Huntington's disease

Summary

- Huntington’s disease is a neurological (nervous system) condition caused by the inheritance of an altered gene.
- Symptoms often don’t appear until the person is in their thirties or forties and not all people with Huntington's disease experience the same symptoms.
- Behavioural problems are thought to be caused by a combination of events, including damage to the brain as the disease progresses and the understandable frustration and depression that people feel when challenged by chronic illness.

Huntington’s disease (also known as Huntington disease) is a neurological (nervous system) condition caused by the inheritance of an altered gene. The death of brain cells in certain areas of the brain results in a gradual loss of cognitive (thinking), physical and emotional function. Huntington's disease is a complex and severely debilitating disease, for which there is no cure.

The most common symptom is jerky movements of the arms and legs, known as ‘chorea’. Chorea usually starts as mild twitching and gradually increases over the years. A person with Huntington's disease may also have difficulties with speech, swallowing and concentration.

Genetics of Huntington’s disease

Huntington's disease is caused by an altered gene. This gene is passed on from parent to child, but the condition isn’t obvious at birth. The symptoms usually, but not always, first appear when the person is approaching middle age. Huntington's disease is a slow, progressive condition that affects people differently.

A person with Huntington's disease may live for 15 to 25 years after developing the first symptoms. Diagnosis is based on a family history of Huntington's disease (when known), genetic testing, plus assessment of physical, neurological and emotional symptoms. There is no cure for Huntington's disease.

Gene testing for Huntington’s disease

A child born to a person who carries the Huntington's disease gene has a 50 per cent chance of inheriting the gene and developing the disease. People at risk can take a test to see whether they have inherited this gene. A person must be at least 18 years old and want to know their gene status before they can have the test. Deciding whether to take the test is a personal choice. Counselling is available to help the person with Huntington's disease and their family, carers and friends to cope with the genetic result.

Find out more about **genetic services** and **genetic counselling**.

Symptoms of Huntington’s disease

Huntington's disease symptoms fall into three types, being physical, cognitive and emotional.

Physical symptoms include:

- Mild twitching of the fingers and toes

- Lack of coordination and a tendency to knock things over
- Walking difficulties
- Dance-like or jerky movements of the arms or legs (chorea)
- Speech and swallowing difficulties.

Cognitive symptoms include:

- Short-term memory loss
- Difficulties in concentrating and making plans.

Emotional symptoms include:

- Depression (around one third of people with Huntington's disease experience depression)
- Behavioural problems
- Mood swings, apathy and aggression.

Depression

It is thought that around one third of people with Huntington's disease experience depression. Symptoms of depression, such as lack of drive, should be medically investigated and not simply assumed to be part of the disease process.

Suggestions for family members, friends and carers include:

- See your doctor for diagnosis. Medications are available to treat depression.
- Psychotherapy may be an option.
- Regular exercise and moderate sunlight exposure can help ease depression.
- Try to incorporate more activities that the person particularly enjoys into their daily schedule.

Huntington’s disease and behavioural problems

Behavioural problems associated with Huntington's disease are thought to be caused by a combination of events, including damage to the brain as the disease progresses, and the understandable frustration and depression that people feel when challenged by chronic illness.

Not all people with Huntington's disease will experience the same behavioural problems, since the disease affects people differently. The severity of behavioural changes can range from mild and barely noticeable to enormously disruptive. It is important for family, friends and carers to appreciate that the person’s behavioural changes are part of the disease and are not under their conscious control.

Loss of motivation

The sections of the brain that help us to plan, organise and commence actions are affected by Huntington's disease. The person may appear lazy, because they will do nothing (except, for example, lie in bed or watch television) if left to their own devices.

Suggestions for family members, friends and carers include:

- Realise that yelling or arguing can’t motivate the person.
- The person may respond well to doing things with others, so take the lead and encourage them to follow.
- Helping the person to participate boosts their sense of worth, which is vitally important to reduce the risks of depression.

Loss of task sequencing

Tasks have to be performed in a certain order. For example, washing the dishes requires filling the sink with hot

water and detergent, cleaning the dishes, drying them and putting them away. A person with Huntington's disease can remember the parts of the task, but not the correct order. They might fill the sink with hot water, but then put the dirty plates away without washing them.

