decreasing IgG recycling and reducing pathogenic IgG autoantibody levels. ADHERE assessed the efficacy and safety of efgartigimod PH20 subcutaneous (SC; co-formulated with recombinant human hyaluronidase PH20) in chronic inflammatory demyelinating polyneuropathy (CIDP). Methods: ADHERE enrolled participants with CIDP (treatment naive or on standard treatments withdrawn during run-in period) and consisted of open-label Stage A (efgartigimod PH20 SC once weekly [QW]), and randomized (1:1) Stage B (efgartigimod or placebo QW). Primary outcomes were clinical improvement (assessed with aINCAT, I-RODS, or mean grip strength; Stage A) and time to first aINCAT score deterioration (relapse; Stage B). Secondary outcomes included treatment-emergent adverse events (TEAEs) incidence. Results: 322 participants entered Stage A. 214 (66.5%) were considered responders, randomized, and treated in Stage B. Efgartigimod significantly reduced the risk of relapse (HR: 0.394; 95% CI: 0.25–0.61) versus placebo ($p$=0.000039). Reduced risk of relapse occurred in participants receiving corticosteroids, intravenous or SC immunoglobulin, or no treatment before study entry. Most TEAEs were mild to moderate; 3 deaths occurred, none related to efgartigimod. Conclusions: Participants treated with efgartigimod PH20 SC maintained a clinical response and remained relapse-free longer than those treated with placebo.

## D.2

### Cost-effectiveness analysis of efgartigimod vs chronic IVIg for treatment of patients with generalized myasthenia gravis in Canada

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Background: Efgartigimod is a human IgG1 antibody Fc fragment recently approved by Health Canada for patients with acetylcholine receptor antibody positive (AChR-Ab+) generalized myasthenia gravis (gMG). We assessed cost-effectiveness of efgartigimod vs chronic IVIg for adult patients with AChR-Ab+ gMG. Methods: A Markov model estimated costs (treatment and administration, disease monitoring, complications from chronic corticosteroid use, exacerbation and crisis management, adverse events, end-of-life care) and benefits (quality-adjusted life-years [QALYs]). The analysis was conducted from the Canadian healthcare system perspective. Health state transition probabilities were estimated using data from ADAPT, ADAPT+, and a network meta-analysis comparing efgartigimod with chronic IVIg. Utility values were obtained from MyRealWorld MG. Patients with MG-ADL ≥5 who did not die/discontinue were assumed to receive treatment every 4 weeks or every 3 weeks over the lifetime horizon. Results: Over the lifetime horizon, efgartigimod and chronic IVIg were predicted to have total discounted QALYs of 16.80 and 13.35, and total discounted costs of $1,913,294 and $2,170,315, respectively. Efgartigimod dominated chronic IVIg with incremental QALYs of 3.45 and cost savings of $257,020 over the lifetime horizon. Conclusions: Efgartigimod may provide greater benefit at lower costs than chronic IVIg for Canadian patients with AChR-Ab+ gMG, with substantial healthcare system savings over the lifetime horizon.

## D3

### Interim results for Myasthenia Gravis-resource utilization, epidemiology, survival & treatment patterns (MG-REST) study in Ontario, Canada

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Background: Reliable real-world data on the burden of MG is needed to inform Canadian clinical and policy decisions in the era of new MG therapeutics, including FcRn inhibitors. Given the lack of recent Canadian data on MG disease burden, the MG-REST Study aims to estimate the clinical burden of MG in Ontario. Methods: Ontario administrative data from ICES were utilized for a retrospective population-based cohort study of adults with MG identified through a validated algorithm (April 2013-March 2019) and followed for up to seven years (March 2020) to determine myasthenic crisis characteristics and overall survival (OS). Results: The MG cohort (n=2,601) had an average age of 65.7 years and 53.3% were males. Incidence of first myasthenic crisis was 9%, with 87% of events occurring at/after diagnosis. MG OS was 89%, 85% and 75% at 1-year, 2-years and 5-years, respectively, while OS after first crisis was 60%, 52%, and 39% for the same years. Conclusions: Despite the availability of conventional therapies throughout the study, MG crisis remains a serious, common complication of MG, with decreased survival at 1-year post-crisis (29% difference versus 1-year OS following MG diagnosis). Study highlights MG burden and unmet need for new effective therapies for MG treatment.

