40 Intellectual Disability

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HISTORICAL OVERVIEW

While centuries of experience have demonstrated that stigma will not disappear with a name change, the field continues to wrestle with how to capture the condition now referred to as "intellectual disability" or "intellectual developmental disorder." For decades, the field has also worked hard to encourage "people first" language. Individuals should not be defined by their illnesses or conditions—for example, John is not a "leper," or a "schizophrenic," or "retarded." Instead, people have illnesses or conditions—for example, John has schizophrenia. Although these lexical and grammatical sensitivities may seem trivial for some, there is no faster way to lose credibility in some settings than to characterize one's patient as "retarded."

Why such heightened sensitivity in this field? While it is well recognized that intellectual gifts have been distributed on a continuum for as long as mankind has walked the earth, the understanding and treatment of persons with intellectual disability has moved like a pendulum between extremes. At one end, persons with intellectual disability have been exalted, and considered "Les enfants du Bon Dieu" ("children of the Good God"). Certain media may convey the message that persons with intellectual or developmental disability are somehow blessed with simpler, more straightforward understandings of basic human truths. And at the other end of the pendulum swing, such persons have also been vilified. The word "cretin" (congenital hypothyroidism), takes its origin from "Christian or Christ-like," even as its modern dictionary definition includes descriptors like "stupid, vulgar, and insensitive." In the introduction to his text on "mental defectives" a century ago, Barr (1904), the chief physician at the Pennsylvania training school for the feeble-minded, observed that: "The terms idiot and imbecile indiscriminately applied for centuries to a despised and neglected class, have naturally, through the mere force of association, become synonymous with and even expressive of opprobrium and reproach. So closely welded has been this association of word and idea, that even the advance of the nineteenth century with all its revelation of true conditions has failed to sunder them" (p. 17).

Evidence for the recognition and treatment of intellectual disability

dates to the earliest of medical writings. Hippocrates described microcephaly and craniostenosis, and Galen actively explored causes of cognitive disability. In the Middle Ages, Avicenna proposed treatments for meningitis and hydrocephalus, and even defined levels of intellectual function.

Esquirol (1838) is credited as the first medical writer to define the disorder and was careful to distinguish intellectual disability from mental illness or disease. He focused on congenital onset in noting that: “Idiocy is not a disease, but a condition in which the intellectual faculties are never manifested; or have never been developed sufficiently to enable the idiot to acquire such an amount of knowledge as persons of his own age, and placed in similar circumstances with himself, are capable of receiving. Idiocy commences with life, or at that age which precedes the development of the intellectual and effective faculties, which are from the first what they are doomed to be during the whole period of existence.”

The modern history for the field of intellectual disability begins in the late 18th and early 19th centuries. At that time, Jean-Marc Itard attempted a natural experiment to educate Victor, a “wild-child” discovered in the forests of Aveyron, France. Although Itard himself judged his work with Victor a failure, this famous experiment marked the important consideration that persons with disabilities might be educable. Itard’s efforts generated interest in educational and other interventions for persons with disabilities. In some ways, Itard and those that followed provided a bridge from beliefs that individuals with intellectual disability were subhuman (even suggested by Linnaeus to be a separate species in the genus Homo with names like: Juvenis Lupinus Hessensis—a young man found in Hesse among wolves; or Juvenis Hannoverianus—a young man found in Hanover), to a more enlightened appreciation of their humanity and their potential.

Even so, in the preface to his text, Barr (1904) still straddles this divide as he categorically writes off the severely affected, while advocating for the less impaired:

“In endeavoring to emphasize the utter hopelessness of cure, and also the needless waste of energy in attempting to teach an idiot, I have sought to make clear the possibilities that may be attained in the training of the imbecile, the urgent need of preventing the backward child from degenerating into imbecility, and of safeguarding the absolutely irresponsible ammoral imbecile from crime and its penalty.”

Howe and Wilbur are credited with introducing the enlightened approach to educating individuals with intellectual disability to the United States with the establishment of training schools like those that were appearing in Europe.

By the turn of the century, there were 30 such institutional settings in 21 states, both public and private, and some reserved for women, and for

individuals with epilepsy. Many of these institutions had residents numbering in the hundreds and gradually the focus shifted away from education and more toward custodial care.

During their peak usage, from 1950 until the late 1960s, institutions were home to approximately 1 per 1,000 Americans.

Since the mid-1960s, many fewer persons with intellectual disability have been institutionalized in the United States. This move toward "deinstitutionalization" has arisen from many sources. First, the overcrowding and neglect common in many large institutions came to light in various exposés during the late 1960s. Blatt and Kaplan's photo story, "Christmas in Purgatory" (reproduced in Look magazine), Geraldo Rivera's television reports, and Robert F. Kennedy's attacks on the Rome and Willowbrook institutions in New York all shocked the nation.

Deinstitutionalization

Other forces also led to deinstitutionalization. Perhaps the most important of these forces was the philosophy of "normalization," the idea that individuals with intellectual disability were entitled to a more normal lifestyle, including a culturally normal rhythm to one's day (school or work, leisure time, sleep), week (weekdays and weekends), and year (vacations, holidays). In 1972, Wolfensberger extended the idea of normalization to the service delivery system itself, calling on all residences, schools, and other services for persons with intellectual disabilities to be as normative as possible. Parent and professional advocacy groups also fought hard for legislative and legal victories to decrease the size of large institutions.

