Overview: Huntington disease is a progressive neurological genetic disorder that causes changes in brain nerve cells. These changes result in symptoms that can affect movement, thought, behavior, and mood of those with the disorder.

Huntington Disease

Inheritance: Huntington disease is an autosomal dominant disorder. This means that only one copy of the altered HTT gene is needed for the disease to be present.

Symptoms: Symptoms of Huntington disease usually start to appear in a person’s 30s or 40s. Early symptoms can include small involuntary movements – a hallmark of HD known as chorea – as well as irritability, depression, poor coordination, trouble learning new things, and difficulty with decision making. As the disease progresses, the involuntary movements become more pronounced. Difficulty with speaking, walking, and swallowing are other possible symptoms. People with Huntington disease may also have personality changes and declines in thinking and reasoning. Most people live 15-20 years after they are diagnosed.

Causes: The disease is caused by a defective HTT protein, which is made from an HTT gene with an increased number of CAG tri nucleotide repeats within the gene. The normal HTT gene has 26 or fewer CAG repeats, while the abnormal version has more than 26 repeats. The additional repeats result in a protein that is toxic to cells.

Treatment: Huntington disease has no known cure. Currently, treatment focuses on treating the symptoms of the disease via medication, therapy, and counseling.