LEARNED OBJECTIVES

After reading this chapter, you should be able to do the following:

5.1. Describe the key features of intellectual disability (ID) and the way in which children with this condition can vary in terms of their adaptive functioning.

- Differentiate ID from global developmental delay (GDD).
- List and provide examples of challenging behaviors shown by some children with developmental disabilities.

5.2. Distinguish between organic and cultural–familial ID.

- Explain how genetic, metabolic, and environmental factors can lead to developmental disabilities in children.

5.3. Identify evidence-based techniques to prevent and treat developmental disabilities.

- Apply learning theory to reduce challenging behaviors in youths with developmental disabilities.

Once there was a craftsman who used all his skill and effort to create a wonderful new pot. The pot was made of clay, crafted by his weathered hands, and baked into a beautiful form. The man glazed and decorated the pot, using colors and designs that were as unique as they were beautiful. When it was finished, the man carried the pot to a nearby well to fetch some water for his home. To his surprise, he discovered the pot had developed a small crack from the kiln, which caused water to leak from the bottom. At first, the crack was small, but over time it became larger and more noticeable.

One day, the man’s friend said, “That pot has a crack. By the time you get home, you’ve lost half of your water. Why don’t you throw it away and get a new one?” The man paused, turned to his friend, and replied, “You don’t understand. Yes, it’s true that this pot leaks more and more every day. But every day it also waters more and more flowers on the path from the well to my home.” Sure enough, along the path had sprung countless wildflowers of all varieties, while in other areas, the land was barren. His friend simply nodded in approval (see Image 5.1).

The story of the broken pot illustrates the dignity and value of every person. Each person has unique gifts and talents, although sometimes they are hard to recognize. When studying children with developmental disabilities, it’s easy to focus on limitations and lose sight of the children themselves. Many of these youths face significant challenges performing everyday activities like bathing and dressing. Others have difficulty with communication and language. Still others struggle in school or exhibit challenging behaviors in social settings. Too often, these problems overshadow their abilities.

Regardless of his or her disability, disorder, or diagnosis, each of these children has intrinsic worth. A challenge facing parents, teachers, and all people who interact with these youths is to not lose sight of the child when we focus on

1Adapted from a story by Kevin Kling.
his or her problem. One of my clients, Will, was born with Down syndrome. Although he struggled with reading and math, he taught his classmates to be patient, to act with empathy, and to respect others who are different. Another client, Camden, a boy with intellectual disability (ID) and attention-deficit/hyperactivity disorder (ADHD), could not stay focused in class; however, he had an excellent sense of humor and loved to play soccer. Still another client, Chloe, had Williams syndrome (WS). Although she had serious cognitive deficits and health problems, she was also one of sweetest girls I have ever met. Consider Rosa, a girl with Down syndrome, whose family changed the way we think about ID today.

### 5.1 DESCRIPTION AND EPIDEMIOLOGY

#### What Is Intellectual Disability?

Intellectual disability (ID) is a term that describes the behavior of 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 their peers. These children also have diverse outcomes. Most are integrated into general education classrooms, many participate in educational and recreational events in their communities, and some raise families of their own. In this chapter, we will explore this heterogeneous group of individuals, explore the causes of their disabilities, and learn evidence-based strategies to help them achieve their highest potentials (Witwer, Lawton, & Aman, 2014).

All individuals with ID have significantly low intellectual functioning (see Table 5.1). They experience 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 5 years of age and older, intellectual functioning is measured using a standardized, individually administered intelligence test. Recall that IQ scores are normally distributed with a mean of 100 and a standard deviation of 15. IQ scores approximately two standard deviations below the 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% to 3% of the population.

Individuals with ID also 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 (Sturmey, 2014b).

The Diagnostic and Statistical Manual of Mental Disorders (DSM-5; American Psychiatric Association, 2013) identifies three domains of adaptive functioning: conceptual, social, and practical. To be diagnosed with ID, individuals must show impairment in at least one domain. Usually, children with ID experience problems in multiple areas:

**Conceptual skills:** understanding language, speaking, reading, writing, counting, telling time, solving math problems, having the ability to learn and remember information and skills.

**Social skills:** having interpersonal skills (e.g., making eye contact when addressing others), following rules (e.g., turn-taking during games), engaging in social problem-solving (e.g., avoiding arguments), understanding others (e.g., empathy), making and keeping friends.

**Practical skills:** activities of daily living including taking personal care (e.g., getting dressed, grooming), practicing safety (e.g., looking both ways before crossing street), doing home activities (e.g., using the telephone), having school/work skills (e.g., showing up on time), participating in recreational activities (e.g., clubs, hobbies), and using money (e.g., paying for items at a store).

### Table 5.1 Diagnostic Criteria for Intellectual Disability (Intellectual Developmental Disorder)

<table>
<thead>
<tr>
<th>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:</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>A.</strong> 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.</td>
</tr>
<tr>
<td><strong>B.</strong> 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.</td>
</tr>
<tr>
<td><strong>C.</strong> Onset of intellectual and adaptive deficits during the developmental period.</td>
</tr>
</tbody>
</table>

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

*Table 5.2 provides a description of each type of severity.
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 adaptive functioning. For example, the Diagnostic Adaptive Behavior Scale (DABS) is a semi-structured interview that is administered to caregivers of children with developmental disabilities (see the following Research to Practice section). Based on caregivers' reports, the interviewer rates children's adaptive behavior across the conceptual, social, and practical domains. 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 (Balboni et al., 2014; Schalock, Tassé, & Balboni, 2015).

Note that ID is characterized by low intellectual functioning and problems in adaptive behavior. Many people incorrectly believe that ID 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 ID (Sturmey, 2014a).

Finally, all individuals with ID show limitations in intellectual and adaptive functioning early in life. Although some people are not identified as having ID until they are adults, they must have histories of intellectual and daily living problems beginning in childhood. This age-of-onset requirement differentiates ID from other disorders characterized by problems with intellectual and adaptive functioning.

**Review:**

- ID is characterized by significant deficits in intellectual and adaptive functioning that emerge early in life. Both intellectual and adaptive functioning deficits are necessary for the diagnosis.
- Adaptive functioning refers to a person's ability to cope with day-to-day tasks. DSM-5 identifies three dimensions of adaptive functioning: (1) conceptual, (2) social, and (3) practical.

### How Does Intellectual Disability Differ Based on Severity?

Clinicians specify the severity of ID based on the person's level of adaptive functioning. Children with mild deficits in adaptive functioning (i.e., standard scores 55–70) in only conceptual skills are classified as having mild ID.

**How Do Clinicians Assess Adaptive Functioning?**

Clinicians assess adaptive functioning by administering semistructured interviews to caregivers of children suspected of intellectual disability (ID). 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 typically developing children.

Here are some areas of adaptive functioning that might be assessed in younger children, older children, and adolescents.

<table>
<thead>
<tr>
<th>Younger Children</th>
<th>Older Children</th>
<th>Adolescents</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Conceptual</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Can count 10 objects,</td>
<td>States value of penny,</td>
<td>Sets a watch or clock to</td>
</tr>
<tr>
<td>one by one; knows day,</td>
<td>nickel, dime; uses</td>
<td>correct time; can complete</td>
</tr>
<tr>
<td>month, year of birth;</td>
<td>mathematical operations</td>
<td>a job application</td>
</tr>
<tr>
<td>Knows “hi” and “bye”</td>
<td></td>
<td></td>
</tr>
<tr>
<td>when coming and going;</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Asks for help when</td>
<td></td>
<td></td>
</tr>
<tr>
<td>needed</td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Social</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Uses the restroom;</td>
<td>Answers the telephone;</td>
<td>Travels to school or work</td>
</tr>
<tr>
<td>Drinks from a cup</td>
<td>Can safely cross busy</td>
<td>by themselves; washes</td>
</tr>
<tr>
<td>without spilling</td>
<td>streets</td>
<td>clothes, dishes</td>
</tr>
<tr>
<td><strong>Practical</strong></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Based on the DABS (Schalock et al., 2015).
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 5.2 provides an overview of children’s adaptive functioning at each level of severity.

**Mild Intellectual Disability (Adaptive Functioning Scores 55–70)**

As infants and toddlers, children with mild ID 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

### Table 5.2 Describing the Severity of Intellectual Disability

<table>
<thead>
<tr>
<th>Severity</th>
<th>Conceptual Domain</th>
<th>Social Domain</th>
<th>Practical Domain</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild</td>
<td>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.</td>
<td>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.</td>
<td>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.</td>
</tr>
<tr>
<td>Moderate</td>
<td>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.</td>
<td>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.</td>
<td>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.</td>
</tr>
<tr>
<td>Severe</td>
<td>The child generally has little understanding of written language or numbers. Caretakers must provide extensive support for problem solving throughout life.</td>
<td>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.</td>
<td>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.</td>
</tr>
<tr>
<td>Profound</td>
<td>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.</td>
<td>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.</td>
<td>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.</td>
</tr>
</tbody>
</table>

*Source: Based on DSM-5, 2013.*
Moderate Intellectual Disability (Adaptive Functioning Scores 40–55)

Children with moderate ID 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 ID 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 35–40)

Children with severe ID are usually first identified in infancy (Jacobson & Mulick, 1996). They almost always show early delays reaching early 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. 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 ID 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 1-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 ID 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 ID is determined by the child’s intelligence and adaptive functioning. Two people can show ID 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 “ID” tells us only about a person’s general intellectual and adaptive functioning; the diagnosis says nothing about causes, course, or outcomes (Witwer et al., 2014).

