Status Dystonicus: A Rare Manifestation of Cerebral Palsy

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To the Editor: Status dystonicus (SD), also known as dystonic storm or dystonic crisis, is a rare life-threatening movement disorder emergency [1] characterized by severe, generalized and continuous hyperkinesias. The course and outcome of SD is highly variable, but early recognition facilitates aggressive intervention, which could potentially prevent progression and favor a better outcome [1].

We herein describe the clinical presentation and management of an episode of SD in a 1.5-y-old patient with dyskinetic cerebral palsy. The child was born through non-consanguineous marriage with no significant birth history, however requiring exchange transfusion on day 5 of life. The mother noticed looseness of all four limbs at 3 mo of age and sporadic abnormal movements with developmental delay at 9 mo of age for which the child was started on anti-epileptics and physiotherapy. The child presented to our hospital with relentless dystonic spasms, severe opisthotonus with retrocollis, excessive drooling and irritability for 1 day accompanied by an upper respiratory tract infection. The investigations revealed metabolic derangements including deranged kidney function tests, elevated creatine kinase and metabolic acidosis. A possibility of grade 4 SD was kept and the child was started on pacitane, clonazepam and oral clonidine. In view of poor control, syndopa, midazolam infusion and tetrabenzine were added sequentially. Midazolam infusion was tapered and stopped once dystonias improved. However, her sensorium continued to be depressed which showed a marked improvement on stopping tetrabenzine. The child was discharged on pacitane and clonazepam.

In the longest series published on SD, majority of patients (37.8%) were affected by secondary dystonia with cerebral

palsy being the most common cause [2]. There are no established guidelines or standardized therapeutic approaches for the treatment of SD [1]. Pharmacological intervention has been established as the first-line strategy in majority of the episodes (tetrabenzine and benzhexol being most effective) followed by a stepwise approach: sedation (benzodiazepines and propofol followed by barbiturate anesthesia), neurosurgery, and rarely muscle paralysis or intrathecal baclofen (ITB) [3–5]. Trial with syndopa should always be given in children with dystonia of unknown etiology due to the clinical variability of the dopa-responsive dystonias [1].

Compliance with Ethical Standards

Conflict of Interest None.

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