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Summary

EASL-ERN Clinical Practice Guidelines on Wilson’s disease

Standards and Guidelines

Hepatobiliary

November 8, 2024

Wilson's disease is an autosomal recessive disorder of copper metabolism affecting the liver, brain and other organs, requiring diagnosis through clinical, biochemical and molecular criteria and management with chelators, zinc salts, and in severe cases liver transplantation.

KEY POINTS

- Diagnosis is based on clinical features, biochemical tests including plasma ceruloplasmin concentration, 24-hour urinary copper excretion, liver copper content, molecular analysis, Leipzig score, and relative exchangeable copper determination.
- Pharmacological therapy comprises chelating agents such as penicillamine and trientine, and zinc salts, with only chelators recommended for significant liver disease.
- Monitoring uses clinical symptoms, liver tests, and copper metabolism markers including urinary copper excretion and exchangeable copper to detect poor compliance and over- or under-treatment.
- Acute liver failure is challenging to diagnose and pharmacological therapy may not be sufficient to save life.
- Liver transplantation has a well-defined role in Wilsonian acute hepatic failure and may also be considered in neurological disease.

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