RARE DEMENTIA SUPPORT

INFORMATION BOOKLET

Providing information on

Frontotemporal Dementia (FTD)

Behavioural variant FTD (bvFTD)

Primary Progressive Aphasia (PPA)

(semantic dementia, logopenic aphasia and progressive non-fluent aphasia)

Rare Dementia Support runs specialist support group services for individuals living with, or affected by, a rare dementia diagnosis. Our vision is for all individuals with or at risk of one of these forms of dementia to have access to information, support and contact with others affected by similar conditions.

Rare Dementia Support is a fund held by The National Brain Appeal (registered charity number: 290173).

Foreword

Rare Dementia Support (RDS) runs specialist support group services for individuals living with, or affected by several rare dementia diagnoses. Our vision is for all individuals with or at risk of frontotemporal dementia (FTD) to have access to information, support and contact with others affected by similar conditions.

Rare Dementia Support seeks to:

- Provide access to reliable disease specific information
- Facilitate contact between people with similar diagnoses and those who live with and care about them
- To represent the views of people affected by rare dementia diagnoses across a range of strategy and policy influencing platforms

This booklet brings together information that we hope will be of use to people embracing a range of FTD diagnoses. There are many symptoms described in this booklet which fall under the spectrum of an FTD diagnosis. Not everyone will experience every symptom listed in this booklet.

Contents

Medical Information

Clinical Information and Management

General Management

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Medical Information

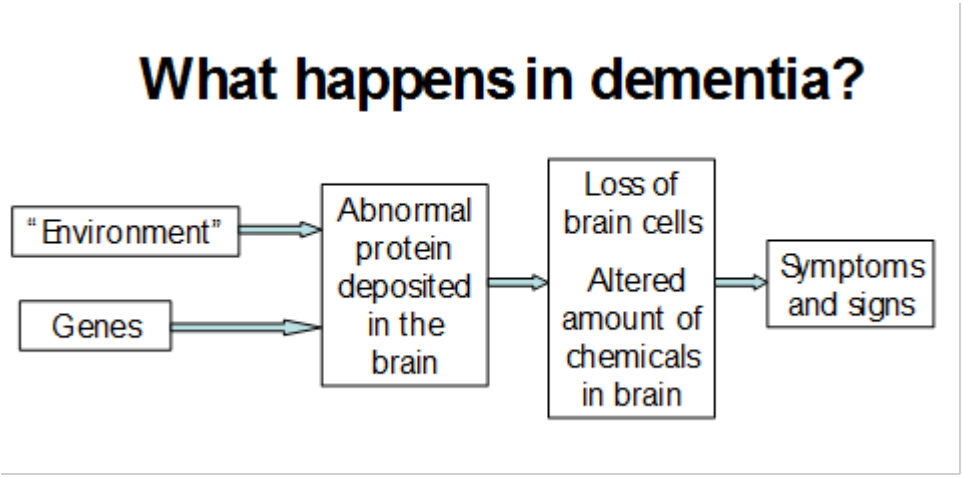
Introduction to Dementia

"Dementia" is a term for progressive problems in thinking or behaviour – this may be:

- Memory, the main problem in Alzheimer's disease
- Behaviour, the main problem in behavioural variant frontotemporal dementia
- Language, the main problem in semantic dementia and progressive non-fluent aphasia (primary progressive aphasia is a term used to describe dementias where loss of speech and language abilities are the leading and most prominent problems)
- Other things, such as arithmetic, planning and problem solving

Different problems are seen in different dementias – some dementias affect the whole brain, such as Alzheimer's disease, whereas others appear to affect only parts of the brain, for example the area for speech production in progressive non-fluent aphasia.

The most common form of dementia is Alzheimer's disease and this accounts for about 60-70% of all cases. Vascular dementia caused by damage to the blood vessels in the brain is probably the next most common cause. The 3rd and 4th most common causes are frontotemporal dementia (which includes behavioural variant frontotemporal dementia and primary progressive aphasia) and Dementia with Lewy bodies. In total these four causes account for about 95% of all cases of dementia. There are many rare causes of dementia and these account for the final 5% of cases.



The diagram above is a simple way of thinking about what is happening in people's brains as they develop dementia as a result of a neurodegenerative disease such as Alzheimer's disease or FTD.

- For some people an abnormal gene triggers the disease (see the section "Is Dementia Inherited?" for more information). This is the case in up to 40% of people with frontotemporal dementia but only a very small amount (less than 1%) of people with Alzheimer's disease.
- However, for the people who do not have an abnormal gene a mixture of things probably trigger the disease – a mix of something from the "environment" and a number of other genes. We currently know very little about what the triggers in the "environment" are but these are likely to include problems such as high blood pressure and diabetes.
- Once the disease is triggered, abnormal proteins become deposited in the brain. In frontotemporal dementia, these are proteins known as tau, TDP-43 and FUS, in Alzheimer's disease it is tau and amyloid and in dementia with Lewy bodies it is a protein called alpha-synuclein. These abnormal proteins are pathologists use to make a definitive diagnosis when they examine the brain after death.

These abnormal proteins cause brain cells to die, and so brain cells from certain areas of the brain are slowly lost. In frontotemporal dementia the cells are particularly lost from the frontal and temporal lobes of the brain while in Alzheimer's disease cells are typically first lost from memory areas but over time involves the whole brain.

Once cells are lost, the amount of chemicals in the brain becomes lower. Brain cells (also called neurons) produce these brain chemicals or "transmitters" are use them to pass signals to pass from one brain cell to another. For example, in Alzheimer's disease there is less of a chemical called acetylcholine. The aim of drugs such as Donepezil (ARICEPT), Rivastigmine (EXELON) and Galantamine (REMINYL) is to try to increase the amount of this chemical. Loss of cells and decreased amount of chemicals cause the symptoms and signs of the disease, which are different in different types of dementia.

Research in dementia takes place all over the world and more and more is becoming known about each of the different types of dementia. As our knowledge continues to increase, the hope is that we will be able to design treatments that will help combat the symptoms and signs of dementia, and eventually hopefully cure it.

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Frontotemporal Dementia (FTD)

Frontotemporal dementia (FTD) is a group of conditions caused by loss of cells mainly in the frontal and temporal lobes of the brain. The main symptoms are a progressive change in personality and behaviour and/or progressive deterioration in language abilities.

It can affect both men and women and usually starts in the 40's, 50's or 60's. However it can also affect older people, and rarely, even younger people.

It was originally described by Arnold Pick in 1892 after whom it was named. Most doctors now prefer to reserve the name 'Pick's disease' for just one of the types of changes in the brain tissue that may be seen with the disease.

FTD is classically subdivided into two types which are described in more detail in this booklet:

1. Behavioural variant FTD (bvFTD)
2. Primary progressive aphasia (PPA), which includes progressive nonfluent aphasia (PNFA), semantic dementia (SD) and logopenic aphasia (LPA)

What causes FTD?

FTD is caused by loss of brain cells in the frontal and temporal lobes of the brain. However, the processes that lead to the loss of these brain cells are not well understood. There are a number of different underlying pathological changes that are recognised and several different pathological varieties of FTD. It is known that there is an abnormal accumulation in the brain cells of certain proteins (these include the proteins known as 'TDP-43', 'FUS' and 'Tau') but we do not yet fully understand why this accumulation occurs, or how it leads to the loss of cells.

In somewhere between 30 and 40% of cases a person with the disease may have a family history of the disease in one of their parents, brothers or sisters. In these cases the cause is more likely to be genetic. Problems in the tau (or MAPT) gene, progranulin (or GRN) gene or a gene called C9ORF72 are the cause in some of these cases, but not all abnormal genes have been discovered yet. Genetic testing is available in some centres. Other genes known as VCP, TARDBP, FUS and CHMP2B are extremely rare causes of FTD. Testing for these is not generally clinically available.

Links to other diseases

In a small number of patients FTD can overlap with one of a number of diseases that affect movement of the body: motor neurone disease (MND, sometimes called amyotrophic lateral sclerosis, ALS), progressive supranuclear palsy (PSP) or corticobasal degeneration (CBD). Symptoms of MND can include weakness of the limbs or problems with swallowing. PSP causes problems with movements of the eyes as well as problems with thinking and

behaviour. There are often other physical problems such as falls, difficulty walking and stiffness. Some patients with FTD will later develop symptoms similar to Parkinson's disease such as slowing of movements, tremor and stiffness of the limbs.

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Behavioural variant frontotemporal dementia (bvFTD)

Behavioural variant (bvFTD) is caused by loss of brain cells mainly affecting the frontal and temporal lobes of the brain. These areas control behaviour, personality and complex thinking such as planning or problem-solving.

Symptoms

The first symptom is usually a change in personality or behaviour (which is out of character for the person) – the symptoms may come on very slowly and not be noticed as definitely abnormal at first.

The symptoms include the following:

- Loss of inhibitions or increased extroversion. The person may talk to strangers, make inappropriate remarks in public or be rude or impatient. They may also become aggressive.
- They may spend money excessively.
- Apathy or withdrawal from social activities.
- Loss of empathy.
- Changes in sexual behaviour: either more/less or inappropriate interest.
- People may be very easily distracted.
- They often develop fixed routines or become obsessive about things, particularly time ('clock watching'). Some people begin to hoard things.
- People may also develop a sweet tooth or a preference for unusual foods. They may also overeat leading to a gain in weight or drink excessive amounts of alcohol. In the later stages people with the illness may compulsively put objects in their mouths.
- Decreased amount of speech or repetitive speech.
- Often the person will be unaware of the true extent of the problems and lack insight.

In the early stages memory is often well maintained on psychological testing (unlike in Alzheimer's disease) but difficulties in organisation and concentration often lead to an apparent memory problem in daily life and so this is also a common complaint.

Medical tests

Behaviour and aspects of thinking (cognitive functions) will be assessed, initially by a doctor, and often followed by a more detailed assessment by a psychologist. Brain scans can show the loss of brain cells in FTD (shrinkage of the affected parts of the brain) but there is no single test that can specifically diagnose FTD with complete reliability during a person's lifetime. Furthermore, in the early stages of the disease the scan may look normal. Diagnosis

is therefore largely based on clinical judgment and FTD can be confused with other disorders in which there are problems with behaviour (e.g. some psychiatric disorders) and with other dementias. The doctor will often arrange blood tests or other tests (usually including detailed brain scans, in particular MRI, and sometimes a lumbar puncture or other specialised tests) to help confirm the clinical diagnosis and rule out diseases that can produce similar symptoms to FTD.

Treatment

Unfortunately, there are no medications presently available which can treat the disorder or slow its progression. Treatment therefore focuses on helping people to manage their symptoms, including the behavioural symptoms and treating problems such as mood changes that may contribute to the difficulties that people experience. Medication for behavioural symptoms and mood changes may be needed as the disease progresses.

