JUVENILE HD

Huntington disease (HD) is a genetic neurodegenerative illness and when HD appears in someone under the age of 20, the illness is recognized as Juvenile Huntington disease (JHD).

- “genetic” means it runs in families and the HD mutation is passed on from a parent to a child
- “neuro” has to do with cells in the nervous system, specifically in the brain
- “degenerative” tells us that it gradually gets worse over time
- “disorder” is another word for disease

For more information about the genetics of JHD, speak with the regional genetics counsellor or see the link to the HSC fact sheet below (Genetic Testing and Huntington Disease).

Symptoms:

Changes to the body in JHD tend to occur faster than with those who have adult-onset HD. The speed and the order in which these changes happen varies from person to person. Just like in adult HD, symptoms of JHD cause changes to three main areas: physical (movement), psychiatric (emotion) and cognitive (thinking). However, there are also some key differences, which will be discussed below.

Physical symptoms:

- Children and young people affected by JHD are less likely to experience the involuntary, jerky movements known as chorea. They are more likely to be affected by muscle contractions, rigidity and stiffness.
- There may be symptoms including tremor, slowness and stiffness that are similar to those seen in patients with Parkinson’s disease. This may be referred to as the Westphal Variant.
- The diminished coordination of voluntary movements can result in awkwardness in walking, clumsiness, loss of balance and falls. The young person with JHD may begin to walk on their toes or develop a stiff legged or scissored gait.
- There may be difficulties with chewing and swallowing, as well as speaking, and this can occur very early on or as the condition progresses.
- In about 25% of JHD cases, there is a tendency for epileptic seizures, something rarely seen in adult HD.

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Psychiatric symptoms:

- 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.
- Obsessive thoughts or irrational fears can add additional stressors. At times, the young person may express negative emotions through aggressive behaviour, compromising the safety of the child or youth with JHD and others.
- Challenging behaviour tends to be more common in individuals who develop the disease in their teens, but many young people are not affected in this way.
- It is important to remember that challenging behaviour is not always related to JHD. Children of families affected by HD may also be experiencing disruption and difficulties in their home life, which may also impact their behaviour, or they may be facing other challenges in their life.

Cognitive symptoms:

- JHD is a progressive disease and symptoms can occur slowly – possibly a number of years before an official diagnosis can be made. Children or youth may begin to struggle with performing tasks they have mastered before (e.g., writing, reading and counting). When attention and concentration decline and it becomes more difficult to complete a task, the child or youth 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.
- Changes in behaviour or a drop in school performance are often the first noticeable symptom of JHD.
- A diagnosis of attention deficit disorder (ADD) may be discussed but it is important to consider that the cognitive symptoms of JHD may be interfering with the child's ability to concentrate.

Another consideration for those living with JHD is 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 may affect the youth's 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 may not be in control of his or her behaviour. A pediatric psychiatrist and a behavioural therapist would be important care team members to include if possible.

Diagnosis

Diagnosing JHD can take time; symptoms can resemble those of other diseases and since it is so rare, there can be a lengthy assessment process.

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Sharing a complete and accurate family history with the doctor can be invaluable. The doctor may confirm a diagnosis of JHD using genetic testing. In the situation when a parent has been diagnosed with HD, this may lead to an earlier diagnosis. In some situations, there may be no awareness of HD in the family, or the family medical history may be unknown due to an adoption or other reasons. Sometimes a youth may be diagnosed before their parent who is at risk, seemingly skipping a generation. This is called genetic anticipation and should be discussed with the doctor and genetics counsellor.

Receiving a diagnosis of JHD or waiting for a diagnosis to be confirmed can be stressful. The HSC Family Services team is here to provide support and resources along the way.

Treatment:

Important research focused on the treatment of HD is ongoing. There is much hope that this will lead to new treatments that could slow or halt the progression of JHD and HD. At this time, there are medications that help alleviate some of the symptoms. It is very important to build a care team. Many professionals can provide help and support for someone with JHD along their journey.

Who Can Help?

Health Professionals: Depending on where you live, you might find your care team at a local children's hospital or children's treatment centre. This care team might include a neurologist, physiotherapist, occupational therapist, speech and language pathologist, therapeutic recreation worker, psychiatrist, dietician, social worker, behavioural therapist and others. In addition to important medical care, they can offer suggestions, strategies and make recommendations for equipment such as assistive devices, or useful modifications for home and school.

School Community: Education about JHD will provide tools to teachers and other school staff so they can provide the support needed and make the necessary accommodations as the disease progresses. This will create a more inclusive learning environment for a child or youth with JHD. Social connections through friendships and school activities continue to be important for the person living with JHD.

Financial Support: Financial assistance through provincial and federal funding sources for things like medical equipment or services such as respite are available. A Huntington Society of Canada Family Services team member can help you access these professionals and resources.

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For the Family...

Living with JHD is challenging in many ways, as every family member will face JHD their own way and within their role in the family. Some considerations for carers include increased care needs as JHD progresses, financial implications due to the increased costs involved with care, and the sense of isolation family members may feel as this rare disease affects the family. It can be very helpful to connect with others in similar circumstances.

The HSC Family Services team offers a variety of support groups and youth programs such as Young People Affected by Huntington Disease (YPAHD) and the Youth and Young Adult Mentorship Program. Online information sites such as HDYO (Huntington Disease Youth Organization) are great ways to connect and learn about JHD as well.

Sibling support groups may also be available through other community-based services. Respite services are available for in home or residential settings when needed for taking some time away from caregiving. Individual counselling is another support option that may be helpful.

Beyond the practical aspects of care, it is important for families to enjoy time together. There are activities the family can do that are safe, enjoyable and fulfilling. It is possible to still have fun and make memories.

RESOURCES

What is Juvenile Huntington's disease?

What is Huntington disease?

Genetic Testing and Huntington disease - Fact Sheet

Kids Sometimes Get Huntington's disease Too - HDBuzz

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