our newborns received therapeutic hypothermia. We have proposed levetiracetam as an effective and safer alternative to phenobarbitone as a first line drug in neonatal seizures, and not in neonates with HIE [1]. We agree about the need for long term studies to look for neurodevelopmental outcome of these neonates, and the same has been acknowledged already as a limitation of our study.

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Early-onset Fulminant Subacute Sclerosing Panencephalitis in a Toddler

A 22-month-old boy born to non-consanguineous parents with pre-morbid normal development, presented with loss of previously acquired developmental milestones and recurrent head drops for the past 3 months. He was completely unimmunized and had a history of exanthematous febrile illness resembling measles at the age of 11 months. On examination, he was in a minimally conscious state, with generalized dystonia, intermittent choreoathetosis and repetitive myoclonic jerks.

Electroencephalography showed generalized periodic epileptiform discharges, with bursts comprising of high amplitude spike and slow-wave complexes. MRI brain showed patchy periventricular white matter signal changes. CSF measles specific IgG levels were elevated (1:625), confirming the diagnosis of subacute sclerosing panencephalitis (SSPE). He was started on isoprinosine and antiepileptic drugs. At 6 week follow up, myoclonic jerks had subsided; however, he was in vegetative state and had persistent extrapyramidal features.

Neurological syndromes caused by measles virus include primary measles encephalitis, acute post-measles encephalitis, inclusion-body encephalitis and SSPE [1]. SSPE is caused by latent smoldering infection of the brain by wild-type measles virus which has variable presentation and is frequently misdiagnosed [2]. The earliest documented case of SSPE following a postnatally acquired measles infection was at 10 months of age [3]. A total of 15 cases of SSPE have been diagnosed before three years of age [1] of which seven cases occurred following postnatally acquired measles infection.

The clinical course of SSPE was atypical, did not follow the classic four stages of the Jabbour Classification [1] and had history of pre-existing developmental delay or seizures. As compared to older children, course of the disease was fulminant with rapid progression to a vegetative state and fatal outcome [4]. Genetically determined immune dysfunction in the first 2 years of life preventing a successful cell-mediated immune clearance of measles virus has been implicated in this short latency and fulminant course [5]. Other putative genetic factors include genetic polymorphisms of Toll-like receptor 3, programmed cell death-1, MxA, interleukin-4, and interferon-1 genes [5]. Clinicians need to be aware of these important clinical observations to suspect atypical presentation of SSPE in young children. Although neuronal ceroid lipofuscinosis and other lysosomal storage diseases remain the most plausible clinical differentials for progressive myoclonic epilepsy with onset less than two years of age, SSPE should be considered in an unimmunized toddler who presents with cognitive decline, extrapyramidal signs and symptoms, myoclonus and a rapidly progressive fulminant course particularly in developing countries.

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Allgrove Syndrome and a Novel Mutation of *AAAS* Gene in a Boy

A 2.5-year-old boy presented with progressive darkening of lips for the past four months, and absence of tears noticed since early infancy. There was no history of dysphagia or neurological problems. Born to non-consanguineous parents, his two elder sisters had died at age 5 and 6 years after similar manifestations. On examination, he was underweight (9.2 kg, –3.07SDS), stunted (82.4 cm, –2.68SDS) and had small head (45.5 cm, –2.42SDS). Oral mucosa was deeply pigmented. The systemic examination was unremarkable. Laboratory investigations showed normal serum sodium and potassium but low morning serum cortisol (89.2 nmol/L) and elevated adrenocorticotropin levels (1470 pg/mL). Reduced tear production was confirmed by Schirmer test. Barium swallow and endoscopic evaluation showed no achalasia. Clinical exome sequencing showed a previously undescribed homozygous frameshift deletion c.762delC (p.Ser255Valfs* 36) at Exon 8 of the *AAAS* gene, further confirmed by Sanger sequencing. The mutation is considered pathogenic by prediction tools such as SIFT, Mutation Taster and Phenolyzer. The child was initiated on lubricant eye drops and hydrocortisone and showed improved growth over one year (weight 10.8 kg, –2.12 SDS and height 89.4 cm, –2.54 SDS).

Allgrove syndrome (or Triple A syndrome) is an autosomal recessively inherited disorder caused by mutations in the *AAAS* gene that encodes for a protein ALADIN involved in the movement of molecules into and out of the nucleus, probably affecting DNA repair mechanisms leading to cell death [1]. The syndrome may present with any one of the four cardinal features that include achalasia, Addison disease, alacrima and progressive neurological dysfunction, and the symptoms may evolve over a period of time [1]. Alacrima is often the earliest and most consistent finding. Presence of one more symptom warrants molecular analysis of the *AAAS* gene [2]. Although a lower prevalence of neurological dysfunction has been noted in Indian patients, this feature is known to manifest later [1]. The poor head growth in our patient probably indicates the beginning of neurological dysfunction. These patients are also prone to dermatological abnormalities such as hyperkeratosis. The *AAAS* gene mutations are known to affect siblings and may explain sibling deaths with adrenal failure [1,3]. A high index of clinical suspicion is required due to the rarity and presentation as incomplete triad of symptoms [4,5]. Molecular analysis of the *AAAS* gene helps in confirming diagnosis and prognostication.

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