

CHIEF COMPLAINT:, Intractable epilepsy, here for video EEG.,HISTORY OF PRESENT ILLNESS: , The patient is a 9-year-old male who has history of global developmental delay and infantile spasms. Ultimately, imaging study shows an MRI with absent genu of the corpus callosum and thinning of the splenium of the corpus callosum, showing a pattern of cerebral dysgenesis. He has had severe global developmental delay, and is nonverbal. He can follow objects with his eyes, but has no ability to interact with his environment to any great degree. He has noted if any purposeful use of the hands. He has abnormal movements constantly, which are more choreiform and dystonic. He has spastic quadriparesis, which is variable at times. The patient is unable to sit or stand, and receives all his nutrition via G-tube.,The patient began having seizures in infancy presenting as infantile spasms. I began seeing him at 20 months of age. At that point, he had undergone workup in Seattle, Washington and then was seeing Dr. X, child neurologist in Mexico, who started Vigabatrin for infantile spasms. The patient had benefit from this medication, and was doing well at that time with regard to that seizure type. He initially was on phenobarbital, which failed to give him benefit. He continued on phenobarbital; however, for a long period time thereafter. The patient then began having more tonic seizures after his episodic spasms had subsided, and failed several medication trials including valproic acid, Topamax, and Zonegran at least briefly. Upon starting Lamictal, he began to have benefit and then actually had 1-year seizure

freedom before having an isolated seizure or 2. Over the next 6 months to a year, he only had few further seizures, and was doing well in a general sense. It was more recently that he began having new seizure events that have not responded to higher doses of Lamictal up to 15 mg/kg/day. These events manifest as tonic spells with eye deviation and posturing.

Mother reports flexion of the upper extremities, extension with lower extremities. During that time, he is not able to cry or say any sounds. These events last from seconds to minutes, and occur at least multiple times per week. There are times where he has none for a few days and other times where he has multiple days in a row with events. He has another event manifesting as flexion of the upper extremities and extension lower extremities where he turns red and cries throughout. He may vomit after these episodes, then seems to calm down. It is unclear whether this is a seizure or whether the patient is still responsive.

MEDICATIONS: The patient's medications include Lamictal for a total of 200 mg twice a day. It is a 150 mg tablet and 25 mg tablets. He is on Zonegran using 25 mg capsules 2 capsules twice daily, and baclofen 10 mg three times day. He has other medications including the Xopenex and Atrovent.

REVIEW OF SYSTEMS: , At this time is negative any fevers, nausea, vomiting, diarrhea, abdominal complaints, rashes, arthritis, or arthralgias. No respiratory or cardiovascular complaints. He has no change in his skills at this point.

FAMILY HISTORY: , Noncontributory.

PHYSICAL EXAMINATION:

GENERAL: The patient is a slender male who is microcephalic. He has EEG electrodes in place and is

on the video EEG at that time.,HEENT: His oropharynx shows no lesions.,NECK: Supple without adenopathy.,CHEST: Clear to auscultation.,CARDIOVASCULAR: Regular rate and rhythm. No murmurs.,ABDOMEN: Benign with G-tube in place.,EXTREMITIES: Reveal no clubbing, cyanosis, or edema.,NEUROLOGICAL: The patient is alert and has bilateral esotropia. He is able to fix and follow objects briefly. He is unable to reach for objects. He exhibits constant choreiform movements when excited. These are more prominent in the upper extremities and lower extremities. He has some dystonic posture with flexion of the wrist and fingers bilaterally. He also has plantar flexion at the ankles bilaterally. His cranial nerves reveal that his pupils are equal, round, and reactive to light. Extraocular movements are intact other than bilateral esotropia. His face moves symmetrically. Palate elevates in midline. Hearing appears intact bilaterally.,Motor exam reveals dystonic and variable tone, overall there is mild spasticity both upper and lower extremities as described above. He has clonus at the ankles bilaterally, and some valgus contracture of the ankles. His sensation is intact to light touch bilaterally. Deep tendon reflexes are 2 to 3+ bilaterally.,IMPRESSION/PLAN: , This is a 9-year-old male with congenital brain malformation and intractable epilepsy. He has microcephaly as well as dystonic cerebral palsy. He had a re-emergence of seizures, which are difficult to classify, although some sound like tonic episodes and others are more concerning for non-epileptic phenomenon, such as discomfort. He is admitted for video EEG to hopefully capture

both of these episodes and further clarify the seizure type or types. He will remain hospitalized for probably at least 48 hours to 72 hours. He could be discharged sooner if multiple events are captured. His medications, we will continue his current dose of Zonegran and Lamictal for now. Both of these medications are very long acting, discontinuing them while in the hospital may simply result in severe seizures after discharge.