WAI Memory Diagnostic Clinic
Network
Assessment of Dementia in
Individuals with Intellectual Disability
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Objectives
- Identify the clinical manifestations of AD dementia in adults with ID with an emphasis on Down Syndrome
- Provide an overview of the Baseline Comparison Model of assessment and its application to the diagnosis of dementia in ID
- Provide an overview of cognitive and functional assessment measures appropriate for use with adults with ID

Why is this important?
- Population prone to develop age-related d/o such as dementia
- Well recognized association between DS and Alzheimer Disease
- Increasing life expectancy of people with ID
- Increasing pressure on carers, support system
- ID people represent particularly vulnerable population

Dementia Rates in General Population
Brookmeyer et al. (2011)

<table>
<thead>
<tr>
<th>Age Group</th>
<th>All Dementia</th>
<th>AD</th>
</tr>
</thead>
<tbody>
<tr>
<td>71-79</td>
<td>4.97</td>
<td>2.32</td>
</tr>
<tr>
<td>80-89</td>
<td>24.19</td>
<td>18.10</td>
</tr>
<tr>
<td>90+</td>
<td>37.20</td>
<td>29.60</td>
</tr>
<tr>
<td>Total</td>
<td>13.67</td>
<td>9.51</td>
</tr>
</tbody>
</table>

Average Age Onset: 75 years
Average Duration: 8-10 years (3-20+)

Dementia Rates in DS
Ball et al. 2006

<table>
<thead>
<tr>
<th>Age Group</th>
<th>All Dementia</th>
<th>AD</th>
</tr>
</thead>
<tbody>
<tr>
<td>30-39</td>
<td>&lt;3%</td>
<td></td>
</tr>
<tr>
<td>40-49</td>
<td>10-25%</td>
<td></td>
</tr>
<tr>
<td>50-59</td>
<td>20-50%</td>
<td>&gt;59</td>
</tr>
<tr>
<td>&gt;59</td>
<td>30-75%</td>
<td></td>
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</tbody>
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Average Age of Onset: 51.67 yrs (31-68)
Average Duration: 3.5 – 10.5 yrs

Dementia Rates in Non-DS ID
Strydom et al. 2007

<table>
<thead>
<tr>
<th>Age Group</th>
<th>All Dementia</th>
<th>AD</th>
</tr>
</thead>
<tbody>
<tr>
<td>&gt;59</td>
<td>13.1</td>
<td>8.6</td>
</tr>
<tr>
<td>&gt;64</td>
<td>18.3</td>
<td>12.0</td>
</tr>
</tbody>
</table>

Average age of Onset: 67 years
Susceptibility to other causes of Cognitive Decline

- Conditions that produce a (potentially) reversible dementia
  - Hypothyroidism
  - B12/Folate Deficiency
  - Depression and pseudodementia (and life stress or other MH disorder)
  - Medications

Conditions that Mimic Dementia

- Sleep Apnea
- Cardiac abnormalities
- Infections
- Pain/Joint problems
- Sensory impairments
- Malignancy

Diagnostic Challenges

- Premorbid Cognitive Deficits
- Heterogeneity of skills
- Dx Overshadowing
- Atypical Presentation

Need for Coordinated Screening Process Tailored to Adults with ID

- Follow longitudinally
- Conduct baseline assessments when healthy
- Monitor for signs and symptoms of decline
- Document declines from “best level of performance”

Baseline Comparison Approach

- Screen when healthy (DS <36 yoa; Non-DS ID <50 yoa)
- Use direct assessment measures appropriate for use with ID
- Use indirect assessments with carers who know the individual well
- Periodic re-screening and referral for formal dx work-up when change noted

Diagnostic Work-up Includes

R. Pary (1992)

- Vitals
- Mental Health
- Physical Exam
- Sleep/appetite/weight
- Blood Count
- Neuropsych Testing
- Urinalysis
- ECG
- Hearing/Vision
- Consider: CT/MRI; EEG
### Differential Dx: Pay special attention to these common conditions
- Depression or other MH impairment
- Sensory Impairments
- Thyroid; B12/Folate Deficiency
- Acute/Chronic Medical Conditions - infection, pain, epilepsy, OSA/CSA, cardiac abnormalities
- Medications
- Major Life Stressors

### Diagnostic criteria
- Currently there are no criteria specific to the diagnosis of dementia in individuals with ID
- Consider use of ICD-10 criteria as they place greater emphasis on non-cognitive aspects of dementia and make use of a two-step diagnostic process

### Clinical Manifestations

### Atypical Presentation
- Core Sxs of dementia in ID adults involves progressive loss of function in multiple cognitive domains similar to the general population
- Presentation of dementia in ID is different (especially in DS) as personality and behavior change mark the early stages of decline

### Personality and Behavior Change
- Emotional lability
- Irritability
- Apathy/Inactivity
- Stubbornness and coarsening of social behavior

