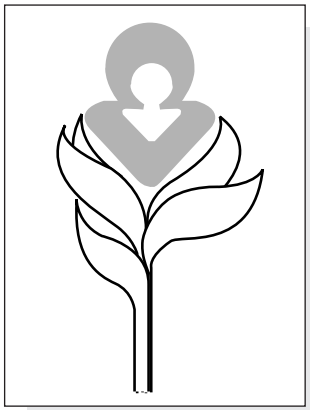
Huntington's Disease Association



General Information About Huntington's Disease

Fact Sheet 2

General Information about Huntington's Disease

Huntington's disease, which is often called HD, is an hereditary disorder of the central nervous system. It used to be known as Huntington's Chorea or HC. Huntington's disease usually develops in adulthood and can cause a very wide range of symptoms. It affects both men and women.

This fact sheet describes:

- what causes Huntington's disease
- the early symptoms of Huntington's disease
- how Huntington's disease progresses
- the treatment and help which is available

What causes Huntington's disease?

Huntington's disease is caused by a faulty gene on chromosome 4. The gene, which produces a protein called Huntingtin, was discovered in 1993. In some way - which is not yet understood -the faulty gene leads to damage of the nerve cells in areas of the brain including the basal ganglia and cerebral cortex. This leads to gradual physical, mental and emotional changes.

Each person whose parent has Huntington's disease is born with a 50:50 chance of inheriting the faulty gene. Anyone who inherits the faulty gene will, at some stage, develop the disease. A genetic test is available from Regional Genetic Clinics throughout the country. This will usually be able to show whether someone has inherited the faulty gene, but it will not show the age at which they will develop the disease.

The early symptoms of Huntington's disease

The symptoms of Huntington's disease usually develop when people are between 30-50 years old, although they can start much earlier or much later and can differ from person to person, even in the same family. Sometimes, the symptoms are present for a long time before a diagnosis of Huntington's disease is made. This is especially true when people are not aware that Huntington's disease is in their family.

The early symptoms include slight, uncontrollable muscular movements, stumbling and clumsiness, lack of concentration and short-term memory lapses, depression and changes of mood, sometimes including aggressive or anti-social behaviour. Great strain is put on relationships if unexpected temper outbursts are directed towards the partner. The time before a diagnosis is made can be very confusing and frightening because people do not understand what is happening and why.

Some people who know they are at risk spend time searching for the first signs that they are developing the disease. They may worry about simple things like dropping a cup, forgetting a name or becoming unusually bad-tempered. Most people do these things occasionally - whether they are at risk from Huntington's disease or not - so they could be worrying unnecessarily. Anyone who is concerned should have a word with their GP who may refer them to a neurologist for tests. These tests could include a number of simple assessments and possibly a brain scan. The genetic test referred to earlier may also be used to aid diagnosis.

How Huntington's disease progresses

Later on in the illness people experience many different symptoms but these may include involuntary movements, difficulty in speech and swallowing, weight loss, emotional changes resulting in stubbornness, frustration, mood swings and depression. Cognitive changes that people experience result in a loss of drive, initiative and organisational skills. This may result in the person appearing to be lazy. There may be difficulty in concentrating on more than one thing at a time.

Sometimes psychological problems, rather than the physical deterioration, cause more difficulties both for the person with Huntington's disease and their carer. Some changes are definitely part of the disease process although they may be made worse by other factors. It is depressing to have a serious illness and extremely frustrating not to be able to do things which previously seemed simple.

In the later stages of the disease full nursing care will be needed. Secondary illnesses, such as pneumonia, are often the actual cause of death.

What treatment and help is available?

Currently there is no cure for the illness but there are many ways to manage symptoms effectively Medication can be used to treat symptoms such as involuntary movements, depression and mood swings. Speech therapy can significantly improve speech and swallowing problems. A high calorie diet can prevent weight loss and improve symptoms such as involuntary movements or behavioural problems.

Social services in your local area can assist with practical issues like appropriate adaptations to your home if necessary and also assist with care at home or respite care. They can also assist with the provision of equipment if necessary. A referral can be made through your GP.

The Huntington's Disease Association produces a full range of literature that looks at these problems in more detail. We also produce literature for professionals who are involved in care.

The Association has a team of Regional Care Advisers who will be happy to offer you support, advice and information. Please contact Head Office for more details on 020 7223 7000.

Fact sheets available from the HDA:

1. All about the Huntington's Disease Association
2. General Information about Huntington's Disease
3. Predictive Testing for Huntington's Disease
4. Talking to Children about Huntington's Disease
5. Information for Teenagers
6. Eating and Swallowing Difficulties
7. Huntington's Disease and Diet
8. The Importance of Dental Care
9. Communication Skills
10. Behavioural Problems
11. Sexual Problems
12. Huntington's Disease and the Law
13. Huntington's Disease and Driving
14. Advice on Life Assurance, Pensions, Mortgages etc.
15. Seating, Equipment and Adaptations
16. Checklist for Choosing a Care Home
17. Advance Directive or "Living Will"

Booklets

Huntington's Disease in the Family (1997)
A booklet produced for young children

For a publication order form, membership form, details of our Regional Care Advisers and local Branches and Groups, please telephone or write to:

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Registered charity no. 296453

May 2006