Valproate induced encephalopathy treated with carnitine in an adult

Hepatotoxicity due to valproate often necessitates discontinuation of the drug. We report a patient with unstable epilepsy in whom valproate was an irreplaceable component of anticonvulsant treatment. Hepatic encephalopathy was reversed and maintained with continued carnitine supplementation. The icteric episode was associated with May-Hegglin anomaly. This illustrates that carnitine supplementation should be considered in adults with epilepsy at risk for hyperammonaemia.

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Increased serum neopterin concentrations in a patient with Creutzfeldt-Jakob disease

Spongiform encephalopathies or prion diseases affect both humans and animals. The human transmissible spongiform encephalopathies include Kuru, Creutzfeldt-Jakob disease, and the Gerstmann-Straussler-Scheinker syndrome (GSS), and are caused by specific prion proteins; these diseases are associated with the accumulation of β-pleated amyloid protein in the brain.5

Besides genetic abnormalities, epidemiological studies disclosed that a transmissible agent is involved in the spread of Creutzfeldt-Jakob disease, and small viral-like particles were considered at increased risk due to suspected mitochon- dridial disorder, malignancy, mental retardation, high dose valproate treatment, or a history of hepatotoxicity to the valproate.4 Treatment with carnitine has also been reported to reverse this hepatotoxicity in children despite continued valproate.1,4 Less is known about the relation between valproate induced hepatotoxicity and carnitine deficiency in adults. Reduced free carnitine concentrations have been reported in 76-5% of adults receiving anticonvulsant drug regimens including valproate compared with 21-5% of adults on schedules without valproate.4 Coma from valproate induced carnitine deficiency in adults is reported to respond rapidly to discontinuation of the drug.1

This case shows that certain adults receiving valproate are, like children, at risk for carnitine deficiency and hyperammonaemia with its clinical accompaniment. The incidence among encephalopathy in children on valproate is unknown. This patient further illustrates that identification and correction of a drug induced deficiency may allow continuation of treatment without disturbing control of an unstable seizure disorder. Much as folate is used in chronic phenytoin administration, supplemental carnitine should be considered in adult patients with epilepsy at risk for hyperammonaemia.

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