Dementia  (or whatever you call it)

Robert W. Keefover, M.D. – Physician Director, BBHHF
What is it?

Merriam-Webster Dictionary: “DEMENTIA”

1. Mad, Insane

2. Suffering from or exhibiting cognitive dementia
What is it?

Merriam-Webster Dictionary: “DEMENTIA”

1. Progressive condition marked by deteriorated cognitive function

1. Insanity
What is it?

DSM-IV

“DEMENTIA”
What is it?

**DSM-III R: “DEMENTIA”**

*Acquired:*

1. Short- and long-term memory impairment +

2. Impairment in abstract thinking, judgment, other higher cortical function or personality change

3. Cognitive disturbance interferes significantly with work, social activities or relationships with others

4. These cognitive changes do not occur exclusively in the setting of delirium
**DSM-5:** Neurocognitive Disorder

**Definition:**

**Acquired:**

Cognitive impairment in domains such as:

- Memory (amnesia)
- Language (aphasia)
- Execution of purposeful movement (apraxia)
- Recognition/familiarity (agnosia)
- Visuospatial function (topographical disorientation)
- Self-control/management (executive function impairment)

**Mild NCD:** 1 or more **minor** impairments, independent

**Major NCD:** 1 or more **significant** impairments, independence lost
DSM-5: Why no dementia?

• Move away from “dementia’s” negative connotation

• Better distinguish disorders in which cognitive impairment is the primary feature

• More accurately reflect the diagnostic process:

  1. Explore symptoms
  2. Identify diagnostic syndrome(s)
  3. Find the cause
NCDs: How do they happen?

- Injury to specific locations in the brain
- Where injury occurs depends on underlying disease

Frontal
- Impulsivity
- Poor Judgment
- Inattention
- Abulia
- Expressive Aphasia

Parietal
- Receptive Aphasia
- Topographical Disorientation, Somatagnosia

Temporal
- Receptive Aphasia
- Amnesia
- Dyscalculia

Occipital
- Cortical Blindness
- Anton’s Syndrome
Lewy Body Disease
- New in DSM 5
- 2nd most common?
- Overlaps: AD & PD

Parkinson’s Disease
- 25% eventually develop dementia

Fronto-temporal Dementia
- Pick’s Disease in DSM-IV-TR

All Others
- Maybe as many as 100 other conditions can produce the syndrome of dementia
Amyloid Plaques

Neurofibrillary Tangles

- Abnormally formed protein produced in neurons
- Excessive production or impaired clearing leads to accumulation
- Cause or effect? (Diabetes Type III?)
- Toxic to nearby brain cells

What's the problem?

- Misshapen malfunctioning cellular transport tubes
- Twisting and kinking occur due to abnormal “Tau” protein
- No longer deliver nutrients and remove waste from distant parts of neuron
VaD: What’s the problem?

- Tiny vessels = small injury
- Affects deep brain areas
- Slight or no immediate signs or symptoms

- Larger vessel = Larger injury
- Mostly outer portions affected
- Immediate signs & symptoms
- Isolated cognitive impairment

- Gradual accumulation = gradual progression of multiple cognitive deficits
Lewy Body

- Initially in Substantia nigra
- Disrupts dopamine synthesis
- Gradual spread causes symptoms worsening

Substantia Nigra

- α-synuclein protein aggregates
- NCDs appear as Basal nucleus & cortex affected
- Dementia

PD: What’s the problem?

? Reason

Displaces normal cell structures
LBD: What's the problem?

Lewy Body

• Initially in limbic areas & cortex

• Parkinson areas involved

Diffuse Lewy Bodies

Alzheimer Plaques

• Initiated

• Parkinson areas involved

Cortical Lewy Bodies

Lewy Bodies
FTD: What’s the problem?

Pick Bodies

- Shrinking of brain tissue in areas controlling memory, emotions, and executive functions
- Pick’s Disease (Tau 3R predominates) in DSM-IV-R
- Slightly more common in women

Frontal & Temporal Lobe Atrophy

- Tangled tau protein aggregates
- The tau variant that predominates determines the form of FTD
- All are rare
- No known genetic component
**Other DSM Dementias:**

**Huntington’s Disease**
- Widespread viral-induced neurotoxins kill neurons
- Fungal lesions, tumors, and other masses also cause focal NCDs

**Prion Disease**
- Violent brain movement snaps neuron connections leading to “retraction balls”
- Longer frontal lobe-directed fibers most vulnerable

**Autosomal dominant mutation of Huntingtin gene (50% inheritance)**
- Degeneration of cells in basal ganglia striatum

**Creutzfeld-Jacob Disease in DSM-IV-TR is human form of Mad Cow Disease**
- Non-viral infectious agents cause “spongiform” injury
Other DSM Dementias:
Substance-Medication Induced: Alcohol

DSM-IV-TR

Alcohol Persisting Amnesia

No distinction

Alcohol-Induced Persisting Dementia

a.k.a. Korsakoff’s Dementia (thiamine deficiency)

a.k.a. Alcoholic Dementia (chronic alcohol toxicity)

