Pick's Disease - *Factsheet*

Dementia web
Information resource for carers, professionals and you

www.dementiaweb.org.uk

Pick's Disease

Dementia is not an illness in itself, but a term for a collection of signs and symptoms which are caused by damage to the brain by a number of illnesses, conditions or traumas. Pick Complex is a term for a group of overlapping conditions all known as **fronto-temporal dementias**. These are a group of rare neurological disorders mostly affecting the frontal and anterior temporal lobes of the brain. These areas control "executive functions" such as reasoning, personality, social behaviour, movement, speech, language, and certain aspects of memory. Among these are included Pick's disease, semantic dementia and dementia associated with motor neurone disease. Those between forty and sixty are most likely to develop fronto-temporal dementias, although they can affect both younger and older people.

What is Pick's disease?

Pick's disease was named by Alois Alzheimer in 1923 after Arnold Pick, the physician who studied and wrote about this rare, neurodegenerative disease.

Whereas Alzheimer's disease causes damage to cells across the brain, Pick's disease affects only the frontal and temporal lobes. There are often specific structural changes in the nerve cells, which take the form of bundles of useless proteins. Sometimes these bundles are quite similar to the neurofibrillary tangles seen in Alzheimer's disease, but on other occasions they are quite different, and are known as Pick bodies. Research is ongoing, but it appears that the death of brain cells may be caused by the toxic effect of the build-up of these unusable proteins.

What are the symptoms?

The loss of brain cells leads to shrinkage of the affected parts of the brain and this will result in slowly developing changes in character and social behaviour, or impairment of language. Because these changes are likely to be very gradual, it may take some time before it is realised that something is seriously wrong.

The frontal lobes of the brain help to control incoming information, to make sense of it, and to process it in order to guide our actions. When the frontal and temporal lobes are damaged and cease to work normally, it becomes impossible to make sense of the information we are receiving.

The first symptom is usually a change in behaviour or personality. Over the first two years the person begins to behave in a way which is "out of character" for them. For example, they may go on a spending spree, lose their inhibitions, make tactless comments, or exhibit aggressive or rude behaviour. They may become extrovert when they were previously introverted or vice versa. They may lose interest in sex – or begin to make unreasonable demands on their partner. They may develop a sweet tooth and experience a huge increase in appetite – or they may be reluctant to eat and become faddy.

Pick's Disease

The person may appear totally unaware of the feelings of others, lacking any insight or empathy. Their family may therefore begin to see them as very cold, selfish and inconsiderate. They may develop fixed routines or become obsessive, and they may experience severe mood swings. They may find concentration impossible and become apathetic.

Initially there may be few problems with speech or with memory, although people may appear forgetful because of faulty attention and concentration. Problems with memory tend to develop because of damage to the part of the brain which enables us to recall what words mean, who faces are, or what objects are for. This is known as semantic memory. Other forms of memory, for example, being able to remember how to get somewhere, what time to have lunch and events in the past are often undamaged.

Depression may be diagnosed because the person exhibits apathy, withdrawal and general lack of interest and motivation. Incontinence tends to develop earlier than it does in Alzheimer's disease. There is likely to be a lack of understanding of risk and danger – for example the person may willingly invite strangers into the home.

Another feature of fronto-temporal dementia is language disturbance. There may be difficulty in speaking or understanding although spatial skills and memory remain intact. This can be a separate disorder known as semantic dementia, or it can be an addition to the features described above. Speech may be fluent and effortless, but the person's language may be nonsensical and odd because they have a reduced ability to grasp the meaning of words and concepts. They perceive something normally, but the damage to the frontal lobes means that they cannot understand what it means. This results in impaired comprehension of words and an inability to name things.

All these changes in behaviour are a direct result of damage to the brain cells, and are not under the conscious control of the person. Apparent coldness or selfishness is not deliberate –the person is no longer able to put themselves in another's shoes. Indeed, people are usually unaware of the changes in themselves and even if they do notice changes, they do not show concern.

Symptoms will gradually worsen and most people have difficulty maintaining normal daily living skills after two to five years, and will need extra care and support. Some individuals will come to need twenty-four hour care and monitoring at home or in a care setting.

There is a much slower form of the disease that can, in some cases, progress over as much as ten years. Studies have shown people to live with the disease an average of eight years, but the range is from three to seventeen years.

Pick's Disease

How is Pick's disease diagnosed?

