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# Reliable Count of Seizures using Diaries versus Video Electroencephalography for Seizures of KCNQ2 Developmental and Epileptic Encephalopathy (KCNQ2-DEE)

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# Disclosures

- Drs. Grayson, Harden, Luzon, Butterfield, Pimstone, and Aycardi are employed by Xenon Pharmaceuticals Inc. (“Xenon”) They receive salaries and may hold stock or stock options in Xenon.
- The KCNQ2 Cure Alliance received funds from Xenon for this patient survey and other projects/events.
- Pursuant to a Consulting Agreement between Ann and Robert H. Lurie Children’s Hospital of Chicago and Xenon, it is anticipated that Dr. Millichap will serve as the Global Coordinating Investigator for Xenon’s planned Phase 3 study of XEN496. The Ann and Robert H. Lurie Children’s Hospital of Chicago receives financial compensation from Xenon in exchange for Dr. Millichap’s services. In addition, Dr. Millichap, in a personal capacity, is a consultant for Xenon and also serves as a member the Xenon Steering Committee for XEN496. Dr. Millichap receives financial compensation from Xenon in exchange for his services.
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# Introduction

## Objective

- We sought evidence as to whether seizures of infants and young children with KCNQ2 developmental and epileptic encephalopathy (KCNQ2-DEE) could be reliably counted using diaries, with seizure behaviors previously confirmed using video electroencephalography (VEEG) at initial diagnosis

## Background

- The FDA has recommended VEEG to measure the outcome of antiseizure mediation (ASM) clinical trials for infants and young children
    - Reasons for this include difficulty with seizure recognition due to clinically subtle seizure behaviors, and occurrence of subclinical electrographic seizures in this age group
- However, use of VEEG presents technical and feasibility challenges, and limits enrollment to subjects with very high seizure frequencies

# Design/Methods

- All reports of seizures with KCNQ2-DEE were reviewed for seizure and EEG correlate descriptions
- A recent survey of caregivers[1] conducted in collaboration with a patient advocacy group (the KCNQ2 Cure Alliance) was reviewed for information regarding recognition of seizure occurrence

[1] Please also refer to: Harden *et al.* Poster No. 4775, "An Online Survey of Caregivers of Patients with KCNQ2 Developmental & Epileptic Encephalopathy (KCNQ2-DEE). 2020 American Academy of Neurology (AAN) Meeting.

# Results

- Sixteen reports were included, encompassing 124 KCNQ2-DEE patients
- Each described stereotyped focal tonic seizures beginning on days 1-5 of life
- Focal tonic seizure was the predominant and often only seizure type present
- The seizures were readily observed and were associated with a clear EEG correlate of contralateral amplitude suppression
- There were no reports of electrographic subclinical seizures
- To understand the ability of caregivers to record seizures, a survey of KCNQ2 families was undertaken, with 55 evaluable responses obtained
  - On a scale from 1-10 (low-high) confidence in recognizing seizures after training using VEEG, **75% reported at least 8 or above**

# Conclusions

- We conclude that the seizures of KCNQ2-DEE:
  - are stereotyped, clinically evident and do not occur without an observable clinical correlation, and
  - families are recognizing seizures confidently.
- A strategy of training families to count seizures accurately using VEEG mitigates the need to use VEEG directly as the method to assess primary outcome in a KCNQ2-DEE antiseizure medicine treatment trial