Prognosis

Behavioural and personality problems continue over time and other aspects of thinking may become affected. There is wide variation in the tempo of the disease between individuals, and some people have a slower form of bvFTD that progresses over a number of years (in some cases over ten). However, in the majority of patients behavioural problems continue to progress such that by around two to five years after the onset of symptoms people generally have problems carrying out their normal activities (particularly where these involve interactions with other people, such as working and driving) and will need extra care and support.

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Primary Progressive Aphasia (PPA)

The term Primary Progressive Aphasia (PPA) refers to a group of disorders in which people develop progressive loss of their language abilities.

PPA falls within the frontotemporal dementia (FTD) group of disorders caused by loss of cells in the frontal and temporal lobes of the brain.

Within PPA three main patterns of language loss are recognised:

1) Semantic dementia (SD): people gradually lose their knowledge of words
2) Progressive non-fluent aphasia (PNFA): people's speech becomes effortful and they might say the wrong word or use the wrong grammar
3) Logopenic aphasia (LPA): people start to develop pauses in their speech as they try to find the right word

Semantic Dementia (SD)

This may also be called the semantic variant of primary progressive aphasia.

Symptoms

The first symptoms of SD are usually problems with language. These may include:

- Difficulty finding the right word - often substituting another word or a vague term such as 'thing' instead of the specific word.
- Loss of knowledge of what words mean or what objects are for.
- People may talk about things in a vague or 'roundabout' manner – referred to as 'circumlocutory' speech.
- Difficulty understanding what other people are saying.
- Problems with reading and writing.

Problems with language increase over time. There is a slow but progressive loss of vocabulary and the ability to understand what people are saying. Speech becomes increasingly vague and the quantity of speech tends to diminish.

With progression of the disease, changes in personality are more common in SD than the other types of PPA. These may include the development of a sweet tooth or other changes in eating, becoming obsessive and disinhibited behaviour.

Later in the illness non-language functions become affected. In particular, the person may have increasing difficulty recognising familiar people or household items. There are also increasing problems carrying out normal daily living activities.

There is no single test that allows doctors to make a diagnosis of SD. Usually the diagnosis is made using a combination of clinical assessment, psychology testing and a brain scan. MRI scanning shows loss of brain cells in the temporal lobe, usually more on the left than the right.

Progressive Non-Fluent Aphasia (PNFA)

This may also be called the nonfluent or agrammatic variant of primary progressive aphasia.

Symptoms

The first symptoms of PNFA are usually difficulties producing speech. These may include:

Difficulty producing words - although the person knows what they want to say, speech may be effortful and words may come out distorted. This is due to difficulties in the co-ordination of the movements of speech and is sometimes called 'apraxia of speech'.

Difficulty organising words - the structure of the sentences may be affected with words being missed out and errors in the grammar.

Together these factors make the speech sound distorted, slow and hesitant and difficult to understand.

As the disease progresses, people may develop other problems including:

- Reading, writing and spelling may become more difficult.
- Understanding of language may become affected and the disease may spread to other areas of the brain causing problems such as changes in behaviour, organisational skills and memory.
- Some people with PNFA develop problems similar to Parkinson's disease such as a tremor, difficulty using their hands, or falls. These problems may be part of conditions called corticobasal degeneration (CBD) or progressive supranuclear palsy (PSP).

As with SD, there is no single test that allows doctors to make a diagnosis of PNFA. Usually the diagnosis is made using a combination of clinical assessment, psychology testing and a brain scan. MRI scanning shows loss of brain cells in the speech areas of the brain, particularly the frontal lobe on the left side.

In a small number of people, PNFA is caused by a genetic problem. The genes that are known to cause problems are called progranulin (or GRN) and C9orf72.

Logopenic Aphasia (LPA)

This may also be called the logopenic variant of Primary Progressive Aphasia.

Symptoms

- The main symptoms of LPA are:
- Difficulty finding the right word. Speech contains pauses where the person stops what they are saying as they try to find the right word.
- Speech may become slow and hesitant and the pronunciation of words may be affected.
- As the problem progresses other cognitive functions such as calculation and memory are affected.

There is no single test that allows doctors to make a diagnosis of LPA. Usually the diagnosis is made using a combination of clinical assessment, psychology testing and a brain scan. Brain scanning shows loss of brain cells in areas further towards the back of the brain than the other PPA subtypes, particularly the area where the temporal and parietal lobes meet.

Like the other forms of PPA, why LPA develops in some people is unclear. Unlike the other types of PPA, in the majority of people where brain tissue has been examined under the microscope, the same pathology as Alzheimer's disease has been found. LPA is therefore often also considered as an unusual form of early onset Alzheimer's disease.

Frequently Asked Questions

How is a diagnosis of PPA made?

At most specialist clinics you will have a neurological assessment usually followed by a neuropsychological assessment by an experienced team of cognitive neurologists and neuropsychologists. Although these initial assessments can point towards a diagnosis of PPA, an MRI scan of the brain will also help with the diagnosis. In some circumstances a lumbar puncture (spinal tap) may be performed.

Is there a treatment for PPA?

There is no cure for PPA at the moment despite research efforts around the world. However, speech therapy to help with communication strategies, particularly in PNFA, is useful. Co-existing depression or behavioural problems can be treated symptomatically.

What causes PPA?

The symptoms of PPA are caused by loss of brain cells in the frontal and temporal lobes of the brain. However, the processes that lead to this loss are not well understood. It is known that there is an abnormal accumulation in the brain cells of certain proteins (known as TDP-43, and tau). What makes these proteins deposit, why they occur in particular areas and how their deposition leads to brain cell damage is still not known.

Does PPA run in the family?

Some people with PNFA have a family history of the same condition or of frontotemporal dementia. Problems (mutations) in genes called progranulin and C9orf72 have been shown to be associated with PNFA. SD and LPA only rarely run in families.

What is the prognosis for people with PPA?

This is a difficult question to answer as there has been very little research into it and it can be extremely variable from person to person. However, we know that from the onset of symptoms many people will live over 10 years and for SD this can be over 15 years.

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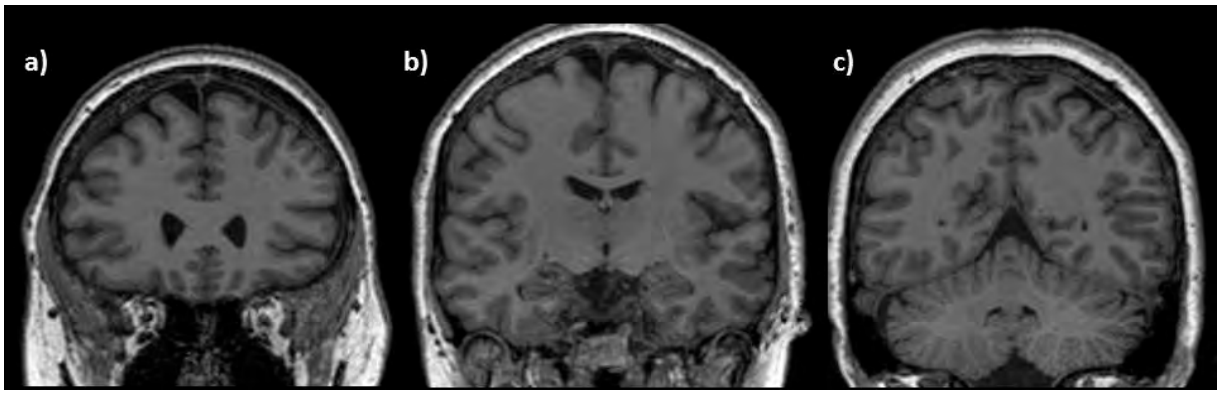
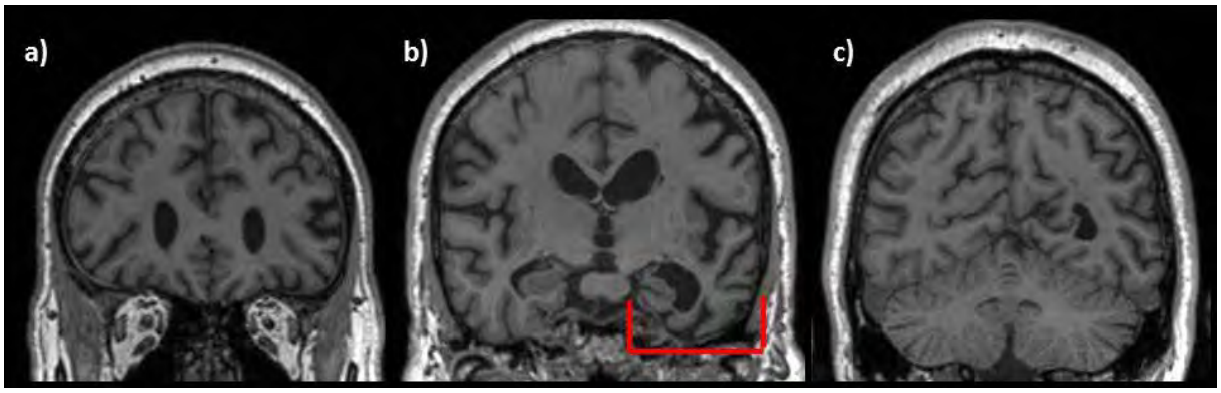
Brain Imaging

These images are MRI (magnetic resonance imaging) scans from four different individuals. They show a few sections throughout the brain starting from a) the anterior (frontal) regions, moving to b) about half way through the brain and finally c) more posteriorly (towards the back) of the brain.

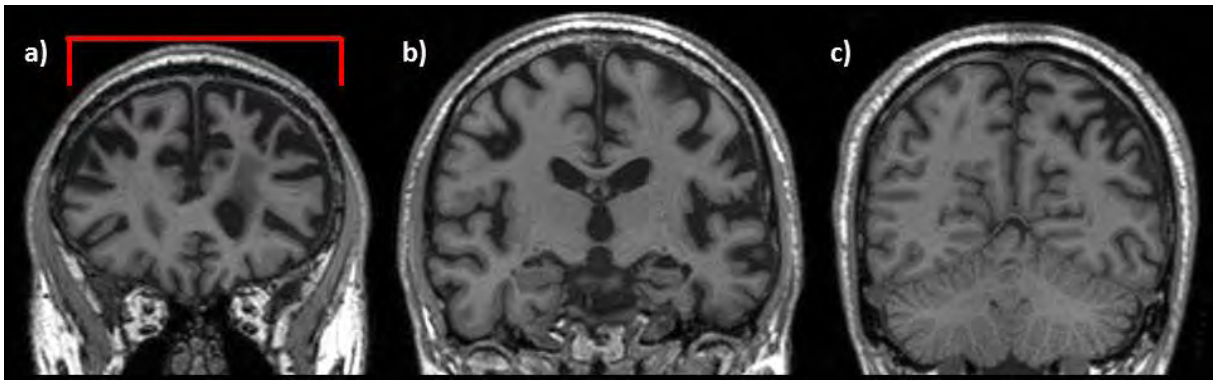
The first scan shows how the normal healthy brain of a 60 year old looks.