Over the past 50 years, changes in US residential institutions have been dramatic. Several states have closed their institutions, and remaining facilities have become much smaller. Typically, individuals remaining in large or even medium-sized institutions have the most severe impairments; many of these residents also have severe behavior problems, or motor or sensory disabilities in addition to their severe-profound intellectual disability. Overall, from 1967 to 1994, the total institutional population—defined as state-operated institutions with 16 or more residents—decreased by almost two-thirds, from 194,650 to 65,818. In California, the number of persons served by the Department of Developmental Services increased 61.6 percent between January 1992 and December 2001. During this same period however, the State developmental center population decreased by 44.4 percent. In many states, Medicaid funding waivers (the ability to access Medicaid dollars in circumstances in which family income criteria or other eligibility criteria are waived) are in place explicitly to support children with intellectual disabilities who meet "institutional level of care" standards. These so-called "deeming waivers," or "Katie Beckett waivers" are meant to ensure that Medicaid support can follow an individual out of an institutional setting if such a move can be done safely and cost

effectively. Katie Beckett was a young lady in Iowa who became ventilator-dependent following a catastrophic CNS infection, and whose parents discovered that if they took her home from the skilled nursing facility, she would lose her Medicaid coverage. As the costs associated with institutionalization are invariably in the tens of thousands for any individual (ranging from a low of $104,025 per year in Arkansas to a high of $375,000 in Tennessee in 2009), the cost effectiveness criterion is rarely a limiting factor, but clinicians may be called upon to attest to the level-of-care criterion for their patients who are seeking support under this waiver.

School. Schooling is the other major service for persons with intellectual disability. Following the move to "school" all American children that began in the mid-1800s, teachers and administrators soon discovered that a subset of children were having difficulties in performing school lessons. As a result, classes for "problem children" began in many cities and towns in the late 1800s. In 1894, the first classes specifically for children with intellectual disability were formed in Providence, Rhode Island. In rapid succession, special education classes were begun in Springfield, MA (1894), Chicago (1898), Boston (1899), New York (1901), Philadelphia (1901), and Los Angeles (1902).

Local school districts have historically held an ambivalent attitude toward special education classes. Often housed within the least desirable buildings and rooms, these classes were frequently furnished with scarce equipment and materials, and had few specially trained teachers. By 1968, Lloyd Dunn declared that most children with intellectual disability could be "mainstreamed" in classes with age-mates; he questioned the need for segregated special education classes for most children with intellectual disability. Dunn's article took note of the Supreme Court's ruling that "separate could not be equal" in educational settings, that research had not shown that children with mild intellectual disability learn better in special education versus regular education classes, and that educational techniques had advanced sufficiently to allow for the effective schooling of most children with retardation alongside other children.

In both education and living settings, then, the post-1970 era witnessed the strong influence of mainstreaming, community living, and normalization. On the whole, such movements have proven beneficial as persons with intellectual disability increasingly take their rightful place within modern society. Many professionals do, however, question whether normalization has sometimes gone too far. Not every child with intellectual or other disabilities may be able to be schooled optimally with typically developing age-mates, nor might every adult be able to live independently in the community. Indeed, some families are advocating for specialized schools or residential settings for children with autism even as state agencies fear a return to institutional approaches. These issues continue to

be hotly debated among professionals, families, and organizations associated with persons with intellectual disability.

Research

Like service delivery, the modern history of research also begins in the mid-19th century. In 1838, Esquirol differentiated intellectual disability from mental illness, and proposed several levels of impairment. Later in the century, various different classification systems were proposed. Probably the most well-known was a system of "ethnic classifications" proposed by J. Langdon Down in 1866—hence the term "Mongolism" for persons with Down syndrome, the type of intellectual disability he first identified.

In addition to classifying persons with intellectual disability, genetics plays an important historic role in intellectual disabilities research. Unfortunately, early in this century, that role was mainly negative. Spurred by the rediscovery of Mendel's genetic findings, various workers attempted to show that intellectual disability was inherited. Most notorious were Dugdale's (1877) study of the Jukes, and Goddard's (1913) study of the Kallikaks. In both cases, multi-generational families were used to argue that intellectual disability was inherited and that sterilization was necessary for the "preservation of the race." Such eugenic scares led to court decrees and sterilization laws in California and other states during the 1920s.

Also playing an important role in intellectual disability research was the new science of psychological testing. Henry Goddard, the research director at the Vineland Training School in New Jersey, was the first to use Binet and Simon's IQ tests in the United States. After testing Vineland residents over a several-year period, Goddard concluded that "the vast majority of feeble-minded children are not changing and are not improving in their intelligence levels," a finding that another leading worker, Walter Fernald, called "the most significant … and the most discouraging that we have ever known."

Since early in this century, research has progressed in both the behavioral and biomedical fields. Behaviorally, recent work has allowed for better diagnoses and classification. Nearly a century ago, psychologists invented tests of motor, nonverbal intelligence, achievement, adaptive behavior, and other skills. Following similar work with typically developing children, psychologists have recently learned much about the development of cognitive, linguistic, social, and adaptive skills in persons with intellectual disability. In addition, many studies now examine the presence of psychiatric disorders in children and adults with intellectual disability, and families, schools, group homes, and workshops have all received research attention. Much of this work has occurred from the 1960s until today, a time of strong, often federally supported, progress in intellectual disability behavioral research.

But arguably the most important progress has occurred in the

biomedical and genetic fields. As early as 1934, the Norwegian physician, Folling, hypothesized that several of his patients with retardation were unable to metabolize phenylalanine. Such insights later led to a dietary treatment of this inborn error of metabolism; the phenylalanine-free diet, though not perfect, has significantly limited PKU as a cause of intellectual disability in most industrialized countries. In addition, PKU can now be screened at birth through the Guthrie test, after which dietary treatments can be instituted. PKU remains one of the major success stories in intellectual disability even as new treatments for this condition, for example tetrahydrobiopterin, are still being developed.

