Intellectual and Developmental Disabilities Overview

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Developmental Disability
Federal Definition

• Developmental Disability means a disability that is manifested before the person reaches twenty-two (22) years of age,
• is likely to continue indefinitely,
• results in substantial functional limitations,
• is attributable to intellectual disability or related conditions which include cerebral palsy, epilepsy, autism or other neurological conditions, and
• reflects the individual’s need for assistance that is lifelong or extended duration that is individually planned and coordinated.
Developmental Disabilities may include:

- Intellectual Disability
- Autism Spectrum Disorder
- Muscular Dystrophy
- Cerebral Palsy
- Fetal Alcohol Syndrome
- TBI
- Some genetic disorders (Down Syndrome, Prader-Willi, Fragile X)
Intellectual Disability

- Intellectual disability is a disability characterized by significant limitations both in **intellectual functioning** and in **adaptive behavior**, which covers many everyday social and practical skills.
- Generally an IQ score of around 70 or less indicates a limitation in **intellectual functioning**
- **adaptive behavior** includes three skill types:
  - 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.
Diagnosis of Intellectual Disability (ID) DSM-4

- 1. IQ below 70
- 2. Adaptive skills markedly delayed, equivalent to an IQ below 70
- 3. Occurred during the developmental period, administratively age 18
- Overall Clinical judgment
  - Made by physician or psychologist
  - *Never based on IQ alone
  - *Cannot really obtain IQ below 45
Normal distribution
(photo source: Wikipedia)

Number of scores

Intelligence Quotient
(Score on Wechsler Adult Intelligence Scale)

0.1% 2% 14% 34% 68% 34% 14% 2% 0.1%

55 70 65 100 115 130 145

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The diagnostic criteria for ID was never just based on an IQ test alone in DSM-4. Nonetheless, it is better to expand the diagnostic criteria to fully represent the breadth of cognitive and adaptive functioning, not just for diagnosis but so that we can understand the person better and therefore provide more appropriate supports.
In 2014, DSM-5 officially changed the diagnosis from “mental retardation” to “intellectual disability”

The most of the world has been using this term since the 1992

In concert with the rest of the world, the complexity of diagnosis was extended to be much more comprehensive and with less reliance on an “IQ” test as the most important measure
DSM-5 Chapter Organization

- Neurodevelopmental Disorders
- Schizophrenia Spectrum and Other Psychotic Disorders
- Bipolar and Related Disorders
- Depressive Disorders
- Anxiety Disorders
- Obsessive-Compulsive and Related Disorders
- Trauma- and Stressor-Related Disorders
- Dissociative Disorders
- Somatic Symptom and Related Disorders
- Feeding and Eating Disorders
- Elimination Disorders
• **Neurodevelopmental Disorders:**
  o Intellectual Disability (*Intellectual Development Disorder*), Global Developmental Delay (children < 5)
  o Communication Disorders –
    • Language Disorder, Speech Sound Disorder, Childhood-Onset Fluency Disorder, Social Communication Disorder
  o Attention Deficit Hyperactivity Disorder
  o Specific Learning Disorder
  o Autism Spectrum Disorder
  o Motor Disorders
    • Developmental Coordination Disorder, Stereotypic Movement Disorder, Tic Disorders/Tourette’s Disorder
Intellectual Disability vs. Intellectual Developmental Disorder

- Every medical condition is a disorder
- AAIDD advocated to keep it disability and use supports…
- ICD-11 intends to use Disorder of Intellectual Development (DID)
DSM-V Criteria

DSM-5 emphasizes the need to use both clinical assessment and standardized testing of intelligence when diagnosing ID, with the severity of impairment based on adaptive functioning rather than IQ test scores alone.

DSM-5 ensures that IQ – neuropsychological tests are not overemphasized as the defining factor of a person’s overall ability, without adequately considering functioning levels.
DSM 5  ID-IDD Criteria

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 socio-cultural standards for personal independence and social responsibility. Without ongoing support, the adaptive deficits limit functioning in one or more activities of daily life, such as communication, social participation, and independent living, across multiple environments, such as home, school, work, and community.

C. Onset of intellectual and adaptive deficits during the **developmental period**.

*Specify severity (based on adaptive function, not IQ): Mild, Moderate, Severe, Profound*
IQ test cognitive areas

- Language (verbal)
- Visual spatial skills
- Visual motor skills
- Social Reasoning
- Memory, verbal, visual, visual motor
Wechsler Scales areas average score 100

- Verbal IQ 69
- Performance IQ 65
- Full Scale IQ 66

- Typical mild ID
Verbal skills significantly higher

- VIQ 74  PIQ 63
- *These individuals will be judged to be more capable than they are due to our perceptions based on superficial verbal ability*
- Hands-on knowledge & practical reasoning can be poor, visual - motor skills less effective
Verbal ability lower than performance

• VIQ 63  PIQ 71
• *These individuals have good “hands-on” skills but may be hampered in tasks requiring verbal ability*
• Or, a specific verbal difficulty influenced performance on the IQ test
Adaptive Behavior

• “Gold standard” for diagnosis of ID
• Includes all our abilities to function in life, e.g., go to the store and buy an item
• Ideally, a formal scale of AB is used
• **Intellectual Disability** *(Intellectual Developmental Disorder)*:

• **Severity based on functional ability, not IQ, or adaptive functioning** in comparison with same age norms has been added as a criteria and must be assessed in 3 domains.

