Dementia:
It’s Not Always Alzheimer’s

A Caregiver’s Perspective
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Mid-America Institute on Aging and Wellness
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My Background
- Caregiver for my husband who had Lewy Body Dementia
- Difficult to get specific diagnosis
- Difficult for me to find information on what I should know, what I should do, and what I should expect
- Now volunteer with Alzheimer’s Association, Lewy Body Dementia Association, and the Lexington Area Parkinson’s Disease Support Group
- Hope to provide useful information to others dealing with the challenge of caring for a person with dementia

Purpose of this presentation
Provide information to familiarize you with some symptoms and treatments of dementias not caused by Alzheimer’s Disease
What is dementia?

Cognitive decline sufficient to interfere with daily living

Includes much more than just memory problems

- Impaired cognitive function
- Poor executive function
- Personality and behavior changes
- Hallucinations and delusions
- Visuo-spatial deficiencies
- Many others

What is dementia?

- Dementia is NOT synonymous with Alzheimer’s Disease
- Alzheimer’s Disease is the most common cause of dementia, but not the only one
- Other diseases can also result in dementia

Four most common causes of irreversible dementia (account for more than 90% of dementia)

- Alzheimer’s Disease (AD) is the most common cause

- Lewy Body Dementia (LBD; includes DLB and PDD)

- Vascular dementia (VaD) (strokes)

- Frontotemporal degeneration (FTD)
Other irreversible causes of dementia

• Huntington’s Disease (HD - rare)

• Creutzfeldt-Jakob disease (CJD - very rare – fast progression)

• Chronic Traumatic Encephalopathy (CTE – from head trauma)

Causes of dementia – potentially treatable

• Infections of the nervous system (such as meningitis)
• Chronic drug or alcohol use
• Depression
• Hydrocephalus
• Brain tumors

• Interactions of prescribed medications
• Metabolic disorders (such as a vitamin B12 deficiency)
• Underactive thyroid
• Low blood sugar
• PTSD

Why talk about “other” dementias?

• The more the caregiver and patient know about what’s going on the better they can plan and maximize quality of life
• The symptoms, treatment, and prognosis for these “other dementias” aren’t necessarily the same as those for Alzheimer’s
• Getting an accurate diagnosis can help improve treatment
• Can identify people who might participate in clinical trials
The “other dementias” are getting more attention

- At the national policy level
- By the medical community

National Alzheimer’s Project Act (NAPA)

- Law passed in 2011
- A National Plan to Address Alzheimer’s Disease was first developed in 2012; updated each year
- Among the five foundational goals of the plan are these two:
  - Expand Supports for People with Alzheimer’s Disease and their Families
  - Enhance Public Awareness and Engagement

Alzheimer’s Disease and AD-Related Dementias (AD-ADRD) Research Summits

- Organized in response to NAPA
- The 2013 Summit was the first time that a national research strategy was developed for LBD
- There were also recommendations for FTD, VaD, and mixed dementias
- The 2016 Summit included speakers from “non-governmental organizations” such as the nonprofit disease organizations and patient advocacy groups
  - This should increase the input from caregivers and patients
National Plan Annual Report 2016

Many recommendations in the report, including these:

• Expanding research and clinical trials specifically related to LBD, FTD, VaD

• Developing a standardized nomenclature
  ➢ This would help patients and caregivers
  ➢ Lack of standardized terms impairs communication with patients, caregivers, and decision makers
  ➢ Causes confusion when one term is used (e.g., AD) but a broader applicability is intended (e.g., all forms of dementia);
  ➢ Can result in frustration when families receive different diagnoses from different clinicians.

Caregiver Issues Getting More Attention Too

National Research Summit on Care, Services and Supports for Persons with Dementia and Their Caregivers will be held Oct 16-17, 2017 at NIH

RAISE Family Caregivers Act (S 1028) introduced in Senate

Increased attention to other dementias from the medical community

• Increase in published work about other dementias in the past few years
• One example: Journal of Geriatric Psychiatry and Neurology, September 2016
• Special issue devoted to non-Alzheimer dementias

“Together they (LBD/PDD, FTD, VaD) represent half of all dementia cases either alone or often in combination with each other or in combination with AD. Despite their high prevalence, these disorders often get little attention and patients are still often misdiagnosed.”