Modern genetics constitutes another success story in intellectual disability research. In 1959, Lejeune, Gautier, and Turpin discovered that most cases of Down syndrome were due to trisomy of the 21st chromosome. Fragile X syndrome, the second most common genetic cause of intellectual disability, was discovered in 1969, and its cytogenetic diagnosis was refined in the mid-1970s. With the banding techniques of the 1970s and the molecular genetic techniques of the past two decades, exact causes are being described for many of the nearly 1,500 different genetic disorders associated with intellectual disability. The genetic—and, increasingly, the behavioral—characteristics of many of these disorders are now known (see below).

Organizations

The field of intellectual disability is also a history of organizations. The first such organization was the Association of Medical Officers of American Institutions for Idiotic and Feeble-Minded Persons, begun in 1876. As the first professional group devoted exclusively to persons with intellectual disability, the Association of Medical Officers was the forerunner to the American Association on Intellectual and Developmental Disabilities (AAIDD, formerly AAMR), the field's most influential organization. Through its various journals and committees, AAIDD has been at the forefront of research, policy, and legislative advances for persons with intellectual disability.

In addition to AAIDD, other groups have also played important roles, particularly concerning changes in federal policy toward people with disabilities. The Council for Exceptional Children (CEC) was begun in 1922, and continues to champion the profession of special education and the education of all children with disabilities. The Arc, formerly the National Association for Retarded Citizens (NARC) is the main parent-led advocacy group. Arc was particularly influential in the passage of the Education for All Handicapped Children Act of 1975 (PL 94–142), the federal law that for the first time mandated the "free, appropriate public education" of all children with disabilities throughout the United States. Each of these groups was also influential in the passage of the Americans with

Disabilities Act, federal legislation that took effect in 1992, and outlawed discrimination of persons with disabilities. The National Association for the Dually Diagnosed, now simply NADD, was established in 1983, specifically in recognition of the lack of attention by the professional community to the needs of individuals with intellectual disability who also experience psychiatric illness. Over the ensuing years the organization has grown to be the principal association in North America to advance mental wellness for persons with developmental disabilities through the promotion of excellence in mental health care.

Federal agencies constitute an additional category of influential organizations. Begun during the presidency of John F. Kennedy (himself a sibling to a woman with intellectual disability), the National Institutes of Child Health and Human Development (NICHD) have long supported disability research. Other federal programs, including the nationally distributed network of nearly 70 University Centers for Excellence in Developmental Disabilities Education, Research, and Service (UCEDDs), and NICHD-funded Mental Retardation Research Centers (MRRCs), have also played important roles in research and policy. Taken together, professional, parent, and federal organizations provide an important infrastructure to the intellectual disability field.

Before discussing definitional issues, it is worth revisiting terminology. In different countries throughout the world, different terms are used for intellectual disability. In Europe, the term "intellectual disabilities" has long been used, and a major international journal is the Journal of Intellectual Disabilities Research. In Great Britain, alongside "intellectual disability," the term "learning disability" (or learning disabilities) is often used—a term with a very different meaning in the United States.

DEFINITION OF INTELLECTUAL DISABILITY

Over the years, the American Psychiatric Association (APA) has adopted definitions of intellectual disability from the American Association on Intellectual and Developmental Disabilities (AAIDD), previously the American Association on Mental Retardation (AAMR). While the definitions in the third and fourth editions of the *Diagnostic and Statistical Manual of Mental Disorders* (DSM; American Psychiatric Association) are very similar, DSM-5 (American Psychiatric Association, 2013) includes substantive changes to the definition of intellectual disability. For the first time in DSM, for example, the term "intellectual disability" replaces "mental retardation," and the language of DSM-5 is more closely aligned with that of the AAIDD. (The term proposed for ICD-11 [2015] is "intellectual developmental disorders.") Further, severity levels are now classified by adaptive rather than intellectual function or IQ scores. This section reviews the changes (see Table 40–1 for summary).

Intellectual and Adaptive Functioning

Intellectual Functioning. Standardized intelligence tests that meet appropriate psychometric criteria for reliability and validity are commonly used to identify intellectual disability. These instruments are summarized in Table 40–2, along with each test's age range and cognitive domains.

Both in terms of the testing situation itself and in the choice of an appropriate measure, administering such tests to people with intellectual disability is quite challenging. As people with intellectual disability have increased risks of comorbid psychiatric or behavioral disorders, examiners need to ensure that difficulties such as hyperactivity or poor frustration tolerance do not impede optimal test performance. Even in persons without behavioral challenges, certain personality characteristics may interfere with testing. Many people with intellectual disability, for example, look to others for solutions to difficult problems, and are quick to acquiesce, or become easily discouraged by failure. To minimize these problems, examiners can begin with easier tasks to provide success and incentives or rewards for effort.

Table 40–1.
A Comparison of DSM-IV and DSM-5 Diagnostic Criteria for Intellectual Disability

DSM-IV Diagnostic Criteria		
A. Intellectual Function	**B. Adaptive Function**	**C. Onset**
Significantly subaverage intellectual functioning: an IQ of approximately 70 or below on an individually administered intelligence test	Concurrent deficits or impairment in present adaptive functioning (i.e., the person's effectiveness in meeting the standards expected for his or her age or cultural group) in at least two of the following skill areas: communication, self-care, home living, social/interpersonal skills, use of community resources, self-direction, functional academic skills, work, leisure, health, and safety	The onset is before age 18 years.
DSM-5 Diagnostic Criteria		
A. Intellectual Function	**B. Adaptive Function**	**C. Onset**
Deficits in intellectual function, 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	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, school, work, and community.	Onset of intellectual and adaptive deficits during the developmental period

Data from American Psychiatric Association. *Diagnostic and Statistical Manual of Mental Disorders*. 4th

ed. Washington, DC: American Psychiatric Association; 1994; American Psychiatric Association. *Diagnostic and Statistical Manual of Mental Disorders*, 5th ed. Washington, DC: American Psychiatric Association; 2013.

