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To study the efficacy of Penicillamine followed by Zinc in treating symptomatic predominantly hepatic WD.

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Background: Experience with Zinc in treating symptomatic hepatic Wilson's disease (WD) is limited.

Methods: We studied symptomatic WD patients in whom Penicillamine was changed to Zinc sulfate either due to financial constraints (in 25 patients) or due to adverse effects of Penicillamine (in 2). Disease severity scores (Child's, MELD, Nazer's and New Wilson Index score) and 24-hour Urinary copper were calculated at 3 time points – baseline at diagnosis, at transition from Penicillamine to Zinc and at end of follow-up.

Results: 27 patients were studied, 18 had hepatic WD, 8 had neurological and hepatic WD and 1 had hepatic and neuropsychiatric manifestations. Child's grade was A in 6 patients, B in 3 and C in 15. Duration of Initial Penicillamine chelation therapy was 132 weeks (range: 2-320), and of subsequent Zinc therapy was 366 weeks (range 35-728). Three patients died at 284, 112 and 437 weeks. No patient underwent liver transplantation. There was significant improvement in liver function tests and disease severity scores (Nazer's score, New Wilson index score, Child's and MELD score) at transition from Penicillamine to Zinc compared to baseline which was maintained till end of study period. Nine patients had received Penicillamine for less than 1 year (35 weeks; range: 2-52) and 15 patients had decompensated cirrhosis with Child Grade-C at presentation who improved until end of follow-up.

Conclusions: Penicillamine followed by Zinc maybe a safe and effective treatment in resource constrained setting for symptomatic (predominantly) hepatic WD patients with all grades of baseline disease severity. Our data also shows that patients with decompensated cirrhosis due to Wilson's disease can be managed with medical treatment avoiding liver transplantation.

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