



CHAPTER 4

Intellectual Disability and Developmental Disorders in Children

LEARNING OBJECTIVES

After reading this chapter, you should be able to answer these questions:

- What is Intellectual Disability and Global Developmental Delay?
- What is adaptive functioning, and why is it critical to understanding and helping children with Intellectual and Developmental Disabilities?
- How common are Intellectual and Developmental Disabilities? How does their prevalence vary by age, gender, and socioeconomic status (SES)?
- What are some known causes of Intellectual Disability in children? Why is studying children with specific behavioral phenotypes important?
- How might Intellectual and Developmental Disabilities be prevented?
- What educational interventions are available for children with Intellectual and Developmental Disabilities?
- How can behavioral and pharmaceutical treatments be used to reduce challenging behaviors in children in Intellectual Disability? What ethical issues are important to consider when selecting and implementing treatment?

If you were asked to imagine a child with an Intellectual Disability,¹ what picture would come to your mind? You might imagine a boy with very low intelligence. He might speak using simple sentences, or he might be unable to speak at all. Maybe he looks different from other boys: He has a flatter face, lower set ears, a protruding tongue, and short stature. He might be clumsy, walk in an awkward manner, or need a wheelchair to move about. He might not interact much with other children, and when he does, he might appear unusual or act inappropriately. In school, he might have a classroom aide to help him, but he still might have trouble reading sentences, learning addition and subtraction, and writing. He might be friendly but still seem “different” from most other boys his age.

For most people, our image of children with Intellectual Disability is formed by our personal experiences. We might have attended school with children who had Intellectual Disability, tutored children with developmental delays, volunteered for the Special Olympics or other recreational programs for youths with disabilities, or seen children with Intellectual Disability at the mall, where we work, or elsewhere in the community.

¹Intellectual Disability was called “Mental Retardation” in previous editions of the *DSM*. The term *Mental Retardation* is no longer used because of its negative connotation.

CASE STUDY

DONTRELL: A FRIENDLY BOY

Dontrell was a 5-year-old African American boy referred to our clinic by his pediatrician. Dontrell showed delays in understanding language, speaking, and performing daily tasks. His mother had used alcohol and other drugs during pregnancy. She did not receive prenatal care because she was afraid that an obstetrician would report her drug use to the police. Dontrell was born with various drugs in his system and had respiratory and cardiovascular problems at birth. Shortly after delivery, Dontrell's mother disappeared, leaving him in his grandmother's care.

Dontrell was slow to reach many developmental milestones. Whereas most children learn to sit up by age 6 months and walk by their first birthday, Dontrell showed delays mastering each of these developmental tasks. Most striking were Dontrell's marked delays in language. Although he could understand and obey simple commands, he was able to speak only 15 to 20 words, and many of these were difficult to understand. He could not identify colors, was unable to recite the alphabet, and could not count. He also had problems performing self-care tasks typical of children his age. For example, he could not dress himself, wash his face, brush his teeth, or eat with utensils.

Dontrell showed significant problems with his behavior. First, he was hyperactive and inattentive. Second, Dontrell showed serious problems with defiance and aggression. When he did not get his way, he would tantrum and throw objects. He would also hit, kick, and bite other children and adults when he became upset. Third, Dontrell's grandmother said that he had "an obsession for food." Dontrell apparently had an insatiable appetite and was even caught hoarding food under his bed and stealing food from relatives.

Dr. Valencia, the psychologist who performed the evaluation, was most struck by Dontrell's appearance. Although only 5 years old, Dontrell weighed almost 85 lbs. He approached Dr. Valencia with a scowl and icy stare. Dr. Valencia extended her hand and said, "Hello." Dontrell grabbed Dr. Valencia's hand and kissed it! His grandmother quickly apologized, responding, "Sorry . . . he does that sometimes. He's showing that he likes you."

Although our image of Intellectual Disability, generated from these experiences, might be accurate, it is probably not complete. Intellectual Disability is a term that describes an extremely diverse group of people. They range from children with severe developmental disabilities who need constant care to youths with only mild delays who are usually indistinguishable from others (Hodapp, Zakemi, Rosner, & Dykens, 2006).

WHAT IS INTELLECTUAL DISABILITY?

The *DSM-5* Definition of Intellectual Disability

According to the *DSM-5*, Intellectual Disability is characterized by significant limitations in general mental

abilities and adaptive functioning that emerge during the course of children's development. Limitations must be evident in comparison to other people of the same age, gender, and social-cultural background. (See Table 4.1, Diagnostic Criteria for Intellectual Disability [Intellectual Developmental Disorder].)

All individuals with Intellectual Disability must show significantly low intellectual functioning. These individuals show problems perceiving and processing new information, learning quickly and efficiently, applying knowledge and skills to solve novel problems, thinking creatively and flexibly, and responding rapidly and accurately. In children approximately five years of age and older, intellectual functioning is measured using a standardized, individually administered intelligence test. IQ scores are normally distributed with a mean of 100 and a standard deviation of 15. IQ scores approximately two standard deviations below the

Table 4.1 Diagnostic Criteria for Intellectual Disability (Intellectual Developmental Disorder)

Intellectual disability (intellectual developmental disorder) is a disorder with onset during the developmental period that includes both intellectual and adaptive functioning deficits in conceptual, social, and practical domains. The following three criteria must be met:

- A. Deficits in intellectual functions, such as reasoning, problem solving, planning, abstract thinking, judgment, academic learning, and learning from experience, confirmed by both clinical assessment and individualized, standardized intelligence testing.

(Continued)

Table 4.1 (Continued)

- B. Deficits in adaptive functioning that result in failure to meet developmental and socio-cultural standards for personal independence and social responsibility. Without ongoing support, the adaptive deficits limit functioning in one or more activities of daily life, such as communication, social participation, and independent living, across multiple environments, such as home, work, and community.
- C. Onset of intellectual and adaptive deficits during the developmental period.

Specify current severity: Mild, Moderate, Severe, Profound*

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* Table 4.2 provides a description of each type of severity.

mean (i.e., IQ < 70) can indicate significant deficits in intellectual functioning. The measurement error of most IQ tests is approximately 5 points; consequently, IQ scores between 65 and 75 are recommended as cutoffs in determining intellectual deficits (American Psychiatric Association, 2013). IQ scores below this cutoff are seen in approximately 2.5–3.0% of the population (Durand & Christodulu, 2006).

Second, individuals with Intellectual Disability show significant deficits in adaptive functioning. Adaptive functioning

refers to how effectively individuals cope with common life demands and how well they meet the standards of personal independence expected of someone in their particular age group, social-cultural background, and community setting (American Psychiatric Association, 2013). Whereas intellectual functioning refers to people's ability to learn information and solve problems, adaptive functioning refers to their typical level of success in meeting the day-to-day demands of society in an age-appropriate manner.

DSM-IV → DSM-5 CHANGES

A DIAGNOSIS BY ANY OTHER NAME

There has been considerable controversy regarding the name of the disorder "Intellectual Disability." In *DSM-IV*, this disorder was called "Mental Retardation" to reflect the below-average intellectual ability of individuals with this condition. However, the developers of *DSM-5* agreed to abandon this term because of its negative connotation. When revising *DSM-IV*, the American Psychiatric Association's Neurodevelopmental Disorders working group considered renaming the disorder "Intellectual Developmental Disorder." Their proposal drew considerable criticism from the leading professional organization of individuals who work with people with developmental disabilities, the American Association of Intellectual and Developmental Disabilities (AAIDD; Gomez & Nygren, 2012). Instead, The AAIDD argued that the name "Intellectual Disability" (not *Developmental*) be adopted in *DSM-5* for several reasons:

- *Intellectual Disability* is the most commonly used term in the United States and internationally to refer to people with intellectual and adaptive skills deficits.
- The term reflects the World Health Organization's conceptualization of low intelligence and adaptive functioning as a "disability."
- It implies deficits in both intelligence and adaptive functioning, not only low IQ.
- It is less offensive than the often pejorative term *Mental Retardation*.

In 2010, Rosa's Law (PL 111-256) replaced the term *Mental Retardation* with *Intellectual Disability* in federal education, health, and labor laws. The law was named after 9-year-old Rosa Marcellino, a girl with Down Syndrome, whose family worked to have the term *retardation* removed from the educational code in her home state of Maryland.

The APA decided to adopt the term *Intellectual Disability* in *DSM-5* yet retains the term *Intellectual Developmental Disorder* in parentheses.

DSM-5 identifies three domains of adaptive functioning: conceptual, social, and practical. These domains were identified by using a statistical procedure called factor analysis to determine groups of skills that tend to co-occur in individuals with developmental disabilities. To be diagnosed with Intellectual Disability, individuals must show impairment in at least one domain. Usually, children with Intellectual Disability experience problems in multiple areas:

- *Conceptual skills:* understanding language, speaking, reading, writing, counting, telling time, solving math problems, the ability to learn and remember information and skills
- *Social skills:* interpersonal skills (e.g., making eye contact when addressing others), following rules (e.g., turn-taking during games), social problem-solving (e.g., avoiding arguments), understanding others (e.g., empathy), making and keeping friends
- *Practical skills:* activities of daily living including personal care (e.g., getting dressed, grooming), safety (e.g., looking both ways before crossing street), home activities (e.g., using the telephone), school/work skills (e.g., showing up on time), recreational activities (e.g., clubs, hobbies), and using money (e.g., paying for items at a store)

Adaptive functioning can be assessed by interviewing caregivers about children's behavior and comparing their

reports to the behavior of typically developing children of the same age and cultural group (Tassé et al., 2012).

Often, psychologists administer a norm-referenced interview or rating scale to caregivers to collect information about children's functioning. For example, the Diagnostic Adaptive Behavior Scale (DABS) is a semi-structured interview that is administered to caregivers of children with developmental disabilities. Based on caregivers' reports, the interviewer rates children's adaptive behavior across the conceptual, social, and practical domains (see text box Research to Practice: Adaptive Functioning Examples). The DABS provides standard scores much like IQ scores, which indicate children's adaptive functioning relative to their peers. Scores more than two standard deviations below the mean (i.e., < 70) on at least one domain could indicate significant impairment in adaptive functioning (Tassé et al., 2011).

It is important to keep in mind that Intellectual Disability is characterized by low intellectual functioning *and* problems in adaptive behavior. Many people believe that Intellectual Disability is determined solely by IQ; however, deficits in adaptive functioning are equally necessary for the diagnosis. A child with an IQ of 65 but with no problems in adaptive functioning would not be diagnosed with Intellectual Disability.

RESEARCH TO PRACTICE

ADAPTIVE FUNCTIONING EXAMPLES

Clinicians assess adaptive functioning by administering semi-structured interviews to caregivers of children suspected of Intellectual Disability. Adaptive functioning scales allow clinicians to assess children's conceptual, social, and practical skills. Caregivers' reports are converted to standard scores which can be used to determine if children have deficits compared to other children their age in the general population. Below are some areas of adaptive functioning that might be assessed in younger children, older children, and adolescents.

	<i>Younger Children</i>	<i>Older Children</i>	<i>Adolescents</i>
Conceptual	Can count 10 objects, one by one; Knows day, month, year of birth	States value of penny, nickel, dime; Uses mathematical operations	Sets a watch or clock to correct time; Can complete a job application
Social	Says "hi" and "bye" when coming and going; Asks for help when needed	Reads and obeys common signs (e.g., stop, do not enter); Knows topic of group conversations	Has satisfying friendships; Keeps personal information private
Practical	Uses the restroom; Drinks from a cup without spilling	Answers the telephone; Can safely cross busy streets	Travels to school or work by themselves; Washes clothes, dishes

Finally, all individuals with Intellectual Disability show limitations in intellectual and adaptive functioning early in life. Although some people are not identified as having Intellectual Disability until they are adults, they must have histories of intellectual and daily living problems beginning in childhood. This age-of-onset requirement differentiates Intellectual Disability from other disorders characterized by problems with intellectual and adaptive functioning, such as Alzheimer's Dementia (i.e., cognitive deterioration seen in older adults).

Severity of Impairment

In the past, children with Intellectual Disability were categorized into one of four subtypes based on their IQ. This practice was abandoned in *DSM-5* for three reasons. First, the developers of *DSM-5* wanted to give equal importance to IQ and adaptive functioning in describing children with Intellectual Disability, rather than focus exclusively on IQ alone. Second,

children's IQ scores were less helpful than their level of adaptive behavior in determining their need for support and assistance at home, at school, and in the community. Third, IQ scores tend to be less valid toward the lower end of the IQ range.

Consequently, in *DSM-5*, clinicians specify the severity of Intellectual Disability based on the person's level of adaptive functioning. Adaptive functioning can be assessed using standardized rating scales, clinical interviews, and observations at home and school. Children with mild deficits in adaptive functioning (i.e., standard scores 55–70) in only one domain would presumably need less support from caregivers than children with profound deficits in adaptive functioning (i.e., standard scores <25) across multiple domains. Furthermore, by specifying the domains most in need of support, clinicians can begin to plan interventions to improve children's adaptive functioning or compensate for deficits that might be less responsive to treatment. Table 4.2 provides a general overview of children's adaptive functioning at each level of severity.

Table 4.2 Describing the Severity of Intellectual Disability

Severity	Conceptual Domain	Social Domain	Practical Domain
Mild	Preschoolers may show no obvious conceptual differences. School-aged children show difficulties in acquiring academic skills (e.g., reading, writing, arithmetic, telling time, using money). Abstract thinking and planning may be impaired; thinking tends to be concrete.	Communication, conversation, and language are more concrete or immature than the skills of peers. The child may have difficulty accurately understanding the social cues of others. There may be difficulties regulating emotion and behavior compared to peers.	The child may function in an age-expected manner with regard to personal care. In adolescence, assistance may be needed to perform more complex daily living tasks like shopping, cooking, and managing money.
Moderate	Preschoolers' language and pre-academic skills develop slowly. School-age children show slow progress in academic skills. Academic skill development is usually at the elementary school level.	The child shows marked differences in social and communicative skills compared to peers. Spoken language is simplistic and concrete. Social judgment and decision making are limited. Friendships with peers are often affected by social or communicative deficits.	The child needs more time and practice learning self-care skills, such as eating, dressing, toileting, and hygiene, than peers. Household skills can be acquired by adolescent with ample practice.
Severe	The child generally has little understanding of written language or numbers. Caretakers must provide extensive support for problem solving throughout life.	There are limited spoken language skills with simplistic vocabulary and grammar. Speech may be single words/phrases. The child understands simple speech and gestures. Relationships are with family members and other familiar people.	The child needs ongoing support for all activities of daily living: eating, dressing, bathing, elimination. Caregivers must supervise at all times. Some youths show challenging behaviors, such as self-injury.
Profound	Conceptual skills generally involve the physical world rather than symbols (e.g., letters, numbers). Some visual-spatial skills, such as matching and sorting, may be acquired with practice. Co-occurring physical problems may greatly limit functioning.	The child has limited understanding of symbolic communication. The child may understand some simple instructions and gestures. Communication is usually through nonverbal, non-symbolic means. Relationships are usually with family members and other familiar people. Co-occurring physical problems may greatly limit functioning.	The child is dependent on others for all aspects of physical care, health, and safety, although he or she may participate in some aspects of self-care. Some youths show challenging behaviors, such as self-injury. Co-occurring physical problems may greatly limit functioning.

Source: Based on *DSM-5*, 2013.

Mild Intellectual Disability (Adaptive Functioning Scores 55–70)

As infants and toddlers, children with mild Intellectual Disability usually appear no different than other children (Jacobson & Mulick, 1996). They achieve most developmental milestones at expected ages, learn basic language, and interact with family members and peers. Their intellectual deficits are usually first identified when they begin school. Teachers may notice that they require more time and practice to master academic skills, such as letter and number recognition, reading, and math. As they progress in school and their schoolwork becomes more challenging, these children fall further behind and may repeat a grade. Some children grow frustrated with traditional education and display behavior problems in class. By middle school, these children master basic reading and math but seldom make further academic progress. After school, they typically blend back into society, perform semiskilled jobs, and live independently in the community. They usually require only occasional support from others to overcome their intellectual deficits. For example, they may need help completing a job application, filing a tax return, or managing their finances.

Moderate Intellectual Disability (Adaptive Functioning Scores 40–55)

Children with moderate Intellectual Disability often show signs of their intellectual and adaptive impairments as infants or toddlers (Jacobson & Mulick, 1996). Their motor skills usually develop in a typical fashion, but parents often notice delays in learning to speak and interacting with others. These children often seem less interested in their surroundings compared to their age mates. They are often first identified as having Intellectual Disability as toddlers or preschoolers, when they show little or no language development. Instead, they rely mostly on gestures and single word utterances. By the time they begin school, these children usually speak in short, simple phrases and show self-care skills similar to typically developing toddlers. However, they display problems mastering basic reading, writing, and mathematics. By adolescence, these children are able to communicate effectively with others, have basic self-care skills, and have simple reading and writing abilities. They may continue to have trouble with reading a newspaper, performing arithmetic, or handling money. As adults, some may perform unskilled jobs if they are given training and supervision. They usually live with family members or in residential care facilities.

Severe Intellectual Disability (Adaptive Functioning Scores 25–40)

Children with severe Intellectual Disability are usually first identified in infancy (Jacobson & Mulick, 1996). They almost always show early delays in basic developmental milestones, such as sitting up and walking. They also usually show one or

more biological anomalies that are indicative of a genetic or medical disorder. These children often have health problems, are at risk for long-term motor disorders, or have seizures. They require ample supervision from parents and caregivers. By the time they begin school, they may be able to move on their own and perform some basic self-care skills, such as feeding, dressing, and using the toilet. They may communicate using single words and gestures. As adults, their speech continues to be limited and difficult to understand, although their ability to understand others is often better developed. They are usually unable to read or write, but they may be able to perform simple daily living tasks under close supervision. They typically live with family or in residential care.

Profound Intellectual Disability (Adaptive Functioning Scores <25)

Children with profound Intellectual Disability are first identified in infancy (Jacobson & Mulick, 1996). They almost always show multiple biological anomalies and health problems indicative of neurological damage. By the time they reach school age, their skills are similar to those of typically developing one-year-olds. They may be able to sit up, imitate sounds, understand simple commands, and recognize familiar people. About half of the children with profound Intellectual Disability will continue to require help from others throughout their lives. The other half will show slow development of adaptive skills. They may learn to walk, develop some communication skills, and be able to perform some self-care activities. As adults, they usually continue to require constant support and supervision from family and caregivers. They may also show chronic medical problems and sensory impairments.

Remember that the diagnosis of Intellectual Disability is determined by the child's intelligence and adaptive functioning. Two people can show Intellectual Disability but look and act very differently. For example, one person might be a child with Down Syndrome. Another child with the same IQ might have no identifiable cause of their impairments. The label "Intellectual Disability" tells us only about a person's general intellectual and adaptive functioning; the diagnosis says nothing about etiology, symptoms, course, or outcomes (Baumeister & Bacharach, 2000).

The AAIDD Definition of Intellectual Disability

The American Association on Intellectual and Developmental Disabilities (AAIDD) is the oldest professional organization devoted to the study and assistance of individuals with impairments in intellectual and adaptive functioning. The AAIDD consists of professionals and laypersons who research, help, and advocate on behalf of people with intellectual disabilities. Since 1910, they have offered guidelines for the identification of Intellectual Disability and the best methods to help children and

adults with this condition. In years past, the *DSM* and AAIDD definitions of Intellectual Disability had differed considerably. Currently, however, the *DSM-5* and AAIDD definitions overlap considerably, which will likely improve communication between members of these two professional organizations (Schalock et al., 2010).