For individuals who are nonverbal, there are several measures that can be selected that do not rely on expressive language (see Table 40–2). People with certain genetic syndromes (e.g., Williams syndrome, Down syndrome) also present unique test challenges, as many show syndrome-specific profiles of cognitive strength and weakness that are not easily described by a global IQ score (see phenotype section). Persons with intellectual disability and co-occurring sensory or motor deficits also require adaptation to routine test procedures. Finally, when testing individuals from some minority groups, examiners need to be sensitive to language and cultural issues, and use a multi-method test approach, especially when diagnosing persons with mild intellectual disability.

Adaptive Functioning. Edgar Doll (1935) was the first to develop a formal definition and measure of adaptive behavior. Two decades later, the AAMR (now AAIDD) officially included deficits in adaptive behavior in its definition of intellectual disability. Since that time, deficits in adaptive behavior have been formally included in all definitions of intellectual disability. However, until DSM-5, IQ score alone was used to classify individuals in terms of *severity* of intellectual disability. Table 40–3 illustrates the substantive nature of the changes in severity classification from DSM-IV to DSM-5.

Table 40–2.
Measures for Assessing Intellectual Ability

Measure	Age Range (years)	Domains Assessed
Wechsler Preschool and Primary Scale of Intelligence, 4th edition (WPPSI-IV) (Wechsler, 2012)	2.6 to 7–3	Verbal IQ, performance IQ, full scale IQ
Wechsler Intelligence Test for Children, 5th edition (WISC-V) (Wechsler, 2014)	6 to 16–11	Verbal IQ, performance IQ, full scale IQ
Wechsler Adult Intelligence Scale, 4th edition (WAIS-IV) (Wechsler, 2008)	16 to 90	Verbal IQ, performance IQ, full scale IQ
Stanford–Binet Intelligence Scales, 5th edition (Roid, 2003)	2 to 85+	Verbal, quantitative, abstract/visual, short-term memory, composite score
Kaufman Assessment Battery for Children, 2nd edition (K-ABC-II) (Kaufman & Kaufman, 2004)	3 to 18	Sequential and simultaneous processing, mental processing composite
Kaufman Adolescent and Adult Intelligence Test (KAIT) (Kaufman & Kaufman, 1993)	11 to 85+	Crystallized and fluid scales, composite IQ

Differential Ability Scales, 2nd edition (DAS-II) (Elliott, 2006)	2–6 to 17–11	Verbal, nonverbal reasoning, spatial abilities, general conceptual ability
Raven's Coloured Progressive Matrices (CPM) (Raven, 1998, 2003)	5 to elderly	Figural reasoning; group test
Test of Nonverbal Intelligence (TONI)-2 (Brown, Sherbenou, & Johnsen, 1990)	5–85	Abstract problem-solving: simple matching, analogies, classification, intersections, and progressions
Leiter International Performance Scale, Revised (Leiter-R) (Roid & Miller, 1997)	5–0 to 85–11	Reasoning ability, similarities, differences, relationships
Universal Nonverbal Intelligence Test (UNIT) (Bracken & McCallum, 1998)	5–0 to 7–11	Reasoning, memory, symbolic, and nonsymbolic tasks

Table 40–3.
DSM-IV Severity Criteria for Intellectual Disability (APA, 1994)

Severity Criteria

Mild	Moderate	Severe	Profound	Unspecified
IQ level 50–55 to approximately 70	IQ level 35–40 to 50–55	IQ level 20–25 to 35–40	IQ level below 20–25	When there is a strong presumption of mental retardation but the person's intelligence is untestable by standard tests.

DSM-5 Severity Criteria for Intellectual Disability (APA, 2013)

Severity Criteria	Conceptual Domain	Social Domain	Practical Domain
Mild	For preschool children, there may be no obvious conceptual differences. For school-age children and adults, there are difficulties in learning academic skills involving reading, writing, arithmetic, time, or money, with support needed in one or more areas to meet age-related expectations. In adults, abstract thinking, executive function (i.e., planning, strategizing, priority setting, and cognitive flexibility), and short-term memory, as well as functional use of academic skills (e.g., reading, money	Compared with typically developing age-mates, the individual is immature in social interactions. For example, there may be difficulty in accurately perceiving peers' social cues. Communication, conversation, and language are more concrete or immature than expected for age. There may be difficulties regulating emotion and behavior in age-appropriate fashion; these difficulties are noticed by peers in social situations. There is limited understanding of risk in social situations; social judgment is immature for age, and the person is at risk of being	The individual may function age-appropriately in personal care. Individuals need some support with complex daily living tasks in comparison to peers. In adulthood, supports typically involve grocery shopping, transportation, home and childcare organizing, nutritious food preparation, and banking and money management. Recreational skills resemble those of age-mates, although judgment related to well-being and organization around recreation requires support. In adulthood, competitive employment is often seen in jobs that do not emphasize conceptual skills. Individuals generally need support to make health care–related and legal decisions, and to learn to perform a skilled vocation competently. Support is typically needed to raise a family.

