



Intellectual disability: definitions, evaluation and principles of treatment

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Contributions: (I) Conception and design: All authors; (II) Administrative support: All authors; (III) Provision of study materials or patients: All authors; (IV) Collection and assembly of data: All authors; (V) Data analysis and interpretation: All authors; (VI) Manuscript writing: All authors; (VII) Final approval of manuscript: All authors.

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Abstract: Intellectual disability is clinically recognized in childhood and adolescence by deficits in cognitive and adaptive abilities. The prevalence of intellectual disability is 1% in the general population. While most individuals have mild intellectual disability, 6 per 1,000 individuals have severe intellectual disability. In persons with mild intellectual disability a specific underlying etiology is generally not recognized; whereas, a specific genetic or biologic etiology is more likely to be recognized in persons who have severe intellectual disability. The diagnosis of intellectual disability requires clinical evaluation and judgment as well as a formal testing of the cognitive and adaptive functions. Such formal testing is done by individually administered standardized tests. The treatment of persons with intellectual disability at an individual level depends on the identified underlying cause, if any, and appropriate support to allow optimal functioning and independent living. Persons who have severe intellectual disability require life-long intensive supports. In addition to general medical care, persons who have intellectual disability also need interventions in the educational settings, provision of educational remediation and accommodations, and appropriate level of community based support. The management, quality of life and longevity vary depending up on the severity of intellectual disability and adequacy of community based support systems.

Keywords: Intellectual disability; intellectual developmental disorder; adaptive function; cognitive function

Received: 09 November 2018; Accepted: 05 December 2018; Published: 17 December 2018.

doi: 10.21037/pm.2018.12.02

View this article at: <http://dx.doi.org/10.21037/pm.2018.12.02>

Introduction

Deficits in cognitive and adaptive functioning are the hallmarks of intellectual disability. With advances in scientific research and our understanding of how societal and cultural factors impact a person's cognitive and adaptive functioning, the terminology has evolved from idiocy to mental retardation to intellectual disability or intellectual developmental disorder (1-3). By the definitional criteria, intellectual disability must be identified during developmental years (childhood through adolescence). However, intellectual disability has life-long implications for an individual's growth and development in all functional domains. Persons with

intellectual disability require a variable degree of life-long support in education, ability to live independently, access health care, employment, and community participation and integration. The physician plays an essential role in the diagnostic assessment and medical treatment of intellectual disability. The physician is also directly involved in facilitating and coordinating ongoing and life-long management of various non-medical aspects of management for persons who have intellectual disability.

The setting within which services are delivered to persons with intellectual disability, the cost of caring for persons with intellectual disability and how the services and healthcare are funded vary across countries because

of differences in healthcare systems. In the United States, services for persons with intellectual disability are delivered by a combination of resources that include both public and private agencies and funding mechanisms. The public funding for the evaluation, educational and other support services to persons with intellectual disability is regulated by various Federal and State regulations.

Definition

Intellectual disability (intellectual developmental disorder) as defined by the World Health Organization (WHO), the American Association for Intellectual and Developmental Disabilities (AAIDD), and the Diagnostic and Statistical Manual of Mental Disorders, all include as criteria, a significant impairment in general cognitive functioning, social skills, and adaptive behavior (1-6). Significant impairment is characterized as performance that is 2 or more standard deviations below the mean based on normed, individually administered standardized tests of cognitive and adaptive function. Scores on the standardized tests should not be the sole criteria to determine the severity of intellectual disability. Clinical judgment is integral to the delineation of the severity of impairment in the cognitive and adaptive function (1,2).

According to the International Classification of Disorders, 11th edition (ICD11), disorders of intellectual development are considered as a group of conditions with different causes that begin during developmental period (6). According to the American Association on Intellectual and Developmental Disabilities (AAIDD), intellectual disability “is a disability characterized by significant limitations both in intellectual functioning and in adaptive behavior as expressed in conceptual, social, and practical adaptive skills” (1). The administration and interpretation of standardized, individually administered psychometric testing for cognitive and adaptive functioning should take into account a person’s age and cultural background (2). Other factors, including a person’s sensory, motor, and communication ability may also modulate the administration and interpretation of such testing (1,2,4).

In the United States a widely used definition of intellectual disability is the one from the Individuals with Disabilities Education Act that defines intellectual disability as “significantly sub-average general intellectual functioning, existing concurrently with deficits in adaptive behavior and manifested during the developmental period that adversely affects a child’s educational performance” (7).

