Top Tips for Maximizing Treatment and Quality of Life in Lewy-Body Dementia, Frontotemporal Dementia, and Vascular Dementia

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“Top Tips” Toolbox

LBD Tools

FTD Tools

General Tools

VD Tools
Lewy Body Dementia (LBD)

- Named for abnormal protein deposits within neurons (alpha-synuclein)
  - Alpha-synuclein = abundant in normal brain; unfolded
  - “Lewy Body” = abnormal folding, located WITHIN the neurons
α-Synucleinopathies

- The LOCATION and type of cell impacted by α-synucleinopathy leads to different symptoms
- Primary α-Synucleinopathies
  - Parkinson’s
  - LBD
  - Multiple Systems Atrophy
- Secondary α-Synucleinopathies
  - May be present in AD
Association Cortex

- Broca's Area
- Wernicke's Area

Primary Areas:
- Auditory
- Motor
- Touch
- Visual

Assoc.

Higher Order Association
Lewy Body Dementia Case Example

- JB is a 75-year-old married man with behavioral changes, including irritability, anxiety, and aggressiveness. About two years ago, he began to report visual hallucinations (seeing small animals and people in his home). He also began to fall frequently and began acting out his dreams. His wife wonders why sometimes his memory is "sharp" and sometimes he is very confused and does not recognize familiar locations.

Adapted from https://www.jscimedcentral.com/NeurodegenerativeDisorders/neurodegenerativedisorders-1-1003.pdf
Lewy Body Dementia - Primary Symptoms

- Consensus criteria include:
  - Presence of dementia (gradual onset, progressive course)
  - At least two of the following
    - Parkinsonism
    - Visual hallucinations
    - Fluctuations in consciousness ("switch-like")
- LBD = 10-30% of all dementias (2nd or 3rd most common after Alzheimer’s, possibly vascular); 1.3 million Americans
Lewy Body Dementia - Secondary symptoms

- Other clinical features may also be supportive of diagnosis
  - Hallucinations in other sensory modalities
  - Repeated falls
  - General visual problems, illusions
  - Syncope
  - Delusions
  - Increased sensitivity to classic antipsychotic medication
  - REM sleep disturbance (occurs in 72%)
LBD symptom pattern

Fluctuations

V
H

Parkinsonism

6 18 13

35 18

7 2

Hallucinations

- Vivid, colorful, complex
  - Case examples
- Often people and animals
- May be free-standing or embedded in a visual scene
- May occur more around awakening, sleep
- May be miniature/Lilliputian
- Often involve delusions as person tries to make sense of what they are seeing
- Person may attempt to interact with them
- Possibly linked to Lewy Bodies in the right temporal lobe
Assessing Visual Hallucinations

- Management recommendations differ depending on assessment findings
  - What is the content of the hallucination, and how frequently does it occur?
  - Is there any pattern to the occurrence (time of day, specific room, etc)?
  - Is the person bothered by the hallucinations?
  - Does the person have intact reality testing?
Tools to treat Visual Hallucinations - Part 1

- **Education**
  - Pt education depends on reality testing
    - If intact = movie analogy
    - If not intact = distraction, “balanced validation,” manage distress
  - Family education about the reason for visual hallucinations

- **Other tools**
  - Ensure corrected vision
  - Experiment with more light vs. no light; night lights
Tools to treat Visual Hallucinations - Part 2

- Medication
  - Acetylcholinesterase inhibitors (memory medication) may be helpful

- Antipsychotic meds are rarely used
Treatment for cognitive symptoms

- Memory medication
  - Rivastigmine (Exelon) has greatest evidence base
  - Consider avoiding nighttime doses if increase in vivid dreams (possible medication side effect)
  - Memantine (Namenda) shown helpful in 1 of 4 studies
Treatment for psychiatric symptoms

- Anxiety and depression may predate cognitive and motor symptoms by decades

- Depression
  - May be helped by Exelon
  - SSRIs = mixed results
  - Electroconvulsive therapy and transcranial magnetic stimulation have been shown effective, but are rarely used

