

## Dementia - frontotemporal dementia with Parkinsonism-17

Frontotemporal dementia with Parkinsonism-17 (FTDP-17) is one of a group of dementias that affect the frontal (front) and temporal (side) regions of the brain. Symptoms usually appear between the ages of 40 and 60 years and get progressively worse over time.

Symptoms may include personality changes with loss of social skills, loss of cognitive (thinking-related) skills and Parkinsonism (slowed movements, rigidity, lack of facial expression and problems with balance).

FTDP-17 is rare and accounts for only about three per cent of all cases of dementia. It includes some instances of what was once called 'Pick's disease' and is sometimes confused with Alzheimer's disease.

FTDP-17 is usually caused by alterations (mutations) in a gene on chromosome 17. There is currently no cure but treatment may ease some of the symptoms.

### Symptoms

Symptoms can vary from person to person, but may include:

- **Parkinson-like symptoms** – these may develop early in the disease. Symptoms include lack of facial expression, slow movements, rigidity and balance problems.
- **Behavioural changes** – the inability to behave appropriately in different settings is typical of this condition.
- **Lack of awareness** – the person fails to realise that their behaviours are changed and inappropriate.
- **Diminished cognitive skills** – functions such as logical thinking, reasoning, problem solving and planning are impaired. Memory problems may occur as the disease progresses.
- **Psychiatric problems** – these may include delusions and hallucinations.
- **Slips in personal hygiene** – these may include neglecting to wash or dress properly.
- **Displays of excitable emotions** – these may include aggressive outbursts and angry pacing.
- **Inappropriate sexual behaviour** – the person may engage in inappropriate or disinhibited sexual behaviour.
- **Emotional coldness** – emotions such as love, empathy and sympathy may be lost. The person may appear indifferent to family and friends.
- **Repetitive actions** – these may include hand wringing, clapping or singing the same song over and over.
- **Impulsive behaviours** – the person may engage in behaviours such as impulse buying or shoplifting.
- **Unusual eating habits** – binge eating, limiting the diet to particular foods, excessive drinking of liquids including alcohol and trying to eat non-food items are some of the unusual habits the person may develop.
- **Language problems** – these may include difficulties finding the right word to say, and difficulties with reading and writing.

### The role of tau proteins

FTDP-17 is caused by abnormalities or imbalances in the tau proteins. These proteins are naturally found in the human brain and provide essential support to cell structures (microtubules) that contribute to cell building, movement and division. The production of tau proteins is controlled by a gene on chromosome 17, known as the MAPT (microtubule-associated protein tau) gene. A variety of alterations (mutations) in the MAPT gene cause the tau protein irregularities that lead to FTDP-17.

## FTDP-17 is usually inherited

The majority of people with FTDP-17 have an affected parent. There is a one in two chance that the children of an affected parent will inherit the genetic alteration and are therefore very likely to also develop FTDP-17. This pattern of inheritance is called autosomal dominant. However, in many cases the affected parent dies before onset of the disease in their children, so recognition of the family history of this condition may be delayed.

Where the occurrence of this genetic alteration in a family **is** known, testing is available for family members before symptoms develop. Prenatal testing for at-risk pregnancies is also possible if the genetic alteration is known.

MAPT gene changes are found in about half of cases that present with one of the FTDP dementias, where there is a family history of the condition. They are rarely found in isolated cases.

## Disease progression

The onset of symptoms usually occurs between the ages of 40 and 60 years. The disease may take from five to 30 years to progress, with most cases progressing within 10 years. The affected person eventually reaches a profound state of dementia and can no longer talk. Their life span is significantly reduced as a result of FTDP-17.

## Diagnosis

Diagnosis methods can include:

- Medical history, including symptoms
- Family history, particularly if the person has two or more first-degree relatives with dementia or Parkinsonism that indicates an autosomal dominant pattern of inheritance
- Tests such as CT scan and MRI scan to check for atrophy (shrinking) of the frontal and temporal lobes of the brain
- Single photon emission computed tomography (SPECT) test to measure the pattern of blood flow in the brain
- A genetic test to confirm whether the MAPT gene alteration is present.

## Treatment

There is no cure for FTDP-17. Treatment aims to ease some of the symptoms and can include:

- **Levodopa drug treatment** – helps to dampen some of the motor symptoms. It is traditionally used in the treatment of Parkinson's disease.
- **Sedatives** – can help relieve symptoms such as agitation or wandering.
- **Antipsychotic drugs** – to help reduce the incidence of hallucinations.
- **Anti-epileptic drugs** – may be helpful if the person has epileptic seizures.

## Genetic counselling and support

It can be distressing to be told that you or a family member has a genetic disorder or is at risk of having one. Genetic counselling provides individuals and families with information about a genetic disorder and its likely impact on their lives. This can assist a person with FTDP-17 to make informed medical and personal decisions about how to manage their condition and the challenges it presents to their (and their family's) health and wellbeing. Prenatal genetic counselling is also available for parents to help them decide about a pregnancy that may be at risk of FTDP-17.

## Where to get help

- Your doctor
- Neurologist
- Clinical geneticist
- Genetic Health Services Victoria, Royal Children's Hospital Tel. (03) 8341 6200
- The National Dementia Helpline Tel. 1800 100 500
- Carer Support and Respite Coordination Centre Tel. 1800 059 059
- Carer's Resource Centres Tel. 1800 242 636
- The Aged Care Information Line Tel. 1800 500 853

## Things to remember

- Frontotemporal dementia with Parkinsonism-17 (FTDP-17) is one of several progressively worsening dementias that target the frontal and temporal regions of the brain.
- The cause is usually an alteration in the MAPT gene on chromosome 17.
- There is no cure for FTDP-17 but treatment can ease some of the symptoms.

**This page has been produced in consultation with, and approved by:**

Genetic Health Services Victoria

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