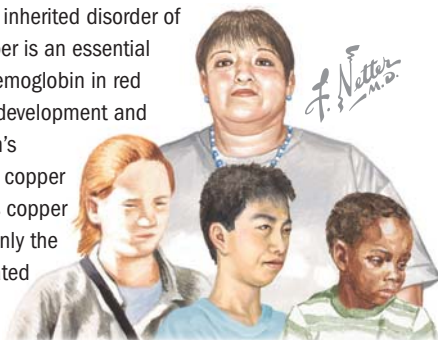


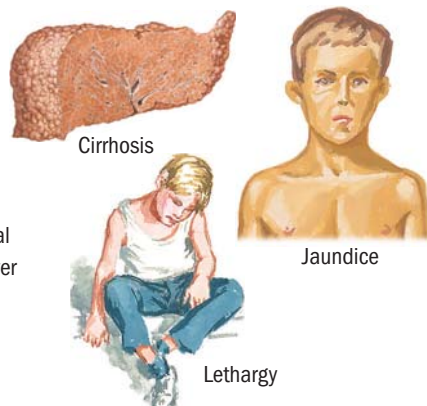
MANAGING YOUR WILSON'S DISEASE

Wilson's disease is a rare inherited disorder of copper metabolism. Copper is an essential metal for production of hemoglobin in red blood cells and for bone development and connective tissues. Wilson's disease causes too much copper to collect in the body. This copper gets into vital organs, mainly the liver and brain. The untreated disease is fatal.

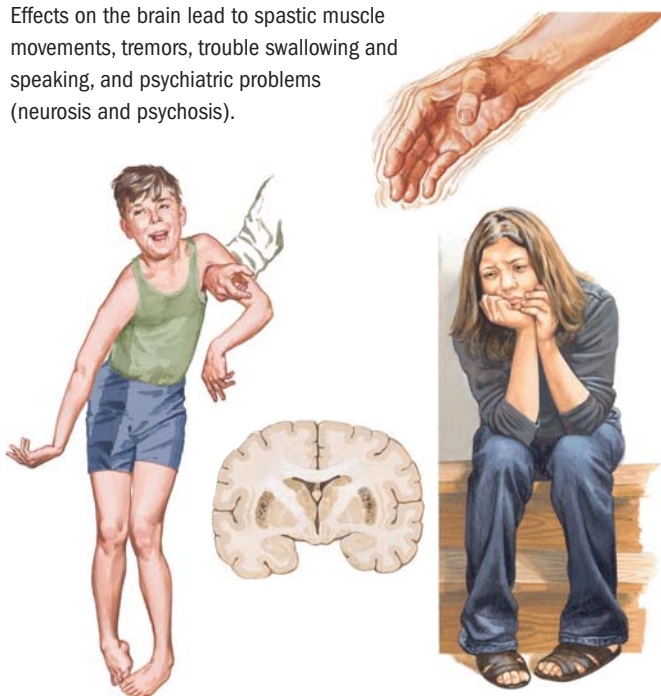


A mutation of a gene that passes from parents to children causes the disease. Both parents must have the mutation for the child to have the disease.

Symptoms usually start after a child is 3 years old, usually with signs of liver disease such as jaundice, fluid in the stomach, mental confusion, and lethargy. Liver cirrhosis may also develop.



Effects on the brain lead to spastic muscle movements, tremors, trouble swallowing and speaking, and psychiatric problems (neurosis and psychosis).



What Is Wilson's Disease?

Wilson's disease is a rare inherited disorder of copper metabolism. Copper is a metal found in small amounts in the blood. Copper plays an essential role in production of hemoglobin. Hemoglobin is a large protein molecule in red blood cells that attaches to oxygen and takes it to body tissues. Copper is also important for bone development and connective tissues.

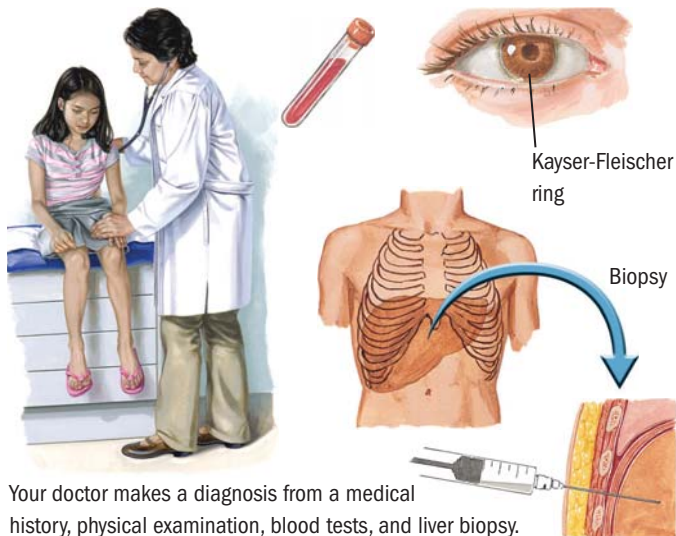
Wilson's disease causes too much copper to collect in the body due to insufficient or defective copper excretion in the liver. The excess copper accumulates in vital organs, mainly the liver and brain. Untreated Wilson's disease is usually fatal. The disease affects 1 in 30,000 people.