One difference in the AAIDD conceptualization of Intellectual Disability is its emphasis on needed supports (Luckasson et al., 2002). **Needed supports** refer to a broad array of assistance that helps the individual function effectively in society. Supports can be formal assistance provided by health care providers, mental health professionals, teachers, educational specialists, professional caregivers, or human service agencies. Supports can also refer to informal help from parents, friends, or members of the community. The AAIDD designates four possible levels of supports, based on how much and how long assistance is needed: intermittent (i.e., occasional, in time of crisis), limited (i.e., short-term), extensive (i.e., long-term), and pervasive (i.e., constant).

Rather than categorize clients into mild, moderate, severe, and profound impairment, the AAIDD recommends that professionals describe individuals' need for supports across various areas of functioning. For example, a child with Intellectual Disability might be described as needing "extensive" educational support, such as a full-time classroom aide, for all academic activities, but only "intermittent" support in areas of social functioning, such as one-time training to help him learn to make friends.

The AAIDD has published a semi-structured interview to help clinicians identify the type and intensity of supports needed for adolescents and adults with Intellectual Disability

(Schalock et al., 2008). The Supports Intensity Scale measures support needs in the areas of home living, community living, lifelong learning, employment, health and safety, social activities, and protection and advocacy. It ranks each activity according to *frequency* (e.g., none, at least once a month), *amount* (e.g., none, less than 30 minutes), and *type* (e.g., monitoring, verbal gesturing) of support needed.

The AAIDD's approach to classifying individuals with Intellectual Disability in terms of needed supports has two main advantages (Schalock et al., 2008). First, this approach conveys more information about clients than simply classifying them with Intellectual Disability alone. Second, it focuses on clients' abilities rather than on their impairments. The main drawback to the AAIDD approach is that it is complex. Describing clients on so many dimensions of functioning is cumbersome and can hinder communication among professionals. The AAIDD approach can also make research difficult; with so many combinations of needed supports and areas of functioning, it is difficult to identify homogenous groups of individuals for study.

WHAT IS GLOBAL DEVELOPMENTAL DELAY?

Definition and Description

The diagnosis of Intellectual Disability requires significant deficits in intellectual functioning and adaptive skills. Typically, intellectual functioning is assessed using norm-referenced IQ tests. However, it is difficult to obtain an IQ

Image 4.1 The AAIDD conceptualization of Intellectual Disability emphasizes needed supports — assistance from others that helps the person with a disability function effectively in society.



Source: Courtesy of Kiona Miller.

score for very young children. Some tests can be administered to very young children. For example, the Wechsler Preschool and Primary Scales of Intelligence Fourth Edition (WPPSI-IV) can be given to children as young as 2 and a half years, whereas the Bayley Scales of Infant and Toddler Development Third Edition (BSID-III) is appropriate for children aged 1 to 42 months. However, these tests are usually considered measures of children’s cognitive, motor, and social development rather than intelligence per se.

The most commonly used intelligence test, the Wechsler Intelligence Scale for Children, Fourth Edition (WISC-IV), can only be administered to children 6 years old or older. Other true IQ tests, like the Stanford-Binet Intelligence Scales Fifth Edition (SB-5), can be administered to 2-year-old children. However, IQ determined prior to age 4 or 5 years is a poor predictor of IQ in childhood or adolescence (Tirosh & Jaffe, 2011). How, then, should infants and toddlers with delays be classified?

Global Developmental Delay (GDD) is a neurodevelopmental disability that is only diagnosed in children less than 5 years of age. GDD is diagnosed when the infant or child fails to meet developmental milestones in several areas. The infant or child’s physician or psychologist

suspects Intellectual Disability; however, because the child is so young, an individually administered IQ test cannot be administered. Consequently, GDD is assigned as a temporary diagnosis to indicate developmental delays until the child is old enough to participate in standardized IQ testing (American Psychiatric Association, 2013).

GDD is usually diagnosed in infants and toddlers who show significant delays in two or more of the following developmental domains: (a) fine/gross motor skills, (b) speech/language, (c) social/personal skills, and (d) daily living. *Significant delays* are defined by scores two or more standard deviations below the mean (Table 4.3). Typically, children with GDD show delays across most or all domains of functioning (Shevell, 2010).

Children with GDD are usually identified in the first year of life. Some children show physical abnormalities at birth indicative of a developmental disorder. Other children’s delays become apparent only when parents notice that their children are not developing in the same way as their peers. For example, a parent might wonder, “Why is my son not sitting up by himself at 9 months or walking at 15 months? Why hasn’t my daughter learned to say ‘mama’ and ask for her cup by 18 months?” (Shevell et al., 2003).

Table 4.3 Developmental Milestones Shown by Infants and Toddlers

Age	Motor	Language	Social	Daily Living
2 mo.	Raises head up in prone position	Differentiated cries	Smiles, follows caregiver w/ eyes	—
3 mo.	Raises head and chest; grasps object	Coos	Laughs	—
4 mo.	Rolls, stretches	—	Social smile in response to others	—
6 mo.	Sits up with support	Babbles, turn to sounds	—	Mouths objects
8 mo.	Sits up without support	Turn to the sound of own name	Stranger anxiety	—
10 mo.	Pincer grasp, crawls	Waves “bye-bye”	Peek-a-boo	Holds bottle with both hands
12 mo.	Walks but falls easily	First words	Separation anxiety	Drinks from a cup
15 mo.	Walks steadily, scribbles	Points to objects, uses single words	—	Uses spoon, helps to dress self
18 mo.	Walks up/down stairs with help; throws ball	Points to body parts when asked, two-word phrases	Plays with others	Builds small tower with blocks
24 mo.	Walks up/down stairs, kicks ball	Uses pronouns, three-word phrases	Says “no” frequently	Tries to feed self without help

Source: Based on Centers for Disease Control (2012).

Note: A pediatrician may consult a table of developmental milestones to determine if an infant is delayed in motor, language, social, or daily living skills. If she suspects delays in a particular domain, she might administer a norm-referenced test to determine the severity of the delay.

CASE STUDY

SILENT SAM

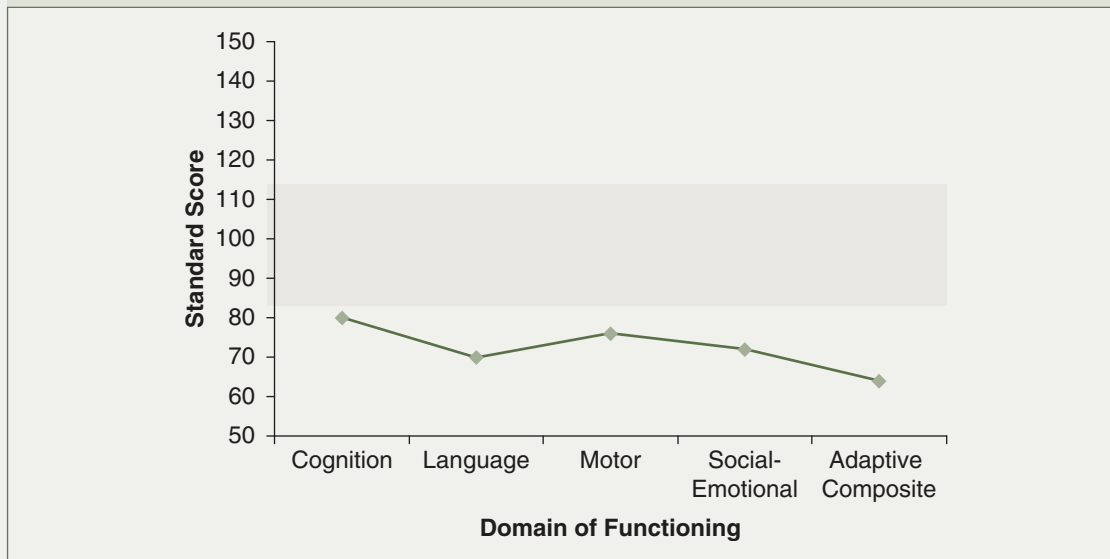
Sammy was a 34-month-old boy who was referred to our clinic by his pediatrician because of significant language delays. "I'm mostly concerned about his speech," his mother said. "Sammy has never been much of a talker. He only says a handful of words. I'm concerned because most of the kids his age in the neighborhood speak in complete sentences and he doesn't."

Dr. Baer learned that Sammy was born approximately seven weeks premature and continues to be small for his age. His motor skills tended to lag behind his peers throughout his life. When other children were learning to walk, Sammy was just beginning to crawl; when his peers began using a spoon and fork during meals, Sammy used his fingers.

Dr. Baer administered the Bayley Scales of Infant and Toddler Development to Sammy. She observed Sammy complete a series of tasks to assess his functioning and asked questions of Sammy's mother about his development and behavior at home.

Sammy's performance on the Bayley Scales showed delays in language, motor, and social-emotional skills (Figure 4.1). Overall, Sammy's functioning was more than two standard deviations lower than other children his age. Sammy's most prominent delays were in language. He showed problems with receptive vocabulary; for example, he could not correctly point to the parts of his body that Dr. Baer named or demonstrate how to use a cup, shoe, or scissors when asked. Sammy also showed delays in expressive language; he usually spoke in two-word sentences, had difficulty naming pictures of common objects (e.g., apple, bed, car), and did not use pronouns when speaking. Testing also showed similar delays in fine motor skills (e.g., putting coins in a slot), gross motor skills (e.g., climbing stairs, kicking a ball) and social-emotional functioning (e.g., pretend play, interest in peers).

Figure 4.1 Sammy's Scores on the Bayley Scales of Infant and Toddler Development-III



Note: Standard scores have a mean of 100 and standard deviation of 15. Scores between 85 and 115 (shaded area) are within normal limits. Scores approximately two standard deviations below the mean (≤ 70) indicate significant delays. Sammy would likely qualify for a diagnosis of General Developmental Delay.

"I'm really glad that you brought Sammy to see me," Dr. Baer said to Sammy's mother. "His language and motor skills are lower than what we'd expect from a boy his age. Let's work together to find some ways we can help him develop these skills."

The word "delay" in the name of this diagnosis implies that children with GDD will eventually catch up to their typically developing peers. Unfortunately, longitudinal studies of children with GDD indicate that is not always the case (Shevell, 2008). Many infants and toddlers initially diagnosed

with GDD eventually meet criteria for Intellectual Disability by the time they begin preschool. Furthermore, retrospective studies indicate that most older children with Intellectual Disability showed delays in early development that would have merited the diagnosis of GDD. Consequently, some

researchers consider GDD to be a “placeholder” diagnosis for children too young to be diagnosed with Intellectual Disability (Shevell, 2010).

Not all children with GDD have concurrent deficits in intellectual functioning, however. In one recent study, researchers examined the WPPSI scores of preschoolers with GDD. Children’s scores ranged widely and nearly 20% of children earned scores within the average range (Riou et al., 2009).

Furthermore, some young children with GDD do not develop Intellectual Disability later in life. For example, cerebral palsy is a lifelong, developmental disorder that causes marked delays in fine motor skills, gross motor skills, and (sometimes) eating, speech, and cognition (Hanna et al., 2009). Children with cerebral palsy usually show abnormal muscle tone (e.g., slouching), muscle spasms (e.g., rapid tightening of muscles that control the limbs), involuntary movements (e.g., jerks of the head, facial expressions), unsteady gait, poor balance, and noticeable joint or bone deformities (see Image 4.2). The disorder ranges in severity from mild (i.e., general clumsiness) to severe (i.e., no coordinated motor activity). Although many children with cerebral palsy also show intellectual deficits, approximately one third of children show normal intelligence (Shevell et al., 2003).

Similarly, children exposed to social deprivation or severe economic hardship can show early delays in motor, language, and cognitive development. For example, some infants adopted from developing countries have been raised in orphanages or “baby centers” with very high caregiver-to-child

ratios (Van IJzendoorn et al., 2011). Many of these children were provided with inadequate nutrition, cognitive stimulation (e.g., access to books, toys), and interactions with others. Their opportunities to develop motor skills through play and exploration may also be limited. Many of these infants and toddlers show marked delays in development across multiple domains. However, sensitive and responsible care, especially if provided before age 9 months, can remedy these skills deficits. Children who receive care early enough are usually indistinguishable from their typically developing classmates by the time they begin school.

Identifying the Causes of Global Developmental Delay

GDD is not uncommon; between 1% and 3% of infants and toddlers have the disorder (Shevell et al., 2003). In some cases, the cause of GDD can be determined based on physical examination. For example, children with Down Syndrome can be identified by certain physical attributes, such as enlarged and rounded face, wide nasal bridge, and low-set ears. In most cases, however, pediatricians or pediatric neurologists must order blood tests to screen for genetic disorders (Image 4.3). **Chromosomal microarray (CMA)** is a standard test for infants with GDD; this test identifies copy number variants (i.e., unusual duplications or deletions) in major regions of the genome. CMA can be used to create a

Image 4.2 Children with cerebral palsy, like this boy, show significant delays in fine and gross motor development. They may also display problems with speech and language and daily living skills. As infants, they often show Global Developmental Delay. However, approximately one third of these children have IQ scores within the normal range.

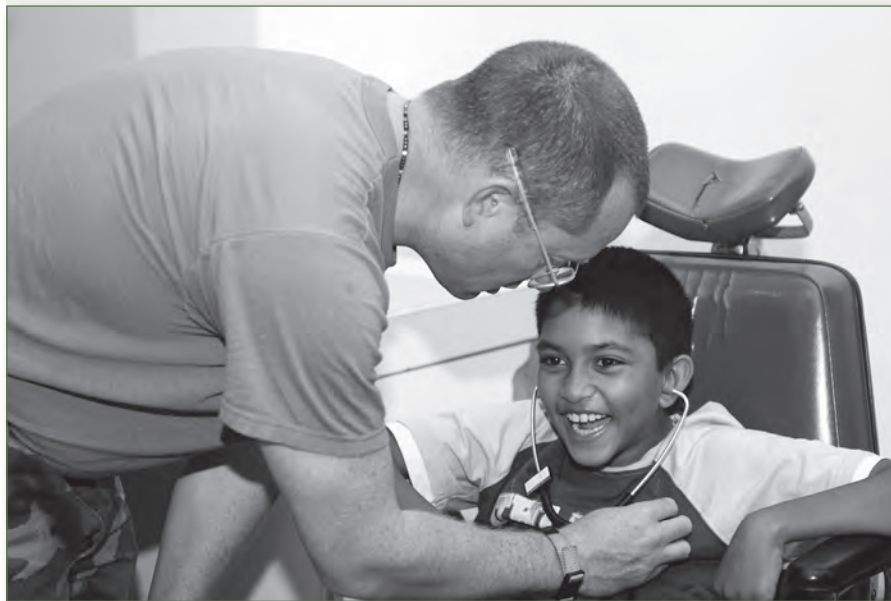
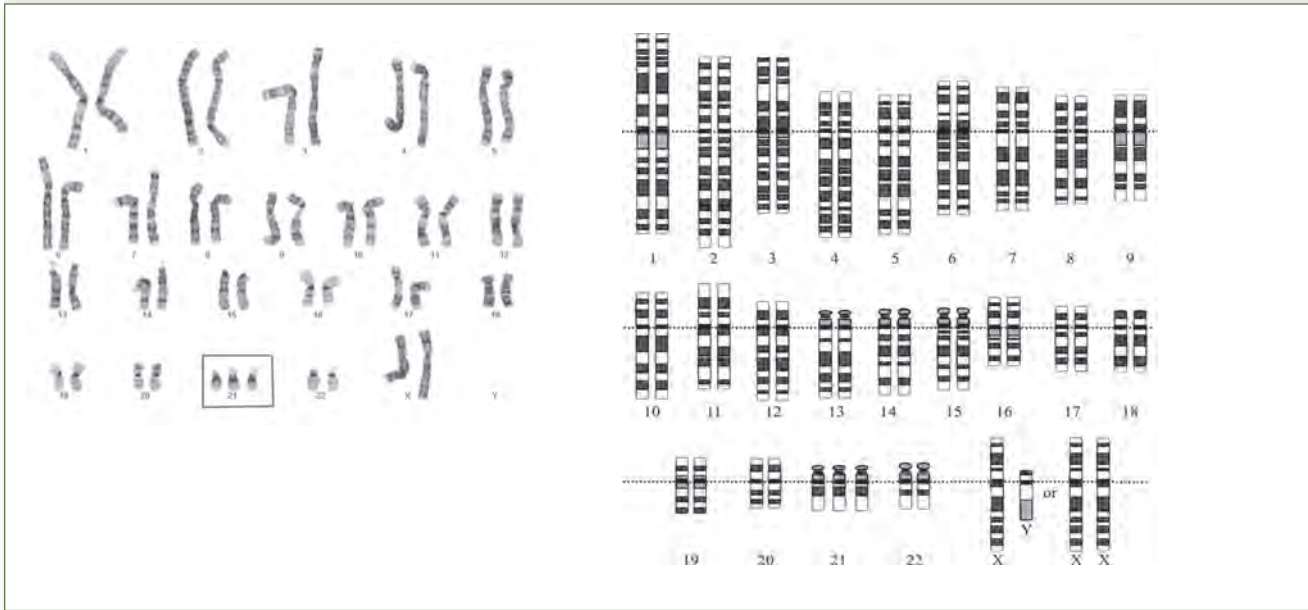


Image 4.3 Two types of genetic testing.



Note: Genetic testing is used to determine the cause of GDD. G-banded karyotyping (*left*) produces a stained image of the child's chromosomes; it is useful for detecting gross genetic abnormalities. In contrast, chromosomal microarray (CMA; *right*) screens the child's genome and generates a high-resolution "virtual karyotype" that can detect small repetitions or deletions of genetic material. These tests above show a child with three (rather than two) 21st chromosomes, the defining feature of Down Syndrome.

"virtual karyotype" of the child's chromosome structure to identify abnormalities. CMA has replaced older forms of genetic testing, such as G-banded karyotyping, which has poorer resolution and may be unable to detect more subtle chromosomal abnormalities (Flore & Milunsky, 2012). The most common genetic disorders that cause GDD are Down Syndrome, Fragile X Syndrome, Rett Syndrome (another X-linked disorder), and subtle translocations or deletions of portions of the genome. Approximately 4% of children with GDD have an identifiable genetic disorder that explains their delays (Stevenson et al., 2012).

Physicians may also order blood or urine tests to screen for metabolic disorders that can cause developmental delays. Some metabolic disorders are phenylketonuria (described later), hypothyroidism, and lead poisoning. These disorders are relatively rare; only about 1% of youths with GDD have identifiable metabolic problems.

If the results of genetic and metabolic testing are negative, physicians may try to determine the source of children's delays using neuroimaging. MRI is usually able to locate structural abnormalities in 30% to 40% of children with GDD. These abnormalities include central nervous system malformation, cerebral atrophy, problems with myelination (i.e., the coating of axons that promotes neural conduction), or cellular damage and lesions (Image 4.4).

Of course, physicians must also rule out the possibility that sensory deficits underlie children's developmental delays. Approximately 13% to 25% of children with GDD also

Image 4.4 MRI abnormality.



Note: MRI is often used to detect the cause of GDD. This newborn has a malformation of the cerebellum (i.e., Chiari malformation). Notice how the infant lacks a prominent bulge at the base of neck below the occipital lobe where the cerebellum should be. This condition is associated with delays in gross and fine motor development, problems with eating (because of nausea), weight gain, and cognition.

show vision problems, whereas 18% of children with GDD display significant hearing problems (Shevell et al., 2003). Visual and auditory deficits can greatly interfere with the acquisition of children's speech, language, and social skills.

In most cases, the causes of GDD can be determined on the basis of a parental interview, physical exam, genetic and/or metabolic testing, and neuroimaging. In order of prevalence, the most common identifiable causes are (a) genetic disorders or chromosomal abnormalities, (b) perinatal asphyxia (i.e., oxygen deprivation, perhaps during gestation or delivery), (c) brain malformation during gestation, (d) early, severe psychosocial deprivation (e.g., severe neglect), and (e) toxin exposure including maternal alcohol and other drug use (Shevell, 2008).

