

TABLE I—Referral Patterns of Neonatal Cholestasis Syndrome.

	January, 1992 - July, 1995		May 1999 - September 2002	
	IHG*	EHBA**	IHG*	EHBA**
Age at onset (days)	17	13	12.31	15.72
Age at presentation (days)	84	132	76.54	122.28
Delay (days)	66	121	64.23	106.56

*Intrahepatic neonatal cholestasis group, ** Extrahepatic biliary atresia.
(All value are expressed as mean).

urgent referral of babies particularly who look well and have pale stools”.

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Wilson's Disease Presenting as Status Epilepticus

Wilson's disease (WD) is an inherited disorder of copper metabolism affecting 1:40,000-100,000 live births(1), characterized by neuropsychiatric and hepatic involvement. Status epilepticus (SE) occurring in WD is rarely reported(2). We report an adolescent with SE as the initial manifestation of WD.

A 16-year-old girl was brought with history of 20-25 episodes of generalized tonic-clonic seizures (GTCS) of one-day duration, without regaining consciousness in between. There was no associated fever, headache or vomiting. Decline in scholastic performance

and disinhibited behavior were noted over the past eight months. A psychiatrist evaluated her and started on lithium 300 mg/day one week ago. There was no history of jaundice. Family history was unremarkable. On examination, vital signs were normal. She was stuporous, optic fundi were normal and there were no meningeal signs. All limbs were rigid, deep tendon reflexes were exaggerated and plantars were extensor. Kayser-Fleischer rings were present in cornea. Other systemic examination was normal. Investigations showed a normal hemogram. Serum electrolytes, blood sugar and creatinine were normal. Liver enzymes revealed AST 124 U/L and ALT 145 U/L. Serum ceruloplasmin was 6 mg% (normal >30 mg%). Twenty-four hour urinary copper excretion was 280 µg/day (normal <100 µg/day). Magnetic resonance imaging showed

bilateral symmetrical T2W hyperintense lesions in lentiform nuclei, thalami, and bilateral frontal regions (left more than right). Electroencephalography showed diffuse slow waves. A diagnosis of WD was made and d-penicillamine 750 mg/day and pyridoxine 20 mg/day were started. Seizures were treated with phenytoin (800 mg loading and 200 mg/day) and midazolam infusion (5 µg/min). She became seizure-free within eight hours of admission. At a follow-up 14 months later, she was seizure-free, and rigidity and cognition had markedly improved.

Epileptic seizures occur in about 6% of cases of WD(3). They can occur at any stage, but often begin shortly after the start of chelating therapy and respond well to therapy(3). SE in WD is extremely uncommon. It has earlier been reported only once in WD (after starting penicillamine therapy)(2). Our patient was not on chelating agents. To the best of our knowledge, this is the first reported case of SE occurring in WD before starting treatment.

Several mechanisms may be responsible for increased seizure activity in WD. Firstly, copper deposition in various parts of brain including cerebral cortex can lead to seizures by inhibition of membrane ATPase(4). Chelating therapy contributes to copper

mobilization. Secondly, pathological lesions in WD include neuronal loss, gliosis, laminar necrosis, spongy degeneration and cavitation of cerebral cortex, which may lead to focal seizure activity(5). Thirdly, penicillamine causes pyridoxine deficiency, which may precipitate seizures.

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