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Commentary

Wilson's Disease- Recent Advancements

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DESCRIPTION

Wilson's disease is a hereditary disease in which copper accumulates excessively in the body, especially in the liver, brain, and eyes. Signs and symptoms of Wilson's disease usually first appear between the ages of 6 and 45, but most commonly begin in the teens. Characteristics of this condition include a combination of liver disease and neurological and psychiatric problems.

Liver disease is usually the first feature of Wilson's disease in affected children and young adults. People diagnosed at an older age may have very mild liver disease, but usually there are no signs of liver damage. Signs and symptoms of liver disease include yellowing of the skin and white of the eyes (jaundice), malaise, loss of appetite, and abdominal distension.

Nervous system or psychiatric problems are often the first signs of an individual diagnosed in adulthood and are common in young adults with Wilson's disease. Signs and symptoms of these problems include clumsiness, tremors, difficulty walking, speech disorders, thought disorders, depression, anxiety, and mood swings.

CAUSES OF WILSON'S DISEASE

Wilson's disease is caused by a mutation in the ATP7B gene. This gene provides instructions for making a protein called copper transport ATPase2, which is responsible for transporting copper from the liver to other parts of the body. Copper is required for many cellular functions, but it is toxic when present in excess. The copper transport protein ATPase2 is especially important for eliminating excess copper from the body. Mutations in the ATP7B gene prevent transport proteins from functioning properly. Due to the lack of functional protein, excess copper is not removed from the body. As a result, copper accumulates at toxic levels and can damage tissues and organs, especially the liver and brain.

TREATMENT

Controlled trials were not possible at the time treatment became available.

WD has historically evolved from intramuscular administration of BAL to easier-to-administer oral penicillamine. Although there are studies showing the dose response of penicillamine and the resulting cupulresis, initial clinical use was limited by the availability of the drug itself. An empirical dose was selected because no formal dose-response study of efficacy over time has been performed.

Interestingly, when these treatments were first available, treatment was initially reserved for symptomatic patients because there were no good diagnostic tests to identify presymptomatic conditions. At the same time as the progress of diagnostic tests for WD in the new era.

It began with the recognition that treating asymptomatic patients can prevent significant morbidity and mortality 149. The development of alternatives to penicillamine was facilitated by intolerance to this drug in some patients.

Designed and introduced specifically for patients who develop the side effects of penicillamine. Zinc was developed individually, similar to the TM used by veterinarians for copper poisoning in animals. Today, lifelong pharmacological therapy continues to be central to the treatment of WM. Liver transplants that correct the underlying liver defect in WD are severe or modest resistant case. In general, the therapeutic approach is whether there is evidence of a clinically detectable disease or laboratory or histologically aggressive inflammatory injury, whether neurological or hepatic, or before the onset of clinical symptoms. It depends on whether the patient is identified. No studies have systematically examined this approach, but we believe that this distinction will help guide treatment choices and dosages of the drugs used.

AVAILABLE THERAPIES

D- penicillamine

Penicillamine was introduced in 1956 as the first oral preparation to treat WD. Although identified as a degradation product of penicillin, it is actually a methyl group-substituted sulfhydryl-containing amino acid cysteine. Like the dimer captopropanol (or BAL), it has a free sulfhydryl group that acts as a chelating agent for copper. Penicillin is now so synthesized and contamination with penicillin is not a problem. Similarly, racemic mixtures that tend to interfere with the action of pyridoxine are no longer used. Nevertheless, additional pyridoxine is provided at Oral administration of 25-50 mg daily.

Trientine

Triethylenetetramine dihydrochloride or 2, 2, 2-tetramine, also known as the formal abbreviation triene belongs to the family of chelating agents. A polyamine-like structure that is chemically

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different from penicillamine. It lacks a sulfhydryl group and copper is chelated by forming a stable complex with the four components.

Nitrogen in a planar ring. Trientine was introduced in 1969 as an alternative to enisylamine. There are few data on the pharmacokinetics of trientine. It is poorly absorbed from the gastrointestinal tract, and what is absorbed is metabolized and inactivated. Usually about 1% of administered trientine and about 8% of the in vivo converted trientine metabolite acetyltrien eventually appear in the urine. Acetyltrien is a less effective chelating agent than trientine. The amount of copper, zinc, and iron in the urine increases in parallel with the amount of trientin excreted in the urine.