ASSOCIATED CHARACTERISTICS

Challenging Behavior

Approximately 25% of individuals with Intellectual Disability show challenging behavior. Experts in the field of developmental disabilities use the term *challenging behavior* to describe children's actions which are of such intensity, frequency, or duration that their physical safety (or the safety of others) is placed in jeopardy. Challenging behavior also includes actions that limit the child's access to educational or social opportunities (Didden et al., 2012).

Challenging behavior is problematic because it can affect children's health and development. Specifically, it can adversely affect children and families in several ways:

- It can be physically harmful.
- It can strain relationships with parents and cause children to be rejected by peers.
- It can limit children's access to developmentally appropriate social experiences, such as birthday parties, sleepovers, and participation in sports.
- It can interfere with learning and cognitive development.
- It can place a financial burden on families and the public.

Given their seriousness, challenging behavior is a main target for treatment.

Although children with Intellectual Disability can show many types of challenging behavior, we will focus on the most common: stereotypes, self-injurious behaviors, and aggression.

Stereotypies

Some children with Intellectual Disability show **stereotypies**, behaviors that are performed in a consistent, rigid, and repetitive manner and that have no immediate, practical significance (Carcani-Rathwell, Rabe-Hasketh, & Santosh, 2006). Stereotypies often involve repeated movements of the hands, arms, or upper body. For example, some children flap their hands, repeatedly move their fingers, twirl, fidget with objects,

or rock back and forth. Other common stereotypes are facial grimacing, face and head tapping, self-biting, and licking.

Typically developing infants and toddlers sometimes show stereotyped behaviors, such as arm waving, kicking, or swaying. Some healthy older children and adolescents continue to engage in repetitive behaviors, such as hair twirling, body rocking, and repetitive object manipulation (e.g., twirling a pencil). These behaviors are not problematic unless they come to dominate the youths' behavior, persist over time, and interfere with functioning. *DSM-5* permits clinicians to diagnose children with Stereotyped Movement Disorder when stereotypies become sufficiently impairing.

Stereotypies are fairly common among children with Intellectual Disability. In one large study, 18% of higher functioning and 31% of lower functioning children with developmental disabilities also displayed stereotypies. Moreover, 71% of youths with both Intellectual Disability and Autism Spectrum Disorder showed stereotyped behaviors (Goldman et al., 2009).

Children engage in stereotypies for many reasons (Didden et al., 2012). Certain genetic disorders are characterized by stereotyped movements. More commonly, children engage in stereotypies because these behaviors are self-reinforcing. For example, spinning in place or rocking back and forth can be pleasurable, especially in situations that might otherwise be boring (e.g., sitting at a desk, waiting in line). Still other children engage in stereotypies to regulate anxiety or frustration. For example, a child might suck his fingers or flap his arms to soothe himself or express agitation or excitement.

Self-Injurious Behaviors

Self-injurious behaviors (SIBs) involve repetitive movements of the hands, limbs, or head in a manner that can, or do, cause physical harm or damage to the person. SIBs can be classified in three ways. First, they can be described in terms of their *severity*, from mild (e.g., head rubbing, finger picking, thigh slapping) to severe (e.g., eye gouging, self-scratching, head banging). Second, SIBs can be described in terms of *frequency*, from low-occurrence acts with high potential for harm (e.g., head banging once per day) to high-occurrence acts that may cause harm over time (e.g., hand rubbing). Third, SIBs can be classified in terms of their *purpose*. Some actions seem reinforced by the reactions they elicit in others. For example, a child might gain attention from his teacher by picking his skin. Other actions appear to be reinforcing by themselves. For example, a child might insert objects into his mouth or ears because they produce positive sensations.

Approximately 10% to 12% of children with Intellectual Disability engage in SIBs (Didden et al., 2012). The prevalence of SIBs, like stereotypies, is directly related to the severity of children's intellectual and adaptive impairments. SIBs are most commonly seen in children with severe and profound impairments, children in institutional settings, and children with Autism Spectrum Disorder (Thompson &

Caruso, 2002). Indeed, children with Intellectual Disability and autism may be five times more likely than children with Intellectual Disability alone to show SIBs. Head banging and self-biting/scratching are the two most common SIBs (Kahng, Iwata, & Lewin, 2002).

SIBs usually occur in episodes or “bouts,” often occurring many times each day. Children with SIBs usually show the same behaviors in each episode (Kahng et al., 2002). In some children, episodes last only for a few seconds. These episodes are usually triggered by the environment, such as when a child with Intellectual Disability is reprimanded by a caregiver. In other children, episodes last for minutes or hours, more or less continuously. During these episodes, the child may not eat or sleep. Although these episodes may be triggered by environmental events, they are usually maintained over time by neurochemical or other biological factors (Holden & Gitlesen, 2006; Thompson & Caruso, 2002).

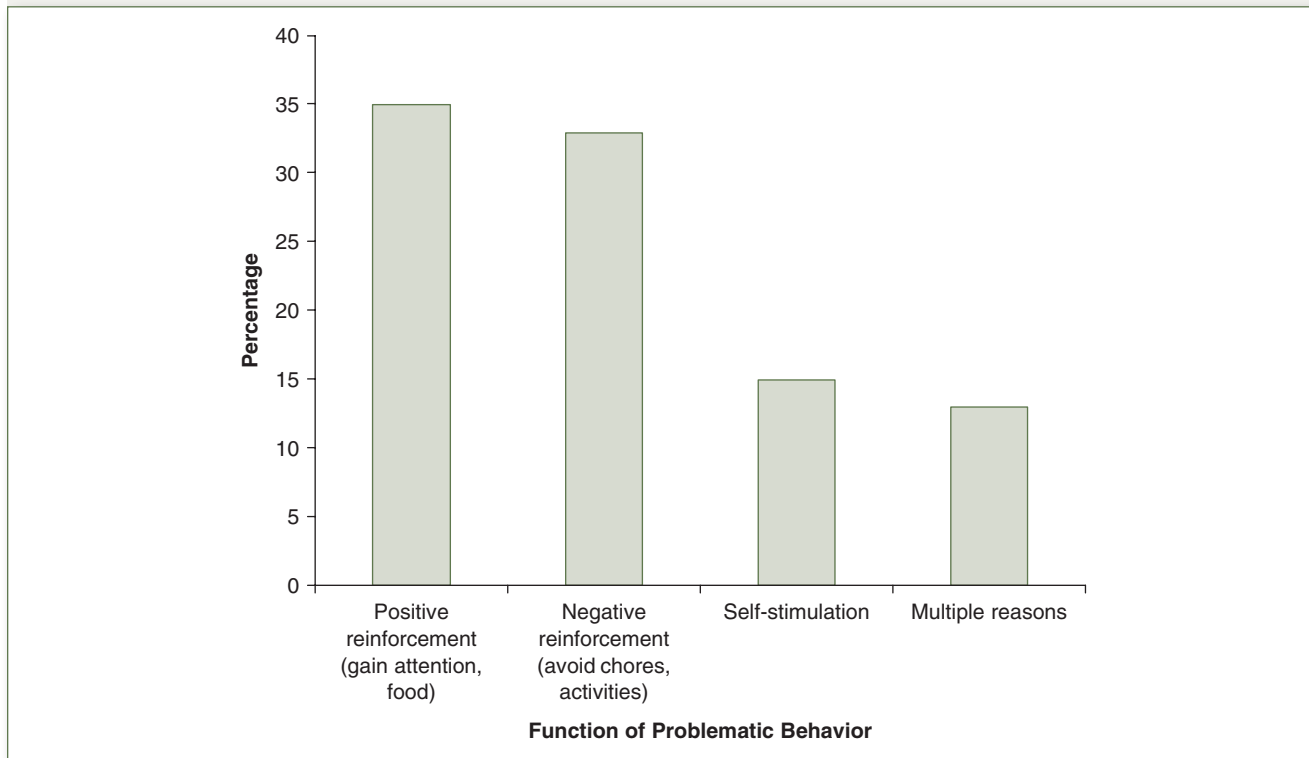
There are at least three possible explanations for SIBs in children with Intellectual Disability (Thompson & Caruso, 2002). One explanation is that children show SIBs because these behaviors serve a certain purpose or function. Carr, Levin, McConnachie, Carlson, Kemp, and Smith (1994) have suggested that individuals engage in SIBs when they lack communication or social skills to effectively interact with others. Head banging may be a way of communicating

“I don’t like this!” or “I’m bored!” To test this hypothesis, Hanley, Iwata, and McCord (2003) reviewed 536 cases of self-injurious or problematic behavior among people with Intellectual Disability. In 95.9% of cases, the SIBs served some identifiable purpose. These purposes included (a) gaining attention, food, or specific items; (b) escaping a chore, activity, or social interaction that they disliked; (c) providing stimulation or enjoyment; or (d) some combination of these three functions (Figure 4.2).

A second explanation is that SIBs are caused by a hypersensitivity to the neurotransmitter dopamine. Three lines of evidence support this hypothesis. First, destroying dopamine receptors in the brains of neonatal rats causes them to develop a hypersensitivity to dopamine. If these rats are then injected with drugs that activate dopamine in the brain (e.g., dopamine agonists like amphetamine or cocaine), they display severe self-injury. Second, healthy rats given chronic high dosages of dopamine agonists also show self-injury. Third, some antipsychotic drugs, which bind to dopamine receptors, decrease SIBs in humans.

A third possibility is that SIBs are maintained by high levels of endogenous opioids or endorphins (Schroeder et al., 2001). These naturally occurring chemicals bind to certain receptors in the brain and produce analgesia and feelings of pleasure. Children and adults who show SIBs may

Figure 4.2 Functions of Self-Injurious and Problematic Behavior Shown by People With Intellectual Disability



Source: Based on Hanley et al. (2003).

Note: More than 95% of cases of problematic behavior have some identifiable function, usually maintained by positive or negative reinforcement.

be better able to tolerate the pain because of these analgesic properties. Some individuals who show SIBs may actually derive pleasure from self-injury.

Evidence for the endorphin hypothesis comes from three sources. First, people with Intellectual Disability who show SIBs display a dramatic increase in endorphins immediately after engaging in self-harm; this increase in naturally occurring opioids is much faster than in individuals who do not show SIBs (Sandman, Hetrick, & Taylor, 1997). Second, many people with Intellectual Disability and SIBs have abnormalities in the functioning of opioid receptors and levels in their brains (Sandman, Spence, & Smith, 1999). Third, some studies indicate that SIBs can be reduced by administering drugs that block opioid receptors (i.e., opioid antagonists; Thompson & Caruso, 2002).

Physical Aggression

Youths with Intellectual Disability, like their typically developing peers, sometimes engage in aggression (Farmer & Aman, 2011). **Aggression** refers to behavior that causes (or can cause) property destruction or injury/harm to another person. Aggressive acts include throwing objects, breaking toys, ruining furniture, hitting, kicking, and biting others. Some experts also consider name-calling, screaming, and yelling a form of aggression. By definition, aggressive acts are done deliberately, not by accident. However, it is sometimes very difficult to determine the intentions of children with severe or profound Intellectual Disability (Didden et al., 2012).

Approximately one fifth of children with Intellectual Disability also show recurrent problems with aggression. Most, but not all, studies indicate that children with both Intellectual Disability and Autism Spectrum Disorder are especially likely to display aggression. Furthermore, there is often an inverse relationship between children's IQ scores and the frequency of their aggressive acts (Didden et al., 2012). Interestingly, several studies have shown that deficits in verbal IQ, communication skills, and social skills were especially predictive of aggression. Many children with Intellectual Disability engage in aggression because they lack the language and social skills to convey their thoughts and feelings in more prosocial ways (Kanne & Mazurek, 2011).

Matson and colleagues (2011) reviewed the published literature on the causes of aggression in individuals with Intellectual Disability. Most individuals showed aggression in order to avoid or escape a task, assignment, or chore. For example, a child might push, throw an object, or yell at his teacher because she asked him to put on his coat. Often, the child's aggression is negatively reinforced by the teacher withdrawing her request and allowing the child to avoid the task. Other children engage in aggression for instrumental reasons, that is, to obtain an item or privilege that they want. For example, a child might shove a classmate to access a toy. To a lesser degree, children might engage in aggression, especially property destruction, because it is self-reinforcing.

Some youths find it pleasurable to destroy objects, toys, and furniture.

Comorbid Mental Health Problems

The term *dual diagnosis* refers to the presence of mental disorders among individuals with Intellectual Disability. Until recently, many mental health professionals believed that people with Intellectual Disability could not suffer from other psychiatric disorders. Some experts believed that low intellectual functioning somehow immunized these individuals against depression, anxiety, and psychological distress. Other professionals simply did not differentiate Intellectual Disability from other mental illnesses. Gradually, clinicians became aware that people with Intellectual Disability could suffer from the full range of psychiatric disorders. In fact, the prevalence of psychopathology among individuals with Intellectual Disability may be four to five times greater than the prevalence of mental disorders in the general population (Brereton, Tonge, & Einfeld, 2006; Wallander, Dekker, & Koot, 2006).

The exact prevalence of psychiatric disorders among individuals with Intellectual Disability is unknown because their behavioral and emotional problems are often overlooked. *Diagnostic overshadowing* refers to the tendency of clinicians to attend to the features of Intellectual Disability rather than to the symptoms of coexisting mental disorders. Why might clinicians miss anxiety, depression, and even psychotic symptoms in people with Intellectual Disability? Some mental health professionals simply do not have much experience in assessing and treating people with intellectual disabilities. Others erroneously attribute psychiatric problems to the person's low intelligence or problems in adaptive functioning (Einfeld et al., 2006; Koskentausta, Iivanainen, & Almqvist, 2007).

EPIDEMIOLOGY

Prevalence

Experts disagree about the prevalence of Intellectual Disability. If we assume that IQ scores are normally distributed in the population, we would expect approximately 2.5% of individuals in the general population to earn IQ scores less than 70. Consequently, some people estimate the prevalence of Intellectual Disability to be between 2.5% and 3% of the general population (Hodapp et al., 2006).

Other experts argue that the prevalence of Intellectual Disability is lower (Tirosch & Jaffe, 2011). A meta-analysis suggested that approximately 1.83% of individuals have Intellectual Disability (Yeargin-Allsop, Boyle, & van Naarden, 2008). There are several reasons for this lower estimate. First, Intellectual Disability is not determined by the individual's IQ score alone; the diagnosis also requires impairment in adaptive functioning. Many people with IQ scores in the 55–70 range do not show significant deficits in adaptive

functioning. Consequently, they are not diagnosed with Intellectual Disability.

Second, a person's IQ can fluctuate over time. Although IQ scores are quite stable for people with severe and profound impairments, IQs are less stable for individuals scoring on the higher end of the Intellectual Disability continuum (i.e., IQ 55–70). Someone might earn an IQ score below 70 when assessed as a child but earn a score above 75 in adolescence. Consequently, he or she would no longer qualify for the diagnosis (Keogh, Bernheimer, & Guthrie, 1997).

Third, the life expectancy of individuals with severe and profound impairment is less than the life expectancy of typically developing individuals. Because of this reduced longevity, the number of people with Intellectual Disability is likely lower than expected based on the normal curve.

Age and Gender

The prevalence of Intellectual Disability varies by age. Intellectual Disability is more frequently diagnosed among school-age children and adolescents than among adults (Hodapp & Dykens, 2006). If all adults in a town are screened for Intellectual Disability, the prevalence is approximately 1.25% (McLaren & Bryson, 1987); if only school-age children are assessed, the prevalence increases to 2.5% (National Center for Educational Statistics, 2003). Why are more school-age children classified as having Intellectual Disability than people in the general population? The answer

seems to be that the cognitive impairments associated with Intellectual Disability are more noticeable when people are in school. After a person leaves school, these impairments are less noticeable, and people with them are less likely to be identified.

Intellectual Disability is slightly more common in males than in females. The gender ratio is approximately 1.3:1. Experts disagree on why males are more likely to show Intellectual Disability than females. Some people believe the male central nervous system is more susceptible to damage. Others believe that males are more likely to show Intellectual Disability than females because some forms of Intellectual Disability are caused by abnormalities on the X chromosome. Because boys have only one X chromosome, they may be more susceptible to disabilities caused by damage to this chromosome (Hodapp et al., 2006; Stromme & Hagberg, 2000).

ETIOLOGY

Organic Versus Cultural-Familial Intellectual Disability

Edward Zigler (1969) proposed one of the first methods to classify children with Intellectual Disability based on the cause of their impairments. Zigler divided children with Intellectual Disability into two groups (Table 4.4). The first group consisted of children with identifiable causes for their

Table 4.4 Intellectual Disability Classified Into Organic Versus Cultural-Familial Types

	<i>Organic</i>	<i>Cultural-Familial</i>
Definition	<ul style="list-style-type: none"> Child shows a clear genetic or biological cause for his/her Intellectual Disability 	<ul style="list-style-type: none"> Child shows no obvious cause for his/her Intellectual Disability Biological relatives may have low IQ
Diagnosis	<ul style="list-style-type: none"> Usually diagnosed at birth or infancy Frequent comorbid disorders 	<ul style="list-style-type: none"> Usually diagnosed after beginning school Few comorbid disorders
Intelligence and Adaptive Functioning	<ul style="list-style-type: none"> IQ usually ≤ 50 Siblings with normal IQ Greater impairment in adaptive functioning Often dependent on others 	<ul style="list-style-type: none"> IQ usually > 50 Siblings with low IQ Lesser impairment in adaptive functioning Can live independently with support
Associated Characteristics	<ul style="list-style-type: none"> Similar across ethnicities and SES groups Associated with health problems and physical disabilities Higher mortality rate Unlikely to mate, often infertile Often have facial abnormalities 	<ul style="list-style-type: none"> More prevalent in ethnic minorities and low-SES groups Usually few health problems and no physical disabilities Normal mortality rate Likely to marry and have children with low IQ Normal appearance

Source: Based on Iarocci and Petrill (2012).

Note: Although these names are somewhat misleading, they are useful for broadly differentiating people with intellectual disabilities.

impairments. He classified these children with **organic Intellectual Disability** because most of the known causes of Intellectual Disability at that time involved genetic disorders or biological abnormalities, such as Down Syndrome. As a group, children with organic Intellectual Disability had IQ scores less than 50, physical features indicating neurological problems, and medical complications associated with the disorder. Children with organic Intellectual Disability usually had parents and siblings with normal intellectual functioning and came from families of all socioeconomic backgrounds (Iarocci & Petrill, 2012).

Children in the second group showed no clear cause for their cognitive and adaptive impairments. They tended to earn IQ scores in the 50 to 70 range, had normal physical appearance, and showed no other health or medical problems. They were more likely to have parents, siblings, and other biological relatives with low intellectual functioning. Furthermore, they often came from low-income families. Zigler referred to individuals in this second group as experiencing “familial” Intellectual Disability because children and family members often had low levels of intellectual and adaptive functioning. Today, many experts refer to people in this category as experiencing cultural-familial Intellectual Disability because children in this group are believed to experience Intellectual Disability due to a combination of environmental deprivation (e.g., low levels of cognitive stimulation, poor schools) and genetic diathesis toward low intelligence (Iarocci & Petrill, 2012).

The terms *organic* and *cultural-familial* can be misleading. A child with organic Intellectual Disability does not necessarily have a genetic cause for his impairments. Similarly, the deficits shown by a child with familial Intellectual Disability are not necessarily caused by environmental factors. The organic/familial distinction is based solely on whether we can identify the cause of the child’s Intellectual Disability. For example, some cases of organic Intellectual Disability are caused by environmental factors, such as mothers’ consumption of alcohol during pregnancy. Similarly, some types of familial Intellectual Disability may be due to genetic anomalies that we have not yet identified. For example, the cause of Fragile X Syndrome, the second-most common genetic cause of Intellectual Disability, was not identified until 1991. Even today, as many as 80% of people with Fragile X do not know they have the disorder (Dykens, Hodapp, & Finucane, 2000). As genetic and medical research progresses, it is likely that more causes of Intellectual Disability will be uncovered (Hodapp et al., 2006).