Needed Supports

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 IDs. Since 1910, they have offered guidelines for the identification of ID and the best methods to help children and adults with this condition. In years past, the DSM and AAIDD definitions of ID had differed considerably. Currently, however, the DSM-5 and AAIDD definitions overlap to a great degree, which will likely improve communication between members of these two professional organizations (Harris & Greenspan, 2016).

One remaining difference in the AAIDD conceptualization of ID is its emphasis on needed supports (Schalock & Luckasson, 2015). Needed supports refer to a broad array of assistance that helps the individual function effectively in society (see Image 5.2). 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 ID 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 semistructured interview to help clinicians identify the type and intensity of supports needed for adolescents and adults with ID (Buntinx, 2016). 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 approach to classifying individuals with ID in terms of needed supports has two main advantages (Schalock & Luckasson, 2015). First, this approach conveys more information about clients than simply classifying them with ID 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.

Review:

• DSM-5 allows clinicians to classify individuals with ID based on their adaptive functioning: mild, moderate, severe, or profound.

• In contrast, the AAIDD classifies individuals with ID based on their needed supports—that is, assistance that helps these individuals function in society.

• Levels of needed supports include intermittent, limited, extensive, or pervasive.

What Is Global Developmental Delay?

Description

The diagnosis of ID requires significant deficits in intellectual and adaptive functioning. Typically, intellectual functioning is assessed using norm-referenced IQ tests. Although assessing IQ is relatively straightforward among school-age children, it can be tricky in toddlers and preschoolers—especially those children suspected of developmental delays.

Clinicians who want to measure the intellectual ability of very young children have several options (Witwer et al., 2014). The Bayley Scales of Infant and Toddler Development, Third Edition (BSID-III) are appropriate for children aged 1 to 42 months. However, the BSID-III is usually considered a measure of children’s cognitive, motor, and social development rather than intelligence per se. The Stanford-Binet Intelligence Scales, Fifth Edition is a true IQ test that can be administered to children as young as two-years old. Similarly, the Wechsler Preschool and Primary Scale of Intelligence, Fourth Edition (WPPSI-IV) can be given to children as young as 2.5 years. However, IQ scores assessed prior to age 4 or 5 are often not good predictors of intelligence in later childhood or adolescence (Tirosch & 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 ID; 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 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: (1) fine/gross motor skills, (2) speech/language, (3) social/personal skills, and (4) daily living. Significant delays are defined by scores two or more standard deviations below the mean (see Table 5.3). 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 caregivers 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). Consider the case of Sam, a preschooler with GDD (see next page).

The word delay in the name GDD implies that children 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 ID by the time they begin preschool. Furthermore, retrospective studies indicate that most older children with ID 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 ID (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

| Table 5.3 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 | Babble, 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.
GLOBAL DEVELOPMENTAL DELAY

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. 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 six 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 (BSID) 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 BSID showed delays in language, motor, and social–emotional skills. 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).

“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.”

with GDD. Children’s scores ranged widely, and nearly 20% of children earned scores within the average range (Riou, Ghosh, Francoeur, & Shevell, 2009).

Furthermore, some young children with GDD do not develop ID later in life. For example, cerebral palsy is a life-long, developmental disorder that causes marked delays in fine motor skills, gross motor skills, and (sometimes) eating, speaking, 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. The disorder ranges in severity from mild clumsiness to a lack of 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 deficits.

Identification

Between 1% and 3% of infants and toddlers have GDD (Srour & Shevell, 2014). 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 and wide nasal bridge. In most cases, however, pediatricians must order blood tests to screen for genetic disorders. The American Academy of Pediatrics recommends chromosomal microarray (CMA) as a standard test for infants with GDD (Moeschler et al., 2014). 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 “virtual karyotype” of the child’s chromosome structure to identify abnormalities (Flore & Milunsky, 2012). 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 (see Image 5.3). 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, Schwartz, & Rogers, 2012).

Physicians may also order blood or urine tests to screen for metabolic disorders that can cause developmental delays. Some metabolic disorders are phenylketonuria (PKU; described later), hypothyroidism, and lead poisoning. These disorders are relatively rare; only about 1% of youths with GDD have identifiable metabolic problems (Gilissen et al., 2014).

If the results of genetic and metabolic testing are negative, physicians may try to determine the source of children’s delays using neuroimaging. Magnetic resonance imaging (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, or cellular damage and lesions.

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 show vision problems, whereas 18% of children with GDD display significant hearing problems (Shevell et al., 2003). Untreated visual and auditory deficits can greatly interfere with the acquisition of children’s speech, language, and social skills (Shevell, 2008).

Review:

- GDD is characterized by significant delays in several developmental domains (e.g., motor, language, social, or daily living skills) prior to age 5 years. It is a temporary diagnosis used when clinicians suspect ID but the child is too young to administer an intelligence test.
- Between 1% to 3% of infants and toddlers meet criteria for GDD.
- The American Academy of Pediatrics recommends CMA as a first-line test to identify genetic abnormalities in children with GDD.

What Challenging Behaviors Are Associated With Intellectual Disability?

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
they significantly interfere with their safety or social functioning. Approximately one-fourth of youths with ID engage in challenging behavior (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 sports.
- It can interfere with learning and cognitive development.
- It can place a financial burden on families and the public.

Given its seriousness, challenging behavior is a main target for treatment. Although children with ID can show many types of challenging behavior, we will focus on the most common: stereotypies, self-injurious behaviors (SIBs), and aggression (Sturmey, 2014b).

### Stereotypies

Some children with ID show stereotypies, behaviors that are performed in a consistent, rigid, and repetitive manner and that have no immediate, practical significance (Vollmer, Bosch, Ringdahl, & Rapp, 2014). 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 (Machalicek et al., 2016).

Stereotypies are fairly common among children with ID. 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 ID and autism spectrum disorder (ASD) showed stereotyped behaviors.

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). 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 their 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 responses they elicit from 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 pleasant sensations (Sigafos, O’Reilly, Lancioni, Lang, & Didden, 2014).

Approximately 10% to 12% of children with ID 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 (Thompson & Caruso, 2002). Indeed, children with ID and autism may be 5 times more likely than children with ID 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” several 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 ID 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 biological factors (Machalicek et al., 2016).

There are at least three possible explanations for SIBs in children with ID (Thompson & Caruso, 2002). One explanation is that children show SIBs because these behaviors serve a certain purpose or function. For example, some youths with ID engage in SIBs because they lack communication skills (Sigafos et al., 2014). 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 SIB or problematic behavior among people with ID. 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 (see Figure 5.1).

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, they display severe self-injury. Second, healthy rats given high dosages of dopamine also show self-injury. Third, some antipsychotic drugs, which bind to dopamine receptors, decrease SIBs in humans.

A final 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 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 ID who show SIBs display a dramatic increase in endorphins immediately after engaging in self-harm; this increase is much faster than in individuals who do not show SIBs. Second, many people with ID and SIBs have abnormalities in the functioning of endorphin receptors and levels in their brains. Third, some studies indicate that SIBs can be reduced by administering drugs that block endorphin receptors (Thompson & Caruso, 2002).

Physical Aggression

Youths with ID, like their typically developing peers, sometimes engage in aggression (Farmer & Aman, 2011). Physical aggression refers to behavior that causes (or can cause) property destruction or harm to another person. Aggressive acts include throwing objects, breaking toys, ruining furniture, hitting, kicking, and biting others. By definition, aggressive acts are done deliberately, not by accident. However, it is sometimes difficult to determine the intentions of children with severe or profound ID (Didden et al., 2012).

Approximately 20% to 25% of youths with ID show chronic problems with aggression. Aggressive acts are most common among boys, children with comorbid autism, and youths with poor communication skills (Healy, Lydon, & Murray, 2014). Furthermore, there is often an inverse relationship between children’s IQ
scores and the frequency of their aggressive acts (Didden et al., 2012). Many children with ID engage in aggression because they lack communication skills to share their thoughts and feelings in more prosocial ways (Kanne & Mazurek, 2011). Indeed, interventions that help children improve their communication skills are frequently effective in reducing aggression (Sturmey, 2014a).

Matson and colleagues (2011) reviewed the published literature on the causes of aggression in individuals with ID. 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 (Machalicek et al., 2016).

**Comorbid Disorders**

Until recently, many mental health professionals believed that people with ID 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 ID from mental illnesses. Gradually, clinicians became aware that people with ID could suffer from the full range of psychiatric disorders (Pandolfi & Magyar, 2016).