What Causes Wilson's Disease?

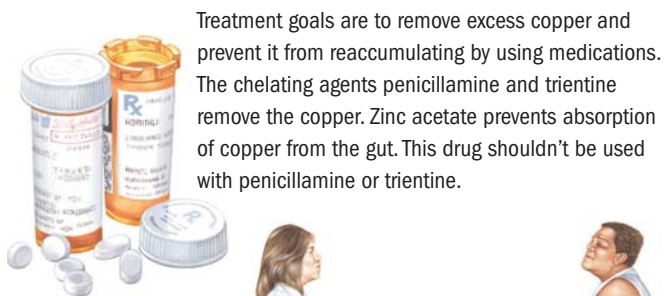
A mutation of a gene that passes from parents to children causes the disease. The disease is autosomal recessive, which means that both parents must have the mutation and give it to a child for the child to have the disease. It's not contagious.

What Are the Symptoms of Wilson's Disease?

Symptoms don't usually start until after a child is 3 years old and may not start until the teenage years or later. Usually, the liver is affected first. Liver inflammation (hepatitis) develops and can be mistaken for hepatitis caused by a virus. Other possible symptoms are jaundice (yellow skin), fluid in the belly (ascites), mental confusion, and lethargy. Liver cirrhosis may also develop. Effects on the brain lead to spastic muscle movements, tremors, and trouble swallowing and speaking.



Your doctor makes a diagnosis from a medical history, physical examination, blood tests, and liver biopsy. High blood ceruloplasmin and liver copper levels confirm the diagnosis. A key effect in the eye is a brownish Kayser-Fleischer ring.



Treatment goals are to remove excess copper and prevent it from reaccumulating by using medications. The chelating agents penicillamine and trientine remove the copper. Zinc acetate prevents absorption of copper from the gut. This drug shouldn't be used with penicillamine or trientine.

Keep follow-up doctor appointments. Once treatment begins, your doctor needs to watch your condition and test your blood (blood counts, liver function tests, and copper levels).



Call your doctor if you have side effects from your medicine. Don't stop taking your medicine. Treatment must continue for life. If you must stop taking it, your doctor should supervise this and can change the drug to another one.

How Is Wilson's Disease Diagnosed?

The doctor makes a diagnosis from a medical history, physical examination, blood tests, and liver biopsy. High blood ceruloplasmin and liver copper levels confirm the diagnosis. Ceruloplasmin is an oxygen-carrying protein in the blood. A key effect in the eye is a brownish color where the cornea and sclera (white part of the eye) meet, called a Kayser-Fleischer ring. Psychiatric problems may occur, including neurosis and psychosis.

How Is Wilson's Disease Treated?

The doctor may suspect the disease in anyone younger than 40 with abnormal liver blood tests, abnormal behavior, and unexplained liver cirrhosis and nervous system symptoms. Treatment goals are to remove excess copper and prevent it from reaccumulating by using medications. Penicillamine is a chelating agent, which means a drug that binds to copper, so it can remove it. It's usually given for life. Drug side effects may include rash, fever, swollen lymph glands, kidney problems, and lower red and white blood cell counts. If penicillamine cannot be tolerated, another chelating drug called trientine can be used. Zinc acetate also helps, by preventing absorption of copper from the gut. This drug shouldn't be used with penicillamine or trientine.

DOs and DON'Ts in Managing Wilson's Disease:

- ✓ **DO** keep follow-up doctor appointments. Once treatment begins, your doctor needs to monitor your condition and blood tests (blood counts, liver function tests, and copper levels).
- ✓ **DO** call your doctor if you have side effects from your medicine.
- ⊘ **DON'T** stop taking the medicine. Treatment must continue for life. If you must stop taking it, your doctor should supervise this and can change the drug to another medicine.

FROM THE DESK OF

NOTES

FOR MORE INFORMATION

Contact the following source:

- Wilson's Disease Association International
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Website: <http://www.wilsonsdisease.org>