



# Information Sheet

## Dementia with Lewy bodies

This information sheet is for carers of people with dementia with Lewy bodies (DLB). Some people with early DLB may also find it useful. If you require further information about other issues regarding dementia call the freephone Dementia Helpline on 0808 808 3000, or see our website at [www.alzscot.org](http://www.alzscot.org). DLB may also be known as Lewy body dementia, diffuse Lewy body disease, dementia of Lewy body type or Lewy body variant of Alzheimer's disease.

This information sheet includes both medical information about the diagnosis and treatment of DLB, and practical information for carers, including some of the experiences of individual carers.

### What is dementia with Lewy bodies?

Dementia is the general term for a gradual progressive decline in a person's memory and other mental abilities. There are many different illnesses which can cause dementia, but only a few common ones. The best known are Alzheimer's disease and vascular dementia.

It is only in the last 10 to 15 years that dementia with Lewy bodies (DLB) has been recognised, as the result of research by psychiatrists and pathologists. This does not mean that it did not exist before. People with the condition would have been said to have a vascular dementia or Alzheimer's disease. This misdiagnosis still occurs, partly because not all doctors are yet aware of the condition, but also because it is not always easy to distinguish between these three common types of dementia.

Dementia with Lewy bodies is thought to be the second or third most common cause of dementia, accounting for 15% to 20% of cases of dementia which start after the age of 65.<sup>1</sup> This means that there could be about 11,000 people with DLB in Scotland.

People who have DLB have microscopic changes called Lewy bodies<sup>2</sup> in the nerve cells of the brain. These were first described by Frederick Lewy in 1912. He looked at the brains of people with Parkinson's disease, where Lewy bodies are mostly in one tiny area of the brain called the substantia nigra. In people with DLB they are much more widely spread throughout the brain.

It is not yet clear what the Lewy bodies signify, but it is likely that they are a

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<sup>1</sup> Campbell S, Stephens S and Ballard C (2001) Dementia with Lewy Bodies: clinical features and treatment. *Drugs Ageing*; 18(6): 397-407

<sup>2</sup> Lewy bodies are round deposits *within* damaged nerve cells, which are probably formed as the cells try to protect themselves and they are distributed in varying degrees throughout all areas of the brain.



sign of damage to the nerve cells. They contain various proteins, some of which have been identified. When it is clear whether these substances are damaging to the nerve cells or are the result of the damage, and how these processes happen, we are likely to know a lot about how Parkinson's disease and DLB develop, and therefore what treatments could prevent or treat these conditions effectively.

### Symptoms

Dementia with Lewy bodies shares the common features of all types of dementia. There is a gradual loss of mental abilities, including orientation and memory, reasoning and intelligence, with a decline in ability to carry out daily tasks. However there are certain specific characteristics of the decline which are much more likely in DLB, and less common in Alzheimer's disease or vascular dementia.

#### Patchy mental impairment

The person's memory impairment and disorientation may be patchy, with some quite normal memory function. He or she may communicate clearly, but may have difficulties with what are called the 'executive functions': problem solving, planning and the like. However, the mental decline in some people with DLB may be just like it is in someone with Alzheimer's disease.

#### Hallucinations

Seeing or hearing things which are not there is not very common in Alzheimer's disease, but 90% of people with DLB have hallucinatory experiences. Usually these will be visual hallucinations such as people, animals or inanimate objects. Occasionally the hallucinations are auditory – of music or voices. Most people with DLB find their hallucinations are puzzling, rather than upsetting or frightening. Unlike hallucinations in many mental illnesses, the person may have quite a lot of insight, realising at times that the visitors are imaginary. However some

people have no insight and may develop delusional beliefs about the hallucinations.

#### Neurological symptoms

People with DLB quite often have vague neurological problems which can include symptoms that are like those of Parkinson's disease, but which may not respond well to the usual treatment given to people with Parkinson's disease, the drug L-Dopa. These symptoms are often in just one part of the body, and may include:

- rigidity and stiffness
- difficulty starting movements (known as 'bradykinesia')
- slowness of movement
- tremor.

This pattern is different from other kinds of dementia. Neurological problems are rare in the early stages of Alzheimer's disease, although people in the very late stages develop problems with mobility and other neurological problems. People with vascular dementia may have some of the neurological problems associated with having small strokes.