Experts use the term *dual diagnosis* to refer to the presence of mental disorders among individuals with ID. Emerging evidence suggests that approximately 40% of children and adolescents with ID have a dual diagnosis. The most common co-occurring disorders are disruptive behavior disorders (25%), ADHD (9%), and anxiety (9%; Witwer et al., 2014).

Unfortunately, clinicians frequently overlook the presence of mental disorders in youths with ID, a phenomenon called *diagnostic overshadowing*. Why might clinicians miss anxiety, depression, and even psychotic symptoms in people with ID? Some mental health professionals simply do not have much experience in assessing and treating people with ID. Others erroneously attribute psychiatric problems to the person’s low intelligence or problems in adaptive functioning (Einfeld et al., 2006; Koskentausta, Iivanainen, & Almqvist, 2007).

**Review:**

- Approximately 25% of youths with ID exhibit challenging behaviors such as stereotypies, SIBs, or physical aggression.
- Challenging behaviors can be harmful to the child or others, strain social relationships, limit children’s access to educational or social opportunities, and place a financial burden on families.
- Approximately 40% of youths with ID have a comorbid mental disorder. Comorbid conditions are easily overlooked in youths with ID.

### How Common Is Intellectual Disability?

Experts disagree about the prevalence of ID. If we assume that IQ scores are normally distributed in the population, we would expect approximately 2.2% of individuals in the general population to earn IQ scores less than 70. Consequently, some people estimate the prevalence of ID to be between 2% and 3% of the general population (Hodapp, Zakemi, Rosner, & Dykens, 2006).

Other experts argue that the prevalence of ID is lower (Tirosch & Jaffe, 2011). A meta-analysis suggested that approximately 1.83% of individuals have ID (Yeargin-Allsopp, Boyle, & van Naarden, 2008). There are several reasons for this lower estimate. First, ID 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 ID.

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 ID 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.

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 ID is likely lower than expected based on the normal curve.

The prevalence of ID varies by age. ID 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 ID, 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, 2015). Why are more school-age children classified as having ID than people in the general population? The answer seems to be that the cognitive impairments associated with ID 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.

ID 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 ID than females. Some people believe the male central nervous system is more susceptible to damage. Others believe that males are more likely to show ID than females because some forms of ID 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).

**Review:**
- Meta-analyses indicate that approximately 1.8% of the population has ID.
- ID is more commonly diagnosed in school-age children (compared to adults) and boys (compared to girls).

### 5.2 CAUSES

**What Is the Difference Between Organic and Cultural–Familial Intellectual Disability?**

**Zigler’s Classification**

Edward Zigler (1969) proposed one of the first methods to classify children with ID based on the cause of their impairments. Zigler divided children with ID into two groups (see Table 5.4). The first group consisted of children with identifiable causes. He classified these children with **organic ID** because most of the known causes of ID at that time involved genetic disorders or biological abnormalities, such as Down syndrome. As a group, children with organic ID had IQ scores less than 50, physical features indicating neurological problems, and medical complications. Children with organic ID 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” ID because relatives often had low levels of intellectual and adaptive functioning. Today, many experts refer to people in this category as experiencing cultural–familial ID because children in this group are believed to experience ID 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 ID does not necessarily have a genetic cause for his impairments. Similarly, the deficits shown by a child with familial ID are not necessarily caused by environmental factors. The organic/familial

<table>
<thead>
<tr>
<th>Table 5.4 Intellectual Disability Classified Into Organic Versus Cultural-Familial Types</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Organic</strong></td>
</tr>
<tr>
<td><strong>Definition</strong></td>
</tr>
<tr>
<td>• Child shows a clear genetic or biological cause for his/her Intellectual Disability</td>
</tr>
<tr>
<td>• Biological relatives may have low IQ</td>
</tr>
<tr>
<td><strong>Diagnosis</strong></td>
</tr>
<tr>
<td>• Usually diagnosed at birth or infancy</td>
</tr>
<tr>
<td>• Frequent comorbid disorders</td>
</tr>
<tr>
<td><strong>Intelligence and Adaptive Functioning</strong></td>
</tr>
<tr>
<td>• IQ usually ≤ 50</td>
</tr>
<tr>
<td>• Siblings with normal IQ</td>
</tr>
<tr>
<td>• Greater impairment in adaptive functioning</td>
</tr>
<tr>
<td>• Often dependent on others</td>
</tr>
<tr>
<td><strong>Associated Characteristics</strong></td>
</tr>
<tr>
<td>• Similar across ethnicities and SES groups</td>
</tr>
<tr>
<td>• Associated with health problems and physical disabilities</td>
</tr>
<tr>
<td>• Higher mortality rate</td>
</tr>
<tr>
<td>• Unlikely to mate, often infertile</td>
</tr>
<tr>
<td>• Often have facial abnormalities</td>
</tr>
</tbody>
</table>

*Source:* Based on Iarocci and Petrill (2012).

*Note:* Although these names are somewhat misleading, they are useful for broadly differentiating people with intellectual disabilities.
distinction is based solely on whether we can identify the cause of the child’s ID. For example, some cases of organic ID are caused by environmental factors, such as mothers’ alcohol use during pregnancy. Similarly, some types of familial ID 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 ID, was not identified until 1991. Even today, as many as 80% of people with Fragile X do not know they have the disorder. As genetic and medical research advances, it is likely that more causes of ID will be uncovered (Karmiloff-Smith, Doherty, Cornish, & Scerif, 2016).

**Similar Sequence and Similar Structure Hypotheses**

Typically developing children progress through a series of cognitive stages in a reliable order (Carey, Zaithchik, & Bascandziev, 2015). 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 ID is similar to the sequence of cognitive development seen in typically developing children. His **similar sequence hypothesis** posits that children with ID 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 ID 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 ID and the other without ID) will show similar abilities. According to the similar structure hypothesis, a 16-year-old with ID 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 ID has generally supported the similar sequence and similar structure hypotheses. Children with cultural–familial ID 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 ID generally show similar cognitive abilities as children without ID of the same developmental age (Weisz, 1990).

Subsequent research involving children with organic ID has yielded mixed results. The cognitive development of children with organic ID does follow an expected sequence, similar to the development of typically developing children. However, children with organic ID often show different cognitive abilities than typically developing children of the same mental age. Specifically, children with organic ID 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 ID. 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 ID show characteristic patterns of cognitive abilities is important. If scientists could identify the cognitive and behavioral characteristics associated with each known cause for ID, this information could be used to plan children’s education and improve their adaptive functioning (Carlier & Roubertoux, 2015).

Consequently, researchers have moved away from lumping all children with known causes of ID into one large “organic” category. Instead, researchers study children with ID 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 ID. 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 ID. 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 evidence-based interventions (Carlier & Roubertoux, 2015).

**Review:**

- The term **organic ID** was used to describe children who had identifiable causes for their intellectual and adaptive disabilities. Usually, they had genetic disorders, earned very low IQ and adaptive functioning scores, experienced medical complications, and had no family history of ID.
- The term **cultural–familial ID** was used to describe children with no identifiable cause for their intellectual and adaptive disabilities. Usually, they earned IQ and adaptive scores in the 50–70 range, were physically healthy, and had family members with ID.
- Most research supports the similar sequence hypothesis—that is, youths with ID progress through the same stages of cognitive development as typically developing peers, albeit at a slower pace.
In contrast, there is limited support for the similar structures hypothesis. Some causes of ID are associated with behavioral phenotypes—that is, specific patterns of behavior and cognitive strengths and weaknesses.

How Can Chromosomal Abnormalities Cause Intellectual Disability?

Researchers have identified more than 800 unique causes of ID. They can be loosely organized into five categories: (1) chromosomal abnormalities, (2) X-linked disorders, (3) metabolic disorders, (4) embryonic teratogen exposure or illness, and (5) complications during or after delivery. Altogether, these conditions explain approximately 70% of cases of ID (see Figure 5.2).

Down Syndrome

Down syndrome is a genetic disorder characterized by moderate to severe ID, 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 (see Figure 5.3).

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. 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 (see Image 5.4). Other physical features include short stature and poor muscle tone. Children with Down syndrome show small overall brain size and fewer folds and convolutions than in the 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 ID; 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.

Figure 5.2 The Causes of Intellectual Disability. Genetic and metabolic disorders explain 50% of cases.

<table>
<thead>
<tr>
<th>“CULTURAL-FAMILIAL”</th>
<th>Unknown Causes 30%</th>
</tr>
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<tbody>
<tr>
<td>Genetic Disorders 40%</td>
<td></td>
</tr>
<tr>
<td>Perinatal Problems or Injury 10%</td>
<td></td>
</tr>
<tr>
<td>Teratogens 10%</td>
<td></td>
</tr>
<tr>
<td>Metabolic Disorders 10%</td>
<td></td>
</tr>
</tbody>
</table>

“ORGANIC”

Source: Based on Toth, deLacy, and King (2015).
Youth 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 ID (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 40, 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 65 years.

