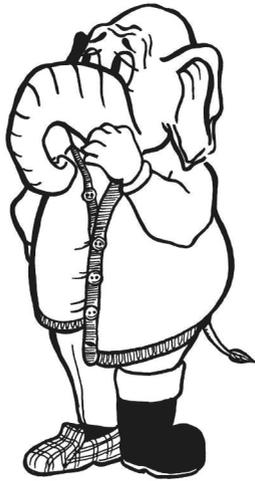


1 | What is dementia?



There are many different types of dementia. This chapter will give you information on what dementia is and on most types of dementia other than Alzheimer's disease. Because Alzheimer's disease is the most common type, we describe it separately in Chapter 2. In all types there is a decline in mental function, especially memory, and disturbance of daily and social activities.

DEFINING DEMENTIA

What is dementia?

'Dementia' is a term used to describe various different brain disorders that have in common loss of brain function, which is usually progressive and eventually severe. There are over 100

different types of dementia. The most common are Alzheimer's disease, vascular dementia and dementia with Lewy bodies (sometimes abbreviated to DLB). The different types are discussed further later in this chapter.

People with dementia usually have particular problems with their short-term memory. They often forget things that they have just said or done, even though they can often clearly recall events that happened many years ago. Their sense of time and place is frequently disturbed as dementia progresses. They may develop problems with finding words, and it becomes increasingly difficult for them to learn new information and to do new things. As time goes on, people with dementia are likely to need help to do even the most basic everyday tasks, including washing, dressing and eating. Eventually, people with dementia may become incontinent and sometimes there can be severe behavioural and communication problems. Most people with dementia eventually require 24-hour care, either at home or in supported accommodation. Whatever the type of dementia (see below), the disease often goes on for many years, and sometimes people die of something else.

My family tell me that my grandmother became 'senile' before she died. Does this mean she had senile dementia or could she have had Alzheimer's disease?

People quite often use the term 'senile' to describe older people who have become confused in their thoughts. Strictly speaking, the word 'senile' just means old, but people have often used it in everyday terms to mean senile dementia (an unhelpful term that should not be used).

If your grandmother's 'senility' lasted for months or years before she died, it is likely that she did have some form of dementia, and the most common form of dementia is Alzheimer's disease (see Chapter 2). However, if your grandmother's mental problems developed only in the last weeks or months of her life, it may be that she did not have dementia but that her brain function was being affected by a disease elsewhere in her body, such as the liver, kidneys or heart.

As people get older they tend to become more forgetful, but this is sometimes part of the natural ageing process rather than dementia (see the section ‘Symptoms and signs’ in Chapter 2 for information on how Alzheimer’s disease and other dementias differ from ordinary forgetfulness). Some people refer to any older person who is a bit forgetful as being senile, but this term has unhelpful and even unpleasant connotations so shouldn’t be used to describe dementia.

I’m 52 and have been told I have early-onset Alzheimer’s disease. What is this? Is it the same as ‘pre-senile dementia’?

‘Pre-senile dementia’ is a term that sometimes still appears in textbooks and other sources of information but it should not be used. In the past, the distinction tended to be made around the age of 65 and ‘senile dementia’ was used as another term for dementia appearing in older people. People described as having pre-senile dementia and senile dementia in fact probably had Alzheimer’s disease or some other form of dementia.

At 52 you are very young to be diagnosed with dementia. In fact, fewer than 2 in 100 cases of dementia occur in people under 65, so it is quite possible that, in the past, your dementia would have been referred to as ‘pre-senile’.

Having dementia so young can bring its own set of problems because people may still be at work and have young children. Another problem is that many areas of the UK do not have specific services for younger people with dementia. Although Alzheimer’s is the most common cause of dementia in people under 65, there are many other, less common, causes which may require specialist assessment. For this reason, if you haven’t already done so, it may be best for you to have an opinion from a neurologist or psychiatrist specialising in this field.

I have been told that my next-door neighbour has dementia but that she does not have Alzheimer's disease. What other sorts of dementia are there?