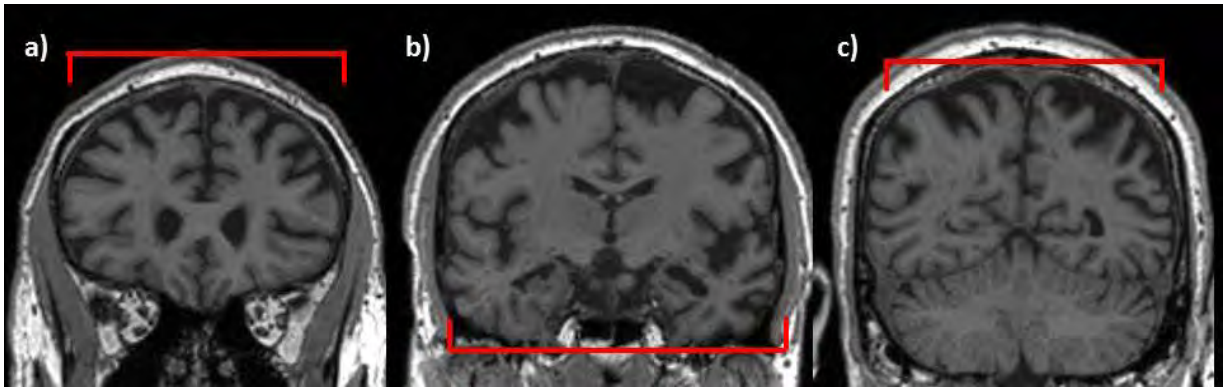
The second image series is taken from someone with semantic dementia. Note the more marked atrophy (shrinking, due to loss of brain cells) of the left hand side of the brain. This is particularly affecting the left temporal lobe (the bottom right of the picture – red bracket). This part of the brain is heavily involved in knowledge about things and is key for language processing in keeping with the symptoms seen with this disease.



The third set of images are taken from someone with a diagnosis of behavioural variant frontotemporal dementia (bvFTD). While there is more variability within this patient population in terms of atrophy (cell loss) pattern across the brain, in general, there is more pronounced atrophy in the anterior (front) part of the brain with the posterior (back) areas relatively more preserved. These frontal brain regions are involved in co-ordinating and monitoring complex behaviours, personality and planning, which are often primarily affected in bvFTD.



The final images show someone with Alzheimer's disease. There has been loss of cells all over the brain rather than the focal loss of cells in the frontal and/or temporal lobes in FTD.



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Clinical Information and Management

Is Dementia Inherited?

In most patients (50-70%), frontotemporal dementia (FTD) is a sporadic disease. This means that FTD occurs in an individual person by chance, without anyone else in the family having been affected. Relatives of a patient with sporadic disease have the same risk of developing FTD as the general population, i.e. they are not at increased risk.

However, in some cases FTD appears to run in families, suggesting there is a genetic component. The genetics of FTD are complex, and not yet fully understood, but research groups around the world are continuously advancing our knowledge with many new developments in recent years.

The largest groups of patients with a genetic component (20-40%) will have other family members who have also had a diagnosis of either FTD or a related neurodegenerative disorder, such as Alzheimer's disease or Motor Neuron Disease. However, the disease is not passed down from parent to child in these families. This type of dementia is termed familial, and these patients may carry genetic risk variants that increase the risk of developing disease but are not sufficient to cause FTD alone. The inheritance pattern in familial disease is complex and likely due to a combination of genes, lifestyle and environment. Some family members may carry the risk gene and remain completely unaffected. The genes that increase the risk of developing disease are only just starting to be discovered, and because the link between these genes and dementia is not clear there is currently no genetic testing available for risk genes.

In a smaller number of families (~10%), dementia is caused by a genetic fault. This type of dementia is called inherited dementia, and there is a clear family history of disease being passed from parent to child. Specifically, every patient will have an affected parent, and each child of an affected person will have a 50% chance of developing the disease. Several genes have been identified with genetic faults that cause FTD.

Understanding Inherited Dementia

A gene is like a set of instructions, and each gene instructs the cell how to make a particular protein. Every cell of our bodies contains two copies of every gene, one inherited from each parent. Genetic diseases can occur when there is a mistake (a mutation) in the gene, resulting in the production of a faulty protein that cannot carry out its normal function. Depending on the protein, and the nature of the mistake, one or both copies of the gene may need to be faulty to cause a disease.

All the mutations discovered so far which cause FTD are "autosomal dominant", meaning that only one faulty copy of the gene is needed to develop the disease. The risk of passing

the faulty copy to children in autosomal dominant disorders is 50% (1 in 2 chance) for each pregnancy. Typically, in inherited FTD the patients become affected at an earlier age and have a more severe form of disease than in sporadic patients. However, with some mutations, for reasons we don't fully understand, it is possible to have the gene and still not develop signs of the disease.

There are 3 main genes that have been identified that can cause Frontotemporal dementia in an autosomal dominant way: tau, progranulin and C9ORF72.

Understanding why changes in tau, progranulin and C9ORF72 cause dementia is the focus of research in laboratories around the world, because understanding how these proteins cause brain cells to die could be the key to developing new treatments for FTD. Some families may have genetic disease without carrying a mutation in tau, progranulin or C9ORF72 and research groups worldwide are also trying to identify new genes that with cause or increase risk for dementia.

Does it skip a generation?

There is usually a strong family history of the illness and you may know of cousins, aunts/uncles and grandparents who are affected. The disease may appear to skip a generation if a person with the faulty gene happened to die of another cause before the illness developed.

If I inherit the gene, will I get the disease?

If you have an abnormal gene, you will probably develop the disease if you live long enough. However, having the abnormal gene does not always predict when you will get symptoms, or how rapidly the disease may progress. A minority of people with an abnormal gene may never get symptoms.

Genetic counselling and testing

The medical team will ask about family history of similar illnesses. If they are concerned that the illness may be inherited, they can test for known mutations. Where tests are available, it often takes several months to get a result. The person's next of kin would be included in the counselling and testing process. Specific consent is needed to perform genetic tests.

If the precise mutation is not known, researchers may be interested in identifying it, and will need DNA from several affected family members. Identifying a new mutation takes many months or years and is not always possible.

If the precise mutation affecting someone is known, it is sometimes possible to test the person's adult children to see whether they have inherited it, to give an idea of whether they will develop the disease. Being tested is a very difficult decision with no right or wrong answer, and counselling and support is available throughout the process.

If you are thinking about finding out whether you have inherited a faulty gene, please ask your GP or hospital consultant to refer you to a Regional Genetics Clinic. It will be helpful to discuss this with the medical team caring for you first.

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Reviewed February 2017 by **Dr Selina Wray,** *UCL Institute of Neurology*

Questions Neuropsychologists Ask (and why they ask them)

Neuropsychology is the branch of psychology that deals with the relationship between the nervous system, especially the brain, and cerebral or mental functions such as language, memory, and perception. In other words, neuropsychologists are interested in how aspects of our behaviour and mental abilities alter following damage to or degeneration of the brain. By testing and measuring our behaviour and abilities, neuropsychologists try to make inferences about the nature, extent and location of the underlying brain cell loss or dysfunction. In this article we explore the motivation for the questions which neuropsychologists ask, and in particular consider the characteristic symptoms associated with damage to different parts of the brain. The role of the neuropsychologist. When someone presents to their doctor with concerns about their behaviour or mental abilities, there are four main ways in which a neuropsychologist might be able to contribute to the diagnosis, care and support which that person receives.

First, neuropsychologists work to identify and corroborate cognitive problems (difficulties with memory, language and other higher brain functions). The patient, their family and other healthcare professionals may be well aware that the person has a problem with their memory for example, but a neuropsychologist tries to measure that deficit and to place it on a quantitative scale using standardised tests.

Second, they assess whether the person meets established diagnostic criteria for a particular degenerative disease. For example, some criteria for Alzheimer's disease require a person to demonstrate evidence of impairments not only in memory but also in at least one other area such as language, perception or decision-making. Therefore it is important to assess not only the mental ability for which the person complains of difficulties, but also other skills in order to determine how focal or widespread the problem may be.

Third, particularly for patients with degenerative conditions, neuropsychologists attempt to measure change in cognitive abilities. Having detailed scores can be particularly important in the earliest stages of a disease, when the difficulties are relatively mild. It is sometimes not possible to determine from a single assessment whether someone's performance on a test at or below the level they would have achieved prior to their illness. So two or more assessments are required in order to determine whether their scores on a particular test stay the same or improve (as might be expected for a healthy individual) or decline (as might be expected for someone for a degenerative condition).

Fourth, neuropsychologists have a role to play in helping people with dementia and their families, friends and carers to better understand their difficulties. Sometimes understanding why and how certain problems or difficult behaviours occur can help the person with dementia or particularly those caring for them to accept and adapt to those problems better than when they remain unexplained. For example, people with Posterior Cortical Atrophy

(PCA), a rare form of Alzheimer's disease which affects the back of the brain causing problems with vision, might fail to pass something across the dinner table when asked, not because they are being deliberately unhelpful, but because they simply cannot perceive where the item they have been asked for is located. Similarly, someone with FTD might swear at a stranger in the street, not because they are being deliberately rude, but because the part of the brain which usually acts to inhibit such socially-unacceptable behaviour has been damaged.

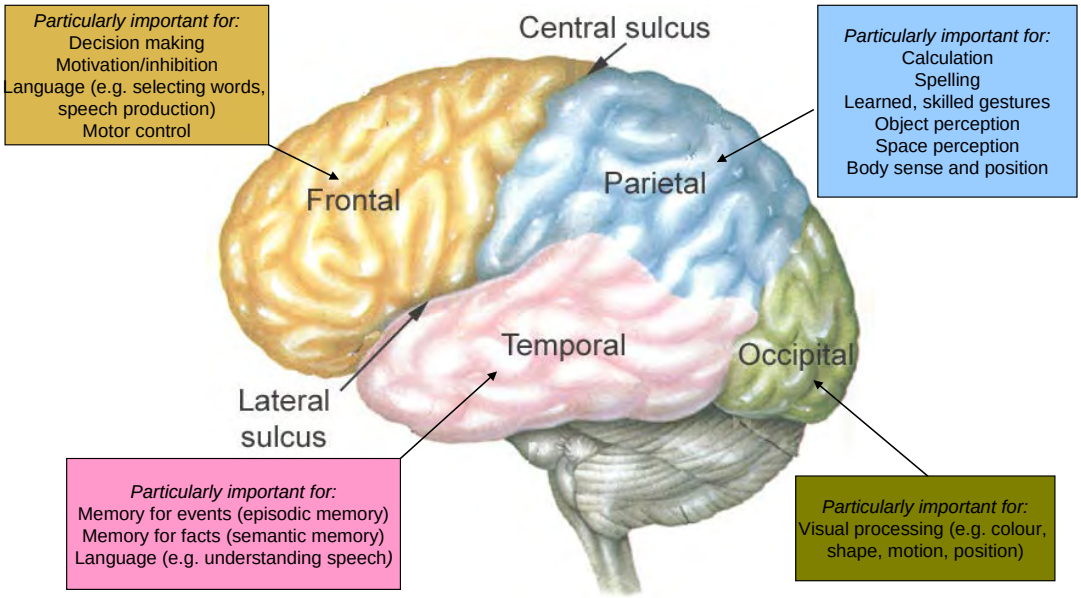

Figure 1. The lobes of the brain and their specialised functions

The specialisation of the brain

We look here at how the four lobes of the brain (shown in Figure 1) contribute to our experience of the world, starting at the back of the brain with the occipital lobes.

The occipital lobes

Although the eyes are the source of visual information about the world around us, it is actually the brain which does most of the hard work. The eyes convert sensory information about light into electrical impulses, but these are passed to the brain for interpretation. The occipital lobes are particularly important in the processing of form, colour, motion and location, skills which underpin the brain's ability to provide us with a complete 3D picture of the world. Therefore, the processing carried out by the occipital lobes enables the brain to perceive the identity of objects (e.g. faces, cars, animals) and to determine their location (e.g. on the left; far ahead). Tests for occipital lobe function include detecting or discriminating shapes and colours, and detecting patterns of coherent movement among

arrays of moving dots. Poor performance on tests sensitive to occipital lobe function are common in Alzheimer's disease but rare in FTLD.

The parietal lobes

The functions of the parietal lobe are somewhat more diverse, and there is a significant difference between the dominant side (the left in most people) and the non-dominant side. The dominant parietal lobe can be thought of as being concerned with things we have to put together into an order or structure. So tasks such as writing and spelling (which require putting letters and words together) and calculation (which involves ordering and combining numbers) are critically dependent on the dominant parietal lobe. This side of the parietal lobe has also been heavily implicated in a condition known as apraxia, an impairment of learned purposive movements, which is tested for by asking people to imitate or pantomime gestures and movements. The non-dominant parietal lobe could be thought of as our '3D centre'. One function of this area is to combine visual information from the occipital lobes into a 3D representation of the object being viewed. Damage to this area leads to a symptom known as visual agnosia, an inability to recognise objects, faces or surroundings. This is the reason the neuropsychologists often ask people to try to identify pictures and degraded or distorted images. The parietal lobes also contribute to our understanding of space, both in terms of our sense of body and personal space (e.g. knowing where our hand is relative to our body), and in terms of calculating the location of objects in external space (e.g. when we are reaching to pick something up). Spatial skills are often tested by asking people to count dots, arrange blocks in a particular order or to perceive which of two squares has a dot exactly in the centre.

Figure 2. Example tests of parietal function

The temporal lobes

The temporal lobes deal primarily with memory and language functions. Neuropsychologists test for the status of at least two types of memory. Episodic memory which, as its name suggests, is our memory of events or episodes which are recorded with a reference to the time when they occurred (for example: 'I ate eggs for breakfast this morning'). Tests for

episodic memory include asking someone to remember as many words as they can from a word list read aloud, or showing someone a series of photos of unfamiliar faces and then asking them to identify those same faces from among a larger sample. By contrast, semantic memory can be thought of as our encyclopaedia for facts and figures about the world (for example: 'Eggs have a shell, are laid by hens, and can be eaten boiled, scrambled or fried'). Episodic memory is compromised to some extent in most forms of dementia but perhaps most profoundly in Alzheimer's disease. Semantic memory is most selectively and seriously affected in individuals with semantic dementia.

The frontal lobes

The frontal lobes perform multiple different functions yet work in unison to form our executive or management centre. The lateral or outer surfaces of the frontal lobe appear to be critical for organising and planning our actions and learning new tasks. In learning to drive, for example, these brain areas help us put together a very complex sequence of movements, which at first seem difficult and clumsy but gradually become more smooth and automatic. For someone with damage to this area, it is like being a learner all over again with many multi-stage tasks such as cooking and shopping becoming very difficult because the pattern or plan of action has been lost. Damage to these lateral areas can also cause people to get stuck on what they are doing (known as 'perseveration'). The middle portion of the frontal lobe generates our motivation and general impetus. If this part of the brain is affected, people can lose their 'get-up-and-go', becoming lethargic and reluctant to get out of bed or perform a particular activity.

Again, it is important to realise that what might be perceived as laziness by some could be a direct consequence of the loss of cells in this area of the brain. The regulation of our behaviour appears to be governed by a third area of the frontal lobes, the orbitobasal area, located in the curvature at the very front of the brain. In healthy people these parts of the brain help to monitor, control and moderate our behaviour: for example, preventing us from saying something rude when someone has really annoyed us. Neuropsychologists probe the integrity of frontal lobe functions with a range of tasks which require attention, decision-making, persistence and inhibition. These include sorting items into categories (e.g. colours and shapes), alternating rapidly between two tasks (e.g. finding numbers and letters), and suppressing pre-potent responses (e.g. naming the colour of the ink a word is printed in rather than reading the word itself). These tasks eventually become difficult for most people with a progressive degenerative condition, but are particularly affected in FTD in the early stages of the disease.

Challenges facing neuropsychologists

Unfortunately of course it's not quite that simple. Although different parts of the brain are undoubtedly specialised for different tasks, the brain works as a network. Some parts are particularly important for individual skills, but most tests at the neuropsychologist's disposal in fact require the person being tested to use more than one skill at a time. Take for

instance picture naming, a common test used for assessing language abilities and in particular word retrieval skills. In showing someone a picture of a dog and asking them to name it, the neuropsychologist is in fact requiring the person to perceive the picture clearly (its shape, colour, features, etc.), know what it is (i.e. access correct semantic knowledge about dogs), retrieve the correct name, and then to say the name aloud (requiring appropriate pronunciation and coordination of speech muscles to create the correct sound). Many stages underlie such an apparently simple task.

This is one of the reasons why neuropsychologists often ask a lot of questions; they need to assess as many different skills as possible in order to derive the most accurate cognitive profile of the person being tested. Numerous other factors also have to be taken into account, such as the previous abilities of the person being tested, their education, age, mood, tiredness, how long they have had the disease for, whether they have tried the test before, and also how difficult the test is for other people of a similar age and background.

Conclusions

Overall neuropsychology offers a non-invasive window into the brain, providing information which, in coordination with clinical interviews, brain scans and blood tests, can help health professionals to reach a prompt and accurate diagnosis. At its best, neuropsychological assessments can also be used to inform patients and carers about the condition they are facing, and to guide coping strategies and care planning.

Fundamentally, neuropsychologists should be asking questions which increase understanding of how and why challenging behaviours occur, and thus enable carers in particular to focus more upon the person with dementia than the dementia itself. Or, as the Canadian physician William Osler (1849-1919) put it, “Ask not what disease the person has, but rather what person the disease has.”

Dr Sebastian Crutch
Professor of Psychology, Dementia Research Centre, UCL Institute of Neurology

Communication

Communication is a two-way process which affects people with dementia as well as those caring for them. Loss of language is caused by damage to the left temporal lobe. The patient with semantic dementia may lose not only the ability to find the right word to express what they want to say but they also may lose the ability to understand the meaning of words and phrases spoken by others. The vocabulary they are able to use and understand becomes increasingly limited. Loss of concentration is also a contributory factor.

When communicating with a person with semantic dementia it is important to speak slowly and use short sentences. Not offering options may help, "would you like a cup of tea or a cup of coffee" is more complicated than "would you like a cup of tea" It is important to face the person with dementia and to be on the same level.

Wordbooks with pictures and dictionaries sometimes help but reading them may develop into on obsession. Making ones own wordbook is a strategy that can be used in the early stages. For instance stamps can be entered under Post Office. The ability to read and write is usually maintained to about the same level as spoken language, but writing messages is occasionally effective.

Gestures may be understood or misunderstood and can sometimes appear threatening to the person with dementia. Using the thumbs up or thumbs down sign sometimes works. Gentle pats and hugs may take the place of words. People with frontotemporal lobar degeneration sometimes develop expressionless faces, often seen in Parkinson's disease. Again this hinders communication.

It can be beneficial to use all forms of communication to get the message across; writing, drawing, speech, gesture, facial expression, and always to have a pen and paper for both the person with FTD and the communication partner. A speech therapist may be able to offer guidance (see below for a description of services offered at the National Hospital for Neurology and Neurosurgery [NHNN] UCL).

Sometimes other forms of communication appear and several members of the FTDSG have shown amazing abilities to paint and draw.

<u>Support Organisations</u>

Whilst we recognise that the communication difficulties associated with FTD diagnoses often carry a particularly unique range of symptoms, there are support organisations for people with aphasia, which offer information, advice and resources which our members may find helpful:

http://www.aphasiaalliance.org - The Aphasia Alliance is a coalition of key organisations from across the UK that share a common purpose – to improve the lives of, and support, people with aphasia and their families.

Penelope Roques
Rare Dementia Support

What can Speech Therapy Offer?

Speech and language therapy at the NHNN aims to provide a timely, 'evidence-based' service (a service based on research) for people with Primary Progressive Aphasia (PPA) and other forms of this condition and their communication partners. We also offer advice and guidance for services provided by community speech and language therapists if required. Everyone with PPA can access NHNN SLT services regardless of where they live. We also aim to share as much information available for people with PPA.

At the NHNN, a standard of 6 SLT sessions will be offered for new clients referred. (This may vary). Clients may opt to be seen by their local service at any stage and long-term follow up is usually by informal review at the PPA groups. These groups are usually run 3 times a year.

The treatment plan or "pathway" usually includes the following; Session 1 is usually an assessment of communication strengths and weaknesses to identify how the client usually communicates i.e. using speech, writing, pointing to pictures, a speaking device or a combination of all the above. This assessment also identifies who the client usually communicates with and their roles. i.e. partner, friend, family member, co-worker, customers (if still working). A range of speech and language tests both formal and informal can be used.

