Diagnosing Dementia

MODULE 2

LIGHT
LAKE ERIE INTEGRATED GERIATRIC HEALTH TEAM

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Outline

- Defining dementia
- Early detection of dementia
- Recognizing and diagnosing dementia
- Diagnosing Alzheimer's Disease and Related Dementias (ADRD)
- ADRD

Learning Objectives

After completing this module, you will be able to:

- Discuss the benefits of early detection and diagnosis of dementia.
- Identify the differences between routine screening and assessing/diagnosing.
- Identify the most common types of dementia.
- List tests used in the diagnosis of dementia.
- List tools used in the diagnosis of dementia.

Key Take-Home Messages

- There are several brief validated tests that can detect dementia.
- Dementia is a group of symptoms and not a part of normal aging.
- Dementia is caused by many diseases and conditions affecting the brain.
  - The most common type of dementia is Alzheimer's disease, followed by vascular dementia, dementia with Lewy bodies, Parkinson's disease dementia, frontotemporal degeneration, and mixed dementia.
- Early diagnosis of dementia and its underlying causes allows for appropriate medical management, access to resources and clinical trials, and future planning with input from the persons living with dementia (PLwD).
- Use of biomarkers for Alzheimer's disease is an emerging field – brain amyloid PET scans are available with FDA-approved radioactive tracers.
- Early diagnosis allows individuals to prepare for years of productive and meaningful life ahead.

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What is Dementia?

- Dementia is an acquired syndrome and not part of normal aging (Dumas, 2015; Galvin, 2012; Gold, 2012; Robinson et al., 2015).
  - There are many causes and etiologies.
  - Once dementia is diagnosed, need to determine specific etiology.
- For a diagnosis of dementia, the 2011 revised National Institute on Aging (NIA) and Alzheimer's Association guidelines require evidence of impairments in at least 2 of the following domains that interfere with the ability to function at work or socially (McKhann et al., 2011):
  - The ability to acquire and recall new information
  - Reasoning and handling of complex tasks
  - Poor judgment (such as impairments in instrumental activities of daily living [IADL])
  - Visuospatial ability
  - Language function
  - Changes in personality, behavior, or comportment

What is Dementia (continued)

- Person Living with Dementia (PLwD) generally have memory impairment and one of the following (Wakefield et al., 2014; Wilson, et al., 2011; Cooper & Greene, 2005):
  - Aphasia: loss of ability to understand or express speech, caused by brain damage.
  - Apraxia: inability to perform particular purposive actions, as a result of brain damage.
  - Agnosia: inability to interpret sensations and hence to recognize things, typically as a result of brain damage.
  - Impaired executive function

APA DSM-5: Redefining Dementia as "Neurocognitive Disorders"

- The 2013 updated American Psychiatric Association's (APA's) Diagnostic and Statistical Manual of Mental Disorders, 5th Edition (DSM-5) made important changes, replacing the category of "delirium, dementia, amnestic and other cognitive disorders" (APA, 2013).
- The new category is neurocognitive disorders (NCD):
  - Mild neurocognitive disorders (mNCD)
  - Major neurocognitive disorders (MNCD)
    - Identifies different types including Alzheimer's disease (AD), vascular neurocognitive disorder (vascular dementia; VaD), Lewy body dementias (LBDs), frontotemporal neurocognitive disorder (or frontotemporal degeneration; FTD), Parkinson's disease (PD), or human immunodeficiency virus (HIV)

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Value of Early Detection and Diagnosis

- Diagnosis of dementia is life changing (McCarten & Borson, 2014).
- Early detection and diagnosis affords many benefits to PLwD and their care partners (Cordell et al., 2013; Johnson et al., 2013; McCarten & Borson, 2014):
  - Involves PLwD in decision-making
  - Can help preserve functioning
  - Allows optimization of other medical conditions
  - Allows for long-term care planning
  - Allows for development of interprofessional care team (Johnson et al., 2013)
- Need to balance benefits of routine screening of asymptomatic patients and early detection against costs of routine screening and early diagnosis (Boustani, 2013; McCarten, 2013)
- Currently, there is insufficient evidence as to the benefits or harms associated with routine screening for cognitive impairment in older adults (Moyer & USPSTF 2014).
- Medicare covers a free Annual Wellness Visit for every beneficiary.