There are a number of different types of dementia. All of these affect the brain and cause a progressive loss of memory and other brain functions that may eventually make it impossible for the affected person to perform even the simplest everyday tasks without help.

Alzheimer's disease (see Chapter 2) is the most common type of dementia, accounting on its own for over half of all cases. There are, however, quite a number of other types of dementia and it seems that your neighbour has been diagnosed with one of these. Among these other types of dementia, all but vascular and dementia with Lewy bodies are rare.

Types of dementia other than Alzheimer's disease include:

- vascular dementia, which is usually the result of brain damage due to tiny strokes;
- dementia with Lewy bodies, which has some features in common with Parkinson's disease;
- fronto-temporal dementia, for example Pick's disease, in which there are often striking changes in behaviour before the memory problems appear;
- Huntington's disease, also sometimes called Huntington's chorea, which is characterised by jerky movements in addition to dementia;
- AIDS-related dementia;
- dementia that sometimes occurs together with Parkinson's disease;
- Creutzfeldt–Jakob disease (CJD);
- dementia due to a brain tumour;

- normal pressure hydrocephalus, due to a build-up of fluid in the brain;
- dementia due to an excessive intake of alcohol over an extended period of time;
- dementias due to various treatable causes, including vitamin deficiency, hormone deficiency and syphilis.

These different types are discussed later in this chapter.

WHO WILL GET DEMENTIA?

How common is dementia, and is it more common in some groups of people than others?

The chances of developing some form of dementia increase with age, but dementia does occur very rarely in people under the age of 60. Over the age of 65, dementia affects approximately 6 people in 100. For people over 80, the number affected rises to around 20 in 100. Similar rates are seen in other countries across Europe. It is estimated that there are currently about 800,000 people with dementia in the UK, and this is projected to rise to 1.7 million by 2051.

In general, dementia seems to affect all groups in society equally. It is not known to be particularly linked with gender, social class, ethnic group or geographic location.

CAUSES OF DEMENTIA

Can stress or worry cause dementia?

There is no evidence that stress or worry is responsible for causing dementia. However, stress or worry can lead to forgetfulness and confusion that may sometimes be mistaken for early dementia. It is also true that a diagnosis of dementia is sometimes made only after a

period of stress or worry has made the disease more apparent, even though the dementia has in fact been present for some time. Anxiety is a common symptom of depression in older people, and the presence of depression can result in severe memory problems that may be mistaken for dementia. Also, of course, the symptoms of dementia may cause stress and worry for people experiencing them, as well as for those around them.

My husband has been told that he has some sort of dementia and that he should give up smoking. Can smoking cause dementia?

Smoking is not thought to be a direct cause of dementia, but it can contribute to heart disease and atherosclerosis (narrowing of the arteries), which often leads to strokes. One form of dementia, called vascular dementia, is often caused by strokes, which cause brain damage by cutting off the blood supply to areas of the brain. Some research has also found that smoking may be one of many risk factors for developing Alzheimer's disease.

Not all doctors agree that giving up smoking is likely to have much effect on the course of dementia once this disease is already apparent, and you may find that your husband has difficulty stopping smoking. However, stopping smoking can be recommended on many other health grounds, and may help to prevent strokes which could make his dementia worse. A lot of help is now available for people who want to stop smoking. Your husband's GP will be able to help or you could ask Quit – a charity dedicated to help people stop smoking – for advice (contact details in Appendix 1).

My father, who is 72, has recently had an operation on his bowel. He was a bit forgetful before he went into hospital but now he is home again he is very confused. Might the operation have given him dementia?

Some people, particularly the elderly, may have temporary confusion after an operation. The after-effects of the anaesthetic and taking pain-relieving drugs may increase confusion for a short time.

Confusion may also be made worse by having to cope with the unfamiliar environment of the hospital.

If your father's confusion persists, it is possible that he already had dementia before the operation, which has made it worse. Rarely, dementia can be triggered by an operation: during an operation; older people may be vulnerable to having little strokes or temporary problems with the blood supply to the brain, which can cause lasting confusion. We still do not understand much of the link between surgery and subsequent memory problems.

