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Short Notes on Copper Storage Disease

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Introduction

Copper builds up in the liver, brain, and other important organs in Wilson's disease, a rare genetic condition. Wilson's disease can affect adults as well as children, but it is most commonly diagnosed in people between the ages of 5 and 35. Copper is needed for healthy neuron development, bone growth. collagen synthesis, and melanin production. After being absorbed from food (bile), excess copper is typically eliminated through a chemical produced by the liver. However, Wilson's disease prevents copper from being adequately eliminated and causes its concentration to reach potentially fatal levels. Wilson's disease [1-3] can be treated early, and many people who suffer from it lead normal lives.Wilson's disease is present at birth, but symptoms do not appear until copper accumulates in the brain, liver, or another organ. The disease has different signs and symptoms depending on which parts of your body are affected. They might be tiredness, a lack of appetite, or pain in the abdomen. a discoloration of the eyes that is golden-brown in color (Kayser-Fleischer rings), also known as jaundice. Wilson's disease is an autosomal recessive trait, which means that each parent must give you one copy of the defective gene to get the disease. You will not get sick if you only have one abnormal gene, but you will be a carrier and have the ability to pass the gene on to your children.

Description

There may be and continue to be a number of neurological problems in Hunter syndrome children. There are a number of neurological issues that can arise from excess fluids in your child's brain. Your child's mental state, vision, and severe headaches may all be altered by the pressure from these fluids. Additionally, there is a possibility that your child will develop a condition in which the membranes that surround the spinal cord become scarred and thickened [4,5]. The upper spinal cord is compressed and put under pressure as a result. Your child may, as a result, experience leg fatigue, gradually become weaker, and become less active.

Wilson's ailment is available from birth, yet side effects don't appear until the cerebrum, liver, or another organ begins to aggregate copper. The disease has distinct signs and symptoms that vary depending on the affected areas of the body. They could incorporate weariness, hunger misfortune, or stomach throb. Jaundice is characterized by a golden-brown discoloration of the eyes as well as a yellowing of the skin and whites of the eyes (Kayser-Fleischer rings). Wilson's disease is inherited as an autosomal recessive trait, which means that each parent must give you one copy of the defective gene. If you only have one defective gene, you won't get sick, but you will be a carrier and can pass the gene on to your children.

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Conclusion

Liver dysfunction can begin quickly (acute liver failure) or progress slowly over time. Liver transplantation is one treatment option. Neurological symptoms like tremors, involuntary muscular movements, clumsy gait, and speech impairments often improve with treatment for Wilson's disease. Wilson's disease can harm the kidneys, resulting in issues such as kidney stones and an abnormally high level of amino acids secreted in the urine, even if some people receive treatment for their condition. Psychological wellness issues such side effects incorporate, yet are not restricted to, character changes, sadness, peevishness, bipolar ailment, or psychosis. Anemia and jaundice are both symptoms of blood disorders like hemolysis, which can also lead to the destruction of red blood cells.

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