

UNDERSTANDING WILSON'S DISEASE IN CHILDREN

What is Wilson's Disease?

Wilson's Disease is a rare inherited disease in which excess copper builds up in the body, damaging the liver, brain, and other organs. It can be challenging to diagnose, especially in children, as the symptoms can be vague or mimic other illnesses. Early diagnosis and treatment are essential to managing the disease and preventing severe damage.



Understanding Wilson's Disease What it is and why it matters

Wilson's Disease occurs when the body is unable to eliminate excess copper, which then accumulates in organs like the liver, brain, kidneys, and corneas. In children, the condition is often diagnosed between ages 5 and 15, although it can appear at any age. If left untreated, Wilson's Disease can lead to liver failure, neurological damage, and psychiatric issues.

Symptoms to watch out for



The symptoms of Wilson's disease

in children can vary depending on which organs are affected.

Liver-related symptoms

Fatigue, jaundice (yellowing of the skin), abdominal pain, and swelling.

Neurological symptoms

Tremors, difficulty with coordination, changes in behaviour, and speech difficulties.

Psychiatric symptoms

Personality changes, irritability, depression, or mood swings.

Eye-related symptoms

A characteristic brown or greenish ring around the iris (Kayser-Fleischer rings), visible with a slit-lamp examination.



Wilson's Disease is caused by a amutation. For a child to develop the condition, both parents must carry and pass on the altered gene. If both parents are carriers, there is a 25% chance their child will inherit Wilson's Disease, and a 75% chance they will not. If the child does not inherit the disease, it could be because one parent passed on the mutated gene (50% chance), or because neither parent did (25% chance).

This genetic change impacts a protein that helps the liver eliminate excess copper by incorporating it into bile, aiding digestion and waste removal. In children with Wilson's Disease, copper does not enter the bile properly and accumulates in the liver, eventually spreading to other organs.

Read more https://www.cincinnatichildrens.org/health/w/wilsons-disease

Early diagnosis - Key to better outcomes

Early detection is critical for preventing long-term damage. A comprehensive approach to diagnosing Wilson's Disease in children typically involves:



Blood test

These measure ceruloplasmin

(a protein that helps carry copper) and copper levels in the blood.



Urine test

24-hour urine copper excretion test helps determine the body's copper elimination efficiency.



Liver biopsy

In some cases, to assess the degree of liver damage.



Genetic testing

A 24-hour urine copper excretion test helps determine the body's copper elimination efficiency.

Treatment options for children with Wilson's Disease



Once diagnosed, Wilson's Disease can be effectively managed with treatment focusing on reducing copper buildup and preventing further damage. Key treatment options include:

Chelating agents

Medications like penicillamine or trientine help remove excess copper from the body.

Zinc therapy

Blocking the absorption of copper from the digestive tract.

Low-copper diet

Limiting copper intake through a low-copper diet can help manage the condition.

Liver transplant

In severe cases of liver failure, a transplant may be necessary.

Living with Wilson's Disease: Family and lifestyle considerations

A diagnosis of Wilson's Disease can bring significant challenges for both the child and their family. Children may require adjustments in their daily routines, particularly in managing their medications and maintaining a copper-restricted diet. Here are a few important tips for

Education

Understanding the condition and its treatment options is essential for helping children manage their health.

Dietary changes

Copper-rich foods, such as shellfish, nuts, chocolate, and certain grains, should be avoided or restricted. A nutritionist can guide families in creating a balanced, copper-free diet.

Mental health support

Emotional and psychological support can be important for children dealing with the neurological and psychiatric symptoms of Wilson's Disease.







Looking ahead Research and advancements in treatment

Ongoing research into Wilson's Disease is focused on improving early diagnostic methods and developing new treatments. Recent advances include the investigation of gene therapies, which could potentially offer a cure for the genetic aspects of the disease in the future. Clinical trials are underway to explore these cutting-edge treatments, and results in the coming years may dramatically change the course of Wilson's Disease management.

References

Pediatric Wilson's disease

https://childliverdisease.org/liver-information/childhood-liver-conditions/wilsons-disease/

https://www.mayoclinic.org/diseases-conditions/wilsons-disease/symptoms-causes/syc-20353251

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