



INTELLECTUAL DISABILITY

A REVIEW FOR DENTAL PROFESSIONALS

Purpose of this Module

The information presented in this module is intended to familiarize dental personnel with the definition, etiology, descriptive terminology, communication deficits, prevalence, and specific learning characteristics of individuals with intellectual disability.

Learning Objectives

After reviewing the written materials, the participant will be able to:

1. Define three factors necessary for a correct diagnosis of intellectual disability.
2. Review and explain the changing terminology for the term intellectual disability.
3. Describe the significance of Rosa's law for those with intellectual disabilities.
4. Compare the adaptive functioning abilities of persons with different levels of intellectual disability.
5. State the prevalence of intellectual disability.
6. Describe major categories of etiologies of intellectual disability and provide several examples from each group.
7. Describe learning characteristics of individuals with intellectual disability.

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INTRODUCTION

The intent of this review is to distill from the vast amount of information on intellectual disabilities those facts and concepts that would be considered important for dental providers treating this population. As with any broad review, generalizations are common. It must be remembered that although the following discussion centers on the differences between individuals with intellectual disabilities and those without intellectual disabilities, there are far more similarities than differences between the two groups.

Definition of Intellectual Disability

Intellectual disability (ID), also known as intellectual developmental disorder (IDD), refers to deficits in intellectual and adaptive functioning that begin during the developmental period. It is considered one of the neurodevelopmental disorders, a group that also includes autism spectrum disorder and attention deficit/hyperactivity disorder (ADHD). The definition from the American Psychiatric Association's fifth edition of the Diagnostic and Statistical Manual of Mental Disorders (DSM-5) consists of three components, all of which must be present in order to correctly diagnose ID:

- (a) Deficits in intellectual functions: This includes deficits in "reasoning, problem-solving, planning, abstract thinking, judgment, academic learning, and learning from experience." The evaluation is based on a combination of clinical assessment and standardized intelligence testing.
- (b) Deficits in adaptive functioning: This includes deficits in conceptual functioning (e.g., reasoning and memory), social functioning (e.g., interpersonal communication and self-regulation), and practical functioning (e.g., personal care

and managing one's behavior). The severity of ID is based on these criteria.

- (c) Onset during the developmental period

Severity of Intellectual Disability

The severity of intellectual disability is based on deficits in adaptive functioning in the conceptual, social and practical domains, and is classified as mild, moderate, severe, or profound. The following examples are based on the DSM-5 guidelines. In the conceptual domain, children with mild ID may have difficulty with skills involving writing, math, money, or time. Adults with mild ID may have difficulty in planning, priority setting, and short term memory. In the social domain, those with mild ID may be delayed in speech and language while those in the severe domain may only use a few words in addition to non-verbal communication. In the practical domain, someone with severe ID might require support for all activities of daily living whereas someone with profound ID may be completely dependent in these areas.

Intelligence Testing

The DSM-5 notes that IQ tests are "approximations of conceptual functioning." The DSM-IV-TR included standardized intelligence testing (IQ tests) in the diagnostic criteria, requiring an IQ score two standard deviations below the mean (by definition a score of 70 with an error of +/-5). While IQ tests are used in the DSM-5, they are used in combination with clinical assessment to evaluate the diagnostic criteria of deficits in intellectual functions. The changes also de-emphasize IQ tests by measuring the severity of ID with assessments of adaptive functioning as opposed to intelligence testing. Additionally, the DSM-5 specifies that the interpretation of IQ tests should take into account other disabilities as well as cultural and linguistic differences.

CHANGING TERMINOLOGY

As older terminology referring to people with intellectual disabilities resulted in stigmatizing language and attitudes, alternative terminology was proposed. Historically, the concept of intellectual disabilities has undergone multiple changes in the United States. In the early 1900s ID was referred to as feeble-mindedness. Those deemed feeble-minded were categorized into severity levels of moron (mental age seven to twelve years), imbecile (mental age two to seven years) and idiot (mental age less than two years).

More recently, the term mental retardation was used widely throughout the United States. Categories of mental retardation included mild (IQ of 55-70), moderate (IQ of 40-55), severe (IQ of 25-40), and profound (IQ of 0-25). In 2010, Rosa's Law eliminated the term mental retardation from United States federal documents, substituting the term with intellectual disability. The American Association of Intellectual and Developmental Disabilities (AAIDD) also defines and promotes the usage of the term intellectual disability as opposed to mental retardation.

