



**Huntington's
Disease
Association**

Huntington's disease

**General information about
Huntington's and the
Huntington's Disease
Association**

General information about Huntington's disease

Huntington's disease (HD), is a 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 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. 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 more care and support will be needed.

What treatment and help are 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 Specialist HD Advisers who will be happy to offer you support, advice and information. Please contact the Operations team for more details on 0151 331 5444.

All about the Huntington's Disease Association

The Huntington's Disease Association exists to support people affected by Huntington's disease. The HDA also provides information and advice to professionals whose task it is to support Huntington's disease families.

Management Structure - The governing body of the Association is its Executive Council. This is made up of family members and professionals from England and Wales. There are separate organisations for Scotland, Northern Ireland and Eire. In addition, there is an external advisory committee, the Medical Advisory Panel, which advises on projects submitted by researchers for funds.

The Association is committed to supporting people affected directly or indirectly by Huntington's disease. Literature is produced on many aspects of the disease for families and professionals. Awareness raising events, such as conferences, are organised nationally and locally, together with social fun events, fund-raising activities and summer camps for young people.

Local branches and support groups form a significant part in these events. These groups also provide an invaluable role in creating opportunities for individuals to get together on a social basis and to learn more about the disease and the support and help available.

The HDA is financed through the generosity of Trusts, Foundations, the statutory and corporate sectors, not to mention the dedication and commitment of individuals fundraising for us. Our local branches do an amazing job in raising funds to support us.

The Association has:

- A central information, advice and support service - which operates from its registered office in Liverpool. The Operations Team consists of three paid staff. The office is open on weekdays from 9.00am to 17.00pm, Monday to Friday.
- A Specialist HD Advisory Service - which consists of a team of Specialist HD Advisers who provide information and advice to families; answer crisis calls; advise and liaise with other professional service providers; promote and develop a full range of local services; identify suitable respite and residential care facilities; liaise with local branches and self-help groups; give talks and organise seminars and training days in various parts of the country; provide speakers for training sessions and workshops for service providers and users such as health, social

services, nursing homes and residential care staff teams.

- Local branches and support groups throughout the country - which provide a local, informal setting for families and individuals to mix socially; offer support and advice; share experiences and ideas; form local links with professionals and other groups in their community; fundraise; organise speakers etc., and, above all, provide a forum to share a common purpose and reduce the feeling of isolation.

Who can provide help and advice?

Although there is currently no cure, there is much that can be done to help people affected by Huntington's disease to cope. The involvement of the Specialist HD Adviser, Dietitian, Speech Therapist and other professionals, can enable those affected by the disease to manage the changes effectively and positively.

Specialist HD Adviser:

As Huntington's disease is relatively rare, many health care professionals may only come into contact with one family. It is very important for these professionals to get specialist help and advice from the Specialist HD Adviser. The Specialist HD Adviser has acquired much of his/her knowledge and understanding of the disease from the shared experiences of families and health care professionals across the country.

The key role of the Specialist HD Adviser is to:

- Provide a listening ear.
- Help families receive support, care and advice that they want and need to enable them to cope.
- Access information on benefits, specialist equipment, respite, holidays and so on. Increase knowledge and skills. Assist in co-ordinating care through contact with health and social care professionals and non-statutory services.
- Work with our local branches and contacts, to raise awareness of Huntington's disease and the Association so that families and professionals will know where to get help, support and advice.
- To promote a better understanding of the disease among the general public.

The Association offers:

- A Helpline service - to provide help and support. A Specialist HD Adviser is available from Monday to Friday.
- Publications and information - we publish a range of booklets, leaflets, fact sheets, information packs for professionals etc., on various aspects of Huntington's disease.

- A Welfare grant fund - for providing small grants to families and individuals in particular need.
- Membership - If you join the Association, you will receive the twice-yearly national newsletter and, if your local branch or support group produces its own newsletter, you may opt-in to receive a copy of that too.

Membership for families is free but, as a charity, we do rely heavily on donations. Once you have been on the mailing list for 6 months you can also apply for voting membership. Voting members receive a copy of the HDA Annual Report and are eligible to vote at the Annual General Meeting. The AGM is usually held in the autumn.

- A Research programme - which promotes and funds both medical and social research. New applications from researchers are encouraged
- Fundraising - We have a range of fundraising, publicity and promotional items from collection boxes to publicity posters, sponsorship forms etc., (which can be personalised to your specific needs for activities and events). We have a dedicated events team to support you in your fundraising endeavours – please contact them on 0151 331 5445 to discuss your fundraising event.

Fact sheets available from the Huntington's Disease Association:

- General information about Huntington's disease and the Huntington's disease Association
- Predictive testing for Huntington's disease
- Talking to children about Huntington's disease
- Information for teenagers
- A young adult's guide
- Eating and swallowing difficulties
- Huntington's disease and diet
- The importance of dental care
- Communication skills
- Behavioural problems
- Sexual problems
- Huntington's disease and the law
- Huntington's disease and driving
- Advice on life assurance, pensions, mortgages etc.
- Seating equipment and adaptations
- Checklist for choosing a care home
- Advance Decision to Refuse Treatment (ADRT)
- A carer's guide
- Challenging behaviour in Juvenile Huntington's disease
- A brief guide to Juvenile Huntington's disease for children's hospices and palliative care services
- A teacher's guide
- A young person with Juvenile Huntington's disease at school

All fact sheets can be downloaded free of charge from our website www.hda.org.uk or ordered by phone 0151 331 5444 or email info@hda.org.uk

For a publication price list/order form, membership form, details of our Specialist Huntington's disease Advisers and local Branches and Support Groups, please phone 0151 331 5444 or email info@hda.org.uk