
3. repetitive and ritualistic behaviors and interests (that is, seemingly odd or purposeless behaviors combined with an intense interest in relatively few topics or activities).

Autism, or autism spectrum disorder (ASD)—the official name in DSM-5—is a disorder that occurs on a spectrum that ranges from mild to severe. "Spectrum" refers to the range of findings and behaviors in people with autism. People on the milder end of the spectrum have fewer symptoms or symptoms that are less disabling, while people on the more severe end of the spectrum have more symptoms that are more debilitating.

Autism is not a mental illness. It does, however, usually contribute to emotional and behavioral problems that complicate life at home, school, and in the community, and these problems will not get better without appropriate treatment. For this reason, we have chosen to include autism in this volume. We will limit our discussion here because we have written about autism in teens and adults with Down syndrome in greater detail in the book When Down Syndrome and Autism Intersect (McGuire, Manghi, & Chicoine, 2013). For more information on the topic, we recommend consulting this book as well as the organization described at the end of this chapter.

Until recently, many professionals believed that autism could not coexist with Down syndrome (McGuire, Manghi, & Chicoine, 2013; Ghaziuddin, Tsai, & Ghaziuddin, 1992). It was assumed that these individuals had a more severe form of cognitive impairment rather than autism. As discussed below, there have also been questions about the diagnosis related to age at onset.

There is now greater recognition that some people with Down syndrome also have autism. This understanding is due in good part to the efforts of a number of

parents, advocates, and professionals who have worked tirelessly to educate others on the coexistence of this disorder with Down syndrome.

Because the recognition that Down syndrome and autism can coexist has only occurred relatively recently (and because some professionals still doubt that the dual diagnosis is possible), we have seen some adults with Down syndrome who have autism but were never diagnosed as children. The diagnosis of autism is often extremely important even for adolescents and adults. For families, the diagnosis gives an explanation for why their family member is different, and why he may have had a dramatic loss of function in childhood. There are other major benefits as well. State agencies often allocate additional funding for individuals with this diagnosis to receive special programs and services because of their greater needs. For example, funding may be available for behavior management and communication and social skills training, which may greatly reduce the more debilitating and isolating symptoms of autism.

Characteristics of People with Down Syndrome and ASD

Researchers have found that people with Down syndrome and autism show somewhat less impairment in social relatedness compared to people with only ASD (Lord et al., 2000). Still, most people with the dual diagnosis have major deficits in social skills, particularly in the areas of empathy and sensitivity to others. In this respect, they are more like others with ASD and less like people with Down syndrome alone, who are generally quite sensitive to others.

We have found, too, that people with Down syndrome and ASD tend to have more significant deficits in intellectual functioning and expressive language compared to those with just ASD. For example, most people with Down syndrome and ASD have intellectual disabilities, which is not the case for everyone whose sole diagnosis is ASD. Additionally, most people with Down syndrome and ASD have speech articulation problems in addition to the other communication problems, which again is not necessarily the case for individuals with just ASD.

Another difference between children who just have ASD and those who have Down syndrome and ASD is that those with Down syndrome and ASD tend to be diagnosed with autism later. According to the DSM-5, symptoms of autism must be present before the age of three for the diagnosis to be made. Parents of children with Down syndrome, however, often report observing symptoms of autism for the first time at around age five or six and even as late as seven or eight years of age.

Sometimes, this may be because parents and professionals assumed that the child's delays in communication and social skills were due to "just" the Down syndrome, and only later were the delays recognized as being more extreme than usual for a child with Down syndrome. Because of the comparatively late recognition of autistic symptoms in children with the dual diagnosis, there has been discussion as to whether this is autism or childhood disintegrative disorder (CDD), which has the same cluster of symptoms as ASD but is diagnosed later than three years of age and involves the child seemingly "losing" some skills (e.g., verbal skills) he had at an earlier age.

Does this mean that the children with Down syndrome do not have "classic" autism? Do they have childhood disintegrative disorder instead? There are no definitive answers to these questions. However, one thing to consider is that DSM-5 criteria are not always 100 percent applicable to people with Down syndrome. For example, we have had to adapt DSM-5 criteria to diagnose depression, anxiety, and psychotic disorders in people with Down syndrome. Perhaps we need to adapt the criteria for ASD to a later age of onset for individuals with Down syndrome to better fit their pattern of symptoms.

Practically speaking, as clinicians treating teens and adults, we are less concerned with age of onset and more concerned with how to help people get the treatment and services they need. We are also aware that the diagnosis of autism spectrum disorder is understood by far more people than is childhood disintegrative disorder and is thus more likely to lead to appropriate services and treatments. For these reasons, we diagnose adolescents and adults who have shown symptoms of autism since childhood as having autism spectrum disorder.

Symptoms of Autism Spectrum Disorders

As mentioned above, people with autism spectrum disorders have particular difficulty in three areas: (1) communication skills, (2) social skills, and (3) repetitive and ritualistic interests and behavior. Since people with Down syndrome who do not have autism also may have problems in these areas, it is important to understand what is and is not normal for someone with Down syndrome.

Impaired Communication Skills

People with autism spectrum disorders have impairments in both expressive and receptive language. That is, they have difficulties in expressing themselves to

others and in understanding what others are saying. People with Down syndrome and ASD also have problems with both expressive and receptive language, but they tend to have more problems with speech articulation than those with ASD alone do, making it more difficult for them to be understood when they are able to communicate. People who just have Down syndrome often have similar speech articulation difficulties, but they do not have the same problems with receptive language and are generally quite good at picking up social cues.

People who have autism spectrum disorders usually have difficulty understanding other people's thoughts and perspectives. Problems in this area can be particularly noticeable in a person with Down syndrome because many people with Down syndrome—even those with limited communication skills and significant intellectual impairments—have an innate ability to sense the feelings of others. This sensitivity to others is missing or at least deficient to some degree in people with Down syndrome and ASD. In other words, people who have a dual diagnosis have pronounced difficulties in social skills that they do not share with those who just have Down syndrome, regardless of the person's level of skill and functioning.

Often people with autism spectrum disorders have deficits in basic social skills. They may have difficulty maintaining eye contact. Many do not like to be touched or to be near others. They may feel unsure and even afraid in social situations. They may also be sensitive to sensory stimuli (see below). For example, they may have difficulty tolerating the sound of people talking in groups.

People with Down syndrome and ASD may have limited abilities to respond to parents or siblings in a caring and affectionate way. They may also have great difficulty interacting with peers and even more trouble establishing and maintaining friendships. In short, social exchanges are fraught with challenges and stress for many people with the dual diagnosis.

Restricted and Repetitive Interests/Behavior

Typically, people with Down syndrome and ASD have repetitive motor behaviors. For example, many flap their hands or make similar body movements that are commonly seen in people with autism. They may also repeat unusual vocalizations such as humming or make raspberry-like sounds. In addition, many are preoccupied with inanimate objects such as shoelaces, paper bags, and other items people do not normally play with. They may also play with toys in very restricted and compulsive ways such as by placing cars or other toys in perfect lines. Many people with Down

syndrome and ASD watch the same movies over and over or do certain tasks repetitively, such as putting their desk in order, opening and closing the doors in the house, or arranging furniture in idiosyncratic ways. Many also collect or save unusual items such as lint or specific paper products.

It is important to note that people with Down syndrome who don't have ASD may also have these kinds of repetitive movements or compulsive behaviors. However, they usually do not engage in these behaviors as intensely as people with Down syndrome and ASD. More importantly, they do not also have the significant problems with communication skills and social skills that individuals with the dual diagnosis have.

Other Symptoms

When Nicole transitioned from elementary school to middle school and then high school, she found the differences enormously stressful. Suddenly she was expected to change classes several times each day, and the overall setting was far larger and more complex. She came in contact with many more teachers and students, and the sensory overstimulation was very high and often intolerable for her. For example, the noises created by students walking, talking, and slamming lockers during the changes in class were extremely aversive. Added to this was her need to maintain more rigid and set patterns and routines in her life, and the changes were intolerable for her.

Sensory Issues. The ability to take in and organize sensory input may be impaired in people with Down syndrome and ASD. Difficulties in these areas may be of particular importance: (1) the ability to understand where the body is in relation to the environment; (2) balance control; and (3) tactile input through the skin. The person may be sensitive to the environment in ways that are difficult to understand. For example, while a gentle touch from someone may be reassuring to most people, it may be frightening to someone with an autism spectrum disorder. The person may stare at lights, crave spinning sensations, or show other unusual sensory responsiveness may be present. See chapter 12 on sensory issues for an in-depth discussion of this topic.

Behavioral Issues. People with a dual diagnosis of autism and Down syndrome may have self-injurious behavior, as discussed in chapter 23. There may also be increased anxiety, irritability, hyperactivity, attention problems, significant sleep disturbances not related to sleep apnea or another medical problem, compulsive behaviors and rituals, and difficulty with transitions.

Diagnosing Autism in an Adult with Down Syndrome

If autism is suspected in an adolescent or adult with Down syndrome, the diagnostic process is similar to diagnosis of any disorder. A complete and thorough evaluation requires that all other explanations of the person's behavior be assessed and excluded. For example, a complete physical exam should be conducted to rule out any health problems that may cause the behaviors. Extreme stress in the person's environment may also lead to behavior that is characteristic of autism. There may be other less obvious explanations as well, such as those related to the person's culture and language. For example, an English-speaking psychologist or physician may attribute social and communication difficulties to autism when an individual simply has problems with English comprehension (Manghi, 2013).

Having ruled out other possible explanations, the next step is for a trained and experienced professional or team of professionals to assess the presence of the three primary deficits of autism spectrum disorders. Optimally, families should try to locate professionals who have experience with people with autism, and preferably, the dual diagnosis of autism and Down syndrome. If you have a family member who displays the behaviors described in this chapter, local parent organizations serving people with ASD and Down syndrome may be able to provide names of local centers or practitioners who diagnose and treat autism.

One challenge in diagnosis is to obtain a detailed history of the childhood of the adult with Down syndrome who is suspected of having autism. If you are a parent bringing your adult child for an evaluation, your observations and experience with your child are essential to any diagnosis. Additionally, records or observations documented by teachers and other caregivers over the years may be very helpful. The same is true of tests or evaluations by medical or psychological professionals. These records often help to show a pattern of behavior that can be very helpful in diagnosing the disorder.

In addition, some families may want to bring in videotapes or family films showing the person's behavior. This may be particularly helpful if the person displays

behavior at specific times and locations, or if the behavior occurs intermittently or in response to certain stimuli such as those at shopping centers, social gatherings, etc. Films/videos may also be helpful if the person with Down syndrome tends to act normal in certain situations, such as when his parents want him to engage in a problem behavior for the doctor.

Treatment: Behavioral Approaches

Many volumes have been written on strategies for managing behavior and assisting the learning of individuals with autism spectrum disorders. We can only begin to touch on some of the methods that, in our experience, can be helpful for adolescents and adults with a dual diagnosis of autism and Down syndrome.

One broad way of helping to reduce the impact of the dual diagnosis on teens and adults is through the use of behavioral approaches. These strategies involve improving or shaping the person's behavior without the use of medication—for example, by teaching or modeling desired behavior or by adapting the environment to better support the individual's needs.

Applied Behavior Analysis

The "gold standard" behavioral approach for teaching people with autism new behaviors is applied behavior analysis (ABA). This approach has been extensively researched and found to be effective in teaching skills to people with all sorts of developmental disabilities. ABA practitioners use a variety of techniques to shape a person's behavior, all of which are rooted in the idea that rewards and consequences can be used to teach specific, measurable skills.

As the name suggests, applied behavior analysis involves *analyzing* a person's behavior and then *applying* the results of that study to help the person learn desirable behaviors. For example, to address an individual's challenging behavior, ABA practitioners often conduct a functional behavior assessment (FBA). To complete an FBA, behavior analysts first determine the antecedents of the behavior (what happens before it). They then carefully observe what happens during the problematic behavior. Finally, they examine the consequences of the behavior: What does the person "get" by engaging in the behavior? Why does he keep doing it? After this analysis is completed, the behavior analyst then moves on to choosing a more appropriate replacement behavior that would enable the person to achieve the same

results and then systematically teaches the person that behavior. Throughout the process, data are regularly collected on how the person's behavior changes or does not change to ensure that treatment strategies are effective.

Here is a brief example of how an FBA might work: Dillon has recently moved to a group residence. When housemates sit down near him at meals, he sometimes shoves them or pushes their plates forcefully away. (This is the behavior.) Upon observation, it appears that Dillon only shoves people when they are sitting less than two feet away from him. (This is the antecedent.) When Dillon shoves his housemates, they scoot their chairs farther away from Dillon, giving him more space. (This is the consequence.) Every time a housemate moves away from Dillon when shoved, it reinforces Dillon's shoving behavior (Dillon is getting what he wants: more space). In effect, he is being rewarded for the inappropriate behavior. To help Dillon learn not to shove people when they sit too close, the behavior analyst would teach him an alternative way to get his need for more space met, such as by using a picture card or an app on his tablet to request more space.

If you think ABA methods might be helpful in addressing the challenging behaviors of a teen or adult with a dual diagnosis of Down syndrome and autism, you can seek out the services of a board-certified behavior analyst.

Helping the Adult Deal with Sensory Issues

As mentioned above, people with autism may have unusual reactions to sights, sounds, smells, and other sensations in the environment. Appreciating this difference can be very important to understanding and helping the individual. A sensory integration assessment by a qualified occupational therapist (OT) can be very helpful in developing an understanding of the person's needs and in choosing useful strategies for dealing with these issues. For example, many individuals with sensory integration issues may benefit from a "sensory diet," which is a list of sensory-related tasks (such as using a weighted blanket or brushes for the skin).

Recommended strategies and activities may go a long way to make someone comfortable and increase his willingness to cooperate with daily living and learning tasks at home, work, or school. It may also be useful to consult with an OT about ways to adapt the environment to make it more palatable to the person with autism—for instance, by getting rid of lights that make a humming sound or figuring out what sounds are calming to him (see chapter 12).

Providing Structure and Consistency

Adults with Down syndrome and autism typically benefit from a structured environment. Routine is important. The structure helps them manage their day. They often respond better to pictures or visual cues than to spoken or written words. Picture calendars and schedules help them understand and appreciate what will be happening in their day. Without this understanding, the person may become more frustrated and irritable. You may find the book Activity Schedules for Children with Autism by Patricia Krantz and Lynn McClannahan useful in learning ways that picture schedules can help individuals with autism (Woodbine House, 2010).

Consistency in the way tasks are done is also beneficial and comforting. For example, if different family members or caregivers are teaching a given task, it may be helpful for each teacher to follow the same written task list to ensure consistency in instruction. It is also important to allow the person adequate time to process requests and to figure out what it will take to perform a task. Limiting other stimuli also encourages the person to focus on the task.

Sharing information with all people who regularly interact with the person is very important. This increases the likelihood that the environment will be responsive in a consistent manner. Here is an example of how one family ensured consistency:

> *Adam's parents wrote a short book about their son. They recorded a list of typical behaviors, what he is usually trying to communicate with the behavior, and how the family usually responds. The staff at his day program regularly reviews this information, and new staff members are asked to read the book before working with Adam. The use of this book is very beneficial for Adam, as he is happier and less frustrated when staff members are consistent. In addition, less staff time is spent dealing with challenging behaviors and more time is spent helping Adam and other people in the day program to learn and participate in the activities.*

Handling Transitions

Making transitions from one event or task to another can be very challenging for most people with autism spectrum disorders. It is very important for others to

appreciate and respect this characteristic. Providing warnings that it is going to be time soon to switch to a new activity helps with transitions. Then, explaining the transition to the person (with pictures, if necessary) as it occurs may make it less likely he will resist the transition.