In sessions 1-2, an assessment of other ways (augmentative, alternative strategies) to help communication is made. I.e. using an iPad or an alphabet chart to help with expression of words and ideas, wants, needs etc. Sessions 1-6 would include practice and integration of any appropriate strategies to enhance communication, through modelling, rehearsing and home tasks involving significant communication partners. Information regarding appropriate high-tech communication aids and applications/software is also provided. Evaluation of strategy take-up, through re-assessment of base-line tests is also done.

Sessions 1 and 2, and on-going sessions aim to introduce the concept of creating a communication notebook or profile. This profile includes information and photographs, pictures about the client e.g. family and friends, likes and dislikes, food and drink, leisure activities, holidays and interests, conversation topics etc. This can also include information about day to day things/activities. Sessions may aim to help the client and their communication partners use this profile in everyday situations with a view to independent use if appropriate (e.g. the client independently initiating needs/wants, ordering food at a restaurant, choosing a TV programme to watch etc. This profile may be created using an Ipad with an application called Proloquo2go and has been helpful for many clients with PPA. As client's abilities vary, this technology is not appropriate for everybody as it requires a sound level of literacy and dexterity to access it.

By session 5, provision of a communication card consisting of information about the client's skills and any strategies to facilitate their communication may also be beneficial. This aims to maintain a client's independence with communication. This might mean that a client can independently go to a café to order a coffee, or to buy a list of groceries from the shop for example.

Sessions 1-6 aim to provide any education about PPA and its related conditions and communication partner training to practice strategies to support a client's functional communication. These may include role plays and specific individualised training around levels of assistance/support needed for a person with PPA to use their communication strategies.

The pathway may also include impairment-based therapy (working on the specific difficulties e.g. word finding). This is called neuro-protective therapy and consists mainly of drilling of functional words (written and/or spoken as appropriate) This is only generalised for the words practiced. These might include family names, names of objects used in everyday activities.

This is a general outline of what SLT can offer at the NHNN, but it can vary based on a person's strengths, weakness and level of participation.

Luke de Visser
Specialist Speech and Language Therapist, Therapy Outpatients, NHNN

Recommended book:

Volkmer, A. (2013) Assessment and Therapy for Language and Cognitive Communication Difficulties in Dementia and Other Progressive Diseases. J&R Press, UK.

Adapting to Changes in Behaviour

Some of the behavioural changes that accompany a diagnosis of frontotemporal dementia can be difficult to adapt to and understand.

Personality changes

Along with obsessional parts of behaviour, speech problems and adherence to strict routines, aggression and disinhibition can be difficult to manage both inside and outside the home.

People may find it useful to carry our credit card sized information cards to show/give to members of the public when the need to explain atypical behaviour is quickly but subtly required.

The card states:

"This person has a brain disease. There may be problems with speech, behaviour and confusion. Your help and patience would be appreciated."

Triggers

Working on the premise that 'prevention is better than cure' it is useful to look for the triggers that pre-empt particular behaviours. It may be helpful to keep a diary document to help establish whether a cause can be determined and managed.

Potential causes may be:

Medical (and includes):

- Reaction to medication, in particular the drugs prescribed for Alzheimers' disease
- Reaction to infection, e.g. urinary or respiratory tract
- Pain: It is important to try and identify whether pain is the trigger for a particular behaviour. It may be helpful to keep an incident chart in an attempt to identify a trigger. In one case, badly fitting dentures had caused ulcerated gums which meant any attempt to eat was incredibly painful. This pain translated into aggressive behaviour at meal times but was resolved by dealing with the cause.

Environmental (this includes anything that impacts upon the senses):

- Too much noise or too many people
- Intrusion of personal space
- Frustration due to change of routine, lack of communication
- Loss of impulse control
- The need to control (often hiding incompetence with accusations)
- Over sensitivity and reaction to touch

Avoid making matters worse

- It helps if family, friends and carers are not confrontational and do not take personal offence or raise their voice. Hurrying or crowding heightens feelings of threat and alarm.
- Try to avoid reacting adversely or trying to reason with the person with the diagnosis. These diagnoses make it difficult for people to follow logic in the way they may previously have been able to do.

Dealing with the problem

- Stay calm, respect personal space and be reassuring.
- Keep a sense of humour.
- Use distraction techniques. Refocus – perhaps with a change of subject or treat.
- Music can sometimes provide a helpful distraction.
- Keep decision making to a minimum. Avoid open ended questions.
- Try to find purposeful activities, including exercise where appropriate.
- Be aware that you may need to actively initiate activities. People with these diagnoses often find it difficult to start an activity, but will take part once it's happening.
- Accommodate routines, behaviour and desires wherever possible.
- Assess the risk, and decide whether this is an issue you need to challenge or not. If no harm can come, maybe it is safer to let the behaviour run its course. Only 'take on' the issues that need to be addressed'.

Adapting to changes in behaviour can be exhausting for all concerned. Make as many efforts as appropriate to understand the disease and to share how you feel. Try to remember that the reason behind the changes in behaviour are being cause by a disease - not intentional direction. Be aware that research has shown that some of the symptoms of FTD cause significant strain on close and marriage relationships.

Conclusion

Be prepared where possible to walk away from the situation, to try later, and be flexible.

Jill Walton
Rare Dementia Support

Obsessions – Frontotemporal Dementia (FTD)

People with FTD may develop obsessions. This list highlights some of them, but nobody develops them all!

A-Z OF 'FTD'

A	Alcohol	increase in consumption, hiding alcohol sometimes works
B	Buying	anything and everything but particularly soft toys
C	Children	talking to children can cause embarrassment
D	Disinhibition	takes many forms, loss of social graces are common
E	Eating	changes in taste often occur, sweet things being preferred
F	Fiddling	and touching everything around - a symptom called utilisation behaviour
G	Giggling	often for no reason or at inappropriate times
H	Hoarding	refusing to throw anything away
H	Hugging	sometimes anybody and everybody
H	Humming	constantly humming, often on one note. The distraction of music, perhaps through headphones, sometimes helps
J	Jealousy	suspicion about anybody or everybody particularly the nearest, dearest and most helpful
K	Kissing	sometimes anybody or everybody
K	Kleptomania	acquiring things from shops, explaining the illness to local shops may help
L	Licking	patients may put anything in their mouths, a symptom called hyperorality
M	Money	it may unaccountably disappear, Enduring Power of Attorney recommended
P	Picking up	almost anything but particularly little bits on carpets

Q	Quantities	counting objects, arranging things in patterns or having to do a task a certain number of times
R	Religion	over and above previous interest
S	Sex	some very sad changes often occur in sexual habits
S	Switches	obsessively switching things off, even the fridge
T	Time	strict adherence to routines, eating, walking, sleeping etc. must be carried out at the designated time
T	Tiredness	may insist in staying in bed or having fixed rest periods
V	Vocalisations	clearing the throat, grunting or shouting out repeatedly for no reason
W	Walking	long walks, often the same route at the same time of day
W	Wordbooks	constantly reading and repeating words from the book
ZZZ	Sleeping	often spending long periods in bed

Penelope Roques
Rare Dementia Support

Apathy and How to Deal with it

Definition of apathy

The meaning of this word has ancient roots - apathy is a term coined by the Greek Stoic philosophers to refer to the condition of being free from emotions and passions, such as fear, pain, desire, and pleasure. Nowadays the term “apathy” (which is also known as a “negative” symptom) primarily refers to a loss of motivation, interest, and concern and is amongst the most commonly complained of symptoms in dementia.

Most research and treatment is focussed on “positive” symptoms of dementia such as aggression and violence. Studies of carers consistently show that apathy causes more burden and stress than any of these behaviours. There are a number of theories as to why this should be. Positive symptoms come and go and sometimes the triggers can be predicted, patients show reactions to their carer, even if sometimes these reactions are violent and distressing.

Patients with apathy tend to be withdrawn and unresponsive on a constant basis and may rarely even acknowledge the presence or intervention of their carer. Apathy paves the way for learned helplessness (the cycle where the sense of failure to take action in day to day activities leads to a feeling of not being in control and loss of ability which then leads to further lack of activity) and thus feelings of hopelessness in both patient and carer.

Causes of apathy

These are complex and are related to the underlying pathology and atrophy of the frontal lobes, their connections to the temporal lobes and the psychological consequences of this, both on the patient and the carer.

The frontal lobes act as the “motivator” of the brain, causing us to plan, initiate and then carry through activities. Damage to the frontal lobes not only takes away the “spark” needed for us to decide what we are to do, but prevents us from being able to sequence the task appropriately and see it through to the end.

Patients with FTD are often unaware of the fact that they have these problems. They lose confidence in their abilities and are unable to divert their thoughts and interests to areas where they may actually still be quite capable.

The important feature of apathy is that it is easy to descend into a downwards spiral of lack of activity, loss of confidence and further loss of skills. This then further adds burden to the carer who has to take over more of the tasks.

There is no medication available to treat apathy and sedatives and neuroleptic medication which are sometimes used to control behavioural problems in FTD will actually make it worse.

What can we do about it?

1. **Exclude the presence of depression**

Clinical depression is rare in bvFTD and is associated with the symptoms of tearfulness, poor appetite, sleep and self esteem. It is easy to confuse clinical depression with apathy but vital to distinguish between the two as this can be treated with antidepressant medication

2. Be creative!

There are no studies about the best way to manage apathy; most ideas have come from carers themselves and sometimes, paradoxically from using some of the other behavioural symptoms of bvFTD.

- Obsessions and rigid patterns of behaviour are a common problem in bvFTD. By establishing a routine such as a daily walk or trip out, this can be capitalised upon- the apathy will be overcome by the patients need to adhere to their daily schedule.
- Patients often show "utilisation behaviour"- the need to pick up, explore and use everything in their environment. This symptom can be used by enriching the environment with picture books or activities the patient enjoyed in the past such as knitting or simple model making.
- Certain visual and practical skills are retained in patients with bvFTD. Many patients enjoy repetitive activities such as word search puzzles and Jigsaws and are given confidence by their ability to be able to complete these.
- Speak to other carers and find out how they cope with apathy. There is helpful information (some of which has been used in this article) available from the Alzheimer's society and the FTDSG.

3. Break tasks down into manageable chunks

This is a very powerful technique used by psychologists. The person may be able to complete a task when it is broken down into sections, even if they can't follow the whole procedure through. An example of this is getting dressed. Putting the clothes out in the order they are put on may make it possible for the person to continue to dress themselves. Achieving only one or two steps of a task may give them a sense of achievement.

- Make eye contact and give instructions slowly, clearly and one at a time. Sometimes pointing or miming what you would like the patient to do also reinforces the information.
- One of the biggest problems experienced by patients with bvFTD is initiating activities. By for example, gently holding a spade with the patient and starting the digging off, they will be able to carry on with the task.
- Try simpler tasks for example, rather than asking the patient to "clean the lounge" break the job up into "could you dust the shelves" and then "will you vacuum the floor".