USPSTF Recommendations: A Lack of Evidence to Support Universal Screening

2014 U.S. Preventive Services Task Force (USPSTF) recommendation:

- Current evidence is "insufficient to assess the balance of benefits and harms of *screening* for cognitive impairment" (USPSTF, 2014).
- Applies to universal screening with formal instruments
- Is currently undergoing reevaluation
- Does not apply to persons with signs or symptoms of dementia
  - Recognizes value of tools to increase detection in these persons (Cordell et al., 2013)
  - Notes that early detection does not improve decision-making or influence outcomes (Lin et al., 2013)

The Medicare Annual Wellness Visit: The Requirement to Assess for Cognitive Impairment

- A Medicare benefit from the Centers for Medicare & Medicaid Services (CMS) is the Annual Wellness Visit (AWV) (CMS, 2014; Cordell et al., 2013).
  - It includes a requirement to assess for cognitive impairment, regardless of the presence/absence of signs of dementia.
  - Older adults must complete a Health Risk Assessment (HRA), which could suggest cognitive impairment (e-CFR, 2016; Cordell et al., 2013).
- Providers can use direct observation, informant information, and objective tools (Cordell et al., 2013; NIA, 2014; AGS 2011).

The Benefits of Early Recognition of Cognitive Impairment: The IAGG and GARN Consensus Conference

- Recent expert consensus panel of the International Association of Gerontology and Geriatrics (IAGG) and its Global Aging Research Network (GARN) focused on improving brain health (Morley et al., 2015).
  - Identifies benefits of early recognition of cognitive impairment
  - Supports CMS in recommending use of Medicare AWV
  - Recommends validated screening tests that take 3–7 minutes to administer plus older adult and informant-based information
- Early cognitive impairment may have treatable components (Köbe et al., 2016; Moon 2016; Morley et al., 2015).

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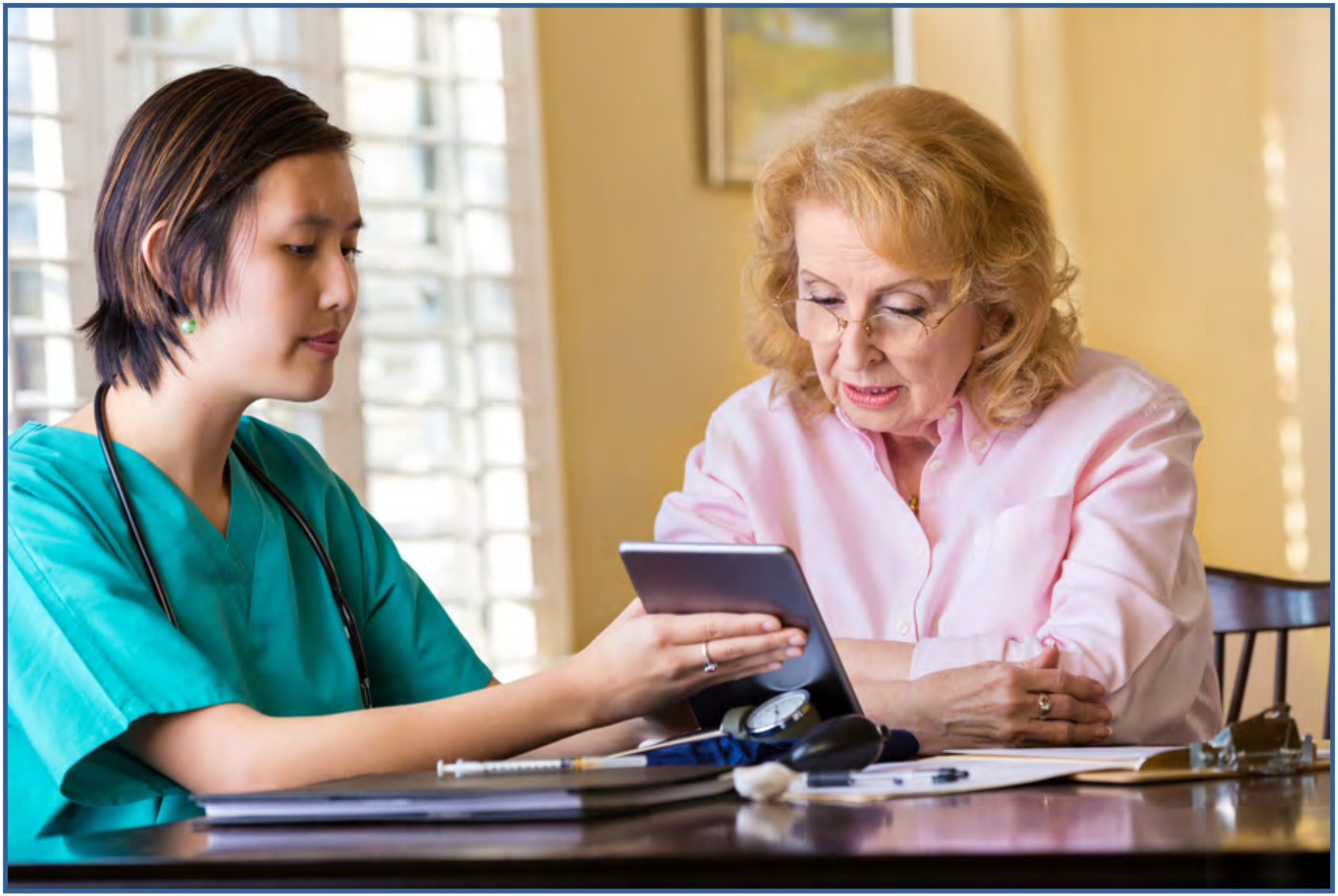
When to Consider Dementia in a Differential Diagnosis

