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Diagnosis and Treatment of Wilson's Disease With an Update on Anticopper Treatment for Other Diseases

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Abstract:
A major key to the diagnosis of Wilson's disease is recognition of the possibility that this diagnosis is a possibility in a given patient. The rare patient with neurologic Wilson's tends to be hidden among much more common patients with essential tremor, Parkinson's disease, and other neurologic conditions. The rare patient with hepatic Wilson's tends to be hidden among much more common patients with viral hepatitis, chronic active hepatitis, alcoholic cirrhosis, and other liver disease. And, of course, patients with initial psychiatric disease only are rarely recognized. Kayser-Fleischer ring examination and 24-hour urine copper assays are excellent screening tools. Liver biopsy with quantitative measurement of copper is still the gold standard for diagnosis. Modern anticopper treatment involves use of different drugs for different presentations and stages of the disease. We recommend tetrathiomolybdate plus zinc for initial therapy of neurologic Wilson's, trientine plus zinc for initial therapy of hepatic Wilson's, and zinc alone for maintenance therapy, the presymptomatic patient, and the pregnant patient.
The final part of this review discusses new findings in which copper-lowering therapy with tetrathiomolybdate is remarkably effective in treating or preventing inflammatory and fibrotic damage in animal models of lung and liver injury. The postulated mechanism is inhibition of overexpressed and dysregulated profibrotic and proinflammatory cytokines.

Key words: Wilson's disease, Tetrathiomolybdate, Anticopper treatment, Fibrotic diseases, Inflammatory diseases

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