It is said that every night, Ronald Reagan's security guards sprinkle a bag of dead leaves on the swimming pool so that when he walks out the next morning he can rake the water; a job the former president enjoys and still obtains a sense of achievement from. The security guard then puts the leaves back in a bag, ready to throw on the pool again in the evening!

4. Remain positive

This is so much easier said than done. There is a lot of evidence however that patients with dementia can sense the mood (and frustrations) of those around them -the more relaxed and positive you are the more the patient will feel able to do.

- Try to encourage the person with dementia to do whatever they can for themselves and only offer as much help as is necessary. If they are struggling with a task, avoid the temptation to take over completely, even though it may seem easier and quicker. If you need to offer assistance, try to do things with the person rather than for them. The person will then be more likely to feel involved.
- It is more important that the person feels useful than that they complete the task perfectly. Praise, encouragement and humour are much more effective than criticism.
- Take up any offers of a place at a day centre, offers of help from friends or sitting services. This will stimulate the patient and engage them in new activities.
- Sometimes the disinhibition and socially inappropriate behaviour of patients with bvFTD can prevent you from wanting to eat out or go on trips. Sometimes a quiet word with the manager or waiter can enable you to establish a regular visit or a table in the corner or nearer the bathroom where you will feel able to relax and enjoy your meal

If you have any other techniques that you have found useful for overcoming apathy, please share them with us and other carers.

Dr Liz Sampson MD MRCPsych
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Consultant in Liaison Psychiatry (Older People), North Middlesex University Hospital

N.B. Editorial note: April 2017 – the original term Pick's Disease has been replaced with bvFTD in the section above.

Swallowing Problems

Swallowing is a complex process, which involves many different aspects of thinking as well as the muscles of the mouth and throat (pharynx and larynx) and the route to the stomach (oesophagus). Fronto-temporal dementia can affect a person's ability to concentrate on their meals, to recognise the foods, to feed themselves or to co-ordinate chewing and swallowing. This may mean they are unable to finish a meal, they may not know what to do with food or drinks and they may not be able to manage foods they have always eaten. You may notice that a person starts avoiding certain foods, they chew food for a long time, or that they cough when they eat and drink.

Difficulties with swallowing can have serious consequences; people may be at risk of losing too much weight, chest infection and pneumonia or choking on food. Many co-occurring medical issues can also cause swallowing difficulties. If someone is concerned about their swallowing it is important to seek advice from a speech and language therapist, as strategies recommended will vary according the areas of difficulty. You can obtain a referral through your Consultant, GP or most other medical professionals you may be involved with. Other professionals, such as a dietician, occupational therapist, physiotherapist, or district nurse, may be able to provide advice to help also.

Common problems that you may notice:

- being unaware of food when it arrives
- failing to do anything with food in the mouth, just holding it there
- difficulty chewing and/or difficulty moving food to the back of the mouth
- spitting lumps of food out
- eating very fast or putting too much into the mouth
- eating insufficient amounts or refusing food and/or drink
- talking with food or drink in the mouth and forgetting to swallow causing coughing
- coughing/choking on food and /or liquids
- complaints of food not going down or getting stuck in their throat
- a 'wet' or 'gurgly' voice after swallowing
- difficulty swallowing tablets
- dribbling
- chronic chestiness or recurring chest infections

<u>Ways to promote safe eating that may be suggested by a speech and language therapist.</u>

1. Strategies

A speech and language therapist may make specific suggestions about what will help so it is important to see a speech and language therapist if you are concerned about swallowing, however the following may be useful:

- A quiet environment, with few distractions (no television or radio).
- Sitting at the dinner table with the table set to prompt the person that it is a mealtime.
- Sitting upright, keeping the chin down.
- Take small sips of drink, perhaps from a teaspoon. Avoid the use of lidded beakers that can encourage the head to tip back.
- Take small mouthfuls of food.
- Alternate food and drink to help clear the mouth of food: this should be discussed with a speech and language therapist.
- Try encouraging the swallowing of each mouthful twice to clear any food or drink that may remain in the mouth or in the throat after the first swallow.
- Frequent swallows to counteract dribbling.
- Check mouth after finishing eating to ensure no food or fluid remains.
- You may have to sit with the person to remind them to use these strategies, and you may find that mealtimes take much longer.

2. Changes to diet

Speech and language therapists may recommend the person with swallowing difficulties have thickener in their drinks or a special diet (modified texture diet such as fork mashed or puree). It is important to see a speech and language therapist, and perhaps a dietitian about this rather than just avoiding foods, otherwise you may miss out important food groups (and so miss out on important vitamins and nutrients). Dietitians can recommend supplements to help this. However the following foods may be particularly difficult and worth looking out for:

- Small hard foods such as nuts, dried fruit, chewy sweets (toffee)
- Stringy foods such as bacon, sausage skins or runner beans
- Husks on foods such as sweet corn, or grapes and cherry tomatoes
- Mixed textures e.g. food in a lot of fluids like minestrone soup, or cornflakes and milk
- Floppy textures e.g. lettuce

People often find tablets can be difficult to swallow, taking tablets with yoghurt, or a teaspoon of jam can make it easier. Check with your doctor or pharmacist if you would like to try crushing tablets or using a syrup form, as this can change the way the medicine is absorbed.

Anna Volkmer
Highly Specialist Speech and Language Therapist, South London and the Maudsley NHS Trust.

General Management

Professional and Voluntary Support Organisations

It is difficult to ensure that people receive the advice and information they require at the time they need it and in a way in which is most suitable for them. Rare Dementia Support has a collated information sheet which outlines general advice and information and is provided to new members, or upon request.

Rare Dementia Support

www.raredementiasupport.org - Our website provides disease specific information. It is regularly updated with forthcoming support group meetings/events. You can access past support group meeting recordings and newsletters.

FTD Talk

www.ftdtalk.org - A site which provides jargon free research updates on FTD for all people in the FTD community.

YoungDementia UK

www.youngdementiauk.org – Young Dementia UK is a charity that exists to help people whose lives are affected by young onset dementia.

Young Dementia Network

www.youngdementiauk.org/young-dementia-network - A community that includes people living with young onset dementia, their family and friends, as well as organisations and professionals who work in the fields of dementia and social care.

Association for Frontotemporal Degeneration (AFTD)

www.theaftd.org - An American organisation, the AFTD envision a world where frontotemporal degeneration is understood, effectively diagnosed, treated, cured and ultimately prevented and have a mission to improve the quality of life of people affected by FTD and drive research to a cure.

Eastern Cognitive Disorders Clinic (ECDC)

www.ecdc.org.au - An Australian organisation, the Eastern Cognitive Disorders Clinic (ECDC) was established in 2006 and is based at Box Hill Hospital in Melbourne, Victoria. ECDC is a diagnostic clinic specifically for patients with focal onset dementias and for those with cognitive syndromes that pose diagnostic difficulties. ECDC clinicians have a particular interest and expertise in the diagnosis and management of Frontotemporal Dementia (FTD). The site contains some helpful resources.

Alzheimer's Society

https://www.alzheimers.org.uk - Provides support for anyone affected by dementia.

Alzheimer's Research UK (ARUK)

www.alzheimersresearchuk.org - ARUK are the UK's leading research charity aiming to defeat dementia.

Supporting Children

FTD often affects people in middle age, or even younger, meaning that it is likely that there will still be children at home. When any parent faces a serious illness, their children need support and understanding. It is important to help young people manage and understand the disease they are witnessing in their parent. Children are very perceptive. They are likely to be confused by the symptoms of this disease in a parent or family member, and without being given factual information they may fill the gaps in their knowledge with their imagination or with incorrect facts. It is therefore very important to facilitate conversation, provide information and allow for emotions and feelings to be discussed.

Change and loss often bring emotional reactions that parallel those we experience when we grieve. Anger, denial and the eventual acceptance of a change in circumstance will present differently for each of us.

Children are not immune from these reactions, and don't always have access to the information they need in order to process their understanding and feelings appropriately. In an attempt to protect children from harsh realities, we can at times deny them access to the facts they need. Some children accept changed circumstances quite easily, while others may find it difficult. It is important that children's questions are answered honestly.

Being aware of children in families affected by these diagnoses helps us to notice changes in their behaviour which may be a result of all they are trying to deal with and come to terms with. It is possible that someone outside the family may notice changes that occur – a teacher or friend. There may be support services accessible via schools or colleges and it is important that appropriate staff are informed.

What to look out for:

- Children may become quiet and withdrawn. They may feel afraid to ask questions or unable to talk to anyone at home. 'Sparing' them may be protecting ourselves. Anger, guilt and sadness can build up
- Lack of concentration – forgetting where they put things – not paying attention
- Wander aimlessly
- Habits may increase – nail biting, twiddling hair etc.
- Attitude to food may change
- Bedtimes may become more difficult
- Phobias may develop
- Psychosomatic responses – tummy pains, lethargy
- Regression or childish behaviour
- Cries for attention may manifest in a variety of ways

Questions children want answering:

- Why did it happen?
- Did I cause it?

- Will it happen to me?
- Will the person get better?
- Who will take care of me?
- Who will take care of the remaining parent?
- Will I be able to leave home/go to university?

Why children need information:

- Information gives people a sense of control.
- Try to find someone who can answer questions if you don't know the answer, but 'I don't know' is acceptable as an answer.
- Without information, children may fear the worst and develop unreasonable concerns about what is wrong and who is responsible.
- Without information children feel left out, and may be likely to isolate themselves further.
- They need to know what is happening in order to make informed decisions about what they want to do in the present and in the future.

Support and Information for children:

There are several links which provide helpful information in respect of providing young people with the information and opportunities for discussion that they need. The links are not all FTD specific, but provide information that is transferrable and relevant:

The links are not all FTD specific, but provide information that is transferrable and relevant:

Understanding dementia: a guide for young people (PDF download). Also available through Dementia Helpline Alzheimer Scotland Action on Dementia: Tel 0808 808 3000

- What about the Kids? (AFTD)
- 'Frank and Tess': A children's activity book about FTD (AFTD)
- Carers Trust: Young Carers
- Information for Young Carers from the NHS
- When Dementia is in the House - for parents and teens
- Alzheimer's Association; Living with Alzheimer's - just for kids and teens
- Alzheimer's Research UK: https://kids.alzheimersresearchuk.org
- Young Onset Dementia Book for Children: http://www.ypwd.info/shop/young-onset-dementia-book-children

Jill Walton
Rare Dementia Support

Legal and Funding Matters

RDS subscribes to CareAware advocacy service to deal with financial and legal queries. CareAware is a non profit making public information, advisory and advocacy service specialising in care funding advice in the UK.

