Dementia: How a diagnosis is made

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alzheimer's association

KNOW the 10 SIGNS

EARLY DETECTION MATTERS

http://www.alz.org/alzheimers_disease_10_signs_of_alzheimers.asp#signs

Objectives

- How do we diagnose dementia?
- Subtypes of dementia
- Case Studies

1. **Memory loss that disrupts daily life**
   - Forgetting important dates or events (appointments, birthdays, anniversaries)
   - Asking the same questions over and over
2. **Challenges in planning or solving problems**
   - Having trouble keeping track of monthly bills. They may have difficulty concentrating and take much longer to do things than they did before.
3. **Difficulty completing familiar tasks at home, at work or at leisure**
   - Having trouble driving to a familiar location
   - Having trouble making a familiar recipe
   - Can't remember the rules to a favorite game

WHY would a person be screened for dementia?

- Close contacts are usually the first to notice a decline in level of functioning and memory and prompt presentation to a healthcare provider
- Concerns by healthcare providers in a closely supervised environment (hospital, nursing home)

4. **Confusion with time or place**
   - Losing track of dates, seasons and the passage of time
   - May not remember where they are or how they got there
5. **Trouble understanding visual images and spatial relationships**
   - Many different things can cause visual problems with aging
   - Difficulty reading, judging distance and determining color or contrast (can cause difficulty with driving)

6. **New problems with words in speaking or writing**
   - Having trouble following or joining a conversation.
   - Word finding difficulties, calling objects by the wrong name (i.e. "watch" becomes a "hand clock")
7. **Misplacing things and losing the ability to retrace steps**
   - Frequently accompanied by accusing others of stealing

Learning and Memory

- Repetitive conversations
- Difficulty learning new information

8. **Decreased or poor judgment**
   - Falling victim to scams
   - Less attention to personal hygiene
9. **Withdrawal from work or social activities**
10. **Changes in mood and personality**
    - Suspicion, depressed, anxious
    - More apparent in situations when outside of their typical activities or "comfort zone"

Language

Official Criteria for Diagnosis of Dementia

- Dementia (also called major neurocognitive disorder) - evidence from the history and clinical assessment that indicates significant cognitive impairment in at least one of the following cognitive domains:
  - Learning and memory
  - Language
  - Executive function
  - Complex attention
  - Perceptual-motor function
  - Social cognition

American Psychiatric Association. Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM-5). American Psychiatric Association, Arlington, VA 2013.

Executive function

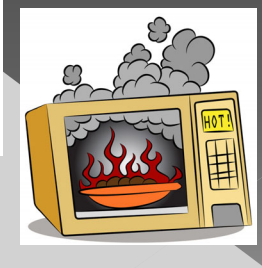
Complex attention

Official Criteria for Diagnosis of Dementia

- The impairment must be acquired and **represent a significant decline from a previous level of functioning**

American Psychiatric Association. Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM-5). American Psychiatric Association, Arlington, VA 2013.

Perceptual –motor function



Official Criteria for Diagnosis of Dementia

- The cognitive deficits **must interfere with independence in everyday activities**

American Psychiatric Association. Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM-5). American Psychiatric Association, Arlington, VA 2013.

Social Cognition

AD8 Dementia Screening Interview

Patient ID#: _____
CS ID#: _____
Date: _____

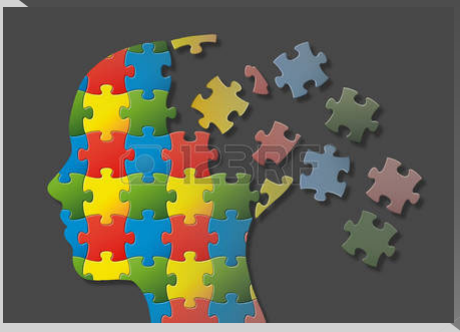
Remember, "Yes, a change" indicates that there has been a change in the last several years caused by cognitive (thinking and memory) problems.	YES, A change	NO, No change	N/A, Don't know
1. Problems with judgment (e.g., problems making decisions, bad financial decisions, problems with thinking)			
2. Less interest in hobbies/activities			
3. Repeats the same things over and over (questions, stories, or statements)			
4. Trouble learning how to use a tool, appliance, or gadget (e.g., VCR, computer, microwave, remote control)			
5. Forgets correct month or year			
6. Trouble handling complicated financial affairs (e.g., balancing checkbook, income taxes, paying bills)			
7. Trouble remembering appointments			
8. Daily problems with thinking and/or memory			
TOTAL AD8 SCORE			

