

## **Wilson`s disease- liver presentation**

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There are two major clinical presentations of Wilson disease- liver and neurological (rare in children). Liver symptoms may vary in severity- from asymptomatic forms, to cirrhosis and acute liver failure. There are adult guidelines and recently elaborated pediatric position paper on diagnosis and therapy of Wilson disease. The diagnostic criteria for adult, pediatric, neurological and liver presentations are similar and use the same scoring system, which is a combination of different clinical symptoms and laboratory tests (ceruloplasmin concentration, 24h urinary copper excretion, copper content in the liver and molecular analysis). WD should be considered in the differential diagnosis of children already above the age of 1 year presenting with any sign of liver disease ranging from asymptotically increased serum transaminases to cirrhosis with hepatosplenomegaly and ascites or acute liver failure. Biochemical tests may be less sensitive in very young children. Pharmacological therapy is mainly based on chelating agents like penicillamine and trientine and zinc preparations. It was proven to be very effective but the major problem on long term is poor compliance. Chelating agents should be preferably used in patients with signs of significant liver disease, such as cirrhosis or abnormal INR. Zinc salts could be used in pre-symptomatic children identified through family screening, or as maintenance therapy after de-coppering with chelators as long as serum transaminase levels remain normal. Liver transplantation is indicated only in selected cases, mainly with acute liver failure and the medical decision can be based on a special prognostic scoring system.