Other symptoms which people with DLB may get include vague weaknesses of arms or legs, clumsiness or falls. These symptoms may come and go.

#### Variability

The person's degree of confusion can vary from moment to moment or day to day. He or she may have episodes of quite sudden severe confusion, which then pass. The person's insight is also likely to vary: at times he or she may realise that things are not right, that the hallucinations are imaginary, etc, but at other times he or she may have no apparent understanding of his or her illness.

#### Sleep disturbance

People with DLB may be wakeful at night, sometimes not sleeping at all for night after night. The person may not realise this; but it can of course be very



distressing for carers. Some people with DLB seem to experience an unusual condition in which they lose the normal paralysis that we normally have when we sleep. This means that they may be physically active, or act out dreams in the night. There has been a suggestion that particular drugs (clonazepam and desipramine) can help this.

### Neuroleptic sensitivity

At least 50% of people with DLB are over-sensitive to the side-effects of neuroleptic drugs (a type of drug used for illnesses such as schizophrenia and also used to treat hallucinations and delusions or as tranquillizers)<sup>3</sup>. Even a tiny dose may cause the person to become sedated, unsteady or develop symptoms like those of Parkinson's disease mentioned above. For this reason it is most important that there is someone else there if any of these drugs are given to *anyone* with early dementia. If signs of neuroleptic sensitivity develop, the drug should not be given again.

### Diagnosis and tests

One problem with DLB is getting a correct early diagnosis, as this dementia can often be mistaken for Parkinson's disease, depression or another dementia. Often relatives and the person with DLB do not expect a connection between hallucinations and dementia and will therefore not mention this to the GP. Many GPs will not recognise early signs

<sup>3</sup> Neuroleptics were originally developed to treat schizophrenia and other major mental illnesses. They have been found to be sometimes useful for other conditions where hallucinations or delusions occur and have also been used as general tranquillisers, to calm disturbed behaviour in dementia and other conditions. Examples are chlorpromazine (Largactil), risperidone (Risperdal), olanzapine (Zyprexa), haloperidol, trifluoperazine (Stelazine) and pericyazine (Neulactil). Thioridazine (Melleril) used to be given very commonly to people with dementia, but has now largely been withdrawn from use.

of DLB and will therefore not pick up on the diagnosis quickly. Ask to be referred to a specialist in order to obtain an accurate diagnosis.

To diagnose DLB, a specialist (usually an old age psychiatrist) will take a precise history of the pattern of the person's symptoms. This is so that he or she can be sure first of all that the person has dementia, as there are other reasons why a person may experience symptoms such as visual hallucinations (for example, people who are in delirium, the confusion that is caused by a physical illness). The specialist will then exclude other possible diagnoses, including vascular dementia and Alzheimer's disease.

A CT or CAT scan<sup>4</sup>, or magnetic resonance imaging (MRI)<sup>5</sup> may reveal degeneration, or shrinkage, of the brain. A SPECT scan<sup>6</sup>, may be able to show up some of the changes that are happening, but this is still being investigated. However the Lewy bodies themselves can only be seen by looking at brain tissue after death.

Early accurate diagnosis is particularly important with DLB because of the risks caused by over-sensitivity to neuroleptic drugs and because other drug treatments may help.

### Course of the illness

Unlike Alzheimer's disease, which usually begins very gradually, DLB often starts quite rapidly or acutely, with quite a fast decline in the first few months, although later there may be

<sup>4</sup> CAT or CT scans are like x-rays but more detailed and allow the specialist to see inside the brain.

<sup>5</sup> MRI uses radio waves and a strong magnetic field to give a clear and detailed picture of the brain.

<sup>6</sup> SPECT scans look at the pattern of blood flow in the brain.



some levelling off. DLB tends to progress faster than Alzheimer's disease and can last from 5 – 7 years, although this will vary from person to person.

## Caring for someone with DLB

### Information and support

If you need help with a particular problem, specific information or access to a service it is vital that you ask for it. Speak to your GP or social worker or Alzheimer Scotland – Action on Dementia for information and advice. Contact a local carers group to find out about other people who are caring or have cared for someone with DLB, as they will often be a vital source of information and support.