Teaching about Social Situations

As mentioned above, people with autism usually have great difficulty putting themselves in another's shoes. We recommend helping adults with Down syndrome and autism improve their understanding of others' feelings by teaching them how these feelings relate to their experiences. Basic instruction on what to do and say in social situations is also beneficial. Many autism experts recommend using Social Stories (Gray, 2015) to help individuals with autism learn new social skills. Social Stories are custom made to help someone learn to handle a situation that is problematic for him. A parent, teacher, or other service provider writes a simple story describing the situation and showing how to appropriately deal with it. The story might then be illustrated with photos or drawings of the person engaged in the situation. The story is read to the person with autism before he encounters the situation and at other times when he can learn from the story.

Preventing Information Overload

Slow processing is another feature of autism spectrum disorder (and of Down syndrome). People whose sole diagnosis is autism may have problems understanding others because of receptive language limitations, difficulties picking up social cues, etc. People with Down syndrome and ASD have these difficulties, too, but they also have a slower processing speed related to their Down syndrome. Therefore, it is important to give the person an opportunity to process what has been said before giving him additional instruction or input. Presenting the material in a concrete form (especially in picture form) is beneficial. Waiting for an answer or giving the person a chance to answer at a later time helps prevent information overload. In addition, you can reduce information overload by limiting the number of people asking questions or giving instructions at once. Limiting the input to one person is generally best.

Teaching to the Individual's Strengths

Learning new information is often difficult for a person with autism. If the information is presented in a way that enables him to use his strengths, the learning process is much more likely to be successful. Use of abstract thinking, imagination, social intuition, interpretation, and rapid responding are not typically learning strengths of people with autism spectrum disorders. Learning will more likely be easier and more complete under these circumstances:

1. New information is presented ***visually*** (such as by showing the person how to brush his teeth).
2. The information is presented in as ***concrete*** a form as possible. For example, it is best to simply show the steps for brushing teeth and not to discuss the benefits of oral hygiene.
3. ***Hands-on*** learning is best. Information may be easier to absorb if the person can first observe and then do the task himself.
4. The information is broken down into ***sequential steps***. For example, toothbrushing could include such steps as grasping the toothbrush, putting on toothpaste, turning on the water, moving the toothbrush up and down, etc.
5. Even if there are multiple teachers, the ***order of the sequence of steps*** is not altered in the learning process. This is why it is useful to use pictures showing the sequence of steps.
6. ***Rote learning*** or repeating the task numerous times helps to ensure that the task is learned.

These methods of learning may be particularly strong in a person with an autism spectrum disorder, even beyond what might ordinarily be expected based on the person's other abilities. See chapter 6 for more information on visual supports.

Medications

People with autism spectrum disorders may have challenging behaviors, including aggressive or self-injurious behavior. If the behavioral approaches discussed above are not sufficient, medications may be necessary. The atypical antipsychotics can be beneficial in reducing aggression and irritability. Aripiprazole (Abilify) and risperidone (Risperdal) have an FDA approval for irritability in children (ages five to seventeen) with autism. While not FDA-approved for this use, antiseizure medications

may help with the behavior challenges. They can be particularly helpful with aggressive behavior (see chapter 22). Clonidine (Catapres) may also reduce agitation and aggression and help improve sleep.

Sometimes adults with autism have other mental illness diagnoses, so other medications as discussed in chapters 17–24 may also have a role in treatment. For example, when there is an accompanying mood disorder, antidepressants may be helpful. Likewise, antianxiety medications are also sometimes beneficial when anxiety symptoms complicate autism.

In-Home Behavior Support

Because life with a child or adult with a dual diagnosis of ASD and Down syndrome can be extremely challenging, caregivers have a critical need for support. The most successful group homes and worksites for adults who have the dual diagnosis have a high ratio of staff to residents. Equally important, employees in these settings have training in positive behavior management techniques and other useful strategies for helping people with ASD with daily living.

Without at least some outside assistance, families may be overwhelmed with the challenges of caring for a person with Down syndrome and autism. We have written a number of letters to state funding sources to secure in-home behavior support for families. We recommend that parents who have an adolescent or adult with the dual diagnosis find a professional in their community who is willing to do the same for them. This may include the physician, psychologist, or other professionals who were involved in the diagnosis of your son or daughter, or similar professionals with whom you are in contact. The following is an example of one such letter we wrote for Tony, a fourteen-year-old boy with ASD and Down syndrome:

> *Tony's behavior challenges are so severe that, if left unsupported, his family would be severely taxed and possibly at risk for adverse effects to their own health and well-being. At home, Tony is extremely demanding of his parents' time and attention. This is limiting to them but also detrimental to his three siblings, who are 9, 11, and 15 years of age. At home, he is frequently not cooperative with activities, which then diverts attention away from the others. Perhaps more frustrating to his*

siblings is that he will also not cooperate with normal and typical family activities outside the home, such as outings to a sibling's sports event, church, evenings out to a restaurant, etc. He often reacts to these outings by dropping on the ground and refusing to move. If his parents attempt to force him to move, he may escalate his behavior, which then compounds the problem.

Of importance, Tony is usually more cooperative with teachers and experienced respite staff than with his own parents. In this way, he is no different than any other teenager. Unfortunately, unlike most teenagers who revel in their freedom, Tony is extremely resistant to leaving his own house. Thus, at a time when most children become less demanding of parental time and attention, he is even more demanding, while still possessing the uncooperative attitude of a teenager.

In-home behavior support would have a number of critically important benefits for Tony and his family. A trained in-home behavior analyst would help to build on Tony's compulsive tendencies to help establish a functional routine for the completion of daily living tasks. Additionally, important skills such as safety training could be reinforced in the home environment. This would free his parents from having to micromanage his behavior, while giving him independence and a sense of pride. Equally important, in-home behavior support would allow his family to return to a more normal pattern of family life. For example, if Tony refused to go out to a community function with his family, he could stay with his trainer. The trainer would also be able to accompany Tony when they do go on outings. It would also be extremely beneficial to Tony's parents and siblings to have more quality time with Tony, especially if they are not responsible for his care 24 hours per day.

States may have funding available for in-home behavior support, although the amount of support varies. More information can be obtained by contacting your state

human services department, autism organizations, your local chapter of ARC, and Down syndrome groups.

Given enough time and contact, staff who provide in-home behavior support can successfully implement picture schedules and behavior management programs and assist with general caregiving tasks. These support people are usually trained by agencies and centers to serve the needs of individuals with autism. Usually a psychologist or behavioral analyst will develop behavioral management strategies, which are then used by the in-home support staff. This professional may also provide ongoing consultation to support staff to help better tailor the programs to meet the person's needs.

Finding Support for Your Family

Support groups for families who have children with autism spectrum disorders may be helpful to you even if your child is now an adult. No one quite knows and understands the challenges of living with someone with autism like another family who is facing the same challenges.

Of course, not all local organizations geared to parents of children with autism are equipped to offer support and useful information to families of teens or adults with Down syndrome and autism. The members of some autism groups may be completely unfamiliar with the dual diagnosis or may focus their efforts on helping only those whose sole diagnosis is autism. In this case, you will probably want to find an organization outside your local area that can help. Fortunately, there is now an organization specifically for people with the dual diagnosis: the Down Syndrome Autism Connection, whose motto is "Providing education, support, and inspiration for the journey."

The Down Syndrome-Autism Connection is an important resource for families of children and adults with both Down syndrome and autism or for those who suspect a dual diagnosis. The group is composed of parents and professionals in Denver who maintain a highly informative and up-to-date website: https://ds-asd-connection.org/. The group offers long-distance support via phone calls, emails, and monthly online chats and offers workshops specific to the dual diagnosis during the National Down Syndrome Congress's annual national conference. In addition, the Down Syndrome-Autism Connection is developing a national database of resources.

If at all possible, we recommend at least one visit to a doctor or clinic with expertise in treating people with Down syndrome and autism. The Global Down

Syndrome Foundation lists clinics serving the needs of children and adults in the United States: https://www.globaldownsyndrome.org/research-medical-care/medical-care-providers/. Staff at many of these clinics may have expertise in both Down syndrome and autism. We recommend contacting the clinic to learn about their specific services. Additionally, there are major medical centers around the country that specialize in evaluating autism, and they would most likely be able to diagnose autism for a person with Down syndrome. These centers can often be located by referring to the Association of University Centers on Disabilities (AUCD) at the following website: https://www.aucd.org.

Conclusion

Autism spectrum disorders begin in childhood. However, there are many benefits to assessing and treating adults with Down syndrome who have not been previously diagnosed with autism. It can also be very useful for families of teenagers and adults with Down syndrome who were diagnosed with autism in childhood to consult experts on the dual diagnosis when the person faces new challenges or experiences stresses that lead to behavioral changes. Understanding the unique issues that autism spectrum disorder presents can significantly affect the care of someone who has both Down syndrome and ASD. Behavioral approaches, medications, and occupational and other therapies should all be considered.

Return to Table of Contents

Chapter 26

Alzheimer's Disease and Decline in Skills

Ken, age fifty-two, came to us for an evaluation due to concerns about his memory. He was talking more about events of long ago and people who had died years before. In addition, he was having difficulties remembering his schedule, something he had always been able to do well. One year prior, Ken had had his first seizure ever. His family reported that one to two years before this seizure, Ken had seemed to become less precise in completing his tasks and in doing them in the order he had for years. He seemed to be "losing his groove," per his family. More recently they had observed he was having more trouble with multistep tasks. For example, once when he went upstairs to brush his teeth, get his shoes, and put on his watch, his family found him standing in the bathroom (without his shoes or watch), holding his toothbrush in one hand and toothpaste in another but confused as to what he should do next.

The pattern of Ken's symptoms was consistent with Alzheimer's disease, and thorough medical and psychological evaluations did not find some other, reversible cause. Ken was diagnosed with Alzheimer's disease, and we discussed his prognosis, anticipated course, and treatment options with him and his family.

Alzheimer's disease is one of the most commonly diagnosed and misdiagnosed mental disorders in adults with Down syndrome. Alzheimer's disease is more common in people with Down syndrome and occurs at a younger age than in people without Down syndrome. However, diagnosing Alzheimer's disease in people with Down syndrome is often not clear-cut, and testing is less precise. Because of this higher incidence and less precise testing, any decline in skills is often assumed to be due to Alzheimer's disease.

Unfortunately, without thoroughly assessing for a variety of mental and physical health conditions that can cause a person with Down syndrome to lose skills,

a potentially reversible or treatable condition may be missed. For example, Caroline was initially misdiagnosed with Alzheimer's disease at age twenty-four due to a reduced ability to do her daily activities, memory loss, and behavioral changes. Our evaluation, however, uncovered that she had sleep apnea, and her abilities returned to normal with appropriate treatment. Miles, age seventeen, was also misdiagnosed when he developed similar symptoms after his brother went away to college and Miles began to anticipate high school graduation. In our evaluation, we diagnosed depression, and after Miles received counseling and treatment with an antidepressant, he returned to his previous level of function. Since decline in skills and/or Alzheimer's disease can greatly complicate the care of any adult, including one with Down syndrome, thorough assessment is very important to optimize diagnosis and treatment.

What Is Alzheimer's Disease?

Alzheimer's disease is a progressively degenerative neurological condition that affects the brain. It is a form of dementia. Dementia has many causes, some of them reversible and some not reversible (at least at this time). It is associated with changes in memory and personality and impaired reasoning. In Alzheimer's disease, there is progressive destruction of brain cells, especially in certain parts of the brain. People with Alzheimer's disease experience increasing impairment of memory, cognitive skills, and daily living skills, as well as psychological changes. There is presently no cure for Alzheimer's disease or known treatment to slow its course. However, there are a variety of ways to lessen or improve some symptoms.

Alzheimer's disease is characterized by plaques and tangles in the brain. The plaques (hard, flat patches) consist of amyloid protein that builds up between neurons (nerve cells). The tangles develop from accumulation of tau protein, which plays a role in the health of microtubules in brain cells (neurons). The microtubules are a normal part of the nerve cell, whose function is to transport nutrients and other substances in the cell. In Alzheimer's disease, tau protein accumulates abnormally. Clumps are formed, and then cell dysfunction and ultimately cell death occurs.

Research is ongoing to unravel the underlying cause(s) of these changes, exactly what mechanism they play in Alzheimer's disease, and how, or if, reversing them can treat or potentially prevent Alzheimer's disease symptoms. The most precise way to study these changes is by examining a piece of brain tissue (generally only done after a person's death) under a microscope. However, as the disease

progresses, computerized tomography (CT) or magnetic resonance imaging (MRI) scans of a living person's brain can depict the destruction of many cells because the brain begins to atrophy or shrink. Positron-emission tomography (PET) scans, a nuclear medicine scan, are also now being used to study changes in the brain.

The cause of Alzheimer's disease remains unclear, and the reason it is more common in people with Down syndrome is not definitely known. However, the reason for the increased incidence is believed to be related to the presence of the extra chromosome. Due to the extra 21st chromosome in people with Down syndrome, some genes are "triplicated" (three instead of the usual two). The amyloid precursor protein (APP) gene is one of these triplicated genes. As noted above, amyloid appears to play a role in Alzheimer's disease, and there is compelling evidence that increased gene dose for APP (and its products) is related to Alzheimer's disease in Down syndrome (Prasher et al., 1998; Korbel et al., 2009; Doran et al., 2017). Other chromosome 21 genes may also play a role in Alzheimer's disease in Down syndrome, as may genes on other chromosomes, including variants that also increase the probability of Alzheimer's disease. However, a uniquely important role is thought to be played by the increased dose for APP (Wiseman et al., 2015; Ballard et al., 2016).

How Common Is Alzheimer's Disease?

In individuals without Down syndrome, the incidence of Alzheimer's disease is increasing as the population ages. The overall incidence of Alzheimer's disease among people in the United States is estimated to be 4 percent for those under sixty-five, 15 percent for those between sixty-five and seventy-four, and 44 percent for those seventy-five to eighty-four. Interestingly, the rate declines a bit, to about 38 percent, for those eighty-five years and older. Presumably, a large percentage of those who will get Alzheimer's have developed it before reaching age eighty-five, and most of those who survive to eighty-five without getting it are those who will never get it.

Nearly all people with Down syndrome develop the physical changes of Alzheimer's disease in their brain (plaques and tangles) by the age of forty, and all do so by the age of sixty (Wisniewski et al., 1985; Wiseman et al., 2015; Ballard et al., 2016). Despite the prevalence of these brain changes, however, not all adults with Down syndrome appear to develop symptoms of the disorder. Still, the incidence is higher than in people without Down syndrome, and the rate increases with age. Between the ages of 35 and 49, an estimated 9 to 23 percent of individuals with Down syndrome develop symptomatic Alzheimer's disease; between 50 and 59 years, the

incidence is 55 percent, and estimates place it as greater than 75 percent in those sixty years of age and above (Mann et al., 1984; Coppus et al., 2006; McCarron et al., 2014; Strydom et al., 2018). Some researchers did find a lower prevalence in those under forty years of age.

Not all the older adults we have evaluated have gone on to develop the symptoms of Alzheimer's disease. If those individuals without Alzheimer's disease had lived longer, would they eventually have developed Alzheimer's disease? Some of the data from the studies above suggest that 100 percent of individuals with Down syndrome above age seventy years develop Alzheimer's disease.

To date there is no information that demonstrates a lower rate of Alzheimer's disease in people with mosaic Down syndrome, although that would be understandable if it were found to be the case. The oldest person we ever evaluated died at age eighty-three, had mosaic Down syndrome, and reportedly had no symptoms of memory impairment, decline in skills, or seizures.

In unpublished data from the Adult Down Syndrome Center, we have found the cause of death in our patients to support these concerns about the higher incidence of Alzheimer's disease with age. As noted in the table, Alzheimer's disease was the most common primary cause or significant contributing cause to death in our older patients and became more common with age. We should note, however, that the data may be higher at our site than in the general Down syndrome community because we see many people who come to us specifically to assess for a decline in skills and Alzheimer's disease.

Age Range	**# in Age Range Who Died**	**Leading cause or contributing factor**	**Alzheimer's Disease N (%)**
40-49	23	Alzheimer's Disease	11 (48%)
50–59	76	Alzheimer's Disease	58 (76%)
60+	37	Alzheimer's Disease	34 (92%)

Leading cause or contributing factor to death at the Adult Down Syndrome Center (unpublished data). The percentage noted in the column on the far right is the percentage of individuals in our practice who died in that decade from Alzheimer's disease. (It is not the percentage of people in that decade of life in our practice who died.)