Similar Sequence and Similar Structure

Typically developing children progress through a series of cognitive stages in a reliable order across their development. Infants learn to represent people in their minds and engage in pretend play, preschoolers show mastery of language,

school-age children develop knowledge of conservation and concrete problem solving, and adolescents show higher level abstract thinking. Zigler (1969) suggested that the sequence of cognitive development among children with Intellectual Disability is similar to the sequence of cognitive development seen in typically developing children. His **similar sequence hypothesis** posits that children with Intellectual Disability progress through the same cognitive stages as typically developing children, albeit at a slower pace.

Zigler (1969) also suggested that the cognitive structures of children with Intellectual Disability are similar to the cognitive structures of typically developing children of the same mental age. His **similar structure hypothesis** indicates that two children of the same mental age (one with Intellectual Disability and the other without Intellectual Disability) will show similar abilities. According to the similar structure hypothesis, a 16-year-old with Intellectual Disability whose intellectual functioning resembles that of a 5-year-old child should show the same pattern of cognitive abilities as a typically developing 5-year-old child.

Subsequent research on children with cultural-familial Intellectual Disability has generally supported the similar sequence and similar structure hypotheses. Children with cultural-familial Intellectual Disability show the expected sequence of cognitive development, although they reach stages at a slower rate than typically developing children (Zigler, Balla, & Hodapp, 1986). Furthermore, children with cultural-familial Intellectual Disability generally show similar cognitive abilities as children without Intellectual Disability of the same developmental age (Weisz, 1990).

Subsequent research involving children with organic Intellectual Disability has yielded mixed results. The cognitive development of children with organic Intellectual Disability does follow an expected sequence, similar to the development of typically developing children. However, children with organic Intellectual Disability often show different cognitive abilities than typically developing children of the same mental age. Specifically, children with organic Intellectual Disability often show characteristic strengths and weaknesses in their cognitive abilities; their cognitive abilities are not uniformly low. Furthermore, these cognitive strengths and weaknesses depend on the cause of the child’s Intellectual Disability. For example, children with Down Syndrome often show one pattern of cognitive abilities, whereas children with Fragile X Syndrome show different cognitive profiles.

Behavioral Phenotypes

The finding that children with different types of organic Intellectual Disability show characteristic patterns of cognitive abilities is important. If scientists could identify the cognitive and behavioral characteristics associated with each known cause for Intellectual Disability, this information could be used to plan children’s education and improve their adaptive functioning (Hodapp & DesJardin, 2002).

Consequently, researchers have moved away from lumping all children with known causes of Intellectual Disability into one large “organic” category. Instead, researchers study children with Intellectual Disability in separate groups in order to better understand the strengths and weaknesses associated with each disorder. For example, some researchers study the abilities of children with Down Syndrome while others focus on the strengths and weaknesses of children with Fragile X Syndrome (Hodapp et al., 2006).

Stated another way, researchers are interested in determining a **behavioral phenotype** for children with each known cause of Intellectual Disability. According to Dykens (1995), a behavioral phenotype involves “the heightened probability or likelihood that people with a given syndrome will exhibit certain behavioral or developmental sequelae relative to those without the syndrome” (p. 523). Behavioral phenotypes include the appearance, overall intellectual and adaptive functioning, cognitive strengths and weaknesses, co-occurring psychiatric disorders, medical complications, and developmental outcomes of children with specific causes for their Intellectual Disability. Behavioral phenotypes are probabilistic. Although not every child will show all of the features associated with the disorder, a general description might help organize and guide research and assist practitioners in developing empirically based interventions (Dykens, 2001; Hodapp & Desjardin, 2002). In the next section, we examine some of

these known causes of organic Intellectual Disability and the characteristic abilities and behaviors shown by children with each disorder.

Causes of Intellectual Disability

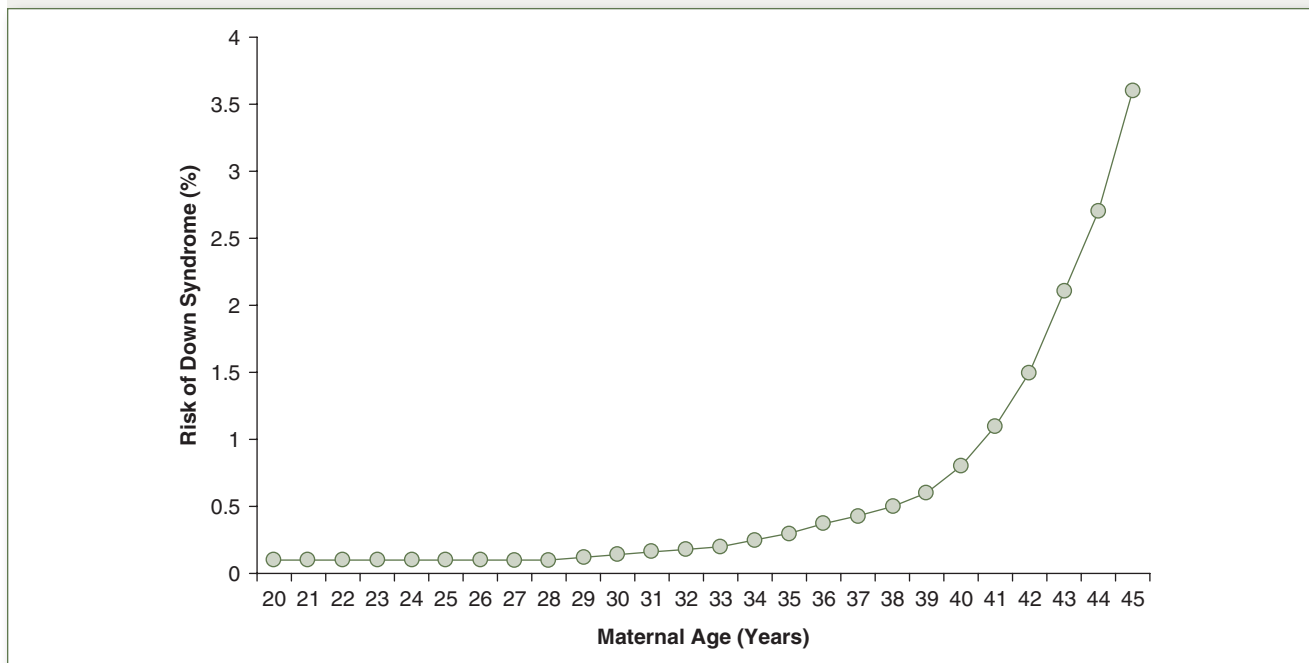
More than 750 different causes of Intellectual Disability have been identified. They can be loosely organized into five general categories: (a) chromosomal abnormalities, (b) metabolic disorders, (c) embryonic teratogen exposure, (d) complications during delivery, and (e) childhood illness or injury.

Chromosomal Abnormalities

Down Syndrome. Down Syndrome is a genetic disorder characterized by moderate to severe Intellectual Disability, problems with language and academic functioning, and characteristic physical features. The disorder was first described by John Langdon Down in 1866. It occurs in approximately 1 per 1,000 live births. The likelihood of having a child with Down Syndrome depends on maternal age (Figure 4.3).

Approximately 95% of cases of Down Syndrome are caused by an extra 21st chromosome. This form of the disorder is sometimes called “trisomy 21” because the child shows three chromosome 21s rather than the usual two. Trisomy 21 is not inherited. Instead, it is due to a **nondisjunction**, that is, a failure of the chromosome to separate during meiosis.

Figure 4.3 Risk of Down Syndrome Increases as a Function of Maternal Age



Source: Based on Cuckle, Wald, and Thompson (1987).

Note: After a woman reaches age 35, most physicians recommend prenatal screening due to increased risk.

In most cases, the mother contributes two chromosomes instead of one, but cases of paternal nondisjunction have also been reported.

Down Syndrome can also occur when the child inherits one chromosome 21 from each parent and an abnormally fused chromosome (usually consisting of chromosomes 21 and 15) from one of the parents. This abnormality, called a **translocation**, results in additional genetic material passed on to the child. It occurs in approximately 3% of youths with Down Syndrome. Down Syndrome caused by translocation is inherited. Usually, the parents are unaffected carriers of abnormally fused chromosomes, and they unknowingly pass them on to their children.

Finally, Down Syndrome can occur when some cells fail to separate during mitosis. This causes the child to have some normal cells and some cells with an abnormal amount of genetic information. The mix of normal and abnormal genetic information is called **chromosomal mosaicism**. Just as a mosaic is made up of different colored tiles, people with chromosomal mosaicism have cells of different genetic makeups. Chromosomal mosaicism accounts for approximately 2% of cases of Down Syndrome.

Children with Down Syndrome have characteristic facial features including flattened face, slanting eyes, wide nasal bridge, and low-set ears (Image 4.5). Other physical features include short stature and poor muscle tone (hypotonia). In most typically developing children, the palms show two or more large, horizontal creases. In contrast, children with Down Syndrome often show a single large crease, known as a simian crease, extending from the thumb across the palm. Children with Down Syndrome show small overall brain size and fewer folds and convolutions than in brains of typically developing children. Fewer folds suggest reduced surface area of the cortex and may be partially responsible for low intelligence (Key & Thornton-Wells, 2012).

Children with Down Syndrome are almost always diagnosed with Intellectual Disability; few of these children earn IQ scores greater than 60. Cognitive development progresses in a typical fashion for the first few months of life. After the child's first birthday, however, intellectual development slows and falls further behind typically developing peers. As a result, the delays of children with Down Syndrome become more pronounced with age.

Children with Down Syndrome show significant deficits in language (Dykens & Hodapp, 2001). They often have simplistic grammar, limited vocabulary, impoverished sentence structure, and impaired articulation. In fact, 95% of parents report difficulties understanding the speech of their children with Down Syndrome (Kumin, 1994). These children also show problems with auditory learning and short-term memory. Consequently, they often struggle in traditional educational settings where teachers present most lessons verbally (Chapman & Bird, 2012).

In contrast, children with Down Syndrome show relative strengths in visual-spatial reasoning (Dykens & Hodapp, 2001). For example, these children can repeat a series of

Image 4.5 Michelle is a 7-year-old girl with Down Syndrome, preparing for her first communion. She attended religious education classes with other children her age and received communion with her classmates.



Source: Photo is courtesy of the National Association for Down Syndrome, www.nads.org. Used with permission of her mother.

hand movements presented visually more easily than they can repeat a series of numbers presented verbally (Dykens, Hodapp, & Evans, 2006). Some experts have suggested that teachers should capitalize on these children's visual-spatial abilities in the classroom. For example, Buckley (1999) taught children with Down Syndrome how to read by having children visually match printed words with pictures, play word-matching games with flashcards, and manipulate flashcards with words printed on them into sentences. These techniques, which relied heavily on their propensity toward visual learning, led to increased reading skills. Furthermore, their advances in reading spilled over into other areas, such as speech and language.

Young children with Down Syndrome are usually described as happy, social, and friendly. It is extremely rewarding to volunteer as a tutor for a child with Down Syndrome because these children are often socially

outgoing and affectionate. Children with Down Syndrome are less likely to develop psychiatric disorders than other children with Intellectual Disability (Dykens et al., 2006). However, in adolescence they may experience emotional and behavioral problems due to social isolation or increased recognition of their impairments (Reiss, 1990).

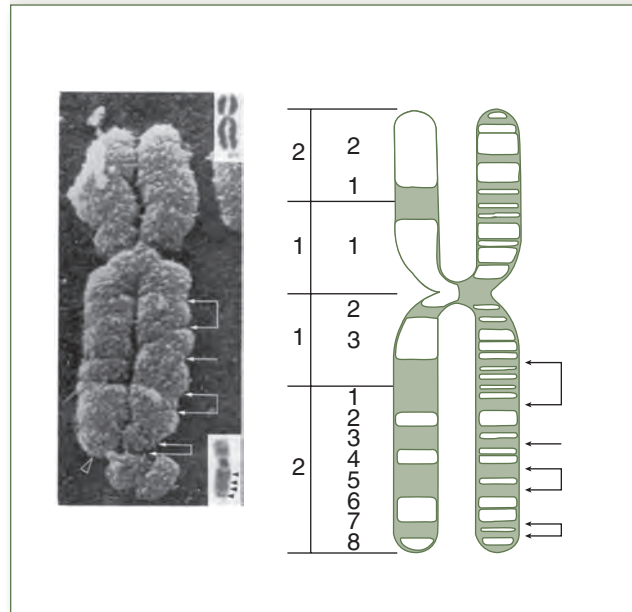
Medical complications associated with Down Syndrome include congenital heart disease, thyroid abnormalities, respiratory problems, and leukemia (Chase, Osinowo, & Pary, 2002). After age 35, many adults with Down Syndrome show early symptoms of Alzheimer’s Disease (Coppus et al., 2006). Postmortem studies of their brains have shown a high incidence of neurofibrillary tangles and plaques, similar to those shown by older adults with Alzheimer’s Disease. The life expectancy for individuals with Down Syndrome is approximately 60 years.

Fragile X Syndrome. Fragile X Syndrome is an inherited genetic disorder that is associated with physical anomalies, moderate to severe intellectual impairment, and social/behavioral problems. It occurs in 1 per 1,500 live births—about 1 per 1,000 males and 1 per 2,000 females (Sadock & Sadock, 2003).

Fragile X Syndrome is caused by a mutation in a gene on the X chromosome, called the **Fragile X Mental Retardation 1 (FMR1) gene** (Cornish, Bertone, Kogan, & Scerif, 2012). In healthy individuals, this gene contains a three-nucleotide sequence of cytosine-guanine-guanine (CGG) that repeats a small number of times. It produces Fragile X Mental Retardation Protein (FMRP), which assists in normal brain maturation and cognitive development (Comery, Harris, Willems, Oostra, & Greenough, 1997). Children with Fragile X Syndrome show an unusually high number of CGG repeats. Children who inherit 50 to 200 repeated sequences usually show no symptoms. They are typically unaware that they carry the genetic mutation, but they may pass this mutation on to their offspring. Children who inherit more than 200 repeated sequences usually show symptoms. The repeated sequences interfere with the functioning of the FMR1 gene and, consequently, decrease the amount of FMRP produced (Pieretti et al., 1991). The disorder is called “fragile” X because the X chromosome appears broken (Image 4.6). In general, the less FMRP produced, the more severe children’s cognitive impairments (Tassone et al., 1999). Brain scans of children with Fragile X show abnormalities of the prefrontal cortex, caudate nucleus, and cerebellum, presumably from less FMRP production (Stevenson et al., 2012).

Boys and girls differ in their presentation of Fragile X Syndrome, with boys showing greater intellectual impairment, more severe behavior problems, and more physical anomalies. Boys show relatively greater impairment because they inherit only one affected X chromosome. Girls, on the other hand, inherit one affected X chromosome and a second X chromosome, which is typically unaffected. The additional unaffected X chromosome produces normal amounts

Image 4.6 Fragile X Syndrome is caused by the FMR 1 gene located at the lower long arm of the X chromosome. This genetic abnormality gives the chromosome a “fragile” or broken appearance.



of FMRP and contributes to higher cognitive functioning (Cornish et al., 2012).

Boys with Fragile X tend to have elongated heads, large ears, hyper-flexible joints, and large testicles after puberty (Sadock & Sadock, 2003). They also tend to be shorter than other boys. Medical problems sometimes associated with Fragile X include heart murmur and crossed eyes (Image 4.7).

Image 4.7 Blake is a preschool-age boy with fragile X syndrome. His parents describe him as “a sweet and spunky little guy.”



Source: Used with permission from the National Fragile X Foundation, www.fragilex.org.

Boys with Fragile X Syndrome tend to show moderate to severe Intellectual Disability (Abbeduto, McDuffie, Brady, & Kover, 2012). Additionally, they show a curious pattern of strengths and weaknesses in the way they process information and solve problems (Alanay et al., 2007). They perform relatively well on tasks that require **simultaneous processing**, that is, perceiving, organizing, and interpreting information all at once. Solving puzzles or completing mazes demands simultaneous processing. Alternatively, boys with Fragile X Syndrome show relative deficits in **sequential processing**, that is, the capacity to arrange and process information in a certain order. Reading a sentence or following instructions on how to assemble a toy requires sequential processing. These boys also show weakness in planning and organizing activities in an efficient manner (Loesch et al., 2003).

Boys with Fragile X also tend to show characteristic patterns of behavior. Most notably, many show autistic-like behavior, such as a reluctance to make eye contact or be touched by others. However, only about 25% of boys with Fragile X meet diagnostic criteria for autism. The rest appear extremely shy in social situations. Many boys with Fragile X also display hyperactivity and inattention. Perhaps as many as 90% have ADHD (Dykens & Hodapp, 2001; Sullivan, Hooper, & Hatton, 2007).

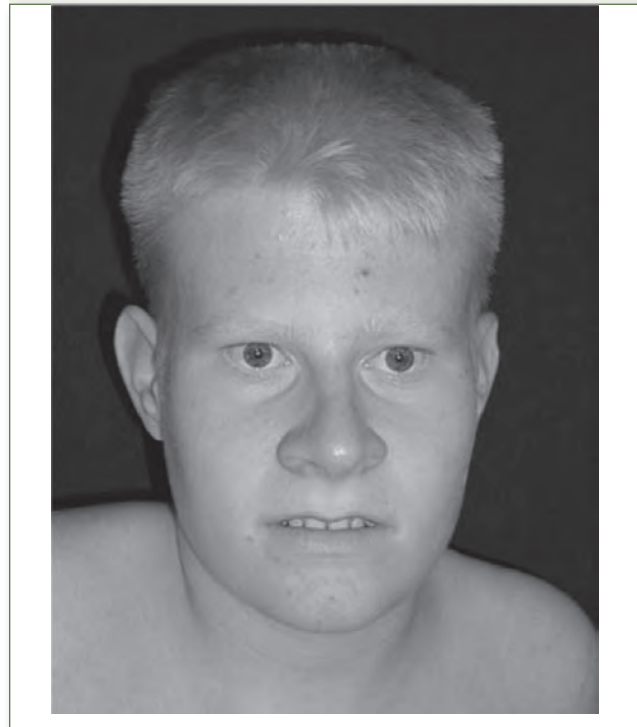
Girls with Fragile X tend to show higher IQs, less noticeable physical anomalies, and less severe behavior problems than do boys with the disorder. Like boys, girls may have problems with visual-spatial abilities (Cornish, Munir, & Cross, 1998) and inattention (Mazzocco, Pennington, & Hagerman, 1993). They may also show excessive shyness, gaze aversion, and social anxiety (Hodapp & Dykens, 2006).

Prader-Willi Syndrome. Prader-Willi Syndrome (PWS) is a non-inherited genetic disorder characterized by mild Intellectual Disability, overeating and obesity, oppositional behavior toward adults, and obsessive-compulsive behavior (Image 4.8). PWS occurs in 1 per 20,000 live births (Dykens & Shah, 2003).

PWS is usually caused by the deletion of genetic information on portions of chromosome 15. In 70% of cases, the father's information is deleted, so the child inherits only one set of genetic information, from the mother. In most of the remaining cases, the mother contributes both pairs of chromosome 15 (called **maternal uniparental disomy** [UPD]). In both instances, the father does not contribute the significant portion of chromosome 15, resulting in missing paternal genetic information.

Individuals with PWS show either mild Intellectual Disability or borderline intellectual functioning. Average IQs range from 65 to 70 (Dykens & Shah, 2003). These children show characteristic strengths and weaknesses on various cognitive tasks. For example, children with PWS show relative strengths on visual-spatial skills. Indeed, some children with PWS may be able to solve jigsaw puzzles faster than the psychologists who test them (Dykens & Cassidy, 1999). On the other hand, these children show weaknesses in short-term memory. Their adaptive behavior is usually much lower than

Image 4.8 This 15-year-old boy has Prader-Willi Syndrome, a genetic disorder characterized by Intellectual Disability, narrow temples, elongated face, thin upper lip and prominent nose.