The Diagnostic and Statistical Manual of Mental

Disorders-Fifth Edition (DSM-5) classifies Intellectual Disabilities under the category of Neurodevelopmental Disorders and describes three diagnoses, (I) Intellectual Disability (Mild, Moderate, Severe, and Profound), (II) Global Developmental Delay, and (III) Unspecified Intellectual Disability (2). Global Developmental Delay is a diagnosis given to children under the age of 5 who are not able to participate in standardized assessment procedures due to typical developmental limitations for the age or delays in development (2). Unspecified intellectual disability is a diagnosis reserved for children over 5 years of age who could not be assessed due to multiple factors, such as a physical disability or co-occurring mental illness. These two diagnoses require reassessment at a later date (1). The DSM-5 diagnostic criteria include deficits in intellectual functions such as reasoning, problem solving, planning, abstract thinking, judgment, academic learning, and learning from experience (2). Deficits in adaptive function affect communication, social participation, and independent living activities (1-4).

Severity classification

The classification of intellectual disability is based on a person’s intellectual and adaptive functioning, and the intensity of supports needed (*Table 1*) (1-5,8). It is often not possible to assess the severity of intellectual disability in many cases solely based on standardized testing. In these instances, a diagnosis of intellectual disability is made based on clinical findings and judgment (1-3). It is also not always possible to determine the severity of intellectual disability or the severity may evolve over time. In such cases a diagnosis of intellectual disability is made without specifying the level of severity (1-3).

Prevalence

The variability in the reported prevalence of intellectual disability is explained on the basis of the differences in the definitions used in different surveys, how data was collected, and the characteristics of the populations studied. The distribution of the measured intellectual quotient (IQ) in a given population follows the typical bell shaped curve. Based on the typical distribution of the measured IQ in the population and applying 2 standard deviations below the mean as the cut-off, intellectual disability is identified in 2.5% of the general population (1,2,4,5,9-11).

According to DSM 5, the prevalence of intellectual

Table 1 Severity levels of intellectual disability

Severity	Communication and language	Basic skills	Supports needed
Mild	Difficulty in the acquisition and comprehension of complex language concepts and academic skills. Able to do simple multiplications/divisions; write simple letters, lists	Most can do basic self-care, home activities. Able to complete job application; basic independent job skills (arrive on time, stay at task, interact with co-workers); use public transportation; may qualify for recipes	Support as needed basis, episodic or short-term Can achieve relatively independent living and employment as adults with appropriate support
Moderate	Language and capacity for acquisition of academic skills of persons affected vary but are generally limited to basic skills. Abilities include: sight-word reading; copy address from card to job application; match written number to number of items	Some may master basic self-care, and home activities. Abilities include: some independence in self-care; housekeeping with supervision or cue cards; meal preparation, can follow picture recipe cards; job skills learned with much repetition; use public transportation with some supervision	Most require consistent support in order to achieve independent living and employment as adults
Severe	Very limited language and capacity for acquisition of academic skills	May also have motor impairments. Require daily support in and supervision. Some may acquire basic self-care skills with intensive training	Regular, consistent, lifetime support in school, work or home activities. Care dependent
Profound	Very limited communication abilities. Capacity for acquisition of academic skills is restricted to basic concrete skills	May also have motor and sensory impairments. Require daily support and supervision	High intensity support needed, across all environments. Limitations of self-care, continence, communication, and mobility; may need complete custodial or nursing care. Care dependent

disability is 1% of the general population; with 6 per 1,000 persons reported to have severe intellectual disability (2). Most epidemiological surveys generally categorize the severity of intellectual disability as mild ($IQ \geq 50$) or severe ($IQ \leq 50$), with 75% of individuals recognized to have mild intellectual disability (1,2,4,9-11). In the United States, the prevalence of severe intellectual disability has been reported to be between 0.3% and 0.5% of the population, which has remained unchanged for past several decades (4).

The worldwide reported prevalence of intellectual disability is 16.41 per 1,000 persons in low income countries; 15.41 per 1,000 persons in middle income countries; and, 9.21 per 1,000 persons in high income countries (4,10,11).

The male to female ratio for intellectual disability is 2:1 (1,2,4,9-11). In a family with one child affected with severe intellectual disability, the recurrence risk for subsequent child to have intellectual disability is between 3% and 9% (1,4,9-11).

Causes

A specific etiology is likely to be identified in less than

50% of cases with mild intellectual disability; whereas, an underlying biologic etiology is likely to be identified in more than 75% of cases with severe intellectual disability (1,9,11-26). Chromosomal disorders, genetic syndromes, congenital brain malformations, neurodegenerative diseases, congenital infections, inborn errors of metabolism, and birth injury are the most common identified causes of severe intellectual disability (1,9,11-17).