	management), are impaired. There is a somewhat concrete approach to problems and solutions compared with age-mates.	manipulated by others (gullibility).	
Moderate	All through development, the individual's conceptual skills lag markedly behind those of peers. For preschoolers, language and pre-academic skills develop slowly. For school-age children, progress in reading, writing, mathematics, and understanding of time and money occurs slowly across the school years and is markedly limited compared with that of peers. For adults, academic skill development is typically at an elementary level, and support is required for all use of academic skills in work and personal life. Ongoing assistance on a daily basis is needed to complete conceptual tasks of day-to-day life, and others may take over these responsibilities fully for the individual.	The individual shows marked differences from peers in social and communicative behavior across development. Spoken language is typically a primary tool for social communication but is much less complex than that of peers. Capacity for relationships is evident in ties to family and friends, and the individual may have successful friendships and sometimes romantic relations in adulthood. However, individuals may not perceive or interpret social cues accurately. Social judgment and decision-making abilities are limited, and caretakers must assist with life decisions. Friendships with typically developing peers are often affected by communication or social limitations. Significant social and communicative support is needed in work settings for success.	The individual can care for personal needs involving eating, dressing, elimination, and hygiene as an adult, although an extended period of teaching and time is needed for the individual to become independent in these areas, and reminders may be needed. Similarly, participation in all household tasks can be achieved by adulthood, although extended teaching and ongoing supports are needed for adult-level performance. Independent employment in jobs that require limited conceptual and communication skills can be achieved, but considerable support from coworkers and others is needed to manage social expectations, job complexities, and responsibilities such as scheduling, transportation, health benefits, and money management. A variety of recreational skills can be developed but require supports and learning opportunities over an extended period. Maladaptive behavior is present in a significant minority and causes social problems.
Severe	Attainment of conceptual skills is limited. The individual generally has little understanding of written language or of concepts involving	Spoken language is quite limited in vocabulary and grammar. Speech may be single words or phrases and may be supplemented through augmentative means. Communication is	The individual requires support for all activities of daily living (meals, dressing, bathing, and elimination), requires supervision at all times, and cannot make responsible decisions regarding well-being of self or others. In adulthood,

	numbers, quantity, time, and money. Caretakers provide extensive supports for problem solving throughout life.	focused on the here and now. Language is used for social communication more than explication. Individuals understand simple speech/gestures. Relationships with familiar others are a source of pleasure and help.	tasks at home, recreation, and work require ongoing support/assistance. Skill acquisition in all domains involves long-term teaching and support. Maladaptive behavior, including self-injury, is present in a significant minority.
Profound	Conceptual skills generally involve the physical world rather than symbolic processes. The individual may use objects in goal-directed fashion for self-care, work, and recreation. Certain visuospatial skills, such as matching and sorting based on physical characteristics, may be acquired. However, motor and sensory impairments may prevent functional use of objects.	The individual has very limited understanding of symbolic communication in speech or gesture. He or she may understand some simple instructions or gestures. The individual expresses his or her own desires and emotions largely through nonverbal, nonsymbolic communication. The individual enjoys relationships with familiar others, and initiates and responds to social interactions through gestural and emotional cues. Sensory and physical impairments may prevent many social activities.	The individual is dependent on others for all aspects of daily physical care, health, and safety, although he/she may participate in some of these activities as well. Individuals without severe physical impairments may assist with daily work tasks at home, like carrying dishes to the table. Simple actions with objects may be the basis of participation in some vocational activities with high levels of support. Recreational activities may involve music, movies, walks, or participating in water activities, all wit h support. Physical and sensory impairments are frequent barriers to home, recreational, and vocational activities. Maladaptive behavior is present in a significant minority.

Table 40–4. Common Measures of Adaptive Behavior

Measure	Age Range	Domains Tested
Vineland Adaptive Behavior Scales, 2nd edition (Sparrow et al., 2005)	Birth to 18–11	Communication: receptive, expressive, written; daily living skills: personal, domestic, community socialization: interpersonal relations, play & leisure, coping skills; motor skills: fine, gross
Adaptive Behavior Assessment System, 2nd edition (ABAS-II); (Harrison and Oakland, 2003)	0–89	Assesses conceptual, social, and practical aspects of adaptive behavior, as well as 10 adaptive skill areas.

Scales of Independent Behavior, Revised (SIB-R) (Bruininks, Woodcock, Weatherman, & Hill, 1996)	Birth to 80+	Motor: fine, gross Social interaction & communication: social interaction, language comprehension, language expression Personal living skills: eating & meal preparation, toileting, dressing, community living skills: time & punctuality, money & value, work
Diagnostic Adaptive Behavior Scale (DABS) (AAIDD release 2015)	4 to 21	Focuses on the critical "cut-off area" for the purpose of ruling in or out a diagnosis of ID or related disability.
Street Survival Skills Questionnaire (SSSQ) (Linkenhoker & McCarron, 1993)	9 to 40+	Administered to person; basic concepts, functional signs, tools, health

Though meanings vary, adaptive behavior is typically viewed as the performance of behaviors required for social and personal sufficiency. Further, adaptive behavior is an inherently developmental and social construct. Adaptive behavior changes as children grow into adolescence and adulthood, and demands for social adaptation are also defined by expectations from others—from one's family, society, and culture. Adaptive skills typically change across various settings; one's adaptive performance on the job or at school may differ from one's performance with friends or at home. Measurements of adaptive behavior, then, need to have a developmental orientation, to be socially and culturally sensitive, and to represent the many settings in which people live, work, and play.