### Atypical Presentation
- There may be a corresponding decline in frontal-executive function (and related adaptive function) prior to the onset of memory impairment or full blown DAT
- This has led some authors to conclude that a FTD represents the pre-clinical stage of AD, particularly in adults with DS
Memory Loss: Mild-Mod ID (IQ 40-70)

- Forgetfulness for names and recent events
- Forgetfulness for location of everyday items
- Disorientation to time and temporal sequence of events
- Spatial disorientation in getting around home, neighborhood or work

Memory Loss: Mild-mod ID

- Frequent reminders required to carry out daily tasks
- Difficulty remembering the steps necessary to perform previously mastered tasks or directions

Memory Loss: Severe to Profound (IQ <40)

- Ability to assess depends on premorbid verbal skills and ability to meet task demands
- Profound ID (MA<2) may preclude detection on standardized tests
- Need to rely on informant report
- Use of neuro signs-myoclonic jerks, Sz activity, abnormal posture/gait, rigidity, incontinence

Cognitive Loss: Mild –Mod ID

- If verbal skills well developed will see word-finding difficulty, dysnomia and diminished command following
- If premorbid verbal skills poor will see decreased use of language progressing to total loss of verbal expression
- Loss of previously acquired skills to read, write, count, draw and color

Cognitive Loss: Mild- Mod ID

- Decreased ability to carry out skilled purposeful motor acts necessary for basic ADLs-dressing, grooming, bathing, toileting, self-feeding
- Inappropriate use of everyday objects

Cognitive Loss: Severe to Profound ID

- General slowing in all areas
- Greater impairments to attention
- Decreased temporal and spatial orientation
Everyday Functioning

Functional declines are first evident in more complex skills with progression to more fundamental basic skills later on.

Everyday functioning-early changes

• Decline in work productivity
• Household chores skills-table setting, washing dishes, making meals, making bed, use of appliances
• $ handling, banking and budgeting, ability to shop
• Sense of direction, use public transportation
• Dependability with assigned activities

Everyday functioning-later changes

• Initiative, persistence and perseverance in activities to structure pass time
• Basic arithmetic, counting and time telling
• Toileting, Bathing and personal hygiene
• Ability to eat, understand spoken language and ambulate last to be affected

Common Physical Signs

Lai & Williams, (1989)

• Late onset seizure or new type of seizure
• Urinary incontinence
• Slowness and shuffling of gait
• Pathological reflexes
• Postural abnormalities
• Limb rigidity
• Pathological reflexes

Assessment Measures

MMSE and Floor Effects

Meyers (1987)

<table>
<thead>
<tr>
<th>Age Range</th>
<th>Ave. IQ (Range)</th>
<th>Ave. MMSE (Range)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hosp. DD  (13-58)</td>
<td>60.9 (42-69)</td>
<td>22.6</td>
</tr>
<tr>
<td>Wksp (19-57)</td>
<td>53 (36-75)</td>
<td>22.7 (18-28)</td>
</tr>
<tr>
<td>Act.Ctr. (19-57)</td>
<td>35.8 (15-63)</td>
<td>17.5 (5-29)</td>
</tr>
</tbody>
</table>
Floor Effects
Of 62 demented and non-demented ID adults, Deb and Braganza (1999) found:
1) Only 34 cases (55%) could perform a MMSE
2) 30 of those cases (95.2%) scored <24
3) 23 Ss(77%) did not have a dx of dementia
4) Informant ratings (DMR;DSDS) proved far superior in detecting dementia

Test of Severe Impairment
Albert and Cohen (1992); Cosgrove et al. (1998)
- Validated in persons with ID
- Assesses a range of skills-motor, language, memory, conceptualization, general knowledge
- 8 Questions require verbal respons
- Brief and easy to use
- Yields range of scores

ToSI
- Psychometric properties are good
- Correlated with tools that measure functional decline (e.g. DLSQ) at baseline and dx
- >sensitivity than MMSE to measure change

Other Tests and Measures
- In a systematic review Zeilinger, Stiehl & Weber (2013) found 114 different instruments for assessment of dementia in ID
  - 79 direct assessment measures
  - 35 informant based measures
  - 4 Test batteries

Assessment Measures
- Select only those designed or adapted for use in detecting dementia in ID
- Must be reliable/valid sensitive to change early and through out the later stages of dementia and measure a wide range of cognitive domains
- Consider Practical Issues-time requirements, level of expertise required to administer, score and interpret tests

NTG-Early Detection Screen for Dementia (NTG-EDSD) 
Esralew et al. (2013)
- Offers carers and staff a resource to record changes in cognitive and adaptive function known to be associated with dementia
- Not an assessment/dx instrument but provides info to begin conversations with healthcare providers
- Adapted from DSQIID (Deb, 2007)
NTG-EDSD
- Relevant Demographics
- Ratings of Health, MH and life Stressors
- Review multiple domains of function
- Survey of chronic health conditions
- Signal items

Observer-Rated Scales
- Dementia Scale for Down Syndrome (DSDS)
- Dementia Questionnaire for Mentally Retarded Persons (DMR)
- Camdex-DS
- Dementia Screening Questionnaire for Individuals with Intellectual Disabilities (DSQIID)