Prader-Willi Syndrome

Prader-Willi syndrome (PWS) is a noninherited genetic disorder characterized by mild ID, overeating and obesity, oppositional behavior toward adults, and obsessive–compulsive behavior (see Image 5.5). 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. 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 ID or borderline intellectual functioning. Average IQs range from 65 to 70 (Dykens & Shah, 2003). Children with PWS show relatively strong 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. Furthermore, their adaptive behavior is usually much lower than their IQ.

The most striking feature of many children with PWS is their intense interest in food (Dykens, 2003; Dykens & Cassidy, 1999). Infants with the disorder show problems with sucking, feeding, and gaining weight. 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 become obese. Medical complications associated with obesity are a leading cause of death among adults with PWS (Dykens & Shah, 2003). Consider the case of Dontrell, a boy with ID caused by PWS (see next page).

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 & Kasari, 1997). Approximately 42% of children with PWS destroy property during their disruptive outbursts, and 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
CASE STUDY

INTELLECTUAL DISABILITY (PRADER-WILLI SYNDROME)

Obsessed With Food

Dontrell was a 6-year-old boy referred to our clinic by his pediatrician because of significant delays in language and self-care skills. Dontrell recently entered the public school system after his maternal grandmother assumed caregiving responsibilities for him. His mother, a migrant worker, returned to Mexico but wanted Dontrell to remain in the United States and attend school.

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. He had limited vocabulary in English and Spanish, struggled to recite the alphabet or recognize letters, and had difficulty counting. He also had problems performing self-care tasks typical of children his age, such as dressing, bathing, and grooming. School officials attributed these developmental delays to a history of poor medical care and nutrition and impoverished learning experiences as a toddler and preschooler.

Dontrell also 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 throw a tantrum. 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. Indeed, Dontrell weighed nearly 85 lbs!

Dr. Valencia, the psychologist who performed the evaluation, recognized that many of Dontrell’s behaviors were consistent with a genetic disorder called Prader-Willi syndrome (PWS). Although most children with this disorder are recognized in infancy or toddlerhood, Dontrell was likely overlooked because he did not receive regular medical care. She suggested genetic testing to confirm her diagnosis.

Angelman Syndrome

Angelman syndrome is a genetically based developmental disorder characterized by ID, speech impairment, happy demeanor, and unusual motor behavior. The disorder was identified by the 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 ID, 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 (see Image 5.6). 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. 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 syndrome. Approximately 1 per 15,000 to 20,000 children have the disorder.

Both PWS and Angelman 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 syndrome occurs when children inherit genetic information on chromosome 15 only from the father. In 70% of cases of Angelman 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 the remaining cases of Angelman syndrome, the child shows other genetic mutations in chromosome 15 or the cause is unknown.
The most striking feature of Angelman syndrome is the persistent social smile and happy demeanor shown by children with the disorder. Many infants with Angelman syndrome begin this persistent smiling between 1 and 3 months of age. Later in development, it is accompanied by laughing, giggling, and happy grimacing. Facial features of children with Angelman syndrome often include a wide smiling mouth, thin upper lip, and pointed chin (Williams, 2005).

Despite children's social smiling, Angelman 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, intellectual functioning is generally in the range of severe or profound ID. Most children with Angelman 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 syndrome show hyperactivity and inattention. Parents usually describe them as "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 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 syndrome have hypopigmentation—that is, they may appear pale and have light-colored eyes. Hypopigmentation occurs when the gene that codes for skin coloring is deleted along with other information on chromosome 15.

More than 90% of children with Angelman syndrome have seizures. Sometimes, seizures are difficult to notice because of these children's sporadic movements. In most cases, physicians prescribe anticonvulsant medications to reduce the number and severity of seizures. Adults with Angelman syndrome have life expectancies approximately ten to fifteen 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 overall intellectual functioning, hyperactivity, impulsivity, inattention, and unusual strengths in spoken language and sociability (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 (see Image 5.7). 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. 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 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. They are especially good at remembering faces and inferring a person’s mental state and emotions based on his or her affect. 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 ghosts, storms, and vaccinations. However, unlike typically developing children, older children with WS continue to fear these stimuli and show a marked increase in generalized anxiety with age. In particular, older children with WS often worry 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 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 ID 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 place them at risk for developing fear 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 the disorder.

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 and other heart problems.

**22q11.2 Deletion Syndrome**

Children with 22q11.2 DS are at risk for developing ID because of missing genetic material on a portion of one pair of the 22nd chromosome. In approximately 10% of cases, the child inherits an abnormal chromosome from a parent. In most instances, however, genetic information is lost during fertilization. Approximately 1 in 2,000–4,000 children have this condition (Toth, deLacy, & King, 2015).

Children with 22q11.2 DS can show a wide range of physical and behavioral features (see Image 5.8). Physical characteristic usually include cleft lip and/or palate, small ears, and small mouth and chin. These youths often experience health problems including congenital heart problems, middle ear infections or hearing loss, immune problems, and seizures. The cleft lip and/or palate and hearing loss often cause these children to experience language delays and learning problems. Their average IQ is approximately 70 with significantly higher verbal skills than nonverbal skills (Butcher et al., 2012).

Children with 22q11.2 DS are at risk for psychiatric disorders. Young children with this condition often experience problems understanding social situations and show impaired social problem-solving. Some develop autistic-like symptoms. Older children with 22q11.2 DS often display problems with attention and may be diagnosed with ADHD. Approximately one-half of these youths

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**Image 5.8** Children with 22q11.2 DS often have cleft palate and small ears. Their average IQ is 70 with higher verbal than nonverbal skills. They are at risk for ADHD in childhood and schizophrenia in adulthood.
develop schizophrenia or psychotic symptoms, making 22q11.2 DS one of the few known causes for schizophrenia (Vorstman et al., 2015).

Review:

- Down syndrome (trisomy 21) is associated with moderate ID, characteristic appearance, weakness in verbal skills and language, strength in visual–spatial reasoning, and sociability.
- PWS is caused by missing paternal genetic material on chromosome 15. It is associated with mild ID, weakness in short-term memory, strength in visual–spatial reasoning, hyperphagia, and obsessive–compulsive behavior.
- Angelman syndrome is caused by missing maternal genetic material on chromosome 15. It is associated with moderate to severe ID, sporadic/jerky movements, lack of spoken language, hyperactivity, and persistent social smile.
- WS is caused by deletions on chromosome 7. It is associated with mild ID, well-developed spoken language, strengths in auditory memory, weakness in visual–spatial reasoning, hyperactivity, anxiety, and friendly demeanor.
- 22q11.2 DS is caused by deletions on chromosome 22. It is associated with mild to moderate ID, cleft lip/palate, ADHD, and risk for schizophrenia later in life.

How Can X-Linked Disorders Cause Intellectual Disability?

X-linked disorders are a specific class of genetic abnormalities that occur on the X-chromosome. Because girls have two X chromosomes and boys have only one, these disorders typically manifest differently across gender.

Fragile X Syndrome (FMR-1 Disorder)

Fragile X syndrome is an inherited genetic disorder that is associated with physical anomalies, moderate to severe intellectual impairment, and social behavior problems. It occurs in 1 per 4,000 boys and 1 per 8,000 girls (Toth et al., 2015).

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, Cole, Longhi, Karmiloff-Smith, & Scerif, 2013). 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. Children with Fragile X syndrome show an unusually high number of CGG repeats. Children who inherit 50 to 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. In general, the less FMRP produced, the more severe children’s cognitive impairments. Brain scans of children with Fragile X show abnormalities of the prefrontal cortex, caudate nucleus, and cerebellum, presumably from less FMRP production. The disorder is called Fragile X because the X chromosome appears broken (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 their only X chromosome is affected. 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 of FMRP and contributes to higher cognitive functioning (Cornish et al., 2013).

Boys with Fragile X tend to have elongated heads, large ears, hyperflexible joints, and large testicles after puberty (Sadock & Sadock, 2015). They also tend to be shorter than other boys. Medical problems sometimes associated with Fragile X include heart murmur and crossed eyes (see Image 5.9).

Boys with Fragile X syndrome tend to show moderate to severe ID (Abbeduto, McDuffie, Brady, & Kover, 2012). Additionally, they show a curious pattern of strengths and weaknesses in the way they process information and solve problems. 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.
processing. Alternatively, boys with Fragile X syndrome show relative deficits in sequential processing, or 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 (Karmiloff-Smith et al., 2016).

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. As many as 90% have ADHD (Dykens & Hodapp, 2001; Sullivan, Hooper, & Hatton, 2007).

Girls with Fragile X tend to have higher IQs, less noticeable physical anomalies, and less severe behavior problems than do boys with the disorder. Like boys, girls may have problems with attention. They may also show excessive shyness, gaze aversion, and social anxiety (Hodapp & Dykens, 2006).