In contrast to Doll's time, many instruments now exist that measure adaptive behavior across multiple domains (e.g., community skills, personal grooming). Table 40–4 summarizes commonly used measures of adaptive behavior, their age ranges, and various domains. Although most of these measures are administered as interviews with parents or other care providers, some measures are administered directly to persons with intellectual disability. Central to all measures of adaptive function is the idea that adaptive behavior is measured by performance as opposed to ability. If people with intellectual disability are able to (possess the skills to) perform certain behaviors, but do not regularly do so or do not do so without prompts, reminders, and assistance, then they necessarily have compromised adaptive functioning.

Severity of Intellectual Disability

There are remarkably variable strengths and competencies of people with intellectual disability. DSM-5 (and similar to AAIDD 2002) provides a classification system based on adaptive functioning across three domains: conceptual, social, and practical. Table 40–3 summarizes the classification

system in DSM-IV (by IQ score) and the DSM-5 system based on characteristics across the three domains and associated with mild, moderate, severe, and profound intellectual disability. As people with intellectual disability can also be classified by the etiology of their conditions, later sections detail how adaptive functioning changes across diverse genetic and other etiological groups.

Mild Intellectual Disability . Mild intellectual disability constitutes the largest group of people with intellectual disability, possibly as many as 85 percent of all persons with ID. These individuals appear similar to unaffected individuals and often blend into the general population in the years before and after formal schooling. Many achieve academic skills at the sixth grade level or higher, and some graduate from high school. As adults, many of these individuals hold jobs, marry, and raise families—yet at times they may appear slow or need extra help negotiating life's problems and tasks.

Robert was a full term infant, the last of three children born to his 38-year-old mother, a high school music teacher, and 40-year-old father, a science teacher. Pregnancy was unremarkable, and Robert's two older sisters were healthy and developing nicely. The family lived in a rural town in the Midwest.

Robert was an extremely fussy newborn, and had extended periods of crying that the pediatrician described as "colic." When Robert was 2 months old, his parents were told that he had supravalvular aortic stenosis, a case that warranted monitoring but no surgeries. Although Robert became less fussy over time, he was a picky eater, refusing solid foods. Robert's parents also noted that he was more "high-strung" than his siblings, often quick to cry or cringe when his sisters played too loudly.

Milestones were slightly delayed, with Robert sitting unassisted at 10 months and walking at 18 months. Language was also delayed, and although his first words appeared at 20 months, Robert had always made his wants and needs known. Although his parents were concerned that he was delayed compared to his sisters, they were reassured by the explanation that boys often had slight delays, and that he was a lively, social boy who would quickly catch up.

When Robert was 3, his parents insisted on a developmental evaluation, which revealed modest delays in cognitive, language, and motor functioning. His developmental quotient was 74 (average range is roughly 85 to 115). He was described as friendly and engaging, a real "charmer," with a cute face that endeared him to many. Robert was enrolled in a special kindergarten, and he remained in a combination of special education and mainstream (general education) classes throughout his academic career. Like the rest of his family, Robert enjoyed listening to music and singing, and he took an active interest in the piano.

At age 7 years, the school psychologist evaluated Robert and felt that he fit a "learning disability" profile. Robert had an overall IQ of 66, but with close to average functioning in short-term memory and expressive language, and pronounced deficits in visual-spatial functioning. He struggled with writing tasks and arithmetic, but loved science and music, and was amazingly conversant to anyone who would listen to him. Indeed, his parents feared he was "too friendly," as well as too active, and with transient, intense interests in unusual items such as vacuum cleaners.

As he entered adolescence, Robert became increasingly anxious, so much so that he occasionally rubbed his hands or rocked, and he "fretted" about day-to-day issues and what would happen next. His long-standing sensitivities to loud sounds seemed to wane slightly, but he developed fears of storm clouds and dogs, and refused to ride on elevators. He became tearful and upset after one of his older sisters left for college, and worried about her health and her ability to watch the weather at college. Although Robert suffered from nightmares, and

would occasionally pace with worry and complain of stomachaches, he attended school, and had a small group of friends in the Special Olympics bowling league. Further, he enjoyed singing with the high school chorus and he was delighted to be frequently selected to play the piano at school concerts.

When Robert was 17 years old, his parents happened to watch a television documentary on Williams syndrome. They were overwhelmed by the similarities between Robert and the people described in the program. They later described the experience like a "jolt." They had always accepted Robert, quirks and all, and had stopped pushing their doctors for reasons "why" when Robert was a preschooler. Nevertheless, they immediately called the informational number offered in the show, and within 2 months, they had the genetic tests done that confirmed their strong suspicion that Robert had Williams syndrome.

While Robert's day-to-day life did not change dramatically after his diagnosis, his parents reported a big difference in Robert's outlook. He met new friends with Williams syndrome at a conference; he applied to go to a summer music camp for young adults with Williams syndrome; and he reported that he felt less alone. Robert's parents reported a mixture of feelings at having such a late diagnosis—disappointment in their doctors, relief in finally knowing, and twinges of guilt. They were energized by having a new community of Williams syndrome families to share their feelings and worries with; like their son, they too felt less alone.

