Care of Patients with Intellectual and Developmental Disabilities

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Objectives

- Define intellectual and developmental disabilities (ID/DD)
- Epidemiology of ID/DD
- Health Disparities
- Primary Care Considerations – Basics, Routine Care, Legal
- Psychiatric Considerations – Psychopathology, Diagnostic Challenges, Assessment Tips, Polypharmacy

Definitions

Developmental Disability (DD)

Intellectual Disability (ID)

Definitions – Developmental Disability

- Any condition that involves an impairment in one's physical condition, learning, language, or behavior
- Begin during the developmental period (most commonly before birth) and typically are lifelong
- Causes: genetics, parental health/behavior during pregnancy, birth complications, congenital or neonatal infections, environmental toxins
- Examples: autism, cerebral palsy, vision or hearing impairment, fragile X syndrome, learning disorder, *intellectual disability*

Definitions – Intellectual Disability

- 3 criteria must be met
  - Limitation in intellectual functioning
  - Limitation in adaptive behaviors
  - Onset during developmental period

Definitions – Intellectual Disability

- 3 criteria must be met
  - *Limitation in intellectual functioning*
    - Measured by IQ testing
    - IQ 50-69 – mild ID (85% of cases)
    - IQ 35-49 – moderate ID (10% of cases)
    - IQ 20-34 – severe ID (4% of cases)
    - IQ < 20 – profound ID (1% of cases)
  - Limitation in adaptive behaviors
  - Onset during developmental period

Definitions – Intellectual Disability

- 3 criteria must be met
  - Limitation in intellectual functioning
  - *Limitation in adaptive behaviors*
    - How well a person meets community standards of personal independence and social responsibility
    - Conceptual skills – memory, language, practical knowledge (money, time, etc.)
    - Social skills – interpersonal communication, social judgment, gullibility, ability to follow rules
    - Practical skills – personal care (ADLs), iADLs, occupational skills
  - Onset during developmental period

Definitions – Intellectual Disability

- 3 criteria must be met
  - Limitation in intellectual functioning
  - Limitation in adaptive behaviors
  - *Onset during developmental period*
    - Before age 22

Definitions – Intellectual Disability

- 3 criteria must be met
  - Limitation in intellectual functioning
  - Limitation in adaptive behaviors
  - Onset during developmental period
- Causes – genetics, trauma, metabolic abnormalities, toxin exposure, infection, unknown
  - Severe ID most likely genetic
  - Mild ID most likely non-genetic

Epidemiology

- 1 in 6 children aged 3-17 in the US have a DD
- 1% of the global population has ID
  - 10-16 million people in the US
- UN Development Programme – 80% of all people with a disability live in a low income country

Health Disparities

- Shorter life expectancy
- Increased rates of medical problems
  - Obesity
  - Diabetes
  - Cardiovascular disease
  - Epilepsy
  - Covid-19 related deaths
- Decreased rates of routine preventive health screenings

Health Disparities – Why?

- Genetic factors
- Communication barriers
- Systematic barriers

Primary Care Considerations

Primary Care Considerations – The Basics

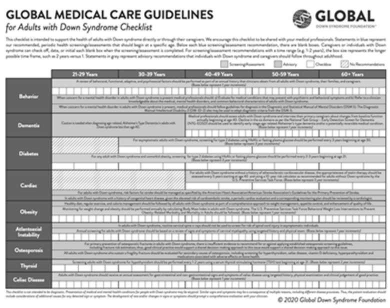
- Importance of knowing the social/living situation
- Allowing the patient to consent to examination and testing regardless of verbal communication skills
- Increased vulnerability for abuse
- Need for collateral information
- Prioritizing quality over quantity
- Consideration of sedation to facilitate exams/testing

Primary Care Considerations – Routine Care

- Follow general screening and immunization recommendations
  - Focus on sexual/reproductive health – one of the greatest disparities
- Higher rates of mental illness
- Screening labs
- If living in a group setting, screening for infectious diseases and vaccinating against hepatitis A and B

Primary Care Considerations – Syndrome Specific Concerns

- Down syndrome – National Down Syndrome Society


GLOBAL MEDICAL CARE GUIDELINES
for Adults with Down Syndrome Checklist

GLOBAL
Screening/Assessment Advisory Checkbox No Recommendations

	21-29 Years	30-39 Years	40-49 Years	50-59 Years	60+ Years
Behavior					
Dementia					
Diabetes					
Cardiac					
Obesity					
Atlantoaxial Instability					
Osteoporosis					
Thyroid					
Celiac Disease					

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Primary Care Considerations - Legal Issues

- Guardianship vs supported decision making
- Healthcare power of attorney
- Advanced directives

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Psychiatry Considerations

Psychiatry and ID

- "Dual diagnosis"
  - Co-existence of ID and mental illness
- Paradigm shift in field (~1980s)
  - Historically, believed persons with ID lacked cognitive capacity to develop psychiatric disorders

Rates of Psychopathology

- Estimates in literature vary considerably
- Prevalence 10-80% (most literature supports 30-50%)
  - 27% in general population
- Population sampled (gen vs psych outpatient, hospitalized, administrative samples)
- Definition of mental ill-health (AOD, challenging behaviors?)

Psychopathology, cont.

- Cooper et al (2007)
- Pop-based study of 1023 adults with ID in Greater Glasgow area
- Assessed by study RN with ID qualifications, discussed with GP
  - "possible, probable, or definite" mental ill-health ID psychiatry assessment
  - Dx: clinical judgment, DC-LD, ICD-10, DSM-IV

Findings (Point Prevalence)

	Clinical	DC-LD	ICD-10	DSM-IV
Psychotic	4.4%	3.8%	2.6%	3.4%
Affective	6.6%	5.7%	4.8%	3.6%
Anxiety	3.8%	3.1%	2.8%	2.4%
Mental Ill Health	40.9%	35.2%	16.6%	15.7%

Diagnostic Criteria for Psychiatric Disorders for Use with Adults with Learning Disabilities (DC-LD)

Psychopathology, cont.

- Schizophrenia spectrum psychosis
  - Prevalence in pt with ID ~ 3% (1% general population)
- Depression
  - Point prevalence of 3-4% (1.7% in gen pop)
- Bipolar
  - 1.5-2x rate of general population

Psychopathology, cont.

Genetic Syndrome	Psychiatric Condition
Down Syndrome	Depression, Anxiety, OCD, Alzheimer's
Velocardiofacial Syndrome	Psychosis
Fragile X	Anxiety, ADHD
Prader-Willi	Bipolar Disorder, Psychosis

Diagnostic Considerations/Challenges

- Accurate psychiatric dx is challenging
  - Underdiagnosis
  - Inaccurate diagnosis
  - Inadequate treatment of mental health issues
- Knowing special considerations for diagnosis → improved diagnosis, development of appropriate treatment plans

Diagnostic Challenges

- Sovner (1986) - 4 aspects of ID which increase difficulty of assessment

1) Baseline exaggeration: increase in severity/frequency of challenging behaviors during psychiatric illness (behavior becomes the focus)

2) Intellectual distortion: Pt cannot understand questions asked, nor formulate accurate response; deficits in abstract thinking, receptive/expressive language skills (Ex. "Do you hear voices?")

Diagnostic Challenges, cont.

3) Psychosocial masking: Due to developmental delay, pt may present with symptoms that occur within a developmental framework common in a young child, vs same age peer. (Ex. Imaginary friend mistaken for delusion)

4) Cognitive disintegration: Due to decreased ability to cope with stress, pt may become grossly disorganized/regress to more primitive behaviors and thus appear "psychotic" (Ex. Become mute, lose skills)

Diagnostic Challenges, cont.

- **"cloak of competence:"** tendency for pt with ID to attempt to hide disability
- **"acquiescence bias" or "yessing:"** tendency to please evaluator by answering falsely or in a manner that is inaccurate
- **Diagnostic overshadowing:** tendency for clinicians to overlook presence of psychopathology, attributing behavioral problems to being an artifact of underlying ID/DD

General Assessment Modifications

- Patient interview
  - Limit Y/N questions
    - Ask follow up questions to augment responses
  - Simple vocab/short sentences
  - Ask 1 question at a time, allow time to formulate response
  - Comprehension checks to ensure they understood the question
  - Use visual materials to complement interview; communication assistive devices

