Wilson's Disease

Wilson's disease is an inherited disorder. With it, large amounts of copper build up in the body. The accumulation of copper begins at birth. But symptoms of the disorder appear later in life. This is between the ages of 6 and 40. The primary effect for about 40 percent of patients with Wilson's is liver disease. In other patients the first symptoms are either neurological or psychiatric or both.

SYMPTOMS

Neurological and psychiatric symptoms may include:

- Tremor.
- Rigidity.
- Drooling.
- Difficulty with speech.
- Abrupt personality change.
- Grossly inappropriate behavior.
- Unexplained deterioration of school work.
- Neurosis.
- Psychosis.

TREATMENT

Treatment of Wilson's disease generally consists of anti-copper agents. These are to remove excess copper from the body and to prevent it from reaccumulating. Most cases are treated with the drugs zinc acetate, trientine, or penicillamine. Penicillamine and trientine increase urinary excretion of copper. But both drugs can cause serious side effects. Zinc acetate blocks the absorption of copper, increases copper excretion in the stool and causes no serious side affects. It is often considered the treatment of choice. Tetrathiomolybdate is an experimental drug. It also shows promise in treating Wilson's disease. In rare cases in which there is severe liver disease, a liver transplant may be needed.

WHAT WILL HAPPEN

Without proper treatment, Wilson's disease is generally fatal, usually by the age of 30. If treatment is begun early enough, symptomatic recovery is usually complete. A life of normal length and quality can be expected.

RESEARCH BEING DONE

NINDS supports research to find ways to treat and prevent inherited disorders such as Wilson's disease. The ultimate goal of this research is to discover new treatments for the many genetic disorders that strike the brain and nervous system. This includes Wilson's disease.

Research is continually being done to learn more about this problem.

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