Eight-item Informant Interview to Differentiate Aging and Dementia. 2005

Official Criteria for Diagnosis of Dementia

- The impairment must be acquired and **represent a significant decline from a previous level of functioning**
- The cognitive deficits **must interfere with independence in everyday activities**
- The impairments are not occurring exclusively during the course of delirium/are not better accounted for by another mental disorder (eg, major depressive disorder, schizophrenia)

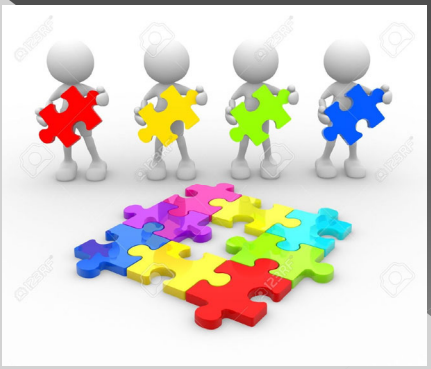
American Psychiatric Association. Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM-5). American Psychiatric Association, Arlington, VA 2013.



So, how do we make a diagnosis?

So, how do we make a diagnosis?

- **Careful history taking** (preferably from both the person affected, and people familiar with them)
  - Concerns from both patient and caregiver
  - How long have the symptoms been present and has there been progression?
  - Family history of dementia
  - Medications
  - Risk factor history
- **Physical exam**



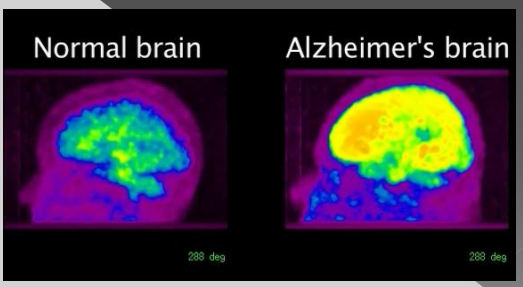
So, how do we make a diagnosis?

- Memory testing
  - Simple (10 minutes or less)
    - Mini-Cog
    - Mini Mental Status Exam (MMSE)
    - Montreal Cognitive Assessment (MoCA)
    - St. Louis University Mental Status exam (SLUMS)
  - Complex (45 minutes – 1 hour)
    - Neuropsychological testing

So, how do we make a diagnosis?

- Screening for depression
- Brain imaging
- Labs (thyroid hormone, B12 levels; in some cases testing for HIV and syphilis)

Amyloid PET imaging



http://coloradodementia.org/tag/pib-pet/

So, how do we make a diagnosis?

Screening for depression

- Cognitive dysfunction in depression is generally mild
  - More complaints by the patient of memory problems rather than objective findings on testing.
  - Apathy
  - "I don't know" vs making something up

Other testing

- Biomarkers
  - Levels of protein found in cerebrospinal fluid that can correlate with Alzheimer's pathology
  - Can be helpful as a tool in conjunction with clinical testing and imaging to make a diagnosis of Alzheimer's (not a stand alone test)
  - Limited primarily to investigational studies and clinical trials
  - Testing is not universally available nor paid for by insurance companies

Morris JC, et al. J Intern Med 2014

Other testing

- Amyloid PET scans
  - Currently not a part of the diagnostic process in patients who meet clinical criteria and are of the typical age for Alzheimer's
  - Testing is not universally available nor paid for by insurance companies

