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)

doi: 10.1017/cjn.2021.264

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)

doi: 10.1017/cjn.2021.265

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)

doi: 10.1017/cjn.2021.266

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