## D.4

### Safety and efficacy of delandistrogene moxeparvovec versus placebo in Duchenne muscular dystrophy (EMBARK): Pivotal Phase 3 primary results

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Background: Duchenne muscular dystrophy (DMD) is caused by DMD gene mutations. Delandistrogene moxeparvovec is an investigational gene transfer therapy, developed to address the underlying cause of DMD. We report findings from Part 1 (52 weeks) of the two-part EMBARK trial (NCT05096221). Methods: Key inclusion criteria: Ambulatory patients aged ≥4-<8 years with a confirmed DMD mutation within exons 18–79 (inclusive); North Star Ambulatory Assessment (NSAA) score >16 and <29 at screening. Eligible patients were randomized 1:1 to intravenous delandistrogene moxeparvovec (1.33×1014 vg/kg) or placebo. The primary endpoint was

change from baseline in NSAA total score to Week 52. Results: At Week 52 (n=125), the primary endpoint did not reach statistical significance, although there was a nominal difference in change from baseline in NSAA total score in the delandistrogene moxeparvovec (2.6, n=63) versus placebo groups (1.9, n=61). Key secondary endpoints (time to rise, micro-dystrophin expression, 10-meter walk/run) demonstrated treatment benefit in both age groups (4-5 and 6-7 years; $p<0.05$). There were no new safety signals, reinforcing the favorable and manageable safety profile observed to date. Conclusions: Based on the totality of functional assessments including the timed function tests, treatment with delandistrogene moxeparvovec indicates beneficial modification of disease trajectory.

## D.5

### Low density scalp electrical source imaging of the ictal onset zone network using source coherence maps

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Background: This study investigates the utility of low-density scalp electrical source imaging (LD-ESI) of the Ictal Onset Zone (IOZ) and interictal spike ripple high frequency oscillation (ISRHFO) networks using Source Coherence Maps (SCM) in the surgical evaluation of children with medically refractory epilepsy. Invasive intracranial monitoring, the gold standard for determining epileptogenic zones, has limited spatial sampling. SCM presents a promising new non-invasive diagnostic technique. Methods: This was a retrospective review of 11 patients who underwent focal resections. SCMs were generated using Standardized Low Resolution Electromagnetic Tomography (sLORETA). SCM concordance to resection margins was assessed, noting outcomes at 3 years. Results: For 7/11 cases, ictal SCMs included the resection, and 5/7 achieved seizure freedom, indicating inclusion of the epileptogenic zone. For the 2/7 not seizure-free, the IOZ networks on the SCMs extended beyond resection margins, suggesting the epileptogenic zone also extended beyond the resection. Interictal spike ripple ESI and ISRHFO SCM were performed for 7/11, with 3/7 included in the resection and all 3 seizure-free. Conclusions: These findings may support LD-ESI of the IOZ and ISRHFO network using SCM as promising methods complementary to ictal and interictal ESI in pediatric epilepsy surgical workup, guiding electrode placement for intracranial monitoring to identify the epileptogenic zone.

## D.6

### Neurological care and outcomes of pregnant patients with epilepsy in a Canadian tertiary care center (2014-2020)

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Background: Limited data exists on neurological care and outcomes of Canadian pregnant patients with epilepsy (PPWE). This study provides Canadian data to inform practice patterns and observed outcomes for PPWE at a tertiary care center. Methods: PPWE receiving care at the University Health Network (Toronto, Canada) epilepsy clinic from January 1, 2014 to November 20 2020 were retrospectively identified with demographics and neurological data and outcomes collected. Results: A total of 195 cases were identified, with a median maternal age of 32 years (SD 4.58), a median age at first seizure of 17 years (range 1 month – 36 years old), 52% were diagnosed with genetic generalized epilepsy and 50% endorsed 6 months of seizure freedom prior to conception. In pregnancy, 93% took ASM(s) with 77% receiving therapeutic drug monitoring (TDM) and drug dose adjustments reported in 69%. Most cases (73%) maintained a stable seizure frequency. Conclusions: This study provides new Canadian data on PPWE at a tertiary care center. PPWE are overall well controlled, more likely to have young adult onset, genetic generalized epilepsy with nearly all taking ASM(s) during pregnancy. While high rates of TDM and drug dose adjustments were observed, most experienced seizure stability in pregnancy.

# Neuroradiology (CSNR)

## E.1

### Focal leptomeningeal vascular anomalies on brain MRI: a mimic of leptomeningeal metastatic disease

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Background: The diagnosis of leptomeningeal metastatic disease has major prognostic implications. We report 13 patients with a radiologically distinct, focal, enhancing leptomeningeal lesion on brain MRI mimicking leptomeningeal metastatic disease. Methods: These patients were assessed at University Health Network between January 2001 and December 2023. Results: Median age was 68 years and 10 patients were women. All patients had brain MRI including contrast-enhanced T2-weighted FLAIR and T1-weighted spin echo sequences. MRI in all patients showed a focal enhancing lesion along the leptomeningeal surface of the brain. The MRI exams were reported as possible metastatic disease for the majority (9/13) of patients. Each lesion was curvilinear rather than sheet-like, and some consisted of multiple connected/branching curvilinear structures with the appearance of abnormal vessels. Some lesions had visible connection with a nearby cortical vein. The lesions were distinct from normal blood vessels. Follow-up contrast-enhanced brain MRI for 8/13 patients at a median of 3.9 years showed all lesions were unchanged over time. Conclusions: We describe a distinct kind of focal, enhancing leptomeningeal lesion on brain MRI that mimics metastatic disease. These lesions are likely a type of low-flow vascular anomaly. Their curvilinear/branching shape and intense enhancement particularly on T2-weighted FLAIR images distinguishes these lesions from tumor.