Understanding and Living with Lewy Body Dementia

Tracy Sherman MD
The ride on the Lewy Body Roller coaster ended. The seat belt that had us held prisoner was removed and we exited the car.

Living with a Thief named Lewy Body Dementia Blog - Kathy Lawrey
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What is Lewy Body Disease?

What are the symptoms and why do they occur?

What are the current treatments available?

How can we best live with Lewy Body Dementia

What does the future hold?
Lewy What?
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86 billion neurons in the brain

Every neuron communicates with up to 10,000 other neurons via neurotransmitters, degenerative dementias cause disruption of neurotransmitters.

Neurons do not replicate (decrease with age)

Are the cells most disrupted and are the cause of symptoms of degenerative dementias
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Alzheimer’s disease          Frontal Temporal Dementia          Lewy Body Dementia

Plaques  Neurofibrillary Tangles
First discovered by Dr. F H Lewy in 1912 with studying with Dr. Alois Alzheimer

Made up of protein known as alpha synuclein

Also the abnormality found in Parkinson’s disease
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• Understanding the Term “Lewy Body Disease”

• Dementia with Lewy Bodies (what we refer to as Lewy Body Dementia) – second most common form of dementia

• Parkinson’s disease
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Timeline for Lewy Body Dementia

?RBD  MCI  DLB
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Criteria for diagnosing probable Lewy Body Dementia

Dementia
Core symptoms
  fluctuating attention and concentration
  recurrent, well formed visual hallucinations
  spontaneous signs of parkinsonism
(? REM sleep behavior disorder?)
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Suggestive symptoms of Lewy Body Dementia

*REM sleep behavioral disorder

*Sensitivity to neuroleptic medications

Delusions, Illusions (misidentification) and other forms of Hallucinations

Autonomic Dysfunction

Frequent Falls
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**Lewy Body Dementia: SPECT and PET**

FDG PET demonstrating decreased uptake in the occipital lobes in addition to posterior temporoparietal lobes, bilaterally [arrows]


Baystate Medical Center
Education & Research

The Western Campus of Tufts University
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DaTSCAN image - transverse slice through the striatum

**AD**
If dopamine transmission is normal, DaTSCAN will be distributed in a ‘comma’ shaped striatum

**DLB**
Loss of dopamine neurons in DLB reduces DaTSCAN uptake, resulting in a ‘full stop’ shape
# Understanding and Living with Lewy Body Dementia

## Table 1

Lewy body composite risk score

<table>
<thead>
<tr>
<th>Does the patient...</th>
<th>Yes</th>
<th>No</th>
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<tbody>
<tr>
<td>Have slowness in initiating and maintaining movement or have frequent hesitations or pauses during movement?</td>
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<tr>
<td>Have rigidity (with or without cogwheeling) on passive range of motion in any of the 4 extremities?</td>
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<td>Have a loss of postural stability (balance) with or without frequent falls?</td>
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<tr>
<td>Have a tremor at rest in any of the 4 extremities or head?</td>
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<td>Have excessive daytime sleepiness and/or seem drowsy and lethargic when awake?</td>
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<td>Have episodes of illogical thinking or incoherent, random thoughts?</td>
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<tr>
<td>Have frequent staring spells or periods of blank looks?</td>
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<tr>
<td>Have visual hallucinations (see things not really there)?</td>
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<tr>
<td>Appear to act out his/her dreams (kick, punch, thrash, shout or scream)?</td>
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<tr>
<td>Have orthostatic hypotension or other signs of autonomic insufficiency?</td>
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</tr>
</tbody>
</table>

Total score

Copyright 2013 *The Lewy Body Composite Risk Score* James E. Galvin, MD, MPH.
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Symptoms can be broken down into different categories:

- Cognitive
- Neuropsychiatric
- Motor
- Sleep
- Autonomic
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Cognitive symptoms, deficits in:
- executive function (planning, organizing)
- speed of thinking
- spatial orientation
- visual identification
- attention and concentration
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Cognitive symptoms:
related to deficits in acetylcholine and dopamine