Elisabeth is a 29-year-old widowed Caucasian mother. Elisabeth was the youngest of three children and born to her parents after an unremarkable pregnancy and delivery. Early development was similarly without distinction as she met her developmental milestones as expected. However, in the context of her enrollment in a Head Start program, some cognitive delays were noted. More strikingly, it became clear that Elisabeth had some visual problems evidenced by her falling a lot and stumbling into things. An examination revealed that Elisabeth had significant visual problems prompting patching and other interventions. Elisabeth was routed into special education classes from kindergarten, and although she was described as somewhat of a loner, never really playing with any of her many cousins, behavioral concerns did not really surface until puberty. At that time, she became more withdrawn, into a "fantasy world." Her family recalls that imaginary friends took on new importance such that Elisabeth would carry on animated conversations with them. She would talk to posters on the wall—even kiss them on occasion. Elisabeth began to have increasing difficulties in school around this time as well. Her mother recalls that Elisabeth seemed always to have a level of discomfort with sexuality and, after menarche, Elisabeth would refuse to attend to personal hygiene. Elisabeth was also incapable of eating in the cafeteria with the rest of the students. Her sister recalls that Elisabeth would invariably "start eating like a pig," and because of these behaviors she would be placed in a separate lunchroom by herself.

At the age of 17, a psychological evaluation was done to determine the level of Elisabeth's intellectual ability. The narrative at that time described how Elisabeth had been coded as having mild intellectual disability but that there were other factors that were of concern, notably frequent absences from school and the need to build in extra incentives to get her to attend to tasks. Elisabeth was described in the evaluation at that time as being a cooperative test subject, appearing "healthy in all respects, but she was quiet, somewhat shy and seemed to have difficulty articulating," which was attributed to word-finding difficulties.

Following her graduation from high school Elisabeth found herself in and out of counseling, she became estranged from her family, and at 24 years of age presented to a mental health clinic where she was described as a young woman "with developmental disability who had recently given birth (2 months earlier) to a son." She was described as experiencing symptoms of depression related to the suicide of her husband approximately a year earlier. The story that Elisabeth told at that time was described as chaotic and confusing, including issues relating to her relationship with her parents, sisters and other individuals in her life.

With the birth of Raymond, Elisabeth's first child, the family recalls how they took Elisabeth back into the home to support her and the new grandchild and that they were very concerned about the behaviors they witnessed, including Elisabeth's absolute refusal to change diapers or to wash the baby's genitalia, claiming that such behavior would constitute sexual abuse. They observed her force-feeding the child, and inappropriately dressing him for the

weather. Ultimately, her father overheard a conversation from an adjacent room in which Elisabeth said that she was going to do the same thing that the child's father did (commit suicide) but that she would take Raymond with her. This prompted a call to the authorities and was the proximal event that led to the removal of Raymond from Elisabeth's custody.

Over the next several years, her family describes numerous and volatile interactions with Elisabeth that were consistently characterized by irritability on Elisabeth's part. The family is quite clear in being unable to identify discreet periods of time during which Elisabeth's behavior cycled up or down into specific depressive or manic episodes. The most prominent recollection is of stable and durable preoccupations with fantasy, misinterpretations and even paranoid accusations, recalling recently how she refused to let her brother turn on the lights because she knew that the CIA would be observing the family if the lights were on.

During a recent involuntary hospitalization, the psychiatrist noted that Elisabeth made sporadic eye contact, that her speech was slurred, and that she smiled or giggled inappropriately as though responding to some private thoughts or perhaps "voices." While she denied hallucinations, she was convinced that both her mother and sister intended to kill her if they got the chance, that both had murdered others or had people do the killing for them, and that they were conspiring to make sure that Elisabeth never got her son back. During this admission, Elisabeth was started on risperidone, her first medication trial, at 1 mg per day and subsequently the dose was increased to 3 mg. Elisabeth agreed that the new medicine helped her to think clearly and it was observed that the delusional thinking about her neighbors and the Mental Health Center gradually disappeared during the course of her hospitalization.

Currently, Elisabeth lives on her own in an apartment with close monitoring from community supports. Elisabeth has been cooperative in participating in various programs that have been offered including weekly swimming opportunities as well as participation in an art studio. Staff report that they have not been aware of any changes in Elisabeth's sleep or appetite, no significant changes in her weight, no changes in mood, and no psychotic symptoms.

Historically, people with mild intellectual disability were thought to show relatively few clear-cut organic causes for their delay. However, in recent years there has been an increase in the number of people with genetic syndromes who function in the mild range of intellectual disability. Examples include most people with Prader–Willi syndrome (PWS), as well as some males and most females with Fragile X syndrome.

A more striking finding is that more people with mild intellectual disability come from minority groups and low socio-economic (SES) backgrounds than would be expected from their percentages in the general population. This overrepresentation of minority groups has been used to criticize standardized intelligence tests that are used for placement in special education classroom settings, as well as to highlight the importance of environmental-cultural influences on intellectual development and assessment.

Moderate Intellectual Disability. Moderate intellectual disability is seen in approximately 10 percent of the population with ID, and includes people with more impaired cognitive and adaptive functioning. People with moderate intellectual disability are typically diagnosed in their preschool years, and some show a clear organic cause for their delay. Many persons with Down syndrome, the most common chromosomal cause of ID, often function in this range, as do many adolescents and adults with Fragile X

syndrome. Most children with moderate intellectual disability require special education services, achieving academic skills at the second to third grade level. Need for supportive services continues throughout the lifespan, and with proper supports, many live, work, and thrive in their local communities. In a study by Ross and colleagues, 20 percent of persons with IQs from 40 to 49 lived independently, whereas 60 percent were considered partially dependent and 20 percent totally dependent on others. In a similar way, some individuals in this range are employed in the competitive job market, and need minimal job supervision, whereas others require more extensive supervision on the job, and may work in sheltered workshops or other, more segregated settings.

