

What is dementia with Lewy bodies (DLB)?

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 around 10 per cent of all cases of dementia. DLB tends to be mistakenly diagnosed as other conditions (that is, DLB is under-diagnosed). This factsheet describes the symptoms of DLB and how it is diagnosed, as well as the treatment and support available.

DLB is sometimes referred to by other names, including Lewy body dementia, Lewy body variant of Alzheimer's disease, diffuse Lewy body disease, cortical Lewy body disease, and senile dementia of Lewy body type. All these terms essentially refer to the same disorder.

Lewy bodies

Lewy bodies, named after the doctor who first identified them, are tiny deposits of protein in nerve cells. Researchers don't have a full understanding of why Lewy bodies appear in the brain, or exactly how they contribute to dementia. However, their presence is linked to low levels of important chemical messengers (mainly acetylcholine and dopamine) and to a loss of connections between nerve cells. Over time, there is progressive death of nerve cells and loss of brain tissue.

Lewy bodies are the underlying cause of several progressive diseases affecting the brain and nervous system, notably DLB and Parkinson's disease. Together, these are sometimes called Lewy body disorders.

The symptoms a person experiences 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 are characteristic of DLB.

Movement problems and changes in mental abilities can occur together. About one third of people diagnosed with Parkinson's disease eventually develop dementia (Parkinson's disease 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

DLB accounts for around 4 per cent of all recorded dementia, but there is good evidence that the condition is under-diagnosed. Based on studies of brain tissue after death, scientists think DLB may represent as much as 10 per cent of all dementia.

DLB appears to affect men and women about equally. As with Alzheimer's disease and vascular dementia, DLB becomes more common over the age of 65. However, in certain cases people under 65 develop DLB.

Other than age, there are few risk factors (medical, lifestyle or environmental) which are known to increase a person's chances of developing DLB. Most people who develop DLB have no clear family history of the disease. A few families do seem to have genetic mutations which are linked to inherited Lewy body disease, but these mutations are very rare.

Symptoms

As with most types of dementia, the initial symptoms of DLB are often subtle, but gradually worsen to 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.

Problems with attention and alertness are very common. It is a feature of DLB that these problems vary (fluctuate) widely over the course of the day, by the hour or even a few minutes. There may also be difficulties with judging distances and perceiving objects in three dimensions, and with planning and organising. Some also experience depression. Day-to-day memory is often affected in people with DLB, but typically less in the early stages than in early Alzheimer's disease.

Visual hallucinations (seeing things that are not there) occur in most people with DLB, and can be distressing. These are often of people or animals, and are experienced as detailed and convincing. Auditory hallucinations (hearing sounds that are not real, such as knocking or footsteps) can happen but are less common.

Hallucinations and visual difficulties partly explain why many people with DLB have delusions (thinking things that are not true). Someone may believe they are being persecuted, that there are strangers living in the house, or that a spouse has been replaced by an identical imposter. Relatives and carers may find such delusions very distressing.

Up to two thirds of people with DLB have movement problems when the condition is diagnosed, and this proportion increases as it progresses. These symptoms are those of Parkinson's disease, and include slowness and rigidity of movement with a blank facial expression. Walking is often stooped and shuffling, with problems balancing. Trembling of the limbs is also sometimes seen.

Motor symptoms are one reason why a person with DLB is prone to falls. They may also faint or have unexplained episodes when they lose consciousness for a few minutes.

Sleep disorders are another common symptom of DLB. The person may fall asleep very easily by day, but have restless, disturbed nights.

Common problems include confusion, hallucinations and violent movements as the person tries to act out nightmares. This night-time sleep pattern is called rapid eye movement sleep behaviour disorder. For bed partners it can be very distressing or even physically harmful.

Later stages

DLB is a progressive disease. Symptoms become worse and tend to become more numerous with time, generally over a period of several years.

As the disease progresses, problems with day-to-day memory and other mental abilities come to resemble more closely 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. 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.

Eventually, someone with DLB is likely to need extensive nursing care. The rate of progression of the condition and the life expectancy of a person with DLB are very variable. On average someone might live for about eight years after the first symptoms. This is similar to Alzheimer's disease.

Diagnosis

It is always important to get an accurate diagnosis of dementia, but a proper diagnosis 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 diagnose, 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. Tests for conditions other

than dementia that can cause similar symptoms, including infection or side effects of medication, will also need to be carried out.