Source: Used with permission.

their IQ because their disruptive behavior often interferes with their acquisition of daily living skills.

The most striking feature of many children with PWS is their intense interest in food (Dykens, 2000; Dykens & Cassidy, 1999). Infants with the disorder show problems with sucking, feeding, and weight gain. However, between 2 and 6 years of age, children with PWS eat enormous amounts of food (i.e., **hyperphagia**). Some evidence suggests that these children have abnormal neural functioning in the paraventricular nucleus of the hypothalamus, the area of the brain that controls hunger and satiety. Since these children never feel full, they eat to excess and are often preoccupied by food. Some children steal food, hoard food, or obtain food from the garbage in an attempt to satisfy themselves. If their diet is not monitored, they will eat to excess and become obese. Medical complications associated with obesity are a leading cause of death among adults with PWS (Dykens & Shah, 2003).

The onset of hyperphagia is also associated with changes in behavior. Many (70%–95%) children with PWS become argumentative, defiant, and throw temper tantrums (Dykens & Cassidy, 1999; Dykens & Kasari, 1997). Approximately 42% of children with PWS destroy property during their disruptive outbursts, while 34% physically attack others (Dykens, Cassidy, & King, 1999).

Most (71%–98%) children with PWS show obsessive thoughts or ritualistic, compulsive behaviors (Dykens & Cassidy, 1999). The most common obsessions concern food (Dykens & Cassidy, 1999). They may eat foods in a certain order or according to color, texture, type, or caloric content (Dykens, 2000). Children with PWS often show nonfood obsessions and compulsive behavior, too. They may hoard paper and pens; order and arrange toys and household objects by color, size, or shape; repeat information or questions; appear overly concerned with symmetry; and redo activities (e.g., untying and tying shoes, rewriting homework) until the behavior is done exactly right (Dimitropoulos, Feurer, Butler, & Thompson, 2001). Most individuals with PWS pick their skin, usually on their head or legs (Dykens & Cassidy, 1999).

In early adulthood, some individuals with PWS show psychotic symptoms, including distorted thinking and hallucinations. In one study, 12.1% of parents of children with PWS reported auditory or visual hallucinations in their children (Stein, Keating, Zar, & Hollander, 1994). Life expectancy among adults with PWS is usually somewhat reduced because of obesity.

Angelman's Syndrome. Angelman's Syndrome is a genetically based developmental disorder characterized by Intellectual Disability, speech impairment, happy demeanor, and unusual motor behavior. The disorder was identified by the English physician Harry Angelman when three children (later diagnosed with the syndrome) were admitted to his hospital at the same time. All three children showed severe Intellectual Disability, an inability to speak, and problems with gait and balance. Their movements were sporadic, jerky, and irregular. They tended to walk with arms uplifted, sometimes on their toes, lurching forward with abrupt starts and stops. Most strikingly, all three children frequently smiled and laughed (Clayton-Smith, 2001; Dykens & Shah, 2003).

Later, while visiting the Castelvecchio Museum in Verona, Angelman saw a painting titled *Boy With a Puppet* that reminded him of the happy disposition of his three young patients (Image 4.9). Angelman subsequently wrote a scientific paper describing his three "Puppet Children," which slowly attracted the attention of clinicians throughout the world (Angelman, 1965). Today, professionals refer to the disorder as Angelman's Syndrome. Approximately 1 per 15,000 to 20,000 children have the disorder.

Both PWS and Angelman's Syndrome are caused by abnormalities on portions of chromosome 15. Healthy children inherit two chromosome 15s, one from each parent. PWS occurs when children inherit genetic information on chromosome 15 only from the mother. In contrast, Angelman's Syndrome occurs when children inherit genetic information on chromosome 15 only from the father. In 70% of cases of Angelman's Syndrome, genetic information from the mother is deleted. In another 3% to 5% of cases, the father contributes two chromosome 15s and the mother contributes none. In

Image 4.9 *Boy With a Puppet*, Giovanni Francesco Caroto (1520). This oil painting prompted Angelman to write the first scientific account of children with the disorder that now bears his name. The smile of the boy in the painting reminded Angelman of his three patients.



the remaining cases of Angelman's Syndrome, the child shows other genetic mutations in chromosome 15 or the cause is unknown.

The most striking feature of Angelman's Syndrome is the persistent social smile and happy demeanor shown by children with the disorder. Many infants with Angelman's Syndrome begin this persistent smiling between 1 and 3 months of age. Later in development, it is accompanied by laughter, giggling, and happy grimacing. Facial features of children with Angelman's Syndrome often include a wide smiling mouth, thin upper lip, and pointed chin (Williams, 2005).

Despite children's social smiling, Angelman's Syndrome is usually not recognized until toddlerhood. Parents and physicians often suspect the disorder when children continue to show cognitive impairment, lack of spoken language, and movement problems. By childhood, children's intellectual functioning is generally in the range of severe or profound Intellectual Disability. Youths with the disorder show levels of functioning similar to a 2-and-a-half to 3-year-old child.

Most children with Angelman's Syndrome are unable to speak, although some can use a few words meaningfully. They usually can understand other people and are able to obey simple commands.

Nearly all children with Angelman's Syndrome show hyperactivity and inattention. Parents usually describe them as constantly "on the go." Children may flap their arms, fiddle with their hands, and become easily excited. Hyperactivity often interferes with their ability to sleep. Children with Angelman's Syndrome often have difficulty sustaining their attention on one person or task for long periods of time. Problems with hyperactivity and inattention continue throughout childhood but decrease somewhat with age.

Some children with Angelman's Syndrome show skin and eye hypopigmentation; that is, they may appear pale and have light-colored eyes. Hypopigmentation occurs when the gene that codes for skin pigmentation is deleted along with the other information on chromosome 15 that causes Angelman's Syndrome. Other children with Angelman's Syndrome show feeding problems in infancy. They may thrust their tongues outward when fed, have difficulty sucking and swallowing, or drool. In most cases, these problems resolve over time.

More than 90% of children with Angelman's Syndrome have seizures. Sometimes, seizures are difficult to notice because of these children's sporadic motor movements. In most cases, physicians prescribe anticonvulsant medications to reduce the number and severity of seizures. Adults with Angelman's Syndrome have life expectancies approximately 10 to 15 years shorter than typically developing individuals. Life expectancy is dependent on the severity of comorbid medical problems, especially seizures.

Williams Syndrome. Williams Syndrome (WS) is a genetic disorder characterized by low intellectual functioning, unusual strengths in spoken language and sociability, hyperactivity, impulsivity, and inattention (Elsabbagh & Karmiloff-Smith, 2012). Children with WS can be identified by their facial features. They often have broad foreheads; full lips; widely spaced teeth; star-shaped patterns in their irises; and elfin-like noses, eyes, and ears (Image 4.10). Their facial features suggest a mixture of joy and mischievousness. WS is caused by a small deletion in a portion of chromosome 7. The disorder occurs in approximately 1 per 20,000 live births.

Despite their low IQ scores, children with WS show curious strengths in certain areas of language (Mervis, 2012). They have unusually well-developed lexicons (Hodapp & Dykens, 2006). They can tell relatively complex stories with advanced vocabulary and sophisticated grammar (Reilly, Klima, & Bellugi, 1990). They may even use sound effects when telling stories to add emphasis. Some children with WS show relative strengths in auditory memory and music (Hodapp & DesJardin, 2002). Teachers sometimes alter their instructional methods to play to the strengths of children with WS. For example, children with WS might respond best

Image 4.10 Mary is a preschool-age girl with Williams Syndrome. She shows a persistent happy demeanor, characteristic of children with the disorder.



Source: Reprinted with permission of Mary's family.

to verbal instruction rather than to reading and may prefer to work with partners or in groups, rather than independently (Hodapp & DesJardin, 2002).

Children with WS do poorly on visual-spatial tasks. They have great difficulty copying pictures or figures. This relative deficit in visual-spatial abilities is likely due to the genetic deletion that causes WS. Specifically, the portion of chromosome 7 that is deleted contains a gene that codes for an enzyme called LIM kinase. This enzyme is necessary for brain development and functioning, especially in brain regions responsible for visual-spatial processing. Deficits in this enzyme likely underlie the visual-spatial problems shown by children with the disorder (Elsabbagh & Karmiloff-Smith, 2012).

Children with WS are described as friendly and sociable (Rosner, Hodapp, Fidler, Sagun, & Dykens, 2004). They are especially good at remembering faces and inferring a person's mental state and emotions based on his or her affect (Tager-Flusberg, Boshart, & Baron-Cohen, 1998). Sometimes, they are overly trusting of strangers, placing them at risk for exploitation by others.

Children with WS often show problems with high-rate behavior and are easily excitable. They display inattention, hyperactivity, and impulsivity; many are diagnosed with ADHD (Einfeld, 2005). Furthermore, children with WS show hyperacusis, that is, an unusual sensitivity to loud noises. Truck engines, fire alarms, and school bells can cause them considerable distress.

Most children with WS show problems with anxiety. Like typically developing children, young children with WS fear tangible and imposing images and events such as storms, vaccinations, and ghosts. However, unlike typically developing children, older children with WS continue to fear these stimuli and show a marked increase in generalized anxiety. In particular,

older children with WS often fear that something bad is about to happen. Many are extremely sensitive to failure and criticism by others (Dykens, 2003). In a sample of 51 individuals with WS, 35% showed full-blown phobias for objects or social situations while 84% showed subthreshold problems with anxiety. In contrast, the prevalence of phobias among children with other types of Intellectual Disability is only about 1% (Landau, 2012)

Dykens and Hodapp (2001) have suggested that the characteristic features of WS may place them at increased risk for developing anxiety problems. For example, their hyperacusis may make them especially sensitive to developing fears of loud noises. Early problems with balance and gait might contribute to fears of falling from high places. Their social sensitivity may place them at increased risk for social anxiety. Consequently, the fears of children with WS may stem from the interaction of genotype, early experiences, and the behavioral characteristics of WS (Dykens & Hodapp, 2001).

Children with WS are at risk for cardiovascular problems. The portion of chromosome 7 that is deleted in WS

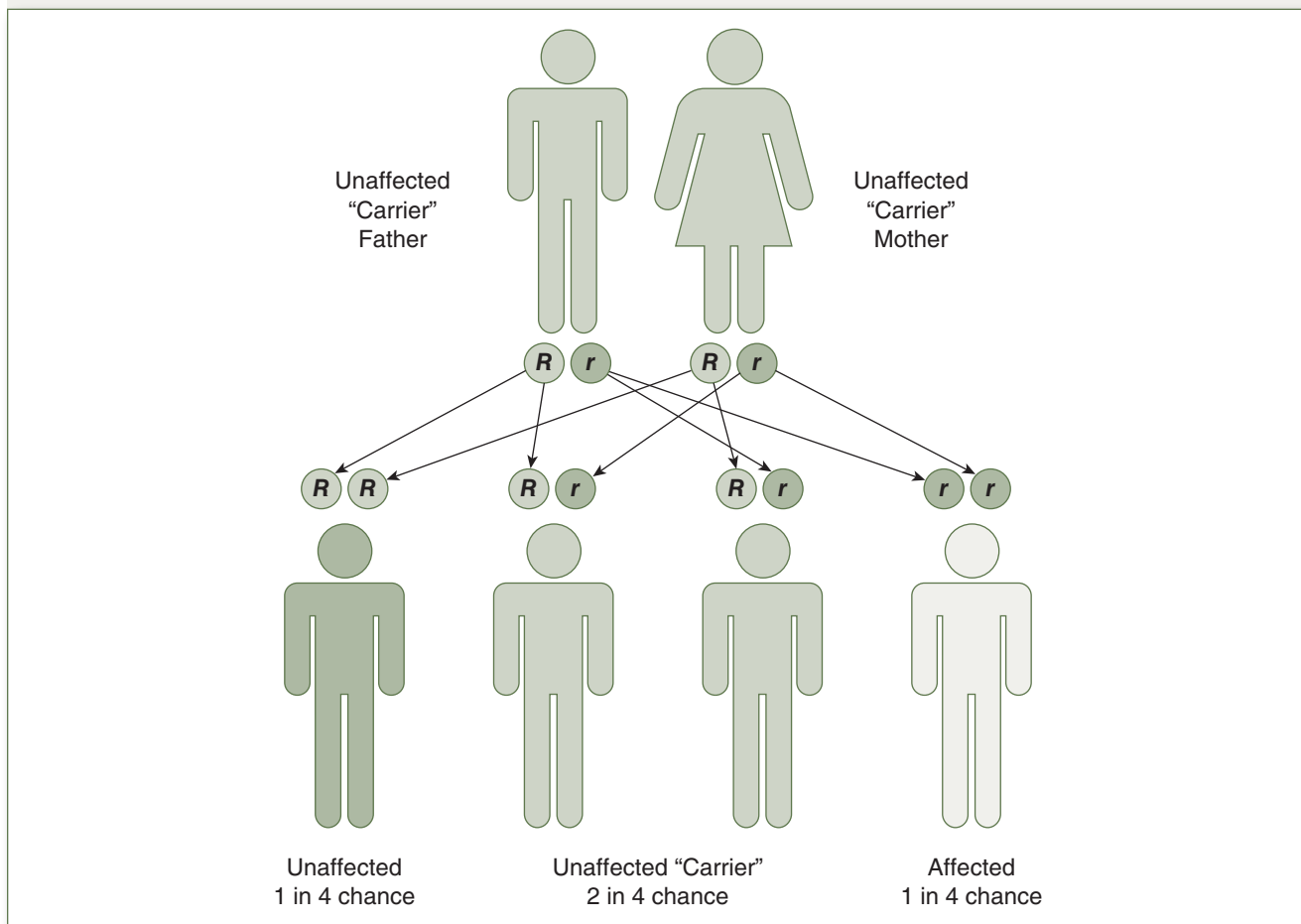
also contains a gene that codes for elastin. Elastin is used by the cardiovascular system to give connective tissue its elastic, flexible properties. Insufficient elastin can cause hypertension, other cardiovascular diseases, and early death.

Metabolic Disorders

Phenylketonuria (PKU) is a metabolic disorder that is caused by a recessive gene inherited from both parents. In most cases, PKU is characterized by the body's inability to convert **phenylalanine**, an essential amino acid found in certain foods, to paratyrosine. In PKU, the enzyme that breaks down phenylalanine (phenylalanine hydroxylase) is not produced by the liver. As the child eats foods rich in phenylalanine, such as dairy, meats, cheeses, and certain breads, the substance builds up and becomes toxic. Phenylalanine toxicity eventually causes brain damage and Intellectual Disability.

PKU is caused by a recessive gene (Figure 4.4). In order for a child to show PKU, he or she must inherit the gene from

Figure 4.4 PKU Is an Autorecessive Metabolic Disorder



Note: Children must inherit the recessive gene from both parents to develop the disorder. If children inherit only one recessive gene, they will carry the disorder but not show symptoms.

both mother and father. Children who inherit the gene from only one parent are carriers of the disorder but do not display symptoms. If a carrier mates with another carrier, each offspring has a 25% chance of showing PKU. The disorder occurs in approximately 1 per 11,500 children.

Newborns are routinely screened for PKU through a blood test conducted shortly after birth. If the disorder is detected, the child is placed on a diet consisting of foods that are low in phenylalanine. The diet decreases the chances of toxicity; consequently, adherence to the diet results in normal intellectual development. Most physicians suggest the diet should be continued indefinitely. Since phenylalanine is an essential amino acid, children on the diet must be monitored by their pediatricians. They are at risk for low red blood cell count (anemia) and low blood glucose levels (hypoglycemia).

Youths with PKU who do not diet show symptoms several months after birth. By childhood, they often develop severe intellectual impairment. Children with untreated PKU are often hyperactive, show erratic motor movements, and throw tantrums. They also may vomit and have convulsions. They usually cannot communicate with others. These impairments are irreversible, even if a phenyl-free diet is initiated later in childhood.

Embryonic Teratogen Exposure

Intellectual Disability can also occur when children are exposed to certain environmental toxins during gestation. The placenta is a bag-like membrane that partially surrounds the fetus during gestation. It delivers oxygen and nutrients from the mother to the fetus and allows the fetus to excrete waste. The placenta is porous; substances ingested by the mother can pass directly to the fetal system. **Teratogens** are environmental substances that cause maldevelopment in the fetus, often resulting in Intellectual Disability.

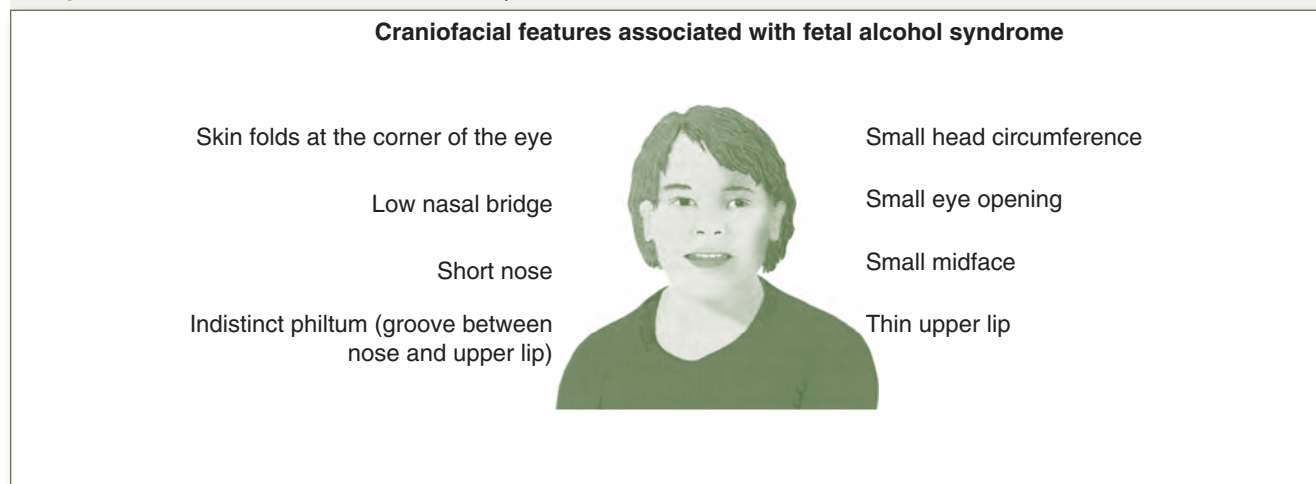
Maternal Illness. Viruses acquired by the mother during pregnancy used to be a leading cause of organic Intellectual Disability. Exposure to the **rubella virus**, especially during the first few months of pregnancy, often caused severe Intellectual Disability, cataracts, and deafness. Similarly, **maternal syphilis** was associated with fetal maldevelopment and Intellectual Disability. Other diseases, such as measles, mumps, diphtheria, tetanus, and poliovirus, can also cause Intellectual Disability. Today, these illnesses are largely prevented by childhood immunizations and regular medical care.

Infants can acquire **human immunodeficiency virus** (HIV) from an infected mother either in utero or through breast feeding. HIV causes damage to the child's central nervous and immune systems. The risk of transmission from an infected mother is approximately 30%. However, zidovudine (Retrovir), taken prenatally by mothers, can reduce the likelihood of transmission to less than 5% (Sadock & Sadock, 2003). The progression of HIV in newborns is more rapid than in adults. Most infants born with HIV show progressive brain degeneration, Intellectual Disability, and seizures during their first year of life. Most affected children die before age three.

