associated with better outcomes. Cerebrospinal fluids (CSF) white blood cell (WBC) count and protein concentration measured early on in the disease process is often used, in combination with other clinical factors, to evaluate the likelihood that a patient has AE. **Methods:** CSF characteristics (WBC count, protein concentration, and oligoclonal banding) measured in a first AE presentation, prior to results of autoantibodies being available, were retrospectively analyzed at two tertiary care centers. **Results:** Ninety-five patients were included in the study. CSF WBC counts and protein levels were within normal limits for 27% (CI95%: 19–37) of patients with AE. When results of oligoclonal banding were added, 14% (CI95%: 6–16) of patients had "normal" CSF. The median CSF white blood cell count was 8 cells/mm3 (range: 0–544) and the median CSF protein concentration was 0.42 g/L (range: 0.15–3.92). **Conclusions:** A substantial proportion of patients with early active AE had a CSF WBC count or protein concentration within the normal. Inclusion of CSF oligoclonal banding may help identify a higher proportion of patients with an inflammatory CSF profile early in the disease process.

A.2

Clinical application of T1-w/T2-w ratio images for in vivo comparisons of myelin content in patients with trigeminal neuralgia

C Li (London)* *PP Chu* (Toronto) *P Hung* (Toronto) *D Mikulis* (Toronto), *M Hodaie* (Toronto)

Background: Novel magnetic resonance (MR) imaging techniques prompted the emergence of T1-w/T2-w images or "myelin-sensitive maps (MMs)" to measure myelin *in vivo*. However, acquisition-related variations in MR intensities prevent meaningful quantitative comparisons between MMs. We propose an improved pipeline to standardize MMs that is applied to patients with classic trigeminal neuralgia (CTN) and trigeminal neuralgia secondary to multiple sclerosis (MSTN). **Methods:** 3T scanner was used to obtain T1-w and T2-w images for 17 CTN and 17 MSTN patients. Template images were obtained from ICBM152 database. MS plaques and normal-appearing white matter (NAWM) were labelled. A Gaussian curve-fit was applied to the histogram of the intensity distribution of each patient image, and transformed to match the Gaussian curve-fit of the template image. **Results:** MM intensities were decreased within MS plaques, compared to NAWM in MSTN patients (*p<0.001*) and its corresponding regions in CTN patients (*p<0.001*). Qualitatively, the standardized patient image and its histogram better resembled the ICBM152 template. **Conclusions:** MM analysis revealed reduced myelin content in MS plaques compared to corresponding regions in CTN patients and surrounding NAWM in MSTN patients. The standardized MM serves as a non-invasive, clinical tool for quantitative analyses of myelin content between different brain regions and different patients *in vivo*.

A.3

Use of diffusion-weighted imaging to distinguish seizure-related change from limbic encephalitis

A Budhram (Rochester)* *JW Britton* (Rochester) *GB Liebo* (Rochester) *A McKeon* (Rochester) *SJ Pittock* (Rochester), *NL Zalewski* (Rochester)

Background: Limbic encephalitis (LE) classically causes medial temporal lobe T2-hyperintensity on magnetic resonance imaging (MRI), but this can also occur with seizure activity. Identifying neuroimaging patterns that can distinguish between LE and seizure activity may help avoid diagnostic confusion in such challenging cases. **Methods:** Through retrospective review of Mayo Clinic patients who had medial temporal lobe T2-hyperintensity on MRI, we identified non-LE patients with seizure-related medial temporal lobe T2-hyperintensity. Their diffusion-weighted imaging (DWI) was reviewed to look for diffusion restriction patterns potentially unique to seizure activity. Next, a control cohort of LE patients with medial temporal lobe T2-hyperintensity was identified, and their DWI was reviewed to see if these diffusion restriction patterns could help distinguish seizure activity from LE. **Results:** We identified 10 non-LE patients who had medial temporal lobe T2-hyperintensity due to seizure activity; 9/10 had one of two medial temporal lobe diffusion restriction patterns we uncovered as being potentially unique to seizure activity. In contrast, only 5/57 LE patients had one of these diffusion restriction patterns identified, all of whom had seizures reported. **Conclusions:** We report two diffusion restriction patterns that may help distinguish seizure activity from LE. Recognition of these diffusion restriction patterns should prompt evaluation for possible seizure activity.

A.4

A Novel Recessive TNNT1 Congenital Core-Rod Myopathy in French Canadians

D Pellerin (Montreal)* *A Aykanat* (Boston) *B Ellezam* (Montreal) *EC Troiano* (Boston) *J Karamchandani* (Montreal) *M Dicaire* (Montreal) *M Petitclerc* (Lévis) *R Robertson* (Montreal) *X Allard-Chamard* (Montreal) *D Brunet* (Quebec city) *CG Konersman* (San Diego) *J Mathieu* (Sherbrooke) *J Warman Chardon* (Ottawa) *VA Gupta* (Boston) *AH Beggs* (Boston) *B Brais* (Montreal), *N Chrestian* (Quebec city)

Background: Mutations in the slow skeletal muscle troponin T (*TNNT1*) gene cause a congenital nemaline myopathy resulting in death from respiratory insufficiency in early infancy. We report on four French Canadians with a novel congenital *TNNT1* myopathy. **Methods:** Patients underwent lower extremity and paraspinal MRI, quadriceps biopsy and genetic testing. *TNNT1* expression in muscle was assessed by quantitative PCR and immunoblotting. Wild type or mutated *TNNT1* mRNAs were co-injected with morpholinos in a

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)

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)

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)

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.