stimulation (VNS). Methods: A retrospective study was completed examining the effectiveness of VNS and CC in children with SYNGAP1-DEE using the SynGAP Research Database and an additional child followed at our centre. Results: Fifteen patients from the SynGAP Database were included. Of those who had VNS (n=11), 7 children had an >50% reduction in seizure frequency (n=7/11, 64%), 2 had worsening (n=2/11, 18%), 1 had no change (n=1/11, 9%), and 1 had an unknown response (n=1/11, 9%). Two children had CC only, 1 had complete seizure freedom, and 1 had a >50% reduction. Two children underwent VNS and CC, 1 had a >50% reduction in seizure frequency and the other had no change. One child followed at our centre experienced a sustained >80% reduction in seizure frequency following CC (i.e., after 1.5 years). Conclusions: We provide the first in-depth description of the response to VNS and CC in children with *SYNGAP1*-DEE, and provide insight into the use of of palliative surgical procedures in this population.

P.053

Biallelic SCN1A variants with divergent epilepsy phenotypes

R Pentz (Hamilton) R Hough (Hamilton) C Li (Hamilton) M Tarnopolsky (Hamilton) K Jones (Hamilton) R Ramachandran Nair (Hamilton) R Whitney (Hamilton)*

doi: 10.1017/cjn.2025.10226

Background: Dravet syndrome and genetic epilepsy with febrile seizures plus (GEFS+) are associated with pathogenic variants in *SCN1A*. While most such cases are heterozygous, there have been 16 reported homozygous cases. We report two new biallelic cases associated with divergent phenotypes. Methods: We performed a chart review for two patients with different homozygous SCN1A variants and reviewed all previously published biallelic *SCN1A* pathogenic variants. Results: Our first patient exhibited early afebrile seizures and severe developmental delay, without febrile seizures or status epilepticus. A homozygous c. 1676T>A, (p. Ile559Asn) variant of uncertain significance was identified, carried by asymptomatic parents. The second patient exhibited early, recurrent, and prolonged febrile seizures, moderate developmental delay, and motor dysfunction; a homozygous pathogenic c. 4970G>A, (p. Arg1657His) variant carried by asymptomatic parents was identified.

Of 18 known cases of biallelic SCN1A pathogenic variants, 15/18 (83%) have diagnoses of Dravet or GEFS+. The remaining 3/18 (17%) had pharmacoresponsive epilepsy with prominent GDD. Cognitive phenotypes ranged from intact neurodevelopment to profound developmental delay. Eleven out of 18 cases (61%) had motor concerns. Conclusions: These cases expand the phenotypic spectrum of biallelic *SCN1A* variants. While some patients present typically for Dravet/GEFS+, others present with developmental delay and controllable epilepsy.

P.055

Novel pipeline for triage of variant of uncertain significance reclassification in epilepsy genomics

A Freibauer (Vancouver) W Alotaibi (Vancouver) M Vas (Vancouver) K Burke (Vancouver) C Boelman (Vancouver)*

doi: 10.1017/cjn.2025.10227

Background: Increased availability of genetic testing has led to increased burden of follow up of variants of uncertain significance (VUS). As of January 2025, 327 VUS were identified patients at BC Children's Hospital. We propose a pipeline to triage and follow up of patients with identified VUS to clarify diagnosis through paternal testing. Methods: Of the 327 patients with VUS, 13 patients with high clinical suspicion for a genetic disorder were identified by their neurologist. Initial chart review for each patient was performed. Clinical phenotype data and the patient's variant were inputted into the online tool Franklin. This program generates a variant interpretation based on 17/ 28 criteria in ACMG scoring. For each patient the variant would be assumed to be de novo in order to determine if parental testing could change variant classification. Results: 5/13 of the patients had suggested reclassification of variants. 6/13 of the patients would have reclassification of variant to likely pathogenic/pathogenic if the variant was found to be de novo, suggesting a need for paternal testing. Conclusions: This highlights a novel clinical pipeline to improve expediency and triaging of VUS reclassification for paternal testing in epilepsy genomics.

P.057

A review of a twenty-seven year experience with the Ketogenic diet: lessons learned and moving forward

H Davies (Montreal) K Myers (Montreal)*

doi: 10.1017/cjn.2025.10228

Background: Although the history of the ketogenic diet dates back centuries, with the advancement of anti-seizure medications, the use of the diet for epilepsy declined. It was not until the early 1990s that there was a resurgence of the diet as an adjunct therapy to anti-seizure medication. In 1998, the Montreal Children's Hospital introduced the ketogenic diet to a child with drug resistant epilepsy. Shortly after, a presentation of the ketogenic diet at hospital Grand Rounds met much skepticism. However, over time the diet has developed into a well-established treatment option for children with drug resistant epilepsy. Two hundred children have since utilized the diet at the Montreal Children's Hospital. Methods: A review of patient files since the initiation of the program was undertaken. Data was extracted regarding adverse effects, common errors in both hospital and home setting, risks for unfavorable outcomes and parental concerns Results: