

Short Notes on Hepatolenticular Degeneration

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Introduction

Hepatolenticular degeneration accumulates in the liver, brain, and other important organs in Wilson's disease, a rare genetic condition. Wilson's illness can affect both children and adults, however it is most frequently diagnosed in patients between the ages of 5 and 35. The development of strong bones, healthy neurons, collagen, and the skin colour melanin all depend on copper. Normally, copper is absorbed from meals, and excess copper is eliminated through a substance made in your liver (bile). However, copper is not effectively removed in those with Wilson's disease and instead builds up, possibly to a degree that is life-threatening. If identified early, Wilson's illness is curable, and many affected individuals lead normal lives [1].

Description

Wilson's illness is present from birth, but symptoms don't show up until the brain, liver, or another organ starts to accumulate copper. Depending on which body areas are afflicted, the disease manifests itself differently in terms of signs and symptoms. They might include fatigue, appetite loss, or stomach ache. Jaundice, a yellowing of the skin and whites of the eyes, and a golden-brown discoloration of the eyes (Kayser-Fleischer rings). Wilson's illness is inherited as an autosomal recessive trait, which means that in order to get the condition, you must get one copy of the faulty gene from each parent. You won't get sick if you only have one faulty gene, but you will be a carrier and can spread the gene to your offspring [2].

Numerous neurological issues may exist in children with Hunter syndrome and continue to worsen. Having too much fluid in your child's brain can result in a number of neurological issues. These fluids' pressure can lead to severe headaches, blurred vision, and changes in your child's mood. In addition, your child can get a problem called thickening and scarring of the membranes that surround the spinal cord. The upper spinal cord is compressed and put under pressure as a result. Your youngster may as a result grow weaker and less active over time, develop leg weariness, and experience leg fatigue [3,4].

If either of your parents or siblings has Wilson's disease, you are more susceptible to the disorder. To find out if you have Wilson's disease, ask your doctor if you ought to undergo genetic testing. Early diagnosis of the illness increases the likelihood of effective treatment. If untreated, Wilson's disease can be fatal. One of the most dangerous side effects is liver scarring (cirrhosis). As liver cells try to heal the damage brought on by too much copper, scar tissue accumulates, making the liver's function more challenging [5].

Conclusion

Dysfunction of the liver this may occur suddenly (acute liver failure), or it may develop gradually. One possible course of treatment is a liver transplant. With Wilson's disease treatment, neurological problems like tremors, involuntary muscular movements, awkward gait, and speech difficulties often get better. However, some patients still experience neurological issues even after receiving treatment. kidney problems Wilson's disease can damage the kidneys, which can lead to issues including kidney stones and an unusual amount of amino acids being discharged in the urine. Psychiatric problems Examples of such symptoms include personality changes, depression, irritability, bipolar disorder, or psychosis. Blood-related issues can cause hemolysis, which results in anaemia and jaundice by destroying red blood cells.

Acknowledgement

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Conflict of Interest

None.

References

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