Suggestions for family members, friends and carers include:

- Supervise and help the person to perform tasks in their proper sequence.
- Encourage the habit of doing one thing at a time.

Inability to block out distractions

Eating a meal while watching television or listening to music can be very difficult for a person with Huntington's disease, because they can't concentrate on both things at once. That's why it is recommended that meals should be eaten in a quiet environment.

Suggestions for family members, friends and carers include:

- Encourage the habit of doing one thing at a time.
- Remember that activities we take for granted, such as walking, can require concentration for the person with HD. They may not be able to carry on a conversation at the same time.

Reduced abilities

A person with Huntington's disease may seem more careless. For example, they may not clean the house properly or may fail to maintain their usual standards of personal hygiene.

Suggestions for family members, friends and carers include:

- Appreciate that the person is trying their very best. It is the disease that is affecting their performance, not laziness.
- The person may not even realise they have made errors. Make it a rule to check on their behalf.
- Try to establish strict routines for bathing.
- Don't deny them tasks simply because it is easier to do things yourself – continuing to make a contribution is important to the person's sense of worth.

Inappropriate social behaviour

The awareness of social conventions may decrease, resulting in (for example) lewd or rude comments to others. Some people with Huntington's disease no longer have the 'inhibiting' emotions of shame, embarrassment and fear that help keep social behaviour in check.

Suggestions for family members, friends and carers include:

- Explaining the inappropriateness of their behaviour may be lost on them. Yelling and arguing probably won't work.
- The person may not understand the inappropriateness of their behaviour, but they may adhere to rules if you set them.
- You may need to consider limiting your social events.

Irritability and aggression

Some people with Huntington's disease become easily irritated or angered. This can be partly caused by an inability to see things from another person's point of view. Some people with Huntington's disease may come across as self-centred and selfish.

Suggestions for family members, friends and carers include:

- Remember that the disease prevents the person from thinking in a flexible manner. They may be more

comfortable and easy-going in familiar environments and situations.

- Make sure they have enough control over their options. For example, a person may become stressed and irritable if they aren't allowed to choose their own clothes for the day.
- Look at what the behaviour may represent. For example, the person may be spitting out their food because they have too much in their mouth, not because they are deliberately trying to annoy.
- Remember the general rule of one thing at a time. Trying to do two things at once can cause agitation.
- Focus on positive behaviours and try as much as you can to ignore the rest.
- The give-and-take of a loving relationship is disrupted by Huntington's disease, since the person may have lost crucial emotions to the disease.

Difficulties with communication

As the disease progresses, the parts of the brain that help control the muscles of the face, throat and tongue are increasingly affected. This can cause the person to have considerable speech difficulties. The person may not initiate conversations either, as the sections of brain responsible for this are also impaired.

Suggestions for family members, friends and carers include:

- Don't assume they can't understand what you're saying just because they can't form an answer. Understanding is usually unaffected by the disease.
- Don't rush them. Allow plenty of time for their answer.
- If you see the person is having trouble expressing themselves, ask if they want help rather than doing it automatically. For example, the person may not like you finishing their words or sentences without their permission.
- Whenever you can, offer them choices instead of open-ended questions. For example, 'Would you like pasta or fish for dinner?' is easier to answer than 'What do you want to eat?'
- Flash cards with common responses such as 'yes' and 'no' may be useful.
- If the person progresses to the point of losing speech, continue talking to them as normal. Otherwise, you risk making them feel isolated and invisible.

Support for people with Huntington's disease

Individual and family support workers are employed in all regions to support people with Huntington's disease, those at risk and their families.

Services include:

- Information and counselling
- Assessment and referral
- Practical help with in-home support
- Practical help with accommodation and respite
- Ongoing support for carers
- Holiday and volunteer programs.

Where to get help

- Your **GP (doctor)**
- **Huntington's Victoria** Tel. (03) 9818 6333
- **Victorian Clinical Genetic Services**

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

Huntington's Victoria