Another possible explanation for your father's increased confusion is that he has picked up a chest or urinary tract infection. This possibility is less likely if the operation was some time ago. However, do speak to his doctor about your concerns, as treatment of an infection or a change in your father's medication can sometimes bring about a dramatic improvement.

Can head injuries cause dementia?

Serious head injuries can cause memory and other difficulties but these do not usually get worse, unlike dementia. Some people who suffer severe or repeated head injuries, however, may be at increased risk of developing dementia. It is possible that a head injury may trigger the disease process in susceptible individuals. People who have sustained serious head injuries through boxing are prone to a type of dementia known as 'dementia pugilistica', which is similar to Alzheimer's disease.

TYPES OF DEMENTIA

What are the types of dementia?

There are many different types of dementia, with different possible causes. In many cases, the causes are not fully understood. For example, dementia in general is much more common in older people,

but the ageing process as such is not considered to be an actual cause of dementia.

A few types of dementia, such as Pick's disease and Huntington's disease (see later in this section) and rare cases of Alzheimer's disease (see Chapter 2), are known to be inherited, passed on from one generation to the next in the genes. Other types of dementia, including most cases of Alzheimer's disease and also vascular dementia, are thought to be caused by a combination of genetic and other factors (sometimes called 'environmental' factors).

Some types of dementia occur as a feature of some other disease, such as AIDS, Parkinson's disease and syphilis. Long-term heavy alcohol consumption can also cause dementia.

What is vascular dementia?

Vascular dementia (which used to be known as multi-infarct dementia) is caused by insufficient blood supply to the brain. It is the second most common type of dementia in the UK, after Alzheimer's disease. Vascular dementia is estimated to account on its own for about one in five dementia cases, and to occur with Alzheimer's disease in another one in five cases. Some doctors and researchers now believe that the two types of dementia occur together much more often than previously thought.

There are various causes of vascular dementia, the most common probably being a series of 'mini strokes' (known as infarcts). A stroke destroys an area of cells in the brain by cutting off the blood supply. A stroke can be so slight that it causes no immediate symptoms, or may just cause a brief spell of dizziness, weakness or confusion. Eventually this damage accumulates sufficiently to cause dementia.

Strokes may occur because of high blood pressure, which can cause blood vessels in the brain to burst, or because of blood clots, which can cause blockages in the vessels supplying blood to the brain. People who smoke, have high blood pressure or diabetes or are overweight seem to be at increased risk of strokes.

Even though the damage caused by an individual stroke may be slight, the cumulative effect is often sufficient to cause dementia.

Sometimes people can get dementia after a single stroke if it affects a crucial part of the brain. Some people develop dementia because the blood supply to the whole brain is reduced for a short period of time; for example, because of a heart attack or during an operation. The brain only needs to have a poor blood supply for a couple of minutes for lasting damage to result.

The main symptoms of vascular dementia, like most other dementias, are loss of short-term memory, loss of sense of time and a progressive decline in other abilities. However, in vascular dementia, the memory loss is typically much more variable than in Alzheimer's disease, and people with this type of dementia can be much better some days than others. There may be long periods when a person's memory loss does not seem to get worse, and can even improve; and then there may be an episode of acute confusion (often associated with a new mini stroke) followed by a step down in the person's memory. Doctors sometimes describe vascular dementia as having a 'step-like' progression.

Another feature of vascular dementia is that people may have a greater degree of awareness of their difficulties than is the case in Alzheimer's disease. There may be an increased likelihood of problems with unpredictable behaviour or changeable emotions.

My father never used to use bad language and now he swears all the time. I've been told he has 'frontal lobe symptoms'. What does this mean?

