TAB 10: Intellectual Disability

Excerpt from American Psychiatric Association, Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (2013) (DSM-5):

Intellectual Disability (Intellectual Developmental Disorder)

Diagnostic Criteria

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

A. Deficits in intellectual functions, such as reasoning, problem solving, planning, abstract thinking, judgment, academic learning, and learning from experience, confirmed by both clinical assessment and individualized, standardized intelligence testing.
B. Deficits in adaptive functioning that result in failure to meet developmental and sociocultural 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.
C. Onset of intellectual and adaptive deficits during the developmental period.

Note: The diagnostic term intellectual disability is the equivalent term for the ICD-11 diagnosis of intellectual developmental disorders. Although the term intellectual disability is used throughout this manual, both terms are used in the title to clarify relationships with other classification systems. Moreover, a federal statute in the United States (Public Law 111-256, Rosa's Law) replaces the term mental retardation with intellectual disability, and research journals use the term intellectual disability. Thus, intellectual disability is the term in common use by medical, educational, and other professions and by the lay public and advocacy groups.

Specify current severity (see Table):

- 317 (F70) Mild
- 318.0 (F71) Moderate
- 318.1 (F72) Severe
- 318.2 (F73) Profound

Diagnostic Features

The essential features of intellectual disability (intellectual developmental disorder) are deficits in general mental abilities (Criterion A) and impairment in everyday adaptive functioning, in comparison to an individual's age-, gender-, and socioculturally matched peers (Criterion B). Onset is during the developmental period (Criterion C). The diagnosis of intellectual disability is based on both clinical assessment and standardized testing of intellectual and adaptive functions.

Criterion A refers to intellectual functions that involve reasoning, problem solving, planning, abstract thinking, judgment, learning from instruction and experience, and practical understanding(Evans 2008; Gottfredson 1997; Harris 2006; King and Kitchner 2002; Margolis 1987; Schalock 2011; World Health Organization 2011). Critical components include verbal comprehension, working memory, perceptual reasoning, quantitative reasoning, abstract thought, and cognitive efficacy. Intellectual functioning is typically measured with individually administered and psychometrically valid, comprehensive, culturally appropriate, psychometrically sound tests of intelligence. Individuals with intellectual disability have scores of approximately two standard deviations or more below the population mean, including a margin for measurement error (generally ± 5 points). On tests with a standard deviation of 15 and a mean of 100, this involves a score of 65–75 (70 ± 5). Clinical training and judgment are required to interpret test results and assess intellectual performance.

Factors that may affect test scores include practice effects and the "Flynn effect" (i.e., overly high scores due to out-of-date test norms). Invalid scores may result from the use of brief intelligence screening tests or group tests; highly discrepant individual subtest scores may make an overall IQ score invalid. Instruments must be normed for the individual's sociocultural background and native language. Co-occurring disorders that affect communication, language, and/or motor or sensory function may affect test scores. Individual cognitive profiles based on neuropsychological testing are more useful for understanding intellectual abilities than a single IQ score. Such testing may identify areas of relative strengths and weaknesses, an assessment important for academic and vocational planning.

IQ test scores are approximations of conceptual functioning but may be insufficient to assess reasoning in real-life situations and mastery of practical tasks(Greenspan and Granfield 1992; Harris 2006; Schalock 2011; Yalon-Chamovitz and Greenspan 2005). For example, a person with an IQ score above 70 may have such severe adaptive behavior problems in social judgment, social understanding, and other areas of adaptive functioning that the person's actual functioning is comparable to that of individuals with a lower IQ score. Thus, clinical judgment is needed in interpreting the results of IQ tests.

Deficits in adaptive functioning (Criterion B) refer to how well a person meets community standards of personal independence and social responsibility, in comparison to others of similar age and sociocultural background(Tassé et al. 2012). Adaptive functioning involves adaptive reasoning in three domains: conceptual, social, and practical. The *conceptual (academic) domain* involves competence in memory, language, reading, writing, math reasoning, acquisition of practical knowledge, problem solving, and judgment in novel situations, among others. The *social domain* involves awareness of others' thoughts, feelings, and experiences; empathy; interpersonal communication skills; friendship abilities; and social judgment, among others. The *practical domain* involves learning and self-management across life settings, including personal care, job responsibilities, money

management, recreation, self-management of behavior, and school and work task organization, among others. Intellectual capacity, education, motivation, socialization, personality features, vocational opportunity, cultural experience, and coexisting general medical conditions or mental disorders influence adaptive functioning.