Worsened by medications that block these chemicals

- acetylcholine- antihistamines (Benadryl), bladder medications (Oxybutynin), muscle relaxants (Cyclobenzaprine), tricyclic antidepressants (Amitriptyline)
- Dopamine- antipsychotics (Haldol), antiemetics (metoclopramide), melatonin
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Medications that can improve cognition:

- cholinesterase inhibitors (donepezil, rivastigmine, galantamine)
- Sinemet (carbidopa-levodopa)
- Memantine
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Neuropsychiatric symptoms:
- hallucinations
- illusions (misidentification)
- delusions
- Capgras syndrome (misidentification of spouse, primary care giver or home)
- Depression
- Anxiety
- Agitation/Aggressive behaviors
Neuropsychiatric symptoms associated with decrease in acetylcholine and serotonin (5HT3) and increase in dopamine and epinephrine

Worsened by medications that effect these neurotransmitters

anticholinergics (see previous list)
5HT3 blockers- antiemetics such as ondansetron
DA agonists- Parkinson’s medications
Epinephrine agonists- stimulants
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Treatment of neuropsychiatric symptoms

Cholinesterase inhibitors

Antidepressants (SSRI, NSRI, mirtazapine, bupropion)

Atypical Antipsychotics (***black box warning) most often used is quetiapine, probably most effective is clozapine (but higher risk of mortality due to neutropenia and myocarditis)

Pimavanserin (Nuplazid)

Anxiolytics- Avoid use of benzodiazepines (ex: lorazepam, alprazolam except for RBD)
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Pimvanserin

Approved 4/2016 for Parkinson’s disease with psychosis

Acts as an 5HT2A inverse agonist (rather than direct agonist of 5HT2A and D2 receptors as atypical antipsychotics do)

Most common side effects: nausea, peripheral edema and confusion

Estimated annual cost: $23,400 ($1950 per month)
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Motor/Movement symptoms (Parkinsonism)
- tremor
- bradykinesia (slowing)
- rigidity
- myoclonus (limb jerking)
- shuffling gait
- stooped posture
- fine motor skills disrupted
- masked facies
- drooling/swallowing difficulties
- hypophonia (soft voice)
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Associated with decrease in dopamine

Worsened by medications that block dopamine (see previous slide on cognition)

Improved by medications that increase dopamine—Parkinson’s medications (sinemet, Mirapex, Requip, Neupro patch)
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For speech and swallow symptoms (drooling, difficulty chewing, pocketing food, coughing on liquids/foods, food sticking in the throat, “forgetting to swallow”)

Speech amplifier (Spokeman)
Speech therapy
“Chin tuck”
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Sleep symptoms
  REM sleep behavior disorder (video)
  Daytime somnolence
  Insomnia
  Obstructive Sleep Apnea
  Central Sleep Apnea
  Restless legs syndrome
  Periodic Limb Movements
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Different symptoms associated with different neurotransmitter issues and treated differently

RBD- clonazepam-increase in GABA (can cause daytime confusion)

Daytime Somnolence- Provigil, Ritalin – increase in dopamine, epinephrine and histamine

Insomnia- trazadone – increase in serotonin
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Autonomic dysfunction

orthostatic hypotension (drop in blood pressure when changing position)

impotence

urinary Incontinence

constipation
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Treatments vary

Orthostatic hypotension- salt, water, compression stockings, midodrine (can be made worse by Parkinson’s meds and BP meds)

Impotence- PDE5 inhibitor such as Viagra

Urinary Incontinence- muscarinic antagonist Sanctura (but can make cognition worse)

Constipation- water, sinekot, miralax
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Special Issue: Falls

gait changes
rigidity
move too slow/too fast
deconditioning
autonomic dysfunction
flexibility and balance
change in vision/interpretation
lack of proprioception
deficit in attention
deficit in integration/planning
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Treatment for one symptom may worsen another so it is important to use individual symptoms effecting quality of life to guide individualized treatment plan.
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On The Other Side of Suffering is Joy
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Acceptance