The doctor will talk to the person, and someone who knows them well, about the person's medical history, when the symptoms started, and how these are affecting their life now. Tests of mental abilities are used but can be complicated by the fluctuating nature of DLB and its effect on visual and spatial abilities (eg drawing a shape). A neurological examination (looking at reflexes and balance, for example) should also be done.

A diagnosis of DLB is largely based on the symptoms – particularly fluctuating attention or alertness, persistent detailed visual hallucinations, and movement problems associated with Parkinson's disease. Brain scans help to clarify the diagnosis: computed tomography (CT) or magnetic resonance imaging (MRI) scans may rule out brain conditions (eg brain tumour, increased pressure) with overlapping symptoms. They may also help to distinguish DLB from 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 reduced density 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. However, with support it is possible to live well with DLB, and management of the condition aims to maximise the person's overall quality of life. Given the range of symptoms, input from a variety of professionals is needed at different times. When caring for someone with DLB, it is important to be as flexible as possible, bearing in mind that many of the symptoms will fluctuate.

Treatment of DLB with drugs is often problematic: everyone responds differently, there is only limited evidence about what works, and there are few approved treatments. Also, medication given to improve mental abilities or 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 with use of antipsychotic drugs for a person with DLB (see ‘Hallucinations and delusions’ below).

Management of the condition starts by focusing on the symptoms that the person and carer identify as the most troublesome. Non-drug approaches should be tried before drug treatments, where options are available.

Problems with mental abilities

A person with DLB will benefit from individualised and meaningful occupation (eg tasks, hobbies), social interaction, reminiscence or life story work and strategies to cope with memory loss and visual hallucinations. For more information see factsheet 526, Coping with memory loss.

There is some evidence that the drugs galantamine, donepezil and especially rivastigmine (which are routinely prescribed for Alzheimer’s disease) can help with DLB. They improve fluctuating cognition, alertness and daily functioning. None of these drugs are currently licensed for use in DLB, although one (rivastigmine) is licensed for 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).

There is mixed evidence of benefit from memantine, a drug which is often given to people in the later stages of Alzheimer’s disease.

Hallucinations and delusions

If someone is having hallucinations or delusions, in most cases it is unhelpful to try to convince them that there is nothing 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 a good idea to get any glasses or hearing aids checked. If faulty, these may increase any difficulty with perception. Misperceptions (different from true 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 good evidence that galantamine, 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'.

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 surrounding the use of antipsychotic drugs in people with DLB (in comparison with other types of dementia). In up to half of cases, these drugs cause a severe reaction, with rigidity, immobility, worsening confusion and an inability to perform tasks or communicate. They may even cause sudden death. If someone is admitted to hospital or sees a new doctor, the person or family should check that medical staff know that the person has DLB.

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 dementias, behaviours that challenge (eg agitation, aggression) in a person with DLB should be viewed as a form of

communication – a sign of an unmet need. Approaches tailored to the individual should try to identify and meet this need.

The underlying cause may be a medical condition such as pain or the side effects of drugs. Or the behaviour might reflect frustration, fear or boredom. To manage these, carers should look for specific triggers and make appropriate changes in the person's environment or care. Aromatherapy, massage or talking therapies may also help.

Behaviours that challenge can often be prevented by social interaction, structured activities (eg music or dance therapy, reminiscence and life story work) or physical exercise. For more information see factsheet 509, Dementia and aggressive behaviour.

If behaviours persist and are severe or distressing, drug treatments may be offered. One of the three Alzheimer's drugs – galantamine, donepezil or rivastigmine – should be tried first. If this fails and an antipsychotic is finally offered to someone with DLB, then all of the risks of a severe reaction (and need for extreme caution) mentioned above apply.

Sleep disorders

Steps can be taken to help a person with DLB have more restful nights. Increasing physical exercise and reducing daytime napping can be of benefit. It can also help to avoid alcohol, caffeine and nicotine late at night, and to keep the bedroom quiet and at a comfortable temperature.

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

Movement problems

A person with movement problems will often benefit from working with an occupational therapist or physiotherapist. These professionals can help the person maintain their independence

and mobility, and advise on aids and adaptations in the home. For more information see factsheet 429, Equipment, adaptations and improvements to the home.

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, and can further increase confusion and 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.

Other useful organisations

Lewy Body Society

Hudson House
8 Albany Street
Edinburgh EH1 3QB

T 0131 473 2385
E info@lewybody.org
W 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

T 0808 800 0303 (helpline 9am–8pm Monday to Friday, 10am–2pm
Saturday)
E hello@parkinsons.org.uk
W 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 11 22

9am–5pm Monday–Friday
10am–4pm Saturday–Sunday

alzheimers.org.uk

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