  1. Conceptual deficits: language, reading, writing, math, reasoning, knowledge and memory
  2. Social deficits: interpersonal communication skills, friendships, social judgment, empathy
  3. Practical deficits: personal care, organizing school and work activities, money management, job duties

Severity rating scale for each domain is based on the level of support required. Mild, Moderate, Severe, Profound
Conceptual deficits: language, reading, writing, math, reasoning, knowledge and memory

Assessment would include IQ testing, educational testing, neuropsychological tests
Social deficits: interpersonal communication skills, friendships, social judgment, empathy

Some instruments from Speech-Language pathology, pragmatic, some scales from IQ test, single items on adaptive skills tests, ? projective personality testing
Practical deficits: personal care, organizing school and work activities, money management, job duties

Vineland Social Maturity Scale, Adaptive Behavior Scales, vocational measurements
# IDD Severity Scale

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DSM-5 Specifiers

• Recommends adding useful information such as IDD with Down syndrome, strabismus, congenital heart defect
• These would have been added previously on Axis 3
• Also suggest adding, for example, age of onset, e.g. acquired brain injury, age 5
Developmental Perspective

- Children develop into adults by going through complex but predictable growth
- Development is affected by biological factors and environmental influences
- Generally, people with ID go through developmental stages more slowly and plateau early
• All children develop at different rates
• Within each individual person, abilities will develop at different rates
• For people with ID, we must be careful to see strengths and growth, rather than deficiencies and delays
All of us continue to grow and develop in adulthood, but this is mostly due to life experiences.

Life experiences affect maturity, social skills, and specific areas of expertise.

People with ID also grow and develop as adults, especially if they have rich environments and opportunities to learn new skills and experience new interests.
• Working memory: “brain power” in the moment to work on a problem (executive function)

• Expertise: “brain storage” to store information in a “zip file” (expertise)
Working memory and ID

- People with IDD may have smaller space for working memory
- They may need to work in smaller steps and handle less complex problems
- However, they do develop “zip files”; throughout life they continue to develop expertise.
• Working at a job, using public transportation, shopping – examples of expertise
• Due to expertise in certain areas, we may create higher expectations in other areas
“Falling farther behind is not the same as going backwards”
Multiple Causes of ID

- Genetic causes of ID
- Physical, e.g., birth & prenatal difficulties, injuries or illnesses in childhood
- Some individuals with early onset schizophrenia lose cognitive abilities
• There is a core set of characteristics shared by all people with ID/DD, regardless of cause.

• There are unique characteristics shaped by family, culture, community, and other experiences.

• There are also more specific patterns of abilities, deficits, and other characteristics associated with known genetic syndromes.
Autism Spectrum Disorder and Genetic Syndromes

Why is it important to understand?

• Both ASD and Genetic Syndromes are frequently seen with individuals with IDD
• Both have characteristics that can be misleading during assessment and diagnosing process
• Awareness and understanding of ASD and Genetic Syndromes add to our ability to appropriately support people
Autism Spectrum Disorder

• Incidence: 1 in 68 (males 1 in 30)
• Cause: current research suggests interaction between genetics environmental factors
• Cognition: wide range of intellectual functioning; 60% have IQ above 70.
• Why has incidence risen?
  o Better diagnosing
  o Better early intervention
ASD Characteristics – DSM 5

• Social Communication/ Social Interaction
  o Deficits in social-emotional reciprocity
    o Abnormal social approach, failure of normal back-and-forth conversation, reduced sharing of interests, emotions, or affect; failure to initiate or respond to social interactions
  o Deficits nonverbal communicative behaviors used for social integration
    o Limited eye contact and body language, deficits in understanding and use of gestures, decreased facial expressions and non-verbal communication
  o Deficits in developing, maintaining, and understanding relationships
    o Difficulty adjusting behavior to suit various social contexts, difficulty in sharing imaginative play or in making friends, absence of interest in peers
ASD Characteristics – DSM 5

• Repetitive/restrictive patterns of behavior, interest or activities
  o Stereotyped or repetitive motor movements, use of objects or speech
    o Simple motor stereotypies, lining up toys or flipping objects, echolalia, idiosyncratic phrases
  o Insistence on sameness, inflexible adherence to routines, or ritualized patterns of verbal or nonverbal behavior
    o extreme distress at small changes, difficulties with transitions, rigid thinking patterns, greeting rituals, need to take same route or eat same food every day
  o Highly restricted, fixated interests that are abnormal in intensity or focus
    o Strong attachment to or preoccupation with unusual objects, excessively circumscribed or perseverative interests
  o Hyper- or hypo-reactivity to sensory input or unusual interest in sensory aspects of the environment
    o Apparent indifference to pain/temperature, adverse response to specific sounds or textures, excessive smelling or touching of objects, visual fascination with lights or movement

• Thinking/Learning
  o Cognitive inflexibility
  o Focus on details/miss big picture
Genetic Syndromes

Why do we need to know about genetic syndromes?