Maternal Substance Use. Many drugs, if ingested by pregnant women, are associated with low birth weight, small head circumference, and increased risk for behavioral and learning problems in childhood. Interestingly, "hard" drugs, such as heroin and cocaine, are not as consistently associated with children's Intellectual Disability as are more socially accepted drugs like alcohol.

Fetal Alcohol Syndrome (FAS) is caused by maternal alcohol consumption during pregnancy. FAS is characterized by Intellectual Disability, hyperactivity, and slow physical growth as well as characteristic craniofacial anomalies (Figure 4.5). Children with FAS often have cardiac problems. By school age, they tend to show hyperactivity and learning problems.

Figure 4.5 Children With Fetal Alcohol Syndrome



Note: Children with Fetal Alcohol Syndrome often have facial anomalies as well as low intellectual functioning.

The prevalence of FAS is approximately 1 to 3 per 1,000 live births. However, among children of women who have Alcohol Use Disorder, the prevalence is approximately 1 in 3. Experts disagree on how much alcohol must be consumed to produce FAS. Some data indicate that FAS can occur from only 2 to 3 oz. of alcohol per day during gestation. Furthermore, binge drinking during pregnancy greatly increases the chance of FAS. Although occasional consumption of alcohol during pregnancy may not produce FAS, it may lead to subtle cognitive, behavioral, and physical abnormalities, such as mild learning problems, reduced attention span, or short stature. Most physicians recommend abstaining from alcohol entirely during pregnancy.

The intellectual functioning of children with FAS varies considerably. Most children with FAS show mild to moderate Intellectual Disability, although some earn IQ scores within the borderline to low-average range. These children usually have academic problems and may drop out of school. Many have learning disabilities. The most common behavioral problems associated with FAS are hyperactivity, impulsivity, and inattention. Young children with FAS are often diagnosed with ADHD. Older children and adolescents with FAS report feelings of restlessness and difficulty sustaining attention on reading and other homework.

Children with FAS are at risk for mood problems as they enter late childhood and adolescence. They may become depressed because of their academic deficits, behavior problems, or stigmatization associated with the disorder.

Complications During Pregnancy and Delivery

Complications that occur during gestation or delivery can contribute to Intellectual Disability. Maternal hypertension or uncontrolled diabetes during pregnancy are sometimes associated with Intellectual Disability in children. Delivery complications that interfere with the fetus's ability to obtain oxygen for extended periods of time (**anoxia**) can also lead to central nervous system damage and Intellectual Disability. For example, anoxia can occur when the umbilical cord wraps around the fetus's throat, interfering with oxygen intake.

Children born before 36 weeks gestation are also at risk for deficits in intellectual and adaptive functioning in infancy and early childhood. Two meta-analyses have shown an inverse relationship between premature birth and children's subsequent IQ scores (Figure 4.6). Furthermore, a very large, community-based study found that the risk of developmental delays increased exponentially with decreasing gestational age. Approximately 4.2% of full-term infants showed motor, language, social, or daily living delays compared to 37.5% of infants born at 24 to 25 weeks gestation. Controlling for other biological and social factors, such as mother's age and education, did not affect this relationship between prematurity and developmental risk (Kerstjens et al., 2013).

Preterm birth is a risk factor because of the rapid growth of the central nervous system during the third trimester. Between 24 and 40 weeks gestation, the fetus's cortical volume increases fourfold. There is a dramatic increase in the number of neurons,

CASE STUDY

ANDREW: A BOY WITH FAS

Andrew was a 14-year-old boy with FAS who was referred to our clinic because of a marked increase in disruptive behavior at school. Andrew was a large boy, approximately 5 feet, 10 inches and more than 160 lbs. He displayed many of the physical features of youths with Fetal Alcohol Syndrome, including wide-spaced eyes, upturned nose, low-set ears, and broad face. Andrew's mother had an extensive history of alcohol and other drug dependence. She drank throughout her pregnancy with Andrew and was intoxicated at the time of his delivery.

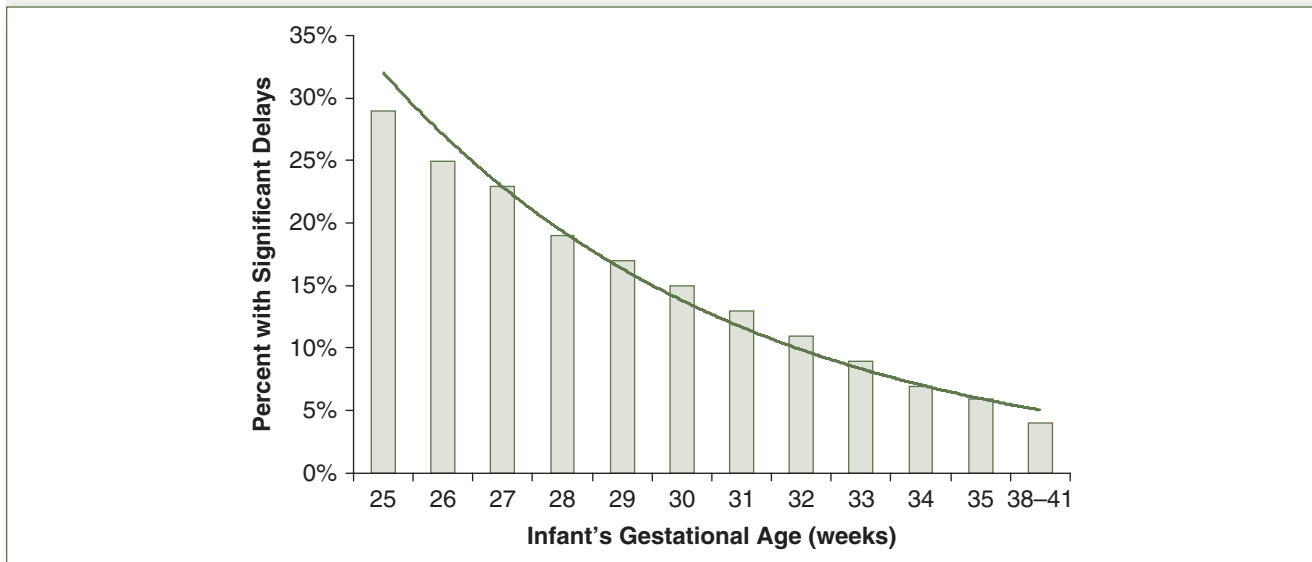
Andrew had long-standing academic problems. His IQ was 67. His reading and mathematics scores were comparable to those of a second- or third-grade child. Andrew received special education services, including remedial tutoring; however, he felt humiliated about receiving these special services.

Since beginning junior high school the previous year, Andrew's behavior became increasingly disruptive. He would often "clown around" in class, play pranks on teachers and other classmates, and get into fights on the playground. Andrew admitted to being teased by classmates because of his appearance, his academic problems, and his family history. "I know I'm slow," he said, "I don't need the other kids to tell me."

Andrew met with a psychologist at our clinic for weekly therapy sessions. Andrew was initially clownish and disruptive during the sessions, but he gradually came to trust his therapist and share his feelings.

During one session, Andrew commented, "You know, if it wasn't for my mom, I wouldn't have all of the problems that I'm having right now." His therapist replied, "I guess you're right. Your mom makes you pretty upset when you visit her. That causes you to get into trouble." Andrew replied, hesitantly, "No. That's not what I mean. I mean, if it wasn't for my mom's drinking—when I was inside her—I wouldn't be so dumb. If only she could have loved me more than drinking."

Figure 4.6 Preterm Birth Places Infants at Greater Risk for Delays



Source: From Kerstjens and colleagues (2013).

Note: Preterm infants are at much greater risk for delays in motor, language, social, and daily living skills than full-term infants. Risk increases exponentially with decreased gestational age.

axons, and synapses; increased myelination; and more complex brain activity. Although maturation can (and does) continue after delivery, the conditions for optimal development occur in utero (Volpe, 2008).

Childhood Illness or Injury

The two childhood illnesses most associated with the development of Intellectual Disability are encephalitis and meningitis. **Encephalitis** refers to the swelling of brain tissue, whereas **meningitis** refers to an inflammation of the meninges, the membrane that surrounds the brain and spinal cord. Both illnesses can be caused by bacteria or viral infections, although viral infections are more serious because they are often resistant to treatment.

All serious head injuries have the potential to cause Intellectual Disability. An obvious source of injuries is car accidents; however, most childhood head injuries occur around the home. Falls from tables, open windows, and stairs account for many injury-related cognitive impairments. Similarly, children who almost drown in swimming pools or bathtubs can experience brain damage and corresponding cognitive problems. Finally, children who are physically abused can experience Intellectual Disability because of environmental neglect or physical trauma (Appleton & Baldwin, 2006).

Lead toxicity is a risk factor for Intellectual Disability. Lead can be found in the paint of older buildings, in lead-soldered water pipes, and in industrial waste. Infants and toddlers may eat paint chips flaking off the walls of older

homes. Older children may inhale dust containing lead-based paint from walls, porches, and window panes. Lead enters the child's bloodstream and produces widespread cerebral damage. Exposure can cause Intellectual Disability, movement problems (ataxia), convulsions, and coma. If poisoning is detected early, children can be treated by flushing the lead from the bloodstream. Damage caused by prolonged exposure is irreversible.

The risk of lead poisoning varies by socioeconomic status (SES) and ethnicity. For example, 7% of middle-class white children are exposed to sufficient amounts of lead to cause poisoning. The risk of lead poisoning among low-income white and black children is much higher: 25% and 55%, respectively. Risk is particularly high among poor children living in urban settings (Dilworth-Bart & Moore, 2006; Phelps, 2005).

Causes of Cultural-Familial Intellectual Disability

Cultural-familial Intellectual Disability results from the interaction of the child's genes and environmental experiences over time. Children inherit a genetic propensity toward low intelligence. Furthermore, these children experience environmental deprivation that interferes with their ability to reach their cognitive potentials. Environmental deprivation might include poor access to health care, inadequate nutrition, lack of cognitive stimulation during early childhood (e.g., parents talking, playing, and reading with children), low-quality

educational experiences, and lack of cultural experiences (e.g., listening to music, trips to the museum). Over time, the interaction between genes and environment contributes to children's low intellectual functioning.

Socioeconomic Status

Familial Intellectual Disability is more prevalent among children from low-income families than middle-class families. The correlation between SES and children's intelligence is approximately .33. Furthermore, the relationship between SES and children's IQ increases when children experience extreme poverty or socioeconomic disadvantage (Turkheimer, Haley, Waldron, D'Onofrio, & Gottesman, 2003).

Both genetic and environmental factors explain this association between SES and children's intelligence. With respect to genetics, low-income parents tend to have lower IQ scores than middle-class parents. Individuals with higher IQs complete more years of schooling and assume more challenging and higher paying jobs. The children of high-income parents inherit their parents' genotypes that predispose them to a higher range of intellectual and adaptive functioning (Stromme & Magnus, 2000).

Furthermore, children from low-income families are exposed to environments that may restrict their intellectual potential. For example, low-income children are more likely to experience gestational and birth complications, have limited access to high-quality health care and nutrition, have greater exposure to environmental toxins, receive less cognitive stimulation from their home environments, and attend less optimal schools (McLoyd, 1998). These environmental deficits or risk factors limit the child's cognitive and adaptive potential. Young children in poverty earn IQ scores approximately nine points lower than children from middle-class families (Brooks-Gunn & Duncan, 1997; Duncan & Brooks-Gunn, 2000).

Ethnicity

Familial Intellectual Disability is also more frequently seen among ethnic minorities, especially African American children (Raghavan & Small, 2004). This association is partially due to the fact that many African American children live in low-income families and experience the same genetic and environmental risk factors as children from other low-income families. Whereas the average IQ score for middle-class whites is approximately 100, the average IQ score for low-income African Americans is approximately 90. Consequently, African Americans and individuals from low-income families are more frequently diagnosed with mild Intellectual Disability than are white, middle-class children (McLoyd, Hill, & Dodge, 2005).

Home Environment

Many studies suggest a relationship between the quality of the home environment and children's intellectual development.

After reviewing the data, Sattler (2002) identified two broad ways parents can enrich their children's home environment and help them achieve their intellectual potentials. First, families who provide their children with ample verbal stimulation, model and provide feedback regarding language, and give many opportunities for verbal learning appear to foster greater intellectual development in their children. Parents should take every opportunity to interact with their children through talking, playing, and reading. Second, encouraging academic achievement, curiosity, and independence in children is associated with increased intellectual functioning. Parents should encourage creative play, arts and crafts, and homemade games and activities, especially with young children, in order to help them develop novel and flexible problem-solving skills.

PREVENTION AND TREATMENT

Prenatal Screening

Shortly after birth, newborns are routinely administered a series of blood tests in order to determine the presence of genetic and other medical disorders that might cause Intellectual Disability. For example, all infants are administered a genetic test to screen for PKU. If PKU is found, a genetic counselor and nutritionist will meet with parents to discuss feeding options for the child.

If parents are at risk for having children with Intellectual Disability or other developmental delays, a physician may recommend genetic screening during gestation. Parents who may be carriers of specific genetic disorders, parents who have other children with developmental delays, or mothers older than age 35 often participate in screening.

At 15 to 18 weeks' gestation, mothers can undergo **serum screening** (Newberger, 2000). This procedure is usually called the "triple test" or "triple screen" because it involves testing mother's blood for three serum markers: alpha-fetoprotein, unconjugated estriol, and human chorionic gonadotropin. These serums are naturally produced by the fetus's liver and the placenta. If the child has Down Syndrome, alpha-fetoprotein and unconjugated estriol may be unusually low, while human chorionic gonadotropin levels may be unusually high. Significant elevations can be a sign of a genetic disorder, but this test has a high rate of false positives. Consequently, if test results are positive, the physician will usually recommend that the mother participate in additional testing.

Amniocentesis is a more invasive screening technique that is usually conducted during weeks 15 to 20 of gestation. The procedure involves removing a small amount of amniotic fluid with a needle inserted into the abdomen of the mother. The amniotic fluid contains fetal cells, which can be cultured and examined for genetic abnormalities. Amniocentesis is invasive; it carries a 0.5% risk of fetal death. Amniocentesis can also be conducted before 15 weeks' gestation, at the beginning of the second trimester, but the risk of fetal death increases to 1%–2%.

Chorionic villus sampling (CVS) is another genetic screening technique that can be done earlier, usually between 8 and 12 weeks' gestation. In CVS, the physician takes a small amount of chorionic villi, the wisp-like tissue that connects the placenta to the wall of the uterus. This tissue usually has the same genetic and biochemical makeup as the developing fetus. The tissue can be analyzed to detect genetic or biological anomalies. CVS is usually performed only when there is greatly increased risk of the fetus having a developmental or medical disorder. The risk of miscarriage associated with CVS is 0.5% to 1.5%.

Recently, physicians have been using ultrasound to detect structural abnormalities in the fetus that might indicate the presence of a developmental disorder (Rissanen, Niemimaa, Suonpää, Rynnänen, & Heinonen, 2007). Ultrasound is believed to be a relatively safe procedure for both mother and fetus. Between 11 and 14 weeks' gestation, embryos with Down Syndrome often show flatter facial profile and shorter (or absent) nasal bones than typically developing fetuses. The presence of these physical abnormalities, revealed by ultrasound, could indicate the presence of a genetic disorder. Based on the findings, mothers can decide whether they want to pursue more invasive testing.

Infant and Preschool Prevention

A number of state- and locally administered programs have been developed to prevent the emergence of Intellectual Disability in children at risk for low IQ. One of the most recent and, perhaps, the best-designed prevention programs for at-risk children is the **Infant Health and Development Program (IHDP)**. Participants in the IHDP were 985 premature infants who showed either low birth weight (weight 2,001 to 2,500 grams) or very low birth weight (weight < 2000 grams). Previous research indicated that these children were at increased risk for developmental delays, including Intellectual Disability and learning problems (Baumeister & Bacharach, 1996). Infants were randomly assigned to either an early intervention group or a control group. The parents of children in the intervention group received regular home visits from program staff. During these visits, staff taught parents games and activities that they could play with their infants to promote cognitive, linguistic, and social development. Staff also served as references to parents, helping parents address problems associated with caring for a preterm, low birth weight infant. When infants turned one year old, parents were invited to place them in a high-quality preschool program. The program was free and transportation to and from the preschool program was provided. The preschool ran year round, 5 days per week, until children were 3 years old. Families assigned to the control group were not given home visits or offered the preschool program.

To evaluate the success of the intervention, children's cognitive development was assessed at the end of the preschool program (age 3 years), at age 5 years, and at age 8

years. Children who participated in the program earned slightly higher IQs than children in the control group at age 3. However, by age 5, these differences in IQ disappeared.

The results of the IHDP indicate that early intervention programs can boost IQ scores among at-risk children, but increases in IQ are not maintained over time. The data are largely consistent with other early intervention programs designed to increase the cognitive functioning of low-income children (Farran, 2000).

Experts have disagreed on how to interpret the findings. Supporters of the IHDP concede that the results of the intervention were "largely negative" (Blair & Wahlsten, 2002, p. 130). However, advocates of the program believe the data speak to the importance of continuing educational enrichment for high-risk children beyond the preschool years. If the program had been extended through elementary school, children in the intervention group might have continued to show higher IQ scores than controls.

Critics of the IHDP argue that early intervention programs do not prevent Intellectual Disability and developmental delays and they should be discontinued (Baumeister & Bacharach, 2000). Instead, the money and time used for early intervention programs could be spent on primary prevention, such as providing at-risk families with better access to health care and nutrition. Critics argue that it is difficult to boost children's IQ because a person's genotype sets an upper limit on his or her intellectual potential (Baumeister & Bacharach, 2000). Furthermore, simply offering intervention services to high-risk families does not mean that they will take advantage of these services. In fact, 20% of children in the intervention group attended the preschool program less than 10 days in 2 years, and 55 children never attended at all (Hill, Brooks-Gunn, & Waldfogel, 2003). Since gains in IQ are directly related to participation in treatment, motivating families to participate in treatment seems to be a critical goal of any effective early intervention program.

Educational Interventions

Mainstreaming and Academic Inclusion

In 1975, Congress passed the Education of All Handicapped Children Act (Public Law 94-142). This act mandated a "free and appropriate public education" for all children with disabilities aged 3 to 18 years. From its implementation in 1977 through the mid-1980s, the practice of mainstreaming became more common in public school systems across the United States. **Mainstreaming** involved placing children with Intellectual Disability in classrooms with typically developing peers, to the maximum extent possible. At first, mainstreamed children with Intellectual Disability were allowed to participate in elective classes, such as physical education, art, and music, with typically developing children. For other subjects, they attended self-contained special education classes for children with developmental delays (Verhoeven & Vermeer, 2006).

In the mid-1980s, many parents argued that children with Intellectual Disability and other disabilities had the right to attend all classes with typically developing peers. This movement, sometimes called the “Regular Education Initiative,” gradually led to the practice of inclusion. **Inclusion** involves the education of children with Intellectual Disability alongside typically developing peers for all subjects, usually with the support of a classroom aide.

In 1997, Congress amended PL 94–142 by passing the Individuals with Disabilities Education Act (IDEA; PL 105–17). IDEA codified the practice of inclusion by demanding that children with disabilities be educated in the least restrictive environment possible:

To the maximum extent appropriate, children with disabilities . . . are educated with children who are not disabled, and special classes, separate schooling, or other removal of children with disabilities from the regular educational environment occurs only when the nature or severity of the disability of a child is such that education in regular classes with the use of supplementary aids and services cannot be achieved satisfactorily. (p. 61)

In addition to providing services for children with disabilities, IDEA also requires local educational systems to identify all infants, toddlers, and children with disabilities living in the community, whether or not they attend school. Once children are identified, a team of educational professionals (e.g., regular education teachers, special education teachers, school psychologists) conducts a comprehensive evaluation of the child’s strengths and limitations and designs a written plan for the child’s education. Infants and toddlers, aged 0 to 3 years, are provided with an **Individualized Family Services Plan (IFSP)**. For preschoolers and school-aged children, school personnel develop an **Individualized Education Program (IEP)** in consultation with parents. Typically, IEPs provide extra support to children while at school; children may be given special education services or a classroom aide. IEPs also usually specify accommodations for children with disabilities that help them achieve their cognitive, social, emotional, or behavioral potentials. IDEA was revised again in 2004 as the Individuals with Disabilities Education Improvement Act (PL 108–446; Williamson, McLeskey, Hoppey, & Rentz, 2006).

