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Health Matters

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Huntington Disease:
The importance of a team approach

HD affects whole family

Although Huntington Disease (HD) is a relatively rare genetic disorder, there is at any given time about 5,000 sufferers in Canada, virtually all of whom will require long-term care during late stage disease.

In general, HD causes progressive degeneration of neurons in the brain leading to changes in a person's ability to control body movements (chorea). Affected individuals will also experience changes in mental processing, learning and memory, which will be accompanied by changes in mood and various psychological disturbances.

Some sufferers of HD and their caregivers say: **"It's like having like having Alzheimer, Parkinson's, schizophrenia and ALS all at once."**

There is no cure for HD, and care requires involvement of a primary care physician in concert with a neurologist, psychiatrist and many allied health professionals from nurses and pharmacists to social workers, physiotherapists, dentists, dieticians, hospice workers, and more.

The key for all who work with individuals that have HD is the need to understand the disease affects whole families and requires a team approach.

HD is an autosomal dominant disorder…

This means that if one parent (either a mother or father) carries a defective allele of the Huntington gene (located on chromosome number four), each successful union of egg and sperm will result in a 50 percent chance of inheritance.

However, offspring who do not inherit the mutation will never develop the disease, nor will their offspring.

In rare instances where both parents carry a defective allele, all offspring (100 percent) will inherit HD.

Disease onset usually occurs between ages 30 and 50…

This means that is some cases, a parent may not know they're a carrier of the mutation, which can subsequently cause great emotional turmoil for those who suddenly realize the implications for their children.

Additionally, children will witness a parent's disease progression, and may know (as a result of genetic testing) that they themselves also have HD.

In some families, due to the multigenerational impact of the disease, there could be several affected individuals within the extended family, all suffering simultaneously.

Note: A small subset of affected individuals will have juvenile onset HD but the disease symptoms and features are somewhat different.

Death usually occurs five to 15 years after disease onset

Death may be caused by medical issues, such as aspiration pneumonia, other types of infections, or malnutrition. Or, death may follow complications of falls or other kinds of trauma.

In addition to loss of life, individuals and family members experience multiple losses over time: Loss of family roles, income, intimacy, identity and so on. Grief and depression extending to suicidal ideation is not uncommon.

Treatment

- Neuroleptic drugs and tetrabenazine are used to treat chorea
- Antidepressants and anxiolytic medications can be used to treat depression
- Explosive behaviors, psychosis, obsessive-compulsive symptoms and apathy can be reduced with medication
- Sleep disturbances and pain can also be treated

Beyond medication, the LTC nursing team's role remains key and involves reassuring the individual with HD (as well as family-members) that they will be safely cared for, and love ones will remain involved and part of their life.