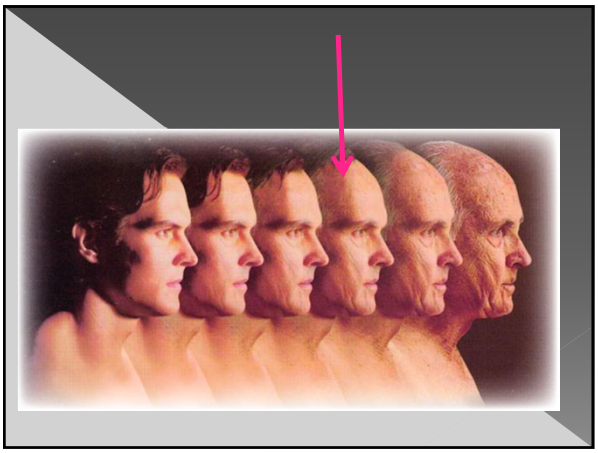
Morris JC, et al. J Intern Med 2014

Other testing

- Genetic testing
  - Not routinely recommended
  - Not perfect: Many false positives and false negatives
  - Can be helpful in people with younger onset of symptoms or a strong family history
  - Referral to a genetic counselor is recommended prior to testing

Subtypes of dementia

Why do we care?



Alzheimer's type dementia

- Most common form of dementia (~ 60 to 80 % of cases)
- Not common under the age of 60
  - Some forms of early onset, more likely related to genetic mutations
- Risk increases with rising age

Alzheimer's type dementia

- Executive dysfunction and visuospatial impairment are often present relatively early (sometimes along with the memory impairment)
  - The person may be less organized or less motivated; multitasking is often particularly compromised.
  - The person may underestimate their deficits and offer alibis or explanations for them when they are pointed out
    - This loss of insight becomes more severe over time and may impair safe decision making (i.e., continuing to drive when recommended not to)

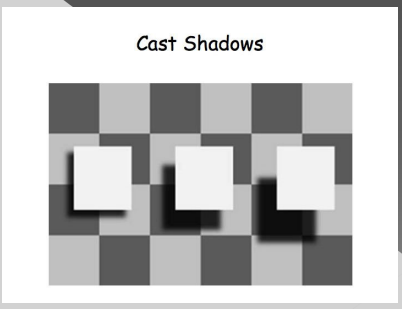
McKhann GM, et al. Alzheimers Dement 2011 .

Alzheimer's type dementia

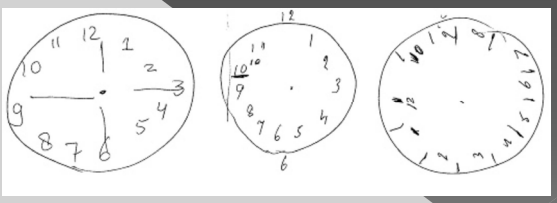
- **Memory impairment** is the most common initial symptom
  - Short term memory more affected (i.e. learning new information)
  - Long term memories are usual available to the person for many years after diagnosis

McKhann GM, et al. Alzheimers Dement 2011 .

