



Wilson's Disease: Nutrition Support

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Abstract:

Wilson's disease (WD) or hepato-lenticular degeneration is a rare autosomal-recessive disorder. A prevalence rate of 30 cases per million and a birth incidence rate of one per 30,000 to 40,000 are often quoted. In 40 to 50% of individuals with WD, hepatic dysfunction is the initial clinical manifestation. With the exception of liver transplantation, treatment of WD is only palliative and intended to restore and maintain copper balance. It does not eliminate the underlying defect responsible for WD. Thus, a lifelong commitment to treatment is required.

Limitation of dietary copper intake is generally ineffective, and pharmacological management is necessary. Administered either as acetate, sulfate, or gluconate, zinc reduces intestinal absorption of dietary copper via induction of metallothionein formation in intestinal enterocytes. Antioxidants, mainly vitamin E, may have a role as adjunctive treatment. Serum and hepatic vitamin E levels have been found to be low in WD. Symptomatic improvement when vitamin E was added to the treatment regimen has been occasionally reported but no rigorous studies have been conducted. No correlation of antioxidant deficiency with clinical symptoms was reported in one study.

A vegetarian diet may be useful as adjunctive therapy; copper is less available. Adherence to a low copper diet is most important during the initial phase of treatment. The recommendation is to avoid the foods highest in copper content: organ meats, shellfish, chocolate, nuts, and mushrooms. Once copper levels have stabilized at normal levels, these foods are allowed occasionally. Copper content of the drinking water you consume should also be tested. If the water is over 0.1 ppm (parts per million) (which is 0.1 mg/L), consider an alternative water source or invest in a good filtering system that removes copper.

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