Dementia Scale for Down Syndrome
Gedye (1995); Prasher (2009)
- An informant-based instrument designed to aid in the dx of dementia in adults with ID, especially DS.
- Also used to establish a baseline for those at risk
- Items grouped into early, middle, late and very late stages of dementia

DSDS
- Informant is asked to classify features as N/A, absent or present; if present, is feature typical or atypical
- New signs are tallied for each stage along with a separate tally of cognitive signs and then compared to criteria for dx and staging
- Provides a Diff’l Dx Screening questions Section to address other potential causes for dementia

DMR
Evenhuis (1992); Prasher (2009)
- Based on observations of caregivers over previous two months
- 50 items (8 subscales) divided into two subcategories
- Cognitive subcategory - stm/ltm,spatial and temporal orientation
- Social subcategory-speech, practical skills, mood, activities, interests, behaviors

DSDS
- Specificity-89%; sensitivity-85%
- Comprehensive but contains no measure of general disability
- Use restrictions apply
- Correlates well with other observer-rated instruments
DMR
- 3 response categories-0 (no deficit) to 2 (severe deficit)
- No use restriction or specialized training required
- Useful measure of general disability
- Quick and easy to use (15-20 minutes), short form available
- Floor effects with advanced dementia

Camdex-DS
Hon et al (1999); Ball et al (2006)
- Modified version of Cambridge Exam for Mental Disorders of the Elderly used to document increasing prevalence with age
- Designed for use in the community by trained healthcare professionals
- Contains informant interview and direct cognitive assessment
- Good reliability, predictive validity; floor effects

DSQIID
Deb (2007)
- Derived from interviews with caregivers of adults with DS and dementia
- Validated on large sample
- 53 items in 3 parts
- Part 1 assesses “best” ability
- Part 2 behaviors and symptoms suggestive of dementia (4 point scale)
- Part 3 ten comparative questions (Yes/No)
- Sensitivity-92%; Specificity-97%
- Quick and easy to score in any setting
- Single fixed cut-off may limit usefulness in adults with more advanced stages of dementia and with varying degrees of baseline ID

Direct Neuropsychological Measures
- Test of Severe Impairment (ToSI)
- CAMCOG-DS
- Institute for Behavioral Research Evaluation of Mental Status (IBREMS)

CAMCOG-DS
Hon et al (1999); Ball et al (2006)
- Self contained neuropsych component of the Camdex
- Validated in subjects with DS
- 7 subscales-orientation, language, memory, attention, praxis, abstraction, perception
- Few floor effects
- Score correlates with age
IBREMS
Wisniewski & Hill (1985); Sikerman et al. (2004)
• Adults respond to 37 items assessing orientation, STM/LTM, Language, Writing, Drawing & General Knowledge
• Recall and Recognition format provided
• If IQ available (when healthy) gives cut-off score for one assessment
• Sensitivity= .89; Specificity=.9

Adaptive Behavior Scales
• Adaptive Behavior Dementia Questionnaire (ABDQ)
• Daily Living Skills Questionnaire (DLSQ)

ABDQ
Prasher et al. (2004)
• Contains 15 items from ABS found to be strongly linked between declines in adaptive skills and aging and dementia in older adults with DS
• Items rated much worse, worse, same, better or much better than normal
• Weighted scores are summed and compared to cut-off
• Sensitivity=89%; Specificity=94%

ABDQ
• User friendly
• Can be done on all older DS adults regardless of level of ID or cooperation level
• No measure of general disability

Daily Living Skills Questionnaire
National Institute of Aging (1989)
• Informants provide info concerning a variety of ADLs including dressing, grooming, eating, manual dexterity and geographical orientation
• No significant floor effect
• High positive predictive value
• Correlates strongly with direct cognitive tests

Summary
• Diagnosis of dementia in ID is challenging and complicated by atypical presentations and baseline intellectual limitations
• Personality and behavior changes often pre-date the onset of memory or other cognitive loss in ID dementia
• There are no criteria specific to the dx of dementia in ID. Criteria commonly used in the general population do not take into account the unique non-cognitive features of dementia in ID
Summary

• Use of the ICD-10 dementia dx criteria is recommended as these place greater emphasis on the non-cognitive aspects of dementia and employ a two-step differential diagnostic process
• Standard assessment measures used “at a single point in time” to document declines in the general population presume a normal level of premorbid functioning and are thus inappropriate for use with this population as many ID adults lack the skills to perform these tasks at baseline.

Summary

• Longitudinal assessment with measures appropriate for use with ID adults is required to document changes in status from a baseline or “personal best” level of function
• Baseline screenings should ideally be done when healthy and periodically repeated
• Comprehensive work-up should be initiated when change is detected.

Summary

• A number of neuropsychological assessment measures have been developed to aid in the clinical diagnosis of dementia in adults with ID
• Combining direct assessment measures and observer rated scales provides the highest sensitivity and specificity

Thank You!

• Next Steps
• Questions/Comments