

What is dementia with Lewy bodies (DLB)?

Factsheet 403LP
June 2016



Dementia with Lewy bodies (DLB) is a type of dementia that shares symptoms with both Alzheimer's disease and Parkinson's disease. It may account for 10–15 per cent of all cases of dementia. DLB can be diagnosed wrongly and is often mistaken for Alzheimer's disease. This factsheet describes the symptoms of DLB and how it is diagnosed, as well as the treatment and support available.

DLB is sometimes known by other names. These include Lewy body dementia, Lewy body variant of Alzheimer's disease, diffuse Lewy body disease and cortical Lewy body disease. All these terms refer to the same condition.

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What is dementia with Lewy bodies (DLB)?

Lewy bodies

Lewy bodies are named after the German doctor who first identified them. They are tiny deposits of a protein (alpha-synuclein) that appear in nerve cells in the brain. Researchers don't have a full understanding of why Lewy bodies appear, or exactly how they contribute to dementia. However, this is linked to two factors:

- low levels of important chemicals (mainly acetylcholine and dopamine) that carry messages between nerve cells
- a loss of connections between nerve cells, which then die.

Lewy bodies are the cause of DLB and Parkinson's disease. They are two of several diseases caused by Lewy bodies that affect the brain and nervous system and get worse over time. These are sometimes called Lewy body disorders.

The way someone is affected by DLB will depend partly on where the Lewy bodies are in the brain:

- Lewy bodies at the base of the brain are closely linked to problems with movement (motor symptoms). These are the main feature of Parkinson's disease.
- Lewy bodies in the outer layers of the brain are linked to problems with mental abilities (cognitive symptoms), which is a feature of DLB.

People with a Lewy body disorder can have problems with movement and changes in mental abilities at the same time. A person with Parkinson's disease is at high risk of going on to develop dementia (Parkinson's disease dementia) as their condition progresses. Dementia may be more likely in a person who has developed Parkinson's later in life or who has been living with it for several years.

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DLB accounts for around 4 per cent of all recorded dementia, but there is good evidence that the condition is not always diagnosed correctly. Based on studies of brain tissue after death, scientists think DLB may account for as much as 10–15 per cent of all dementia.

Similarly, at least two-thirds of people with DLB develop movement problems at some point. The symptoms of DLB and Parkinson's disease dementia become more similar as the conditions progress. Together they are referred to as Lewy body dementias.

Who is affected?

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DLB appears to affect men and women equally. As with Alzheimer's disease and vascular dementia, DLB becomes more common over the age of 65. However, it can also affect people under 65.

Other than age, there are few risk factors (such as medical conditions or lifestyle choices) that are known to increase a person's chances of developing DLB. Most people who develop DLB have no clear family history of the condition. A few families seem to have genetic mutations that are linked to inherited Lewy body disease, but these are very rare. For more on this see factsheet 405, **Genetics of dementia**.

Symptoms

As with most types of dementia, the first symptoms of DLB may affect someone only slightly, but gradually they get worse and cause problems with daily living. Everyone is different, but a person with DLB will usually have some of the symptoms of Alzheimer's disease and some of the symptoms of Parkinson's disease. They will also have some symptoms which are unique to DLB. These are covered in the sections below.

Problems with mental abilities

Problems with attention and alertness are very common in DLB. An important feature of the condition is that these problems may switch

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between being bad and then better – or the other way round – over the course of the day, by the hour or even a few minutes. Someone with DLB may stare into space for a long time or have periods when their speech is disorganised.

People may also have difficulties judging distances and seeing objects in three dimensions. It is common to struggle with planning, organising and decision-making. Some people also experience depression. Day-to-day memory is often affected in people with DLB, but usually less in the early stages than in early Alzheimer's disease.

Hallucinations and delusions

Visual hallucinations (seeing things that are not there) occur frequently in people with DLB, often in the early stages of the condition. They can happen daily. Visual hallucinations are often of people or animals, and are detailed and convincing to the person with dementia. They can last several minutes and may be distressing. (Someone may also have visual misperceptions, such as mistaking a shadow or a coat on a hanger for a person.) Auditory hallucinations – hearing sounds that are not real, such as knocking or footsteps – can happen but are less common.

Hallucinations and visual misperceptions partly explain why most people with DLB have delusions (persistently thinking things that are not true) at some stage. Some common delusions held by people with DLB are believing that someone is out to get them, that there are strangers living in the house, or that a spouse is having an affair or has been replaced by an identical imposter. The person's relatives and carers may find these delusions very distressing.

Visual hallucinations are also a common reason for a person with DLB to stop driving, because it is no longer safe for them to be on the road. For more on this sensitive issue see factsheet 439, **Driving and dementia**.

Movement problems

Up to two-thirds of people with DLB have movement difficulties when the condition is diagnosed, and this number increases as DLB progresses. These symptoms are similar to those of Parkinson's disease, and include slow and stiff (rigid) movement with a blank facial expression.

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The person's posture may be stooped and their walk may be shuffling. They may also have difficulty with their balance, and their limbs may sometimes tremble.

