Dementia With Lewy Bodies

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What is Dementia With Lewy Bodies?

It is a type of progressive dementia that involves a build-up of abnormal protein deposits, called Lewy bodies, in the brain. It is considered the third most common form of dementia, accounting for 20-25% of all dementias (the most common being Alzheimer's Disease Dementia, followed by Vascular Dementia).

What Are Lewy Bodies?

Lewy bodies can be found in the brain. They are composed of misfolded proteins of ubiquitin and alpha-synuclein, which accumulates into clumps. These clumps of toxic protein then interfere with normal cellular function, ultimately leading to the death of nerve cells and the cognitive and physical symptoms of Lewy Body Disease and Dementia.

What is Dementia?

Dementia is a general term used to describe a significant decline in cognitive function such that it affects a person's ability to perform everyday activities, such as directing and controlling attention, learning and memory functions, language, perceptual-motor skills and/or executive function. It is generally accepted that dementia is irreversible, progressive and incurable. Whilst dementia is the traditional term used, the DSM-5 refers to varying levels of Neurocognitive Disorder rather than dementia in an attempt to remove stigma associated with the word dementia.

Is Dementia With Lewy Bodies a Cortical or Subcortical Dementia?

Dementia with Lewy Bodies is a **systemic dementia**, meaning that it affects multiple regions of the brain and the body. It is better classified as a systemic, or cortico-subcortical dementia.

Around What Age Does Dementia With Lewy Bodies Tend to be Diagnosed?

Most develop between the **ages of 50-85** with the mean onset being around 67 years of age.

Does Dementia With Lewy Bodies Appear Gradually or Suddenly?

It tends to develop suddenly and with a rapid decline noted at first, before a levelling off. Overall, Dementia with Lewy Bodies progresses more rapidly than Alzheimer's Disease Dementia (which also has a more insidious onset).

Is Dementia With Lewy Bodies Genetic?

No, DLB is mainly sporadic (although rare cases of autosomal dominant inheritance have been reported).

Where Is the Damage in the Brain With Dementia With Lewy Bodies?

The Lewy Bodies are deposited throughout the **brainstem, cortex and sub-cortical areas**, especially in the **substantia nigra and locus coeruleus**. Degeneration and depletion of dopamine cells is typically observed in the substantia nigra, and **cortical degeneration is found particularly in the temporal and parietal lobes**.

The cingulate island sign is a biomarker that appears in FDG-PET or SPECT scans. It indicates that the metabolism of the posterior cingulate cortex is relatively preserved compared to the parietal and occipital lobes. This helps distinguish patients with Dementia with Lewy Bodies from those with Alzheimer's Disease since the sign is present in the former, but not the latter.

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A "diagnosis certain" case of dementia with Lewy bodies in a 60 year old © Radiopaedia

What Are the Symptoms of Dementia With Lewy Bodies?

Dementia with Lewy Bodies (DLB) can cause a range of symptoms, including:

- **Fluctuating cognition**: There can be significant changes in the person's ability to reason, problem-solve, and remember over the course of a day or even within hours. Fluctuations may also occur in mood and movement. These fluctuations can be confusing and disorienting for both the patient and their caregivers and can also make it difficult to accurately diagnose the condition.
- **Disproportionate visuo-spatial problems**: DLB patients tend to have greater difficulty with visuospatial skills such as drawing or constructing than in other cognitive domains.
- **Visual hallucinations**: People with DLB might see things that are not there, such as animals or people, and they may not recognize familiar faces.
- **Parkinsonian symptoms**: resting tremor (less prevalent {10%} or severe than in Parkinson's Disease, where asymmetry of motor symptoms is also the norm in early stages), stiffness, and difficulty with gait (stooped and shuffling) - which may cause falls.
- **Extrapyramidal symptoms:** akathisia (feeling restless), dystonia (muscles contract involuntarily), tardive dyskinesia: (involuntarily facial movements), bradyphrenia (slowed movement and thinking), parkinsonism (see above).
- **Sleep disturbances**: REM sleep behaviour disorder, where the person acts out their dreams, is experienced by about 15% of DLB patients.
- **Changes in mood and behaviour**: People with DLB may become more anxious, depressed (40% of patients), or apathetic, and they may display impulsive or disinhibited behaviour.
  Not everyone with DLB will experience all of these symptoms, with presentation dependent on where the Lewy Bodies are in the brain, and the severity of symptoms varies from person to person.

