

# Lewy body disease

This Help Sheet has been developed to assist your understanding of Lewy body disease and the disorders that arise from it.

## What is Lewy body disease?

Lewy body disease is a common neurodegenerative disease of ageing. This means that the disease causes gradual brain damage. For reasons not fully understood, it occurs when there is an abnormal build up of a protein called alphasynuclein in brain cells. These abnormalities occur in specific areas of the brain, causing changes in movement, thinking and behaviour.

Lewy body disease includes three overlapping disorders:

- Dementia with Lewy bodies
- Parkinson's disease
- Parkinson's disease dementia

This overlap results in the disease being called a spectrum disease.

## History of Lewy body disease

In 1817, James Parkinson described a group of people with similar problems. He wrote about the movement or physical signs (commonly referred to as motor signs) that these people exhibited, but stated that they did not appear to have any mental impairment.

Forty years later two French doctors, Charcot and Vulpian, assigned the name Parkinson's disease to people showing the signs that Parkinson had described. However, they disagreed that there was no mental impairment.

In 1912 scientists in Germany were identifying abnormalities in brain tissue. Among the team were Alois Alzheimer and Friedrich Lewy. Lewy identified the alphasynuclein deposits in brain samples taken from people who had been diagnosed with Parkinson's disease. In 1919 a researcher called Tretiakoff named these deposits Lewy bodies.

## New understanding

In the early 1990s advances in laboratory techniques enabled researchers in the UK to more easily identify Lewy bodies in brain tissue. Earlier reports from Japanese researchers suggested a clinical pattern of

cognitive (thinking) and physical deficits in people diagnosed with Parkinson's disease and work in the UK confirmed these findings.

The first international meeting on dementia with Lewy bodies was held in the UK in 1995. This resulted in the publication of criteria for the diagnosis of dementia with Lewy bodies. These criteria have been updated at subsequent meetings.

For years, it has been accepted that as Parkinson's disease progresses many people develop cognitive impairment (Parkinson's disease dementia). Now, ongoing research is leading to greater understanding that in some people cognitive impairment can precede the movement symptoms of Parkinson's disease (this is diagnosed as dementia with Lewy bodies).

None of these disorders are new, yet our recognition of all of their features is not well established. Developments in research and clinical practice are providing a better understanding of this disease and its impact – both on the people with it and their carers. The diagnosis of one disorder over another may simply depend on the time the person presents for assessment.

Naming within the spectrum can lead to confusion.

- Lewy body disease is the 'umbrella' term signifying there is underlying alphasynuclein deposits in the brain.
  - Parkinson's disease is usually diagnosed when a person develops significant movement symptoms first.
  - Parkinson's disease dementia is diagnosed when a person with established Parkinson's disease subsequently develops significant cognitive impairment.
  - Dementia with Lewy bodies is usually diagnosed when a person develops significant cognitive symptoms first.
  - Lewy body dementia refers to the cognitive changes typically seen across the spectrum of disorders.
- Remember too that there is overlap between the distinct disorders.

National Dementia Helpline **1800 100 500**

[dementia.org.au](http://dementia.org.au)

## Getting a diagnosis

Lewy body disease and its accompanying disorders can present diagnostic challenges. Lewy body disease is still not well known. Also, people with Lewy body disease can present with a variety of problems in the early stages of the disease. They may have problems with autonomic (the system that automatically regulates bodily functions), cognitive (thinking), behavioural or motor functions. Further explanations of these presentations are provided on subsequent sheets in this Help Sheet series.

One of the Lewy body disorders (Parkinson's disease) does have a public profile and established protocols for its diagnosis and treatment. It is not surprising, therefore, that people who experience the motor or physical symptoms in the early stages of their illness seek advice from medical practitioners who specialise in movement disorders.

If a diagnosis of Parkinson's disease is made, with time a subsequent diagnosis of Parkinson's disease dementia is, in most cases, inevitable.

Getting a diagnosis can be more challenging when the motor signs are not as evident. Often the person dismisses the concerns of others, presents well at a GP consultation and performs well on initial screening tests such as the Mini Mental State Examination (MMSE). A 'watch and review' plan is sometimes suggested or medication is offered for the most pressing complaint.

Family or friends can be advocates. They may express their concerns to the doctor in person, or by telephone or letter. They may visit the doctor with the person and, if still concerned, ask for a referral to a geriatrician or specialised assessment clinic.

A full assessment may include:

- A history from the person
- An interview with a family member
- Screening blood tests
- A full neuropsychological assessment (tests of cognitive abilities)
- Brain imaging
- Other medical tests

Even with this battery of tests, a definitive diagnosis of a Lewy body disorder (particularly dementia with Lewy bodies) may not be possible at the first assessment.

There are established clinical criteria for the diagnosis of all Lewy body disorders including dementia with Lewy bodies. The assignment of one diagnosis instead of another is a function of timing. If the cognitive deficits (dementia) are apparent prior to the development of motor signs then dementia with Lewy bodies is the appropriate diagnosis. If the dementia follows the motor signs then the diagnosis is Parkinson's disease dementia.

The acknowledgement of a pre-motor phase of Parkinson's disease, which can include cognitive deficits, confounds these arbitrary classifications.

It must be acknowledged that some people with Lewy body disease also have signs and symptoms typical of Alzheimer's disease, particularly as the illness progresses, and this can add to difficulties in getting an accurate diagnosis. Brain damage consistent with both diseases is often found in imaging and autopsy studies.

## Resources

Additional Help Sheets on Lewy body disease are available at the Dementia Australia website  
Visit [dementia.org.au](http://dementia.org.au).

Parkinson's Australia is the peak body for advocacy and support of people with Parkinson's disease.  
Visit [parkinsons.org.au](http://parkinsons.org.au) or call **1800 644 189**.

The US Lewy Body Dementia Association  
Visit [lbda.org](http://lbda.org)

The UK Lewy Body Society  
Visit [lewybody.co.uk](http://lewybody.co.uk)

## FURTHER INFORMATION

Dementia Australia offers support, information, education and counselling. Contact the National Dementia Helpline on **1800 100 500**, or visit our website at [dementia.org.au](http://dementia.org.au)



For language assistance phone the Translating and Interpreting Service on **131 450**