angelman SYNDROME FOUNDATION

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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.

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

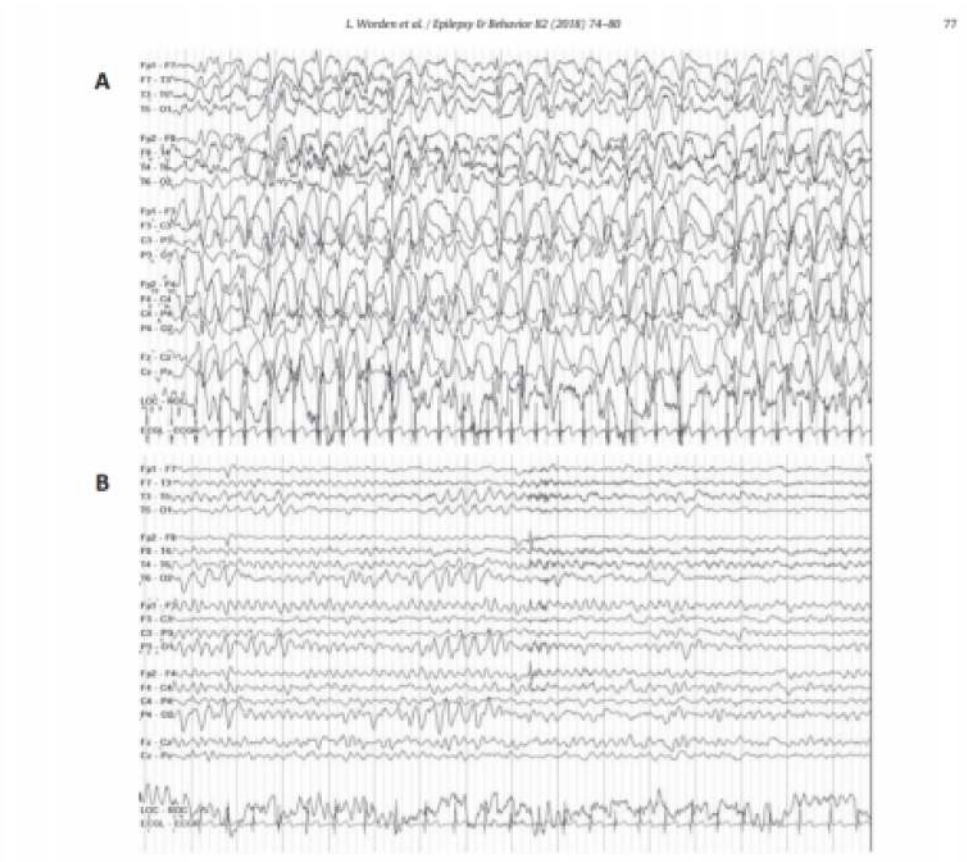

Fig. 2. Electroencephalogram sample for Patient 1 during 3rd episode of NCSE. A. Prediazepam treatment EEG showing 2–2.5 Hz, 700 μV frontally predominant generalized spike–wave activity. B. Postdiazepam treatment EEG showing continuous diffuse irregular 5–6 Hz, 200 μV theta slowing and occasional biposterior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

NSCE should be suspected with the following constellation of clinical findings:

- Somnolence
- Fatigue
- Regression
- Increased seizure activity*
- Cluster of seizure activity at onset*
- Decreased activity

Angelman syndrome is a rare genetic disorder and caregivers are often the first to recognize the signs of clinical decline in their child. Their concerns should be heeded and low threshold to consider valium taper if NCSE is confirmed or 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.