**Rett Syndrome (MECP-2 Disorder)**

Rett syndrome is the most common cause of severe ID in girls. It is almost always caused by a genetic mutation in the MECP-2 gene of the X chromosome; it is rarely inherited. The disorder is almost always seen in girls; boys with the condition often die during or shortly after fertilization. The disorder affects 1 in 8,500 to 10,000 children (Toth et al., 2015).

Infants with Rett syndrome display normal development for the first 6 to 18 months of life. Then, they usually show a rapid deterioration in their language, motor, and social skills. Most will display social withdrawal and develop stereotypic hand-wringing movements (see Image 5.10). Many will also show health problems such as growth failure, breathing problems, loss of movement, and scoliosis (i.e., curvature of the spine). As toddlers, many develop emotion-regulation problems such as episodes of crying and screaming. Approximately 90% have seizures. Youths with Rett syndrome can live into adulthood with support from caregivers (Lyst & Bird, 2015).

**Review:**

- Fragile X syndrome is an inherited, X-linked disorder that adversely affects boys more than girls. It is characterized by mild to moderate ID, characteristic appearance, strengths in simultaneous processing, weakness in sequential processing, and social deficits.
- Rett syndrome is usually caused by a genetic mutation in a portion of the X chromosome. It almost always affects girls. It is characterized by typical development in early infancy followed by rapid deterioration in social functioning and language, severe ID, hand-wringing stereotypies, and seizures.
How Can Metabolic Disorders Cause Intellectual Disability?

Phenylketonuria (PKU) is the most well-known metabolic disorder that can lead to ID if untreated. 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 ID.

PKU is caused by a recessive gene (see Figure 5.4). In order for a child to show PKU, he must inherit the gene from 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 ID and they lack spoken language. Children with untreated PKU are also hyperactive, show erratic movements, and throw tantrums. Additionally, they may experience gastrointestinal problems and have seizures. These impairments are irreversible, even if a phenyl-free diet is initiated later in childhood.

Review:
- PKU is an inherited disorder characterized by an inability to break down phenylalanine, an amino acid in many foods (e.g., dairy, meats).
- Restricting phenylalanine can prevent severe ID, seizures, and other medical problems.

Figure 5.4 PKU is an Autorecessive Metabolic Disorder

![Figure 5.4 PKU Is an Autorecessive Metabolic Disorder](image)

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.
How Can Maternal Illness or Environmental Toxins Cause Intellectual Disability?

**Maternal Illness**

Infections acquired by mothers during pregnancy can result in preterm birth; low birth weight; and, in some instances, ID (Silasi et al., 2015). The five main classes of maternal illnesses that can contribute to cognitive problems in children are represented by the acronym TORCH.

T: *Toxoplasma* infection is caused by a parasite that exists in warm-blooded animals throughout the world. In developed countries, pregnant women are most likely to acquire this parasite by consuming undercooked meat or handling animal droppings. Consequently, pregnant women should avoid cleaning cat boxes. Toxoplasmosis affects the development of the central nervous system. Symptoms may not appear until late infancy or toddlerhood in some children.

O: *Other infections* that can cause ID include the varicella zoster virus (which causes chickenpox), syphilis, hepatitis B, and HIV/AIDS. Mothers can avoid varicella infection with the chickenpox vaccine or by limiting contact with affected individuals. The other conditions can often be avoided by altering sexual behavior. If a mother becomes infected, syphilis can be treated with penicillin. Although there is no cure for hepatitis B or HIV/AIDS, several strategies can reduce the likelihood that the virus will be transmitted to the fetus: the use of antiviral medications during gestation, delivery by Caesarian section, and bottle-feeding rather than breastfeeding. For example, the likelihood of mother-to-infant transmission of HIV is 25% and <1% with and without these prevention strategies, respectively.

R: *Rubella* is a virus that causes German measles. Although maternal symptoms are relatively minor (i.e., a rash), infants infected with rubella typically show moderate to severe cognitive impairment. Infection is most serious during the first trimester of gestation. With the widespread use of the rubella vaccine, fetal infection is relatively rare in developed countries. However, women who refuse vaccination or who live in countries where vaccination is not practiced continue to be at risk.

C: *Cytomegalovirus* (CMV) is an extremely common virus that most people acquire during adolescence or early adulthood. It causes mononucleosis, an illness characterized by a high fever, chills, sore throat, and fatigue. CMV can be transmitted to a fetus if the mother acquires the illness for the first time during gestation. The illness can cause significant damage to the fetal nervous system, hearing loss, ID, and death.

Consequently, pregnant women should avoid individuals who experience symptoms of mononucleosis.

H: *Herpes simplex virus type 2* (HSV-2) is an infection that is typically acquired through sexual contact. Approximately 20% of women aged 14 to 49 have the illness. Fetuses typically acquire the virus in two ways: (1) if a mother first acquires the infection during the final trimester of pregnancy or (2) if a mother has an active herpes lesion that comes into contact with the infant during delivery. Infants who acquire the virus can experience severe damage to their central nervous system, blindness, seizures, and ID. Although there is no cure for HSV-2, mothers can prevent fetal transmission by using antiviral medication and delivering by Caesarian section.

One final illness deserves special mention: *Zika virus disease* (Rasmussen, Jamieson, Honein, & Petersen, 2016). The disease is caused by the Zika virus and is usually acquired through infected mosquitoes or sexual contact. The disease causes fever, rash, joint pain, and conjunctivitis in adults. Symptoms are usually minor. Unfortunately, fetuses can acquire the virus during gestation. Zika can cause severe damage to the developing central nervous system, microcephaly (i.e., small head circumference), cognitive impairment, and death (see Image 5.11; Brasil et al., 2016; Centers for Disease Control and Prevention, 2016f).

**Lead Exposure**

Lead is a neurotoxin that can cause developmental disabilities in children. Fetuses are at risk for lead toxicity if their mothers are exposed to lead during gestation. For example, pregnant women may inadvertently ingest lead through contaminated drinking water or by working in industrial facilities. In high amounts, ingested lead can pass through the placenta and affect fetal development. A mother’s history of lead exposure can also place her fetus at risk. For example, a mother exposed to lead during her childhood and adolescence may store it in her bones. If she does not consume sufficient calcium during pregnancy, her body may substitute this stored lead for the calcium her baby needs. In either case, newborns exposed to lead during gestation are at risk for premature birth, low birth weight, and cognitive problems (Shah-Kulkarni et al., 2016).

Young children can be exposed to lead in three ways. First, children can drink contaminated tap water that contains high lead levels (see Figure 5.5). Second, children can be exposed to lead from playing near industrial sites that use lead in manufacturing. Third, and most commonly, children can ingest lead-based paint. Although lead-based paint was banned in the United States in 1978, approximately 24 million older homes and apartments still contain lead-based paint. Eventually, this paint can chip or flake. Infants may eat paint chips on the floor and children may inadvertently inhale paint dust containing lead (Eid & Zawia, 2016).
Lead exposure is typically measured with a blood test. Levels greater than 5 micrograms per deciliter (>5μg/dL) are considered “elevated” (Centers for Disease Control and Prevention, 2016a). However, no level of lead exposure is considered safe. Even lower lead levels are associated with neurodevelopmental problems in some children.

The effects of lead on development vary as a function of the child’s age and the amount and duration of exposure. In general, lead is most dangerous to youths 0 to 5 years, because of the rapid development of the nervous system that occurs during this developmental period. Lead is strongly associated with cognitive problems,
especially lower intelligence and increased risk for learning disabilities. Recent longitudinal studies indicate that every 1μg/dL increase in lead is associated with a 1- to 2-point IQ reduction. These cognitive deficits typically persist into adulthood (Mazumdar et al., 2011).

Infants and toddlers exposed to lead are also at risk for behavior problems. Most commonly, children experience problems with attention and concentration, hyperactivity, and disruptive behavior including aggression. Early lead exposure is also associated with increased risk for conduct problems in adolescence.

Unfortunately, most children do not show clear signs of lead toxicity until their lead levels approach 40 to 50μg/dL. Symptoms of lead toxicity in young children include cognitive delays and learning problems, irritability, sluggishness, loss of appetite, and gastrointestinal problems. Children with very high levels can receive chelation therapy. Chelation is injected into the child’s bloodstream, where it binds to lead and helps remove it from the body. Chelation therapy is used only in extreme instances of lead toxicity because it can cause kidney damage, heart problems, and other metabolic difficulties.

How can families prevent lead exposure? First, families who live in older homes can inspect walls, windows, and porches for chipping or flaking paint. Deteriorating surfaces can be professionally treated and repainted to protect children (see Image 5.12). Second, parents should regularly clean floors and other surfaces of their home as well as their children’s hands and toys. Parents should limit their children’s exposure to toys that might contain lead-based paint. Third, families should remove their shoes before entering the house to prevent them from bringing in lead-contaminated soil. Children should also avoid playing outside while barefoot. Finally, families whose tap water comes from a public water system can check with their water supplier for information about lead levels. Water suppliers are required to provide customers with a consumer confidence report that describes water quality each year. Families with a private water supply (e.g., a well) can check lead levels using a kit (Centers for Disease Control and Prevention, 2016d).