Movement problems are one reason why a person with DLB is particularly prone to falls.

Sleep disturbance

Sleep disturbance is another common symptom of DLB and may start years before the diagnosis. Someone with DLB may fall asleep very easily during the day, but have restless, disturbed nights. In the most common night-time sleep problem, the person has violent movements (eg yelling, hitting out) as they try to act out nightmares. This is called rapid eye movement sleep behaviour disorder and is found in different Lewy body disorders such as DLB and Parkinson's disease. For bed partners it can be very distressing or even physically harmful.

Other symptoms

Some people with DLB lose their sense of smell, become constipated or have urinary incontinence (passing urine when they don't intend to). They may also faint or have unexplained episodes when they lose consciousness for a few minutes.

Later stages

DLB is a progressive condition. Over time, symptoms increase and get worse. This is generally over a period of several years.

As the disease progresses, problems with day-to-day memory and other mental abilities become more similar to those of middle- or later-stage Alzheimer's disease. People can also develop behaviours that challenge (eg agitation, restlessness, shouting out).

Worsening movement problems mean that walking gets slower and less steady. As a result, falls become more common. In the later stages of DLB, many people have problems with speech and swallowing, leading to chest infections or risk of choking.

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Eventually, someone with DLB is likely to need a large amount of nursing care. How quickly the condition progresses and the life expectancy of a person with DLB vary a lot. On average someone might live for about six to 12 years after the first symptoms, similar to a person with Alzheimer's disease.

Diagnosis

It is always important to get dementia diagnosed, but knowing the type of dementia (eg DLB, Alzheimer's disease) is particularly important if DLB is suspected. This is mainly because people with DLB can benefit from some medications, but also react very badly to others.

However, DLB can be difficult to identify correctly and diagnosis should ideally be made by a specialist with experience of the condition. People with DLB are often mistakenly diagnosed as having Alzheimer's disease or, less often, vascular dementia. Other things that can cause similar symptoms, including infections or the side effects of medication, also need to be ruled out.

The doctor will talk to the person, and someone who knows them well. They will ask about the person's medical history, when the symptoms started, and how these are affecting their life now. The doctor will carry out mental ability tests, but these may not always give a reliable result. This is because symptoms of DLB can vary a lot over time, particularly on tasks that need attention and decision-making. A neurological examination (looking at reflexes, stiffness and balance, for example) should also be done.

Typical features of someone who is diagnosed with DLB are:

- varying levels of attention or alertness
- persistent detailed visual hallucinations
- movement problems similar to those of Parkinson's disease
- rapid eye movement sleep behaviour disorder.

If the person shows signs of DLB, brain scans are used to help to clarify the diagnosis. Computed tomography (CT) or magnetic resonance imaging (MRI) scans may rule out brain conditions (eg a brain tumour or bleed)

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which can have similar symptoms. They may also help to show that the person has DLB rather than vascular dementia. If there is still doubt, a more specialised brain scan might be carried out. This can confirm a diagnosis of DLB if it shows loss of a particular type of cells (dopamine nerve cells) at the base of the brain.

Treatment and support

At present, there is no cure for DLB. If the person is able to manage their symptoms, it will help them to live well. Given the range of different types of symptoms, they are likely to need support from a variety of professionals at different times. With correct diagnosis and support from a team of professionals, it is possible to live well with DLB.

When caring for someone with DLB, it is important to be as flexible as possible, bearing in mind that many of the symptoms will vary over time.

Treatment of DLB with drugs is often problematic. This is because everyone responds differently, there is only a small amount of evidence about what works, and there are few approved treatments. Also, medication given to improve hallucinations can make movement problems worse. Similarly, drugs for movement problems may have a negative effect on mental abilities or make hallucinations worse. There are also serious issues for a person with DLB with use of a specific group of medications known as antipsychotic drugs (see 'Hallucinations and delusions' below).

Managing the symptoms of DLB starts by focusing on those that affect the person the most. Where possible, they should be helped to manage these symptoms in ways that don't use drugs (non-drug approaches), before trying drug treatments.

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Problems with mental abilities

As in other types of dementia, a person with DLB should be offered non-drug approaches to help them to maintain their mental abilities. Examples of these include social interaction, staying mentally active (cognitive

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stimulation), developing a structured routine and using practical strategies to live well with memory loss (see factsheet 526, **Coping with memory loss**).

There is some evidence that the drugs donepezil and rivastigmine (which are routinely prescribed for Alzheimer's disease) can help with DLB. They improve a person's mental abilities, such as attention and alertness, as well as their ability to do day-to-day tasks. There is less evidence to support use of the related drug galantamine. None of these drugs are currently licensed for use in DLB in Europe, although rivastigmine is licensed for use in Parkinson's disease dementia. For this reason, families may find that doctors are reluctant to prescribe one of these drugs for a person with DLB, though they may do so 'off label' (outside the terms of the licence).

Memantine is a drug often given to people in the later stages of Alzheimer's disease. Some evidence shows that it can also help people with DLB, but not all of the evidence shows this.