Very few people know what DLB is, and carers often find that there is little support from other people, as they do not understand what they are going through. As a result carers can feel very isolated. Being able to talk to someone who understands is very important. Alzheimer Scotland's **Dementia Helpline**, on freephone **0808 808 3000** is open 24 hours a day for information and emotional support, and can provide a free information pack and help you find services and support near you; or see the website at [www.alzscot.org](http://www.alzscot.org).

### Dealing with hallucinations and delusions

Hallucinations and delusions are often the most difficult symptoms to deal with. They can cause a great deal of emotional strain for carers. Some people with DLB will know they are experiencing hallucinations, while others will not be able to distinguish between reality and what is happening in their hallucinations and delusions. Being able to deal with these symptoms is important if the carer is to cope effectively. Ask for advice and support from the person's specialist or GP, and seek support for yourself, for example through a local dementia carer support group.

### Insight

Some of the most distressing times for carers can be when the person they care for understands what is happening to them. One carer found it particularly distressing, when her husband was in a care home, on days when he had insight into his condition. He would ask to come home with her, as he did not want to stay in the home anymore. For this carer this seemed worse than when he was unaware of the changes taking place in their lives. Seek support from family, other carers and professionals to help you cope with the distress the fluctuating insight of LBD can cause.

### Sleeping

Some people with DLB are sleepless or physically active in their sleep. (See *Sleep disturbance* on page 2.) One carer reported problems when her husband entered very deep sleeping phases and she could not wake him. This type of deep sleep can seem like a stroke. If the person has any problems related to sleep, always contact the GP to ensure there are no other problems and for treatment and advice.

### Vision

Some people with LBD experience problems with vision, with some people suffering from double vision. This can cause problems with many activities including eating, as the person can no longer see the food clearly to pick it up. Make sure the food is on a plain plate and that the table or tablecloth is a different colour from the plate, as this helps the person to distinguish between the food, plate and table.

### Dealing with dribbling

Dribbling can cause embarrassment and may prevent the person wanting to go out. Some carers find it hard to deal with this problem and may feel they cannot take the person out. This is a symptom which people with Parkinson's disease often have. There are drugs which can be prescribed by the person's GP which help dry up the saliva. However, some



carers have reported that these drugs can cause additional constipation problems because they are dehydrating.

### Other strategies to help

There are strategies that can help, especially in the early stages of DLB. These include keeping a set routine, providing written or 'alarm call' reminders and providing reassurance. People with DLB will often have times when they have a lot of insight into their condition and will need a great deal of support. Speech and language therapists can help people who are having difficulty in finding the right words or in following conversations. Closely monitoring the person's condition and the progression of the dementia will help provide vital information for his or her doctors.

### Services

There are many services that may help the person with DLB and his or her carer to cope more easily. The person with LBD is entitled to a **community care assessment** from the social work department to work out what services might help. The carer is also entitled to a **carer's assessment**.

Contact your local social work department to ask for assessments. The Dementia Helpline (0808 808 3000) can explain about assessments and services and can put you in touch with your local Alzheimer Scotland – Action on Dementia project for support.

### Treatment and drugs

There is no cure yet for DLB. Research continues to search for a specific course of therapy. The present form of treatment usually focuses on treating the symptoms. This means using drugs to control symptoms such as Parkinson type symptoms and hallucinations and delusions. A group of drugs developed for treating Alzheimer's disease ('acetylcholinesterase inhibitors' or 'cognitive enhancers') may also help people with DLB.

### Acetylcholinesterase inhibitor or cognitive enhancer drugs

Acetylcholinesterase (AChE) inhibitor drugs are also known as cognitive enhancers. They include **donepezil (Aricept)**, **rivastigmine (Exelon)** and **galantamine (Reminyl)**. These drugs are considered by many psychiatrists as the first line of therapy in DLB.

Donepezil and rivastigmine studies have shown that AChE inhibitors may be a safe and effective alternative to neuroleptic/anti-psychotic treatment. One study showed that people with DLB given donepezil (Aricept) showed an improvement in understanding, behaviour and psychiatric symptoms, although it is important to recognise that they are not suitable for everybody and will not help everyone who tries them.<sup>7</sup> However, there is a problem with prescribing these drugs for DLB, as they are only licensed in the UK for the treatment of mild to moderate Alzheimer's disease. Speak to the specialist treating the person with DLB to see if one of these drugs might be suitable and if he or she is able to prescribe it.<sup>8</sup>

These drugs prevent the breakdown of acetylcholine, a chemical which carries messages between brain cells. This means that there is more acetylcholine available to transmit messages between nerve cells. This may temporarily improve or stabilise the symptoms of the disease.