- Dementia is an umbrella term encompassing many symptoms that together interfere with daily functioning (Mayo Clinic, n.d.).
- Dementia often is undetected in primary care setting (Cordell et al., 2013; Rabins et al., 2014).
- PLwD may not be aware of or raise issues regarding cognitive impairments (McCarten et al., 2014).
- Dementia should be considered part of differential diagnosis if (USPSTF 2014; Rabins & Blass, 2014):
  - Symptoms of memory difficulty interfere with daily functioning.
  - Unexplained functional decline or new onset psychiatric symptoms are evident.
  - Personal hygiene deteriorates.
  - There is sudden difficulty adhering to a medication regimen.



Describing the Process: From Detection of Cognitive Impairment or Dementia to Diagnosis in Primary Care Setting

- Primary care often first point of contact for PLwD (Robinson et al., 2015)
- Adults who manifest subjective signs and/or symptoms of cognitive impairment or dementia should undergo evaluation.
- Initial evaluation typically involves administration of a brief objective standardized instrument to detect dementia and determine if additional evaluation is warranted (Borson et al., 2006; Rabins & Blass, 2014).
  - Screening alone cannot diagnose dementia.
- Specialized screening necessary when presented by adult with Down syndrome or other intellectual disability (Moran et al., 2013)

Describing the Process: From Detection of Cognitive Impairment or Dementia to Diagnosis in Primary Care Setting (continued)

- Components of the diagnostic evaluation for dementia (Galvin & Sadowsky, 2012):
  - Observation for common warning signs
  - Medical and symptom history (from patient and informant)
  - Cognitive screening test
  - Standard medical tests to rule out reversible causes of dementia or coexisting disorders

- Additional tests as warranted

Detecting Cognitive Impairment in Primary Care Clinical Settings: Observation and Warning Signs

- Factors to observe in all older adult persons (Cooper & Greene, 2005; Galasko, 2013; Galvin & Sadowsky, 2012; Parmar et al., 2014):
  - Ability to interact appropriately with staff
  - Ability to maintain conversation
  - Appropriate clothing and personal hygiene
  - Excessive weight gain or loss over time
  - Changes in gait or mood over time
  - Frequently missed appointments
- Potential warning signs of dementia:
  - Impairments in memory or otherwise routine abilities over time
  - Impairments in ability to perform IADLs over time
  - Increasing reliance on others to make decisions over time

Detecting Cognitive Impairment in Primary Care Clinical Settings: Patient History

- Components of the older adult's history for dementia assessment (Cooper & Greene, 2005; Galasko, 2015; Galvin & Sadowsky, 2012; Olazarán et al., 2011; Parmar et al., 2014; Simmons et al., 2011)
- Symptom history:
  - Nature of symptoms
  - Onset and progression of symptoms
  - Consequence of symptoms with regard to functional abilities
- Medical and psychiatric history:
  - Personal
  - Family
- Current medications, vitamins, and herbal supplements

Detecting Cognitive Impairment in Primary Care Clinical Settings: Patient History (continued)

Other pertinent information can help a provider develop a differential diagnosis:

- Presence of hallucinations or delusions (Simmons et al., 2011)
- Parkinsonian symptoms, which include:
  - Tremor, or trembling in hands, arms, legs, jaw, and face;
  - Rigidity, or stiffness of the limbs and trunk;
  - Bradykinesia, or slowness of movement;
  - Postural instability, or impaired balance and coordination.
- Alcohol or drug use/misuse/abuse
- Sleep changes/Mood/Orientation changes
- Unsafe driving behaviors

