Huntington's disease is a congenital disorder involving degeneration of nerve cells in the brain due to a faulty gene. Symptoms of the disorder include progressive loss of mental functions, speech impairment, psychosis, paranoia, hallucinations, and abnormal facial and body movements. There is no cure for the disease. Treatment involves slowing the progression of the disease. Usually the symptoms do not appear until adulthood. The disorder is usually fatal within 15 to 20 years. See also Birth Defects and Abortion

Huntington's disease is a rare neurodegenerative genetic disorder with psychological, physiological, and social repercussions. It is characterized by the onset of psychiatric symptomology, declining muscle coordination, and loss of cognitive abilities. Although genetic in nature, Huntington's disease is significantly impacted by culture in a number of ways. Among them are risk factors for the disease, variation in prevalence rates among countries, interpretation of symptoms, and treatment and responses to the illness.

Background
Prior to first being identified as a separate disorder, people with Huntington's disease were sometimes thought to be witches or possessed by demons and later were simply lumped together with “the insane.” This resulted in people with the disorder being shunned or exiled from their communities. In 1872, George Huntington noticed that this particular collection of symptoms ran in families, suggesting a genetic origin to the disorder.

Because Huntington's disease is caused by a genetic defect, the increase of prevalence and earlier age of onset are less likely due to cultural influences; rather, the factors include better detection and an increase in incidence of cases. These new cases are attributable to how the genetic defect causing Huntington's disease is passed from parent to child at a rate of approximately 50 percent as well as how the genetic repeat increases with each new generation. In 1993, Huntington's disease was attributed to a genetic defect on chromosome 4. A section of DNA known as a CAG repeat happens significantly more often in people with the disorder. In most people, the CAG repeat occurs between 10 and 28 times; however, for those with Huntington's disease, the repeat occurs up to 120 times. As this genetic repeat is passed from parent to child, the repeats increase, resulting in an earlier age of onset for the next generation.

Because most cases of Huntington's disease have an age of onset during midlife, in the absence of genetic testing, many people with the disorder have already had children by the time they realize that they have the disease. About 10 percent of identified cases have juvenile Huntington's disease with an onset prior to age 20.

Symptoms and Risk Factors
Huntington's disease manifests as various cognitive, behavioral, mental health, and psychomotor symptomology. Symptoms of dementia such as memory loss, confusion, and loss of judgment and speech are common. Behavioral and mental health symptoms characterized by mood changes,
psychosis, hallucinations, and anxiety are reported. In particular, caregivers of those with Huntington’s disease are often warned of the increased risk of depression and suicide among sufferers. Noticeable changes in gait and movement are noted, such as facial grimaces, sudden jerking movements, tremors, and rigid or slow movements. In the later stages of the illness, individuals with the disease no longer are able to walk, communicate, and eat by themselves, but they are still conscious and able to recognize family members.

Folk musician Woody Guthrie in 1943. By the late 1940s, Guthrie’s health was declining and various diagnoses such as alcoholism and schizophrenia attempted to explain his erratic behavior. In 1952, he was diagnosed with Huntington’s disease.

The wide array of symptomology has been differently interpreted by various cultures. It is not uncommon for traditional cultures to attribute such a collection of symptoms and behaviors as resulting from a magic spell or curse.

A child of someone who is inflicted with Huntington's disease runs a 50 percent chance of inheriting the disorder. However, approximately 8 percent of Huntington's disease cases are attributed to new genetic mutations and are not inherited from a parent. Huntington's disease is just as common regardless of gender. However, distinct ethnic trends are partly due to how Huntington's disease has developed. It is commonly believed that the Huntington genetics repetition developed independently in Europe, Japan, and Africa, resulting in distinct varieties of the genetic mutation. However, those with European ancestry have the greatest incidence of Huntington's disease.

**Prevalence Rates by Country**

Although Huntington's disease is found in various countries and cultures around the world, prevalence rates vary by culture of origin. The highest rates of Huntington's disease exist in Europe and countries where Europeans have settled, such as Australia, Canada, and the United States. Countries with the lowest rates of Huntington's disease are China, Japan, and the nations of Africa. More specifically, the comparative prevalence rates per 1 million people are United States (100), Japan (1 to 4), Hong Kong (3.7), South Africa (0.6), and European countries (40 to 100). However, much of the variation in
prevalence rates around the world can be attributed to European migration patterns.

One interesting exception is the drastically lower prevalence rates in Finland, with only six cases per 1 million people, as compared to other European countries. This is attributed to Finland's genetically distinct lineage. Some reports suggest that the largest prevalence rates of Huntington's disease exist in Zulia, Venezuela. Researchers attribute this large concentration of people with the disorder to the isolated and impoverished nature of this area. The geographic isolation of this area increases the likelihood of both parents being genetically predisposed to developing the disease and, due to financial constraints, genetic testing prior to having children is not employed. As a result, children in this area can run a three in four chance of developing the disease.

Treatment and Care

For an adult sufferer, once the early signs of the disease manifest and a person is diagnosed with the disorder, symptoms usually progress over the next 15 to 20 years until death. This progression occurs more quickly for an individual with early-onset Huntington's disease, usually between 10 to 15 years. Currently there is no known treatment that will slow or stop the progression of Huntington's disease, although some medications have been found to manage some of the symptomology.

Given that Huntington's disease is a progressively worsening condition requiring 24-hour care in the later stages, economic resources and cultural values play a significant role in how those with the illness receive care. Traditional cultures with strong family values often provide care themselves, whereas more modern and mobile cultures often seek out residential care for those family members in later stages of the illness.

Economic resources are vital for the extensive care required to manage Huntington's disease. For those individuals without health insurance or who live in countries with poorly established health care systems, the burden of care often falls on family members. This can be devastating to families, given that the genetic nature of the disorder causes many families to have multiple members with Huntington's disease. For example, in the previously mentioned state of Zulia, Venezuela, Huntington's disease is so common that almost every household has members in various stages of the illness. As a result, children are often not able to attend school as they are either inflicted with the disease or are caring for a family member with it.

Since the overall number of people afflicted with the disease is relatively small, research into a cure for Huntington's disease offers little in the way of financial reward. In the United States, it is estimated that about 30,000 people currently have Huntington's disease. Pharmaceutical intervention research has historically been limited because of the small market of people inflicted with Huntington's disease.

Social Challenges

Given the established genetic link to developing Huntington's disease and the particular symptomology, people with Huntington's disease endure significant legal and social difficulties related to the disease. Historically, people with the disease have been unable to secure health insurance due to the genetic nature of this degenerative disorder. With no established treatments to stop the progression of the disease, those diagnosed will eventually require 24-hour care, which is extremely costly. In addition, people with Huntington's disease often endure discrimination in the workplace. Many of the symptoms indicative of Huntington's disease, such as behavioral disturbances, moodiness, slurred speech, and unsteady gait are commonly misinterpreted as intoxication. This can lead to tremendous discrimination.
and stigma for those suffering with Huntington's disease, as well as their families.

Moreover, it is also difficult for the children of sufferers, given that they run a 50 percent chance of developing the disease themselves. Cultural and religious beliefs also impact the decision to genetically test—prior to the development of symptoms—children who are at risk of developing the disorder. Some cultures are more familiar with genetic testing, while in others it is relatively uncommon.

See Also: Board and Care Homes Children Cultural Prevalence Family Support Genetics Incidence and Prevalence Measuring Mental Health Policy: Medical

Further Readings


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