Unfortunately, it appears that Alzheimer's disease not only occurs more frequently in people with Down syndrome but also at a younger age. One group of researchers reported that the average age at which the clinical development of Alzheimer's disease is diagnosed in people with Down syndrome is fifty-five (Strydom et al., 2018). A review of individuals evaluated at the Adult Down Syndrome Center found a slightly younger average age of diagnosis of 52.5 years. The youngest person with Down syndrome we diagnosed with Alzheimer's disease was in his midthirties when his symptoms developed. However, that finding is an outlier, and we have rarely seen individuals whose symptoms developed before age forty. As discussed below, it is important to remember that all people with Down syndrome deserve an evaluation for other causes of a decline in mental abilities rather than assuming it is due to Alzheimer's disease.

After a thorough evaluation, if the diagnosis of irreversible dementia is made, one question is whether this is Alzheimer's disease or another type. Although nonreversible dementia in people with Down syndrome is usually labeled Alzheimer's based on years of research, there are a couple of additional considerations. Some researchers have questioned whether any of the dementia seen in people with Down syndrome is actually Alzheimer's disease. Could it be something similar but not exactly the same? In addition, there are other known types of dementia including Lewy body dementia, Parkinson's disease dementia, and others. Much less is known about these conditions in people with Down syndrome, as very little Down syndrome–specific research has been done to date on any type of dementia other than Alzheimer's disease. In the future, hopefully more study will reveal additional information about these other conditions as well as the link between Down syndrome, dementia, and Alzheimer's disease.

Diagnosing the Cause of Declining Skills

Since many other health problems can cause dementia, it is imperative to evaluate for these other conditions before making the diagnosis of Alzheimer's disease. Unfortunately, this is not always done for people with Down syndrome. One of the concerns expressed by the parents who asked that we start a clinic for adults with Down syndrome was that their sons or daughters were not being given an appropriate workup when a decline in skills was noted.

There is no specific test that definitely diagnoses Alzheimer's disease. The diagnosis is made by assessing for and finding a pattern of decline in neurological and

psychological function consistent with Alzheimer's disease and assessing for and ruling out other possible causes. This diagnostic process is similar for adults with and without Down syndrome, but much of the cognitive/psychological testing for people without Down syndrome is not helpful in diagnosing Alzheimer's disease in people with Down syndrome.

When an individual with Down syndrome is assessed for a decline in abilities, a thorough medical and psychological evaluation is recommended. We recommend assessing for a variety of conditions, particularly those that are more common in adults with Down syndrome (see chapter 2).

Conditions to Rule Out

To rule out causes of cognitive decline other than Alzheimer's disease, we consider the following:

- depression and other psychological concerns
- sleep apnea
- thyroid disease
- vitamin B-12 deficiency
- metabolic diseases such as kidney disease, diabetes, or calcium abnormalities
- celiac disease
- loss of hearing or vision
- atlantoaxial instability or other cervical (neck) problems
- heart disease
- seizure disorder
- normal pressure hydrocephalus
- medication side effect

Additional possible causes that we have not seen in our patients include:

- syphilis and
- Acquired Immune Deficiency Syndrome (AIDS).

Another consideration is chronic, undiagnosed pain. Adults with Down syndrome sometimes have a global decline in function in response to pain and illnesses that do

not directly cause a loss of function. This appears to be an emotional, psychological, or perhaps neurological response to the trauma of pain or illness.

In addition, as discussed in chapter 11, people with Down syndrome seem to age more rapidly than others, so that when they are fifty-five, on average, we consider them to be more like someone without Down syndrome at age seventy-five. It is important to remember that there may be aging changes at a younger age in a person with Down syndrome. We have seen a number of patients who were slowing down because of age and age-associated health issues. Often these factors were not being considered or addressed, and the changes were attributed to behavioral challenges or dementia. Addressing them from an aging perspective put the changes in a whole new light.

Tests for Decline in Function

The tests we recommend for all our adults with Down syndrome who are experiencing a decline in function include

- CBC (complete blood count),
- electrolyte panel including serum calcium,
- thyroid blood tests,
- serum Vitamin B12 level, and
- vision and hearing testing.

Additional tests that may be indicated based on the findings on the history, physical, and lab tests include

- lateral cervical spine x-ray in flexion, extension, and neutral positions,
- liver function tests,
- RPR (for syphilis),
- HIV testing (for AIDS),
- CT scan or MRI of the brain,
- blood testing for celiac disease (anti-tissue transglutaminase antibody or anti-endomysial Ig A and anti-gliadin IgG and IgA),
- EEG, and
- sleep study.

Neuropsychological testing is part of the evaluation for Alzheimer's disease for people who do not have an intellectual disability. However, this testing is more difficult in people with Down syndrome and other intellectual disabilities. The underlying intellectual disability makes the tests used in people without Down syndrome less accurate when used for people with Down syndrome. There are, however, a few tests (see below) that are more specific for people with an intellectual disability. These seem to be most beneficial when done sequentially over time. However, clinically, we find that by the time a cognitive decline is evident on the testing, the decline and diagnosis are often clear from the person's behavior. In our experience, referring our patients for these tests often does not add clinical (diagnostic) benefit. We are able to get similar information by asking parents and other caregivers to update us on symptoms, particularly over time.

There is an excellent but somewhat lengthy questionnaire available through the National Task Group (NTG) on Intellectual Disabilities and Dementia Practices, which is affiliated with the American Academy of Developmental Medicine and Dentistry (https://www.the-ntg.org/ntg-edsd). At the Adult Down Syndrome Center we have built some of the NTG screening into the series of questions we ask at our annual evaluations.

A variety of tests have been developed to assess people with Down syndrome or other intellectual disabilities. These include The *Down Syndrome-Mental Status Examination* (DS-MSE) (Haxby, 1989); The *Dementia Questionnaire for Mentally Retarded Persons* (Evenhuis et al., 1990), now updated to The *Dementia Questionnaire for People with Learning Disabilities* (Evenhuis et al., 2009); *Dementia Scale for Down's Syndrome* (Huxley et al., 2000); The *Dementia Scale for Down Syndrome* (Gedye, 2000); The *Severe Impairment Battery* (SIB) (Fish, 2011); and The *Vineland Adaptive Behavior Scales*, Third Edition (Vineland-3) (Burger-Caplan et al., 2018).

These tests may be an aid to diagnosis for trained mental health or medical professionals, as they help point out key areas that need to be considered when ruling out other causes. However, these tests should not be used as stand-alone measures in the diagnosis of Alzheimer's disease. There is currently no definitive test to make the diagnosis. The diagnosis is still based on a process of excluding other possible causes of the person's skill loss. The tests mentioned can be used to help establish the changes associated with dementia as part of a total assessment including a thorough physical exam, information gathered from caregivers regarding skill and memory loss, environmental and developmental stresses, and lab evaluation.

Symptoms of Alzheimer's Disease in Adults with Down Syndrome

Symptoms of Alzheimer's disease in people with Down syndrome include the following:

- **Memory Impairment.** In early Alzheimer's disease, short-term memory loss is primarily affected, while memories of events and people from the distant past are preserved. In late AD, however, both short- and long-term memory are impaired. Many individuals we have treated for Alzheimer's begin talking frequently about people who are now deceased; they seem to be able to remember them better than people and events that are current, so they talk about those from the past. Sometimes this appears to be a symptom of depression, but most often it seems to be associated with memory impairment and missing those individuals from the past (because that is who they are able to remember best).
- **Decline in Skills.** These include cognitive skills such reading and doing math and the ability to complete activities of daily living such as tooth brushing, showering, etc.. Often the first sign of skill loss is the need for more prompts. Early on, the person may still have the ability but need more guidance or direction. Losing the ability to do multistep tasks is also often an early symptom.
- **Incontinence.** This can affect urine, stool, or both.
- **Gait Disturbance** (gait apraxia). We often seen poor balance, leaning to one side (later this occurs even when sitting), and falling. Ultimately, the loss of the ability to walk is very common as the disease progresses. We have cared for a number of individuals who stopped walking before they actually lost the ability to walk after they seemed to develop a fear of walking and falling. Many people with Down syndrome have weaknesses in balance and depth perception throughout their lives, and as Alzheimer's disease progresses, they experience further difficulties with these abilities.
- **Personality or Psychological Changes**, including
    - depressed mood,
    - aggressiveness,
    - paranoia,
    - loss of interest in activities,

    - compulsiveness,
    - changes in "the groove." (Many of the individuals with Down syndrome who developed Alzheimer's disease we have assessed experienced either an increase or decrease in their groove as part of the symptoms. Sometimes these are subtle early changes that are only recognized as early symptoms in retrospect.)
- **Seizures**. This may be a new onset of seizures or a change in type or pattern (e.g., increased frequency) of seizures.
- **Swallowing Dysfunction.** This may manifest is a variety of ways, including choking, gagging, or coughing with eating or drinking; fear of eating, apparently due to a sense that swallowing abilities are changing; and aspiration pneumonia due to breathing saliva or food/liquid into the lungs. Unfortunately, in our experience, loss of swallowing function is often a major contributor to death in individuals with Down syndrome and Alzheimer's disease.
- **Sleep Changes.** These may include day-night reversal, daytime fatigue, and disrupted nighttime sleeping.
- **Altered Appetite and Thirst.** Most commonly, there is a decrease in eating and drinking. Many people with Down syndrome lose weight, at least partially due to decreased appetite and eating. Some, however, actually gain weight in the early stages, at least partially because their appetite is still good but their activity level has significantly dropped.

Most of the symptoms above are similar to those seen in people with Alzheimer's who don't have Down syndrome. However, there are some differences. It has been observed and reported that the first symptoms that appear in all people with Down syndrome who develop Alzheimer's disease consist of personality, psychological, or behavioral changes. In our experience, these are commonly, but not always, the first changes. Older individuals who develop these changes should be monitored for other symptoms of Alzheimer's disease but should also be evaluated for other (especially those that are potentially reversible) causes. In addition, seizures tend to occur much more frequently and at an earlier stage in Alzheimer's disease in people with Down syndrome. Recurrent, uncontrolled seizures can lead to a more rapid decline. People with Down syndrome are also more likely to lose the ability to walk earlier in the disease and to have earlier difficulty with swallowing and with aspiration.

Particularly earlier in the disease, the functioning level of a person with Alzheimer's disease often fluctuates. These fluctuations may occur over several days or weeks, from day to day, and even within minutes or moments. A skill may come and go over these periods of time. As the disease progresses, the person's skill level declines, and periods of better functioning will be shorter and not as functional as before. This fluctuation of skills can be challenging for families and caregivers.

A frequent question about skill fluctuation is "How do I know if she is just pulling my leg or the disease is just fluctuating that day?" Unfortunately, it can be very difficult to tell. Behavioral and personality changes are part of the disease. You should not be surprised if the person is unwilling to do a task that she can still do, but then neither should you be surprised if she has a neurological inability to do the task. Either way, neurologically or psychologically (behaviorally), these changes are part of the disease and very difficult to differentiate. In the end, the disease, not the individual, is the cause, and the approach to handling behavioral changes is the same, as discussed later in the chapter. Certainly, as the disease progresses, skill loss becomes much more pronounced.

Treatment of Alzheimer's Disease

There is no treatment presently available to cure Alzheimer's disease. Various medications and other treatments, however, may be prescribed in an attempt to address symptoms.

Exercise and Diet

In individuals without Down syndrome, there is limited evidence that regular physical exercise and a healthy diet (fruits, vegetables, whole grains, lean meats, fish, seeds, and nuts) may be helpful in either preventing or slowing down Alzheimer's disease. These studies have not been done in people with Down syndrome. Furthermore, different studies have come to different conclusions. Since regular physical exercise and healthy diets have implications for health in a variety of ways, we encourage these health habits for all people with Down syndrome.

Medications for Alzheimer's Disease

There are a number of FDA-approved medications for Alzheimer's disease. While they don't cure or slow Alzheimer's, researchers found that they assist in improving function, although only temporarily, in adults without Down syndrome. Unfortunately, studies assessing these medications in people with Down syndrome have not demonstrated benefit. The authors of a Cochrane review (Livingstone et al., 2015) concluded that they could not be certain that any of these medications were effective. However, the authors did indicate that the quality of the available research on these medications for people with Down syndrome was low and further research was recommended. In our experience and in discussion with other clinicians, there are differing opinions on the benefit of these medications, but the overriding sense is that these medications provide little, if any benefit, to adults with Down syndrome. In addition, there is significant potential for side effects.

In individuals without Down syndrome, two classes of medications have been studied and received FDA approval for use in people with Alzheimer's disease.

Cholinesterase Inhibitors

Cholinesterase inhibitors are medications that slow down the breakdown of choline (in the brain and in the peripheral nervous system) and have been shown to improve the function of people (without Down syndrome) with Alzheimer's disease. Nerve cells communicate with each other via neurotransmitters (chemicals) that pass from cell to cell. One of these transmitters, choline, is the chemical used for communication by many of the cells that are destroyed in Alzheimer's disease. Medications that slow down the rate of breakdown of choline prolong the ability of the choline to transmit messages to the next cell. Blocking cholinesterase, the chemical that breaks down choline, does this. This improves the function of the cells, and thus the function of the person with Alzheimer's disease. Unfortunately, this improvement is temporary, and the effectiveness of the medications decrease as more cells are destroyed and fewer cells are sending and receiving signals via choline. The medications available include donepezil (Aricept), galantamine (Razadyne), and rivastigmine (Exelon).

All three are available as tablets. Donepezil is also available as a disintegrating tablet placed under the tongue; galantamine is also available as a liquid; and rivastigmine is also available as an oral liquid and a patch that is placed on the skin.

Side effects to watch for include gastrointestinal upset and decreased appetite. Many people with Alzheimer's disease require assistance and encouragement to consume enough calories and appropriate nutrition anyway. If they develop these side effects, it may be even more difficult to maintain good nutrition. In addition, seizures, although not a common side effect, can occur as well. If the person is taking one of the medications and develops seizures, the question arises whether the seizures are secondary to the medication or the Alzheimer's disease. Seizures, as noted above, are more common in people with Down syndrome who develop Alzheimer's disease, and unfortunately, there is no way to determine which is the cause. A decision must then be made as to whether the benefit of the medication exceeds the downside that it could be causing seizures.

Memantine (Namenda)

Memantine (Namenda) belongs to a class of drugs called N-methyl-D-aspartate (NMDA) receptor antagonists that appear to work by slowing calcium influx into cells and nerve damage. Memantine is the only medication in this category approved for Alzheimer's disease. As for cholinesterase inhibitors, there no data have shown benefits in people with Down syndrome and Alzheimer's disease, although some clinicians do find it beneficial. The medication is generally fairly well tolerated, but side effects may include sedation and, conversely, agitation or aggression. After discussing the potential lack of benefit and significant side effects, families will often decide against trying the medication or if they do decide on trying it, will request stopping if the side effects occur.

Treatment of Associated Seizures

The seizures seen in Alzheimer's disease may be tonic-clonic (grand mal) or other types. In addition, myoclonic jerks (intermittent jerking movements) are often seen. Phenytoin (Dilantin), carbamazepine (Tegretol), valproic acid (Depakote), gabapentin (Neurontin), levetiracetam (Keppra), topiramate (Topamax), and other antiseizure medications can be effective, depending on the type of seizure. Unfortunately, antiseizure medications can cause drowsiness and increased confusion. We have found this to be particularly true of phenytoin (Dilantin). Levetiracetam has is quite effective in most people that use it, with limited side effects, but in some it causes drowsiness, and in others, significant agitation or even aggression.

Since seizures occur in about 75 percent of people with Down syndrome who develop Alzheimer's disease, we are sometimes asked whether it is appropriate to start antiseizure medications before they occur. We wait until seizures develop before starting antiseizure medications, in light of the potential side effects and the fact that not all people develop seizures. It is important, however, to bring seizures under control as soon as possible. Uncontrolled seizures seem to contribute to a more rapid rate of decline.