Diagnosis is largely based on clinical judgement. Brain scans can show the shrinkage of the affected parts of the brain, but there is no single test that can specifically diagnose fronto-temporal dementia with total reliability while a person is alive. There can be considerable similarities with Alzheimer's disease, and the two are commonly confused. However, careful psychological testing can often distinguish the pattern of changes in fronto-temporal dementia and establish that the symptoms are not due to psychiatric disorders or other dementias. Diagnosis may include blood tests, MRI and EEG scans, and sometimes a lumbar puncture.

In many cases of fronto-temporal dementia there is a family history of similar illness, with as many as 50% of patients having affected brothers, sisters, cousins, or parent and grandparent. This is known as an autosomal dominant pattern of inheritance and is associated with a mutation in a gene on chromosome 17.

What treatment can be given?

Although research is ongoing, no medications are currently known which will treat or prevent fronto-temporal dementia. Those drugs which are used in the treatment of Alzheimer's disease, such as Aricept, are actually likely to worsen the symptoms of fronto-temporal dementia. Treatment focuses on helping people to manage their symptoms and treating problems such as depression or mood changes. Anti-depressants have been shown to improve some symptoms, and serotonin-boosting medications may alleviate some behaviours.

What to do

It is important to consult a doctor as soon as worrying symptoms appear. There are a number of reasons for this:

- Other possible causes for the symptoms can be investigated.
- An early diagnosis will allow the person and their family to learn about the disease, to come to terms with the situation and to plan for the future.
- The person will be able to state their preferences about future treatment, financial arrangements, care choices etc.
- Carers will be able to seek help and advice for themselves, as well as for the person with fronto-temporal dementia.
- Where there is a strong family history of fronto-temporal dementia, genetic testing should be discussed, but this should not be undertaken without prior genetic counselling.

Pick's Disease

Caring for people with fronto-temporal dementia is not easy and carers need to appreciate that changes in behaviour are an integral part of the disease, and people are not being deliberately difficult. It is really not possible to reason with a person with fronto-temporal dementia, because one of their main problems is that they cannot think about consequences and repercussions. In the early stages the problems may not be evident to others, and it is important to be prepared to tell friends and family about the difficulties being experienced and to be willing to seek support.

Where to get help

Contact your GP for support, advice and information. The GP will rule out reversible or temporary causes for symptoms, carry out first line tests, refer to a specialist for specific tests and assume ongoing responsibility for the person's general health. The GP can be seen as a "gatekeeper" who can provide access to a range of specialist services. Home support, day services and 'talking therapies' may be provided.

Many other people may also be able to help, such as social workers, community psychiatric nurses, district nurses, speech therapists, occupational therapists, counsellors, advocates and carers who may come into the home to assist with personal care.

It may be that the person's illness reaches a stage where their family can no longer cope with their care. If moving into a care home is seen to be the person's best interest, independent reports and quality ratings on all homes in your local area can be found on the website of the Care Quality Commission which regulates them. www.cqc.org.uk (use its webform)

Or telephone 8.30 am to 5.30 pm Monday to Friday on 03000616161

You can also refer to our factsheet: "Moving to a Care Home and Funding Your Care" - coming soon.

Because people with fronto-temporal dementia often do not recognise that they are ill, and because they may become uncaring, cold and unsympathetic about the difficulties their behaviour causes, the carer may feel overwhelmed by their responsibilities. The Pick's Disease Support Group aims to provide information and support to carers. It also welcomes people with fronto-temporal dementias as members, but warns that as its focus is on carers, some content may be unpalatable to them. It was set up by carers of people with various forms of fronto-temporal dementias and includes a Members' Forum, clinical information and helpful tips. There is a network of regional contacts.

The website address is: www.pdsg.org.uk Email: frontotemp@aol.com

Telephone: Penelope Roques on 01297 445488

Pick's Disease

Guideposts Trust provides specialist information and care services for people with dementia and their carers. www.dementiaweb.org.uk

Contact the Helpline number: **0845 1204048** available Monday to Friday office hours, answer service at other times or by email at info@dementiaweb.org.uk

The Alzheimer's Society is a care and research charity for people with Alzheimer's disease (and other forms of dementia) and their families. As well as a national helpline, there are over 250 local branches.

Helpline: **020 7423 3500**

Email: enquiries@alzheimers.org.uk
Website: http://alzheimers.org.uk

Carers UK provides advice and information to carers and the professions who support carers.

Carers UK Adviceline: **0808 808 7777** (Wednesday and Thursday 10am to 12pm and 2pm to 4pm)

Email: adviceline@carersuk.org
Website: www.carersuk.org

Dementia Information Service for Carers

Helpline Number **0845 1204048**

Call in normal office hours. Answer phone at other times.

Email: info@dementiaweb.org.uk

Web: www.dementiaweb.org.uk