Detecting Cognitive Impairment in the Primary Care Clinical Setting: The Importance of Informant or Care Partner Reports

- Importance of informant/care partner reports:
  - Memory impairments interfering with reporting by PLwD (Simmons et al., 2011)
  - PLwD denial (Hildreth & Church, 2015)
- Identifying appropriate informant (Simmons et al., 2011; Hildreth & Church, 2015)
- Obtaining information from informant:
  - Meeting alone with informant
  - Reviewing warning signs (Simmons et al., 2011)
  - Screening tools for informants (Harrison et al., 2014)
- Give respondent time to respond and be sensitive to their feelings of fear, sadness, loss, denial, etc., when asking questions.

Properties of Cognitive Tests Suitable for Primary Care Visits

- Many cognitive tests available
- Suitable tests for primary care (Cordell et al., 2013):
  - Require ≤5 minutes to administer.
  - Can be administered by medical staff members (not only physicians).
  - Are relatively free of biases.
  - Have good to excellent psychometric properties (validated in primary care setting).
  - Are free and readily available to be used in primary care setting
- With non-verbal persons or non-English speakers, may need different tests (Moran et al., 2013).
  - If intellectual disability present, informant interview can help.

Brief Cognitive Assessment Tools

- There are many commonly used brief assessment tools (Cordell et al., 2013; Morley et al., 2015; Simmons et al., 2011; Velayudhan et al., 2014).
  - Ascertain Dementia (AD8)
  - Mental Status Questionnaire (MSQ)
  - Mini-Cognitive Assessment Instrument (Mini-Cog)
  - Saint Louis University Mental Status (SLUS) exam
  - Rapid Cognitive Screen (RCS)
  - Short Blessed Test (SBT)
  - Short Test of Mental Status (STMS)
  - Short Portable Mental Status Questionnaire (SPMSQ)
  - Six Item Screener (SIS)

Brief Cognitive Assessment Tools (continued)

- Not all meet suitable criteria as defined by the AWV recommendations (Arevalo-Rodriguez et al., 2015; Barnes et al., 2014; Cooper & Greene, 2005; Creavin et al., 2016; Dudas et al., 2005; Fage et al., 2015; Malmstrom et al., 2015; Wilding et al., 2016)

- The American Academy of Family Physicians (AAFP) – searchable database of cognitive evaluation tools.

Comprehensive Evaluation If Dementia Is Suspected

- If dementia is suspected, the person should undergo a full evaluation.
  - Neurocognitive testing
  - Laboratory tests and imaging studies to rule out secondary causes (Simmons et al., 2011)
- In some cases, care partner/informant may be administered a screening test such as the Neuropsychiatric Inventory to provide information about PLwD (Lai, 2014).
- Typically, neurological and physical exams are normal unless focal deficits from stroke or Parkinsonism (Cooper & Greene, 2005)
- Persons with atypical presentations (such as Down syndrome or other intellectual disability) and/or are under the age of 65 require additional evaluation by a specialist.

Conducting Evaluation When Intellectual Disability Is Present

- Preliminary screening information can be obtained from family- or staff-administered screen instrument (for example, the NTG-EDSD) (Jokinen et al., 2013; British Psychological Society, 2015).
- Follow-up evaluations include comparing the PLwD to him or herself over time
- Request longitudinal data (screening forms and visual digital record of function) from family or staff
- Identify any extrinsic events that may be contributing to behavioral change, which may exclude dx of dementia
  - Check for drugs, reactive depression (changes in family, friends or staff), physical effects (pain, GI, hearing)
- Seek consult with clinician who is familiar with intellectual disability and aging-related neuropathologies

Recommended Tests for Developing a Differential Diagnosis

- Tests for developing differential medical or psychiatric diagnosis:
  - Laboratory work includes:
    - Typical panels (Rabins & Blass., 2014; AGS 2011)
    - Toxicology screen (Simmons et al., 2011)
  - Neuroimaging recommended if (Simmons et al.,2011):
    - Early onset
    - Abrupt onset with rapid cognitive decline
    - Focal neurologic symptoms
    - Predisposing conditions
  - Genetic tests are rarely required (Goldman, 2012).