Empathy

Communication Skills

Understanding Needs
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Acceptance

Not the same as “giving up”

The disease/behaviors are ALWAYS changing, we must change with them
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EMPATHY

Behaviors are biomedical and a reasonable reaction to the person’s experience

See through their eyes: begin to see solutions

Modify tasks and demands
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**COMMUNICATION SKILLS**

Slow down
Clear and concise
One step requests
Avoid drawn out explanations
Don’t try to convince them of something
Keep voice volume moderate- tone of voice changes with volume
Uninterrupted time to process and respond (don’t talk for them!) but ok to help them get “unstuck”
Environment free of distractions
Look directly at the person when you talk to them
Watch your tone and body language
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Get attention, approach slowly
Face directly
Maintain eye contact
Smile
Appear relaxed
Orient (repeat key words)
paraphrase
give time to comprehend
ask “yes/no” questions
provide two options at a time
one question/instruction at a time
continue same topic for as long as possible
prepare/cue for a new topic
short/simple/direct sentences
***Repeat nouns rather than pronouns
hand signals, pantomimes, pictures
facial expressions
begin conversations with pleasant/normal topics
ask easy questions
give the person clues as to how to answer
Don’t correct but can repeat what they say using correct word
Ask them to “talk around” or describe something
ignore unimportant errors
ask to speak louder if they are getting too soft
Dementia does not rob someone of their dignity, it's our reaction to them that does.

Teepa Snow

AlzheimersCaregiverMinute.com
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UNDERSTANDING NEEDS

Need to feel respected

Need to feel sense of control

Need to feel useful and worthy

Need to give and receive love
MOST EMOTIONAL DISTRESS OCCURS WHEN A BASIC EMOTIONAL NEED IS THREATENED
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SEE STRENGTHS, NOT WEAKNESSES

REMEMBER, YOU ARE A TEAM, YOU NEED EACH OTHER TO “WIN”

IF YOU DON’T LAUGH, YOU CRY
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THE FUTURE

Genetics- very complicated, research into genes that put one at risk. One gene known (alpha SNCA gene) only a few dozen people in the world
Little effect of APOE. Some increased risk with a first degree relative with DLB, AD or PD
Shoot, we'd better hurry up and figure this out.

Aging researchers

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phone: 216.371.8600 / email: ft@funnytimes.com
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**THE FUTURE**

Sleep Research- Almost everyone who develops RBD will go on to develop DLB or PD, how to use that information to identify people decades before they develop the disease.

Imaging- use of MRI, PET, PIB and DaT scans to identify earlier.
The Future

Therapeutics-

RVT101- 5HT6 antagonist in phase 2 trials for cognition (Recruiting)

Nelotanserin- similar to Pimavanserin, phase 2 trials in DLB or PDD with psychosis (Recruiting)
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**THE FUTURE**

Nuvigil - sleep, apathy and quality of life studies, phase 3 trials (completed - no results available)

Donepezil - clinical trial (already being used, but only approved in Japan) - post marketing (Recruiting)

Nilotinib - decrease alpha synuclein in CSF (completed - no results available)
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THE FUTURE

Immunotherapy- Alpha synuclein antibodies, phase 1 trials
  vaccine- D001A and PA03A
  passive- B11B054 and PRX002

Deep Brain Stimulation- currently recruiting (London)
Too often we underestimate the power of a touch, a smile, a kind word, a listening ear, an honest compliment, or the smallest act of caring, all of which have the potential to turn a life around.

LEO BUSCAGLIA
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Resources:

The Lewy Body Dementia Association
https://www.lbda.org

The Alzheimer’s Association
www.alz.org

Living with a thief named Lewy Body Dementia blog
www.thieflewybodydementia.com

Clinical Trials
https://clinicaltrials.gov
THANK YOU!