The 'frontal lobe' refers to the front area of the brain, just behind the eyes, and there are two of them (one left, and one right). If the frontal lobes stop working properly, different symptoms can emerge, including:

- changes in personality; people can become more placid or, occasionally, more aggressive or irritable;
- loss of motivation;

- people just lose their 'get up and go' and become more uninterested in things. This may also happen in depression, Parkinson's disease and other conditions;
- loss of the ability to plan and organise;
- loss of 'restraint' (disinhibition) in that some people may swear, say rude things, laugh inappropriately or become sexually disinhibited.

There is no fixed pattern of symptoms. People with this condition may have any combination of the typical symptoms, and these may change over time. Eventually, any type of dementia can cause frontal lobe symptoms but some types of dementia (for example, Pick's disease – see the next answer) usually begin with frontal symptoms.

It seems that my mother has a type of dementia called fronto-temporal dementia. What is this? How does it differ from Alzheimer's disease?

Fronto-temporal dementia (one type of which is called Pick's disease, after the doctor who first described it) is a rare type of dementia. It has various similarities with Alzheimer's disease (see Chapter 2), but also differs from it in a number of important respects.

Fronto-temporal dementia typically develops at an earlier age than Alzheimer's disease, usually in a person's 40s or 50s. It is thought to have a genetic cause in some families, but it can also occur when there is no family history of this form of dementia.

The most important difference between fronto-temporal dementia and Alzheimer's disease is that the brain tissue changes that occur in fronto-temporal dementia typically affect mainly the front part of the brain (also known as the frontal lobes). It causes marked shrinkage of these lobes while leaving other parts of the brain relatively unaffected.

As with Alzheimer's disease, a definite diagnosis of fronto-temporal dementia disease can be made only at a post-mortem examination (autopsy). However, in practice, the diagnosis can often be made from

a person's symptoms and signs. If a doctor suspects fronto-temporal dementia, he or she will sometimes ask a psychologist to do specific tests that can help the diagnosis.

Many of the early symptoms of fronto-temporal dementia are 'frontal lobe symptoms' (see the previous answer). These can be very subtle in the early stages of the illness, and it may take many months, or even years, for anyone, including doctors, to suspect that the person has dementia.

Another feature of fronto-temporal dementia is that some people with this condition may start by having problems with language. This is because the area of the brain that is involved in creating speech is in the frontal lobe. It may seem like the person can't find the right words, or no longer understands words that were previously familiar to them (known as 'semantic dementia') or some people may progressively lose their speech altogether (known as 'primary progressive aphasia').

Similar to people with Alzheimer's disease, someone with fronto-temporal dementia is likely to experience a progressive decline in abilities.

My boyfriend says he doesn't want to get serious with me because there is Huntington's disease in his family. What does this mean?

Huntington's disease is a relatively rare form of dementia in which mental deterioration is accompanied by problems in controlling body movements. Huntington's disease is an inherited disease. If your boyfriend's mother or father has the disease, there is a one in two chance that he (and also any of his brothers or sisters) will also develop it.

People at risk of developing Huntington's disease can have genetic counselling and then a genetic test to find out whether or not they have inherited the abnormal gene that will give them the disease. However, it is important to respect a person's wishes if he or she prefers not to find out this information in advance.

Huntington's disease usually becomes apparent when someone who has inherited the disease reaches his or her 30s or 40s, although symptoms can start sooner. The illness may begin with either mental or physical symptoms. Progressive loss of memory and concentration, leading to severe dementia, may be accompanied by anxiety, irritability and depression. The movement problems take the form of twitching and spasms of the muscles. There is no specific treatment for Huntington's disease, but drugs do sometimes help the movement problems. The illness typically lasts for 10 to 25 years, leading to severe disability. As the disease progresses, 24-hour nursing care will almost certainly be needed.

Do most people with HIV develop dementia?

Most people infected with HIV – human immunodeficiency virus, the virus that leads to AIDS – do not develop dementia. However, there is a possibility that someone with HIV may develop subtle memory problems before developing late-stage HIV infection.