How Can a Neuropsychological Assessment Help Those With Suspected Dementia With Lewy Bodies?

A neuropsychological assessment can help those with suspected Dementia with Lewy Bodies by evaluating their cognitive abilities (such as memory, attention, language, and executive function), identifying any deficits, and tracking changes over time. This information can be used by the clinical neuropsychologist to develop a neuropsychological rehabilitation plan (where possible), monitor the progression of the disease, and provide support for the individual and their family.

Additionally, a neuropsychological assessment can help differentiate Dementia with Lewy Bodies from other types of dementia, which is important for accurate diagnosis, treatment and support. It

is wiser to achieve this as early as possible in the disease progression where differential diagnosis is more accurate than at later stages where symptoms of dementia subtypes tend to be less distinguishable.

What Neuropsychological Tests Are Recommended When Assessing Those With Suspected Dementia With Lewy Bodies?

Neuropsychological tests that are recommended when assessing those with suspected Dementia with Lewy Bodies include tests of attention, executive function, visuospatial function, memory, language, and perceptual-motor function.

Additionally, tests of fluctuation in cognition, attention, and alertness may also be useful in the assessment of Dementia with Lewy Bodies. Basically, therefore, using something like the WAIS-IV alongside the WMS-IV and the Boston Naming Test and CVLT would suit, and especially given notable differences in performance domains on this battery between those with DLB versus Alzheimer's Disease.

What Is the Neuropsychological Profile of Somebody With Dementia With Lewy Bodies?

- The neuropsychological profile of DLB is characterised by deficits in attention, executive function, visuospatial abilities, and memory.
- Whilst visuospatial deficits can exist in a number of disorders, they tend to be disproportionately impaired relative to other domains in DLB where patients will do poorly on block design, figure copying and clock drawing. Mistakes made in clock drawing tend not to improve on copy.
- Memory deficits vary and tend to be more "frontal", with issues in encoding and retrieval, such that DLB patients are likely to do better than Alzheimer's Disease patients on delayed recognition (retrieval), despite doing similarly on delayed recall.
- Comprehension tends to be generally intact, but there may be syntax errors and perseveration in speech.
- Psychotic symptoms, such as visual hallucinations and delusions, are also common in DLB.
- Additionally, motor symptoms, such as parkinsonism, may be present (rigidity, bradykinesia, gait disorder/slow/shuffling, falls, masked faces, hypophonic speech).

It is important to note that the neuropsychological profile of DLB can vary widely between individuals (depending, to an extent, on where the Lewy Bodies are located in the brain), and may overlap with other neurodegenerative disorders, such as Parkinson's disease and Alzheimer's disease.

A comprehensive neuropsychological evaluation by a clinical neuropsychologist is necessary to accurately diagnose and manage DLB.

What Are Some of the Differential Diagnoses Related to Dementia With Lewy Bodies?

Some of the differential diagnoses related to Dementia with Lewy Bodies include Alzheimer's disease, Parkinson's disease, frontotemporal dementia, vascular dementia, and normal pressure hydrocephalus.

What Are the Main Differentials Between the Neuropsychological Profile for Dementia With Lewy Bodies Versus Alzheimer’s Disease?

The neuropsychological features of DLB are generally considered similar to those of Alzheimer's Disease but DLB has more of a fronto-subcortical profile with greater deficits in:

- attention
- psychomotor speed
- visuospatial skills (early visual deficits tend to be an early sign for DLB rather than Alzheimer's Dementia)
- executive function
- verbal fluency

Orientation is typically intact in DLB and less so in Alzheimer's Dementia.

Patients with Dementia with Lewy Bodies demonstrate more visuo-constructional and visuo-spatial deficits than those with Alzheimer's disease, whilst those with Alzheimer's Disease are more likely to present with memory difficulties. As a result of the former, those with DLB may experience greater difficulty perceiving and naming objects. Research has demonstrated that error analysis on the Boston Naming Test can accurately identify between these 2 groups. Those with DLB tend

to make more visuospatial errors, whilst those with Alzheimer's disease made more semantic errors and had inferior performance overall.