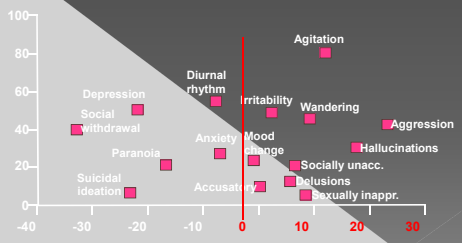
Visuospatial deficits



Clock Drawing Examples in AD

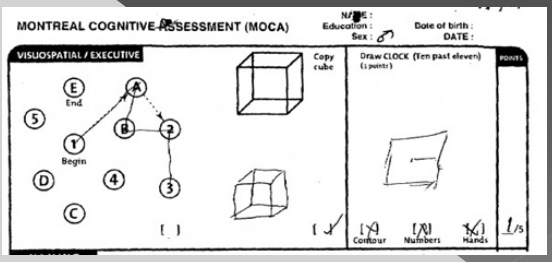


Peak Frequency of Behavioral Symptoms as Alzheimer's Disease Progresses

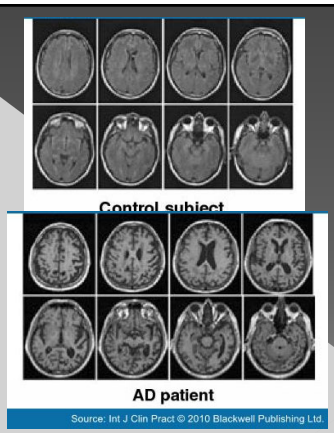


Jost BC, Grossberg GT. J Am Geriatr Soc. 1996;44:1078-1081

Moderate Stage AD



http://www.medscape.com/viewarticle/58145



Alzheimer's type dementia

- Language and behavioral symptoms often manifest later in the disease course

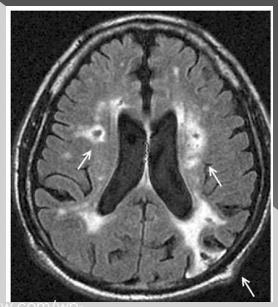
Progressive Disease

- Alzheimer's disease is irreversible, and progressive
- **Advanced care planning is extremely important**
  - Living will
  - Health care power of attorney
  - Financial power of attorney
  - Finances for caregiving as the disease progresses

Vascular Dementia

- 10 to 20 % of cases
- Heterogeneous syndrome rather than a distinct disorder, in which the underlying cause is cerebrovascular disease in some form and its ultimate manifestation is dementia.
- Associated with damage to the brain from strokes (not always clinically apparent)
- "Step-wise" progression

MRI in Vascular dementia



http://what-when-how.com/wp-content/uploads/2012/04/tmp5F39_thumb.jpg

Vascular dementia

- **Cortical syndrome** —cognitive deficits are specific to the areas affected

Staekenborg SS et al. Stroke. 2008

Vascular Dementia vs Alzheimer's Dementia

- Neuropsychological testing
  - Similar deficits on tests of language, construction, and memory registration
  - VaD patients have significantly less impairment on tests of recognition memory
  - VaD patients have more impairment on measures of executive functioning

Looi JC et al. Neurology. 1999

Vascular dementia

- **Subcortical syndrome** - deeper areas of brain affected, usually due to chronic decrease in blood flow
  - Focal motor signs
  - Early presence of gait disturbance
  - History of unsteadiness and frequent, unprovoked falls
  - Urinary symptoms
  - Emotional lability
  - Personality and mood changes, **apathy, depression**
  - Relatively mild memory deficit, psychomotor retardation, and abnormal executive function

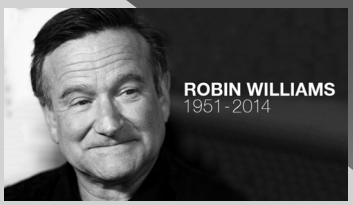
Staekenborg SS et al. Stroke. 2008

Prognosis

- Some patients with VaD can live for years with the same deficits, and never get worse, whereas others may continue to have ongoing progression
- Risk factor control
  - Smoking – STOP!
  - Alcohol use – limit or quit completely
  - Blood pressure, cholesterol, blood thinners – take medications as precribed

Dementia with Lewy Bodies

- Dementia with Lewy bodies (DLB) makes up 4 to 30 % of dementia cases



Vann Jones SA et al. Psychol Med. 2014

Dementia with Lewy Bodies

- **Early impairments in attention and executive and visuospatial function**
- Memory affected later in the course of the disease

Calderon J et al. J Neurol Neurosurg Psychiatry. 2001

Dementia with Lewy Bodies

- Fluctuating cognition
- **Recurrent well-formed, detailed visual hallucinations**
- Spontaneous features of parkinsonism (usually appearing around the time of hallucinations and dementia)
- REM sleep behavior disorder
- Severe sensitivity to anti-psychotics or similar medications

Prognosis

- DLB is irreversible, and progressive
- **Advanced care planning is extremely important**
  - Living will
  - Health care power of attorney
  - Financial power of attorney
  - Finances for caregiving as the disease progresses

Dementia with Lewy Bodies

Suggestive features

- Recurrent falls
- Syncope or transient loss of consciousness
- Severe autonomic dysfunction
- Hallucinations in other senses (hearing, taste, smell, touch)
- Delusions
- Depression

Dementia associated with Parkinson's disease

- 31% of Parkinson's disease (PD) patients diagnosed with dementia (1)
  - 5-6 times higher rate than in people without PD (2)
  - Risk increases with age (estimated to increase to 80-90% by age 90) (3)
- 3.6% of all cases of dementia (1)

(1) Aarsland D, et al. Mov Disord. 2005
(2) Aarsland D, et al. Neurology. 2001
(3) Buter TC, et al. . Neurology. 2008

Dementia associated with Parkinson's disease

- Early impairments in executive and visuospatial function
  - Tests of face recognition impaired early in the course of PD
- Memory deficits related to retrieval of learned information, which is improved by cuing.
- Aphasia, apraxia, and severe memory loss are usually not present

DLB vs PDD

- No differences in memory testing profiles
- PDD occurs in the setting of well-established parkinsonism (at least 1 year before onset of dementia) (1)
- DLB usually occurs before, at the same time, or very shortly after the development of parkinsonian signs.