Alcoholic

Control Subject

Amnestic-confabulatory Type

Non-amnestic Type

a.k.a. Korsakoff's Dementia (thiamine deficiency)

a.k.a. Alcoholic Dementia (chronic alcohol toxicity)
Clinical Features:

Depend on:

- Underlying Disease
  - Brain region(s) most affected
  - Diffuse v. focal injury
  - Rapidity of advancement

- Advancement Stage
  - Even global disease may be localized initially
Clinical Features:

**Symptom Patterns**

**Mixed Diffuse Deficits**
- Aphasias
- Amnestic (Cortical strokes)
- Lewy Body Disease
- Frontal Temporal Dementia
- Prion Disease
- HIV

**Isolated Difficits**
- Aphasias
- Amnesia
- Dyscalculia
- Agnosia
- Apraxia

**Mixed Diffuse Deficits**
- Aphasias + Visuospatial Impairment
- Amnesia + Dyscalculia
- Agnosia + Apraxia

**Isolated Deficits**
- Aphasias
- Amnesia
- Dyscalculia
- Agnosia
- Apraxia

- Alzheimer's Disease
- Multi-infarct (Cortical strokes)
- Lewy Body Disease
- Frontal Temporal Dementia
- Prion Disease
- HIV

- Vascular Dementia (large vessel)
- HIV-related tumors
- Alcohol-Induced (amnestic type)
- TBI (focal injury)
Clinical Features:

Symptom Patterns

- Impulsivity
- Inattentiveness
- Aggressiveness

Frontal Predominant

- Impulsive
- Inattentive
- Aggressive
- Social Imppropriety
- Low motivation
- Poor insight

- Vascular Dementia (small vessel)
- Frontal Temporal Dementia
- Parkinson’s Disease
- TBI
- Huntington’s Disease

Movement Abnormalities

- Bradykinesia
- Chorea
- Startles

- Tremors
- Myoclonus
- Seizures

- Vascular Dementia (small vessel)
- Parkinson’s Disease
- Lewy Body Disease
- TBI
- Huntington’s Disease
- Prion Disease

- Vascular Dementia (small vessel)
- Parkinson’s Disease
- Lewy Body Disease
- TBI
- Huntington’s Disease
- Prion Disease
### 1. Screening

- Forgets appointments, family events, etc.
- Trouble writing checks, paying bills
- Difficulties shopping independently
- Fails to follow medication instructions
- Gets lost walking or driving in familiar places

### 2. Assessing

- Conduct:
  - Standard Medical History
  - Physical Exam
  - Functional Status (FAQ)
  - Mental Status (MMSE, GDS)
  - Labs (CBC, electrolytes, Glucose, BUN – Creatinine, TSH, Drug levels)
  - Caregiver Interview (personal strain, patient behavior changes)

### 3. Diagnosing

- Consider referral:
  - Neurology
  - Psychiatry
  - Neuropsychology

### Family Questionnaire

<table>
<thead>
<tr>
<th>Problem</th>
<th>Caregiver Rating</th>
</tr>
</thead>
<tbody>
<tr>
<td>Repeated Questioning</td>
<td>None, Sometimes, Frequent, N/A</td>
</tr>
<tr>
<td>Forgets appointments, family events, etc.</td>
<td>None, Sometimes, Frequent, N/A</td>
</tr>
<tr>
<td>Trouble writing checks, paying bills</td>
<td>None, Sometimes, Frequent, N/A</td>
</tr>
<tr>
<td>Difficulties shopping independently</td>
<td>None, Sometimes, Frequent, N/A</td>
</tr>
<tr>
<td>Fails to follow medication instructions</td>
<td>None, Sometimes, Frequent, N/A</td>
</tr>
<tr>
<td>Gets lost walking or driving in familiar places</td>
<td>None, Sometimes, Frequent, N/A</td>
</tr>
</tbody>
</table>

Score of 4 or greater suggests need for further evaluation

Any Mental Health/Medical Person

- Consider referral:
  - Neurology
  - Psychiatry
  - Neuropsychology

Conduct:
- Standard Medical History
- Physical Exam
- Functional Status (FAQ)
- Mental Status (MMSE, GDS)
- Labs (CBC, electrolytes, Glucose, BUN – Creatinine, TSH, Drug levels)
- Caregiver Interview (personal strain, patient behavior changes)

Note if client/patient is:
- Odd or poor historian
- Disheveled, inappropriately dressed, dirty
- Repeatedly late for or misses appointments (e.g., wrong time/day)
- Has unexplained weight loss or vague symptoms
- Poorly adaptive to stress
- Defers to family/caregiver to answer questions directed to him/her
**Other Tools:**

**Alzheimer’s Association:**

“Tools for Early Identification, Assessment, and Treatment of People with Alzheimer’s Disease and Dementia”

alz.org/national/documents/brochure_toolsforidassesstreat.pdf
Questions/Comments: