Letters to the Editor

Improving Clinical Course in Congenital Hypomyelinating Neuropathy

I read with interest the recent article by Chandra, *et al.*(1). They report an interesting case of an infant with congenital hypo-myeljnating neuropathy (CHN), who showed clinical improvement with a course of steroids. However, I would like to make certain observations.

CHN is known to have variable clinical presentations ranging from asymptomatic cases to rapid progression and death. Though the neurological deficits are progressive in a majority of infants, some cases are characterized by clinical improvement on follow up(2-4). Chandra, *et al*. attribute the clinical improvement observed in their case to course of oral steroids. However, there is no evidence that steroids are beneficial in the treatment of congenital neuropathies. The reference cited by Chandra, *et al.* to support their use of steroids actually is an evidence against using steroids; as the infant described therein had progressive worsening on corticosteriods and immunoglobulins and improved only after the discontinuation of corticosteriods(2). Moreover, long term follow up studies on two patients (neonatal period to nine years of age and infancy to five years of age respectively) have shown spontaneous recovery in them despite; unchanged findings on nerve conduction velocities(3). Therefore, the most likely explanation of recovery in the case described by Chandra, *et al*. is a spontaneous recovery rather than treatment with steroids, as they suggest. However, it would be interesting to follow up this child for longer periods preferably after discontinuation of cortico-steroids and conduct genetic studies to further characterize the neuropathy. The possible mechanism that could explain reversibility in a genetic disorder such as CHN could be an abnormal expression of a developmental gene, as in reversible cytochrome oxidase deficiency(4).

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