Clinical presentation

The initial symptoms and signs seen in children who have intellectual disability vary depending on the age at presentation, the severity of functional deficits, and the underlying biologic cause especially in cases of severe intellectual disability (1-5,9). In children who have severe intellectual disability, the symptoms and signs are recognized at an early age and may suggest an underlying cause. Children with profound to severe intellectual disability may be recognized clinically during the first 3 years of life (4,5,9). In children who have mild intellectual disability, symptoms and signs are recognized at a later

age and are not suggestive of any specific underlying cause; rather, developmental delay or atypical behavior are common presenting clinical features. Children with mild to moderate intellectual disability may not be recognized until 4–6 years of age and new cases are identified up to 9 years of age (4,5,9,18).

A newborn with intellectual disability may have feeding or breathing difficulty, microcephaly, macrocephaly, dysmorphic facial features, or other congenital anomalies (3-5,9). During infancy, caregivers may notice that infant fails to engage and interact with environmental stimuli. Vision deficits and hearing deficits also become first apparent during infancy (4). A common concern for parents to seek medical attention during infancy is a delay in attaining age expected gross motor skills (3-5,9).

Between 3 and 5 years of age, a delay or difficulty with language acquisition is a more common clinical presentation and a reason for parents to seek medical attention (4,5). There may be deficits in early social play, and fine motor skills, such as, cutting or drawing may be delayed (4,5). As the child enters the early school years, difficulty with school work and concerns about behavior, such as difficulty sustaining attention, become more apparent as presenting symptoms (4,5).

Children and adolescents with intellectual disability may manifest associated behavioral symptoms such as self-injurious behavior, aggression, self-induced vomiting, and difficulty with sleep. Associated or co-morbid mental health conditions are also common in children and adolescents with intellectual disability. These include attention deficit hyperactivity disorder, mood disorders, autism spectrum disorder, anxiety disorders, and obsessive compulsive disorder (1-4,9,12). It is important to recognize behavioral symptoms of co-morbid conditions as part of clinical evaluation of intellectual disability. Children and adolescents who have intellectual disability are 3–4 times more likely to also have associated other mental health conditions when compared to those who do not have intellectual disability (19). In children and adolescents who have symptoms and signs of both intellectual disability and associated other mental health disorder, a dual diagnosis of intellectual disability and co-morbid mental health disorder is appropriate if the diagnostic criteria are met for both.

Mild intellectual disability in adolescents can be difficult to recognize. Although adolescents with mild intellectual disability can engage without limitation in who, what and where discussions, intellectual limitations are more noticeable with why or how discussions (4). Adolescents

are quite cognizant of how others view them and of peer pressure. They do not want to be identified as having intellectual deficits and find different ways to compensate for any deficits (4). This further complicates identification of adolescents with mild intellectual disability (4).

Evaluation

The breadth and depth of evaluation of children and adolescents for intellectual disability will be guided by the age of the child at presentation, severity of symptoms, and the need to pursue an etiological diagnosis. Such an evaluation comprises clinical assessment, psychological testing, genetic and metabolic testing and imaging studies (1,9,18,20-40).

Psychological assessment of intellectual disability involves conducting a clinical interview, administration of standardized intellectual and adaptive assessment measures, and additional assessments to take into account differential diagnoses (1,2,22-32). The psychologist administering the assessments should have training in reliable and valid assessment procedures to help ensure the quality of the results. The psychologist should have sufficient experience working with individuals with intellectual disability to help with accurate identification of severity levels. It is important to consider the impact of culture, gender, stigma, and socioeconomic status on intellectual functioning (1,2,23,26,27).

Clinical history should be obtained from all different relevant sources. For school age children and adolescents, information regarding academic progress over time should be obtained from their school system. Observations, formal evaluations and interventions in the school setting by school psychologist, teachers, or social workers should be obtained.

Assessment of intellectual function

Assessment of intellectual disability using standardized assessment tools is important as it is difficult to infer an individual's intellectual functioning based solely on conversation and observation. For example, confounding variables such as autism spectrum disorder may impact social and behavioral function making identification of intellectual disability difficult (1,2,13,23,29,32). The behaviors associated with autism could cause some people to infer low intellectual functioning, when in fact, individuals with autism might be identified through the use of standardized assessments of intelligence at much higher rate than previously considered (2,29,32).