MAKE SURE TO PLAN FOR A LONGER ASSESSMENT

Assessment: Collateral Information

- Multiple sources of collateral
- Collateral from different settings - (home, school, work, day program)
- Clarify – how well does the informant know the patient?
- Caveats:
  - NOT from the patient's perspective
  - Externalizing symptoms (aggression) >>>> internalizing (withdrawal)

Challenging Behaviors

- SIB, aggression, property destruction
- #1 reason pt with ID brought to mental health attention
- Medical Hx:
  - Medical issues or drug side effects are common causes of behavioral changes BUT medical eval often neglected
  - constipation, UTI, thyroid dysfx, diabetes, dental disease, HA, menstrual pain

Anxiety + Challenging Behaviors

- Aggression is non-specific
  - Impaired psychosocial development → reduced capacity to regulate emotions/responses
  - Unable to articulate distress
  - "final common pathway"
- Provoking events or environments
- Assumption: managed with Rx → overlooking assessment of root cause → missed opportunity to address environmental issues

Developmental Effects on Psychopathology

- Developmental effects influence presentation
- DSM limitations
  - Developed with general population in mind → not reliable for PWID
    - Studies reflect use of DSM consistently results in lower rate of diagnosis
  - Emphasis on self-report - not possible or unreliable in some PWID
- Diagnostic Manual-Intellectual Disability
  - Collaboration between NADD and APA
  - Intended to assist psychiatric dx in PWID based on DSM-5 (DM-ID 2)

DM-ID 2: Depression

- Limited ability to self-report internal mood states/recognize and label feelings
  - Increased reliance on caregiver reports
- Developmental factors – less demonstration of certain cognitive features
- Neurodevelopmental profiles parallel younger, neurotypical peers
  - Ex, anhedonia:
    - NT adult – "I don't care to do things I used to enjoy"
    - PWID – throw 'tantrum' when prompted to engage in previously enjoyed activity

Depression, cont.

- Depressed mood – facial expressions (smile less, cry more), more irritability (angry/grouchy facial expression)
- Anhedonia – refuse activities, social withdrawal, participates in activities but doesn't appear to enjoy
- Feelings of worthlessness – negative self-statements ("I am bad"), reassurance seeking they are "good"
  - Severe to profound ID – do not have cognitive capacity to express these
- Thoughts of death/SI – speak more about death/morbid preoccupations; frequent comments about fears of illness or death; threats of/suicide attempts

Depression, cont.

Mild to Moderate ID

- Easier to dx depression
- Full range of dx criteria
- Mild cognitive difficulty + good expressive lang skills can be assessed much like general adult
  - BE SURE to have a solid understanding of their skills

Severe to Profound ID

- Cognitive symptoms not typically described in persons with little to no verbal ability
- May be unable to express hopelessness/feelings of guilt
- Emphasis on observable features
  - Eating patterns/weight, sleep, motor activity

Polypharmacy

- Significant and growing concern for overmedication
- Atlas on Primary Care of Adults with DD in Ontario (2013)
  - Adults with DD aged 18-64y in Ontario Drug Benefits Program
  - 52,404 people, April 2009-March 2010
- Findings (of entire DD sample):
  - 26% rx'd 2-4 meds concurrently
  - 13% 5-7 meds concurrently
  - 8% 8+ (up to 41 meds concurrently)

Polypharmacy, cont.

- Dual diagnosis increases risk for polypharmacy
  - ~26,504 were DDx, 25,900 were non-DDx
  - 59% of dual dx rx'd 5+ meds concurrently, compared to 35% non-dual dx
- Medication trends (entire DD sample)
  - Of the 10 most commonly rx'd, 5 were psychotropic
    - **21% - antipsychotic**
    - 13% - sedative
    - 12% - SSRI
    - 8% and 7% - VPA and carboxamide derivatives, respectively

Polypharmacy, cont.

- Of the 21% prescribed antipsychotics (AP)
  - 19% dispensed 2+ AP concurrently
  - 11% dispensed 2+ AP concurrently, continuously for 3 months
  - 7% dispensed 2+ AP concurrently, continuously for 6 months

In Summary…

- ID/DD is sizeable portion of population
- Increased rates of physical and mental health conditions
- Health disparities
- Careful consideration of personal and social environments
- Modify assessment/empower!!

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