Treatment of Psychological, Personality, and Behavioral Changes

Psychological, personality, and behavioral changes are common in Alzheimer's disease, and this seems to be particularly true in people with Down syndrome who develop Alzheimer's disease. Changes may include sleep problems, depression, anxiety, agitation, compulsiveness, paranoia, hallucinations, and others. These symptoms can often be reduced through behavioral management. Sometimes medications are also beneficial. Further information regarding specific medications for specific symptoms is provided below.

One key issue for treating people with Alzheimer's disease is to limit the negative impact of medications. People with Alzheimer's disease are often more susceptible to side effects such as sedation, increased confusion, and further loss of skills such as walking or swallowing. Therefore, careful monitoring of the medications and the benefits and negative effects is important. In addition, smaller doses, less frequent dosing, and shorter duration of use may be effective while at the same time limiting side effects.

Caring for Someone with Alzheimer's Disease and Down Syndrome

There are a number of general helpful hints to consider when caring for or interacting with a person with Down syndrome and Alzheimer's disease. While there is not a cure or a highly effective treatment for Alzheimer's disease in people with Down syndrome (as noted above), there are still many steps you can take to help the person and her caregivers cope with the changes that Alzheimer's disease brings.

1. Provide the Best Quality of Life

We recommend that family members or other caregivers follow these general guidelines:

- Emphasize doing what can still be done (focus on the skills present and not on those lost).
- Encourage interaction with friends and family to the point that the person finds it enjoyable and comfortable.
- Encourage physical exercise.
- Encourage healthy eating.
- Work to maintain the ideal body weight, as too little weight and too much weight both can contribute to decreased functioning.
- Encourage social engagement.
- Involve the person in creative arts such as painting, sculpture, singing, and theater.
- Provide sensory stimulation at the right level, as too little or too much could contribute to agitation.
- Consider the "Bingo Pace"—a phrase we have coined to consider the level and pace of the activity. Sometimes activities that are too fast or too intense can cause confusion. Sometimes a slower rate, such as one might experience while playing Bingo, is better. The key is not what the rate is overall but matching the rate to the needs of the individual. This rate will be unique for the individual and will likely fluctuate from day to day and at different times of the day.
- Consider music. The organization Music and Memory (https://musicandmemory.org) recommends using music to reawaken memories and increase enjoyment of life in people with Alzheimer's disease. The organization does so by uploading a playlist of music the individual likes to a portable device (iPod) the person can listen to. The playlist is unique to the individual and includes songs the person enjoyed prior to the development of Alzheimer's disease. While we are aware of limited use for people with Down syndrome and Alzheimer's, we suspect benefit will be seen when used more widely, particularly because music is so important to many people with Down syndrome.

2. Use Effective Communication Strategies

When communicating with someone with Down syndrome and Alzheimer's disease, consider using the following strategies:

- Eliminate background noise.
- Talk one-on-one.
- Make direct eye contact (be at the level of the person; for example, if the person is in a chair, stoop down so your eyes are at the same level as the individual's eyes.
- Don't correct or argue.
- Take time to listen to the person's words and tone.
- Validate feelings.
- Slow down your words and actions.
- Use short, simple sentences.
- Supplement verbal communication with cues and gestures.
- Break down tasks you are asking the person to do into simple steps.
- Use gentle touch to supplement verbal communication.
- Limit choices. When communicating a choice or asking the person to do a task, too many choices can be confusing. Some families have reported success with taking all but two (or even sometimes just one) choices of clothing for the day out of the closet. This allows the person to participate in picking the outfit but reduces the strain of too many choices.

3. Consider Your Approach to Challenging Behaviors

Is there a challenging or change in behavior? Consider possible causes and then take them into account in handling the behavior:

- **Caregiver triggers:** Caregivers, even with the best intentions, can be a trigger to a behavioral change. For example, reasoning with the person who has a declining ability to reason often results in little if any success in the interaction and can cause upset behavior. Short of an immediate safety issue (the person is doing or about to do something dangerous), in most situations, backing off, waiting a few minutes, and trying again is more likely to be successful.

- **Physical triggers:** Pain is a common contributor to challenging behaviors. The person may have a bladder infection, a sore throat, or other causes for pain. It is important to assess for possible painful conditions that may be contributing to a change in behavior.
- **Environmental triggers:** Environmental factors like too much noise can be stressful and result in a change in behavior. Going to a new environment can also be challenging. Are we expecting that the individual can successfully participate in a gathering at a new setting in the presence of fifty relatives she hasn't seen for years? These events and even much less hectic events or settings can be challenging. For individuals who don't live with family, sometimes even going back to the family home is too overwhelming. Their present home/residence is now familiar, and the family home—no matter how long the person lived there—may no longer be "familiar" due to memory loss and visiting the home may become too stressful.

Addressing things in the environment that the individual didn't find stressful prior to developing Alzheimer's disease and adding some changes to the environment to make it more manageable can be very helpful. A good resource is available from Down's Syndrome Scotland: *Living with Dementia: A Families' and Carers' Guide*. (See the references for the link.)

Treatment of Other Disorders

Adults who develop Alzheimer's disease may also develop a variety of other disorders that can affect their mood, behavior, and well-being as well as complicate their treatment. These disorders are all discussed in more detail elsewhere in the book, but the sections below discuss considerations specific to adults with Down syndrome who also have Alzheimer's disease.

Obsessive-Compulsive Disorder

Some degree of compulsiveness is common in people with Down syndrome. However, the development of Alzheimer's disease may increase (or sometimes decrease) this behavior. Some adults we have treated developed obsessive-compulsive disorder that was, in retrospect, the earliest sign of Alzheimer's disease. Chapter 10 discusses how to assist people with compulsive tendencies, but in short, helping

someone use these tendencies in a positive manner is likely to be more effective than trying to use behavioral techniques to eliminate this tendency.

If the problem does not respond to behavioral approaches and affects the person's ability to participate in daily life, we generally recommend the use of medications. We have found the selective serotonin reuptake inhibitors and others to work well. Medications for obsessive-compulsive disorder are discussed in chapter 19.

Depression

Depression is common in people with Alzheimer's disease. Depression can be seen independent of Alzheimer's disease, it can mimic Alzheimer's disease (which is why checking for depression is part of the diagnostic process), and it can be part of the symptoms of Alzheimer's disease. Supportive treatment is essential for a person with depression, whether Alzheimer's disease is part of the depression or not. Offering reassurances, listening to concerns, and encouraging participation in activities are some of the many ways to support a person with depression.

Medications are also necessary sometimes. Use of medications for depression is discussed in chapter 17. Treatment with medications for depression related to Alzheimer's disease is similar for depression not associated with Alzheimer's disease. There are a few particular concerns regarding bupropion (Wellbutrin) and the older antidepressants. There is a theoretical risk of seizures (which are already a concern in Alzheimer's disease) with the use of bupropion, so we generally avoid prescribing it for individuals with Alzheimer's disease.

The older antidepressants such as amitriptyline (Elavil), desipramine (Norpramin), and others are probably effective as well. However, we also tend to avoid these medications because of their greater incidence of anticholinergic side effects. People with Down syndrome seem to be more sensitive to these side effects even when they do not have Alzheimer's disease. There is also the concern that blocking the effect of choline, such as these medications do, will result in a greater decline in skills since choline does play a role in Alzheimer's symptoms.

Sleep Disturbance

Many people with Alzheimer's disease have sleep disturbances. Often, they are confused with regards to typical day-night sleep cycles. The person sleeps during the day and is awake at night. This may not be harmful if the person can get adequate

sleep, just at a different time. If the environment can allow for this pattern, then it is reasonable not to intervene.

There are several reasons to consider intervention, however. Safety is often the most important reason. If care providers sleep at night, the person with Alzheimer's disease is not as well supervised if she is awake at night. In addition, stimulating activities are often only available during the day. Therefore, even if safety could be maintained at night, the person would have no activity to participate in during her wake time, which can lead to further decline. In addition, the person who is up at night may be very disruptive to others who sleep at night. Continued sleep deprivation can also be very stressful for caregivers.

Interventions for sleep changes can include both nonmedicinal and medicinal treatments. We have outlined our nonmedicinal recommendations in the "Sleep Hygiene" section in chapter 2. When these recommendations are unsuccessful, additional measures are available. We have had some success with the natural product melatonin. We generally recommend starting with 2 mg and increasing to 4 mg in a few weeks if 2 mg is not adequate. Sometimes we recommend up to 6 mg. There is a combination melatonin product as well. It has both an immediate release and a delayed release effect and can help some individuals fall and stay asleep. There are several other over-the-counter agents, but many of them contain diphenhydramine (Benadryl), which has anticholinergic side effects. As noted above, anticholinergic side effects can, particularly in someone with Alzheimer's disease, include confusion. Therefore, we don't recommend these products. Magnesium, taken at bedtime, in various forms (magnesium oxide, magnesium sulfate, and others) can also improve sleep. Before an adult begins taking magnesium for sleep, we recommend checking kidney function and a magnesium level to avoid developing a level of magnesium that is too high.

Prescription medications such as zaleplon (Sonata), eszopiclone (Lunesta), or zolpidem (Ambien) can be effective, as can a short-acting benzodiazepine such as oxazepam (Serax). However, all these medications can contribute to daytime drowsiness, confusion, and unsteady walking and should be used with caution. We have also found trazodone (Desyrel) to be helpful. It is an antidepressant that we don't find works well as an antidepressant in people with Down syndrome but can be effective as a sleep aid when used at night.

Anxiety

Anxiety can be part of the psychological decline in Alzheimer's disease. Some anxiety may stem from a direct neurological impairment. We suspect that some of it may result from the person's fear of the inability to understand what is happening to her as she declines. Often, anxiety seems to occur during earlier stages, which would seem to go along with the latter idea. It can be very disconcerting to sense that you are losing skills but not be able to understand why. Ways to reduce anxiety include the following:

- Provide reassurances (gentle verbal reassurance, encouraging and helping the person do the task that she is having difficulty with, etc.).
- Help the person find tasks she can successfully complete.
- Provide written or picture cues that help the person find her way or do things. (We find pictures work best.)
- Remove reminders of things that she can no longer do. (For example, if it frustrates her that she can't cook her own meals anymore, removing the microwave may reduce anxiety.)
- Do not argue with her when she is recalling something incorrectly (unless there is a safety issue involved).

Medications can also be used. A number of antidepressants, discussed above, can also help with anxiety, as can a shorter-acting benzodiazepine. We have used alprazolam (Xanax) and lorazepam (Ativan) with success. We generally prescribe very small doses and use them less frequently than generally recommended. Real care must be taken when prescribing these medications for a person with Alzheimer's disease. Sedation, unsteady gait, depressed mood, and increased confusion are common side effects.

We have generally found that there is a relatively short period of time (weeks to a few months) that the anxiety requires medications, although some of our patients had anxiety for a longer period. We recommend careful observation for side effects and discontinuing the medications if side effects occur. In addition, wean the medication as soon as possible, as anxiety symptoms decrease.

Agitated Behavior

Agitated behavior is another problem that people with Alzheimer's disease can experience. When it occurs, careful assessment is important. An evaluation for medical problems and physical sources of pain may find a cause that is not directly related to Alzheimer's disease. Because of the person's reduced ability to understand or inform others of her discomfort, she may be using behavioral changes to communicate. In addition, depression, increasing obsessive tendencies or compulsivity, anxiety, and sleep disturbance may cause agitated behavior. Treatment for the appropriate condition may reduce or eliminate the agitated behavior. Sometimes, however, no other underlying cause is found.

Sometimes agitated behavior can endanger the person with Alzheimer's disease or other individuals. In addition, it may be associated with hallucinatory behavior or paranoia. If this is disturbing to the person or is a safety issue, medications can be beneficial. We have found the atypical antipsychotics helpful. Risperidone (Risperdal), olanzapine (Zyprexa), ziprasidone (Geodon), aripiprazole (Abilify), quetiapine (Seroquel), and others can all reduce symptoms. But we have also seen unsteadiness and increased sedation, confusion, and incontinence in patients taking these medications. However, we start with very tiny doses—e.g., risperidone (Risperdal) 0.125 mg at bedtime—which reduces the incidence of side effects. More information about medications for psychoses can be found in chapter 21.

Among people without Down syndrome who have Alzheimer's disease, there appears to be an increased incidence of stroke when they take these medications. These medications are not approved for use in dementia-related psychoses, and there is a "Black Box Warning" against their use in people with dementia. Vascular disease, which can lead to strokes, is less common in people with Down syndrome, so this theoretically could be less of a concern in people with Down syndrome and Alzheimer's disease, but no studies have been done to assess this risk in people with Down syndrome. Out of an abundance of caution, we therefore generally avoid using atypical antipsychotics in people with Down syndrome and Alzheimer's disease. But at times, based on symptoms and quality-of-life concerns related to the symptoms, we consider their use after discussion between the patient (if able), family, and health care provider, as long as there is understanding and agreement on taking the potential risk.

Hallucinatory behavior and paranoia can also occur without agitated behavior. If this is a significant problem for the person, treatment as described above can be

beneficial. Again, it is important to weigh the symptoms, the effect on quality of life, and the risks of the medications.

Keeping Activities at the Right Level

Another aspect of caring for a person who has developed Alzheimer's disease is maximizing her level of function. We recommend encouraging her to participate in activities that are of the appropriate cognitive level. Engaging her in activities that are not too easy and not too difficult will help maintain a higher level of function for a longer period of time. Tasks that are too difficult will be frustrating and can lead to a more rapid loss of skills, as well as emotional changes, stress-related behaviors, and unhappiness. Similarly, tasks that are too easy will not allow the person to use the skills that she has and will lead to greater erosion of skills.

Assessing the appropriate skill level of tasks can be difficult, particularly if the person's skill level fluctuates. What was appropriate yesterday may not be appropriate today, but it may again be appropriate tomorrow. As discussed earlier in this chapter, this fluctuation in abilities can be a challenge for caregivers, both from an assessment standpoint and from an emotional standpoint. Caregivers can begin to "take it personally" when the person with Alzheimer's disease can't do a task that she could do just recently. They may feel that the person with Alzheimer's disease is not trying, is being lazy, or is not doing it to spite them. Although the caregiver may have previously helped the person with Down syndrome develop new skills and greater independence, this emphasis on improving skills is no longer appropriate when Alzheimer's disease is diagnosed. The focus must instead shift to maintaining skills or limiting the decline in skills.

The Right Environment

In our experience, it is usually best for the person with Alzheimer's disease to remain in a familiar environment. A change in environment can be confusing, require learning new skills, and be emotionally upsetting. Compare a change in environment to changing the furniture in the house of a person with severe vision impairment. It requires new learning in order to function in that setting. With the declining intellect of a person with Alzheimer's disease, this can be difficult. Instead, the environment should be adjusted as the person's skills decline. (Adjust the environment to meet the needs of the person, not the person to meet the needs of the environment.)

The flexibility of the environment is crucial to optimizing the care of the person with Alzheimer's disease. When she first begins to lose skills, she may continue to do well in the same setting. As her skills further decline, however, her adaptability will too. Often, she will be most comfortable (or perhaps only comfortable) at home. Going to work may become too stressful, especially as Alzheimer's disease progresses. If the person lives somewhere where leaving the building and going to work is a required part of the schedule, this may become a significant problem. A flexible schedule is most helpful to someone with Alzheimer's disease who is declining. Ideally, each day the person's level of function should be assessed to determine whether she would benefit more from working or staying home. An alternative program or activities should be available on days when it is apparent that she would best be served by staying home.

Safety issues may also develop. Loss of judgment related to appliances, hot water, and other potential household hazards can lead to potentially serious accidents. In addition, as the person's walking skills decline, stairs and other obstacles can become safety hazards. Wandering is another potential safety issue that may need to be addressed with alarms on doors, the person's bed, and other sites. Assessing the safety of the environment is critical. A home safety inspection by an occupational therapist can be of significant benefit.