The Role of Biomarkers in Diagnosis of Dementia

- Large body of evidence supports use of biomarkers in diagnosing dementia (Noel-Storr et al., 2013)
- Current biomarkers for AD in the research setting include cerebrospinal fluid (CSF) and brain amyloid-beta protein depositions through CSF or positron emission tomography (PET) amyloid imaging (Ferreira et al., 2014; Jack et al., 2008).
  - CSF is used routinely in Europe and for many patients in the United States.
  - There are currently 3 tracers with FDA approval for brain amyloid PET imaging: florbetapir, flutemetamol, and florbetaben.
- Combining the 2 biomarkers provides greatest sensitivity and specificity (Ferreira et al., 2014).
- No CSF biomarkers can consistently distinguish between the different dementias (Ewers et al., 2015; Ferreira et al., 2014).

Treatable Conditions Causing Cognitive Impairment

- Many treatable conditions can cause cognitive impairment (Downing et al., 2013; Galvin & Sandowsky, 2012; Hildreth & Church, 2015).
- 3D's of geriatric psychiatry: Dementia, delirium, depression (Downing et al., 2013)
- Others (Galvin & Sandowsky, 2012; Hildreth & Church, 2015):
  - Vitamin deficiencies
  - Endocrine disorders
  - Infections
  - Diseases
  - Drug/alcohol abuse
  - Sleep disorders
  - Brain tumors/lesions

Delirium: An Overview

- Delirium is an acute, temporary state of mental confusion, with abrupt onset (Cooper & Greene, 2005; Downing et al., 2013).
- Can occur at any time, and is frequently observed during a hospitalization for older patients (Francis & Young, 2016)
- Altered consciousness, visual delusions, and symptoms worsen at night (Cooper & Greene, 2005; Downing et al., 2013).
- Manifestations can include hyperactivity or hypoactivity or both (Downing et al., 2013).
- Delirium is a risk factor for dementia and can worsen underlying dementia (Davis et al., 2012).
- The Confusion Assessment Method (CAM) tool is used for diagnosis (Inouye et al., 1990).

Depression: An Overview

- Depression is not a normal consequence of aging; it needs to be identified early to prevent progression (Ellison et al., 2012).
- Symptoms are often atypical and include somatic complaints: weight change, sleep disturbances, and behavioral changes (Ellison et al., 2012; Cooper et al., 2005; Downing et al., 2013).
- Depression is associated with both delirium and dementia (Downing et al., 2013).
- Depression can impair cognition, with or without delirium or dementia.
- Depression is commonly observed in older persons in primary care practices (Lyness et al., 2009).
- Three tools are available to assess for depression in geriatric patient (Phillips, 2012).

Dementia, Depression, Delirium

Features	Dementia	Delirium	Depression
Memory problems	Yes (storage and recall)	Yes (storage and recall)	Yes (recall)
Onset	Gradual	Acute	Gradual
Mood disturbance	Possible	Possible	Yes
Disorientation	Possible	Yes	No
Sleep disturbance	Possible	Yes	Yes
Fluctuating symptoms throughout day	Yes	Yes	No
Progression	Gradual	Fast	Either
Somatic complaints	Possible	No	Yes
Apathy or anhedonia	Yes	Yes	Possible

Adapted from Downing et al., 2013.

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Dementia Due to Alzheimer's Disease (AD)

- Diagnostic criteria updated in 2011 by NIA in conjunction with AA based on increased understanding of the disease (Jack et al., 2011):
  - Recognition that AD begins much earlier than previously known
    - 3 stages of disease: preclinical, mild cognitive impairment (MCI), dementia due to AD (Jack et al., 2011)
    - Signs and symptoms of dementia appear only during last stage
  - Incorporate biomarkers into diagnosis of first 2 stages (Jack et al., 2011)
- Notes that not all persons living with MCI will progress to dementia

Diagnosing Mild Cognitive Impairment (MCI)

- Persons living with MCI have cognitive impairments not normal for their age but do not meet criteria for dementia (Albert & DeKorsky, 2011)
- Changes are assessed over time to monitor progression (Barnes et al., 2014).
- As with any cognitive impairment, rule out other brain diseases or systemic causes of impairments (Albert & DeKorsky., 2011).

Appropriate Use of Brain Amyloid PET Scans

- Brain beta-amyloid PET imaging can detect amyloid beta protein plaques, which are one of the defining pathological features of AD.
  - Brain amyloid plaques are associated with AD, but can also be seen with aging and other brain disorders.
  - A negative brain amyloid PET scan can rule out AD.
  - Three FDA-approved radioactive tracers are available for brain amyloid PET scans.
- Appropriate Use Criteria have been developed for patients in a clinical (versus research) setting (Johnson et al., 2013).

