WHAT IS HUNTINGTON DISEASE (HD)?

Huntington disease is a genetic neurodegenerative disorder. What does this mean?

- “genetic” means it runs in families
- “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

Therefore, HD affects the nerves in the brain and it gets worse over time. HD most commonly shows up in adult years and as it progresses, it affects a person’s physical, thinking and emotional abilities.

Symptoms of HD

HD symptoms fall under three main areas, so we refer to a triad of symptoms. Here are the three main areas:

- Physical symptoms: affect how the person moves, such as involuntary dance-like movements (also called chorea); diminished coordination; difficulty walking, talking and swallowing; weight loss
- Cognitive symptoms: these affect how the person thinks, such as difficulty with focus, planning, recall of information and making decisions; impaired insight
- Psychiatric symptoms: affect how the person feels and behaves, such as depression, apathy, irritability, anxiety, obsessive behaviour

It is important to note that every person living with HD will experience symptoms and progress in a different way. Signs and symptoms usually appear between the ages of 35 to 55.

A diagnosis in older adults, after the age of 60, would be considered late onset HD (about 10% of total cases). In youth under 20 years of age, it would be considered Juvenile HD (JHD) (about 10% of total cases). JHD tends to follow a more rapid progression. For more information on JHD, please see the link to the HSC fact sheets below.

Who Gets HD?

Huntington disease is a genetic illness and the HD gene is dominant. This means that only one copy of the HD expanded gene is needed. Each child of a parent with HD has a 50% chance of inheriting the expanded gene and said to be “at-risk” for developing HD. Males and females have the same risk of inheriting the disease. HD occurs in all parts of the world.

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STAGES OF HUNTINGTON DISEASE

Early Stages: In the early stages, people with HD can function well at work and at home. Signs and symptoms of the disease may include:

- Difficulty organizing routine activities or coping well with new situations
- Decreased ability to recall/remember information and make decisions
- Increased difficulty with work activities
- Decreased attention to details
- Mood changes and irritability
- Minor involuntary movements (e.g., "nervous" activity, fidgeting, a twitching of the limbs or restlessness)
- Changes in handwriting or difficulty with daily tasks such as driving

Intermediate Stages: People with HD in the intermediate or middle stage may have more difficulty working and managing a household but can still deal with most activities of daily living. Activities of daily living are routine things people do without help, such as eating, bathing, getting dressed, toileting and mobility. Symptoms may include:

- Increased involuntary movements (chorea)
- Increased difficulty with walking, coordination and balance
- Challenges with speaking (speech may become slurred) and slower in thinking
- Solving problems becomes more difficult
- Difficulties with swallowing
- Weight loss

Advanced Stages: People in the advanced stages of HD can no longer manage the activities of daily living. They often require daily health care support at home (such as Homecare) or they need to live in a supportive care setting. Symptoms may include:

- Decrease in involuntary movements and increase in rigidity (or stiffness)
- Increased difficulties with swallowing
- The ability to speak lessens, but it is likely they continue to understand what is being said
- Significant weight loss
- No longer able to walk
- Completely dependent on others for daily activities

To date, there are no drugs to slow or stop the progression of HD. There are specific drugs available to ease some symptoms. Research within Canada and globally is ongoing and holds promising options for treatments for the future.

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RESOURCES

Ongoing support, education and information is available from the Huntington Society of Canada (HSC). You can find a listing of our Family Services team members at www.huntingtonsociety.ca/family-services-team.

- A Physician's Guide to the Management of Huntington Disease
- Understanding Behaviour in Huntington Disease
- A Carer's Guide to Huntington Disease
- Jimmy Pollard: Easy But Difficult Accommodation In Cognitive Care (Video)
- Amanda's Story About Huntington disease (Video)
- What is Huntington disease? (Video)
- HSC educational modules (especially Responsive Behaviours, Caregiver Overview, Thinking Module). Connect with the regional Resource Centre Director (RCD) for a presentation based on these educational modules.
- Fact sheets on a variety of other topics including Cognitive Changes in HD, Have you Met HD, Responsive Behaviours, Tips When Working with Individuals with HD, and JHD are available at www.hdfactsheets.ca.

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