In general, inclusion improves the functioning of children with developmental disabilities, especially children with mild or moderate impairments. Inclusion seems to work best when (a) students with Intellectual Disability can become active in the learning process, and (b) these children frequently interact and cooperate with typically developing classmates. Inclusion may also have benefits for typically developing peers. Specifically, inclusion may teach typically developing children greater tolerance and understanding of individuals with developmental delays and increase students’ willingness to welcome children with delays into their peer groups.

The educational rights of children with Intellectual Disability have increased greatly over the past 30 years. The trend toward inclusion has allowed these children to have access to educational experiences that their counterparts, a generation ago, were typically not afforded. Today, the focus of attention has shifted from *where* children are educated (e.g., regular versus special education classes) to *how* they are educated (e.g., the nature and quality of services they receive; Zigler, Hodapp, & Edison, 1990). Simply placing all children with disabilities in regular classrooms is not enough. Now, we must learn to tailor regular educational experiences to the needs of these children in order to allow them to benefit from these experiences. Tailoring might involve smaller class size, more classroom aides, greater access to behavioral or medical consultants, specialized teacher training, and more time for teachers to plan lessons for students with disabilities (Hocutt, 1996).

Universal Design in the Classroom

In recent years, universal design has been a primary method of including children with intellectual and physical disabilities. **Universal design** is an educational practice that involves creating instructional materials and activities that allow learning goals to be achievable by children with different abilities and skills (Schalock et al., 2010). All children (with and without disabilities) use and benefit from these educational materials.

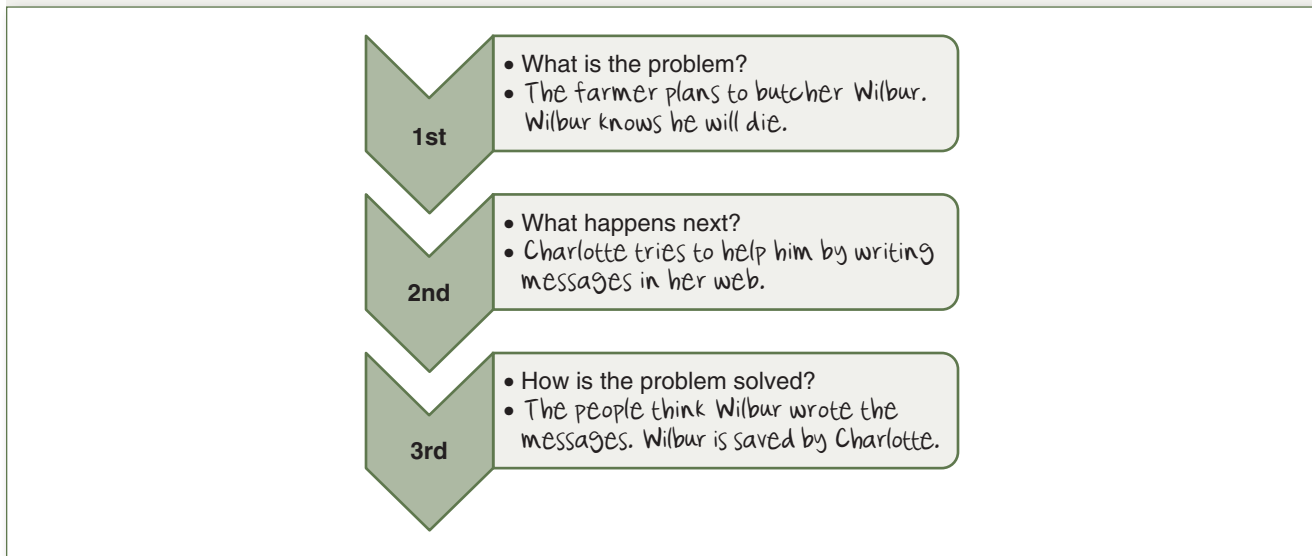
The clearest example of universal design can be seen in accommodations for people with physical disabilities. Many sidewalks now have “curb cuts” or sidewalk ramps that allow people who use wheelchairs to more easily cross the street. Similarly, buses are often built with low floors, rather than steep steps, to allow people with orthopedic problems easier access. These specially designed sidewalks and buses are used by all people; even people without physical disabilities often find them easier to use (Goldsmith, 2012).

Similarly, teachers can design assignments and activities that are accessible to children with a wide range of abilities and skills. These assignments and activities offer alternatives to traditional lecturing, reading, and writing. They can be used to plan (a) the way teachers introduce content to students, (b) the format of instructional material, and (c) the way students demonstrate their learning (Coyne et al., 2012).

First, a teacher might use a wide variety of instructional strategies to match the diversity of students’ skills and abilities. For example, a fourth-grade science teacher might find that all children (with and without disabilities) can learn about human anatomy by tracing their bodies on large sheets of paper and then drawing and labeling major organs. Similarly, a fourth-grade English teacher might demonstrate the steps involved in writing a book report using pictures, symbols, and arrows (i.e., graphic organizers) to help all students understand the temporal relationship of elements in a story (Figure 4.7).

Second, a teacher might modify the instructional technology she uses to present material. *Instructional technology* refers to the educational materials instructors use to teach

Figure 4.7 Using Universal Design at School



Note: Teachers might use a graphic organizer, like this flow chart, to help children write a book report.

ideas and concepts. For example, the science teacher might supplement her lessons with a child-friendly website about human anatomy. The website might allow children to enlarge the size of text, to read text aloud, and to access diagrams, pictures, videos. Similarly, the English teacher might use digital media that allows children to simultaneously listen to and read books online.

Third, teachers can measure students' learning in ways that do not penalize them for their disability. One way to accomplish this task is to rely on assistive technology when assessing student learning. *Assistive technology* refers to educational tools students use to compensate for their disabilities. For example, students with mild deficits in writing might be allowed to use text-to-speech software. Children with more profound problems with writing might use a software package, like Widgit Essentials, that allows children to use symbols and pictures to create sentences. Children with impairments in cognitive processing or fluency might be given extra time to complete tests. Indeed, if speed is not an important skill for a given learning domain (e.g., history), all children might be given extra time.

Overall, instructional strategies that adopt principles of universal design are effective. Students with mild to moderate impairments in intellectual and adaptive functioning seem to benefit the most from modifications to instructional methods and materials. For example, Coyne and colleagues (2012) examined the effectiveness of a computer-based reading comprehension program based on principles of universal design (Figure 4.8). The children in the study were in grades K-2; all had a developmental disability (e.g., Down, Fragile X, Prader-Willi Syndromes) and low intellectual functioning. The computer program allowed children to read books along with a narrator, to read with a partner, or to read silently.

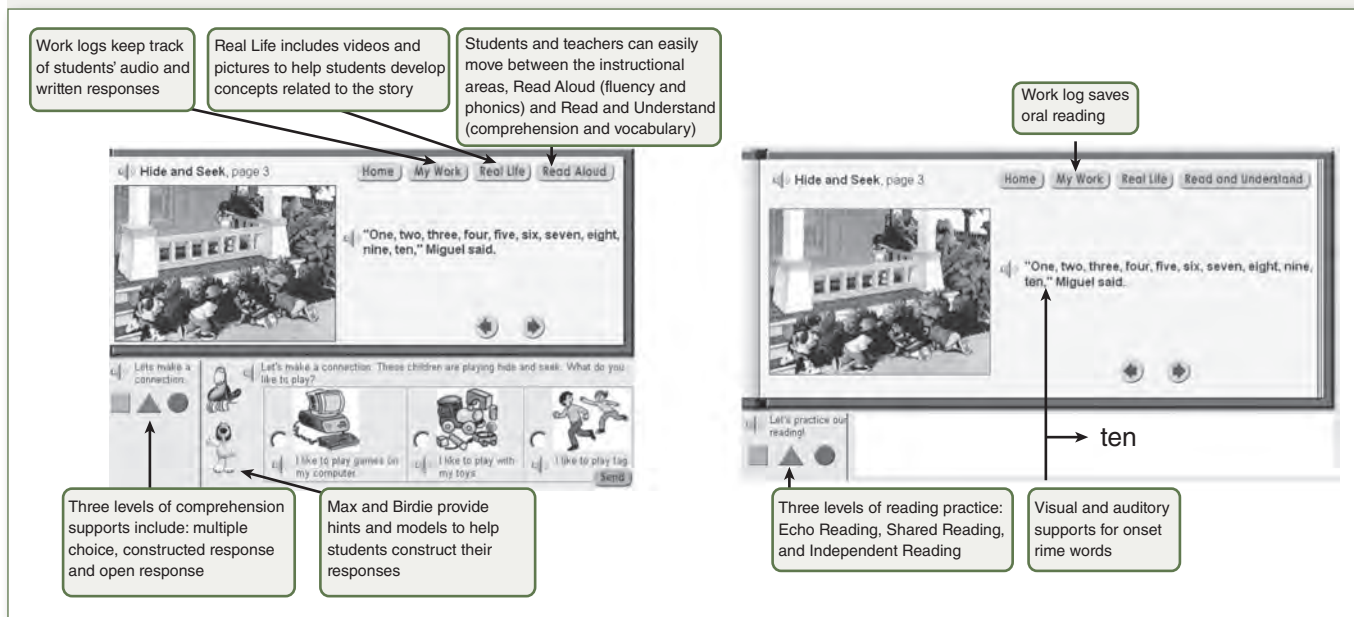
Children could also click on words to hear them pronounced and to access definitions, pictures, and videos related to the words. After listening to and reading the books, children could demonstrate their learning by taking a multiple-choice test, typing their responses on a keyboard, or speaking their responses into a microphone. Children in the control group received regular reading instruction. At the end of the school year, students in the universal design reading intervention showed significantly greater improvement in reading and listening comprehension than controls.

Applied Behavior Analysis

Approximately 25% of children with Intellectual Disability show challenging behavior, such as stereotypies, SIBs, or aggression. These behaviors are the primary reason children with Intellectual Disability are referred for treatment (Matson et al., 2011).

Applied behavior analysis (ABA) is a scientific approach to identifying a child's problematic behavior, determining its cause, and changing it (Feeley & Jones, 2006). The principles of ABA are based largely on the work of B. F. Skinner (1974), who believed that the study of behavior should be based on observable, quantifiable data. Skinner asserted that psychologists do not need to rely on latent (unobservable) constructs to explain and predict behavior. Instead, behavior can be understood in terms of overt actions and environmental contingencies. Rather than viewing behavior as originating from within the person, applied behavior analysts understand behavior primarily as a function of environmental antecedents and consequences (Holburn, 2005).

Figure 4.8 Literacy by Design



Source: From Coyne and colleagues (2012). Used with permission.

Note: Literacy by Design is a universal design approach to teaching reading comprehension. Children can read along with the narrator (i.e., echo reading), read with a partner, or read to themselves. Children can click on text to hear words or access pictures and videos. Children can demonstrate their comprehension by taking a multiple-choice test, typing their answers, or speaking their answers aloud.

A behavior analysts' first job is to operationally define the child's problem behavior, that is, to describe the behavior in a way that it can be observed and measured. For example, if a child repeatedly behaves "aggressively" in the classroom, the behavior analyst might operationally define the child's behavior in terms of one or two discrete, observation actions, such as "throws objects" or "pushes classmates." Whereas *aggression* is a somewhat vague term that cannot be easily observed or measured, throwing and pushing are more concrete actions that are easily identifiable.

Next, the behavior analyst will carefully observe and record the child's challenging behavior (see text box Research to Practice: Three Ways to Monitor Child Behavior). Several methods of behavioral observation are available (Hurwitz & Minshawi, 2012). One technique is to use *event recording*: The clinician observes the child and records the number of times the problem behavior occurs in an allotted period of time (e.g., 15 minutes). Event recording is suitable

for behaviors that occur frequently and have a clear beginning and ending. For example, a school psychologist might record the number of times a child blurts out answers during class. Another technique is *interval recording*. In interval recording, the clinician divides the observation period into brief time segments (i.e., intervals) usually less than 30 seconds in length. Then, the clinician observes the child and notes whether the problem behavior occurred during each interval. Interval recording is useful for frequently occurring behaviors without clear beginnings or endings. For example, a psychologist might use interval recording to determine the percent of class time a child engages in stereotyped rocking or swaying during class. A third technique, *duration recording*, is most appropriate for behaviors that take a long time to resolve. A clinician using this technique would record the duration of a continuously occurring behavior, such as the length of a temper tantrum or the time a child spent out of seat during class.

RESEARCH TO PRACTICE

THREE WAYS TO MONITOR CHILD BEHAVIOR

Event recording is used to assess discrete behaviors that occur frequently. In this case, the clinician records the number of times the child blurts out answers in class.

Child's name: Mark
 Target behavior: Blurts out answer without being called on by teacher

Date	Start time	End time	Count
2/20	10:00	10:20	✓✓✓✓✓✓
2/21	10:05	10:25	✓✓
2/22	9:55	10:15	✓✓✓✓✓✓✓✓

Interval recording is most appropriate for behaviors that occur frequently but do not have clear beginnings or endings. In this case, the clinician records the number of intervals the child showed stereotyped movement during class.

Child's name: Malorie
 Interval: 20 seconds
 Target behavior 1: Rocking or swaying in seat
 Target behavior 2: Flapping arms or hands

Behavior	20	40	60	20	40	60	20	40	60
1	✓	✓	✓		✓	✓	✓		
2					✓	✓			

Duration recording is best used to assess discrete behaviors that last a long time. In this case, the clinician records the amount of time the child leaves his seat during class.

Child's name: Miguel
 Target behavior: Time child leaves seat until seated again
 Start/End time: 9:00 AM–10:00 AM, 60 minutes

Episode	Start time	Stop time	Duration
1	9:05	9:09	4 min
2	9:15	9:24	9 min
3	9:31	9:36	5 min
			Total: 18 min

Observations of children's behavior can help identify the environmental conditions that elicit it or the consequences that maintain it (Lancioni et al., 2012). Is the child's challenging behavior prompted only by certain people or situations? Is it followed by consequences that might be positively or negatively reinforcing? Does the behavior tend to occur at certain times during the day?

Finally, the behavior analyst conducts a functional analysis of the child's behavior in order to identify and alter the causes of the problem behavior (Matson, 2011). **Functional analysis** involves carefully specifying the

child's challenging behavior, identifying the environmental contingencies that immediately precede the behavior (i.e., the antecedents), and identifying the environmental events that occur immediately after the behavior (i.e., the consequences) that likely maintain it. A functional analysis of behavior, therefore, involves identifying A (antecedents), B (the behavior), and C (its consequences). To change the child's behavior, the therapist can either alter the antecedents that prompt the undesirable behavior or change the consequences of the behavior so that it is no longer reinforced.

Brian Iwata and colleagues (1994) have developed a method of functional analysis to determine the causes of children's challenging behavior. This method involves observing the child in four conditions and noting the effect of each condition on the child's behavior:

- Attention condition: Whenever the child engages in challenging behavior in this condition, the therapist provides attention by reprimanding him or showing concern. For example, if the child throws an object, the therapist might respond, "Don't do that."
- Demand condition: In this condition, the therapist asks the child to engage in a moderately difficult task (e.g., sorting objects, cleaning a room).
- Alone condition: The child waits in a room with no people or toys present.
- Play condition: The therapist and the child play together.

The frequency and intensity of children's challenging behavior across the four sessions can indicate the behavior's purpose. Relatively high levels of challenging behavior in the attention condition, compared to the other conditions, might suggest that the behavior is maintained by *positive social reinforcement*, that is, to get attention from others.

Relatively high levels of challenging behavior in the demand condition, compared to the other conditions, suggests that the behavior is maintained through *negative reinforcement*; that is, it allows the child to avoid or escape undesired tasks. It is likely that caregivers negatively reinforce the challenging behavior by backing down from requests.

Relatively high rates of challenging behavior in the alone condition compared to the other conditions indicate that the behavior may be *automatically reinforced*. Children may engage in challenging behavior while alone because the behaviors themselves are reinforcing.

Once the purpose of the child's challenging behavior has been identified, the therapist can either alter the antecedents that elicit the behavior or the consequences that follow the behavior. Typically, therapists rely on reinforcement and punishment to accomplish the second objective.

Positive Reinforcement

Whenever possible, therapists use positive reinforcement to strengthen desirable behavior at the same time they reduce undesirable behavior. In a technique called **differential reinforcement**, therapists provide positive reinforcement only for behaviors that are desirable, while they ignore unwanted actions.

The two most common forms of differential reinforcement are (a) differential reinforcement of incompatible behaviors and (b) differential reinforcement of zero behavior. In **differential reinforcement of incompatible behaviors (DRI)**, the therapist provides positive reinforcement when the child engages in a behavior that is incompatible with the problematic behavior. For example, if a child engages in hand flapping or skin picking, the therapist might reinforce

him for keeping his hands in his pockets or holding onto a special toy or blanket. Since the child cannot flap his hands and keep them in his pockets at the same time, the hand flapping should decrease. In **differential reinforcement of zero behavior (DRO)**, the therapist reinforces the child for not engaging in the problematic behavior for a certain period of time. For example, a therapist might give a child an M&M every 30 seconds he does not engage in hand flapping or skin picking.

Positive Punishment

Reinforcement increases behavioral frequency; punishment decreases it (Singh, Osborne, & Huguenin, 1996). Positive punishment involves the presentation of a stimulus that decreases the frequency of a behavior. A common form of positive punishment used by parents is spanking. However, behavior therapists do not use spanking as a means of reducing behavior. Instead, some behavior therapists rely on other forms of positive punishment. For example, some therapists use aversive tastes, water mists, or visual screens to decrease severe behavior problems (Singh et al., 1996).

Since positive punishment techniques are aversive, they are only used under certain conditions such as when children's behaviors are dangerous or life threatening and other methods of treatment have been ineffective at reducing the problematic behavior. Punishment is only used in combination with positive reinforcement, and its use is carefully reviewed and monitored by independent experts. Parents must consent to the use of punishment before it is used to correct their children's behavior problem (APA, 1996).

Salvy, Mulick, Butter, Bartlett, and Linscheid (2004) describe the use of **punishment by contingent stimulation** to reduce self-injurious behavior in a toddler with Intellectual Disability. The girl, Johanna, would bang her head against her crib and other hard surfaces approximately 100 times each day. She had visible bruises on her forehead because of her behavior. Nonaversive interventions were not effective in reducing Johanna's head banging. The therapists and Johanna's mother decided to use punishment to reduce SIBs. The punisher was a brief electric shock that was administered by a device attached to Johanna's leg. The therapists could administer the shock remotely using a handheld activator. The shock was unpleasant (like being snapped by a rubber band), but it did not cause injury.

Treatment involved two phases. In the first (experimental evaluation) phase, Johanna and her mother played in an observation room in the hospital. Observers counted the frequency of her head banging during the first 10 minutes. This provided a baseline of Johanna's behavior to evaluate the effectiveness of the punishment. Then, the shock device was attached to Johanna's leg but shocks were not administered. Observations continued for another 10 minutes to see whether Johanna's behavior would change merely because she wore the device. Next, therapists began administering a

brief electrical shock contingent on Johanna's head banging. As before, observations were conducted for an additional 10 minutes. Finally, the shock device was removed and Johanna's behavior was observed for another 10 minutes. Results showed that the frequency of Johanna's head banging decreased from 30 times during baseline observation to 4 times after punishment.

