
Huntington's Disease

What is it?

Huntington's disease (HD) is an inherited, progressive degenerative brain disease that causes certain nerve cells to waste away, resulting in a loss of both mental capability and physical control. Over a period of 10-30 years, the ability to think, to speak and to walk is greatly diminished in persons with HD. A person does not die from the disease itself, but from the complications such as pneumonia, heart failure or infection developing from the weakened condition of the body.

Who does it affect?

About 1 in 15,000 people in New Zealand have HD. Onset is usually between ages 35 and 50 years after the person has had children and perhaps already passed on the gene. It affects both males and females and can be passed from one generation to the next by either the mother or father. HD affects all races and ethnic groups throughout the world.

Although rare, HD does appear in children. Younger persons often have a more severe condition.

Symptoms

Early symptoms appear slowly and may present as slight physical, cognitive or emotional changes. They may initially consist of 'nervous' activity, fidgeting and twitching in the extremities, or excessive restlessness. The affected person may notice certain clumsiness, alterations in handwriting, or difficulty with normal daily physical skills such as driving. Personality changes, from moodiness to paranoia, may occur.

As the disease progresses involuntary movements such as jerking and twitching of the head, neck, arms and legs become more marked. Speech becomes slurred, swallowing difficult and walk unsteady. There may be some short-term memory loss, personality changes and intellectual deterioration. Reasoning and judgement become impaired.

Development of the disease is slow and deterioration is gradual. Not every person has all the difficulties mentioned here. Some may have a great deal of involuntary movements, others little. Some may have marked emotional or cognitive difficulty, others very few.

Diagnosis

A family doctor will likely refer an individual to a neurologist for diagnosis, treatment and at least an annual, or more frequent, follow up, depending on each individual situation.

Huntington's disease can now be detected by a simple blood test. The gene that causes HD has been identified and individuals can be tested to see who will develop the disease. Prenatal testing (during pregnancy) is also available.

How it affects the Person

Difficulty with speaking and swallowing

Speech and language therapy will be helpful in providing strategies so that the affected persons will be able to speak as clearly as possible and to verbally express their thoughts and ideas for as long as possible. It is important that the person with HD clearly explains to family and caregivers their care needs and wishes before speaking becomes too difficult. When speaking does become too difficult, prior knowledge of the person's lifestyle, personality and care preferences will enhance their quality of care.

Difficulty swallowing and an increased risk of choking are also part of the disease process. Change in position during eating, varied food textures and adaptive equipment have all been beneficial in maintaining independent eating.

Family

Children face the prospect of inheriting the disorder. Fear about passing the gene on to the next generation influences their decisions about marriage, children and career choice.

Treatment

As yet there is no effective treatment for the condition, but some drugs are now available which are useful in reducing the severity of the abnormal movements. Other drugs are helpful in controlling the depression, moodiness, anxiety and irritability that can occur. The effectiveness of these drugs varies from person to person. It is important to provide accurate and comprehensive information to the affected person in a supportive and caring environment.

Physiotherapy and Occupational Therapy

In the early stages of HD the physiotherapy goal is to improve or maintain the person's functional level within the limits imposed by the disease process, promoting safe mobility for as long as possible. In the later stages the goal is to prevent or delay the complications of immobility such as joint contractures and muscle weakness.

The physiotherapist will assess areas such as balance, coordination, strength, flexibility and the ability to move around and participate in one's self-care. The need for walkers, canes etc. will be assessed, and the person will be trained in their use. The need for specialized seating will be examined to ensure a secure upright sitting posture is achieved. This is important as in the later stages more and more of the day is spent in a sitting position.

Occupational therapy primarily deals with independence in activities of daily living. Assistance from an occupational therapist may help to maintain or retrain such basic activities as cooking, dressing, eating, bathing, grooming, shopping and the use of public transportation which are all geared to keeping the affected person independent as long as possible. The occupational therapist may be able to make other suggestions for daily living such as using adapted equipment or making environmental modifications to the home, e.g., the use of grab bars in the bathroom to prevent falls.

Practical Suggestions for Coping with Huntington's Disease

Support: Get help early - counselling, assistance with care giving duties, etc. Involve your family from the beginning by sharing your concerns with them. Join a support group early.

Education: Access all the information you can about the disease and educate yourself as much as possible about its progression. Have an awareness of the losses to come, such as incontinence and inability to dress, so they are not totally unexpected.

Taking Care of Yourself: Have regular check-ups. Get as much rest and respite as possible. Eat well-balanced meals. Take care of yourself emotionally. Give yourself time to cry. Don't be afraid to acknowledge your feelings of anger, anxiety, helplessness, guilt and despair. Hang on to your sense of self by keeping up with your regular activities as much as possible.

Planning: Take one day at a time but don't neglect to plan for the future. Good planning can include getting a power of attorney, accessing community care early and filling our placement papers.

Communication: Learn how to communicate differently with your loved one if cognitive and language abilities decline. Good communication strategies help to avoid frustration.

Diet: Additional calories are required to prevent weight loss and secondary illness as HD progresses. It is necessary to plan food intake carefully, after assessing the individual's needs.

Where to go for Support

Huntington's Disease Association

The Huntington's Disease Association has newsletters (both national and local), articles, booklets, books and videos which provide information about Huntington's Disease, pre-symptomatic testing, and caring for people with HD. They have regular social get-togethers and Field Officers are also available to support families.

For more information phone 03 358 8470

Caring for Carers

All too often the burden of caring for a person with Huntington's disease falls on one or two of the immediate family members and the emotional, physical and financial hardships involved can be difficult to cope with. Carers can get discouraged and lonely, angry and depressed. Someone they have loved gradually disintegrates as a person. The whole relationship changes and new demands are placed upon the carer.

Caring for Carers has a Newsletter and several support meetings each month and ongoing telephone support. These services are provided to help make your job easier.

For more information phone 03 377 842

Each child of a parent who has HD, is at risk and has a 50% chance of inheriting the disorder.