Nonconvulsive status epilepticus in patient with Angelman syndrome: Are corticosteroids of any benefit?

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Introduction and Aim

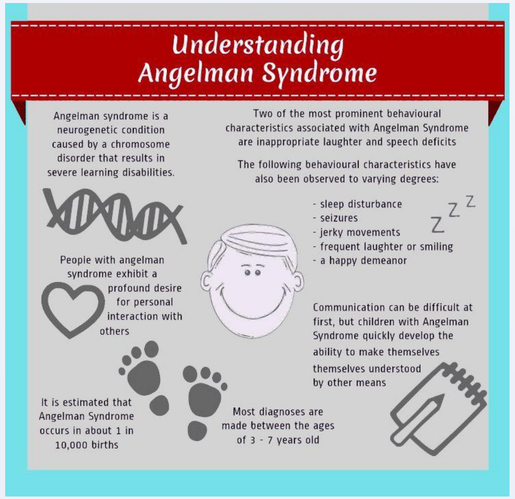
Angelman Syndrome (AS) is a severe neurodevelopmental disorder characterised by severe developmental delay, speech impairment, movement or balance disorders and specific behavioural characteristics with atypical frequent smiling and/or laughter.

Intractable epileptic seizures since early childhood with characteristic EEG abnormalities are present in majority (80-90%) of patients with AS. (1)

AS can present with various seizure types including generalised tonic-clonic, atonic, myoclonic, and atypical absences.

Status Epilepticus may be seen however nonconvulsive status epilepticus happens more often. (2)

Our aim is to assess whether use of corticosteroids is of any benefit in terminating the seizure cycle



Presentation and Method

4-year-old Adam (name altered to maintain confidentiality) who was diagnosed as Angelman Syndrome (Chromosome 15q deletion) and associated pharmaco-resistant epilepsy with developmental delay presented with 2 days history of diarrhoea and vomiting and increased seizure frequency.

Adam was already on multiple anti-seizure medications namely Levetiracetam, Lamotrigine and Sodium valproate and having been tried on Ketogenic diet in the past. Initially in view of his presentation he was started on broad spectrum antibiotic which were discontinued after 36 hours

Adam had different types of seizures namely tonic-clonic, focal and atonic seizures.

Day 5 of admission Adam recovered from his acute illness however he continued to have clusters of focal absences and he was reported to be not his “usual self” being less interactive and playful despite optimising his current anti-seizure medications. The following day he was commenced of a short course of Clobazam with little benefit.

Adam had an EEG after completing course of Clobazam which showed frontally predominant continuous notched delta activity as spike and wave discharges with only a few seconds of normal background consistent with non-convulsive status. Adam was trialled with further loading of Levetiracetam, again with no benefit

Team after discussion with carer decided to commence Adam on 3 days of Methylprednisolone at a dose of 30mg/kg/day

Results

Adam on his second day of corticosteroid therapy was more alert and responsive which was backed with repeat EEG showing improvement with more background activity and by the third day he was reported to be back to his baseline function and discharge from inpatient services after nearly spending two week in a hospital.

Discussion

Managing epilepsy in patients with AS often pose a clinical challenge and multiple therapies have been tried including diazepam(3) and levetiracetam(4) especially in the context of NCSE along with corticosteroids(5) to reduce the seizure burden but there is little evidence to support its use and more evidence/research is needed preferably with a larger cohort however it may still be considered on a case-to-case basis.

References

1- Samanta D. Epilepsy in Angelman syndrome: A scoping review. Brain Dev. 2021 Jan;43(1):32-44. doi: 10.1016/j.braindev.2020.08.014. Epub 2020 Sep 4. PMID: 32893075; PMCID: PMC7688500.
2- Angelman Syndrome | Epilepsy Foundation
3- Worden L, Grocott O, Tourjee A, Chan F, Thibert R. Diazepam for outpatient treatment of nonconvulsive status epilepticus in pediatric patients with Angelman syndrome. Epilepsy Behav. 2018 May;82:74-80. doi: 10.1016/j.yebeh.2018.02.027. Epub 2018 Mar 27. PMID: 29597185.
4- Weber P. Levetiracetam in nonconvulsive status epilepticus in a child with Angelman syndrome. J Child Neurol. 2010 Mar;25(3):393-6. doi: 10.1177/0883073809338626. Epub 2009 Jul 15. PMID: 19605773.
5- Forrest KM, Young H, Dale RC, Gill DS. Benefit of corticosteroid therapy in Angelman syndrome. J Child Neurol. 2009 Aug;24(8):952-8. doi: 10.1177/0883073808331344. PMID: 19666884.