(1) McKeith IG, et al. Neurology. 2005

Dementia associated with Parkinson's disease

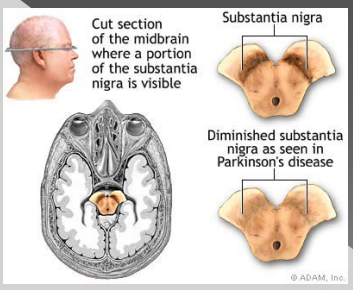
- Symptoms more common with progression of disease
- Visual hallucinations
- Paranoid delusions (can be worsened by Parkinsons' drugs)
- Depression and anxiety
- REM sleep behavior disorder

DLB vs PDD

- **DLB may have a faster clinical decline**, earlier onset of hallucinations and delusions, and are less likely to respond to parkinson's drugs
- Tremor is more common in PDD than DLB (1)
- Significant fluctuations in cognition are common in DLB, but rarely occur in PDD.

(1) Galasko D et al. Brain Cogn. 1996

Loss of neurons in substantia nigra in PD



http://www.nytimes.com/health/guides/disease/parkinsons-disease/print.html

Other dementia syndromes

- Frontotemporal dementia
- Alcoholic related dementia
- Chronic traumatic brain injury
- Prion disease
- HIV dementia

Important for ALL patients with dementia (and everyone else)

- Social connections, Intellectual Activity

Important for ALL patients with dementia (and everyone else)

- Healthy diet
- Exercise

Important for ALL patients with dementia (and everyone else)

- Risk factor control

Case studies

Mrs.S.
Mr. P.

Important for ALL patients with dementia (and everyone else)

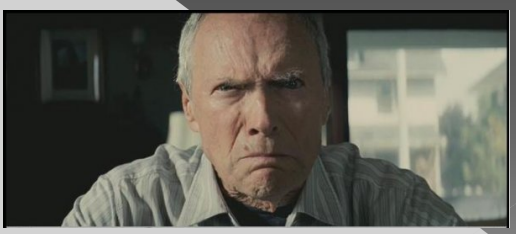
- Advanced care planning

Mrs. S.

Mrs. S.

- An 80 year old woman is brought to the office because she has hallucinations of children and small animals when she is alone in a room. These hallucinations sometimes agitate and disturb her.
- Her family notes she is having more trouble walking and has hand tremors when she sits quietly.

Mr. P



Mrs. S.

- She has a 9 months history of short term memory loss; problems with orientation that sometimes worsen dramatically
- Difficulty managing finances, preparing complex meals, following her "stories" on the television.

Mr. P.

- 83 year old male brought to office by family
- Has been "seeing" and talking to his dead wife and sister for the past 3 months; sometimes they respond back to him
- 3 year history of declining memory and impairment in ability to keep track of finances (family discovered many overdue and unpaid bills) and household tasks (a "hovel")

Mrs. S

- Physical exam:
  - Resting tremors in the hands
  - Rigidity with movement of the arms and wrists by the examiner
- Memory testing
  - Mini Mental Status Exam: 23/30 (mild impairment)

Mr. P

- Mr. P complains of sadness and missing his spouse who died 2 years ago
- Family has noticed that he has been irritable
- No history of alcohol or drug use
- Mr. P self discontinued his medications 2 years ago after spouse died ("what is the point?")
- Mr. P denies a history of falls; family does not know

Mr. P

Physical exam:

- Blood pressure elevated at 165/90
- Mild shuffling gait
- No tremors or other neurologic findings on exam

Memory testing: MMSE 17/30 (moderate impairment); Geriatric depression scale was 2/15 (>6 suggestive of depression)

Lab findings: normal

CT head: mild cortical atrophy

Resources

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Resources

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