Besides assessing how the environment affects the person with Alzheimer's disease, it is necessary to assess how the person with Alzheimer's disease affects the environment. For example, how does she affect the other people who are living with her? For a person of "normal" intelligence, the stress of caring for, or just living with, a person with Alzheimer's disease can be substantial. While we have seen adults with Down syndrome and other intellectual disabilities rise to the occasion when someone they live with develops Alzheimer's disease, we have also seen it become an overwhelming stress. One group of three women who lived with a woman with Down syndrome who developed Alzheimer's disease initially "blossomed" with regards to their own caregiver skills. However, later they found the situation too much to handle, and a different living situation was arranged.

Whenever possible, we encourage letting roommates or housemates with intellectual disabilities try to provide some assistance for the person with Alzheimer's disease. Many people with Down syndrome or other intellectual disabilities get "done for" their whole lives with little opportunity to "do for." Assisting someone with Alzheimer's disease can provide a real boost to self-esteem for the care provider as well as lead to the development of new skills.

However, sometimes the stress of something that seems relatively minor can create problematic tension in the house. This might occur, for example, when the person with Alzheimer's disease is no longer expected to participate in life skills classes, go to work, or follow the daily schedule. The sense of "injustice" can create emotional or behavioral problems for the others. At other times, people with Alzheimer's disease may recurrently yell or talk loudly, have irregular sleep patterns that disturb others' sleep, or need changes in the environment that are stressful for the others. Roommates of individuals with Alzheimer's disease have made comments along these lines: "She doesn't like me anymore"; "He won't do his chores"; and "She yells at me." Clearly, people with Down syndrome and other intellectual disabilities who are living with a person with Down syndrome and Alzheimer's disease are affected by the changes. Sometimes the changes in the person with Alzheimer's disease just become too great a stress. All these issues may create a situation where reassessing the environment becomes necessary.

A Change in Environment

If an environment does not allow for the adult with Alzheimer's disease to stay home during the day when she needs to, this can result in a great deal of stress for her. The continuous expectation to do tasks that are too difficult or too stressful can lead to emotional, behavioral, and cognitive changes. If she feels overwhelmed by expectations, the person might give up and seem to have fewer skills than she actually has. Moving to an environment that allows for the necessary flexibility can be of significant benefit. This benefit often outweighs the negative impact a move to a new residence can have.

Moving may also be advisable if safety issues cannot be resolved. The presence of stairs or potentially dangerous household appliances or the inability to assure that the person does not wander away can all be significant safety issues that may not be correctable in the present living situation.

Finally, it can be best for the person with Alzheimer's disease to move to another residence if her caregivers or the people she lives with are overwhelmed by the situation and appropriate in-home assistance is not available. This can be necessary both for someone who lives at home with her family or someone who lives in a residential facility.

We have participated in a number of successful, appropriate moves to different residences for individuals with Down syndrome and Alzheimer's disease. Nursing

homes, particularly if they offer specialized care for people with Alzheimer's disease, can be appropriate. Some agencies that serve people with disabilities also have residences for "seniors" that are able to provide appropriate care. Moving back home with family (if the person is living in a residential facility) has also worked for some. Generally, however, this requires some additional in-home assistance.

Duration of Alzheimer's Disease

The duration of Alzheimer's disease in adults with Down syndrome is not clear. In those without Down syndrome, the course of Alzheimer's disease is thought to be approximately ten to twelve years. Particularly in people with Down syndrome who have a higher degree of functioning before the onset of Alzheimer's disease, an overall course of ten or so years might be expected. However, our experience suggests that the duration is shorter for many, particularly in those who have a lower level of functioning prior to the onset of Alzheimer's disease. In a sense, the further the person has to fall cognitively, the longer it generally takes. We have seen people live one year from the time of diagnosis to the time of death. On average, though, the time from the development of symptoms to death is usually in the five- to six-year range but has been much longer for some. Note that these time frames are referring to the time that symptoms are noted. It is known that the brain changes in Alzheimer's disease begin a number of years before the individual develops symptoms.

Again, the development of seizures (particularly if they are difficult to control) seems to accelerate the decline in some people. Losing the ability to walk and the complications seen when this skill is lost also seem to increase the rate of decline. The onset of swallowing dysfunction, when associated with recurrent aspiration pneumonias, is the symptom that most commonly contributes to death.

Future Considerations

At present, a great deal of research is addressing the issue of Alzheimer's disease, both for people with and without Down syndrome. People with Down syndrome receive particular attention when it comes to Alzheimer's disease because studies have found that all people with Down syndrome develop the neuropathologic changes that are seen in Alzheimer's disease by the age of sixty. Eventually, the

findings in people with Down syndrome may be important keys in unlocking the mysteries of Alzheimer's disease for all people.

Return to Table of Contents

Chapter 27

Regression

When Jade, age thirty, came to see us for her first evaluation, she initially appeared to have autism. She made little or no eye contact with our staff and made many repetitive motions. She did not speak during the assessment and reportedly had no verbal skills. However, we soon learned that for many years, she had been a typically developing person with Down syndrome. As a child and teenager, she had had good verbal and self-care skills and only mild intellectual disabilities, and she had attended school and participated in a variety of activities.

Jade's mother reported that her daughter had suddenly regressed after being treated with an antibiotic for a cold at age seventeen. She had undergone extensive testing over the past thirteen years, including blood tests, imaging, and a sleep study. She had been diagnosed variously with depression, anxiety, late-onset autism, early Alzheimer's disease, and psychosis. She had been treated with different psychotropic medications and supplements. Although Jade's cognitive skills had not declined further, she had made little to no improvement.

Evan was twenty-three years old when he was evaluated. He had been given prednisone to treat a rash that had not responded to prescription lotions and oral antihistamines, and shortly thereafter, he began showing symptoms of depression. He had been a very social person, and his interactions with others rapidly decreased. His sleep was altered, and then his appetite diminished. Within a few weeks, he was not eating. His verbal skills deteriorated, and he could no longer take care of his own hygiene, clean his room, and go to work; all areas he was quite skilled and independent in previously. He was started on an antidepressant, but his symptoms worsened. He required hospitalization because he became

dehydrated. There it was noted that Evan seemed "rigid," "not moving," and even "frozen," and he was diagnosed with catatonia. Treatment options were discussed, and Evan and his family selected electroconvulsive therapy (ECT). After the first treatment, he had remarkable improvement and after several treatments, he was back to his baseline. He was again taking care of his own needs, working, and enjoying life.

For decades, health professionals with a special interest in people with Down syndrome have noted cases in which teens or young adults with Down syndrome experienced a puzzling decline in abilities. These individuals, usually in their twenties or younger, suddenly lost speech, cognitive, and daily living skills and often had behavioral or psychological changes. This "regression" has been given several names, the underlying diagnosis has varied, and response to treatment has been mixed.

As is evident from reading the two case summaries above, much has been learned about these changes in individuals with Down syndrome. Jade was first evaluated many years ago before regression in young people with Down syndrome was a regular topic of conversation at meetings among Down syndrome specialists. Evan was evaluated and treated more recently, when there was a better appreciation for the condition. However, there is still much that is unknown and there are differences in the understanding of the cause and treatment, and even different names for this cognitive decline. Is "regression" a specific condition from a specific cause or is it a large category that can have many causes from a variety of mental and physical health conditions? A variety of physical and mental health conditions can cause people with Down syndrome to have further cognitive impairment. Is regression a specific diagnosis or a description of symptoms that can be attributed to many of those causes? Answers to those questions are all part of the ongoing work to further understand these changes.

As you will note below, the condition has been given several names. For the purpose of the chapter, we will call it "regression syndrome" or "adult regression syndrome."

What Research Has Found

Reports of regression in young people with Down syndrome go at least as far back as the 1940s, when a researcher (Rollin, 1946) described the development of catatonic psychoses in twenty-eight of seventy-three individuals with Down syndrome living in a residential facility. Many years later, another researcher (Prasher, 2002) observed regression over one to two years followed by a plateau in functioning in young adults with Down syndrome and reported on them in his letter titled *Young Adults with a Disintegrative Syndrome* (YADS).

Recently, there have been more clinical case reports of regression in adolescents and adults with Down syndrome who have shown unexpected and severe regression in cognitive and adaptive functioning, motor function, communication skills, and behavior. This regression is reported to occur following a period of stable functional skill acquisition in young adolescents or adults as described by their families.

In 2015, one group of researchers (Worley et al.) described a new onset of autistic-like regression they labeled "Down Syndrome Disintegrative Disorder." That same year, other researchers (Ghaziuddin, Nassiri, & Miles) described four individuals with Down syndrome and regression along with motor disturbances such as slowing or increased motor activity who were diagnosed with catatonia.

One researcher (Akahoshi, 2012) described a mean age of onset of 21.2 years of age. Another researcher (Mircher, 2017) found an average age of onset of 18 years in girls and 21 years in boys among the thirty individuals he identified. However, the youngest individual was eleven and the oldest was thirty when symptoms were noted.

What Regression Is Not

Both autism and Alzheimer's disease are relatively common in adults with Down syndrome and can cause behaviors and loss of skills similar to those seen in regression syndrome. However, there are also important differences that have led to the current conclusion that regression syndrome is not caused by either autism or Alzheimer's disease. Most importantly, autism and Alzheimer's disease each have a characteristic age of onset and timeline that are distinguishable from the regression syndrome. To determine whether one of these conditions is contributing to a decline in skills in a person with Down syndrome, careful history-taking and diagnostic assessment for autism and Alzheimer's disease are needed to determine the age of onset, preexisting

level of function, and pattern of change over time. Ruling out other contributing conditions or illnesses via medical assessment is also necessary.

One group of researchers (Worley et al., 2015) did describe individuals with regression syndrome as having autism or an autistic-like condition. However, these individuals are "too old" to be diagnosed with childhood autism. This is true even though childhood autism has a later onset in children with Down syndrome than usual. (ICD-10 criteria specify onset by age three.) Even late-onset autism or childhood disintegrative disorder, as referred to by Worley and his colleagues, begin earlier in life than the regression syndrome. Those criteria include onset by ten years of age for late-onset autism, and by age seven years for childhood disintegrative disorder in those without Down syndrome.

On the other end of the age spectrum is Alzheimer's disease. Among adults with Down syndrome, the average age of onset has been reported as 54.7 years (Tyrrell et al., 2001) and 54.2 years (Lai & Williams, 1989). Symptoms of Alzheimer's disease are unlikely to occur in individuals with Down syndrome younger than 35 to 40 years (Moran et al., 2013). The onset of adult regression syndrome in the teens and early twenties is much earlier than the age of onset for Alzheimer's disease. In addition, Alzheimer's disease is marked by progressive impairment in cognitive, adaptive, and motor function. In contrast, regression syndrome often can plateau (sometimes for many years) after a rapid onset of symptoms. Also, Alzheimer's disease is often accompanied by seizures in people with Down syndrome (Zigman & Lott, 2007; Head, Powell, Gold, & Schmitt, 2014). These characteristics are significantly different from the symptoms of regression syndrome.

Due to these differences and the very young age that some individuals develop symptoms of regression syndrome, there is at least some agreement that Alzheimer's disease should be excluded as the cause of regression syndrome. For example, Akahoshi et al. (2012) concluded that regression syndrome has features and a clinical course that is different from those seen in typical Alzheimer's disease in people with Down syndrome.

What Is Known about Regression Syndrome?

Although researchers and clinicians now generally agree that regression syndrome is not due to autism or Alzheimer's disease, there is still much that is unknown or unclear. For example, in some cases, regression in adults with Down syndrome is due to depression, sleep apnea, or another treatable condition—should

those individuals be considered to have the syndrome until they have recovered normal function? Should only those who are more challenging to treat and who never fully recover be considered to have the syndrome? Do the individuals with severe regression in some way form a unique group, or do the behaviors and other changes in regression syndrome occur along a continuum similar to the way symptoms of autism occur along a spectrum (Devenney & Matthews, 2011)?

There is some disagreement as to the answer to those questions. For this chapter, however, we include anyone with Down syndrome who is regressing for a variety of reasons (except for those with Alzheimer's disease or autism) and has a certain pattern of symptoms (see "Clinical Features" below). We will explore the differential diagnosis—how other diagnoses are ruled out—and give special attention to those for whom the diagnosis is less clear or who are more severely affected and provide a greater treatment challenge. Many conditions that cause similar symptoms that should be considered during differential diagnosis often don't include the breadth of symptoms described (although they can). Therefore, those with regression but without the breadth of symptoms are excluded from the diagnosis of regression syndrome. Instead, they could be described as having some regression as part of their primary diagnosed condition (for example, depression accompanied by regression).

Further study is obviously needed to improve our understanding of diagnosis and treatment of individuals with Down syndrome who experience a decline in skills. What is clear, however, is that the brain/cognitive function of people with Down syndrome can be "fragile" and susceptible to further impairment for a variety of reasons. It is also clear that regression syndrome includes symptoms that could be caused by a number of conditions.

Features of Regression Syndrome

There is not agreement, yet, on the name of condition, nor is there agreement on the definition and clinical features—the signs/symptoms that an outside observer or health care provider would be able to note. However, the Down Syndrome Medical Interest Group-USA (DSMIG-USA) working group is developing a working definition to provide a framework for case finding and further study.
The discussions by this group have led to this operational definition:

Core features:

- Cognitive and executive dysfunction (executive dysfunction refers to difficulties with the brain function that helps us manage time, pay attention, switch focus, plan and organize, and perform other higher thought processes)
- social withdrawal
- loss of acquired skills
- loss of functional use of language
- duration of more than three months (see comment below)

Exclusions:

- not autism (although people with Down syndrome and autism can also develop regression)
- not Alzheimer's disease

Variable features:

- maladaptive behavior (new onset or change in behavior)
- psychiatric symptoms (including depression, compulsive behaviors, and others)
- failure to acquire new skills
- inattention-disorganization (new onset or change in behavior)
- motor changes (slowing or increased movements)
- vegetative symptoms (appetite/weight loss, incontinence, sleep pattern disturbance)

Demographics:

- typically, between ages fifteen and thirty years
- equally common in males and females

Based on the consensus of experienced clinicians, all five core features are very common in regression syndrome: cognitive-executive dysfunction, social withdrawal, loss of acquired skills, and loss of functional use of language. Some or all the other variable features are often present as well. Initially, a duration of more than three months was considered necessary for the diagnosis, but more recently it has been recognized that early treatment can make a significant difference in some individuals. Therefore, the DSMIG group recommended against waiting three months

to make the diagnosis and start treatment, as that delay may have a negative effect on recovery, even though the symptoms commonly last more than three months.

People at all cognitive levels may be affected, including many without prior mental health concerns (Akahoshi et al., 2012). These individuals are commonly in their teens or twenties (although older and younger individuals have been described), and their decline occurs mostly in association with other neuropsychiatric symptoms/conditions (e.g. depression, psychosis, obsessive-compulsive disorder, or catatonia) and rarely in isolation from other psychiatric symptoms (Prasher, 2002). The onset of symptoms is typically fairly acute (sudden), but changes often occur over an extended period of weeks to months.

The core findings of regression/loss of skills may not be apparent initially or may be less significant than other symptoms, particularly behavioral or psychological, and, therefore, not immediately recognized. For example, diagnosis and treatment of depression may or may not lead to improvement of mood disturbance, whereas symptoms of skill loss may only become apparent over time.

Assessment of Regression

Darius, a twenty-three-year-old man with Down syndrome was brought to a Down syndrome clinic because he had experienced increasing cognitive impairment over several months (from his baseline of a stable, mild intellectual disability). According to his family, he had the following worrisome symptoms: social withdrawal and reduced verbal communication; periods of motor slowing alternating with times of increased motor activity and emotional agitation; difficulties with self-care tasks he had previously mastered; periods of increased sleep for weeks followed by weeks of difficulty sleeping; and inability to perform at his job. Darius was taking levothyroxine, which had been prescribed eight months ago when he was diagnosed with hypothyroidism. His thyroid blood testing had been normal five months prior, but now his lab results revealed a very low TSH and high free T4, which indicated that his dose of levothyroxine needed to be reduced. Neither adjusting the dose of his

levothyroxine nor subsequently discontinuing it resulted in a return to normal of those lab values.