The DSM-5, released in May of 2013 from the American Psychiatric Association, changed the term "mental retardation" to "intellectual disability (intellectual developmental disorder)." There is some controversy regarding the use of the term intellectual developmental disorder (IDD). During development of the DSM-5, the APA proposed the term "intellectual developmental disorder" in conflict with the AAIDD's use of "intellectual disability" and Rosa's Law which also supports the use of the term "intellectual disability." The APA noted that they considered the term intellectual developmental disorder as opposed to intellectual disability to be consistent with the proposals for the 2015 International Classification of Diseases (ICD-11) utilized by the World Health Organization. The controversy stemmed from, in part, the

classification of ID/IDD as either a health condition and disorder or as a disability, and the conceivable effect on services and stigma for those with the diagnosis. The term IDD is classified as a health condition and disorder whereas ID is classified as a disability. The DSM-5 ultimately chose "intellectual disability (intellectual developmental disorder)" to represent both of these terms. The APA noted that the ICD-11 has yet to be published and that they are using "intellectual developmental disorder" as a bridging term for future DSM publications. As the DSM is a commonly used diagnostic tool for this condition in the United States, future DSM revisions are likely to affect which terms are encountered in a clinical setting.

With the support of the AAIDD and Rosa's law, the use of the term intellectual disability may be more appropriate than the term intellectual developmental disorder in most contexts. In these modules, the term intellectual disability (ID) is primarily used.

Another important change in terminology is the use of person first language. Person first language puts the person term before the disability term. With the new terminology and utilizing person-first language, the phrases "person with an intellectual disability" is preferred over "intellectually disabled person."

LOCATION WITHIN COMMUNITY

Until the mid-1950s many persons with intellectual disabilities at all levels of disability were located in institutions. With widespread deinstitutionalization in the 1970s and mandated school attendance legislation (PL 94-142) enacted in 1978, school age children at all levels of adaptive functioning are provided services within the local school system through extensive special education programs. Day training and service centers established at the community level in most states in the 1970s now provide education and training services for adult clients (age 22 or older) of all levels of intellectual disability.

Through efforts of normalization, only one or two percent of all persons with intellectual disability now reside in institutions. These are primarily individuals with the most severe intellectual disabilities, the greatest deficits in adaptive functioning, and the most severe medical problems.

PREVALENCE OF INTELLECTUAL DISABILITY

Epidemiologists have measured the prevalence of intellectual disabilities in the United States to be approximately one percent.

ETIOLOGY

ID has many different etiologies and may be seen as a common pathway of numerous pathological processes that affect the functioning of the central nervous system. These etiologic factors may be primarily biological, primarily psychosocial or some combination of both. In approximately thirty to forty percent of cases the etiology is unidentifiable even after extensive diagnostic evaluation. In individuals with more severe ID, etiologies are more likely to be identifiable. A review of the prevalence of known causes of ID found wide variation in the literature, with possible attribution to psychosocial or other environmental influences.

Predisposing factors can be separated into a few major groups:

Hereditary

Researchers have identified over 280 genes associated with ID. As additional genetic causes of ID are identified, the proportion of known genetic causes of ID increases. Past studies found a hereditary cause in approximately 8% of cases while more recent studies have found a genetic etiology in up to 25-55% of cases. In general, genetic factors contributing to ID are either chromosomal aberrations (both sex and autosomal) and specific defective genes. The two most

common genetic causes of ID are Down syndrome and Fragile X syndrome.

The major aberration in somatic chromosomes produces Down syndrome (Trisomy 21). Although there are several trisomies associated with ID, Down syndrome is most common. This syndrome affects both males and females, is characterized by the presence of an extra chromosome and is the most common genetic cause of ID. Down syndrome accounts for 4.7-16% of cases of ID. The age of the mother is considered the greatest determinant, i.e., the older the mother, the greater the birth prevalence. A detailed review of Down syndrome and associated dental implications is presented in Module 3.

Aberrations in sex chromosomes can produce Turner syndrome (missing one X chromosome) and Klinefelter syndrome (presence of extra X chromosome). Turner syndrome occurs only in females, with ID occurring in twenty percent of the cases. Klinefelter syndrome occurs in males only, with ID occurring more frequently.

The second most common genetic cause of ID is Fragile X syndrome, which occurs primarily in males. This is caused by a single gene, FMR1, and presents with characteristics of autism in addition to intellectual disability. Other specific defective genes include:

- Congenital ectodermoses such as tuberous sclerosis.
- Metabolic storage disorders such as galactosemia and phenylketonuria (PKU), the former a carbohydrate disorder and the latter a protein disorder.
- Disorders of endocrine function.
- Cranial anomalies such as microcephaly.

Physical

The major physical factors precipitating ID include the following:

- **Prenatal:** Maternal malnutrition, acute maternal infections (e.g., Rubella), chronic maternal infections (e.g., syphilis),

maternal sensitization (e.g., Rh factor), pregnancy complications due to maternal medical conditions (e.g., hypertension, diabetes), anoxia, radiation and drugs (including maternal alcohol consumption). Fetal alcohol syndrome is seen as an etiologic factor in approximately 2% of the population with ID.

- **Neonatal:** Prematurity, hypoxia or birth injury.
- **Postnatal:** Traumatic head injury, brain tumors, infections (e.g., meningitis) or toxins (e.g., lead).

Despite the multitude of specific causes, relatively few specific clinical disorders resulting in damage to the developing brain can be delineated. The most well-known of these is cerebral palsy. Although only approximately fifty to seventy percent of persons with cerebral palsy have an intellectual disability, this condition comprises a large proportion of those with severe to profound ID. A detailed review of cerebral palsy is presented in Module 4. Some forms of neonatal hypothyroidism, microcephaly, and hydrocephaly can also result in damage to the developing brain.

Mental

Mental risk factors for ID include autistic disorder and other pervasive developmental disorders.

Environmental and Psychosocial

Environmental influences include deprivation of nurturance, lack of stimulation, and poverty.

CHARACTERISTICS OF INDIVIDUALS WITH INTELLECTUAL DISABILITIES

Physical

There are no specific physical characteristics that distinguish the population with intellectual disabilities from those without intellectual disabilities. However, some genetic syndromes have particular features

associated with them. For example, individuals with Down syndrome have specific syndrome-associated facial features. While most individuals diagnosed with intellectual disabilities do not necessarily have distinguishing physical characteristics, many do have visible physical differences, including unsteady gait, scoliosis, orofacial abnormalities, and hypotonicity.

Behavioral

There are no specific behavioral characteristics common to all individuals with intellectual disabilities. Sometimes there are behaviors that are associated with the emotional and psychological problems that sometimes accompany intellectual disabilities. Perseveration (continued repetition of words, phrases or certain physical movements) may be characteristic of some individuals with intellectual disabilities, especially those who also have a diagnosis of autism or traumatic brain injury.

Intellectual

It is the area of intellectual or learning difficulties that engenders the term intellectual disabilities. That is, these deficits occur in the areas of acquiring knowledge, storing knowledge, and using this stored knowledge in various situations. Only a brief mention of the major learning characteristics that are important for dental professionals is presented here. One must always remember that this discussion, as well as most information concerning persons with intellectual disabilities, consists of broad generalizations and that obviously individual differences abound.

- People with ID may learn visual and auditory discriminations more slowly than others.
- People with ID often exhibit short-term memory deficit; however, long-term memory may be unaffected.
- Abstract terminology or verbal cues can produce more difficulty in problem solving than the use of concrete

terminology and visual cues in many individuals with ID.

- Separation of a stimulus from a distracting background is often difficult for a person with ID, especially for those who also have autism or ADHD.
- The population with ID often thinks more concretely and may have difficulty generalizing from one stimulus to another. Because persons with ID often have difficulty in predicting outcome, changes in routine can create stress and may trigger stress-related behaviors.
- In addition, the experience and expectation of failure compared to success can greatly influence the intellectual development of those with ID.

Further elaborations of these characteristics and suggestions for coping with these differences in a dental environment will be explored in Module 2.

REFERENCES

Editors' Note:

Much of the information outlined in this section can be found in the excellent text *The Mentally Retarded Child* by Robinson and Robinson. Additional references are used when appropriate.

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