Diagnosing Vascular Dementia (VaD)

- Diagnosis of VaD not as clear-cut as for other forms of dementia
- Many similarities with AD
- Many conditions linked with VaD and vascular cognitive impairment (De Bruijn et al., 2015; Biessels et al., 2015; Sahathevan et al., 2011)
- Diagnosis involves similar procedures as with AD, with testing to determine underlying vascular etiology

Diagnosing Lewy Body Dementias (LBD)

- LBD syndromes include Dementia with Lewy Bodies (DLB) and Parkinson's Disease Dementia (PDD) (McKeith et al., 2005; Mrak & Griffin, 2007; Oliveira et al., 2015; Walker et al., 2015).
  - Both are aging-related dementias (Ajon Gealogo, 2013).
  - Major distinction between DLB and PDD is the temporal sequence of appearance of clinical symptoms (Mrak & Griffin, 2007)
    - DLB if dementia within 1 year after Parkinsonian symptoms
    - PDD if dementia years after PD diagnosed/Parkinsonian symptoms (McKeith et al., 2005; Mrak et al., 2007)

Distinguishing Between Lewy Body Dementias (LBD) and Alzheimer's Disease

- Distinguishing between LBD and AD
  - Memory impairment not prominent feature of early LBD (Mrak & Griffin, 2006; Huang & Halliday, 2013)
  - Similar manifestations between LBD and late-stage AD (Huang et al., 2013)
  - LBD has similar mean age of onset as AD (around age 68) but PD has earlier onset (Ajon Gealogo, 2013)
  - LBD has more rapid course of progression than AD or other dementias (Ajon Gealogo, 2013)

Diagnosing Frontotemporal Degeneration (FTD)

- Symptoms of FTD very different from AD (Cardarelli et al., 2010; Piguet et al., 2011)
- Persons with FTD likely to have:
  - Younger onset, rapid progression, more psychiatric symptomatology
  - Relatively preserved memory and visuospatial abilities
  - Greater apathy, disinhibition, impulsivity
  - Lacks insight into disease and consequences

Diagnosing Frontotemporal Degeneration (continued)

- Speech-related variants of FTD also have different manifestations versus AD (Cardarelli et al., 2010)
- Up to half of persons with behavioral variant FTD (bvFTD) are misdiagnosed with either major depression or another psychiatric disorder (Cardarelli et al., 2010; Cooper et al., 2013; Ducharme et al., 2015; Pose et al., 2013; Riedl et al., 2014)
- International consensus criteria for diagnosis of bvFTD were recently revised (Rascovsky et al., 2011).

Diagnosis of Early Onset Dementia (EOD)

- EOD considered if PLwD under age 65 at time of diagnosis
- Extensive list of conditions to consider in differential diagnosis
- Diagnosis thorough clinical assessment (Kuruppu & Matthews, 2013):
  - The older adults history with informant corroboration
  - Assessment of functional impairment
  - Temporal profile of mode of onset and progression of symptoms
  - Past medical and psychiatric history
- Use “dementia-plus” algorithm: Assessment plus neuroimaging and laboratory analyses (Kuruppu et al., 2013)
- PLwD who have Down syndrome usually manifest dementia in their early 50s (Prasher, 2005)

Evaluation

1. **Early detection and diagnosis of dementia:**
   a. Enables initiation of treatments to prevent progression
   b. Provides an opportunity for the person to make treatment decisions while he/she retains adequate cognitive functioning
   c. Indicates the person should immediately stop driving
   d. Suggests treatments for non-acute medical comorbidities can be stopped

2. **The USPSTF statement notes the evidence is "insufficient to assess the balance of benefits and harms of…":**
   a. Universal screening for cognitive impairment using formal instruments
   b. Assessing persons with symptoms of dementia
   c. Brief screening tools in the initial diagnosis of dementia
   d. Neurocognitive evaluation for persons with cognitive impairment

Evaluation (continued)

3. A person who develops dementia within a year of developing Parkinsonian symptoms is likely diagnosed with:

a. Parkinson disease dementia
b. Dementia with Lewy bodies
c. Alzheimer's disease
d. Mixed dementia

4. Which of the following tools is recommended to be administered to a person suspected of having dementia?

a. CAM
b. PHQ-9
c. Mini-Cog
d. Zarit Burden Interview

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