Results of a Pilot feasibility study to develop reduce wait times strategy in the evaluation of children with new onset epilepsy


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Background: The goal was to understand factors leading to prolonged wait times for neurological assessment of children with new onset seizures. A second objective was to develop an innovative approach to patient flow through and achieve a reduction in waiting times utilizing limited resources.

Methods:
1. Audit of the referrals, flow through, wait times
2. Identification of bottlenecks
3. Development of triaging strategy:
   i. Suspected Febrile seizures and non-epileptic events;
   ii. Suspected benign and absence epilepsies;
   iii. Suspected other Focal epilepsies, generalized epilepsies, epilepsy under 2 years
4. Initiation of early telephone contact and support
5. Development of a ketogenic diet

Results: Using a triaging strategy and focusing on timely access to investigations, wait times for clinic evaluations were shortened despite larger numbers of referrals (mean wait time reductions from 179 to 91 days). Limiting factors such increase in referral numbers, attrition in support staff, interfered with sustainability of reduced wait times achieved in the initial phase of the program. Conclusions: This pilot study highlights the effectiveness of an innovative triaging strategy and improvements in patient flow through in achieving the goals of reduction in wait times for clinical evaluation and timely investigations to improve care for children with new onset seizures. Insights into limitations of such strategies and factors determining sustainability are discussed.

Forced normalization after vagal nerve stimulation in a case of intractable Lennox-Gastaut syndrome

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Background: Forced normalization is the development of psychiatric symptoms in a patient experiencing remission of intractable seizures. The mechanism of this phenomenon is unknown. We present a complex case of Lennox-Gastaut syndrome that experiences forced normalization after vagal nerve stimulation (VNS). Methods: This case details a 31-year-old male with seizures since early childhood. The patient has intractable epilepsy and failed AEDs, VNS, and a partial callosotomy. Results: The patient was in remission from 2-12 years old, when seizures returned at a frequency of 2-5 per day. He has multiple types of seizures including drop attacks, absences, and tonic-clonic seizures. Patient experienced status epilepticus multiple times. Twelve AEDs were failed before VNS was started in 2010, which helped curb the severity of seizures and the potential for clusters. Forced normalization developed over the course of treatment with VNS. The patient behavior was characterized by aggression, paranoia, and hallucinations. VNS was turned off late in 2010 and then re-started in January of 2011. Patient proceeded to cycle between several days of seizures without psychiatric symptoms and several days of psychosis without seizures. Conclusions: Vagus nerve stimulation gave way to forced normalization, characterized here as aggressive behaviour and psychosis. Forced normalization is seen commonly after epilepsy surgery, but rarely following VNS.

Novel GRIN2A variant in family members with variable phenotypic expression of epilepsy


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Background: Epilepsy aphasia spectrum of disorders is characterized by developmental and language regression with EEG abnormalities that include electrical status epilepticus of sleep (ESES). Landau-Kleffner syndrome (LKS) and epileptic encephalopathy with continuous spike-wave during sleep (CSWS) are the most severe presentations. GRIN2A mutations have been recognized as causative. Methods: we present two sisters with different epilepsy phenotypes. A variant of unknown clinical significance (VUS) in GRIN2A gene was found in one of the sisters and her similarly affected father. Results: The first sister presented with focal onset seizures at the age of 3 years accompanied by language and cognitive regression and EEG features consistent of ESES, meeting criteria for LKS. Multiple anticonvulsants were tried until she responded well to steroids regaining developmental milestones. Her 5-year-old sister recently presented with focal onset seizures. Her language development is appropriate. Her EEG showed independent multifocal spikes but no ESES during sleep. Her seizures were controlled on monotherapy anticonvulsants. Conclusions: We observed a variable EEG-clinical phenotype and different severity among these family members as...