Note that brain bank studies show that DLB is often misdiagnosed as Alzheimer's Disease. It's also important to note that some of the deficits noted above may not necessarily be seen during a neuropsychological assessment as a result of the fluctuation prevalent in DLB.

Is Dementia With Lewy Bodies Related to Tau Misfolding?

No, Dementia with Lewy Bodies is not directly related to tau misfolding. It is related to the accumulation of alpha-synuclein protein in the brain, which forms Lewy body clumps. Tau misfolding, on the other hand, is associated with other neurodegenerative diseases such as Alzheimer's disease. However, DLB and Alzheimer's often co-occur, so individuals with DLB may also (but not always) have symptoms of Alzheimer's Disease such misfolding tau.

What Causes Attention to Fluctuate in Dementia With Lewy Bodies?

In Dementia with Lewy Bodies, attention may fluctuate due to changes in the levels of acetylcholine and dopamine in the brain. These are important neurotransmitters involved in attention and other cognitive processes. Additionally, Lewy bodies themselves can affect attention.

What Causes the Sleep Disturbances Seen in Dementia With Lewy Bodies?

The sleep disturbances seen in Dementia with Lewy Bodies are caused by disruption in the brain and brainstem circuitry that affects the regulation of sleep-wake cycles, as well as dystonia and inhibition. As a result, patients often do not experience sleep paralysis, and instead act out their dreams (often quite violently). They are often not aware of this, but their bed partner will be.

Why May People With Dementia With Lewy Bodies See Children or Animals?

People with Dementia with Lewy Bodies may experience visual hallucinations, which can include seeing people, animals or objects that are not actually there. The hallucinations can be quite vivid.

There is evidence to suggest that changes in cholinergic function within the thalamic and visual associative areas of the brain may cause the hallucinations, and this is supported by the fact that anti-cholinergic drugs have been found to reduce visual hallucinations.

Is Dementia With Lewy Bodies Treatable?

While there is no cure for Dementia with Lewy Bodies, there are treatments available that can help manage the symptoms. These treatments may include medications to address cognitive, movement, and psychiatric symptoms, as well as non-pharmacological interventions such as physical therapy, occupational therapy, and speech therapy.

Is Dementia With Lewy Bodies Preventable?

There is currently no known way to prevent Dementia with Lewy Bodies. However, some studies suggest that a healthy lifestyle, including regular exercise and a balanced diet, may help reduce the risk of developing certain types of dementia, including Dementia with Lewy Bodies.

Is Any Research Into Dementia With Lewy Bodies Being Undertaken Right Now?

Yes, research into Dementia with Lewy Bodies is currently being undertaken by various organizations and institutions around the world. There are ongoing studies focused on understanding the underlying causes of the disease, developing better diagnostic tools, and identifying potential treatments.

In terms of potential treatments, there are ongoing clinical trials testing new medications and therapies aimed at slowing or halting the progression of Dementia with Lewy Bodies. Some of these treatments target the underlying pathways believed to contribute to the development and progression of the disease, while others focus on managing the symptoms associated with Dementia with Lewy Bodies.

Regarding diagnostic tools, researchers are working on improving the accuracy of diagnostic tests, such as brain imaging and blood tests, to help identify Dementia with Lewy Bodies earlier in its progression. Early diagnosis is critical to allow for early intervention and treatment.

In terms of understanding the causes of Dementia with Lewy Bodies, researchers are exploring the role of genetics and the presence of certain proteins in the brain. Some studies are looking into how environmental factors such as toxins and infections may contribute to the development of the

disease. There is also ongoing investigation into the relationship between Dementia with Lewy Bodies and other neurodegenerative disorders such as Alzheimer's disease.

Overall, the research being done on Dementia with Lewy Bodies is crucial for both understanding the disease and developing effective treatments. With ongoing efforts, there is hope for better outcomes and improved quality of life for those living with this condition.

Suggested Videos on Dementia With Lewy Bodies

Embedded media: https://www.youtube-nocookie.com/embed/qfXWqlf2WBk?controls=0

Embedded media: https://www.youtube-nocookie.com/embed/vwsmat2tbgc?controls=0

Further Reading

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Neuropsychological Assessment for Dementia With Lewy Bodies

If you or a loved one is experiencing cognitive difficulties similar to those mentioned above, do please contact us for neuropsychological assessment from a clinical neuropsychologist.