zebrafish knockdown model to assess for rescue of the morphant phenotype. **Results:** Four patients shared a novel missense homozygous mutation in *TNNT1*. They developed from childhood slowly progressive limb-girdle weakness with spinal rigidity and contractures. They suffered from restrictive lung disease and recurrent episodes of rhabdomyolysis. Older patients remained ambulatory into their sixties. Lower extremity MRI showed symmetrical myopathic changes. Paraspinal MRI showed diffuse fibro-fatty involution. Biopsies showed multi-minicores. Nemaline rods were seen in half the patients. *TNNT1* mRNA expression was similar in controls and patients, while levels of *TNNT1* protein were reduced in patients. Wild type *TNNT1* mRNA rescued the zebrafish morphants but mutant transcripts failed to do so. **Conclusions:** This study expands the spectrum of *TNNT1*-related myopathy to include a milder clinical phenotype caused by a functionally-confirmed novel mutation.

A.5

Identification of predictors of response to Erenumab in episodic and chronic migraine in a cohort of patients: a preliminary analysis

H Salem-Abdou (Quebec)*, *J Puymirat* (Quebec)

doi: 10.1017/cjn.2021.267

Background: Erenumab is an antibody anti-calcitonin gene related peptide (CGRP) receptor approved for the treatment of episodic (EM) and chronic migraine (CM). In this study, we aimed to identify the predictors of response to the treatment. **Methods:** This is an ongoing retrospective cohort study of 120 patients (49 with cervicalgia) with EM or CM treated with Erenumab. The first endpoint was to identify the success rate of this treatment (at least 50% reduction in monthly migraine days during the third month of the treatment). The second endpoint was to identify the predictors of response to Erenumab treatment. **Results:** Seventy one percent of patients achieved a favorable response (P-value<0.001) to Erenumab. Patients with cervicalgia showed a lower treatment success rate (21.1% with vs 40.8% without cervicalgia) while patients without cervicalgia showed a higher treatment success rate (78. 9% without vs 59.2% with cervicalgia) with a P-value of 0.025 and an odd ratio of 0.388 (95% CI 0.174-0.869, P-value=0.021). A similar trend was observed in patients with occipital neuralgia and obesity (P-value<0.08). **Conclusions:** The preliminary analysis of this study demonstrates that cervicalgia (and to a lesser extend occipital neuralgia and obesity) is a negative predictor of response to Erenumab in patients with migraine.

A.6

Vagus Nerve Stimulation in patients with therapy resistant generalized epilepsy

AS Suller Marti (London)* *SM Mirsattari* (London) *KW MacDougall* (London) *D Steven* (London) *A Parrent* (London) *A Andrade* (London) *S de Ribaupierre* (London) *DC Diosy* (London) *R McLachlan* (London), *JG Burneo* (London)

doi: 10.1017/cjn.2021.268

Background: For patients with generalized epilepsy who do not respond to anti-seizure medications, the therapeutic options are limited. Vagus nerve stimulation (VNS) is a treatment mainly approved for therapy resistant focal epilepsy. There is limited information on the use of VNS on generalized epilepsies, including Lennox Gastaut Syndrome(LGS) and genetic generalized epilepsy(GGE). **Methods:** We identified patients with a diagnosis of Lennox-Gastaut Syndrome or Genetic Generalized Epilepsy, who underwent VNS implantation, between1997 and July 2018. **Results:** A total of 46 patients were included in this study with a history of therapy resistant generalized epilepsy. The mean age at implantation was 24 years(IQR= 17.8-31 years) and 50%(n=23) were female. The most common etiologies were GGE in 37%(n=17) and LGS in 63%(n=29). Median follow-up since VNS implantation was 63 months(IQR:31-112.8months). 41.7%(n=12) of the LGS group became responders, and 64.7%(n=11) in the GGE group. The best response in seizure reduction was seen in generalized tonic-clonic seizures. There was a reduction of seizure-related hospital admissions from 89.7%(N=26) pre-implantation, to 41.4%(N=12) post-implantation (p<0.0001). The frequency of side effects due to the stimulation was similar in both groups(62.1% in LGS and 61.1% in GGE). **Conclusions:** VNS is an effective treatment in patients with therapy resistant generalized epilepsy, especially GGE.

A.7

In vivo hippocampal mGluR5 abnormalities predict MTLE post-surgical outcome

J Lam (Montreal)* *JM DuBois* (Montreal) *J Rowley* (Montreal) *OG Rousset* (Baltimore) *KA González-Otárula* (Montreal) *J Soucy* (Montreal) *G Massarweh* (Montreal) *JA Hall* (Montreal) *M Guiot* (Montreal) *M Zimmermann* (Montreal) *L Minuzzi* (Hamilton) *P Rosa-Neto* (Montreal), *E Kobayashi* (Montreal)

doi: 10.1017/cjn.2021.269

Background: PET imaging of [^{11}C]ABP688 shows reduced hippocampal mGluR5 availability in mesial temporal lobe epilepsy (MTLE) patients, however the relation with post-surgical outcomes is unclear. Here, we tested whether [^{11}C]ABP688 binding in hippocampal subfields vulnerable to glutamate excitotoxicity is related to post-surgical outcome. **Methods:** [^{11}C]ABP688-PET was obtained from 31 unilateral MTLE patients and 30 controls. Hippocampal subfields were automatically segmented into 1) CA1-3, 2) CA4/dentate gyrus (DG), and 3) Subiculum and manually corrected. Partial volume corrected [^{11}C]ABP688 non-displaceable binding potential (BP_{ND}) was calculated in the subfields and compared between seizure-free and non-seizure-free patients. **Results:** [^{11}C]ABP688 BP_{ND} was significantly reduced in ipsilateral CA1-3 & CA4/DG (p<0.001) compared to controls. No difference was seen in Subiculum. Ipsilateral CA1-3 [^{11}C]ABP688 BP_{ND} was lower in seizure-free (p=0.012; Engel Ia, n=13) vs non-seizure-free (Engel Ic-III, n=10) patients, and this effect was independent of subfield volume. In a subset of patients with [^{18}F]FDG-PET, CA1-3 [^{11}C]ABP688 BP_{ND} was significantly lower in seizure-free patients (p=0.03), while no difference was found for [^{18}F]FDG uptake. **Conclusions:** Reduced CA1-3 mGluR5 availability was associated with post-surgical seizure-freedom independent of atrophy and hypometabolism. Thus, [^{11}C]ABP688-PET may offer a potential biomarker for surgical outcomes and may be particularly relevant for pre-surgical workup in MRI- and [^{18}F]FDG-negative MTLE patients.