Evaluating for Dementia with the Intellectually Disabled Population

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Objectives

• Demonstrate understanding of the clinical manifestations of dementia
• Identify the medical/psychiatric issues that can complicate the presentation of this population and impact diagnosis
• Identify tools and materials used to diagnosis dementia with the ID population
Dementia Overview

- Alzheimer’s Disease most common cause
- Vascular Dementia
- Lewy Body Dementia
- Frontotemporal Dementia
- Mixed
- Other - Huntington’s, TBI, Parkinson’s
Alzheimer’s Disease

- Progressive and degenerative
- Plaques – clumps of beta-amyloid protein
- Tangles – tau protein
- Clinical course described by early, middle and late stage features
Mimics dementia

- Infection/immune disorder
- Metabolic problems
- Nutritional Deficiencies
- Medication side effects
- Subdural hematomas
- Poisoning
- Anoxia
- Normal Pressure Hydrocephalus
ICD 10 Diagnostic Criteria

1. Evidence of decline in memory, most evident in the learning of new information.

2. A decline in other cognitive abilities and daily living skills
   - Language comprehension and expression
   - Perception
   - Executive function
   - Usual daytime activities
   - Use of household utensils and equipment

3. Absence of clouding of consciousness/delirium
ICD 10

• 4. Decline in emotional control, motivation or social behavior in at least one of the following: ■ Emotional lability ■ Irritability ■ Apathy ■ Coarsening of social behavior

• 5. The duration in changes in memory must be longer than 6 months
DSM V

• Changed wording to Mild and Major Neurocognitive Disorder
• Focus on decline of functioning, not just memory functioning

• Mild – beyond normal aging, able to engage in compensatory strategies and accommodations to allow for independence

• Major – cognitive deficits interfere with independence
  – At least 2 domains impaired
Early Stage

• Changes appear gradually
• Changes subtle
• Triad of cognitive deficits: memory impairment, expressive language and visual-spatial disturbance
Early Stage

• Memory impairment often presenting complaint
• Difficulty forming new memories as well as rapid forgetting
• Receptive language preserved
• Visual spatial less complained about
Early Stage

• Problem solving changes
  – Unable to use previous strategies
  – Trouble with novel problems

• Emotional changes – subtle
  – Less interest in others, life
  – Restless, irritable
Early Stage

- Awareness of deficit – can trigger denial, anxiety
- Stage lasts 1-5 years
- Neurologic exam may remain within normal ranges
- Input pathways from entorhinal cortex and hippocampal areas disrupted
Middle Stage

• Struggle with disorientation
• Problems understanding instructions
• Episodic memory loss
• Increasing difficulty with language
  – Naming/conversation
  – Word errors
Middle Stage

• Difficulties with self care
• Problems with initiating and organizing motor tasks
• Functional declines
• Stage ranges 5-15 years
Middle Stage

• Neuropathological changes - medial temporal lobes to the parietal and frontal cortex areas

• Eventually consumes most of neocortex
Late Stage

- Global disorientation and confusion
- Remote memory loss
- Basic skills forgotten
- Fragmented speech
- General physical deterioration
- Totally dependent
Late Stage

• Stage lasts 3-5 years
• Medical complications common
  – UTI
  – Pneumonia
  – Aspiration
  – Sepsis
### Dementia Rates General Population

<table>
<thead>
<tr>
<th></th>
<th>All Dementia</th>
<th>Alzheimer’s Disease</th>
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<tbody>
<tr>
<td><strong>Age 71-79</strong></td>
<td>4.97</td>
<td>2.32</td>
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<tr>
<td><strong>Age 80-89</strong></td>
<td>24.19</td>
<td>18.10</td>
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<tr>
<td><strong>Age 90 +</strong></td>
<td>37.20</td>
<td>29.6</td>
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</tbody>
</table>

Dementia Rates ID Population (nonDS)

<table>
<thead>
<tr>
<th></th>
<th>All Dementia</th>
<th>Alzheimer’s Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>&gt;59</td>
<td>13.1</td>
<td>&gt;59</td>
</tr>
<tr>
<td>&gt;64</td>
<td>18.3</td>
<td>&gt;64</td>
</tr>
</tbody>
</table>

Dementia Rates Down Syndrome Population

Age Range
• 30-39
• 40-49
• 50-59
• >59 http://www.wai.wisc.edu/pdf/IDDtoolkit.pdf

Prevalence Rates %
• < 3
• 10-25
• 20-50
• 30-75
Clinical Manifestations

• Changes in personality and behavior
  – Emotional lability
  – Increased irritability
  – Apathy/inactivity
  – Stubbornness
Clinical Manifestations

• Cognitive loss
  – Changes in adaptive functioning
  – Decline in frontal/executive functions
  – Judgement/planning
  – Visual organization
  – Semantic/short term memory
Clinical Manifestations

• Decline in ADL’s
  – Personal hygiene
  – Dressing
  – Eating

• Increase in maladaptive behaviors
  – Decreased social engagement/apathy
Aspirational Goals

• National Task Force on Intellectual Disabilities and Dementia Practices
• Detailed medical history
• Physical/neurological workup
• Psychiatric assessment
• Neuropsychological evaluation
Aspirational Goals

• Routine tests
  – Blood work
  – Urinalysis
  – EEG
  – Chest x ray

• Minimum: annual evaluations and rescreening
Diagnostic Challenges

- Relies on documentation of declines in cognitive function and adaptive skills
- Lack of consistent and reliable documentation
- Often lack ability to complete standard tests
- Atypical presentation
- Other causes of decline
Psychiatric Comorbidity

• Anxiety

• Behavioral disturbance

• Depression
  – Behavioral changes, social withdrawal
  – Loss of adaptive living skills
  – Mood changes
Life Stress

• Death/grief

• Loss of primary caretaker can lead to many other changes
  – Residence
  – Day activities
  – Provision of care
Life Stress

- Disruptions in work routine
- Staff changes
- Disruptions in home environment
- Personal relationships
- Change in family visits
- Anniversaries of losses
Medical Complications

- UTI – delirium
- Problems hearing
- Visual changes
- Dehydration
Challenges to diagnosing

- May not have verbal skills to explain feelings
- Uninformed staff or clinicians may misdiagnose
- Some behaviors/responses can be confusing: self talk, verbal processing of emotion
Observe for symptoms

• Looking sad, preoccupied or tearful
• No longer initiates activities or participates in activities previously enjoyed
• More easily irritable or upset more than usual for a given situation
• Appears restless or agitated
What does that suggest?

- Think of behaviors that are examples of diagnostic criteria
- There are scales: Mood Assessment Scale for Demented Adults (Sunderland et al, 1988)
- Various psychopathology scales for use with ID population
Important Considerations

• Make environment relaxed
• Remove physical barriers
• Position self to allow for communication with patient and caregiver
• Distraction/noise free
• Allow plenty of time
Important Considerations

• Avoid framing questions yes/no
• Ask permission to ask other people present questions
• Build some rapport before going into evaluation
• Allow plenty of time
Important Considerations

• Establish and maintain eye contact
• Be friendly, remember to smile
• Use a gentle tone of voice
• Remain calm, provide reassurance
• Be patient and supportive
• Allow plenty of time
Evaluation for Dementia

• Requires a change in baseline functioning – not a change from “normal” level of age based functioning

• More accurate diagnosis = documenting change over time
Baseline Functioning

• Can vary widely among people
• Great to screen when healthy
• Not the typical practice
• Good informants know the patient well
• Using different informants over time can be helpful
Traditional Methods

• Weschler Memory Scale
• Dementia Rating Scale
• Saint Louis University Mental Status
• Vineland Adaptive Functioning

• Use what works
Dementia Rating Scale for Down Syndrome

• Gedye 1995
• Can be used to establish baseline
• Same form can be used for 10 assessments of same client
• Allows for staging early, middle or late
• Good sensitivity and specificity
Dementia Questionnaire for people with Learning Disabilities

• Designed as a screening instrument
• Also provides a measure of general disability
• Not appropriate for people with severe or profound ID
• Not appropriate for single application
Dementia Screening Questionnaire for Individuals with Intellectual Disability (DSQUID)

Deb et al. 2007

Developed to overcome floor effects
Assesses best ability as well as symptoms
Quick and easy to score
Good sensitivity and specificity
Useful for baseline as well as more advanced
Test of Severe Cognitive Impairment

- Originally designed as a downward extension of the MMSE
- Assesses a range of skills: motor, language, memory, conceptualization and general knowledge
Adaptive Behavior Dementia Questionnaire

• Prasher et al., 2004
• 15 items found to be strongly linked between declines in adaptive skills and aging in older adults with DS
• Good sensitivity and specificity
• No measure of general disability
My evaluation

• Use Dementia Rating Scale for Down Syndrome
• Involve family if available as well as group home caregivers
• Complete a writing sample at each visit
• Complete a clinical interview with each visit
My evaluation

• Review for any medication/health changes, loss, life changes
• Talk to patient directly, even if nonverbal
• Spend time building rapport, make notes of favorites (conversation starters later on)
• After baseline review in 6 months
My evaluation

• If no changes – re-evaluate in 12 months
• If changes – re-evaluate again in 6 months
• Every 6 months until a period of stability
• Challenges with consent
• Scheduling
• Questions?

• Thank you for your time and attention!