• Professionals working with people with IDD need to be aware of the contribution of behavioral characteristics of genetic syndromes to diagnosis and treatment.

Knowing that a person with Down syndrome who is hallucinating is more likely to have major depressive disorder with psychotic features than schizophrenia, and that the cause may be hypothyroidism, is important to treatment.
Genetic Syndromes

Genetic Syndromes include:

- Angelman Syndrome
- Cri-du-Chat Syndrome
- Down Syndrome
- Fetal Alcohol Syndrome & Fetal Alcohol Spectrum Disorders
- Fragile X Syndrome
- Prader-Willi Syndrome
- Rubenstein-Taybi Syndrome
- Tuberous Sclerosis Complex
- Velocardiofacial Syndrome (DiGeorge Syndrome)
- Williams Syndrome
Physical Signs to Notice

- **Height** – small stature in Down Syndrome, Williams, Prader-Willi, Rubenstein-Taybi
- **Head size** – small in Down Syndrome and Angelman
- **Shape of Face** – elongated in Fragile X; small jaw in Williams; broad, square face in Smith-Magenis
- **Eyes:**
  - Small openings in FASD
  - Puffiness around eyes in Williams Syndrome
  - Downward slant to eyes in Cri-du-Chat
  - Upward slant to eyes in Down Syndrome
  - Almond shaped eyes in DiGeorge
Physical Signs to Notice

• **Ears** –
  o Large size in Fragile X
  o Low set ears in Cri-du-Chat and Rubenstein-Taybi
  o Small ears in DS

• **Hands** –
  o Simian crease in DS and Cri-du-Chat
  o Abnormalities seen in FASD

• **Skin** –
  o Pale skin – Angelman, Prader-Willi
  o Skin lesions – Tuberous Sclerosis

• **Low Muscle Tone** –
  o Down Syndrome and Prader-Willi
Behavioral Signs to Notice

- **Specific/Unusual Phobias** – Williams Syndrome
- **Unusual Self-Injury** – Smith-Magenis
- **Overly social** – Williams Syndrome
- **Hyperactivity** – Fragile X, Cri-du-Chat, Angelman, FASD, Tuberous Sclerosis
- **Impulsivity** – Fragile X, FASD, Rubenstein-Taybi, Smith-Magenis, DiGeorge
Medical and Psychiatric Risks

**Medical**
- Heart problems – Down Syndrome, Prader Willi, FASD
- Thyroid – Down Syndrome
- Dementia – Down Syndrome
- Sleep Issues – Prader Willi, Angelman Syndrome, Smith Magenis

**Psychiatric**
- Depression – Down Syndrome
- OCD/Anxiety – Down Syndrome, Fragile X, Prader Willi, Williams Syndrome
- Bipolar Disorder – VCFS
- Psychotic Disorder – Prader Willi, VCFS
Autism Spectrum Disorder and Genetic Syndromes

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Executive Function and IDD

Core Deficit in DD

- Many studies have shown that people with a developmental disability of any cause have a deficit in **executive functions** – set of mental processes that help us with planning, organization and task management.
- Deficits in executive functions affect:
  - attention
  - organization
  - time management
  - impulse control
  - memory
  - self-monitoring
Executive Functioning is a term used to describe a set of mental processes that helps connect past experience with present action.

EF enables us to learn new information, remember and retrieve information we’ve learned in the past, and use this information to solve problems.

This ability allows us to adapt and perform in everyday life by recognizing the significance of unexpected situations and to make alternative plans when unusual events interfere with normal routines.
Development of Executive Functioning

• Emerges in late infancy, goes through marked changes during the ages of 2 through 6, and does not peak until around the age of 25 in people with neurotypical development

• The brain continues to mature and develop connections well into adulthood

• A person’s EF abilities are shaped by both physical changes in the brain and by life experiences
Executive Functioning

- “Conductor of the Orchestra”
- Anticipation
- Goal selection
- Planning
- Initiation of activity
- Self regulation
- Mental flexibility
- Sustained focused attention
- Utilization of feedback-shifting set
Summary

- There are multiple causes of intellectual and developmental disabilities
- Level of severity of intellectual disability is based on adaptive functioning – not on IQ alone
- Executive functioning skills are affected in people with IDD
- Genetic disorders and autism spectrum disorder add to the complexity of assessment and planning
- People with IDD have individual patterns of strengths and deficits
- People with ID grow and develop all their lives, especially if they have rich environments and opportunities to learn new skills and experience new interests
- Appropriate supports are guided by understanding executive functioning, identifying specific patterns of skills and deficits, and focusing on strengths