With further assessment, it became clear that Darius's thyroid was fluctuating from a hyperthyroid (overactive) state to a hypothyroid (underactive) state and back. Ultimately, his thyroid function was stabilized with radioactive ablation and levothyroxine treatment, and his regressive symptoms improved over several months. In Darius's case, fluctuating thyroid function likely contributed to psychological, motor, and cognitive symptoms.

The assessment of a person with Down syndrome who is regressing can require an extensive search for underlying health problems. A number of possible causes must be ruled out as the cause. However, there are conditions that are more common in people with Down syndrome and commonly result in some degree of regression: sleep apnea, celiac disease, depression, adjustment to life changes, and other conditions listed below in the section on differential diagnosis. Some people with Down syndrome with these conditions develop the full range of symptoms described above and thus meet the definition as proposed by DSMIG-USA, and some have milder symptoms or only some of the symptoms. A thorough assessment, initially focusing on the conditions that are more common in individuals with Down syndrome can help direct the diagnosis and treatment in either case.

It is important to remember that the complete picture of regression syndrome is often not evident on the first visit. Patients or families often focus on the symptoms that are particularly problematic and may not recognize or report other symptoms. Over time, medications prescribed for psychiatric symptoms may lead to improvement of some or all these changes, making other symptoms (e.g., psychomotor slowing) more evident. Sometimes all symptoms may intensify over time until they stabilize. Repeated assessment for new symptoms and improvements will often make the clinical picture clearer.

If you are seeking an assessment for a teen or adult with Down syndrome who is experiencing regression, you may have trouble finding a physician with experience evaluating and treating the condition. You are most likely to find health care providers with the expertise at a Down syndrome clinic. To help delineate cause and

guide treatment, you may want to share these recommendations on what to assess for:

- Duration, age onset, duration of symptoms (weeks-months-years): Has the person experienced prior episodes of regression? What is his developmental, behavioral, and psychiatric history?
- Specific adaptive skill loss, based on previously established skills:
    - cognitive-executive skills
    - social skills
    - speech/language skills
    - ability to learn new skills
    - loss of control of bodily functions (e.g., incontinence)
- Psychiatric symptoms:
    - mood, irritability
    - inattention, distractibility, disorganization
    - obsessive-compulsive behavior, perseveration
    - agitation, aggression
    - apathy, mutism, abulia (reduced initiative)
    - stereotypy, mannerisms (e.g., seemingly odd or purposeless repetitive actions)
- Maladaptive behaviors: self-injury, scratching, poking, skin-picking
- Motor symptoms:
    - catatonia-muscular rigidity, immobility, or slowing or conversely, increased movements
    - tics
- Vegetative symptoms: sleep disturbance, loss of appetite and/or weight
- Additional history to obtain:
    - level of function prior to onset
    - snoring, pauses in breathing, daytime drowsiness
    - history of surgery and/or general anesthesia and relationship in time to the onset of symptoms
    - personal or family stressors
    - trauma, victimization
    - life changes (school, family changes, deaths)
    - recent infections
    - menstrual changes, dysmenorrhea

    - puberty
    - vision or hearing changes
    - change in gait, weakness
    - headaches
- Medication review: changes, additions, or subtractions and timing of these changes relative to onset of regression
- Examinations:
    - Observation-Mental Status-Physical-Neurologic examination
    - DSM 5 or DM-ID 2 criteria checklists
    - DSMIG-USA operational criteria checklist
- Neuropsychology assessment (by neuropsychologist or family/care providers completing checklists):
    - adaptive skills
    - cognitive function (executive skills) decline
    - maladaptive behavior
    - psychiatric symptoms
    - motor-movement (catatonia)

Differential Diagnoses

Miranda, age thirty-three, was brought to a Down syndrome clinic due to concerns about her reduced ability to manage her activities of daily living. Due to changes over the last six months, her family and her support staff were very worried that she was developing Alzheimer's disease. Miranda was not participating in activities, was speaking less, and had withdrawn from contact with others. She was not sleeping well and had taken to sleeping on the couch in the family room, even though outside lighting made it brighter than her bedroom. In addition, her appetite had decreased, and family and support staff reported that she seemed "nervous."

At this exam, dense cataracts were diagnosed, although no cataracts had been visible at her eye exam about eighteen months earlier. In addition to the cataracts, Miranda was diagnosed with depression, possibly

related to her emotional response to the rapid loss of vision. The evaluation did not reveal any other causes for Miranda's regression. Several months after cataract surgery and treatment with an antidepressant, Miranda returned to her previous level of function.

As mentioned above, people with Down syndrome are susceptible to a variety of medical conditions that can affect their brain function, leading to further cognitive, behavioral, motor, and communication impairment. For this reason, it is often important to rule out a number of conditions when exploring the diagnosis of regression syndrome in adolescents and adults with Down syndrome.

As noted earlier, some conditions such as depression and sleep apnea are more common in people with Down syndrome and should be assessed for as part of the initial evaluation. It is actually common to find that the person has multiple disorders rather than just one. Additionally, people with Down syndrome often have limited verbal skills, resulting in difficulties reporting physical symptoms. As a consequence, a physical problem often develops secondary mental health or behavioral symptoms. For all these reasons, multiple medical and psychosocial causes must be considered when assessing an individual with Down syndrome with regression.

Some of the conditions to consider include the following:

1. Medical conditions
    a) medication side effects
    b) sleep apnea
    c) seizures
    d) vitamin B12 deficiency
    e) cervical myelopathy: spine injury (subluxation, spinal stenosis)
    f) chronic pain
    g) dental pain/problems
    h) sinus pain/problems
    i) menstrual pain/problems
    j) gastrointestinal diseases or conditions (e.g., celiac disease), severe constipation
2. Cardiovascular disease

a) uncorrected congenital heart disease with pulmonary hypertension, congestive heart failure, or Eisenmenger's syndrome (a form of severe cyanotic heart disease)
b) stroke: ischemic or hemorrhagic

3. Infectious disease
    a) urinary tract infection
    b) pneumonia
    c) sepsis
    d) viral/bacterial meningitis/encephalitis
    e) Lyme's disease
4. Toxic-metabolic
    a) various electrolyte (e.g. sodium, potassium, calcium) abnormalities
    b) ingestion of various toxic substances
5. Neuropsychiatric disorders
    a) catatonia (see section below)
    b) mood disorder (depression, anxiety)
    c) obsessive-compulsive disorder
    d) psychotic disorder
    e) complex tic disorder
    f) post-traumatic stress disorder
    g) Parkinsonism, dystonia
    h) adjustment to life events – transitions and relationships
        i. loss of family, friends, pets
        ii. school graduation, work setting changes, physical relocation
        iii. response to hospitalization or medical condition
6. Visual impairment
    a) glaucoma
    b) retinal detachment
    c) cataracts
    d) keratoconus
7. Hearing impairment
    a) impaired hearing: for example, due to high frequency hearing loss or due to ear infections or fluid in the middle ear
    b) hyperacusis (overly sensitive hearing)
    c) tinnitus (ringing in the ears)
    d) vertigo (sensation of spinning)

8. Endocrine disorders
    a) hypo- or hyperthyroidism
    b) adrenal insufficiency
    c) diabetes mellitus
    d) puberty-associated
    e) menopause-associated
9. Autoimmune disorders (additional evidence required)
    a) Hashimoto's encephalopathy
    b) pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections (PANDAS)
    c) central nervous system manifestation of celiac disease
    d) central nervous system manifestation of systemic lupus erythematosus (SLE)
    e) autoimmune encephalopathy
    f) limbic encephalitis

Recently researchers described a method to guide the assessment of a person with Down syndrome and regression (Jacobs, Schwartz, McDougle, & Skotko, 2016). This approach involves grouping possible causes into tiers and assessing for the more likely diagnoses first, gradually working down the tiers to more obscure possibilities. To decide how far to go down the tiers, practitioners rely on clinical assessment, the conditions diagnosed, and the response to treatment. In practice, most individuals with Down syndrome and regression do not need to be evaluated for all the causes of regression listed above. The further down a practitioner goes in the tiers, the less likely one of those conditions is present.

In addition to assessing for the possible causes of regression listed above, it may also be worthwhile to perform an MRI or CT scan of the brain. This is because regression may be associated with changes visible on MRI/CT. One study (Akahoshi et al., 2012) reportedly found atrophy or hypoplasia in the basal ganglia of individuals with Down syndrome and regression, but there was no data on controls with Down syndrome who did not have regression, so it is not clear if this finding is unique to regression.

Here are a few other considerations when considering causes of regression:

Immune System Dysfunction. It is known that the immune system of people with Down functions differently than that of people without Down syndrome (Kumar,

Rajadhyaksha, & Wortsman, 2001; Pellegrini et al., 2012). This includes a higher rate of autoimmune conditions—conditions in which the person's immune system attacks a part (or parts) of the person's own body. In regression, in theory, the immune system could be functioning against the person's brain (Worley et al., 2015). Ongoing study should help improve the understanding of the role the immune system may play in regression syndrome and how that understanding can improve treatment.

Sleep Disturbance. Sleep is another area that is more commonly a concern in people with Down syndrome, and sleep disturbance can contribute to mental health changes. Sleep disturbances are extremely common in adolescents and adults with Down syndrome who experience new or worsening mental health changes. In one study, researchers (Capone, Aidikoff, Taylor, & Rykiel, 2013) found that sleep apnea was more common in those with depression, and functional decline was found only in those with depression.

Depression. Severe depression can also be a cause of regression syndrome. In one study (Capone, Goyal, and Ayres et al., 2006), researchers suggested that stressors, such as a growing awareness of being different and bereavement, may exacerbate symptoms of depression in adolescents. Some of these depressed individuals appear to have regression in relation to a life event (or events). However, the nature of these events and their potential for triggering neuropsychiatric change is not at all clear and there are no data on life events in individuals without regression.

Although mood disturbances such as depression are often part of the regression syndrome and many individuals are treated with antidepressants, the causal relationship between depression and regression, sleep disturbance, and stressful life events is unclear presently.

Treatment for Regression

Treatments for many of the commonly diagnosed conditions that can lead to regression in people with Down syndrome are covered elsewhere in this book and in our book *The Guide to Good Health for Teens & Adults with Down Syndrome*. Consequently, these treatments will not be discussed in detail here, although we will briefly describe issues related to some of the major causes of regressive symptoms. It is important to note, however, that no one medication or combination of medications has been found to address the symptoms of all individuals with regression syndrome. This is consistent with the finding that there are a number of possible causes. Several

researchers (Worley et al., 2015; Mircher, 2017) have described some medications that were beneficial for some individuals but found that no medication or combination of medications was consistently effective. Likewise, one group of researchers (Akahoshi et al., 2012) described diagnosing a variety of psychiatric diagnoses in people with regression, including anxiety, depression, and psychosis, with variable responses to treatment—from nonresponsive to fully responsive. A combination of medications may be necessary.

Despite, or perhaps due to the lack of universal, clear understanding of the cause of regression syndrome, health care providers often prescribe treatment that targets symptoms such as psychiatric, behavior, attention, motor, sleep, and other medical symptoms, with a focus on the symptoms that cause the most impairment and are most likely to respond to treatment. Treatment often involves an approach that includes medications; changes to the person's home, school, or work environment; and treatment of coexisting medical conditions. Depending on the person's symptoms, occupational, physical, speech, and behavioral therapy may all be helpful. Which treatment to prioritize will depend on the provider's experience, family preferences, and the patient's response to treatments. Support for the individual's caretaker is generally part of the treatment regimen and may include assistance with adapting to a new reality and the future.

A very important consideration during treatment is how to handle the withdrawal from activities or reluctance to leave home that often occurs in people with regression syndrome. Many of these individuals need a gradual, "safe" reentry into activities. To prevent abrupt reversal of improvement, it is essential that the person successfully participate in activities that are unlikely to provoke significant anxiety or fear. Strategies to help the person ease into activities may include the following:

- giving the person lots of reinforcement for small steps;
- initially not expecting him to do things as long or as well as he used to;
- giving the person input into the selected activity, the participants, etc.; and
- carefully observing the individual's response to a gradual increase in the amount and intensity of the activity (to avoid too rapid of an increase that could result in excessive stress).

This gradual resumption of normal activities without fear and anxiety is therapeutic in itself.

Catatonia and Regression

Catatonia is an especially worrisome condition that may occur in people with Down syndrome and regression. Catatonia is a condition that traditionally was associated with psychoses, but it is now recognized that it can occur with a variety of psychiatric or medical conditions such as depression (Daniels, 2009). It has also been proposed that it be a separate diagnosis not associated with any psychiatric or medical conditions (Shorter, 2012; Padhy, Parakh, & Sridhar, 2014). The condition has not been studied much in Down syndrome, but one group of researchers (Ghaziuddin et al., 2015) described catatonia in a small series of people with Down syndrome.

There are two primary treatments for catatonia: medications and electroconvulsive therapy (ECT). ECT involves sedating an individual with anesthesia and then inducing a seizure with a short electrical stimulation of the brain. The authors of the 2015 study (Ghaziuddin et al.) used high dose lorazepam combined with ECT to treat the four individuals with Down syndrome in their study and reported that they all recovered their baseline level of functioning. However, the patients in this study needed prolonged usage of ECT to successfully recover from catatonia. The researchers speculated that this raises the possibility that differences in pathophysiological mechanisms may underlie catatonia in individuals with Down syndrome and other developmental disabilities versus those with typical development. The authors concluded that they suspect catatonia is a common cause of unexplained deterioration in adolescents and young adults with Down syndrome.

More recently, Judith Miles (2017), a coauthor on the paper with Neera Ghaziuddin, described successfully treating patients with Down syndrome and catatonia using medications without ECT. In addition to high dose lorazepam, she reported using N-methyl-D-aspartate receptor (NMDA) antagonists including dextromethorphan-quinidine (Nuedexta), memantine (Namenda), and amantadine. She reported best success with dextromethorphan-quinidine. In patients without Down syndrome, another researcher (Daniels, 2009) successfully used NMDA antagonists to treat individuals whose catatonia did not respond to lorazepam. These medications may be effective because a dysregulation of dopamine, gamma-Aminobutyric acid (GABA), and glutamate is proposed as being involved in catatonia. Lorazepam enhances GABA activity, and the NMDA antagonists reduce the effect of glutamate.

Despite Ghaziuddin's successful use of ECT in treating four patients with Down syndrome and catatonia, further investigation of the use ECT for this purpose is warranted. A recent meta-analysis of the use of ECT in people without Down

syndrome who were diagnosed with catatonia found serious side effects in seven studies, including mental confusion, memory loss, headache, or adverse effects associated with anesthesia (Leroy et al., 2017). The study further concluded that the "literature consistently describes improvement in catatonic symptoms after ECT. However, the published studies fail to demonstrate efficacy and effectiveness." Additional study is needed to understand the role of ECT in people with Down syndrome and catatonia. However, in our experience and the experience other clinicians have reported to us, treatment with ECT does result in dramatic improvement in some individuals.

Prognosis

For people with Down syndrome, recovery from regression syndrome is highly dependent on both the cause and the certainty of the diagnosis. For those with more challenging regression, recovery of mental health changes and loss of skills may take many months or years. One group of researchers (Mircher et al., 2017) found that only 10 percent of the individuals they followed completely recovered from regression syndrome. They also reported that 10 percent actually worsened, 37 percent stabilized but did not recover (they did not keep regressing but did not regain their lost skills or their previous level of mental health), and 43 percent had partial recovery. As we learn more about the causes of regression in people with Down syndrome, the prognosis for recovery will hopefully improve.

Conclusion

Regression in adolescents and younger adults with Down syndrome has only recently begun to be studied in earnest. At present, we do not even have a universally agreed-upon name for the condition, although the name regression syndrome has been proposed by the Down Syndrome Medical Interest Group in the United States. People with Down syndrome can lose skills due to many disorders or combinations of disorders. Clearly, there are significant gaps in our knowledge of regression syndrome, and further study is needed to define and optimize treatment for affected individuals.