Alcohol and Other Drugs

Many drugs, if ingested by pregnant women, are associated with low birth weight, reduced 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 ID as are more socially accepted drugs like alcohol.

**Fetal alcohol spectrum disorder (FASD)** is caused by maternal alcohol consumption during pregnancy. FASD is characterized by lower intellectual functioning, learning disabilities, hyperactivity, and slow physical growth as well as characteristic craniofacial anomalies. Some children with FASD have cardiac problems. FASD is called a spectrum disorder because the severity of symptoms range from mild to severe, depending on the mother’s use of alcohol during gestation (Popova et al., 2016).

The prevalence of FASD 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 FASD. Some data indicate that FASD can occur from only 2 to 3 oz. of alcohol per day during gestation. Furthermore, binge drinking during pregnancy greatly increases the chance of FASD. Although occasional consumption of alcohol during pregnancy may not produce severe symptoms, it may lead to subtle cognitive, behavioral, and physical abnormalities, such as mild learning problems, reduced attention span, or short stature. Consequently, the American Academy of Pediatrics states that no amount of alcohol during pregnancy is safe and that women should abstain from alcohol during gestation (Williams, Smith, & Committee on Substance Abuse, 2015).

The intellectual functioning of children with FASD varies considerably. Most children with FASD show mild to moderate ID, 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 FASD are hyperactivity, impulsivity, and inattention. Young children with FASD are often diagnosed with ADHD. Older children and adolescents with FASD report feelings of restlessness and difficulty sustaining attention on reading and homework.

Children with FASD are at risk for conduct and mood problems as they enter late childhood and adolescence. They may experience peer problems and teasing because of their cognitive impairment. Furthermore, they may become depressed because of their academic deficits, behavior problems, or stigmatization associated with the disorder (Popova et al., 2016). Consider the case of Andrew, a boy with moderate FASD.

**CASE STUDY**

**FETAL ALCOHOL SPECTRUM DISORDER**

**Tough Love**

Andrew was a 14-year-old boy 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 FASD including wide-spaced eyes, upturned nose, low-set ears, and broad face. Andrew’s mother had an extensive history of alcohol and other drug use. She drank throughout her pregnancy 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 silly 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.”
How Can Perinatal or Postnatal Problems Cause Intellectual Disability?

Complications With Pregnancy and Delivery

Complications that occur during gestation or delivery can contribute to ID. Maternal hypertension or uncontrolled diabetes during pregnancy is sometimes associated with ID 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 ID. Anoxia can occur when the umbilical cord wraps around the fetus's throat, interfering with oxygen intake.

One of the greatest predictors of cognitive problems in children is premature birth and/or low birth weight. Children born before 36 weeks' gestation are 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 (see Figure 5.6). 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 (Kerstjensk, DeWinter, Bocca-Tjeertes, Bos, & Reijnveld, 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, axons, and synapses; increased myelination; and more complex brain activity. Although maturation can (and does) continue after delivery, optimal development occurs in utero (Volpe, 2008).

Childhood Illness or Injury

The two childhood illnesses most associated with the development of ID 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 resistant to treatment.

All serious head injuries have the potential to cause ID. 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 (Appleton & Baldwin, 2006). Children exposed to certain types of physical maltreatment are also at risk for ID and other cognitive problems (Shaahinfar, Whitelaw, & Mansour, 2015). Abusive head trauma (more commonly called shaken baby syndrome)

Figure 5.6 Preterm Birth Places Infants at Greater Risk for Delays

Source: From Kerstjensk 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.
occurs when caregivers induce brain injury by forcefully shaking an infant or toddler. Very young children have weak neck muscles; therefore, any amount of back-and-forth shaking can cause trauma to the brain. Usually, caregivers engage in this behavior when frustrated by their baby’s crying. Children’s symptoms range from irritability, lethargy, and poor appetite to vomiting, tremors, and death. Infants who survive such shaking frequently experience cognitive deficits such as ID, learning disabilities, and attention and memory problems.

Review:
- Anoxia during gestation or delivery and preterm birth are risk factors for ID and learning problems in childhood.
- Encephalitis, meningitis, and high fever in childhood can also contribute to ID.
- Head injuries during infancy and childhood, especially abusive head trauma (i.e., shaken baby syndrome) are risk factors for ID.

What Causes Cultural–Familial Intellectual Disability?

Cultural–familial ID results from the interaction of the child’s genes and environmental experiences over time. Some children inherit a genetic propensity toward low intelligence. When these children also experience environmental deprivation that interferes with their ability to reach their cognitive potentials, they are at risk for below-average intelligence and adaptive functioning. Environmental deprivation might include poor access to healthcare, inadequate nutrition, lack of cognitive stimulation during early childhood (e.g., parents talking, playing, and reading with children), low-quality early education, and a general lack of cultural experiences during early childhood (e.g., listening to music, trips outside the home). Over time, the interaction between genes and environment can contribute to ID.

Cultural–familial ID is more prevalent among children from low-income families than middle-class families. The correlation between socioeconomic status (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 & Horn, 2015).

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 such as cigarette smoke or lead, receive less cognitive stimulation from their home environments, and attend less optimal schools. These environmental deficits or risk factors can limit their cognitive and adaptive skills (von Stumm & Plomin, 2015).

Brain imaging studies confirm the risk of poverty on children’s cognitive development (Luby, 2015). In one of the largest studies conducted to date, researchers used MRI to measure brain size in a large group of school-age children. Children living in poverty showed an 8% to 10% reduction in brain volume compared to youths not living in poverty. Reductions were greatest in areas responsible for problem-solving and memory, especially the frontal lobe and hippocampus. Furthermore, these reductions were associated with a 6-point decrease in IQ and a 4- to 7-point decrease in academic achievement (Hair, Hanson, Wolfe, & Pollak, 2015; von Stumm & Plomin, 2015).

Many studies suggest a relationship between the quality of the home environment and children’s intellectual development. After reviewing the data, Sattler (2014) 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 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.

Review:
- Cultural–familial ID is associated with a genetic predisposition toward lower intellectual functioning and environmental experiences that restrict the development of intelligence and adaptive functioning (e.g., low-quality nutrition, health care, schooling).
- Caregivers can promote their children’s intellectual functioning by providing verbal stimulation and encouraging academic achievement, curiosity, and independence at home.

5.3 PREVENTION AND TREATMENT

How Do Professionals Screen for Developmental Disabilities?

Shortly after birth, newborns are routinely administered a series of blood tests to detect genetic and metabolic disorders.
that cause ID. For example, all infants are screened 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 ID 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 (Cuckle, Pergament, & Benn, 2015). 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 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 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% to 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 wisplike 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 disorder. The risk of miscarriage associated with CVS is 0.5% to 1.5%.

Finally, physicians can use ultrasound to detect structural abnormalities in the fetus that might indicate the presence of a developmental disorder (Rissanen, Niemimaa, Suonpää, Ryynänen, & 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 (see Image 5.13).

Review:
- Serum screening is a maternal blood test that can be conducted 15 to 18 weeks’ gestation to detect the presence of some developmental disorders.
Amniocentesis and CVS are more invasive procedures that may be used when there is an elevated possibility of a developmental disability.

Physicians can use ultrasound to detect structural abnormalities in the fetus that might indicate Down syndrome.

Can Early Education Programs Prevent Intellectual Disability?

Infants and Toddlers

A number of state- and locally administered programs have been developed to prevent the emergence of ID in infants at risk for below-average intelligence. The Infant Health and Development Program (IHDP) is one of the largest. Participants in the IHDP were 985 premature, low birth weight infants. Previous research indicated that these children were at increased risk for developmental delays, including ID (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 helped parents address problems associated with caring for a preterm, low birth weight infant. When infants turned 1, parents were invited to enroll them in a high-quality preschool program. The program was free and transportation to and from the preschool 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), at age 5, and at age 8. 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 in at-risk children, but increases in IQ are not maintained over time. Experts have disagreed on how to interpret the findings. Critics of the IHDP argue that early intervention programs do not prevent ID 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.

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. 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 et al., 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 prevention program.

Head Start and Preschool Prevention

Other studies have investigated whether providing early intensive preschool education might improve children's cognitive outcomes (Yoshikawa et al., 2013). Early studies focused largely on young children at risk for lower intelligence and academic problems because of socioeconomic disadvantage. For example, the Perry Preschool Project was administered to low-income children. Most children participated in a high-quality, daily preschool program for 2 years taught by well-trained teachers. Teachers also visited families in their homes to encourage parents to take a greater role in their children's preschool education. Similarly, the Abecedarian Project delivered high-quality, year-round child care and preschool to disadvantaged children. The intervention emphasized language development, literacy, and problem-solving.

Perhaps the best-known prevention program is Head Start, an intervention that provides comprehensive early childhood education, health, nutrition, and parent-involvement services to lower-income preschoolers and their families. Head Start was originally designed as a summer program for at-risk preschoolers to give them a “head start” before beginning kindergarten. Today, Head Start provides preschool services to 1,000,000 children in the United States including preschoolers in Native American communities, the children of migrant workers, and homeless youths.