Adaptive functioning is assessed using both clinical evaluation and individualized, culturally appropriate, psychometrically sound measures. Standardized measures are used with knowledgeable informants (e.g., parent or other family member; teacher; counselor; care provider) and the individual to the extent possible. Additional sources of information include educational, developmental, medical, and mental health evaluations. Scores from standardized measures and interview sources must be interpreted using clinical judgment. When standardized testing is difficult or impossible, because of a variety of factors (e.g., sensory impairment, severe problem behavior), the individual may be diagnosed with unspecified intellectual disability. Adaptive functioning may be difficult to assess in a controlled setting (e.g., prisons, detention centers); if possible, corroborative information reflecting functioning outside those settings should be obtained.

Criterion B is met when at least one domain of adaptive functioning—conceptual, social, or practical—is sufficiently impaired that ongoing support is needed in order for the person to perform adequately in one or more life settings at school, at work, at home, or in the community. To meet diagnostic criteria for intellectual disability, the deficits in adaptive functioning must be directly related to the intellectual impairments described in Criterion A. Criterion C, onset during the developmental period, refers to recognition that intellectual and adaptive deficits are present during childhood or adolescence.

Associated Features Supporting Diagnosis

Intellectual disability is a heterogeneous condition with multiple causes. There may be associated difficulties with social judgment; assessment of risk; self-management of behavior, emotions, or interpersonal relationships; or motivation in school or work environments. Lack of communication skills may predispose to disruptive and aggressive behaviors. Gullibility is often a feature, involving naiveté in social situations and a tendency for being easily led by others(Greenspan et al. 2001;Greenspan et al. 2011). Gullibility and lack of awareness of risk may result in exploitation by others and possible victimization, fraud, unintentional criminal involvement, false confessions, and risk for physical and sexual abuse. These associated features can be important in criminal cases, including Atkins-type hearings involving the death penalty(Greenspan 2009; Tassé 2009).

Individuals with a diagnosis of intellectual disability with co-occurring mental disorders are at risk for suicide. They think about suicide, make suicide attempts, and may die from them(Ludi et al. 2012). Thus, screening for suicidal thoughts is essential in the assessment process. Because of a lack of awareness of risk and danger, accidental injury rates may be increased(Finlayson et al. 2010).

Prevalence

Intellectual disability has an overall general population prevalence of approximately 1%, and prevalence rates vary by age. Prevalence for severe intellectual disability is approximately 6 per 1,000(Einfeld and Emerson 2008; Roeleveld et al. 1997).

Development and Course

Onset of intellectual disability is in the developmental period. The age and characteristic features at onset depend on the etiology and severity of brain dysfunction. Delayed motor, language, and social milestones may be identifiable within the first 2 years of life among those with more severe intellectual disability, while mild levels may not be identifiable until school age when difficulty with academic learning becomes apparent(Reschly 2009). All criteria (including Criterion C) must be fulfilled by history or current presentation. Some children under age 5 years whose presentation will eventually meet criteria for intellectual disability have deficits that meet criteria for global developmental delay.

When intellectual disability is associated with a genetic syndrome, there may be a characteristic physical appearance (as in, e.g., Down syndrome). Some syndromes have a *behavioral phenotype,* which refers to specific behaviors that are characteristic of particular genetic disorder (e.g., Lesch-Nyhan syndrome)(Harris 2010). In acquired forms, the onset may be abrupt following an illness such as meningitis or encephalitis or head trauma occurring during the developmental period. When intellectual disability results from a loss of previously acquired cognitive skills, as in severe traumatic brain injury, the diagnoses of intellectual disability and of a neurocognitive disorder may both be assigned.