Some people (probably less than 5%) who have advanced HIV infection develop severe dementia, which is often characterised by apathy and problems with clumsiness and walking. In some people with advanced HIV infection the dementia that accompanies it is due to a direct effect of the HIV virus on the brain. In others, the dementia is due to an infection or a tumour of the brain that develops because of lowered immunity due to HIV.

There are now treatments – called anti-retroviral therapy – for people with HIV infection which may help to reverse some of the symptoms of dementia. The treatment can be complicated and involves several tablets taken at different times of the day. For this reason it is important to make sure that people who have memory problems associated with HIV infection are able to take their medications properly or have help to do this.

My father has had Parkinson's disease for three years now and seems to be getting quite forgetful. I have heard that people with Parkinson's sometimes develop dementia – is this true?

It is true that people with Parkinson's disease do have an increased risk of developing dementia, usually starting at least two years after the Parkinson's disease begins. This is known as 'Parkinson's disease dementia' and affects an estimated 15–20% of people with Parkinson's disease. However, the fact that your father is showing signs of forgetfulness does not necessarily mean that he is developing dementia. It is possible that your father's memory is working perfectly well and that he only appears to have a memory problem because he is slowed down by the Parkinson's disease. Sometimes this slowness not only affects the body but can also slow down people's thoughts; this is known as 'bradyphrenia'.

Even if he does have a memory problem, this may not be due to dementia. For example, people who have Parkinson's disease are often treated with medicines belonging to a drug group known as anticholinergics (see Glossary). These drugs can reduce the symptoms of Parkinson's disease but they also sometimes make someone's memory worse. Also, people who have Parkinson's disease sometimes become depressed. Depression is a common cause of poor memory, which is sometimes mistaken for dementia (see Chapter 3 for more information on depression and dementia).

Some people develop symptoms of Parkinson's disease at around the same time as they show signs of dementia. If this happens, they are more likely to have a type of dementia called dementia with Lewy bodies (see the next answer for more information).

I thought my husband had Alzheimer's disease but the specialist says that it is dementia with Lewy bodies. What are the differences? Will the fact that my husband has dementia with Lewy bodies affect how I can look after him?

Dementia with Lewy bodies is a form of dementia that is similar to Alzheimer's disease. It takes its name from abnormal collections

of protein, known as Lewy bodies, that occur in the nerve cells of the brain.

Your husband may have symptoms and signs similar to those of someone with Parkinson's disease, such as having a tremor, being somewhat unsteady on his feet and being rather slowed down. He may also be experiencing visual hallucinations (see the section 'Hallucinations' in Chapter 7) and may have started to have falls. Some people with dementia with Lewy bodies have more confusion at night. It is also likely that your husband's condition will vary from day to day, and that on some days he will have a short episode of being very confused, which will then settle. This pattern is typical of dementia with Lewy bodies but is not common in Alzheimer's disease.

In terms of care, your husband's needs will be much the same as if he had been found to have Alzheimer's disease. The main difference is that you may find it more difficult to predict how he is going to feel on a day-to-day basis compared with someone with Alzheimer's disease.

People with dementia with Lewy bodies are very sensitive to tranquillisers known as antipsychotic or neuroleptic drugs, and for this reason use of these drugs should be avoided if at all possible, although there is now evidence that they are probably best avoided in all types of dementia.

My wife, who is 75, started to lose her memory about ten years ago and five years ago was diagnosed with Alzheimer's disease. We have both eaten a lot of beef. Do you think she might have CJD not Alzheimer's?

Your wife is very unlikely to have CJD (Creutzfeldt–Jakob disease). CJD is an extremely rare type of dementia that affects only one in a million people in the UK, whereas Alzheimer's disease (see Chapter 2) affects around one in ten people of your wife's age. However, the main reason for thinking that your wife is unlikely to have CJD is that her illness has now lasted for a number of years. CJD usually progresses very rapidly and is usually fatal within a year.

Many of the early signs of CJD are similar to Alzheimer's disease but there are some differences. People with CJD may become rather withdrawn and forgetful, and soon develop problems in finding the right words and having a conversation. They also become unsteady on their feet and frequently have spasms or jerky movements in their arms and legs.