Most recently, communities have begun to offer universal early childhood education programs—that is, preschool programs designed for all children regardless of socioeconomic risk. For example, the Tulsa (Oklahoma) and Boston Pre-K Programs offered free, voluntary preschool education for one or two years. On average, these programs deliver intensive, high-quality educational programs to approximately 75% of youths in their communities.

Do preschool prevention programs boost children's IQ? Overall, results have been mixed (Yoshikawa et al., 2013). The available data from many different evaluations suggest the following:

1. Early childhood interventions can significantly increase children's cognitive skills. Recent meta-analyses indicate that children who participate in these preschool programs earn IQ scores 4 to 5 points higher than controls. Furthermore, youths (especially girls) who participate in these programs earn higher reading and math achievement scores than controls. On average,
youths who participate in preschool programs are approximately one-third of an academic year ahead of their kindergarten classmates who did not attend these programs. The benefits of preschool are greatest for minority children, youths from low-SES backgrounds, and youths at risk for developmental disability (Magnuson et al., 2016).

2. The highest-quality preschool programs were associated with the greatest increase in children's cognitive skills. For example, children who participated in the high-quality Tulsa and Boston Pre-K programs earned reading scores approximately one academic year ahead of other children who did not participate in these programs (see Figure 5.7). The highest-quality programs were characterized by stimulating interactions between teachers and children; a curriculum that focused on language, literacy, and math; and an emotionally supportive learning environment. Furthermore, teachers received ongoing training and supervision (Phillips & Meloy, 2016).

3. The effects of preschool programs on other aspects of children's functioning were mixed. Overall, these programs were not associated with large improvements in children's behavior. However, programs that targeted specific aspects of children's social–emotional functioning were associated with modest improvement in behavioral inhibition and attention. Furthermore, Head Start, which specifically targets children's health outcomes, is associated with increases in child immunizations. Immunizations, in turn, can prevent serious childhood illnesses that hinder cognitive development or cause youths to miss school.

4. The benefits of preschool programs fade over time. Although children who participate in preschool programs show early gains, most of these gains are lost in elementary school. Although experts do not agree what causes this loss, many believe it is due to low-quality primary education, especially in disadvantaged school districts (Hill, Gormley, & Adelstein, 2015; Yoshikawa et al., 2013).

5. Some children show long-term benefits from preschool prevention programs, however. Recent, longitudinal studies indicate that at-risk boys who participate in preschool programs are less likely to repeat a grade in school, less likely to be referred for special education, and more likely to graduate than boys who do not participate in preschool programs (Magnuson et al., 2016).

Altogether, these data indicate that preschool programs can give young children a head start for kindergarten. However, their effects are typically modest, may disappear over time, and depend on sufficient resources to deliver high-quality services (Jenkins, Farkas, Duncan, Burchinal, & Vandell, 2016).

Review:
- The IHDP showed that early childhood prevention programs can boost IQ among high-risk youths, but most gains are not maintained over time.
- Preschool prevention programs (e.g., Tulsa and Boston Pre-K, Head Start) show boosts of 4 to 5 IQ points compared to controls. Prevention programs are most effective for girls, ethnic minority children, youths from low-SES backgrounds, and children at risk for developmental disabilities.
- Most studies indicate that the IQ benefits of preschool programs fade over time. However, youth who participate in these programs may be less likely to repeat a grade, be referred to special education, or drop out of school than controls.

What Services Are Available to School-Age Children?

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. 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 ID in classrooms alongside typically developing peers,
to the maximum extent possible. At first, mainstreamed children with ID were allowed to participate in 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).

Unfortunately, subsequent research showed that children with ID who were assigned to special education classes actually earned lower academic achievement scores and had poorer adaptive functioning than children with ID who spent most of the school day with typically developing peers (Sturmey, 2014a). Consequently, many parents argued that children with ID and other disabilities had the right to attend all regular education classes. This movement, called the regular education initiative, gradually led to the practice of inclusion. Inclusion involves the education of children with ID alongside typically developing peers for all subjects, usually with the support of a classroom aide.

In 1997, Congress passed 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 required local educational systems to identify all infants, toddlers, and children with disabilities living in the community, whether or not they attended 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, 0 to 3 years, are provided with an Individualized Family Services Plan (IFSP). In contrast, preschoolers and school-aged children receive 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 can also grant accommodations to children with disabilities that help them achieve their cognitive, social, emotional, or behavioral potentials. IDEA was revised in 2004 as the Individuals With Disabilities Education Improvement Act (IDEIA; PL 108–446).

Empirical studies show that 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 ID 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.

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 all children—with and without disabilities (Schalock et al., 2010).

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 universally accessible. These assignments and activities offer alternatives to traditional lecturing, reading, and writing. Universally designed materials can affect (a) the way teachers introduce content to students, (b) the format of instructional material, and (c) the way students demonstrate their learning (Coyne, Pisha, Dalton, Zeph, & Smith, 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.

Second, a teacher might modify the instructional technology she uses to present material. Instructional technology refers to the educational materials instructors use to teach 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, and videos. Similarly, the English teacher might use digital media that allow 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 software that allows them 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 (Coyne et al., 2012).

How Can Clinicians Reduce Challenging Behaviors in Children With Intellectual Disabilities?

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

Applied Behavior Analysis

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 (Vollmer et al., 2014).

A behavior analyst’s 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 actions, such as “throws objects” or “pushes classmates.” Whereas aggression is a vague term that cannot be easily observed or measured, throwing and pushing are easily observed and measured.

Next, the behavior analyst will carefully observe and record the child’s challenging 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 percentage of class time a child engages in stereotyped rocking or swaying. 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.

Observations of children’s behavior can help identify the environmental conditions that elicit it or the consequences that maintain it (Lancioni, Singh, O’Reilly, Sigafoos, & Didden, 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 its causes (Matson et al., 2011). Functional analysis of behavior 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. 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 (see Figure 5.8). 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 to accomplish the second objective (O’Brien, Haynes, & Kaholokula, 2015).

**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 (1) differential reinforcement of incompatible behavior (DRI) and (2) differential reinforcement of zero behavior (DRO). In **differential reinforcement of incompatible behavior (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 a small piece of candy every 30 seconds he does not engage in hand flapping or skin picking.

**Figure 5.8** Clinicians can use functional analysis to identify the cause of a child’s challenging behavior. In this case, 6-year-old Jeoffrey engaged in rocking only when alone. This indicates that his behavior is automatically reinforced. Attention, differential reinforcement, and exercise might be helpful to reduce this behavior. Adapted from Vollmer and colleagues (2014).
**Positive Punishment**

Reinforcement increases behavioral frequency; punishment decreases it. Positive punishment involves the presentation of a stimulus that decreases the frequency of a behavior. 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.

Salvy and colleagues (2004) describe the use of punishment by contingent stimulation to reduce SIB in a toddler with ID. The girl, Johanna, would bang her head against her crib and other hard surfaces approximately 100 times each day. She had 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 phase (hospital implementation), 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 electric 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 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 bed-wetting, 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 bed-wetting, the child might be required to sit on the toilet five times to practice appropriate urination. 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 may temporarily increase. 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.

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 give up a certain number of tokens or points for each problematic behavior.

Behavioral treatment for people with ID 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, time-out) with 83.2% effectiveness, followed by extinction (e.g., planned ignoring) with 82.6% effectiveness, and positive reinforcement with 73.2% effectiveness. Combining behavioral interventions usually resulted in slightly higher effectiveness than the use of any single intervention alone.

**Medication**

Two medications are frequently prescribed to reduce challenging behavior shown by youths with ID: aripiprazole (Abilify) and risperidone (Risperdal). These medications are atypical antipsychotics that were originally intended to treat disorders like schizophrenia and bipolar disorder. However, several double-blind, placebo-controlled studies showed that these medications are also effective in reducing aggression and irritability among youths with low intellectual functioning. In one of the first studies, the Risperidone Disruptive Behavior Study Group (Aman et al., 2002) examined 118 children ages 5 to 12 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 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 (Roth & Worthington, 2015).

Atypical antipsychotics may also be effective in reducing SIBs among youths with ID. Previous studies showed that some youths who displayed SIBs also had hypersensitivity to dopamine. When they engaged in SIBs, they may experience a certain degree of pleasure or reinforcement. Atypical antipsychotics, which block dopamine receptors, likely reduce the reinforcing properties of SIBs, making children less likely to engage in them (Szymanski & Kaplan, 2006).

Antidepressants are often used to treat anxiety and mood disorders in adolescents with ID. Unfortunately, most of the research supporting the efficacy of antidepressants on these disorders was conducted on typically developing youths. Because behavioral interventions are effective in reducing both anxiety and depressive symptoms in older children and adolescents with ID, these behavioral treatments should be used first (Sturmey & Didden, 2014; Sturmey, Lindsay, Vause, & Neill, 2014).

