

section 2.6.2: Neurodegenerative conditions

Huntington's disease (HD)

General description of the disease and progression

Huntington's disease is an inherited brain disorder which is characterised by degeneration of neurons. This degeneration results in uncontrolled movements, loss of intellectual faculties and emotional disturbances. Huntington's disease is a familial disease, caused by a dominant defective gene that may be passed on to a child of a person carrying the defective gene.¹

There is currently no treatment that can halt or reverse the disease. Medications can assist in the management of emotional and movement related symptoms but have associated side effects.

Potential problems

The symptoms of HD may vary in range and severity, age at onset, and rate of clinical progression from patient to patient – including among members of the same family (kindred).

Symptoms of disease may appear between the ages of 2-80 years – the peak age of adult-onset HD is between 35 to 50 years.

Evidence suggests that early onset is associated with increased severity as well as more rapid disease progression. For example, in adult-onset HD, disease duration is typically about 15 to 20 years. However, in juvenile HD, the disease course tends to have duration of approximately 8 to 10 years.

Specific considerations for end stage disease

- disease progression is frequently characterised by the following:
 - impaired concentration with intellectual tasks, slurred speech and decision-making becomes increasingly difficult
 - jerking movements become more uncontrolled
 - feeding, swallowing difficulties occur
- encourage the consideration for end of life care and advance care planning early in disease
- special sensitivity for other family members, such as the children of the patient, is essential given they may fear inheriting the disease – predictive testing and counselling is available² – see Resources at the end of this section.

references

¹ National Institute of Neurological Disorders and Stroke. see www.ninds.nih.gov/

² Huntingtons Association of South Australia and Northern Territory, see www.huntingtonssa.org/