• The Lancet had several articles on non-Alzheimer dementias in October 2015

• “Clearly, underdiagnosis and misdiagnosis of non-Alzheimer's dementias is a key issue. More research is needed, not only for effective pharmacological interventions, but also for differential diagnostic techniques and effective care, to ensure the proper management of patients and to optimise their quality of life.”

www.thelancet.com Vol 386 October 24, 2015    Editorial

In Diagnostic and Statistical Manual of Mental Disorders (DSM)

• Used by mental health professionals for diagnosis
• Most recent version is DSM-5, published in 2013
• This version eliminated the term “dementia” and replaced it with “major neurocognitive disorder”
• It does not require memory impairment for a diagnosis, which it did before
• It recognizes that memory impairment is not always the first thing to be affected in a neurocognitive disorder
• Lewy Body Dementia specifically recognized for the first time

Diagnosing the Different Dementias

• Caregiver observations can help the clinician
• Physicians can perform a variety of tests to help with diagnosis
  – Clinical examinations and quick mental status tests
  – Diagnostic criteria exist for the dementias
  – Neuropsychological testing
  – Medical imaging techniques (CT, MRI, PET, SPECT)
  – Testing for biomarkers in body fluids
• Only examination of brain tissue after death can confirm diagnosis
Mixed Dementias

- Increasing recognition that many people, especially older ones, may have brain changes characteristic of more than one disease
- Complicates diagnosis and treatment

Lewy Body Dementia (LBD)
- Dementia with Lewy Bodies (DLB)
  If cognitive problems apparent before movement disorder usually diagnosed as DLB
- Parkinson’s Disease Dementia (PDD)
  If cognitive problems appear more than a year after movement disorder usually diagnosed as PDD

Cognitive / Behavioral Issues in LBD that may be observed by the caregiver in early stages

<table>
<thead>
<tr>
<th>LBD</th>
<th>Alzheimer’s Disease</th>
</tr>
</thead>
<tbody>
<tr>
<td>REM sleep disorder (acting out dreams) may occur many years before other symptoms</td>
<td>REM sleep disorder not usually present</td>
</tr>
<tr>
<td>Hallucinations and delusions prominent early in the disease</td>
<td>Hallucinations and delusions don’t occur until later stages, if at all</td>
</tr>
<tr>
<td>Fluctuating cognition - variations in attention and alertness (&quot;show time!&quot;)</td>
<td>Cognitive ability more stable</td>
</tr>
</tbody>
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<tr>
<td>Some features of parkinsonism (slow movement, resting tremor, or rigidity)</td>
<td>No symptoms of Parkinsonism</td>
</tr>
<tr>
<td>Significant deficits in visuo-spatial ability</td>
<td>Deficits in visuo-spatial ability typically less severe</td>
</tr>
<tr>
<td>Deficits in executive function are a common early symptom</td>
<td>Deficits in executive function not as noticeable as memory problems</td>
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<td>Severe autonomic dysfunction, e.g., constipation, orthostatic hypotension, urinary incontinence; hypersomnia; hyposmia</td>
<td>Significant autonomic dysfunction not apparent early</td>
</tr>
<tr>
<td>Deficits in attention prominent</td>
<td>Attention deficits less prominent</td>
</tr>
<tr>
<td>Memory relatively spared</td>
<td>Early symptoms include prominent memory problems</td>
</tr>
<tr>
<td>Mood changes (apathy, depression, anxiety)</td>
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Diagnosis and Treatment of LBD

- Dementia that may resemble Alzheimer’s
- Parkinsonism that resembles Parkinson’s Disease
- No drugs are specifically approved by FDA for LBD
- One drug approved in 2016 for PDD psychosis (Nuplazid)
- Two drugs now in clinical trials for LBD
Treatment of LBD

- Use medications designed for Alzheimer’s for cognitive problems
- Use medications for Parkinson's to treat motor problems
- BUT – often the drugs that improve cognition worsen movement disorder; drugs that improve movement disorder can worsen cognition
- LBD patients can react very badly to the traditional antipsychotic (neuroleptic) drugs

Frontotemporal Degeneration (FTD)

- Group of rare diseases that involve shrinkage of specific areas of the brain that regulate behavior, personality, and language
- Recognition of this as distinct from AD began in the 1980's
- Behavioral variants are the most common (bvFTD)
- Language variants (several types)
- Sometimes occurs with motor neuron disease

FTD is a complicated disease with several variants
Cognitive / Behavioral Issues in FTD that may be observed by the caregiver in early stages

<table>
<thead>
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<th>FTD</th>
<th>Alzheimer’s Disease</th>
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<tr>
<td>Typically begins from age 40–60</td>
<td>Usually seen in people over 65</td>
</tr>
<tr>
<td>Inappropriate or bizarre social behavior; may lead to criminal behavior</td>
<td>Typically retain appropriate social behavior in early–mid stages</td>
</tr>
<tr>
<td>Often exhibit lack of concern for social norms or other people, lack of insight into their own behaviors</td>
<td></td>
</tr>
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<td>Retain important features of memory (keeping track of day-to-day events, orientation to space and time, etc)</td>
<td>Memory loss is an early indicator; worsens as disease progresses</td>
</tr>
<tr>
<td>Language variant FTD displays serious difficulties with names and words</td>
<td>Milder problems with recall of names and words in early stages</td>
</tr>
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Cognitive / Behavioral Issues in FTD that may be observed by the caregiver in early stages