Table 2 Measures of cognitive abilities

Instrument	Age range
Bayley Scales of Infant Development III	1 to 42 months
Wechsler Preschool and Primary Scale of Intelligence™-Fourth Edition (WPPSI-IV)	2 years, 6 months to 7 years, 7 months
McCarthy Scales of Children's Abilities	2 years 6 months to 8 years 6 months
Stanford-Binet Intelligence Scale (5th edition)	2 to 85 years
Wechsler Intelligence Scale for Children-Fifth Edition (WISC-V)	6 to 12 years
Wechsler Adult Intelligence Scale-Fourth Edition (WAIS-IV)	16 years, 0 months to 90 years, 11 months
Kaufman Brief Intelligence Test, Second Edition (KBIT-2)	4 years, 0 months to 90 years, 0 months
Leiter International Performance Scale-Revised (Leiter-R)	2 to 21 years

Table 3 Measures of adaptive behavior

Adaptive measures	Age range
Vineland Adaptive Behavior Scales, Third Edition (Vineland-3), Interview Form	Birth to 90 years
Vineland Adaptive Behavior Scales, Third Edition (Vineland-3), Parent/Caregiver Form	Birth to 90 years
Vineland Adaptive Behavior Scales, Third Edition (Vineland-3), Teacher Form	3 to 21 years

The most common intellectual assessment measures are the Wechsler scales (*Table 2*) (31). The most frequently used scale for assessing intellectual function in children and adolescents is the Wechsler Intelligence Scale for Children-Fifth Edition (WISC-V), which can be administered to children ages 6 to 16 years, 11 months (31). The Kaufman Brief Intelligence Test-Second Edition (KBIT-2) is a very brief intellectual assessment measure that can be used if full WISC-V evaluation is not possible (25). It is generally agreed that, although not perfect, appropriately measured IQ provides the best estimate of intellectual functioning (1-4,9). Based on the mean value for IQ of 100, the upper limit of 70 as the cut off represents the value that is two standard deviations below the mean. Because there is a five-point standard error of measurement, it is proposed by some that a range of 70–75 should be considered as the upper limit of

IQ as the cut off value for intellectual disability (1-4,9).

Based on the typical bell shaped curve of distribution of IQ scores, raising the IQ score from 70 to 75 as the upper limit of cut off, will double the number of individuals with intellectual disability from 2.27% to 4.85% of the population (2,5,9). An individual with an IQ score of 75 with significant adaptive disability will be considered to have intellectual disability, whereas an individual with no adaptive disability and an IQ score of 65 may not be considered to have intellectual disability (2,5,9).

The diagnosis of Borderline Intellectual Functioning (IQ Score 71–84) may be appropriate in cases where it is difficult to determine if the patient's functioning (IQ scores and adaptive functioning) meets the upper limits of mild intellectual disability (IQ score of 70) or possibly slightly above these limits (2). When the diagnosis of mild intellectual disability is unclear, under such a circumstance the diagnosis of Borderline Intellectual Functioning is likely most appropriate. This indicates that there are significant concerns with intellectual functioning; however, the evidence is insufficient to make a diagnosis of intellectual disability.

Several challenges are recognized in the assessment of intellectual functioning. These include, measurement error, test fairness, the Flynn Effect, comparability of scores from different tests, practice effect, extreme scores, determining a cutoff score, evaluating the role that an IQ score plays in making a diagnosis, assessor credentials, and test selection (1). All these issues present legitimate concerns regarding the validity and reliability of standardized cognitive testing for intellectual disability assessment. It is imperative that the practitioner have adequate training in psychological assessment, scoring, and interpretation.

Assessment of adaptive function

The Vineland Adaptive Behavior scales are commonly used to measure adaptive functioning of individuals with intellectual disability (1,23). Three versions of the Vineland-3 are available, as listed in *Table 3*, offering an interview form, parent/caregiver form, and teacher form. The Vineland 3 assesses adaptive functioning in communication, daily living skills, socialization, motor skills (optional), and maladaptive behavior (optional). Although, in most cases, there is a correlation between the level of cognitive functioning and adaptive functioning; this can vary depending up on multiple factors.

AAIDD's Diagnostic Adaptive Behavior Scale (DABS) is under development. The DABS provides a comprehensive

standardized assessment of adaptive behavior. Designed for use with individuals from 4 to 21 years old, DABS provides precise diagnostic information around the cutoff point where an individual is deemed to have “significant limitations” in adaptive behavior. The presence of such limitations is one of the measures of intellectual disability (25).