The Advocacy Service can assist with:

- Understanding social services assessment procedures
- Sourcing appropriate care services and support organisations
- Establishing entitlement to state benefits including: Attendance Allowance, Pension Credit, Continuing Care Benefit and Registered Nursing Care Contribution
- Maximising support from the Local Authority including 12 week property disregard and deferred payment agreement
- Paying care fees, protecting assets, funding top-up payments, securing an inheritance and tax planning
- Ensuring your wishes are respected and Enduring/Lasting Power of Attorney (or equivalent) to protect independence and integrity.

The key features of the Advocacy Service are:

- To ensure that care residents and their families have access to all the independent help and information they will need in dealing with funding long term care.
- It complies with National Minimum Care Standards.
- It co-ordinates access to the UK's leading specialists on care issues.
- There is no cost, charge or obligation for the end user.

To contact:

Care Aware, PO BOX 8, Manchester, M30 9NY

Helpline: 0161 707 1107

Email: enquiries@careaware.co.uk

Web: www.careaware.co.uk

Planning for the Future: Advance Care Planning and Lasting Power of Attorney

Planning for the future is an important part of living with a long term condition such as dementia. Everyone's situation and circumstances are individual. Some people want all the information available at the time of diagnosis, while others wish to take things as they come. Some people are in regular contact with health and social care professionals, while others have limited involvement of services. Whatever the personal circumstances or preferences of the person with dementia and their family are, it is important that they are aware of things that can be done to assist future decision making.

The process of future planning allows people with dementia and their families to have time to consider their views on different health and social care situations in advance. This may make future situations more manageable and may help to avoid crisis. It is advisable to start thinking about the future and the possible decisions that may need to be made from the time of diagnosis to allow the person with dementia to be included.

Throughout the disease progression, those affected and their families will need to make many decisions related to the changes that the disease causes in every area of their life. Although no one can predict the exact timeline of the progression, the deteriorating nature of the disease means that with time people affected will gradually lose their capacity to make decisions and become increasingly dependent on others.

A person may have capacity to decide on some issues but not on others. Many difficult decisions such as care home placement and treatments in the late stages of the disease are made at the time when the person with dementia is unlikely to have capacity. This is especially stressful for carers who often are left with a feeling of guilt over the decisions they make under already distressing circumstances.

Advance care planning, including arranging a Lasting Power of Attorney, may help with decision making and management of care in the later stages of the disease. It is essential for these to take place when the person with dementia still has capacity in order for them to be legally valid.

Even if the person does not have capacity, or does not wish to arrange an LPA, it is still useful to plan for the future. Sometimes it may not be necessary to write down the wishes and preferences or draw up legal documents. Just being able to talk about future care options, either within the family or with professionals involved, may be enough.

Advance care planning

Advance care planning is a dynamic process of discussions between a person with dementia, those close to them and professionals providing care for them. The aim is to discuss the views, preferences and wishes of the person with dementia related to future care.

Advance care planning should take place at the time the person affected and their family feel it is right for them. The process is entirely voluntary. The aim is to identify the views, wishes and preferences of the person with dementia and to ensure that those close to

them, and the professionals involved, are made aware of them. It is also possible to refuse a specific treatment. Some of the issues that are included in the advance care planning discussions are listed below.

Identifying wishes and preferences

Some people have strong views about treatments or types of care that may be offered in late stages of the disease. For some people the place of care may be especially important. For example, they may wish to stay at their home (family home or care home) even if they become very unwell. Many care homes discuss these issues with the person and their family at the time when a person moves in.

Some people have spiritual or religious beliefs that they wish to be taken into account in their care. There may be important practical issues that carers need to be aware of. For example, some people may prefer a shower to a bath or it may be important to them to have a carer of the same gender as themselves to attend to personal care tasks.

The person with dementia may wish to identify someone close to them to represent their views. This is different from Lasting Power of Attorney, as it is not legally binding. It is not necessary to have the wishes and preferences written down, however it might be useful for those involved in the care of the person with dementia if their views are in writing.

While health and social care professionals are expected to take into account the views of the person with dementia and their family when planning for care, sometimes it may not be realistic or practical to follow all.

Refusing specific treatment

A person with dementia may have strong views about life-sustaining or life-prolonging treatments at the time when recovery is not possible and quality of life is compromised.

An advance decision to refuse treatment (previously known as a living will or advance directive) is a decision which a person with capacity can make to refuse a specific type of treatment at some point in the future. They may wish to refuse a treatment in some specific situations, but not in others. It is important to specify the details of the circumstances.

There are specific rules in refusal of a life sustaining treatment, such as ventilation. A decision to refuse treatment must be put in writing, signed and witnessed. If a person wishes to refuse a treatment they are advised to discuss this with an experienced care professional who knows the person and their medical history well; for example, their GP.

Further information about advanced care planning and refusal of treatments is available from Alzheimer's Society and NCPC.

The National Council for Palliative Care and Dying Matters have produced a booklet entitled 'Difficult Conversations. Making it easier to talk to people with dementia about the end of life.'

The booklet offers guidance to help people open up conversations about end of life issues and preferences. It is available via:

enquiries@ncpc.org.uk or by telephoning 0207 697 1520.

Lasting Power of Attorney (LPA)

Many people are concerned about how decisions are made if they lose capacity to do so themselves. A Lasting Power of Attorney (LPA) is a legal document which gives the attorney(s) that the person has chosen the right to make decisions on both financial and health care matters in the event they lose capacity to make decisions themselves.

The LPA has two parts; the property and affairs LPA and the personal welfare LPA. It is possible to make both or only one. The LPA gives the person a choice of conferring broad or limited powers to make decisions on their behalf, and a choice of who to appoint. For example, it is possible to appoint relatives to make welfare decisions, but a professional adviser for decisions relating to their property and affairs.

A property and affairs LPA gives the attorney(s) the power to make decisions about financial and property matters, such as selling a house or managing a bank account.

A personal welfare LPA gives the attorney(s) the power to make decisions about health and personal welfare, such as day-to-day care, medical treatment, or where the person should live.

A personal welfare LPA only ever takes effect when the donor lacks the capacity to make decisions. A property and affairs LPA can take effect as soon as it is registered, even while the donor still has capacity, unless the donor specifies otherwise. The donor can, of course, specify that the attorney may only start managing their financial affairs after they lose capacity, at some time in the future.

Planning for the future is an ongoing process which should start at the time of diagnosis. Health and social care professionals as well as voluntary organisations such as Alzheimer's Society can provide assistance and further information.

Further information

Alzheimer's Society

Tel: 020 7423 3500

Email: info@alzheimers.org.uk

Website: www.alzheimers.org.uk

Advance decision factsheet and downloadable form: www.alzheimers.org.uk/factsheet/463

Office of the Public Guardian

Tel: 0845 330 2900

Email: customerservices@publicguardian.gsi.gov.uk
Website: http://www.guardianship.gov.uk

Department of Constitutional Affairs

(Archived) Information Booklets on the Mental Capacity Act 2005

A range of booklets for individuals who may wish to plan ahead or who may not be able to make some decisions for themselves
www.dca.gov.uk/legal-policy/mental-capacity/publications.html

Also see:

https://www.gov.uk/government/collections/mental-capacity-act-making-decisions

The National Council for Palliative Care
This is the umbrella charity for all those involved in palliative, end of life and hospice care in the UK. The charity believes that everyone approaching the end of life has the right to the highest quality care and support, wherever they live and whatever their condition.

Email: enquiries@ncpc.org.uk

Website: www.ncpc.org.uk

Telephone: 0207 697 1520

Ritta Kukkastenvehmas
Dementia Research Centre

Driving and Dementia

"It's difficult to be on the passenger seat after years of driving" – person with dementia

We often think about driving as an autonomic action that we do without thinking, although in fact it is a very complicated task. Driving involves highly complex interaction between senses, thought process and manual skills. A driver must be able to react quickly to other road users' actions, judge distances, be able to read road signs, and to remember where they are going.

Having a diagnosis of dementia does not necessarily mean that the person is unable to drive. Many are able to continue driving for some time after the diagnosis. However, with progression of the disease, abilities such as judgement, attention, concentration, orientation, and memory, all of which are important for driving, will be affected.

Orientation is usually retained for a long time in those with a form of frontal dementia, such as bvFTD. Manual tasks such as changing gear, may become difficult due to loss of the ability to carry out familiar movements (apraxia). Judgement may also be affected, for example, judging the speed of the car, or anticipating moves of the other road users. People with Lewy body disease may experience visual hallucinations, potentially causing difficulties with concentration. These are some examples of how dementia can affect the ability to drive.

Early warning signs
It often is the person in the passenger seat who is the first to notice change in the way the person with dementia is driving. The first sign usually is not feeling as safe in the car as previously. The following list may be helpful. Early signs of driving problems include:

- Incorrect signalling
- Trouble navigating turns
- Moving in the wrong lane
- Confusion at exits
- Parking inappropriately
- Hitting curbs
- Inappropriate speed
- Delayed responses to unexpected situations
- Increased agitation and irritation when driving
- Scrapes and dents on car
- Getting lost in familiar places
- Accidents

Difficulties in giving up driving

"My children had a meeting without me and decided that they want me to stop driving, but they are making a big deal about nothing. I'm very comfortable on the road. I've driven longer than they've been alive" – person with dementia

Driving symbolises freedom and independence, as well as being a practical way of transport. It is not surprising that giving up driving is very hard. It is always best if the person with dementia decides voluntarily to give up driving. However, they may be unable to assess their driving skills, or insist on driving even when it is no longer safe. It is essential to have as much support as possible from family members, friends, and professionals. There is no single right way to deal with the situation. What may work in one situation, may not necessarily work in another. Some possible interventions are listed below:

Arrange support when making and implementing the decision about driving

- Share the responsibility with other family members, friends, and professionals
- A letter from the doctor explaining the reasons for having to give up driving may be helpful
- A respected friend's opinion could be valuable

Make the car inaccessible

- Hide the car keys
- Park the car away from the usual place
- Change the colour or the model of the car. It can be helpful if the person can't recognise the car
- Disable the car
- Sell the car

Alternatives for driving

"I hated to stop driving, but sometimes it nice not to let others worry about the traffic" – person with dementia

If it is possible to find alternatives for getting around, this can make giving up driving less stressful.

- Ask a family member or a friend for a lift

- Taxis. It is possible to set up an account with a local taxi company. The cost can end up more reasonable that way, and managing one's account can support the person's independence
- Using public transport. All local authorities must offer free of reduced bus fares to people of pension age and people with disabilities.
- Community transport. Find out from your local authority about the facilities available (Door-to Door transport, Dial a Ride)

Legal requirements

By law, anyone holding a current driving licence must inform the Driver and Vehicle Licensing Authority (DVLA) if they receive a diagnosis of dementia.

