

Juvenile Huntington Disease

When Huntington disease (HD) appears in someone under the age of 20, the illness is recognized as Juvenile Huntington Disease (JHD). HD is a hereditary, neurodegenerative disease characterized by a progressive loss of control over movement, emotion, and thinking. Approximately 1 in 7000 Canadians has HD. About 10% of these cases are JHD. While the disease has a high variability in presentation of symptoms, the average life expectancy for children and youth affected by JHD after diagnosis is approximately 15 years.

Movement:

- Contrary to adults with HD, children or youth tend to show only mild chorea (involuntary jerky movements of the arms, legs, trunk, neck or face) or no involuntary movements at all; instead they tend to become more rigid and stiff.
- The diminished coordination of voluntary movement can result in awkwardness in walking, clumsiness, loss of balance and falls. Tasks that had been easy to perform (e.g. riding a bike, balancing on a tree trunk) might become challenging. A young person with JHD may begin to walk on their toes or develop a stiff legged or scissored gait.
- Throughout the progression of JHD, there will be difficulties with chewing and swallowing as well as speaking.
- In about 25% of JHD cases, there is a tendency for epileptic seizures – something almost never seen in adults with HD.

Emotion:

- A child or youth with JHD will experience, on an ongoing basis, the loss of abilities, competencies and independence. Understandably, this can cause feelings of frustration, anger, sadness, fear and grief.
- It can become difficult for the child or youth to regulate and prioritize stimuli.
- Obsessive thoughts or irrational fears can add additional stressors, and at times, the young person may express negative emotions through aggressive behaviour, compromising the safety of the child or youth with JHD and others.

Thinking:

- JHD is a gradually progressing disease and symptoms can occur slowly – possibly a number of years before an official diagnosis can be made. Children may begin to struggle performing tasks they had mastered before (e.g. writing, reading and counting). When attention and concentration decline and it becomes more difficult to complete a task, the child may become anxious or frustrated.
- It can become increasingly difficult for a child or youth with JHD to learn new information and form new memories.
- A diagnosis of attention deficit disorder (ADD) may be discussed or the cognitive symptoms of HD may be mistaken for “bad behaviour”, when actually it is JHD that is interfering with the child’s ability to concentrate.

Puberty

- Puberty can become a particularly challenging time because the changes in teenagers’ bodies bring on normal sex drive and the need for independence. However, JHD affects teenagers’ abilities to control these natural behaviours and urges.
- It is important to remember that due to the changes caused by HD, the child or youth is not in control of his or her behaviour. Involvement of a pediatric psychiatrist and a behavioural therapist should be sought.

Diagnosis

Diagnosing JHD can be challenging. A physician may have to see the child several times before being confident that neurological symptoms are apparent. A complete and accurate family history can be invaluable in evaluating a child with symptoms suggestive of Huntington disease. However, there are situations in which parents may not even be aware that HD is in the family, or in other cases, adoption with unknown family history may be involved. If a physician suspects JHD, a confirmation of the diagnosis will be sought through diagnostic genetic testing.

Diagnostic Genetic Testing: Physicians can seek confirmation of a suspected neurological disorder through Diagnostic Genetic Testing. In JHD, the CAG repeat expansion - which is the genetic mutation that leads to HD - involves a larger number of repeats than those found in adult HD.

Predictive Genetic Testing: Predictive Genetic Testing refers to a process whereby an individual with a family history of Huntington disease wishes to know whether or not he or she has inherited the expanded HD gene, even though no symptoms are currently present. Predictive genetic testing is not available in Canada for persons under the age of 18. For youth over 18 years, please consult with a HSC Family Services team member or the local genetics clinic for more information.

First Symptoms

The early signs of Juvenile Huntington disease will represent a noticeable change from the past behaviour of the child. These symptoms may include:

- rigidity
- slowness and stiffness
- awkwardness in walking
- diminished coordination
- personality changes
- changes in behaviour and poor judgement
- slowness in responding
- variable/poor school performance
- difficulty in learning new information
- unable to do things previously learned

Living with Juvenile Huntington Disease - Building Your Team

At School:

- Enable the child or youth with JHD to participate in as many activities of the student population as possible for as long as possible.
- Friendships and experiences made at school give the young person with JHD contacts and memories that will be increasingly important as the disease progresses.
- Re-evaluate the individual teaching plan on an ongoing basis and adjust activities to current level of ability and needs.
- Educate all staff on the symptoms of JHD to foster the establishment of an understanding environment.
- Allow for frequent periods of rest: A child or youth with JHD can be easily exhausted. Every task and activity requires much strength, energy and perseverance to compensate for the physical and cognitive changes occurring in the brain.

The Medical Team:

- Integrate suggestions and strategies from Physiotherapist, Occupational Therapist, Speech Language Pathologist, Behavioural Therapist, Recreational Therapist, Dietitian, Psychiatrist, Social Worker and Neurologist into daily activities at home and at school (e.g. assistive devices, new routines and approaches).

The Family:

- Supporting a child with Juvenile HD can increase emotional, financial and physical stress. Seek professional support and services (e.g. counselling, respite, home care) for all family members as JHD progresses.
- Search for provincial and federal funding sources that may provide respite care and financial relief.
- In addition to the practical aspects of care, it is important for families to enjoy and celebrate life together. Find activities that the child or youth enjoys and make life as safe, happy and fulfilling as possible.

Treatments

Juvenile HD remains incurable, and there are no treatments which can stop or slow the course of the disease at this time. However, there are medications that help alleviate some of the symptoms. Developments in research have given rise to tremendous optimism that new treatments will soon be within sight.

Additional information on JHD can be found on the HSC website (www.huntingtonsociety.ca) and in the following HSC resources: *A Physician's Guide to the Management of Huntington Disease* and the booklet entitled *Juvenile Huntington Disease*. You may also contact your local Family Services team member for more information and support.

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