Although intellectual disability is generally nonprogressive, in certain genetic disorders (e.g., Rett syndrome) there are periods of worsening, followed by stabilization, and in others (e.g., Sanfilipposyndrome) progressive worsening of intellectual function. After early childhood, the disorder is generally lifelong, although severity levels may change over time. The course may be influenced by underlying medical or genetic conditions and co-occurring conditions (e.g., hearing or visual impairments, epilepsy). Early and ongoing interventions may improve adaptive functioning throughout childhood and adulthood. In some cases, these result in significant improvement of intellectual functioning, such that the diagnosis of intellectual disability is no longer appropriate. Thus, it is common practice when assessing infants and young children to delay diagnosis of intellectual disability until after an appropriate course of intervention is provided. For older children and adults, the extent of support provided may allow for full participation in all activities of daily living and improved adaptive function. Diagnostic assessments must determine whether improved adaptive skills are the result of a stable, generalized new skill acquisition (in which case the diagnosis of intellectual disability may no longer be appropriate) or whether the improvement is contingent on the presence of supports

and ongoing interventions (in which case the diagnosis of intellectual disability may still be appropriate).

Risk and Prognostic Factors

Genetic and physiological

Prenatal etiologies include genetic syndromes (e.g., sequence variations or copy number variants involving one or more genes; chromosomal disorders)(Kaufmann et al. 2008), inborn errors of metabolism, brain malformations, maternal disease (including placental disease)(Michelson et al. 2011), and environmental influences (e.g., alcohol, other drugs, toxins, teratogens). Perinatal causes include a variety of labor and delivery-related events leading to neonatal encephalopathy. Postnatal causes include hypoxic ischemic injury, traumatic brain injury, infections, demyelinating disorders, seizure disorders (e.g., infantile spasms), severe and chronic social deprivation, and toxic metabolic syndromes and intoxications (e.g., lead, mercury)(Harris 2006).

Culture-Related Diagnostic Issues

Intellectual disability occurs in all races and cultures. Cultural sensitivity and knowledge are needed during assessment, and the individual's ethnic, cultural, and linguistic background, available experiences, and adaptive functioning within his or her community and cultural setting must be taken into account.

Gender-Related Diagnostic Issues

Overall, males are more likely than females to be diagnosed with both mild (average male:female ratio 1.6:1) and severe (average male:female ratio 1.2:1) forms of intellectual disability (Einfeld and Emerson 2008). However, gender ratios vary widely in reported studies. Sex-linked genetic factors and male vulnerability to brain insult may account for some of the gender differences(Harris 2006).

Diagnostic Markers

A comprehensive evaluation includes an assessment of intellectual capacity and adaptive functioning; identification of genetic and nongenetic etiologies; evaluation for associated medical conditions (e.g., cerebral palsy, seizure disorder); and evaluation for co-occurring mental, emotional, and behavioral disorders. Components of the evaluation may include basic pre- and perinatal medical history, three-generational family pedigree, physical examination, genetic evaluation (e.g., karyotype or chromosomal microarray analysis and testing for specific genetic syndromes), and metabolic screening and neuroimaging assessment.

Differential Diagnosis

The diagnosis of intellectual disability should be made whenever Criteria A, B, and C are met. A diagnosis of intellectual disability should not be assumed because of a particular genetic or medical condition. A genetic syndrome linked to intellectual disability should be noted as a concurrent diagnosis with the intellectual disability.

Major and mild neurocognitive disorders

Intellectual disability is categorized as a neurodevelopmental disorder and is distinct from the neurocognitive disorders, which are characterized by a loss of cognitive functioning. Major neurocognitive disorder may co-occur with intellectual disability (e.g., an individual with Down syndrome who develops Alzheimer's disease, or an individual with intellectual disability who loses further cognitive capacity following a head injury). In such cases, the diagnoses of intellectual disability and neurocognitive disorder may both be given.

Communication disorders and specific learning disorder

These neurodevelopmental disorders are specific to the communication and learning domains and do not show deficits in intellectual and adaptive behavior. They may co-occur with intellectual disability. Both diagnoses are made if full criteria are met for intellectual disability and a communication disorder or specific learning disorder.

Autism spectrum disorder

Intellectual disability is common among individuals with autism spectrum disorder(Mefford et al. 2012; Moss and Howlin 2009). Assessment of intellectual ability may be complicated by social-communication and behavior deficits inherent to autism spectrum disorder, which may interfere with understanding and complying with test procedures. Appropriate assessment of intellectual functioning in autism spectrum disorder is essential, with reassessment across the developmental period, because IQ scores in autism spectrum disorder may be unstable, particularly in early childhood.