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<sup>7</sup> Samuel W et al (2000) Better cognitive and psychopathologic response to donepezil in patients respectively diagnosed as dementia with Lewy bodies: A preliminary study. *International Journal of Geriatric Psychiatry* 15, 794-802

<sup>8</sup> Gurling K, Kelly, J, Ramsay C, Rodriguez C. (2001) Dementia with Lewy Bodies and Dementia in Parkinson's Disease: Special considerations. In *Medication and Dementia*. Dementia Services Development Centre



The effect of these drugs varies from person to person. Some will not notice any effect, while others may find their condition improves slightly or they stay the same when they would have expected to deteriorate. People with DLB may see an improvement in behavioural symptoms such as apathy, hallucinations and delusions. Trials indicate that these drugs may delay the progression of the symptoms for up to nine months on average. Drugs should not be stopped after nine months as some people may benefit for longer than this.<sup>9</sup>

### Atypical anti-psychotic drugs

If treatment with AChE inhibitor drugs fails, you can discuss with the doctor the use of atypical neuroleptic/anti-psychotic drugs. Despite what was said above about neuroleptic drug sensitivity (see *Neuroleptic sensitivity* on page 3) some people with DLB are able to tolerate treatment so that their hallucinations are reduced. The utmost caution is needed. The drugs need to be started in the very lowest doses and the patient should be monitored regularly in the first days of treatment or after any change in dose.

Early reports on some of the more recently developed 'atypical' anti-psychotic drugs have suggested that olanzapine (Zyprexa), quetiapine (Seroquel) or risperidone (Risperdal), may be safe to use.

### Other treatments

If the person cannot tolerate neuroleptic drugs, and the AChE inhibitor drugs are ineffective, but hallucinations or other symptoms are distressing, the doctor will usually try other tranquilliser drugs, but there is no particular evidence that these

give specific help to the symptoms of DLB.

If the person has severe sleep disturbance the doctor will wish to try a hypnotic drug, but these are sometimes ineffective. You may need to suggest trying clonazepam and desipramine to the doctor.

There has been no research into any 'alternative' therapies for DLB.

### Problems with drug treatments

Anti-parkinson drugs such as L-dopa (levodopa) and Sinemet (a combination of levodopa and carbidopa) which may help reduce the tremor and loss of muscle movement may make the person's hallucinations and delusions worse.

Similarly, neuroleptic/anti-psychotic drugs prescribed for hallucinations may in fact make the muscle movement symptoms worse. Sometimes there is poor recovery of the muscle movement even after stopping the medication. In the worst cases, a patient treated with these drugs could become catatonic, lose cognitive function and/or develop more muscle rigidity, which could threaten their life. Some commonly used drugs which should be used with great caution, if at all, for people with DLB are chlorpromazine, haloperidol, or thioridazine.

### What is the cause?

The cause of DLB is uncertain. There are certain similarities with Alzheimer's disease and Parkinson's disease. Genetic studies are trying to discover which genes may contribute to the development of DLB, but these are still at an early stage. Just as the amyloid protein may play a central role in Alzheimer's disease there may be a protein or proteins which are central to the nerve cell damage in DLB. One candidate is alpha-synuclein, which is found within the Lewy bodies themselves.

<sup>9</sup> Australian Alzheimer Association. Feb. 2001  
Website information on drugs.  
[www.alzvic.asn.au](http://www.alzvic.asn.au)



## Useful websites

**Alzheimer Scotland – Action on Dementia:** [www.alzscot.org](http://www.alzscot.org) - over 500 pages about a wide range of issues relating to dementia, including caring, support, treatment, research, campaigns, training, volunteering and local services.

## Lewynet:

[www.nottingham.ac.uk/pathology/lewy/lewyhome.html](http://www.nottingham.ac.uk/pathology/lewy/lewyhome.html) - mainly medical information on DLB, plus links to other organisations' websites.

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