**Review:**

- Clinicians can use functional analysis to identify and alter the antecedents or consequences of challenging behavior. Most challenging behavior is maintained by positive social reinforcement, negative reinforcement, or automatic reinforcement.
- Differential (positive) reinforcement is usually the first-line behavioral treatment to reduce challenging behavior in youths with ID. Negative punishment strategies include extinction, time-out, and response cost. Positive punishment is used only when other interventions are unsuccessful, when the behavior is very problematic, and when parents consent to treatment.
- Atypical antipsychotics like aripiprazole (Abilify) and risperidone (Risperdal) are effective in reducing aggression in some youths with ID.

**How Can Clinicians Help the Caregivers of Children With Intellectual Disability?**

Clinicians should also help support the caregivers of children with ID. After children are initially diagnosed with GDD or ID, parents are at increased risk for mood problems. 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 a significant elevation in parenting stress (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 (Witwer et al., 2014).

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 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 behavioral control, social deficits, and aggression (Tervo, 2012).

Therapists can help children with developmental disabilities by supporting 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 and empathize with parents’ concerns. Therapists 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.

Review:
- Caring for a child with a developmental disability can be stressful. Parents report greater stress when children with ID show challenging behaviors or comorbid disorders.
- Clinicians can support parents by providing evidence-based treatment to their children and encourage problem-focused coping strategies to deal with stress.

KEY TERMS

Abusive head trauma: A form of maltreatment in which a caregiver induces brain injury by forcefully shaking an infant or toddler; can cause learning disabilities, memory problems, ID, irritability, lethargy, vomiting, tremors, and death (sometimes called shaken baby syndrome)

Adaptive functioning: A term used to refer to a person’s ability to cope with common life demands and meet the standards of independence expected of someone in their particular age group and social-cultural background

American Association on Intellectual and Developmental Disabilities (AAIDD): The oldest professional organization devoted to the study and assistance of individuals with ID

Amniocentesis: A moderately invasive procedure to screen for developmental disabilities 15 to 20 weeks’ gestation; involves collecting amniotic fluid from the mother

Angelman syndrome: A genetic disorder caused by missing maternal genetic material on chromosome 15; associated with moderate to severe ID, sporadic/jerky motor movements, lack of spoken language, hyperactivity, and persistent social smile

Anoxia: The absence of oxygen; a potential cause of ID

Applied behavior analysis (ABA): A scientific approach to identifying a child’s problematic behavior, determining its causes, and altering environmental contingencies to change it

Behavioral phenotype: Characteristic features (e.g., appearance, cognitive strengths/weaknesses, comorbid disorders) associated with specific causes for ID

Challenging behavior: Actions shown by some youths with ID that are physically hazardous or that limit their access to educational or social opportunities

Chorionic villus sampling (CVS): A moderately invasive technique to screen for developmental disabilities 8 to 12 weeks’ gestation; involves collecting tissue that connects the placenta to the wall of the uterus

Chromosomal microarray (CMA): A genetic test that identifies copy number variants (i.e., unusual duplications or deletions) in major regions of the genome; used to identify causes of GDD

Cultural–familial ID: A term used by Zigler to describe children with no identifiable cause for their intellectual and
adaptive disabilities; associated with IQ and adaptive functioning scores in the 50–70 range, no health problems, and a family history of ID or low intellectual functioning

**Diagnostic overshadowing:** A term used to describe the tendency of some clinicians to overlook the presence of mental disorders in people with ID

**Differential reinforcement:** A form of positive reinforcement in which therapists reinforce only behaviors that are desired, while they ignore unwanted actions

**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

**Differential reinforcement of zero behavior (DRO):** The therapist reinforces the child for not engaging in the problematic behavior for a certain period of time

**Down syndrome:** Trisomy 21; associated with moderate ID, characteristic appearance, weakness in verbal skills and language, strength in visual–spatial reasoning, and sociability

**Dual diagnosis:** A term used to refer to the presence of mental disorders among individuals with ID

**Education of All Handicapped Children Act:** Federal law that mandated “free and appropriate public education” for all children with disabilities aged 3 to 18

**Extinction:** A form of negative reinforcement: the therapist withdraws reinforcement from the child immediately following an unwanted behavior; sometimes called planned ignoring

**Extinction burst:** A term used to describe a temporary increase in the rate or intensity of children’s behavior immediate after therapists use extinction or “planned ignoring”

**Fetal alcohol spectrum disorder (FASD):** A disorder caused by maternal alcohol consumption during gestation, characterized by lower intellectual functioning or mild ID, learning disabilities, hyperactivity, and characteristic craniofacial anomalies

**Fragile X syndrome:** An inherited, X-linked disorder that adversely affects boys more than girls; characterized by mild to moderate ID, characteristic appearance, strengths in simultaneous processing, weakness in sequential processing, and social communication deficits

**Global developmental delay (GDD):** A DSM-5 disorder, diagnosed in children < 5 years, and characterized by significant delays in several developmental domains (e.g., motor language, social, or daily living skills); a temporary diagnosis used when clinicians suspect ID but the child is too young to determine IQ

**Inclusion:** A term used to describe the education of children with ID alongside typically developing peers for all subjects, usually with the support of a classroom aide

**Individuals With Disabilities Education Act (IDEA):** Federal legislation that extended disability rights to infants and toddlers, mandated IFSP for young children and IEPs for school-age children with disabilities

**Intellectual disability (ID):** A DSM-5 disorder characterized by deficits in intellectual and adaptive functioning deficits in conceptual, social, and practical domains that emerge during infancy or childhood

**Mainstreaming:** Involved placing children with ID in classrooms with typically developing peers, to the maximum extent possible

**Needed supports:** Assistance that helps an individual with ID function effectively in society; an important component of the AAIDD definition of ID

**Organic ID:** A term used by Zigler to describe children who had identifiable causes for their intellectual and adaptive disabilities: associated with genetic disorders, very low IQ and adaptive functioning, medical complications, and no family history of ID

**Overcorrection:** A form of positive punishment in which 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

**Phenylketonuria (PKU):** A recessive disorder characterized by an inability to break down phenylalanine, an amino acid; dieting can prevent severe ID, seizures, and other medical problems

**Physical aggression:** Actions that cause, or can cause, property destruction or injury/harm to another person

**Positive practice:** Usually a form of positive punishment; the therapist makes the child repeatedly practice an acceptable behavior immediately following an unacceptable act; usually paired with overcorrection

**Prader-Willi syndrome (PWS):** A genetic disorder caused by missing paternal genetic material on chromosome 15; associated with mild ID, weakness in short-term memory, strength in visual–spatial reasoning, hyperphagia, and obsessive–compulsive behavior

**Response cost:** A form of negative reinforcement; the therapist withdraws reinforcers from the child immediately following a problematic act

**Repetitive behaviors (SIBs):** Repetitive movements of the hands, limbs, or head in a manner that can, or do, cause physical harm or damage to the person

**Serum screening:** A blood test conducted between 15 and 18 weeks’ gestation to screen for possible developmental disorders in the fetus

**Similar sequence hypothesis:** Posits that children with ID progress through the same cognitive stages as typically developing children, albeit at a slower pace; generally supported by research

**Similar structure hypothesis:** Posits that two children of the same mental age (one with ID and the other without ID) will show similar abilities; has mixed support

**Stereotypies:** Actions performed in a consistent, rigid, and repetitive manner and that have no immediate, practical significance

**Time-out (from positive reinforcement):** A form of negative reinforcement; the therapist limits the child’s access to positive reinforcers (e.g., attention, toys) for a certain period of time, usually by placing the child in a specific setting

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TORCH: An acronym that represents the main maternal illnesses that can cause ID in offspring

Universal design: An educational practice that involves creating instructional materials and activities that allow learning goals to be achievable by children with different abilities and skills

Williams syndrome (WS): A genetic disorder caused by deletions on chromosome 7; associated with mild ID, well-developed spoken language, strengths in auditory memory, weakness in visual–spatial reasoning, hyperactivity, anxiety, and friendly/social demeanor

22q11.2 DS: A genetic disorder caused by deletions on chromosome 22; associated with mild to moderate ID, cleft lip/palate, social communication deficits, and risk for schizophrenia later in life

CRITICAL THINKING EXERCISES

1. When many people think of ID, they think about a child with Down syndrome. To what extent do children with Down syndrome reflect all children with IDs?

2. How does the treatment for PKU illustrate the interaction of genes and environment in child development?

3. Why are children of lower-SES backgrounds at greater risk for certain types of ID? Why might low-SES children with IDs have poorer prognoses than middle-class children with IDs?

4. Why would clinicians probably not use extinction (i.e., planned ignoring) to reduce SIBs in a young child with a developmental disorder?

5. What might be the benefits and drawbacks of academic inclusion on a typically developing child?

TEST YOURSELF AND EXTEND YOUR LEARNING

Videos, flash cards, and links to online resources for this chapter are available to students online. Teachers also have access to PowerPoint slides to guide lectures, case studies to prompt classroom discussions, and exam questions. Visit www.abnormalchildpsychology.org.