Return to Table of Contents

References

Adult Down Syndrome Center (2016). *Advocate Medical Group's Adult Down Syndrome Center* [Video]. https://youtu.be/KjKESkPjXks.

Akahoshi, K., Matsuda, H., Funahashi, M., Hanaoka, T., & Suzuki, Y. (2012). Acute neuropsychiatric disorders in adolescents and young adults with Down syndrome: Japanese case reports. *Neuropsychiatric Disease and Treatment, 8*, 339–345.

Alzheimers.net. 2017 Alzheimer's statistics (2018). Alzheimers.net. Retrieved August 20, 2020, from https://www.alzheimers.net/resources/alzheimers-statistics/.

American Psychiatric Association. (2013). *Diagnostic and statistical manual of mental disorders* (5th ed.). Arlington, VA: American Psychiatric Publishing.

Anderson, L. M., Shinn, C., Fullilove, M. T., et al. (2003). The effectiveness of early childhood development programs: A systematic review. *American Journal of Preventative Medicine, 24* (3 suppl.): 32.

Ayres, A. J. (1989). *Sensory integration and praxis tests*. Los Angeles, CA: Western Psychological Services.

Ballard, C., Mobley, W., Hardy, J., Williams, G., & Corbett, A. (2016). Dementia in Down's syndrome. *Lancet Neurology, 15*, 622–636.

Beck, M. (2011). *Expecting Adam: A true story of birth, rebirth, and everyday magic*. New York: Three Rivers Press.

Breiter, H. C., Rauch, S. I., Kwong, K., et al. (1996). Functional magnetic resource imaging of symptom provocation in obsessive-compulsive disorder. *Archives of General Psychiatry, 53*(7): 595–606.

Brickell, C., & Munir, K. (2008). Grief and its complications in individuals with intellectual disability. *Harvard Review of Psychiatry, 16*(1): 1–12.

Brown, C., & Dunn, W. (2002). *The adolescent/adult sensory profile*. San Antonio, TX: Psychological Corporation.

Brown, R. T., Freeman, W. S., Perrin, J. M., Stein, M. T., Amler, R.W., Feldman, H. M., Pierce, K., & Wolraich, M. L. (2001). Prevalence and assessment of attention-deficit/hyperactivity disorder in primary care settings. *Pediatrics, 107*(3): e43.

Bruininks, R. H., & Bruininks, B. D. (2005). *Bruininks-Oseretsky test of motor proficiency*. 2nd edition. Circle Pines, MN: AGS Publishing.

Bruni, M., Cameron, D., Dua, S., & Noy, S. (2010). Reported sensory processing of children with Down syndrome. *Physical & Occupational Therapy in Pediatrics, 30*(4), 280–293.

Bruni, M. (2016). *Fine motor skills in children with Down syndrome*. 3rd ed. Bethesda, MD: Woodbine House.

Buckley, S., & Le Prevost, P. (2002). Speech and language therapy for Down syndrome children: Guidelines for best practice based on current research. *Down Syndrome News and Update, 2*(2): 70–76.

Buggey, T. (2009). *Seeing is believing: Video self-modeling for people with autism and other developmental disabilities*. Bethesda, MD: Woodbine House.

Burger-Caplan R., Saulnier C. A., & Sparrow S. S. (2018). Vineland Adaptive Behavior Scales. In J. Kreutzer, J. DeLuca, & B. Caplan B. (Eds.), *Encyclopedia of clinical neuropsychology*. New York: Springer.

Capone, G. T., Goyal, P, Ares, W., & Lannigan, E. (2006). Neurobehavioral disorders in children, adolescents, and young adults with Down syndrome. *American Journal of Medical Genetics, Part C: Seminars in Medical Genetics, 142*,158–172.

Carey, W. B., & McDevitt, S. C. (1995). Coping with children's temperament: A guide for professionals. New York: Basic Books.

Champagne, T. (2005). Expanding the role of sensory approaches in acute psychiatric settings. *Mental Health Special Interest Section Quarterly, 28*, 1–4.

Chicoine, B., & McGuire, D. (1997). Longevity of a woman with Down syndrome: A case study. *Mental Retardation, 35*, 477–479.

Chicoine, B., McGuire, D., Hebein, S., & Gilly, D. (1994). Development of a clinic for adults with Down syndrome. *Mental Retardation, 32*(2): 100–106.

Chicoine, B, Parikh, S, Sulo, S, & Chicoine, L. (2014). The utility of anti-tissue transglutaminase antibody-IgA (tTG-IgA) testing for celiac disease in adults with Down syndrome. *American Journal of Clinical Medicine, 10*(2), 56–60.

Cohen, W. I., & Patterson, B. J. (1998). Neurodevelopmental disorders in Down syndrome. In T. Hassold & D. Patterson (Eds.), *Down syndrome: A promising future, together*. New York: Wiley-Liss.

Cohen, W. (Ed.). (1991). Health care guidelines for individuals with Down syndrome. *Down Syndrome Quarterly, 4*(3). (Available online at http://www.ds-health.com/health99.htm.)

Coppus, A., Evenhuis, H., Verberne, G. J., Visser, F., van Gool, P., Eikelenboom, P., & van Duijin, C. (2006). Dementia and mortality in persons with Down's syndrome. *Journal of Intellectual Disability Research, 50*, 768–777.

Cuijpers, P., Sijbrandij, M., Koole, S., Andersson, G., Beekman, A., & Reynolds, C. (2014). Adding psychotherapy to antidepressant medication in depression and anxiety disorders: A meta-analysis. *World Psychiatry, 13*(1): 56–67.

Daniels, J. (2009). Catatonia: Clinical aspects and neurobiological correlates. *Journal of Neuropsychiatry and Clinical Neurosciences, 21*, 371–380.

Devenny, D.A., & Matthews, A. (2011). Regression: Atypical loss of attained functioning in children and adolescents with Down syndrome. In R. M. Hodapp (Ed.), *International Review of Research in Developmental Disabilities* (Vol. 41, pp. 233–264). Oxford: Academic Press.

De Vinck, C. (1990). *The power of the powerless*. New York: Doubleday.

Diaz, R., & Berk, I. (1991). *Private speech: From social interaction to self-regulation*. Mahwah, NJ: Lawrence Erlbaum Associates.

Doran, E., Keator, D. Head, E., Phelan, M. J., Kim, R., Totoiu, M., Barrio, J. R., Small, G. W., Potkin, S. G., & Lott, I. T. (2017). Down syndrome, partial trisomy 21, and absence of Alzheimer's disease: The role of APP. *Journal of Alzheimer's Disease, 56*, 459–470.

Down syndrome & spectrum disorders (September/October 1999). *Disability Solutions 3, 5 & 6*: 1–40.

Down's Syndrome Scotland. *Living with dementia: A families' and carers' guide*. Edinburgh, Scotland: Down's Syndrome Scotland. Retrieved August 25, 2020, from https://www.dsscotland.org.uk/wp-content/uploads/2019/05/DSS_LIVING-WITH-DEMENTIA_AW_WEB.pdf.

Dowrick, P. W. (1991). *Practical guide to using video in the behavioral sciences*. New York: Wiley.

Dunn, W. (2014). *The sensory profile 2*. San Antonio, TX: Psychological Corporation.

Dykens, E. M., Shah, B., Davis, B., et al. (2015). Psychiatric disorders in adolescents and young adults with Down syndrome and other intellectual disabilities. *Journal of Neurodevelopmetal Disorders, 7(*9).

Eddy, M. F., & Walbroehl, G. S. (1998). Recognition and treatment of obsessive-compulsive disorders. *American Family Physician, 57*(7), 1623–1628.

Ekstein, S., Glick, B., Weill, M., Kay, B., & Berger, I. (2011). Down syndrome and Attention-Deficit/Hyperactivity Disorder (ADHD). *Journal of Child Neurology, 26*(10), 1290–1295.

Esbensen, A. J, Seltzer, M. M., & Krauss, M. W. (2008). Stability and change in health, functional abilities, and behavior problems among adults with and without Down syndrome. *American Journal of Mental Retardation, 113*(4), 263–277.

Evans, A. L., & McKinlay, I. A. (1988 Jan.-Feb.). Sexual maturation in girls with severe mental handicap. *Child: Care, Health, and Development, 14*(1), 59–69.

Evenhuis, H., Kengen, M., & Eurlings, H. (2009). The dementia questionnaire for people with intellectual disabilities. In V. Prasher (Ed.), *Neuropsychological assessments of dementia in Down Syndrome and intellectual disabilities*. London: Springer.

Fish, J. (2011). Severe impairment battery. In J. S. Kreutzer, J. DeLuca, & B. Caplan (Eds.), *Encyclopedia of clinical neuropsychology*. New York: Springer.

Fletcher, R., Barnhill, J., & Cooper, S-A. (2017). *Diagnostic manual—Intellectual disability 2 (DM-ID): A textbook of diagnosis of mental disorders in persons with intellectual disability*. Kingston, NY: National Association for the Dually Diagnosed.

Frank, E., Kupfer, D. J., Derel, J. M., Cornes, C., Mallinger, A. G. Thase, M. E., McEachran, A. B., & Grochoneinski, V. J. (1990). Three-year outcomes for maintenance therapies in recurrent depression. *Archives of General Psychology, 47*, 1093–1099.

Gamage, K. I., Hardy, J., & Hall, C. R (2001). A description of self-talk in exercise. *Psychology of Sport & Exercise, 2*(4), 233–247.

Gedye, A. (1995). *Manual for the dementia scale for Down syndrome*. Vancouver, BC: Gedye Research and Consulting.

Geisinger, K. F., & Carlson, J. F. (1992). Assessing language-minority students. *Practical Assessment, Research & Evaluation, 3*(2).

Ghaziuddin, N., Nassiri, A., & Miles, J. H. (2015). Catatonia in Down syndrome: A treatable cause of regression. *Neuropsychiatric Disease and Treatment, 11*, 941–949.

Ghaziuddin, N., Tsai, M., & Ghaziuddin, N. (1992). Autism in Down syndrome: Presentation and diagnosis. *Journal of Intellectual Disability Research, 36*, 449–456.

Ginsberg, Y., Quintero, J., Anand, E., Casillas, M., & Upadhyaya, H. P. (2014). Underdiagnosis of attention-deficit/hyperactivity disorder in adult patients: A review of the literature. *Primary Care Companion for CNS Disorders, 16*(3).

Glasberg, B. (2006). *Functional behavior analysis for people with autism: Making sense seemingly senseless behavior*. Bethesda, MD: Woodbine House.

Gray, C. (2015). *The new social story book: Over 150 social stories that teach everyday social skills to children and adults with autism and their peers*. Arlington, TX: Future Horizons.

Greenspan, S., & Granfield, J. M. (1992). Reconsidering the constrict of mental retardation: Implications of a model of social competence. *American Journal on Mental Retardation, 96*(4), 442–453.

Greenspan, S., & Shoultz, J. (1981). Why mentally retarded adults lose their jobs: Social competence as a factor in work adjustment. *Applied Research in Mental Retardation, 2*, 23–38.

Guralnick, M. (1998). Effectiveness of early intervention for vulnerable children: A developmental perspective. *American Journal on Mental Retardation, 102*, 319–345.

Head, E., Powell, D., Gold. B. T., & Schmitt, F. A. (2012). Alzheimer's disease in Down syndrome. *European Journal of Neurology and Neurogenetic Disorders, 1*, 353–364.

Healey, K. (2017). More than meets the eye: Deafness and invisible disabilities. *Canadian Medical Association Journal, 189*(13), E506–E507.

Heller, T. (1982). Social disruption and residential relocation of mentally retarded children. *American Journal of Mental Deficiency, 87*, 48–55.

Hill, W., & Wehman, P. (1979). Employer and nonhandicapped co-worker perceptions of moderately and severely retarded workers. *Journal of Contemporary Business, 8,* 107–111.

Huxley, A., Prasher, V. P., & Hague, M. S. (2000). The dementia scale for Down's syndrome. *Journal of Intellectual Disability Research, 44*(6), 697–698.

Jacobs, J., Schwartz, A., McDougle, C. J., & Skotko, B. G. (2016). Rapid clinical deterioration in an individual with Down syndrome. *American Journal of Medical Genetics A, 170,* 1899–1902.

Jarrold, C., & Baddeley, A. D. (2001). Short-term memory in Down syndrome: Applying the working memory model. *Down Syndrome Research and Practice 17*(1) 17–23.

Jensen, P. S., & Cooper, J. R. (Eds.). (2002). *Attention deficit hyperactivity disorder: State of science—best practices*. Kingston, NJ: Civic Research Institute.

Johnson, D. (2016). *Differentiating sensory from behavior*. [Video]. https://summit-education.com/course/CSENDJ.1/differentiating-sensory-from-behavior#/onlinevideo/6-ceus.

Kahn, S., Owinowa, T., & Pary, R. J. (2002). Down syndrome and major depressive disorder: A review. *Mental Health Aspects of Developmental Disabilities, 5*, 46–52.

Katerndahl, D.A., & Vande Creek, I. (1983). Hyperthyroidism and panic attacks. *Psychosomatics, 24*(5), 491–496.

Korbel, J. O., Tirosh-Wagner, T., Urban, A. E., et al. (2009). The genetic architecture of Down syndrome phenotypes revealed by high-resolution analysis of human segmental trisomies. *Proceedings of the National Academy of Sciences of the USA, 106*, 12031–12036.

Kranowitz, C. (2016). *The out-of-sync child grows up: Coping with sensory processing disorder in the adolescent and young adult years*. New York: Penguin Random House.

Krantz, P., & McClannahan, L., (2010). *Activity schedules for children with autism: Teaching independent behavior*. Bethesda, MD: Woodbine House.

Kumar, V., Rajadhyaksha, M., & Wortsman, J. (2001). Celiac disease-associated autoimmune endocrinopathies. *Clinical and Vaccine Immunology, 8*, 678–685.

Kumin, L. (2003). *Early communication skills for children with Down syndrome: A guide for parents and professionals*. 2nd ed. Bethesda, MD: Woodbine House.

Lai, F., & Williams, R. S. (1989). A prospective study of Alzheimer disease in Down syndrome. *Archives of Neurology, 46*(8), 849–853.

Landon, T. M., & Barlow, D. H. (2004). Cognitive-behavioral treatment for panic disorders: Current status. *Journal of Psychiatric Practice 10*(4): 211–226.

Lee, H. (1960). *To kill a mockingbird*. Philadelphia: J. B. Lippincott.

Leroy, A., Naudet, F., Vaiva, G., Francis, A., Thomas, P., & Amad, A. (2017). Is electroconvulsive therapy an evidence-based treatment for catatonia? A systematic review and meta-analysis. European Archives of Psychiatry and Clinical Neuroscience.

Levinson, D. (1978). *The seasons of a man's life*. New York: Ballantine.

Livingstone, N., Hanratty, J., McShane, R., & Macdonald, G. (2015). Medications for cognitive decline in people with Down syndrome. Cochrane Database of Systematic Reviews. https://www.cochrane.org/CD011546/BEHAV_medications-cognitive-decline-people-down-syndrome.

Lord, C., Risi, S., Lambrecht, L., et al. (2000). The ADOS-G (Autism Diagnostic Observation Schedule-Generic): A standard measure of social communication deficits associated with autism spectrum disorders. *Journal of Autism and Developmental Disorders, 30*, 205–223.

Luchterhand, C. (1998). Mental retardation and grief following a death loss. Silver Spring, MD: The Arc of the United States.

Manghi, E. (2013). Multicultural challenges: Co-occurring Down Syndrome and autism. In M. Froehlke & R. Zaborek (Eds), *When Down syndrome and autism intersect*. Bethesda, MD: Woodbine House.

Mann, D. M., Yates, P. O., & Marcyniuk, B. (1984). Alzheimer's presenile dementia, senile dementia of Alzheimer type, and Down's syndrome in middle age form an age-related continuum of pathological changes. *Neuropathology and Applied Neurobiology, 10*, 185–207.

