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

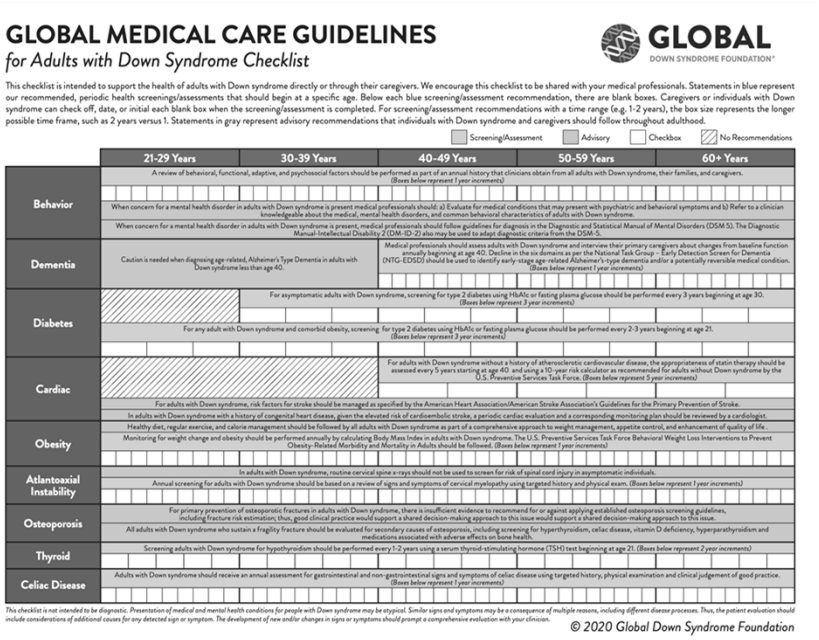
- Importance of knowing the social/living situation
- Allowing the patient to consent to examination and testing regardless of verbal communication skills
- Increased vulnerability for abuse
- Need for collateral information
- Prioritizing quality over quantity
- Consideration of sedation to facilitate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

This checklist is intended to support the health of adults with Down syndrome directly or through their caregivers. We encourage this checklist to be shared with your medical professionals. Statements in blue represent our recommended, periodic health screenings/assessments that should begin at a specific age. Below each blue screening/assessment recommendation, there are blank boxes. Caregivers or individuals with Down syndrome can check off, date, or initial each blank box when the screening/assessment is completed. For screening/assessment recommendations with a time range (e.g. 1-2 years), the box size represents the longer possible time frame, such as 2 years versus 1. Statements in gray represent advisory recommendations that individuals with Down syndrome and caregivers should follow throughout adulthood.

Screening/Assessment | Advisory | Checkbox | No Recommendations

	21-29 Years	30-39 Years	40-49 Years	50-59 Years	60+ Years
Behavior	A review of behavioral, functional, adaptive, and psychosocial factors should be performed as part of an annual history that clinicians obtain from all adults with Down syndrome, their families, and caregivers. *(Boxes below represent 1 year increments)*				
	When concern for a mental health disorder in adults with Down syndrome is present medical professionals should: a) Evaluate for medical conditions that may present with psychiatric and behavioral symptoms and b) Refer to a clinician knowledgeable about the medical, mental health disorders, and common behavioral characteristics of adults with Down syndrome.				
	When concern for a mental health disorder in adults with Down syndrome is present, medical professionals should follow guidelines for diagnosis in the Diagnostic and Statistical Manual of Mental Disorders (DSM 5). The Diagnostic Manual-Intellectual Disability 2 (DM-ID-2) also may be used to adapt diagnostic criteria from the DSM-5.				
Dementia	Caution is needed when diagnosing age-related, Alzheimer's Type Dementia in adults with Down syndrome less than age 40.		Medical professionals should assess adults with Down syndrome and interview their primary caregivers about changes from baseline function annually beginning at age 40. Decline in the six domains as per the National Task Group – Early Detection Screen for Dementia (NTG-EDSD) should be used to identify early-stage age-related Alzheimer's-type dementia and/or a potentially reversible medical condition. *(Boxes below represent 1 year increments)*		
Diabetes		For asymptomatic adults with Down syndrome, screening for type 2 diabetes using HbA1c or fasting plasma glucose should be performed every 3 years beginning at age 30. *(Boxes below represent 3 year increments)*			
	For any adult with Down syndrome and comorbid obesity, screening for type 2 diabetes using HbA1c or fasting plasma glucose should be performed every 2-3 years beginning at age 21. *(Boxes below represent 3 year increments)*				
Cardiac			For adults with Down syndrome without a history of atherosclerotic cardiovascular disease, the appropriateness of statin therapy should be assessed every 5 years starting at age 40 and using a 10-year risk calculator as recommended for adults without Down syndrome by the U.S. Preventive Services Task Force. *(Boxes below represent 5 year increments)*		
	For adults with Down syndrome, risk factors for stroke should be managed as specified by the American Heart Association/American Stroke Association's Guidelines for the Primary Prevention of Stroke.				
	In adults with Down syndrome with a history of congenital heart disease, given the elevated risk of cardioembolic stroke, a periodic cardiac evaluation and a corresponding monitoring plan should be reviewed by a cardiologist.				
Obesity	Healthy diet, regular exercise, and calorie management should be followed by all adults with Down syndrome as part of a comprehensive approach to weight management, appetite control, and enhancement of quality of life .				
	Monitoring for weight change and obesity should be performed annually by calculating Body Mass Index in adults with Down syndrome. The U.S. Preventive Services Task Force Behavioral Weight Loss Interventions to Prevent Obesity-Related Morbidity and Mortality in Adults should be followed. *(Boxes below represent 1 year increments)*				
Atlantoaxial Instability	In adults with Down syndrome, routine cervical spine x-rays should not be used to screen for risk of spinal cord injury in asymptomatic individuals.				
	Annual screening for adults with Down syndrome should be based on a review of signs and symptoms of cervical myelopathy using targeted history and physical exam. *(Boxes below represent 1 year increments)*				
Osteoporosis	For primary prevention of osteoporotic fractures in adults with Down syndrome, there is insufficient evidence to recommend for or against applying established osteoporosis screening guidelines, including fracture risk estimation; thus, good clinical practice would support a shared decision-making approach to this issue would support a shared decision-making approach to this issue.				
	All adults with Down syndrome who sustain a fragility fracture should be evaluated for secondary causes of osteoporosis, including screening for hyperthyroidism, celiac disease, vitamin D deficiency, hyperparathyroidism and medications associated with adverse effects on bone health.				
Thyroid	Screening adults with Down syndrome for hypothyroidism should be performed every 1-2 years using a serum thyroid-stimulating hormone (TSH) test beginning at age 21. *(Boxes below represent 2 year increments)*				
Celiac Disease	Adults with Down syndrome should receive an annual assessment for gastrointestinal and non-gastrointestinal signs and symptoms of celiac disease using targeted history, physical examination and clinical judgement of good practice. *(Boxes below represent 1 year increments)*				

This checklist is not intended to be diagnostic. Presentation of medical and mental health conditions for people with Down syndrome may be atypical. Similar signs and symptoms may be a consequence of multiple reasons, including different disease processes. Thus, the patient evaluation should include considerations of additional causes for any detected sign or symptom. The development of new and/or changes in signs or symptoms should prompt a comprehensive evaluation with your clinician.

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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