Letters to the Editor

Neonatal Cholestasis in India

National consensus meeting on neonatal cholestasis syndrome (NCS) observed that this entity stood ignored in India(1). An average delay of 3 months in referral to medical and surgical centers was observed thus, the document emphasized early referral of jaundiced babies with passage of high colored urine. In order to achieve the goal of early referral, quick workup and cost effective strategy, the consensus document recommended a uniform approach of NCS management(1). It is mandatory to audit the impact of these guidelines.

Sanjay Gandhi Postgraduate Institute of Medical Sciences has a wide referral from most of Uttar Pradesh and adjoining states. Awareness related to NCS was targeted in this part of country by (*a*) taking several lectures at district or state level; and (*b*) circulation of conference brochures highlighting early referral of NCS cases with special reference to EHBA, besides the above meeting on consensus guidelines. To find out the impact on referral of NCS, we therefore evaluated data of NCS cases presenting to our hospital from May 1999 to August 2002 and compared this with previously published data (n = 60; study period: January 1992 to July 1995) from the same center(2). All babies with NCS (n = 70) were studied as per management algorithm(1). Their mean age of presentation was 88.3 (range 5 - 395) days and mean age of onset of symptoms 13.2 (range birth - 75) days; and thus an overall delay of 75 (range 4 - 394) days in referral. Age wise break up of presentation to our center was: 19% by 30 days, 32% between 31 - 60 days, 20% between 61-90 days, and 29% after 90 days. Subgroup analysis showed that age of presentation in EHBA and intrahepatic group (IHG) were 28% and 60% before 60 days of age, 22% and 196/0 between 61-90 days and 50% and 21% after 90 days respectively. Present data (May 1999 to August 2002) revealed early referral of IHG (59.6% cases) as compared to EHBA (27.8%) within first 2 months of age (P = 0.02) and this was due to presence of several risk factors. Among IHG presenting before 60 days of age, 55% NCS cases had identifiable risk factors, as compared to 14% presenting after 60 days of age (P <0.01). Factors identified were those of coagulopathy, sepsis, birth asphyxia, history of sib death, abortions, prematurity, low birth weight, eventful post-natal course, ascites, bronchopneumonia, urinary tract infection and history of jaundice in mother during pregnancy. We compared differences in patterns of referrals of NCS cases between the present and previously published data(2) (*Table I*). We observed an overall declining trend of delay in referral in EHBA (previously 121 days vs. 107 days now) suggesting an impact on early referral by 2 weeks (*Table I*). There was no impact on referral pattern of IHG.

A message forthcoming is an impact of just 2 weeks early referral of EHBA despite multi-directional awareness efforts. This does not in true sense fulfill the desired objective of optimum referral of EHBA by 4-6 weeks of age. Babies having EHBA look well, feed well, and develop normal social smile giving a false impression of well being to parents with the exception of being dressed up in yellow color. Simple additional message is "very

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