

TACKLING THE NEUROLOGICAL WILSON DISEASE WITH OTHER SPECTRUM OF PRESENTATIONS: A CASE SERIES

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Wilson's disease (WD) is an inherited autosomal recessive, potentially treatable disorder of copper metabolism that produces neurologic, psychiatric, and liver manifestations, alone or in combination. It is caused by mutations in the ATP7B gene encoding a P-type ATPase. WD has a worldwide prevalence of ~1 in 30,000, with a mutation carrier frequency of 1 in 90. About half of WD patients (especially younger) present with liver abnormalities. The remainder present with neurologic disease (with or without underlying liver abnormalities), and a small proportion have hematologic or psychiatric problems at disease onset. Pathological copper accumulation causes a range of symptoms, most commonly hepatic and a wide spectrum of neurological symptoms including tremor, dystonia, chorea, parkinsonism, dysphagia, dysarthria, gait and posture disturbances. To reduce copper overload, medications are used that improve liver function and neurological symptoms. However, in some WD patients, treatment introduction leads to neurological deterioration, and in others, neurological symptoms persist with no improvement or improvement only after several years of treatment. Based on case and series reports, current recommendations and expert opinion, WD treatment is focused mainly on drugs leading to negative copper body metabolism (chelators or zinc salts) and copper-restricted diet. Treatment of WD neurological symptoms should follow general recommendations of symptomatic treatment. Patients should be always considered individually, especially in the case of severe, disabling neurological symptoms. Here we discuss 03 cases of WD with neurological and other manifestations treated accordingly, 02 patients responded to treatment while we lost another unfortunately.

Key words: Wilson's disease; dysphagia, dysarthria; dystonia, chorea, parkinsonism; neurological symptoms; symptomatic treatment.

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