- Anxiety
  - Anxiolytic medication may help
Treatment for motor symptoms

- Physical therapy
- Home safety modification (especially fall prevention)
- Medications used in PD (e.g. Levodopa/Carbidopa) are usually less effective in LBD, and could increase neuropsychiatric symptoms, but may be used
Treatment of sleep problems

- REM Sleep Disorder
  - Behavioral strategies
  - Melatonin
  - Clonazepam

- Excessive daytime sleepiness
  - Decrease sedating meds
  - Rule out primary sleep disorders
  - Caffeine may help
  - Mixed evidence for Ritalin
Frontotemporal Dementia
Three subtypes of Frontotemporal Degeneration

- Progressive behavior/personality change: changes in personality, judgment, behavior, emotions

- Progressive language decline: changes in speaking, comprehending, reading, and writing

- Progressive motor decline - difficulties with physical movement such as shaking, difficulty walking, frequent falls, poor coordination
Frontotemporal Diagnoses

- Progressive behavior/personality decline
  - Behavioral variant FTD (bvFTD)
  - Temporal/frontal variant FTD (tvFTD, fvFTD)
  - Pick’s disease
- Progressive language decline
  - Primary progressive aphasia
- Progressive motor decline
  - Corticobasal syndrome (CBS)
  - Progressive supranuclear palsy (PSP)
  - FTD with parkinsonism
  - FTD with amyotrophic lateral sclerosis (FTD-ALS)
Frontotemporal Dementia (FTD)

- Dementia that begins in the frontal lobe, temporal lobe, or both
FTD Overview

- Progressive decline in behavior and personality and/or language, resulting in significant impairment in social and daily activities
- 10-20% of all dementias
- Average age of onset is 57
- Life expectancy = 7-13 years (ranges from 2-20)
- Causes: Unknown in most cases, though some studies show abnormal protein deposits and genetic risk factors
- Often initially misdiagnosed
- No treatments to slow or stop progression, but some treatments help manage symptoms; clinical trials underway

(http://www.theaftd.org/frontotemporal-degeneration/ftd-fast-facts)
Behavioral Variant FTD (BvFTD)

- The most common FTD
- Changes in behavior, personality, judgment
  - Unusual behavior that is out of context for the social situation
  - Person may not know or care that behavior is unusual (loss of insight)
  - Reduced empathy, likely due to impaired ability to recognize facial or vocal emotions
  - Hyperorality (excessive eating, often of sweet foods)
  - Disinhibition
  - Impulsivity
  - Lack of initiation
  - Social withdrawal
- Cognitive domains impacted: executive functioning/EF (planning, mental flexibility, speeded strategic tasks including verbal and nonverbal fluency); memory initially stronger than EF
A 51-year-old male real estate lawyer began to embezzle money at work, regularly listing mysterious expenses on his travel reimbursement forms, which turned out to be purchases of pornographic materials via the internet. At about the time this behavior was discovered by his partners, a few of the female law clerks complained that he often made inappropriate comments about their physique and that he stared at them in a way that made them uncomfortable. His work had dramatically deteriorated, and rather than working with his clients, he spent most of the day at work shuffling papers, reading magazines or downloading pornography onto his computer. He was eventually asked to leave the firm, but made no attempts to find a new job. His wife and children reported that over the past year he lost interest in them and watched television without speaking when at home. He developed a strong desire for potato chips and gained 15 lbs. His manners deteriorated, and he stuffed his mouth, often choking at the dinner table. He insisted on eating food on his plate in a specific order, often with his hands.

http://memory.ucsf.edu/ftd/medical/cases/case1/multiple
FTD Behavioral Management Strategies

- Establish a daily routine; take part in it too!
- Redirection
- Environmental modification is vital (add, take away things)
- Break tasks down; “let’s do this” rather than “don’t do”; restrict disinhibited behavior;
- Family: Depersonalize symptoms
- Use behavioral interventions; start with ABC analysis
- Medications may be helpful with depression, agitation
- Caution: some memory meds (acetylcholinesterase inhibitors) can worsen symptoms
PPA Subtypes

- Non-fluent variant PPA
- Semantic variant PPA
- Logopenic variant PPA
3 Subtypes of Language - related FTD/Primary Progressive Aphasia (PPA)