Comorbidity

Co-occurring mental, neurodevelopmental, medical, and physical conditions are frequent in intellectual disability, with rates of some conditions (e.g., mental disorders, cerebral palsy, and epilepsy) three to four times higher than in the general population(Harris 2006). The prognosis and outcome of co-occurring diagnoses may be influenced by the presence of intellectual disability. Assessment procedures may require modifications because of associated disorders, including communication disorders, autism spectrum disorder, and motor, sensory, or other disorders. Knowledgeable informants are essential for identifying symptoms such as irritability, mood dysregulation, aggression, eating problems, and sleep problems, and for assessing adaptive functioning in various community settings.

The most common co-occurring mental and neurodevelopmental disorders are attention-deficit/hyperactivity disorder; depressive and bipolar disorders; anxiety disorders; autism spectrum disorder; stereotypic movement disorder (with or without self-injurious behavior); impulse-control disorders; and major neurocognitive disorder. Major depressive disorder may occur throughout the range of severity of intellectual disability. Self-injurious behavior requires prompt diagnostic attention and may warrant a separate diagnosis of stereotypic movement disorder.

Individuals with intellectual disability, particularly those with more severe intellectual disability, may also exhibit aggression and disruptive behaviors, including harm of others or property destruction.

Relationship to Other Classifications

ICD-11 (in development at the time of this publication) uses the term *intellectual developmental disorders* to indicate that these are disorders that involve impaired brain functioning early in life. These disorders are described in ICD-11 as a metasyndrome occurring in the developmental period analogous to dementia or neurocognitive disorder in later life(Salvador-Carulla and Bertelli 2008; Salvador-Carulla et al. 2011; World Health Organization 2011). There are four subtypes in ICD-11: mild, moderate, severe, and profound.

The American Association on Intellectual and Developmental Disabilities (AAIDD) also uses the term *intellectual disability* with a similar meaning to the term as used in this manual. The AAIDD's classification is multidimensional rather than categorical and is based on the disability construct. Rather than listing specifiers as is done in DSM-5, the AAIDD emphasizes a profile of supports based on severity(Schalock et al. 2010).

Excerpt from Website of the American Association on Intellectual and Developmental Disabilities (AAIDD):

Definition of Intellectual Disability

Intellectual disability is a disability characterized by significant limitations in both intellectual functioning and in adaptive behavior, which covers many everyday social and practical skills. This disability originates before the age of 18.

Intellectual Functioning

Intellectual functioning—also called intelligence—refers to general mental capacity, such as learning, reasoning, problem solving, and so on.

One way to measure intellectual functioning is an IQ test. Generally, an IQ test score of around 70 or as high as 75 indicates a limitation in intellectual functioning.

Adaptive Behavior

Adaptive behavior is the collection of conceptual, social, and practical skills that are learned and performed by people in their everyday lives.

- Conceptual skills—language and literacy; money, time, and number concepts; and self-direction.
- Social skills—interpersonal skills, social responsibility, self-esteem, gullibility, naïveté (i.e., wariness), social problem solving, and the ability to follow rules/obey laws and to avoid being victimized.
- Practical skills—activities of daily living (personal care), occupational skills, healthcare, travel/transportation, schedules/routines, safety, use of money, use of the telephone.

Standardized tests can also determine limitations in adaptive behavior.

Age of Onset

This condition is one of serveral developmental disabilities—that is, there is evidence of the disability during the developmental period, which in the US is operationalized as before the age of 18.

Additional Considerations

But in defining and assessing intellectual disability, the AAIDD stresses that additional factors must be taken into account, such as the community environment typical of the individual's peers and culture. Professionals should also consider linguistic diversity and cultural differences in the way people communicate, move, and behave.

Finally, assessments must also assume that limitations in individuals often coexist with strengths, and that a person's level of life functioning will improve if appropriate personalized supports are provided over a sustained period.

Only on the basis of such many-sided evaluations can professionals determine whether an individual has intellectual disability and tailor individualized support plans.