Assessment Issues in Detecting Cognitive Impairment Among Persons with ID:

1) Adults with Intellectual Disability

2) Adults with Down Syndrome

3) Adults with Dual Diagnosis (MI/ID)

Broad issues
Key concerns
Recommendations

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Cognitive Assessment of Individuals with Down Syndrome

Broad Issues

- Early onset dementia, with average age of onset in early 50s
- May see rapid deterioration from point of diagnosis
- Decline in executive functioning and increase in behavioral problems more likely to signal early signs of dementia rather than memory loss
- Conditions that are highly co-prevalent with DS may confound differential diagnosis
- Diagnostic overshadowing may obscure differential diagnosis; practitioners without DS-specific training may have tendency to attribute all cognitive, behavioral and functional changes to AD, overlooking other factors that may account for observed decline from baseline

Considerations

Collect data on baseline cognitive, behavioral, and functional skills beginning at age 40

Need for successive short-term intervals to document progressive decline (as in the ***NTG-EDSD***)

Sensory losses, thyroid disorder, sleep apnea, late onset myoclonus, communication (increasing dysarthria with age), catatonia, regression syndrome

Consider use of biomarkers to aid in differential diagnosis

Cognitive Assessment of Individuals with Intellectual Disability (non-DS)

Broad Issues

- Dementia in non-DS adults with intellectual disability less studied and less well understood
- Different neurodevelopmental disorders may be associated with unique trajectories of cognitive, behavioral or functional change as the individual ages
- Measures are neither designed nor adapted for individuals with already documented memory, attention and learning problems
- Assessment among neurotypical adults involves normative comparison; assessment among adults with ID involves comparing an individual against his/her own baseline performance
- No brief in-office assessment comparable to the MMSE, SLUMS, MoCA
- Multiple conditions can confound differential diagnosis: medical, mental health, environmental, psychosocial stressors

Considerations

- Longitudinal studies needed to profile the trajectory of cognitive and functional aging changes for persons with ID (non-DS)
- Autism, Prader-Willi, & Williams syndrome among neurodevelopmental disorders that may have unique trajectories for aging changes
- Need to capture information about baseline cognitive, behavioral and functional status by age 50 in order to identify departures from baseline
- A brief in-office screen needed for persons with ID which can flag the need for formal assessment and comprehensive work-up; this screen needs to be easily administered as part of AWV
- Since observed change likely to occur within family or residential, work or day program settings, need to educate family and staff about indications for referral to PCP/HCP
- Identify a protocol for assessment of individuals with in severe or profound range of intellectual disability

Cognitive Assessment of Individuals with Dual Diagnosis: Intellectual Disability and Mental Health Needs

Broad Issues

- Higher percentage of adults with ID who have co-occurring mental health needs compared with chronological neurotypical peers
- Chronic mental health conditions such as schizophrenia, bipolar disorder, major depressive disorder associated with neurocognitive changes resulting in non-AD dementia
- Differential diagnosis of frontotemporal dementia, pseudodementia, bipolar dementia, behavioral and psychological symptoms of dementia, hallucinations

Considerations

- No standard instrument used in assessment of mental health needs of individuals with ID
- Promote use of the DM-ID-2 (2013)
- High index of suspicion of adults with ID who have a history of SMI: MDD, bipolar disorder, schizophrenia
- Elevated risk for dementia among adults with ID and SMI
- Identify the incidence of non-AD neurocognitive disorders; profile the cognitive, behavioral and functional changes associated with SMI and long-term use of psychoactive medications

Key Takeaways - 1

- Early recognition of neurocognitive changes among adults with ID is delayed because of the lack of a universally accepted brief in-office screening for MCI or neurocognitive disorder that could be used during AWV
- Need for a brief, in-office assessment that can be used to screen adults with ID during their AWV for referral for formal assessment and comprehensive work-up to rule out neurocognitive disorders
- Any brief screen that can be used during the AWV needs to be compatible with electronic medical records to allow for serial comparison of status across visits
- Practitioners would benefit from the development of consensus guidelines for referral to PCP/HCP following observed change in cognitive, behavioral, or functional status

Key Takeaways - 2

- Adults with Down syndrome differ in presentation with early signs of dementia from their neurotypical peers; PCP/HCPs would benefit from education on early onset dementia
- Practitioners need education about the range of condition that may complicate differential diagnosis
- Need for data gathering tool because observation of change is most likely to occur within family or residential setting
- Not enough known about non-AD neurocognitive disorders in this population
- Need to develop both informant- based and direct assessments for adults with ID

Recommendations-1

- Educate families and Direct Support Professionals about the need to gather information about observed changes in cognitive, behavioral and functional status that would warrant referral to the individual with ID's PCP/HCP. The ***NTG-EDSD*** is one such administrative tool that can aid in shared decision-making and help families and staff have conversations with PCP/HCP about observed changes
- Educate health care professionals (medical and non-medical) about early onset dementia in adults with DS and about conditions that may obscure reasons for observed change so that they can knowledgeably advocate for their patients
- Provide training on detection of MCI and dementia among adults with ID for professionals who conduct dementia evaluations

Recommendations-2

- Develop a brief, in-office screen that can alert the PCP/HCP to the need for referral for formal work-up
- Develop relevant tools to gather collateral information and conduct direct assessment during formal batteries
- Develop consensus guidelines for what is to be included within a cognitive and functional assessment
- Identify assessment protocols that would aid differential diagnosis with consideration of unique challenges posed by diverse neurodevelopmental disorder to cognitive aging and function
- Identify protocol to assess individuals within the severe and profound range of intellectual disability