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Commentary

Brief Description of Wilson's Disease

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DESCRIPTION

Wilson disease is a rare hereditary disease marked by the accumulation of copper in numerous human tissues, mainly the liver, brain, and corneas of the eyes. If left untreated, the disease progresses and can lead to liver (hepatic) disease, central nervous system malfunction, and death. Wilson disease signs and symptoms normally develop between the ages of 6 and 45, but they are most common during adolescence. This syndrome is defined by a mix of liver disease, neurological, and psychological issues. Copper is necessary for the development of healthy nerves, bones, collagen, and the pigment melanin in the skin. Copper is normally taken from meals and expelled by a chemical made by your liver (bile). Copper, on the other hand, is not adequately removed in patients with Wilson's disease.

Symptoms

Wilson disease is a rare chromosome abnormality that begins with liver malfunction at the age of six years old and progresses to clinical manifestations in the teenage years or early twenties.

Hepatic: Copper build-up in the liver can cause the following symptoms:

- Asymptomatic hepatomegaly
- Isolated splenomegaly
- Fatty liver
- Acute hepatitis
- Autoimmune hepatitis
- Cirrhosis
- Acute liver failure

Neurological: Copper build-up in the brain can result in symptoms such as:

- Movement disorders (tremor, involuntary movements)
- Drooling
- Dysphagia
- Pseudobulbar palsy
- Migraine headaches
- Insomnia

Eye symptoms: Kayser-Fleischer rings, which are greenish, gold, or brownish rings around the edge of the corneas, are common in persons with Wilson's disease. Kayser-Fleischer rings are caused by a deposit of copper in the eyes. These rings can be seen by a doctor during a slit-lamp examination.

Other symptoms: The build-up of copper in other organs can cause-

- Bluish discoloration in the nails
- Kidney stones
- Premature osteoporosis, or lack of bone density
- Arthritis
- Menstrual irregularities
- Low blood pressure

Causes

Wilson's disease is caused by mutations in the ATP7B gene. The body is unable to remove excess copper due to certain gene abnormalities. The liver normally excretes extra copper into the bile. The copper, along with other poisons and waste items, is carried by the bile. The digestive tract is where bile leaves the body. Wilson's disease causes the liver to discharge less copper into bile, resulting in excess copper remaining in the body. Wilson's disease is caused by ATP7B mutations that are passed down from one generation to the next. Wilson's disease is autosomal recessive, which means that a person must inherit two ATP7B genes with mutations (one from each parent). Wilson's disease does not affect people who have one ATP7B gene without a mutation and one ATP7B gene with a mutation, although they are carriers of the disease.

Treatment

Wilson disease is treated with a lifelong regimen aiming at reducing copper levels to safe levels. Treatment is divided into three parts: first, treatment of symptomatic patients, second, maintenance therapy when copper levels in afflicted tissues have been lowered, and third, maintenance therapy may be employed from the start in asymptomatic patients.

Wilson disease is treated with three different types of medicines. First, penicillamine (cuprimine) and trientine dihydrochloride

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(syprine), which remove copper from the body through urine excretion; second, zinc salts, which prevent the gut from absorbing copper from the diet; and third, tetrathiomolybdate, which both prevents absorbing copper and binds toxic copper in the blood, rendering it nontoxic.