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<tr>
<td>Compulsive behavior (pacing, pointless repetitive activity)</td>
<td>Compulsive behavior not commonly observed</td>
</tr>
<tr>
<td>Hyperorality (binge eating, change in food preference, consumption of inedible objects)</td>
<td>Hyperorality not a common symptom</td>
</tr>
<tr>
<td>Visuo-spatial ability spared</td>
<td>Visuo-spatial deficits commonly observed</td>
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<td>May display early motor problems</td>
<td>Motor problems mild or non-existent</td>
</tr>
<tr>
<td>such as rigidity, tremor, difficulty</td>
<td></td>
</tr>
<tr>
<td>walking</td>
<td></td>
</tr>
<tr>
<td>Executive function impaired</td>
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Treatments

- No FDA-approved drugs specifically for FTD
- Existing drugs can be used to address symptoms and contribute to patient and caregiver quality of life
- Anti-depressant drugs can improve some behavior symptoms
- Cholinesterase inhibitors used for AD [donepezil, rivastigmine, galantamine] may help cognitive symptoms
- Memantine not shown to help FTD patients
- Psychostimulants and antipsychotics drugs should be used with caution
- Movement disorder aspect often doesn’t respond well to Parkinson’s drugs (l-dopa)

Vascular Dementia
Vascular Dementia

- Blood vessels in the brain may become clogged or sometimes they can burst
- The resulting lack of blood flow can kill the cells
  This area of dead tissue is called an “infarct”
- Effects of these infarcts depend on what part of the brain is affected

Vascular Dementia: fast or slow

Fast:
- When the blood flow to the brain is interrupted due to a major blockage or rupture we call it a stroke
- Cognitive problems after a stroke are sometimes called post-stroke dementia

Slow:
- But - small blockages and ruptures also can occur over a long period of time in many small blood vessels
- This is harder to diagnose as a cause of dementia

Vascular Dementia Effects: Variable

- AD, LBD, FTD have definite brain changes in specific areas of the brain
- But any part of the brain could be affected by clots or ruptures
- Effects of stroke or slow damage to blood vessels are unpredictable
- MRI and CT scans can give some information about vascular damage (but they don’t give direct evidence of AD, LBD, FTD lesions)
What a caregiver might observe in VaD

• Trouble paying attention and concentrating
• Reduced ability to organize thoughts or actions
• Decline in ability to analyze a situation, develop an effective plan and communicate that plan to others
• Difficulty deciding what to do next
• Problems with memory
• Restlessness and agitation
• Unsteady gait
• Sudden or frequent urge to urinate or inability to control passing urine
• Depression

Treatments

• No treatment can repair the effects of vascular dementia

• Treatment aimed at preventing future events by controlling cardiovascular risk factors
  - High blood pressure
  - Cholesterol level
  - Diabetes
  - Smoking

• Drugs effective for AD (cholinesterase inhibitors and memantine) don’t benefit most people with VaD

Where to Find Information and Help
Alzheimer’s Association  (alz.org)

- Web site has LOTS of information for both caregiver and patient
- There is a 24-hour help line (800 272 3900)
- Local offices provide community education

Lewy Body Dementia Association (LBDA)  (www.lbda.org)

- LBD Caregiver Link
  - Toll-free: 800-539-9767
  - Email: support@lbda.org
- LBD Support Groups
- LBDA Forums
  - To join, email: forum@lbda.org
- LBD Awareness Movement
  - October of each year

Association for Frontotemporal Degeneration (AFTD)  
http://www.theaftd.org/

- Information about FTD
- Helpline: 866-507-7222
- Email: info@theaftd.org
Vascular Dementia

No specific organization related only to VaD

Information about VaD found on websites related to stroke and heart disease

- http://memory.ucsf.edu/education/diseases/vascular
- http://www.strokeassociation.org/STROKEORG/

Selected References
(all publicly available online)

- Report of the 2016 ADRD Summit
- National Plan to Address Alzheimer’s Disease - 2016 Update
  https://aspe.hhs.gov/national-plan-address-alzheimer’s-disease-2016-update
- National Research Summit on Care, Services and Supports for Persons with Dementia and Their Caregivers
  http://journals.sagepub.com/doi/pdf/10.1177/0891988716654982
- Web site for the Lewy Body Dementia Association
  www.lbda.org
- A 2016 report on the state of the science for LBD
- National Parkinson’s Foundation “mind guides” to cognition, mood, and hallucosis in PD
- The Association for Frontotemporal Degeneration
  http://www.theaftd.org/
- Links to resources for information about frontotemporal degeneration from the NIA
  https://www.nia.nih.gov/health/frontotemporal-disorders-resource-list
- Information on vascular dementia from the American Heart Assn and American Stroke Assn
  http://strokeconnection.strokeassociation.org/Summer-2016/Understanding-Vascular-Dementia/