- Clinical presentation
- Diagnosis
- Treatment
- Pregnancy in WD
- Laboratory services

Treatment

Envoyer | Imprimer

Download booklet

The treatment of Wilson disease was impractical until J.M. Walshe, in 1956, described the favourable effects of this copper-chelating therapy in Wilson disease. In almost all asymptomatic patients and many symptomatic patients penicillamine therapy can prevent or repair the devastating effects of the copper overload. However, side effects and toxic reactions are frequently observed and in 10% of the patients therapy has to be stopped. Also, a significant proportion of the patients with neurological disease experience worsening of their neurological symptoms after starting penicillamine, and some of them will never return to their pre-penicillamine base-line again. Therapeutic alternatives have therefore been sought. The first to appear was trientine, another copper-chelating agent, which probably will give the same results as penicillamine, but with less side effects. However experience with this medication is limited so far.

In 1961 Schouwink introduced zinc as an alternative to penicillamine. He described that during zinc treatment the amount of copper excreted with the stools increased, making overall copper balance negative. This effect is now known to be based on zinc-induced metallothionein synthesis in the small intestinal epithelium. The metallothionein binds copper and the complex is sloughed off into the faeces together with the intestinal cell. Especially during the last 10 years zinc has gained increasing acceptance as it has been shown sufficiently that oral zinc is a suitable alternative to penicillamine as long-term maintenance therapy both for adults and children with Wilson disease. Zinc seems to be safe in presymptomatic patients too. It is currently investigated whether it is also a good alternative for patients with neurological problems.

A special treatment problem is the patient with severe liver disease and impending liver failure. In these patients penicillamine will fail in up to 50%. Nevertheless this medication should be started as soon as the diagnosis has been made. However when, despite therapy, liver synthesis function deteriorates further, liver transplantation should be performed. Liver transplantation is also done in patients with end-stage liver disease due to advanced cirrhosis.

Present therapy will improve symptoms and prevent a fatal outcome in most patients. However, for patients with neurological disease the symptoms may not all be reversible. For patients presenting with mild liver problems, residual morbidity seems to be confined to the presence of cirrhosis in some, while in pre-symptomatic patients, therapy prevents the onset of symptoms altogether. For all patients compliance with the life-long continuous treatment is of paramount importance. Stopping medication will lead to severe organ damage, or even death, within a time-period that can be as short as a couple of months.