

Management of Nonconvulsive Status Epilepticus in Angelman Syndrome

Up to 50% of patients with Angelman syndrome (AS) will have nonconvulsive status epilepticus (NCSE) with myoclonic or atypical absence status. Adapted from the experience of the Angelman group at Massachusetts General Hospital, this document provides guidance on the outpatient management of patients with AS and NCSE treating with a tapering course of oral diazepam (Worden et al, 2018; PMID: 29597185).

While standard clinical EEG can confirm the presence of nonconvulsive status epilepticus (NCSE) in individuals with Angelman syndrome, it is not always necessary.

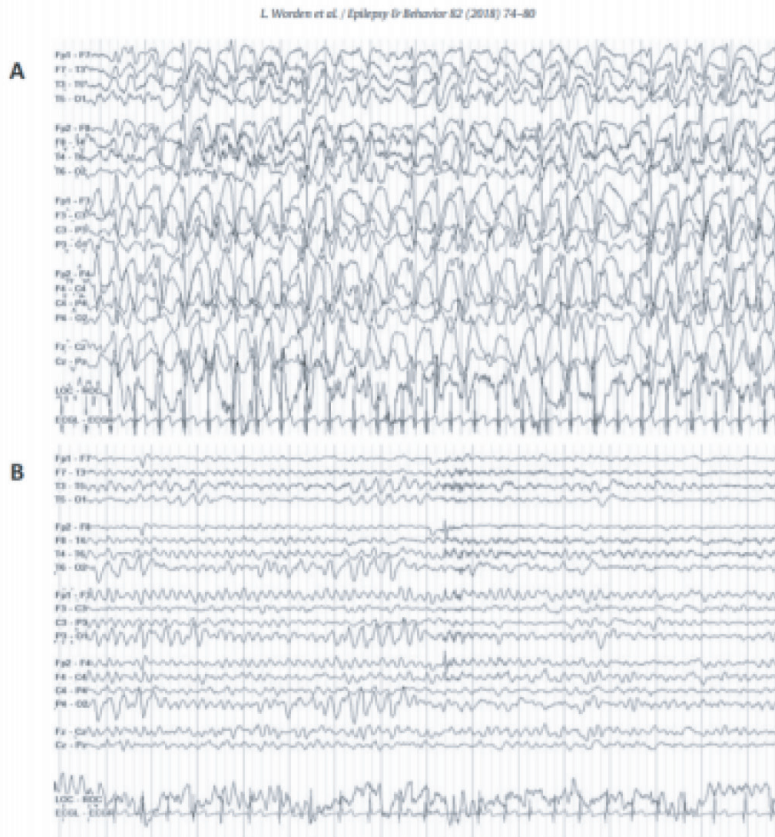


Fig. 2. Electroencephalogram sample for Patient 1 during 3rd episode of NCSE. A. Pre-diazepam treatment EEG showing 2–2.5 Hz, 700 µV frontally predominant generalized spike-wave activity. B. Post-diazepam treatment EEG showing continuous diffuse irregular 5–6 Hz, 200 µV theta slowing and occasional biposterior notched delta activity.

NCSE typically consists of "periods of decreased responsiveness or alertness, which may last hours to days, often with loss of developmental skills (Duis et al, 2022)." NCSE may sometimes occur in the setting of an increase in readily recognized clinically evident seizures, however, preceding clinical seizures may not be present in all cases.

We advise that physicians and other medical providers pay close attention to parents' concerns of such changes or declines in their child's typical behavior, and NCSE be considered as a possible cause of such changes. We recommend a low threshold for implementing the diazepam "reset" therapy regimen when NCSE is confirmed or strongly suspected.

Recommended Valium Taper Regimen:

6 day tapering course of oral diazepam (lorazepam can be used if there is a tolerance issue or sensitivity to diazepam)

Beginning with 0.25-0.5 mg/kg/day divided TID x 2 days

Then tapering to BID dosing for 2 days and then daily for 2 days before tapering off entirely

Example schedule for 20kg child:

0.3mg/kg/day = 6mg/day

DAY 1-2	2MG TAB TID
DAY 3-4	2MG TAB BID
DAY 5-6	2MG TAB DAILY
DAY 7	OFF VALIUM, BACK TO BASELINE MAINTENANCE MEDICATIONS

In addition, consider increasing maintenance dosing of medications or further carb restriction for children on diet therapy for seizure control to prevent future breakthrough seizures and/or recurrent NCSE.

Some children required repeat tapers or prolonged courses up to 2 weeks to break NCSE.



Full reference: 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.

Disclaimer: This document was created and reviewed by the LADDER Learning Network clinicians. This should not be considered medical advice, but a resource that you can provide to your medical care teams to help understand seizure activity and treatment options in Angelman Syndrome.