You may remember that in the 1990s there was a lot of publicity about a link between CJD and BSE (bovine spongiform encephalopathy), a similar disease that affects cows. CJD, BSE and a disease called scrapie (in sheep) are all known to be caused by an unusual infectious particle called a prion. In recent years a number of cases of CJD have occurred in mainly younger people. These cases are thought to be linked to eating beef from cows infected with BSE. Their illness is now considered to be a distinctive new form of CJD known as variant CJD (vCJD). No link has been found between beef eating and the more usual forms of CJD, which typically develops in old age. (More information is available from the CJD Support Network, which provides help for anyone affected by any strain of CJD; see Appendix 1 for contact details.)

Might dementia be due to a brain tumour?

Brain tumours are a rare cause of dementia. One type of slow-growing brain tumour, known as a meningioma, sometimes causes symptoms of dementia. Most brain tumours cause other kinds of symptoms, such as loss of limb function, visual disturbance and loss of balance. In some cases, removal of a meningioma may result in recovery from the dementia.

Can 'water on the brain' cause dementia?

The rare type of dementia known as 'normal pressure hydrocephalus' is due not to water but to an excess of the fluid that bathes the brain, known as cerebrospinal fluid.

Early symptoms of this type of dementia include incontinence of urine and problems with walking. If your doctor suspects this type of

dementia, he or she may order a brain scan to confirm the diagnosis. This type of dementia may be helped by an operation (see the section 'Surgery' in Chapter 12).

Can hormone deficiencies cause dementia?

Under-activity of the thyroid gland (a gland in the neck that regulates our metabolism) can result in a condition called 'hypothyroidism', of which dementia can be a symptom. People with hypothyroidism typically gain weight and develop a hoarse voice, dry skin and thinning hair. Hypothyroidism can be treated with replacement thyroid hormones. Some other hormone deficiencies are very rare causes of dementia.

Does drinking too much alcohol cause dementia?

People who drink too much alcohol over a prolonged period of time may develop dementia in addition to many other health problems.

Some heavy drinkers have a specific problem of loss of short-term memory, known as Korsakoff's syndrome (or sometimes called Korsakoff's psychosis), which develops because of vitamin B₁ (thiamine) deficiency. This results in difficulty in learning new information, although old memories and other aspects of brain function are often preserved. Others develop a wider range of problems that resemble Alzheimer's disease. Drinking moderate amounts of alcohol is probably safe, and some researchers have suggested that small amounts of red wine (which contains anti-oxidants) may actually reduce the chance of developing dementia.

Can dietary deficiencies cause dementia? My wife has been a vegetarian for many years and now has dementia. Could her diet be to blame?

It is true that dietary deficiencies are a rare cause of dementia. However, it is very unlikely that your wife's dementia has been

caused by her vegetarianism. Your wife is much more likely to have one of the more common types of dementia, such as Alzheimer's disease.

Deficiencies of some vitamins, such as vitamin B₁₂ or B₁ (thiamine), have been suggested as rare causes of dementia. These vitamins are present in a wide range of foods and can be stored by the body for long periods, and it is uncommon for anyone to develop a deficiency of either of these vitamins.

People who drink excessive amounts of alcohol over a long period of time sometimes develop a deficiency of vitamin B₁, leading to a condition called Korsakoff's syndrome (discussed in the previous answer). People on a vegan diet, which excludes not only meat but also eggs and milk, may need supplementary vitamin B₁₂.

A few people develop a deficiency of vitamin B₁₂ because their bodies do not absorb this vitamin properly. This is usually because of a condition called 'pernicious anaemia' – due either to a rare stomach problem, known as 'intrinsic factor deficiency', or to previous surgery to the bowel.

Although dementia due to vitamin deficiency is rare, your wife's doctor will probably have already carried out a blood test to exclude this possibility. In cases where a vitamin B₁₂ deficiency is found, the usual treatment is to give the vitamin by injection every three months.