Bryan is a 17-year-old sophomore with moderate intellectual disability associated with Down syndrome. Bryan had significant cardiac problems in infancy, and eventually required surgery to repair a ventricular septal defect. Although his energy level improved significantly thereafter, he still had some delays in motor and other developmental milestones. Nevertheless, throughout his education, Bryan has been placed in general education classrooms with same-aged peers and with the support of classroom aides. Bryan's IQ is 53, he has some difficulties with articulation, and his language mainly consists of two-to-four-word sentences. Recently, he has made progress in reading and can read 50 sight words. Bryan has had some minor problems with managing his anxiety over the years, and can become somewhat stubborn at times, but he is well-liked by his teachers and classmates, and his parents boast that Bryan has been a charming, pleasant child from infancy. In addition to attending classes at the local high school, Bryan also spends some time learning job skills in the community. He enjoys his position as the assistant manager for his high school's cross-country team and actively participates in Special Olympics.

Severe Intellectual Disability. Severe intellectual disability occurs in about 3 to 4 percent of the population of people with intellectual disability. Individuals at this level often have one or more organic causes for their delay, and many show concurrent motor, ambulatory, and neurological problems, as well as poorly developed communication skills. Most persons with severe intellectual disability require close supervision and specialized care throughout their lives. Some individuals learn to perform simple tasks or routines that facilitate their self-care or their ability to perform in a sheltered workshop or pre-workshop type of setting.

Harrison is a 16-year-old resident at a school for children with severe disabilities. His mother raised concerns early on when at the age of 2.5 Harrison lost acquired language skills. He was seen by a pediatric neurologist, who elected to introduce carbamazepine in the presence of an abnormal EEG. His mother recalls that the carbamazepine not only appeared to improve Harrison's overall function but also seemed to have a positive effect on Harrison's sleep and activity level, both of which were problematic. In fact, the hyperactivity and impulsivity later prompted a trial of methylphenidate and Harrison's mother remembers that while there may have been some initial improvement on this drug, over time Harrison became "an animal" on the medication, resulting in its discontinuation. Indeed, methylphenidate is the only medication that his mother lists when given the opportunity to identify drugs that have been tried in the past that were clearly bad.

On the other side of the equation, his mother lists carbamazepine and other anti-seizure medications as having been consistently helpful over the years. Staff at Harrison's school note

that carbamazepine is unique among the anticonvulsants that have been tried in that there seemed to be benefit with respect to reducing seizure frequency but also significant behavioral benefit. Over the years, the dose of carbamazepine has varied and has been periodically discontinued in favor of trials of newer anticonvulsants because of concerns about Harrison's borderline white blood cell counts. However, because of the unequivocal behavioral benefit that staff and family attribute to the carbamazepine, it is being used again, albeit at doses that may be lower than have been utilized in the past. His mother's recollection is that a number of medication trials have been explored over the years and that each of the medications has conferred only temporary benefit. In addition to carbamazepine and methylphenidate, previous medication trials have included melatonin, clonidine, sertraline, aripiprazole, risperidone, and diphenhydramine.

The behavioral difficulties that have prompted medication changes include agitation, hyperactivity, and mood instability in the form of giddiness, and more recently, inappropriate sexual advances towards female staff. There have also been significant aggressive outbursts in recent years. In addition to the periods of heightened irritability and hyperactivity, there have been periods of diminished activity, and loss of appetite such that Harrison's weight has occasionally dropped into the 120s in spite of his 6′3″ frame. Teachers and Harrison's mother report that during these times his interests also diminish, including his participation in activities that he otherwise enjoys. Observers note that Harrison seems to want to eat but just cannot muster the interest or enthusiasm when food is placed in front of him.

On examination Harrison comes quite willingly into the office setting accompanied by staff and family. He is a tall and slender young man dressed in sweat pants that are loose on his thin frame. Harrison extends his hand to acknowledge the presence of the examiner and does make eye contact on occasion. His psychomotor activity is remarkable for fairly constant stereotyped hand movements and occasionally some vocalizations. Harrison takes a piece of paper from his mother when he sees it, and proceeds to flick it in front of his eyes and to tear small strips from it and place them in his mouth. He readily surrendered these bits of paper upon request. Harrison's affect was full with very clear and bright smiles, especially when interacting with his mother. Later during the visit, he seemed to become more agitated and would occasionally place his hand to his mouth in a self-biting gesture or place his face into the palm of his hand while vocalizing and rocking. Harrison followed simple requests and was able to sign "thank you" after having been given some crackers from the staff who accompanied him.

When Harrison's mother was given a description of bipolar disorder she quite readily recalled all of those symptoms in her older brother, reporting, for example, that he had periods of elevated mood noteworthy for reckless and impulsive behaviors. At other times he would withdraw, become more aloof, and had expressed suicidal thoughts at various times.

Lithium was subsequently initiated, and resulted in a significant improvement in Harrison's irritability, sleep disturbance, and hyperactivity. He remains on a relatively low dose of carbamazepine for control of his epilepsy. His antipsychotic is gradually being tapered toward discontinuation.

Profound Intellectual Disability. Profound intellectual disability affects relatively few individuals (1 to 2 percent) with ID, and involves pervasive deficits in cognitive, motor, and communicative functioning. Impairments in sensory-motor functioning are often seen from early childhood on, and most individuals require extensive training to complete even the most rudimentary aspects of self-care such as eating and toileting. The vast majority of people with profound intellectual disability have identifiable organic causes for their delay, and most require total supervision and care throughout life.

Annalise was born following a pregnancy that was unremarkable—her mother's first. There were significant feeding difficulties in the first few weeks of life, and failure to thrive was considered early on. Annalise was quite hypotonic and her milestones were delayed. She did