One of the main concerns seen in practice is rater bias and the subjectivity of adaptive assessment measures. Most adaptive measures are often caregiver and/or teacher self-reports, context specific, and make objective reporting difficult. For example, some parents and caregivers may overestimate their child’s adaptive functioning in an attempt to present their child most favorably, while others will underestimate their child’s adaptive functioning due to many complex factors, which may include parent fatigue and frustration. Another challenge with adaptive assessment is inconsistency between teacher and parent responses. This too is due to multiple factors; teachers observe children at school that is often a much more structured and rule oriented environment. In addition, teachers who teach primarily in special education classrooms may inadvertently base their ratings of adaptive functioning relative to other children in the specialized classroom as opposed to the normative group. When this occurs, artificially high scores can be given to children with significant adaptive delays.

Genetic and metabolic testing

All genetic and metabolic testing should occur in consultation with a clinical geneticist or biochemical geneticist, with an understanding of providing pretesting and post testing genetic counseling services for families. The yield of genetic testing in identifying a specific genetic condition ranges from 2% to 7% (4,9,17,18,20). Newborn screening programs generally identify major inborn errors of metabolism and the yield of metabolic testing done later in infancy and childhood is reported to be $\leq 1\%$ (4,9,17,18,21,23).

Chromosomal microarray is generally recommended as the first test for children and adolescents in whom the cause of intellectual disability is not known (9,40). Studies estimate a 12% yield for CMA in identifying a specific genetic cause in all patients with ID (1-11,16-21). Advanced genetic testing has improved the yield of identifying underlying genetic abnormalities and in some cases have positively affected medical management decisions (33-38). Tests for inborn errors of metabolism should be considered in cases where a clinical diagnosis is not known (9,33,39).

Apart from identifying a genetic etiology, genetic testing

can also help provide guidance into prognosis, associated morbidity, future reproductive options, and clinical management. However, genetic testing does not substitute for astute clinical acumen in describing a phenotype or expected standard of clinical care. It is important to note that many of the complex genetic testing technologies are still being refined and expensive. Often genetic findings may be totally unrelated to individual patient’s phenotype. Evidence is limited to assess whether doing genetic testing improves health outcomes for children and adolescents with intellectual disability (21,35,37).

Neuroimaging studies

The likelihood of abnormal finding on neuroimaging in persons with intellectual disability is between 33% and 63% (9,17,18,21). Multiple abnormalities have been found on magnetic resonance imaging of the brain in persons with intellectual disability; however, abnormal findings on neuroimaging may or may not help in establishing a cause of intellectual disability. The clinical significance of such findings also has not been clearly elucidated. When indicated, based on clinical history and examination findings, magnetic resonance imaging scan is the study of choice.

Diagnosis

Diagnosis of intellectual disability is primarily based on the presenting symptoms and signs, detailed medical history and findings on a detailed physical examination. The physical examination should particularly focus on any abnormal findings on neurological and dysmorphology examination. According to the widely accepted definitional criteria, a diagnosis of intellectual disability requires formal individualized assessment of cognitive and adaptive functioning by standardized tests (*Tables 2,3*). Multiple variables can affect the administration and interpretation of formal testing for intellectual and adaptive functioning; therefore, such testing procedures should take into consideration the age and developmental stage (mental age) of the child, educational background, environmental risk factors, and social and cultural context. A work up should include complete audiological and vision evaluation in all children and adolescents with intellectual disability (3,5,9).

Multiple factors should be considered when assessing the need for identifying as specific cause for intellectual disability. It is not clear if it is necessary to establish an etiological diagnosis in all cases of intellectual disability. Both, physicians and parents or caregivers are divided in

their opinion about the need for an etiological diagnosis. It is reasonable to pursue etiological diagnosis in children with severe to profound intellectual disability, as an underlying cause is likely to be found in about 75% of cases (4,9). Certain symptoms and physical examination findings may suggest specific conditions for which, appropriate laboratory and imaging studies can assist in confirming a diagnosis.

Those who favor pursuing an etiological diagnosis cite several reasons: complications associated with a specific condition can be anticipated and management can be planned, treatment may be available for a specific condition identified, and long-term life planning can be facilitated (1,3,4,9,21).

In the absence of well-defined clinical symptoms and signs, an extensive work up that includes genetic testing, neuroimaging, and metabolic testing, is needed to search for potential cause of intellectual disability. Such an extensive work up should preferably be undertaken in consultation with specialists with expertise in this field. The yield of these tests in identifying a cause varies depending up on presence or absence of associated symptoms and signs.

Differential diagnosis

Intellectual disability should be differentiated from developmental language disorder and autism spectrum disorder. The characteristic deficits seen in children and adolescents who have intellectual disability are in domains of cognitive function and language development; whereas, their social development is commensurate with their mental age (2,3,9).

