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Short Note on Niemann-Pick Neurological Disorder

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Introduction

A rare genetic illness called Tay-Sachs is passed from father to son. It is brought on by a deficiency in an enzyme that helps break down fatty compounds. These fatty compounds, known as gangliosides, build up in the brain and spinal cord to hazardous levels, affecting nerve cell function. The most prevalent and severe form of Tay-Sachs disease first exhibits symptoms between the ages of 3 and 6 months. Growth slows down and muscles become weaker as the condition worsens. This leads to seizures, eyesight and hearing loss, paralysis, and other catastrophic issues [1].

Description

Although Niemann-Pick disease can affect people of any age, it mostly affects children. The illness is potentially deadly and has no known cure. Certain genes involved in fat metabolism are mutated to produce Niemann-Pick disease (cholesterol and lipids). Autosomal recessive inheritance is the method used to transmit the mutations in the Niemann-Pick gene from parents to offspring. This indicates that both the mother and the father must transmit the gene's faulty form for the child to be impacted [2].

There is no treatment for Niemann-Pick disease, which is incurable. At any age, it can occur. Sphingomyelinase is an enzyme that is either absent or defective in Types A and B, resulting in these conditions. Fat builds up in cells as a result of the body's impaired capacity to metabolise fat (cholesterol and lipids). Cellular malfunction and ultimately cell death occur from this. Infants with severe, progressing brain diseases, such as Type A, are most commonly affected. Most children do not survive their first few years since there is no cure. Type B typically appears later in life and has no connection to fundamental brain disorders. The majority of type B patients survive into adulthood. An very uncommon genetic disease is Niemann-Pick type C [3,4].

A rare genetic illness called Niemann-Pick disease makes it difficult for the body to metabolise fat (cholesterol and lipids) inside of cells. These cells degenerate and finally pass away. Niemann-Pick disease can harm the brain, nerves, liver, spleen, bone marrow, and, in extreme cases, the lungs. The symptoms of this disorder are brought on by the progressive loss of brain, nerve, and other organ function. Although Niemann-Pick disease can affect people of any age, it mostly affects children. The illness is potentially deadly and has no known cure. The goal of treatment is to help individuals manage their symptoms [5].

Niemann-Pick disease is identified by its signs and symptoms of clumsiness and trouble walking. Niemann-Pick disease also manifests as excessive eye movements or dystonic muscular contractions. difficulty swallowing and eating, disturbed sleep, pneumonia that persists The three primary Niemann-Pick types are types A, B, and C. Depending on your condition's kind and severity, your signs and symptoms will change. Within the first several months of life, some infants will display signs and symptoms. Patients with type B have a better chance of surviving to adulthood and may not have symptoms for years. Until they are adults, people with type C may not show any symptoms. Certain genes involved in fat metabolism are mutated to produce Niemann-Pick disease (cholesterol and lipids).

Conclusion

Autosomal recessive inheritance is the method used to transmit the mutations in the Niemann-Pick gene from parents to offspring. Accordingly, both the mother and the father must pass on the gene's faulty version for the child to be impacted. There is no treatment for Niemann-Pick disease, which is incurable. At any age, it can occur medication that decreases saliva production and posture strategies are other options to lower the risk of mucus collection and avoid aspiration pneumonia.

Acknowledgement

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Conflict of Interest

None.

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