Hallucinations and delusions

If someone is having hallucinations or delusions, in most cases it is unhelpful to try to convince them that what they are seeing is not there, or that what they believe is untrue. What the person is experiencing is real to them at the time. Instead, carers can offer reassurance that they are there to support the person, and perhaps try distracting them.

It is important to get any glasses or hearing aids checked. Any problems with these can make difficulties with perceiving things worse. Misperceptions (different from hallucinations) can also be triggered by things in the environment such as excess noise and reflective or patterned surfaces. For more information see factsheet 527, **Sight, perception and hallucinations in dementia**.

If hallucinations are distressing or likely to lead to physical harm, drug treatments may be offered. There is evidence that donepezil and rivastigmine (see 'Problems with mental abilities' above) reduce hallucinations and delusions in people with DLB. This treatment is recommended by the National Institute for Health and Care Excellence (NICE) and may be prescribed 'off label'.

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However, some drugs may not help and may even harm the person. In particular, the prescription of antipsychotic drugs for hallucinations or delusions in DLB is very problematic. These drugs do not work in everyone and are known to increase the risk of stroke and death in people with dementia.

There is an additional and much greater risk to the use of antipsychotic drugs in people with DLB (in comparison with other types of dementia). Up to half of people taking these drugs have severe reactions, with difficulty moving (including stiffness), becoming more confused, and being unable to perform tasks or communicate. These drugs may even cause sudden death. If someone goes into hospital or sees a new doctor, the person or family should check that medical staff know that the person has DLB and that this diagnosis is clearly recorded in the person's notes.

If a person with DLB must be prescribed an antipsychotic for distressing visual hallucinations or delusions, this should be done only by a specialist after all other approaches have failed. The drug should be given with the utmost care, under constant supervision and with regular monitoring.

Behaviours that challenge

As in other types of dementia, behaviours that are challenging to cope with – for the person and the carer (eg agitation, aggression) – should be seen as a form of communication or a sign of an unmet need. Carers or professionals should try to identify and meet this need in a way that is tailored to the individual. The cause behind a behaviour may be a medical condition, such as pain, or the side effects of drugs. It may also indicate that the person is frustrated, scared or bored. To manage these, carers should look for specific triggers and try to make appropriate changes in the person's care or environment, such as reducing unnecessary noise and clutter.

This type of behaviour can often be prevented or made better by interaction with other people, or through activities matched to the person's abilities and interests. There is also evidence of benefits from music therapy, physical exercise and hand massage. Life story work or reminiscence can help with low mood. For more information see factsheets 525, **Changes in behaviour**, and 509, **Dementia and aggressive behaviour**.

Sleep disturbance

Steps can be taken to help a person with DLB have more restful nights. Increasing physical exercise, having a regular bedtime and getting-up time, and reducing daytime napping can all help. It can also help to avoid alcohol, caffeine and nicotine close to bedtime, and to keep the bedroom quiet and at a comfortable temperature.

Practical steps can be taken to make the area around the bed safe – remove sharp objects and put a mattress alongside the bed in case the person falls or jumps out.

Carers should also talk to the GP about sleep problems. Drugs such as clonazepam or melatonin can be effective treatments.

Movement problems

A person with movement problems will often benefit from working with an occupational therapist or physiotherapist. These professionals can help the person to move and stay independent, as well as advise on aids and adaptations in the home. For more information see factsheet 429, **Equipment, adaptations and improvements to the home**.

If the person has had falls or is worried about falling, the GP may refer them to a falls prevention service. Their risk of falls may be reduced by strength and balance exercises, a sight test, a medication review and making the home safer (eg removing trip hazards).

Support from a speech and language therapist is often helpful if the person develops problems with swallowing or speaking.

The drug levodopa, routinely used to treat Parkinson's disease, is sometimes given to people with DLB and movement problems. Levodopa is, however, less effective in DLB than in Parkinson's disease. It improves movement problems and falls in only around one third of people with DLB. Levodopa can also further increase confusion, reduce mental abilities (eg alertness) or make hallucinations worse.

With correct diagnosis and support from a team of professionals, it is possible to live well with DLB.

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Other useful organisations

Lewy Body Society

Hudson House
8 Albany Street
Edinburgh EH1 3QB

0131 473 2385
info@lewybody.org
lewybody.org

Charity that aims to raise awareness of DLB for the general public and educate those in the medical profession and decision-making positions about all aspects of the disease. They also support research into the disease.

Parkinson's UK

215 Vauxhall Bridge Road
London SW1V 1EJ

0808 800 0303 (helpline 9am–8pm Monday to Friday,
10am–2pm Saturday)
hello@parkinsons.org.uk
www.parkinsons.org.uk

Charity that provides information, advice, support and publications for people with Parkinson's disease. They can put people in touch with their nearest branch, offering information, support and social contact for those with Parkinson's and their families.

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This factsheet has also been reviewed by people affected by dementia.

A list of sources is available on request.

Alzheimer's Society National Dementia Helpline England, Wales and Northern Ireland:

0300 222 1122

9am–8pm Monday–Wednesday

9am–5pm Thursday–Friday

10am–4pm Saturday–Sunday

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