Deficits in different aspects of language development is the main characteristic in children and adolescents who have specific or developmental language disorder; their development in social, motor, and cognitive domains is similar to that seen in typically developing children (2,3,5).

Deficits in social development and language or communication development are the main characteristics seen in children who have autism spectrum disorder; their motor development is similar to that seen in typically developing children (2,9).

In children and adolescents who present with symptoms suggestive of intellectual disability, hearing and vision impairments should be ruled out. In mild intellectual disability it is important to carefully consider the contribution of environmental and psychosocial conditions as confounding factors in differentiating true intellectual disability from impact of such environmental factors (1-5,9).

Principles of management

Children and adolescents who have intellectual disability require participation of professionals from different medical, social, and psychological disciplines. The medical care should be ideally delivered in one setting by all different disciplines in an integrated and coordinated manner. The physician should provide a lead role in guiding the interdisciplinary team approach to medical care for children and adolescent with intellectual disability. In addition to medical care specific to concerns associated with intellectual disability and deficits in cognitive and adaptive functioning, preventive and health maintenance should be integral component of healthcare for these children and adolescents.

Different behavioral management modalities are the mainstay for treating behavioral symptoms and associated mental health conditions (3-5,9,32,41). In carefully selected cases, use of psychotropic medications is indicated to treat target symptoms or specific mental health disorder (42,43). The medications used include stimulants, antidepressants, mood stabilizers, and antipsychotics (36). In selected cases, a formal child and adolescent psychiatry specialty consultation is indicated for medication management.

The physician should work collaboratively with community based governmental and non-governmental agencies and programs to access appropriate community based interventional for persons with intellectual disability. The physician should facilitate effective coordination and communication between different agencies and the child's family to access services and monitor ongoing needs and supports. The child's primary physician should also help facilitate consultations between other specialist physicians as indicated.

Access to healthcare for individuals with intellectual disability, allocation of resources and their community based or health system wide deployment vary in different health systems. In the United States, developmental interventions and other related services to children and adolescents with developmental and intellectual disabilities are delivered in the educational or school system. Various programs are designed that serve the needs of specific age groups. These programs are administered based on regulatory framework provided by the Federal and State governments (3-5). Individualized Family Service Plan is for children 3 years and younger, that provides early interventions services through local community agencies. For children from 3 years of age to 16 years of age, the services are delivered through development and implementation

of an Individualized Education Plan (IEP). The IEP primarily focuses on educational interventions that includes remediation and accommodations as indicated to allow for optimal and least restricted educational experience for the child or adolescent with intellectual disability.

Both the clinical and educational services need to transition from the educational setting to community and family setting as the adolescent grows into young adult. Between the ages 14 years and 16 years of age and Individualized Transition Plan is developed and implemented. The Individualized Transition Plan comprises access to adult oriented healthcare services, vocational training, and community based independent living arrangements for the individual with intellectual disability. For ongoing support and intervention services, after leaving the educational setting and the school system, an Individual Habilitation (Support) Plan. The Individualized Habilitation Plan services based on person's severity of intellectual disability (1,41,44-48).

Conclusions

Intellectual disability is defined as significant limitations in cognitive and adaptive functioning. The severity is classified as mild, moderate, severe, and profound, based on various measures of functioning and clinical judgment. Mild intellectual disability is primarily due to environmental risk factors. Of individuals with intellectual disability, three fourths are categorized as having mild intellectual disability. On the other hand, severe intellectual disability is more likely to be due to an underlying biologic etiology. Symptoms and signs of severe intellectual disability are seen at an earlier age, generally before 3 years of age; whereas, those of mild intellectual disability are seen at a later age. The diagnosis of intellectual disability is based on clinical features and formal standardized testing for intellectual and adaptive functioning. There is not a general agreement among healthcare professionals and lay public alike on the need for determining specific underlying cause of intellectual disability. The main principles of management of individuals who have intellectual disability include general medical care, treatment of associated conditions, educational interventions, treatment of behavioral symptoms, and community based supports.

Acknowledgements

None.

Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

References

1. Schalock RL, Borthwick-Duffy SA, Bradely VJ, et al. editors. *Intellectual Disability: Definition, Classification, and Systems of Supports*, 11th ed. Washington, DC: AAIDD, 2009.
2. American Psychiatric Association. *Diagnostic and Statistical Manual of Mental Disorders*, 5th ed. Washington, DC: American Psychological Association, 2013:33-40.
3. Patel DR, Greydanus DE, Merrick J, et al. Introduction to intellectual and developmental disabilities. In: Rubin IL, Merrick J, Greydanus DE, et al. editors. *Rubin and Crocker 3rd Edition: Health care for people with intellectual and developmental disabilities across the lifespan*. Dordrecht: Springer, 2016:5-14.
4. Shapiro BK, Batshaw ML. Intellectual disability. In: Kliegman RM, Stanton BF, Schor NF, et al. editors. *Nelson Textbook of Pediatrics*, Philadelphia: Elsevier, 2016:216-22.
5. Shapiro BK, Batshaw ML. Developmental delay and intellectual disability. In: Batshaw ML, Pellegrino L, Roizen NJ. editors. *Children with Disabilities*, 7th edition, Baltimore: Paul Brookes, 2013:291-306.
6. World Health Organization International Classification of Diseases 11. Available online: <http://www.who.int/classifications/icd/revision/en/>, accessed 25 May 2018.
7. Individual with Disabilities Education Act. Available online: <https://sites.ed.gov/idea/>, accessed 25 May 2018.
8. Harris JC. *Intellectual disability: understanding its development, causes, classification, evaluation, and treatment*. New York: Oxford University Press, 2006.
9. Moeschler JB, Shevell M. Committee on Genetics. Comprehensive evaluation of the child with intellectual disability or global developmental delays. *Pediatrics* 2014;134:e903-e918.
10. Maulik PK, Mascarenhas MN, Mathers CD, et al. Prevalence of intellectual disability: a meta-analysis of population-based studies. *Res Dev Disabil* 2011;32:419-36.
11. McKenzie K, Milton M, Smith G, et al. Systematic review of the prevalence and incidence of intellectual disabilities: current trends and issues. *Curr Dev Disord*

- Rep 2016;3:104-15.
12. Srivastava AK, Schwartz CE. Intellectual disability and autism spectrum disorders: causal genes and molecular mechanisms. *Neurosci Biobehav Rev* 2014;46:161-74.
 13. Erbetta A, Bulgheroni S, Contarino VE, et al. Low-functioning autism and nonsyndromic intellectual disability. *J Child Neurol* 2015;30:1658-63.
 14. Bilder DA, Pinborough-Zimmerman J, Bakian AV, et al. Prenatal and perinatal factors associated with intellectual disability. *Am J Intellect Dev Disabil* 2013;118:156-76.
 15. Huang J, Zhu T, Qu Y, et al. Prenatal, perinatal and neonatal risk factors for intellectual disability: a systematic review and meta-analysis. *PLoS One* 2016;11:e0153655.
 16. Ellison JW, Rosenfeld JA, Shaffer LG. Genetic basis of intellectual disability. *Annu Rev Med* 2013;64:441-50.
 17. Moeschler JB, Shevell M; American Academy of Pediatrics Committee on Genetics. Clinical genetic evaluation of the child with mental retardation or developmental delays. *Pediatrics* 2006;117:2304-16.
 18. Shevell M, Ashwal S, Donley D, et al. Practice parameter: evaluation of the child with global developmental delay: report of the Quality Standards Subcommittee of the American Academy of Neurology and The Practice Committee of the Child Neurology Society. *Neurology* 2003;60:367-80.
 19. Iwase S, Bérubé NG, Zhou Z, et al. Epigenetic etiology of intellectual disability. *J Neurosci* 2017;37:10773-82.
 20. Curry CJ, Stevenson RE, Anghon D, et al. Evaluation of mental retardation: recommendations of a consensus conference. *Am J Med Genet* 1997;72:468-77.
 21. van Karnebeek CD, Jansweijer MC, Leenders AG, et al. Diagnostic investigation in individuals with mental retardation: A systematic literature review of their usefulness. *Eur J Hum Genet* 2005;13:6-25.
 22. Patel DR, Taylor CM. Developmental and functional evaluation and assessment. In: Rubin IL, Merrick J, Greydanus DE, et al. editors. *Rubin and Crocker 3rd Edition: Health care for people with intellectual and developmental disabilities across the lifespan*. Dordrecht: Springer, 2016:595-618.
 23. Tassé MJ, Schalock RL, Balboni G, et al. The construct of adaptive behavior: its conceptualization, measurement, and use in the field of intellectual disability. *Am J Intellect Dev Disabil* 2012;117:291-303.
 24. Available online: <https://www.pearsonclinical.com/psychology/products/100001622/vineland-adaptive-behavior-scales-third-edition--vineland-3.html>, accessed 25 May 2018.
 25. Available online: <https://aaidd.org/dabs>, accessed December 12, 2018.
 26. Thomas-Presswood TN, Sasso J, Gin G. Cultural issues in the intellectual assessment of children from diverse cultural backgrounds. *Journal of Social Distress and the Homeless* 1997;6:113.
 27. Werner S, Corrigan P, Ditchman N, et al. Stigma and intellectual disability: a review of related measures and future directions. *Res Dev Disabil* 2012;33:748-65.
 28. McConaughy SH. *Clinical interviews for children and adolescents: assessment to intervention*. Second edition. New York: Guilford, 2013.
 29. Lewis B. *Encyclopedia of autism spectrum disorders*. Springer, 2017:1-6.
 30. Pearson Clinical Assessments. Available online: <https://www.pearsonclinical.com/psychology/products/100000771/wechsler-intelligence-scale-for-childrensupupffifth-edition--wise-v.html>, accessed May 25 2018.
 31. Flanagan DP, Alfonso VC. *Essentials of WISC-V*. Hoboken NJ: John Wiley and Sons, 2017
 32. Lloyd BP, Kennedy CH. Assessment and treatment of challenging behaviors for individuals with intellectual disability: a research review. *JARID* 2014;27:187-99.
 33. Michelson DJ, Shevell MI, Sherr EH, et al. Evidence report: genetic and metabolic testing on children with global developmental delay: report of the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of Child Neurology. *Neurology* 2011;77:1629-35.
 34. Stevenson RE, Schwartz CE. X-linked intellectual disability: unique vulnerability of the male genome. *Dev Disabil Res Rev* 2009;15:361-8.
 35. Sun F, Oristaglio J, Levy SE, et al. Genetic Testing for Developmental Disabilities, Intellectual Disability, and Autism Spectrum Disorder. Technical Brief No. 23. (Prepared by the ECRI Institute–Penn Medicine Evidence-based Practice Center under Contract No. 290-2012-00011-I.) AHRQ Publication No.15-EHC024-EF. Rockville, MD: Agency for Healthcare Research and Quality; June 2015. Available online: www.effectivehealthcare.ahrq.gov/reports/final.cfm
 36. Gilissen C, Hehir-Kwa JY, Thung DT, et al. Genome sequencing identifies major causes of severe intellectual disability. *Nature* 2014;511:344-7.
 37. Vissers LECM. Genetics studies in intellectual disability and related disorders. *Nat Rev Genet* 2016;17:9-18.
 38. Tzschach A, Grasshoff U, Bauer P. Next-generation sequencing in X-linked intellectual disability. *Eur J Hum*