During the second phase of treatment (home implementation), Johanna's mother was taught to punish Johanna's behavior at home. Therapists observed Johanna's behavior in the home for 2 days to obtain baseline data. On the third day, the shock device was attached to Johanna's leg. When Johanna began banging her head, her mother said "No hit, Johanna," retrieved the activator from her purse, and immediately issued a brief shock. The frequency of Johanna's behavior was recorded over the next month, at which time the shock device was removed from the home. Results showed that the frequency of Johanna's head banging at home decreased from 117 times per day at baseline to zero times per day after the contingent administration of shocks. Johanna's mother discovered that the verbal prompt "No hit, Johanna" combined with the action of walking toward her purse was sufficient to stop Johanna's head banging. At one-year follow-up, her mother reported no problems with Johanna's SIBs and no need to use the shock device.

Another form of positive punishment is called **overcorrection**. In overcorrection, the therapist requires the child to correct his problematic behavior by restoring his surroundings to the same (or better) condition than that which existed prior to his disruptive act. Overcorrection is often used when children show chronic problems using the toilet, wetting the bed, or destroying property. In the case of bedwetting, the therapist might require the child to strip his bedding, take his bedding and wet clothes to the laundry, help wash the clothes, and assist in making the new bed. For most children, this procedure is aversive because it is tedious and takes time away from sleep or enjoyable activities.

Overcorrection is often combined with a technique called **positive practice**. In positive practice, the therapist makes the child repeatedly practice an acceptable behavior immediately following his unacceptable act. In the case of bedwetting, the child might be required to sit on the toilet five times to practice the appropriate means of urinating. Positive practice can be aversive to children, but it also teaches children alternative, appropriate behavior.

Negative Punishment

Negative punishment occurs when the therapist withdraws a stimulus from the child, which decreases the recurrence of the child's behavior. Usually, the stimulus that is withdrawn is pleasant to the child. Consequently, the child experiences distress over its removal. Negative punishment is usually less aversive than positive punishment, so it is more often used to reduce problematic behavior.

The most benign form of negative punishment is **extinction**. In extinction, the therapist withdraws reinforcement from the child immediately following an unwanted behavior. Hanley and colleagues (2003) found that some children with developmental delays tantrum in order to obtain attention from caregivers. Caregivers would unknowingly reinforce their children's tantrums by looking at, talking to, and holding them. To extinguish these tantrums, caregivers can withdraw this reinforcement; that is, they can simply ignore their children's bids for attention. This strategy is sometimes called "planned ignoring."

When caregivers begin to extinguish behavior, the rate of children's behavior sometimes temporarily increases. This phenomenon is called an **extinction burst**. Children will usually escalate their problematic behavior in an attempt to gain the reinforcement that was previously provided. Over time, the behavior's frequency and intensity will decrease, as long as reinforcement is withheld. Extinction is a slow, but effective, means of reducing behavior problems. The primary drawback of extinction is that it cannot be used to reduce SIBs because these behaviors cannot be ignored.

A second form of negative punishment is **time out**. In time out, the therapist limits the child's access to reinforcement for a certain period of time. Time out can take a number of forms, but it must involve the complete absence of reinforcement. Children should not be allowed to play, avoid tasks, or gain attention from others while in time out. Time out is usually accomplished by physically removing the child from the reinforcing situation for several minutes.

A final form of negative punishment is **response cost**. In response cost, the therapist withdraws reinforcers from the child immediately following a problematic act. Each problematic behavior "costs" the child a number of tangible reinforcers, such as candy, points, tokens, or other desired objects or privileges. Response cost is similar to time out. In time out, reinforcement is withdrawn *for a specific amount of time*. In response cost, reinforcement is withdrawn *in a specific quantity*. Response cost is often used in combination with token economies. Children may be reinforced with tokens or points for each desirable behavior and required to give up a certain number of tokens or points for each problematic behavior.

Behavioral treatment for people with Intellectual Disability has considerable empirical support. Kahng and colleagues (2002) reviewed 35 years of published research on the effectiveness of behavior therapy to treat SIBs. Data from 706 individuals showed an overall reduction in SIBs of 83.7%. The most effective treatments tended to involve punishment (e.g., overcorrection, shock, time out) with 83.2% effectiveness, followed by extinction (e.g., planned ignoring) with 82.6% effectiveness, and positive reinforcement (e.g., DRI, DRO) with 73.2% effectiveness. Combining behavioral interventions usually resulted in slightly higher effectiveness than the use of any single intervention alone.

Medication

Medication is frequently administered to children and adolescents with Intellectual Disability. Approximately 19% to 29% of people in the community and 30% to 40% of individuals in residential facilities with Intellectual Disability are prescribed at least one psychiatric medication (Singh, Ellis, & Wechsler, 1997). Little research has examined the efficacy of medication in youths with Intellectual Disability, for at least three reasons. First, for a long time, mental health experts did not think that children with Intellectual Disability suffered from psychiatric disorders, or they overlooked their psychiatric symptoms. The problem of dual diagnosis has only recently been recognized. Second, it is difficult to recruit large samples of children with both Intellectual Disability and a psychiatric diagnosis. Consequently, most research has involved very small samples. Third, many research studies have not used adequate experimental designs. Typically, a double-blind, placebo-controlled study is necessary to infer a causal relationship between the use of a medication and symptom reduction, but this type of study has been rare in the research literature (Singh, Matson, Cooper, Dixon, & Sturmey, 2005).

Medications for Disruptive Behaviors

Risperidone (Risperdal) is an atypical antipsychotic medication that blocks certain dopamine and serotonin receptors. It was first released in the United States in 1994 as a medication to treat schizophrenia and other psychotic disorders in adults. Some psychiatrists began using risperidone with developmentally delayed children who showed disruptive behavior problems. Today, risperidone is frequently used to treat oppositional and defiant behavior, destructive behavior, aggression, and SIBs in children and adolescents with Intellectual Disability (Handen & Gilchrist, 2006; Singh et al., 2005).

Evidence supporting the use of risperidone to treat problem behavior comes from a series of double-blind, placebo-controlled studies of children and adolescents who showed behavior problems and low intelligence. For example, Aman, De Smedt, Derivan, Lyons, Findling, and the Risperidone Disruptive Behavior Study Group (2002) examined 118 children aged 5 to 12 years who showed both low intellectual functioning and significant behavior problems. Children were randomly assigned to either an experimental group whose members received a low dose of risperidone or to a control group whose members received a placebo. Six weeks later, 77% of the children in the experimental group showed significant improvement in their behavior, compared to only 33% of children in the control group. Other studies have yielded similar results (Findling, Aman, Eerdeken, Derivan, Lyons, & Risperidone Behavior Study Group, 2004; Snyder et al., 2002).

Medications for Stereotypies and Self-Injurious Behavior

Physicians often prescribe traditional antipsychotic medications to suppress SIBs. Most traditional antipsychotics block dopamine receptors. Interestingly, some individuals who show SIBs also display a hypersensitivity to dopamine. Consequently, there is reason to expect that medications that block certain dopamine receptors would be especially effective at reducing SIBs and stereotypies (Szymanski & Kaplan, 2006).

Research examining the effectiveness of dopamine blockers has been limited, however. Two antipsychotic medications, haloperidol (Haldol) and fluphenazine, appear to be effective in treating adults with SIBs; however, these medications have not been adequately studied with children (Aman, Collier-Crespin, & Lindsay, 2000). Critics argue that these drugs reduce problematic behavior mostly through sedation. Consequently, individuals taking traditional antipsychotics show a general decrease in *all* behaviors, not just SIBs.

Some individuals who engage in self-injurious behavior show high secretions of endogenous opioids. Opioids may reduce pain sensitivity during the SIBs. Consequently, some physicians have used **naltrexone**, a drug that blocks opioid receptors, to curb SIBs. Research investigating the efficacy of naltrexone has been mixed. Unfortunately, some of the largest studies, including those involving children, have not shown naltrexone to be efficacious.

Medication for Anxiety and Mood Disorders

Antidepressants have been used to treat anxiety and mood disorders in adolescents with Intellectual Disability. Unfortunately, research supporting their use has largely involved only case studies and anecdotal reports (Aman et al., 2000). Physicians have also used lithium and valproic acid for children with Intellectual Disability and Bipolar Disorder. However, few large-scale studies have been conducted to investigate their efficacy (Aman et al., 2000).

Support for Parents

Before ending this section on treatment, we should highlight the importance of supporting the parents of children with developmental disabilities. After children are initially diagnosed with GDD or Intellectual Disability, parents are at increased risk for depression. They often report a sense of loss or disappointment associated with their child's diagnosis and apprehension about their child's future or their ability to raise a child with special needs. Over time, however, parents' dysphoria usually decreases (Glidden, 2012). Nevertheless, challenges associated with caring for a child with a developmental disability remain. In one large study of parents of children with GDD, nearly 42% reported significant elevations in stress associated with caring for their children (Tervo, 2012).

Parenting stress can take its toll on the family system (Al-Yagon & Margalit, 2012). Parents of children with

developmental disabilities often experience disruptions in the quality of their marriage and family life. However, the effect of having a child with a disability on marital and life satisfaction is complex. Parents who report socioeconomic stress, a high degree of work and interpersonal hassles, and low support from their spouse, often report a marked deterioration in marital and family life after the birth of a child with special needs. In contrast, parents who feel supported by their spouse and who use active, problem-focused coping techniques to deal with family-related stress often report no change in marital satisfaction or quality of life. Indeed, some families report greater cohesion and satisfaction after the birth of a child with a developmental disability (Glidden, 2012).

Some developmental disorders are not strongly associated with increased parental stress. For example, the parents of children with Down Syndrome often report only moderate levels of stress, perhaps because children with this condition typically show mild cognitive impairment and are usually described by others as affectionate and social. There may be less stigma associated with caring for a child with Down Syndrome; most people can easily recognize a child with this condition and generally have some understanding of the disorder. Support groups for families are also available in many communities.

Other developmental disorders are associated with higher levels of parenting stress. Parents can experience considerable stress when the cause of their child's developmental delay cannot be identified. Parents might erroneously blame themselves for their child's limitations or feel uncertain about their child's prognosis. When the cause of children's delays is unknown, or when the disorder is uncommon, parents may also feel misunderstood or alienated. Regardless of etiology, certain child behaviors seem to increase parenting stress: poor motor control, social deficits, and aggression (Tervo, 2012).

Therapists can help children with developmental disabilities by supporting their parents in times of difficulty and uncertainty. Besides providing informational support about their children's development and suggestions for symptom management, therapists can offer emotional support through their willingness to listen to parents' concerns. Parents can also encourage the use of active, problem-focused strategies to deal with parenting stress (Al-Yagon & Margalit, 2012). Parents who are able to cope with their own stress may be better able to care for their children and implement many of the interventions that will promote their children's intellectual and adaptive functioning in the long run.

CHAPTER SUMMARY

What Is Intellectual Disability?

- Intellectual Disability is characterized by significant limitations in cognitive and adaptive functioning that emerge during the course of children's development.
 - IQ scores <70–75 usually indicate limitations in cognitive ability.
 - Adaptive functioning is measured in three broad domains (a) conceptual skills, (b) social skills, and (c) practical skills. Scores <70–75 usually indicate limitations in adaptive functioning.
 - Cognitive and adaptive limitations are equally important to the identification of Intellectual Disability. Children are not diagnosed based on IQ alone.
- The American Association on Intellectual and Developmental Disabilities (AAIDD) offers its own definition on Intellectual Disability. It is largely consistent with the *DSM-5* definition, but it places greater emphasis on contextual factors and the supports children need to succeed academically and socially.
- Global Developmental Delay is characterized by significant deficits in two of the following domains that emerge prior to age 5 years: (a) fine/gross motor skills, (b) speech/language, (c) social/personal skills, and (d) daily living skills.
 - Global Developmental Delay is usually diagnosed in infants and very young children suspected of Intellectual Disability but who are too young to be administered an IQ test.
 - Not all children with Global Developmental Delay will meet criteria for Intellectual Disability when they become older.

Associated Characteristics

- Many youths with Intellectual and Developmental Disabilities show challenging behavior: actions of sufficient intensity, frequency, or duration that children's physical safety (or the safety of others) is jeopardized. Challenging behaviors include the following:
 - Stereotypies are actions performed in a consistent, rigid, and repetitive manner with no immediate practical significance (e.g., hand flapping, rocking back and forth).
 - Self-injurious behaviors are repetitive actions that can, or do, cause physical harm to the person.
 - Physical aggression are actions that cause (or can cause) property destruction or injury to others.
- Children with Intellectual and Developmental Disabilities can experience other psychiatric disorders and are at greater risk than typically developing children their age.

Epidemiology

- Approximately 1.8% of individuals have Intellectual Disability. The disorder is more common among school-age children than adults and is more common among boys than girls.
- According to Edward Zigler, children with Intellectual Disability can be differentiated into two types:
 - Youths with organic Intellectual Disability have known medical causes for their impairments. They tend to have IQ scores <50, physical abnormalities associated with neurological problems, and parents without impairments.
 - Youths with cultural-familial Intellectual Disability have no identified cause for their impairments. They tend to have IQ scores between 50 and 70, have normal physical appearance, have parents with low intelligence, and come from lower income families.
- According to the similar sequence hypothesis, children with Intellectual Disability show cognitive development in the same sequence as typically developing children, albeit at a slower pace. Most research supports this hypothesis.
- According to the similar structure hypothesis, children with and without Intellectual Disabilities who have the same mental age will show the same level of abilities. Although generally supported, some children with organic Intellectual Disability show abilities different than their typically developing peers.
- A behavioral phenotype refers to the curious pattern of strengths and weaknesses shown by children with specific causes for their Intellectual Disability.

Causes of Intellectual Disability

- Chromosomal abnormalities can cause Intellectual Disability.
 - Down Syndrome is usually caused by an extra 21st chromosome (trisomy 21). It is characterized by Intellectual Disability, relative deficits in language, and relative strengths in visual-spatial skills and social functioning in childhood.
 - Fragile X Syndrome is caused by an X-link genetic mutation. The disorder is more severe in boys. It is characterized by Intellectual Disability, relative deficits in sequential processing, relative strengths in simultaneous processing, and problems with social functioning.
 - Prader-Willi Syndrome is caused by a missing paternal chromosome 15. It is characterized by mild Intellectual Impairment, obesity, and obsessive-compulsive behavior.
 - Angelman's Syndrome is caused by a missing maternal chromosome 15. It is characterized by Intellectual Disability, relative deficits in language, and unusually happy demeanor.
 - William's syndrome is caused by deletions on chromosome 7. It is characterized by Intellectual Disability, relative weakness on visual-spatial tasks, well-developed vocabularies and spoken language, and inattention/hyperactivity.
- Metabolic disorders can also cause Intellectual Disability. PKU is caused by a recessive gene which can cause severe disability if children do not abstain from phenylalanine-rich foods (e.g., cheese, meats).
- Teratogen exposure in utero can cause Intellectual Disability. Common teratogens include bacteria, viruses, alcohol, and other drugs.
- Complications during pregnancy or delivery can contribute to Intellectual Disability, especially when they restrict the fetus's access to oxygen.
- Childhood illnesses or injuries can cause Intellectual Disability including encephalitis, meningitis, high fever, and lead toxicity.
- Cultural-familial Intellectual Disability is usually caused by a genetic propensity toward below-average cognitive functioning and environmental deprivation or socioeconomic disadvantage.

Prevention and Treatment

- Women at high risk may participate in prenatal screening to estimate the likelihood of having a child with a genetic disorder. Screening techniques include (a) serum screening, (b) amniocentesis, and (c) chorionic villus sampling.
- Prevention programs for youths at risk for Intellectual Disability, such as the Infant Health and Development Program, are associated with short-term gains in IQ, but, no long-term differences in children's outcomes.
- The Individuals with Disabilities Education Improvement Act requires children with Intellectual Disability to be identified at an early age and provided with services to help them achieve.
 - School-age children receive an Individualized Education Program (IEP) and usually participate in academic inclusion programs alongside typically developing peers.
 - Universal design is an educational practice that involves instructional materials and activities that allow learning goals to be achievable by children with and without disabilities.
- Applied Behavior Analysis (ABA) is typically used to reduce challenging behavior in children with Intellectual Disability.
 - Clinicians can use event recording, interval recording, or duration recording to identify and monitor challenging behavior.
 - Functional analysis can help determine the cause of the behavior:
 - Positive social reinforcement (i.e., to get attention)
 - Negative reinforcement (i.e., to avoid or escape unpleasant tasks)
 - Automatic reinforcement (i.e., self-stimulation)

- Clinicians can use operant conditioning to correct behavior.
 - Positive reinforcement (e.g., differential reinforcement)
 - Positive punishment (e.g., overcorrection, positive practice)
 - Negative punishment (e.g., time out, response cost)
- Medication, such as risperidone (Risperdal) can be used to reduce some challenging behavior.
- Therapists can provide information to families of children with Intellectual and Developmental Disabilities, alleviate parenting stress, and teach problem-focused coping.

KEY TERMS

aggression	101	maternal syphilis	111
amniocentesis	114	maternal uniparental disomy	107
anoxia	112	meningitis	113
applied behavior analysis (ABA)	117	naltrexone	122
behavioral phenotype	104	needed supports	94
chorionic villus sampling	115	nondisjunction	104
chromosomal microarray (CMA)	97	organic Intellectual Disability	103
chromosomal mosaicism	105	overcorrection	121
conceptual skills	91	phenylalanine	110
differential reinforcement	120	positive practice	121
differential reinforcement of incompatible behaviors (DRI)	120	practical skills	91
differential reinforcement of zero behavior (DRO)	121	punishment by contingent stimulation	120
encephalitis	113	response cost	121
extinction	121	risperidone (Risperdal)	122
extinction burst	121	rubella virus	111
Fetal Alcohol Syndrome	111	self-injurious behaviors (SIBs)	99
Fragile X Mental Retardation 1 (FMR1) gene	106	sequential processing	107
functional analysis	119	serum screening	114
Global Developmental Delay (GDD)	95	similar sequence hypothesis	103
human immunodeficiency virus	111	similar structure hypothesis	103
hyperphagia	107	simultaneous processing	107
inclusion	116	social skills	91
Individualized Education Program (IEP)	116	stereotypies	99
Individualized Family Services Plan (IFSP)	116	teratogens	111
Infant Health and Development Program	115	time out	121
lead toxicity	113	translocation	105
mainstreaming	115	universal design	116

CRITICAL THINKING EXERCISES

1. When many people think of Intellectual Disability, they think about a child with Down Syndrome. To what extent is a child with Down Syndrome an accurate portrayal of all children with Intellectual Disability?
2. Until recently, people mistakenly believed that children and adolescents with Intellectual Disability could not experience other psychiatric disorders. Why? Why might it be difficult for a psychologist or physician to assess anxiety and depression in a child with moderate Intellectual Disability?

3. Some professionals classify Intellectual Disability into two broad categories: organic Intellectual Disability and cultural-familial Intellectual Disability. How can these labels be misleading? Why might it be better to describe children with Intellectual Disability based on their behavioral phenotype?
4. How does the treatment for PKU illustrate the interaction of genes and environment in child development?
5. Why are children of lower SES backgrounds at greater risk for certain types of Intellectual Disability? Why might low-SES children with Intellectual Disability have poorer prognoses than middle-class children with Intellectual Disability?
6. Why do clinicians always try to use other forms of behavioral treatments (e.g., positive reinforcement, negative punishment) before using positive punishment? Under what circumstances might it be permissible to use positive punishment to help a child with Intellectual Disability?

EXTEND YOUR LEARNING

Videos, practice tests, flash cards, study guides, and links to online resources for this chapter are available to students online. Teachers also have access to lecture notes, PowerPoint presentations, suggestions for classroom activities, and possible exam questions. Visit: www.sagepub.com/weis2e.