Mantry, D., Cooper, S. A., Smiley, E., et al. (2008). The prevalence and incidence of mental ill-health in adults with Down syndrome. *Journal of Intellectual Disability research, 52*(Pt. 2), 141–155.

Martin, J. E., Rusch F. R., Lagomarcino, T., & Chadsey-Rusch, J. (1986). Comparison between workers who are nonhandicapped and mentally retarded: Why they lose their jobs. *Applied Research in Mental Retardation 7*, 467–474.

Martinez-Cue, C., Baamondem, C., Lumbreras, M. A., Vallina, F., Dierssen, M., & Flores, J. (1999). Murine model for Down syndrome shows reduced responsiveness to pain. *Neuroreport 10*(5), 1119–1122.

May-Benson, T. A., & Koomar, J. A. (2010). Systematic review of the research evidence examining the effectiveness of interventions using a sensory integrative approach for children. *American Journal of Occupational Therapy*, *64*, 403–414.

McBrien, D. (2012). Attention problems in Down syndrome: Is this ADHD? University of Iowa Stead Family Children's Hospital. Retrieved Sept. 3, 2020, from https://uichildrens.org/health-library/attention-problems-down-syndrome-adhd.

McCarron, M., McCallion, P., Reilly, E., & Mulryan, N. (2014). A prospective 14-year longitudinal follow-up of dementia in persons with Down syndrome. *Journal of Intellectual Disability Research, 58*, 61–70.

McGuire, D. (1999, November). The groove. *NADS: The Newsletter of the National Association for Down Syndrome*.

McGuire, D. (2012). If people with Down syndrome ruled the world [presentation]. National Association for Down Syndrome.

McGuire, D., & Chicoine, B. (1996). Depressive disorders in adults with Down syndrome. *The Habilitative Mental Healthcare Newsletter 15*(1), 1–7.

McGuire, D. E., & Chicoine, B. A. (2002). Life issues of adolescents and adults with Down syndrome. In W. Cohen, I. Nagel, & M. E. Madnick (Eds.), *Down syndrome: Visions for the 21st century*. New York: Wiley-Liss.

McGuire, D., Chicoine, B., & Greenbaum, E. (1997, July/August). "Self talk" in adults with Down syndrome. *Disability Solutions, 2*(1), 1–4.

McGuire, D., Manghi, E, & Chicoine, B. (2013). When autism is suspected for teens and adults with Down syndrome. In M. Froehlke & R. Zaborek (Eds.), *When Down syndrome and autism intersect*. Bethesda, MD: Woodbine House.

Mehra, C., Sil, A., Hedderly, T., et al. (2019). Childhood disintegrative disorder and autism spectrum disorder: A systematic review. *Developmental Medicine and Child Neurology, 61*(5), 523–534.

Miles, J. (2017). Catatonia as a cause of regression in Down syndrome [conference presentation]. Sacramento: Down Syndrome Medical Interest Group-USA.

Mills, S., & Hedderly, T. (2014). A guide to childhood motor stereotypies, tic disorders and the Tourette spectrum for the primary care practitioner. *Ulster Medical Journal, 83*(1), 22–30.

Mircher, C., Cieuta-Walti, C., Marey, I., et al. (2017). Acute regression in young people with Down syndrome. *Brain Sciences, 7*, E57.

Moran, J. A., Rafii, M. S., Keller, S. M., Singh, B. K., & Janicki, M. P. (2013). The National Task Group on Intellectual Disabilities and Dementia Practices consensus recommendations for the evaluation and management of dementia in adults with intellectual disabilities. *Mayo Clinic Proceedings, 88*, 831–840.

Murphy, K. R., & Barkley, R. A. (1996). The prevalence of DSM-IV symptoms of AD/HD in adult licensed drivers: Implications for clinical diagnosis. *Comprehensive Psychiatry, 37*, 393–401.

Myers B.A., & Pueschel, S. (1991). Psychiatric disorders in a population with Down syndrome. *Journal of Nervous & Mental Disorders, 179*, 609–613.

National Alliance on Mental Illness. (2019, Sept.). *Mental health by the numbers*. Retrieved September 21, 2020, from https://www.nami.org/learn-more/mental-health-by-the-numbers.

NADS News: The Newsletter of the National Association for Down Syndrome. (2004, January).

National Institutes of Health. (2020, August). *Health literacy*. Retrieved September 21, 2020, from https://www.nih.gov/institutes-nih/nih-office-director/office-communications-public-liaison/clear-communication/health-literacy.

National Institute on Mental Health. (2019). *Mental illness*. Retrieved March 15, 2019, from https://www.nimh.nih.gov/health/statistics/mental-illness.shtml.

Padhy, S. K., Parakh, P., & Sridhar, M. (2014). The catatonia conundrum: Controversies and contradictions. *Asian Journal of Psychiatry, 7*, 6–9.

Papolos, D., & Papolos, J. (1999). *The bipolar child: The definitive and reassuring guide to childhood's most misunderstood disorder*. New York: Broadway Books.

Parker, R. M., Ratzan, S. C., & Lurie, N. (2003). Health literacy: A policy challenge for advancing high-quality health care. *Health Affairs, 22*(4), 147–153.

Patel, L., Wolter-Warmerdam, K., Leifer, N., & Hickey, F. (2018). Behavioral characteristics of individuals with Down Syndrome. *Journal of Mental Health Research in Intellectual Disabilities, 11*(3), 221–246.

Patt, P., Andiloro, N., & Gavin, M. (2009). Parent/carer ratings of self-talk behavior in children and adults with Down syndrome in Canada and the United Kingdom. Down Syndrome Education International. Retrieved June 8, 2020, from https://library.down-syndrome.org/en-us/research-practice/online/2008/parent-carer-ratings-self-talk-behaviour-adults-down-syndrome-canada-united-kingdom/?_ga=2.33810465.1427435271.1676408262-859960116.1671553740.

Pellegrini, F. P, Marinoni, M., Frangione, V., et al. (2012). Down syndrome, autoimmunity and T regulatory cells. *Clinical & Experimental Immunology, 169*, 238–243.

Pickhardt, C. E. (2013). *Surviving your child's adolescence*. San Francisco: Jossey-Bass.

Pickhardt, C. E. (2015, May 11). Adolescence and goal avoidance. Psychology Today. https://www.psychologytoday.com/blog/surviving-your-childs-adolescence/201505/adolescence-and-goal-avoidance.

Powers, M., ed. (2000). *Children with autism: A parents' guide*. Bethesda, MD: Woodbine

Prasher, V. (2002). Disintegrative syndrome in young adults [Letter to the editor]. *Irish Journal of Psychological Medicine, 19*, 101.

Prasher, V. P., Farrer, M. J, Kessling, A. M., Fisher, E. M., West, R. J., Barber, P. C., & Butler, A. C. Butler. (1998). Molecular mapping of Alzheimer-type dementia in Down's syndrome. *Annals of Neurology, 43*, 380–383.

Reid, J. R., & Wheeler, S. F. (2005). Hyperthyroidism: Diagnosis and treatment. *American Family Physician, 72*, 623–630

Reiss, S., Levitan, G. W., & Szysko, J. (1982). Emotional disturbance and mental retardation: Diagnostic overshadowing. *American Journal of Mental Deficiency, 86*, 567–574.

Rogers, C. (1951). *Client centered therapy: Its current practice implications and theory*. Boston: Houghton Mifflin.

Rollin, H. (1946). Personality in mongolism with special reference to the incidence of catatonic psychosis. *American Journal of Mental Deficiency, 51*, 219–237.

Rosen, L. (2002, December). Family dynamics: In the treatment of Tourette syndrome, *Exceptional Parent, 32*(12).

Saxena, S., Brody, A. L., Schwartz, J. M., & Baxter Jr., L. R. (1998). Neuroimaging and frontal-subcortical circuitry in obsessive-compulsive disorder. *British Journal of Psychiatry, 35*(suppl.), 26–37.

Schwartz, J. M., Stoessel, P.W., Baxter Jr., L. R., et al. (1996). Systematic changes in cerebral glucose metabolic rate after successful behavior modification treatment of obsessive-compulsive disorder. *Archives of General Psychiatry, 53*(2), 109–13.

Seligman, M. (1998). *Learned optimism: How to change your mind and your life*. New York: Pocket Books.

Seligman, M. E. P. (1975). *Helplessness: On depression, and death*. San Francisco: W. H. Freeman.

Seligman, M. E. P., Klien, D. C., & Miller, W. R. (1967). Depression. In H. Leitenberg (Ed.), *Handbook of behavior modification*. New York: Appleton-Century Crofts.

Seligman, M. E., Maier, S. F., & Geer, J. H. (1968). Alleviation of learned helplessness in the Dog. *Journal of Abnormal Psychology, 73*(3), 256–262.

Shorter, E. Making childhood catatonia visible, separate from competing diagnoses (Comment). (2012). *Acta Psychiatrica Scandinavica, 125*, 3–10.

Shott, S. R., Amin, R., Chini, B., Heubi, C., Hotze, S., & Akers, R. (2006). Obstructive sleep apnea: Should all children with Down syndrome be tested? *Archives of Otolaryngology–Head & Neck Surgery, 132*(4): 432–436.

Siperstein, G. N., & Bak, J. J. (1985). Effects of social behavior on children's attitudes toward their mildly and moderately handicapped peers. *American Journal of Mental Deficiency, 90*, 319–27.

Snowdon, D. (2001). *Aging with grace: What the nun study teaches us about leading longer, healthier, and more meaningful lives*. New York: Bantam Dell.

Sovner, R. S. (1986). Limiting factors in the use of DSM-III criteria with mentally ill/ mentally retarded persons. *Psychopharmacological Bulletin, 22*, 1055–1059.

Sovner, R. S., & Hurley, A. D. (1993). Commentary: Psychotoform psychopathology. *Habilitative Mental Healthcare Newsletter, 12,* 112.

Stainton, T., Hole, R., Charles, G., Yodanis, C., Powell, S., & Crawford, C. (2006). *Non-residential supports and intellectual disability: A review of the literature on best practices, alternatives, and economic impacts*. Vancouver, BC: University of British Columbia. https://citizenship.sites.olt.ubc.ca/files/2014/07/Non-residential_Alternatives_Document_nov06.pdf.

Strydom, A., Coppus, A., Blesa, R., et al. (2018). Alzheimer's disease in Down syndrome: An overlooked population for prevention trials. *Alzheimer's and Dementia (NY), 4,* 703–713.

Taylor, M., & Mottweiler, C. (2008, Summer). Imaginary companions: Pretending they are real but knowing they are not. *American Journal of Play*. https://www.museumofplay.org/app/uploads/2022/01/1-1-article-taylor-imaginary-companions.pdf.

Tyrrell, J., Cosgrave, M., McCarron, M., et al. (2001). Dementia in people with Down's syndrome. *International Journal of Geriatric Psychiatry,16*, 1168–1174.

VandenBerg, N. L. (2001). The use of a weighted vest to increase on-task behavior in children with attention difficulties. *Journal of Occupational Therapy, 55*, 621–628.

Vicari, S., Pontillo, M., & Armando, M. (2013). Neurodevelopmental and psychiatric issues in Down's syndrome: assessment and intervention. *Psychiatric Genetics, 23*(3).

Visser, S.N. et al. (2013). Trends in the parent-report of health care provider-diagnosed and medicated attention-deficit/hyperactivity disorder: United States, 2003–2011. *Journal of the American Academy of Child & Adolescent Psychiatry, 53*(1), 34– 46.e2.

Vygotsky, L. (1962). *Thought and language*. Cambridge, MA: MIT Press. (Lev Vygotsky is the Russian psychologist credited with explaining how higher thought and our inner silent dialogues emerge from childhood's private speech or self-talk.)

Walker, J. C., Dosen, A., Buitelaar, J. K., & Janzing, J. G. (2011). Depression in Down syndrome: A review of the literature. *Research in Developmental Disabilities 32*(5):1422–1440.

Wehman, P. (1981). *Competitive employment: New horizons for severely disabled individuals*. Baltimore, MD: Brookes.

Wild, G. (2015). Developing effective sensory diets [Presentation]. Retrieved March 20, 2020, from https://www.summit-education.com/.

Will, E., Daunhauer, L., Fidler, D., Lee, N., Rosenberg, C., & Hepburn, S. (2019). Sensory processing and maladaptive behavior: Profiles within the Down syndrome phenotype. *Physical & Occupational Therapy in Pediatrics, 39*(5), 461–476.

Wiseman, F. K., Al-Janabi, T, Hardy, J, et al. (2015). A genetic cause of Alzheimer disease: Mechanistic insights from Down syndrome. *Nature Reviews Neuroscience, 16*, 564–574.

Whittle, N., Sartori, S. B., Dierssen, M., Lubec, G, & Singewald, N. (2007). Fetal Down syndrome brains exhibit aberrant levels of neurotransmitters critical for normal brain development. *Pediatrics, 120*(6), e1465–471.

Will, E. A., Daunhauer, L. A., Fidler, D. J., Lee, N. R., Rosenberg, C. R. & Hepburn, S. L. (2019). Sensory processing and maladaptive behavior: Profiles within the Down syndrome phenotype, *Physical & Occupational Therapy in Pediatrics, 39*(5), 461–476.

Wisniewski, K. E., Dalton, A. J., McLachlan, C., Wen, G. Y., & Wisniewski, H. M. (1985). Alzheimer's disease in Down's syndrome: Clinicopathologic studies. *Neurology,* 35**,** 957–961.

Wolf, A. (2002). *Get out of my life, but first could you drive me & Cheryl to the mall: A parent's guide to the new teenager*. New York: Farrar, Straus and Giroux.

Worley, G., Crissman, B. G., Cadogan, E., Milleson, C., Adkins, D. W., & Kishnani, P. S. (2015). Down syndrome disintegrative disorder: new-onset autistic regression, dementia, and insomnia in older children and adolescents with Down syndrome. *Journal of Child Neurology,* 30, 1147–1152.

Yapok, M. (1997). *Breaking the patterns of depression*. New York: Doubleday.

Zigman, W. B., & Lott, I. T. (2007). Alzheimer's disease in Down syndrome: Neurobiology and risk. *Mental Retardation and Developmental Disabilities Research Reviews, 13*, 237–246.

Zimmer, M., & Desch, L. (2012). Sensory integration therapies for children with developmental and behavioral disorders. *Pediatrics, 129*(6), 1186–1189.

Zinner, S. H., & Mink, J. W. (2010, June). Movement disorders I: Tics and stereotypies. *Pediatrics in Review, 31*(6), 223–233.

Return to Table of Contents

About the Authors

In 1992, Dennis McGuire, PhD, and Brian Chicoine, MD, founded the Adult Down Syndrome Center of Lutheran General Hospital in suburban Chicago. The Center, now part of Advocate Aurora Health, has served over 6,000 adolescents and adults with Down syndrome since its inception.

Brian Chicoine received his medical degree from Loyola University of Chicago Stritch School of Medicine. He completed his Family Medicine residency at Lutheran General Hospital, where he is now on the faculty. Dr. Chicoine has worked with people with intellectual disabilities in a variety of capacities for more than forty years. He is the father of three, the grandfather of one, and he lives with his wife in Arlington Heights, Illinois.

Dennis McGuire is the former director of Psychosocial Services for the Adult Down Syndrome Center. More recently he served as Senior Consultant with the Global Down Syndrome Foundation in Denver, Colorado. Dr. McGuire received his master's degree from the University of Chicago and his doctorate from the University of Illinois at Chicago. His work experience includes over forty years in the mental health and developmental disabilities field as a clinician, presenter, and writer.

Katie Frank, PhD, OTR/L, is an occupational therapist at the Adult Down Syndrome Center. Dr. Frank received her occupational therapy degree from Saint Louis University and her doctoral degree in Disability Studies from the University of Illinois at Chicago. Most of her work has been with individuals with Down syndrome of all ages. Dr. Frank has experience with treatment and evaluation as well as facilitating groups for people with Down syndrome, conducting trainings for staff, families, and caregivers, and offering a variety of other educational opportunities across the U.S. Her research is published in peer-reviewed journals.

Return to Table of Contents