- Genet 2015;23:1513-8.
39. van Karnebeek CDM, Shevell MI, Zschocke J, et al. The metabolic evaluation of the child with an intellectual developmental disorder: diagnostic algorithm for identification of treatable causes and new digital resource. *Mol Genet Metab* 2014;111:428-38.
 40. Coutton C, Dieterich K, Satre V, et al. Array-CGH in children with mild intellectual disability: a population-based study. *Eur J Pediatr* 2015;174:75-83.
 41. Reichow B, Servili C, Yasamy MT, et al. Non-specialist psychosocial interventions for children and adolescents with intellectual disability or lower-functioning autism spectrum disorders: a systematic review. *PLOS Med* 2013;10:e1001572.
 42. Calles JL Jr. Use of psychotropic medications in children and adolescents with cognitive adaptive disabilities. *Pediatr Clin North Am* 2008;55:1227-40.
 43. McQuire C, Hassiotis A, Harrison B, et al. Pharmacological interventions for challenging behaviors in children with intellectual disabilities: a systematic review and meta-analysis. *BMC Psychiatry* 2015;15:303-13.
 44. Kandel I, Schofield P, Merrick J. Aging and disability. Research and clinical perspectives. Victoria, BC: Int Acad Press, 2007.
 45. Voss H, Vogel A, Wagemans AMA, et al. Advance care planning in palliative care for people with intellectual disabilities: a systematic review. *J Pain Symptom Manage* 2017;54:938-960.e1.
 46. Shogren KA, Shaw LA, Wehmeyer ML, et al. The support needs of children with intellectual disability and autism: implications for supports planning and subgroup classification. *J Autism Dev Disord* 2017;47:865-77.
 47. Santillil S, Nota L, Ginevra C, et al. Career adaptability, hope and life satisfaction in workers with intellectual disability. *J Vocat Behav* 2014;85:67.
 48. Coppus AM. People with intellectual disability: what do we know about adulthood and life expectancy. *Dev Disabil Res Rev* 2013;18:6-16.

doi: 10.21037/pm.2018.12.02

Cite this article as: Patel DR, Apple R, Kanungo S, Akkal A. Intellectual disability: definitions, evaluation and principles of treatment. *Pediatr Med* 2018;1:11.