Drivers Medical Group

DVLA

Swansea SA99 1TU

Telephone: 0870 600 0301 (weekdays 8.15am - 4.30pm)

Website: www.dvla.gov.uk

- If the person wants to continue driving they should request a medical investigation. DVLA will send a questionnaire, which seeks permission to obtain further information from the person's GP and/or specialists. Based on the evidence DVLA may issue a driving licence that is valid for a limited period, after which the situation will be reassessed.
- If the person with dementia does not wish to continue driving they should return their licence to DVLA
- Failure to inform the DVLA of a diagnosis of dementia is a criminal offence punishable by a fine.

Insurance

- When a person receives a diagnosis of dementia they must inform their insurance company, failing this will jeopardise the validity of their insurance
- Driving without a valid insurance is a criminal offence

Arranging a driving test

It is possible to arrange a driving test through a specialist driving centre, to have an up to date assessment. For example, Queen Elizabeth's Foundation Mobility Centre can provide a list of accredited mobility centres in England and Wales. Queen Elizabeth's Foundation Mobility Centre Damson Way Fountain Drive Carshalton Surrey SM5 4NR Tel: 01372 841100 http://qef.org.uk/our-services/mobility-services/assessments

Giving up driving is difficult, both emotionally and in practical terms for all those involved. It often is one of the first concrete and visible losses a person experiences due to dementia.

Support from others and trying to find alternative ways to get around can help. As the needs of the person with dementia change over time, the importance of driving may decrease as well; this could be helpful to keep in mind at difficult times.

Ritta Kukkastenvehmas
Dementia Research Centre

2015 update: In cases where a person refuses to stop driving or to inform the DVLA of their diagnosis, a medical practitioner can contact the DVLA and disclose any relevant medical information, in confidence, to the medical adviser.

See www.dft.gov.uk/dvla/medical/ataglance.aspx for more information.

Police Contact and FTD

Whilst there are many other diagnoses within the dementia spectrum which cause specific and identifiable changes to a person's behaviour, behavioural variant FTD, because of its symptoms, has potential to lead to contact with the police. People with dementia who fall within the precincts of the police or judicial system, rarely attract headline news. Rather, it is often with a sense of shame that families manage the involvement, and with a reticence to discuss the intricacies of the contact.

The consequences of contact with the police or judiciary is typically exhaustive and stressful, encumbering emotional, social and sometimes financial implications as a result.

Given that between them, the police service and the judiciary have the only 24 hour, 7 day a week mobile emergency community response capacity, along with an unparalleled authority, it is clear that an understanding of the implications of a diagnosis of dementia MUST be understood and countered for within these services.

The contact that people with dementia have with the police and judiciary is typically connected to issues of:

- crime or crime prevention public safety
- aggression /violence
- shoplifting
- impulsive behaviour wandering
- memory loss false allegations
- disinhibition (incl. sexually inappropriate behaviour)
- driving offences and accidents
- it may also be because the person with dementia is themselves a victim

Because many people with FTD are not aware of their illness, they may become frustrated at limitations and constraints that they do not understand and consider to be unfair. As a result the person may strike out or resist assistance. Shouting, name calling or physical abuse may result from a frustrating situation or may sometimes occur suddenly, with no apparent reason. RDS has produced a flyer addressing the issue of FTD and Police Contact. Please use any of the contact addresses to request.

Jill Walton
Rare Dementia Support

General Advice

Rare Dementia Support provides credit card sized awareness cards stating:

"This person has a brain disease. There may be problems with speech, behaviour and confusion. Your help and patience would be appreciated."

The FTDSG also provides a summary information sheet. This details typical symptoms and needs of people with FTD and maybe useful for patients going into hospital attending other appointments where little is known about this condition.

Both of these are available upon request.

Medic Alert is a registered charity providing a life-saving system.

One of their medical identification bracelets should ensure that the patient receives rapid and appropriate care.

Freephone: 0800 581 420

Website: www.medicalert.org.uk

GPS Location tracking devices can be a useful means of giving peace of mind and allowing more freedom. There are many on the market, including 'Mindme Locate' which has previously been trialled by service providers in Sussex.

Blue Badge parking scheme provides a range of parking concessions for people with mobility (or awareness) problems throughout the UK and while travelling abroad within the EU. To apply contact your local authority or apply online at http://www.direct.gov.uk

Radar Keys Radar keys provide access to over 8000 public toilets throughout the UK and are available from Radar at a cost of approx. £3.

Tel: 0207 250 3222, Monday - Friday, 9 - 5pm

Website: www.radarshop.org.uk

Email: radar@radar.org.uk

Message in a Bottle

The Lions' 'Message in a Bottle' scheme is a simple idea designed to encourage people to keep their personal and medical details on a standard from and in a common location –the fridge! The scheme is free to the user. It can serve to inform the emergency services about you and your emergency contacts. By providing information about your diagnosis, indicating whether you have special medication or allergies or not, it is a potential life saver and provide peace of mind to users and their families and friends.

Bottles which are free of charge can usually be found in your local Chemist or Doctors Surgeries. Thanks to a number of other organisations, you may also find that you can obtain

one through your nearest Neighbourhood Watch Group, Age UK, Council Offices, Housing Associations and many other places including police stations. If you are unable to find a Bottle, please contact your local Lions Club.

If you have any difficulties in finding your nearest club, or would like to know more about this scheme, please email: mdhq@lions.org.uk , or miab@lions.org.uk , or telephone: 0121 441 4544.

Smoke Alarms
Make sure you have in place.

Security
Make sure security levels are safe and appropriate.

Local Resources

Make a list of local resources in case of emergencies or sudden deterioration. If the person with dementia uses day care, respite or residential care services, or if they need to be admitted to hospital it may be advisable to have an information sheet to describe their problems and idiosyncrasies. Or alternatively provide the staff with a copy of this booklet.

Activities

- Think about activities enjoyed in the past
- If reading is difficult, audio books may be an alternative
- Music can be a great therapy - singing, dancing, keep fit activities
- A personal CD player/iPod may focus the sounds.
- Puzzles of all kinds - particularly jigsaws
- Rug-making, gardening, baking (supervision may be required)
- Walking and swimming are good exercise
- Painting or drawing may become a new hobby – painting by numbers or adult colouring books

Travel

Make sure that all parties have some awareness of the illness and that certain problems could occur e.g. airline staff, coach drivers and tour guides. They will want to know there is going to be adequate supervision. Make sure travel sickness tablets are taken if required. If tablets are prescribed try a dummy run for example a coach trip when one can get off if necessary.Access to toilets for disabled people can be a tremendous help. Most of them are kept locked and a key can be obtained from RADAR .

Tel: 0207 250 3222, Monday - Friday, 9am - 5pm

Website: www.radarshop.org.uk

Email: radar@radar.org.uk

Falls

People with dementia are more liable to fall over. Specialist advice can be obtained from an occupational therapist or physiotherapist. Ensure there is adequate lighting in corridors - dim lights can be plugged into wall sockets near the floor. Make certain rugs are not slippery or rucked up. Check that the person with dementia has shoes that are appropriate and fit properly. If someone falls make them comfortable until they are orientated and able to help themselves stand. Take care of your own back.

Going into Hospital

Day case, emergency or planned admissions to hospital can be problematic for people with dementia. Their behavior can be misinterpreted and misunderstood. With rapidly changing scenarios and inevitable shift changes in staff, the real needs of the person with dementia can be missed.

Rare Dementia Support provides laminated cards which highlight some of the typical behaviours seen in fronto-temporal dementia. These may be useful to give to people responsible for providing primary care or left in places where they can be easily read by staff/carers. The Alzheimer's Society produces a leaflet called 'This is Me' for people with dementia who are going into hospital. 'This is Me' is a simple and practical tool that someone going into hospital can give to staff to help them understand the condition. It provides a 'snapshot' of the person with dementia, giving information about them as an individual, such as needs, preferences, likes and interests. The Royal Collage of Nursing is supporting the 'This is Me' initiative. The leaflet can be downloaded via www.alzheimers.org.uk or ordered by telephoning 01628 529240.

Useful websites

There are several other websites which you may find beneficial:

◊ www.raredementiasupport.org - Rare Dementia Support website.

◊ www.ftdtalk.org - The key goal of this site is to provide jargon free research updates on FTD for all people in the FTD community.

◊ www.theaftd.org - This American site hosts a wealth of material which we are not able to resource ourselves, but which they are happy to share. Of particular note is the 'Changes in Behaviour' chart and the 'What about the Kids?' booklet.

◊ www.ecdc.org.au - This Australian site has produced a very comprehensive toolkit which comprehensively addresses many of the issues that present as a result of a diagnosis of FTD.

◊ http://www.dlf.org.uk and http://asksara.dlf.org - Disabled living sites that provide information about living aids which can be appropriate and helpful for people with FTD.

◊ https://crm.disabilityrightsuk.org/radar-nks-key - RADAR keys are another easily obtainable resource which people find helpful when needing to use accessible toilet facilities in public places. www.ppaconnection.org - is an international site offering PPA specific resources and information about collaborative research.

- ◊ https://extendstore.ucl.ac.uk/product?catalog=UCLXBCA - You may also be interested in accessing the 'Better Conversations with Aphasia' free e-learning resource to improve access to conversation therapy for speech and language therapists/pathologists (SLTs), and for people with aphasia (PWA) and their families.
- ◊ https://uk.pinterest.com/temenuzkhova/primary-progressive-aphasia-support-group is an internet community site that links to various resources for PPA, and is updated by its users.

Some of the iPad applications have been recommended by our members as helpful resources for people with PPA. Specifically they are:

'Predictable'; a text to speech application for iPad, iPhone and iPod touch, for which more information is available at www.therapy-box.com

'Pages'; for which more information is available at www.apple.com/uk/ios/pages

'Proloquo2go'; for which more information is available at: www.assistive.com/product/proloquo2go

Rare Dementia Support (RDS) runs specialist support group services for individuals living with, or affected by, a rare dementia diagnosis. Our vision is for all individuals with or at risk of one of these forms of dementia to have access to information, support and contact with others affected by similar conditions.

Rare Dementia Support seeks to:

- Provide access to disease specific information
- Facilitate contact between people embracing similar diagnoses
- To represent the views of people affected by rare dementia diagnoses across a range of strategy and policy influencing platforms

For more information about our services, please contact:

Rare Dementia Support

Dementia Research Centre | UCL Institute of Neurology

8-11 Queen Square | London | WC1N 3BG

contact@raredementiasupport.org | 07341 776 317