- **Agrammatic PPA** (Progressive nonfluent aphasia) - effortful speech, distorted sounds; omitting words that link nouns and verbs (such as to, from, the); difficulty swallowing
  - May progress to generalized motor problems including CBD or PSP

- **Semantic PPA** ("semantic dementia") - word-finding difficulty, single-word comprehension deficits; repetition and motor speech intact
  - Tau-positive pathology

- **Logopenic PPA** - word finding problems, hesitations/pauses while speaking, intact single-word comprehension; impaired repetition
  - Alzheimer’s pathology may underlie
PPA Case Example

- A 62-year-old female retired executive began having difficulty finding words. She slowly began to lose her ability to express ideas. She became quieter and somewhat socially withdrawn. She also started to have trouble writing. When talking, she took a long time to express her ideas and communicated agrammatically with nouns. Others told her that she had trouble "spitting out her words." Social graces remained preserved, although she expressed profound frustration regarding her speech, and she developed major depression.

http://memory.ucsf.edu/ftd/medical/cases
PPA management strategies

- Speech therapy
- Augmentative communication devices
- Complete advanced directives and other important paperwork early

Behavioral communication strategies

- Speak in short sentences
- Provide multiple choice responses
- Determine whether spelling and math are impacted, and if so, minimize tasks that require it
FTD Resources and Support

- The Association for Frontotemporal Dementias
  http://www.theaftd.org/frontotemporal-degeneration/disorders
  - Monthly support group at Alzheimer’s Association in Milwaukee - third Wed of every month 6-7:30 pm

- Alzheimer’s Association:
  http://www.alz.org/dementia/fronto-temporal-dementia-

- Frontotemporal Dementia Caregiver Support Center
  http://ftdsupport.com/
Vascular Dementia
Blood provides oxygen and nutrients to the brain

- **BLOOD SUPPLY**
  - Every time your heart beats, about 25% of the blood goes to your brain!
  - Blood flow through whole brain (adult) = 750-1000 ml/min

http://faculty.washington.edu/chudler/facts.html
Blood vessels are extensive
Arterial territories

http://instruct.uwo.ca/anatomy/530/530notes.htm
Vascular Dementia (VD)

- A decline in cognitive skills when blood flow is blocked or reduced due to:
  - Stroke
  - Diabetes
  - High blood pressure
  - High cholesterol
  - Anoxia (due to surgery, injury, etc)

- Many prefer the term "vascular cognitive impairment (VCI)" because it expresses the continuum of cognitive changes that can occur

- Vascular compromise causes up to 30% of all dementias
A 65-year-old man presents with difficulty in decision-making and planning, which is of abrupt onset and occurs 3 months after a stroke. He has strong vascular risk factors, including smoking. Over time, there has been a fluctuating stepwise reduction in cognitive function. There is a history of nocturnal confusion and incontinence. Neuro-imaging indicates a probable vascular etiology with white matter changes and infarction.

https://online.epocrates.com/diseases/31922/Vascular-dementia/Common-Vignette
Symptoms depend on cause

- Stroke symptoms may include confusion, trouble speaking or understanding speech, vision loss, motor paralysis/weakness
- VD due to chronic diseases (DM, HTN, high cholesterol) often present with motoric and cognitive slowing, difficulty recalling info (improved with cueing)
- Vascular compromise is a risk factor for the development of AD
VD Management Strategies

- Vascular treatment to reduce impact of further damage
  - If stroke, medical workup to determine etiology
  - Improve management of chronic vascular conditions (med compliance, lifestyle factors, attend appts)
  - Improve diet and exercise under direction of medical professional
- Memory medication may be more helpful in microvascular subtype
- Cognitive tools: encode info with verbal and visual cues, link info, allow extra time, don’t multitask
- Consider antidepressant medication and psychotherapy to treat depression
“Top Tips” Toolbox

- LBD Tools
- General Tools
- FTD Tools
- VD Tools
GENERAL TOOLS to enhance mood and minimize distress in most dementias

- All can be helpful proactively and reactively
  - Routine
  - Distraction
  - Tailored calming strategies
    - Music, family time, reading, praying, television, pictures - customization is key
    - Tactile tools (touch, aromatherapy, objects, music, pets, walking, games)
Questions?