Chapter 5: Intellectual Disability and Developmental Disorders

# 5.1 Description & Epidemiology

## What is Intellectual Disability (ID)?

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

## How does ID Differ Based on Severity?

* DSM-5 allows clinicians to classify individuals with ID based on their adaptive functioning: mild, moderate, severe, or profound.
* In contrast, the AAIDD classifies individuals with ID based on their needed supports, that is, assistance that helps these individuals function effectively in society.
* Levels of needed supports include intermittent, limited, extensive, or pervasive.

## What is Global Developmental Delay (GDD)?

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

## What Challenging Behaviors are Associated with ID and GDD?

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

## How Common is ID?

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

# 5.2 Causes

## What is the Difference between Organic and Cultural-Familial ID?

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

## How Can Chromosomal Abnormalities Cause ID?

* Down Syndrome (Trisomy 21) is associated with Moderate ID, characteristic appearance, weakness in verbal skills and language, strength in visual-spatial reasoning, and sociability.
* Prader–Willi Syndrome is caused by missing paternal genetic material on chromosome 15. It is associated with Mild ID, weakness in short-term memory, strength in visual-spatial reasoning, hyperphagia, and obsessive-compulsive behavior.
* Angelman Syndrome is caused by missing maternal genetic material on chromosome 15. It is associated with Moderate to Severe ID, sporadic/jerky motor movements, lack of spoken language, hyperactivity, and persistent social smile.
* Williams Syndrome is caused by deletions on chromosome 7. It is associated with Mild ID, well-developed spoken language, strengths in auditory memory, weakness in visual-spatial reasoning, hyperactivity, anxiety, and friendly/social demeanor.
* 22q11.2 DS is caused by deletions on chromosome 22. It is associated with Mild-to-Moderate ID, cleft lip/palate, social communication deficits, and risk for Schizophrenia later in life.

## How Can X-Linked Disorders Cause ID?

* Fragile X Syndrome is an inherited, X-linked disorder that adversely affects boys more than girls. It is characterized by Mild to Moderate ID, characteristic appearance, strengths in simultaneous processing, weakness in sequential processing, and social communication deficits.
* Rett Syndrome is usually caused by a genetic mutation in portion the X chromosome. It almost always affects girls. It is characterized by typical development in early infancy followed by rapid deterioration in social functioning and language, Severe ID, hand-wringing stereotypies, and seizures.

## How Can Metabolic Disorders Cause ID?

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

## How Can Maternal Illness or Environmental Toxins Cause ID?

* The acronym TORCH reflects the five most common maternal illnesses that can cause ID.
* Lead exposure during gestation or early childhood is associated with behavior and learning problems. In high elevations, lead toxicity can cause ID.
* Fetal Alcohol Spectrum Disorder is associated with lower intellectual functioning, learning disabilities, hyperactivity-impulsivity, inattention, and characteristic facial appearance. High exposure to alcohol during gestation can cause Mild ID.

## How can Perinatal or Postnatal Problems Cause ID?

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

## What Causes Cultural-Familial ID?

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

# 5.3 Prevention & Treatment

## How Do Professionals Screen for Developmental Disabilities?

* Serum screening is a maternal blood test that can be conducted 15-18 weeks gestation to detect the presence of some developmental disorders.
* Amniocentesis and chorionic villus sampling (CVS) are more invasive procedures that may be used when there is elevated possibility of the developmental disability.
* Physicians can use ultrasound to detect structural abnormalities in the fetus that might indicate a disability (e.g., Down Syndrome).

## Can Early Education Programs Prevent ID?

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

## What Services Are Available to School-Age Children?

* Academic inclusion is the practice of educating children with ID and other disabilities alongside typically-developing classmates for all subjects possible, usually with the support of a classroom aide.
* IDEIA requires school systems to identify infants and children with ID and other disabilities early in life and prepare an IFSP (for infants/toddlers) or IEP (for school-age children) to promote their development.
* Universal design is an educational practice that involves creating instructional materials and activities that allow learning goals to be achieved by children, regardless of their abilities and skills.

## How Can Clinicians Reduce Challenging Behaviors in Children with ID?

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

## How Can Clinicians Help the Caregivers of Children with ID?

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