CBD: 7.5%, 142/1894), other frontotemporal lobar degeneration (FTLD: 5.7%, 32/561), Lewy body disease (LBD: 4.1%, 49/1202), and Alzheimer disease (AD: 1.8%, 48/2687). Average age-at-symptom onset was 69.5±10.4 years. Average disease duration was 2.9±1.0 years. Prion diseases had the most rapid disease course (1.6±1.3 years). Comorbid cerebrovascular disease (25.5%), and clinically symptomatic depression (41.3%), psychoses (37.1%), and sleep disturbances (39.4%) were common across groups. Only psychosis was associated with shorter disease duration (β=-0.31 years, $CI_{95\%}$ -0.53, -0.082, controlling for age-at-symptomatic onset). Conclusions: Although prion disease commonly presented as RPD, atypical presentations of more prevalent neurodegenerative diseases accounted for most cases of RPD. Rapidly progressive variants of typical neurodegenerative diseases warrant consideration in clinical practice.

P.006

Etiologic diagnoses of rapidly progressive dementia in a prospective multicenter cohort

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Background: Accurate etiologic diagnoses are needed in patients with rapidly progressive dementia (RPD) to ensure access to symptomatic and disease-modifying therapies when available. Methods: Patients with RPD were prospectively enrolled and evaluated at Washington University (Saint Louis, MO; 2016-2019) and Mayo Clinic (Jacksonville, FL; 2020-2021). Etiologic diagnoses were independently assigned by two dementia specialists integrating clinical features and the results of diagnostic tests; disagreements were resolved via blinded review by a third specialist. Results: 160 RPD patients were enrolled and followed. Average age-at-symptom onset was 60.0±15.9 years; 50% were female. Inter-rater reliability (91% agreement; Cohen's κ=0.88, p<0.001) and clinicopathologic correlation were excellent (100% agreement in 24 patients with neuropathologic data). Autoimmune encephalitis was the leading cause of RPD (39%), followed by Alzheimer disease and related dementias (29%), Creutzfeldt-Jakob disease (15%), and other causes (15%). Patients with potentially treatable causes of RPD were younger (54.5±18.2 than those with neurodegenerative causes (67.3±9.5; p<0.001), and more likely to present with altered levels of consciousness, seizures, or CSF pleocytosis (p<0.05). Conclusions: Etiologic diagnoses can be reliably established in RPD patients using available clinical data. The prevalence of autoimmune encephalitis in this series justifies routine screening for potentially treatment-responsive causes of RPD, particularly in younger patients.

Epilepsy and EEG

P.008

Functional network reorganization in temporal lobe epilepsy: looking beyond the hippocampus

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Background: Temporal lobe epilepsy (TLE) has been redefined as a disorder associated with network-level dysfunction not limited to the epileptic zone. As such, as resting state (rs) fMRI has been used to evaluate the implicated resting state networks (RSN) and their ensuing functional impairments. However, few studies have analyzed patients with (TLE-HS) and without (TLE-nonHS) hippocampal sclerosis independently. Whereas TLE-HS often warrants surgical intervention, drug-resistant TLE-nonHS might pose challenges for diagnosis and treatment decisions. Methods: This study aimed to investigate functional connectivity changes (FC) of RSNs beyond the hippocampus using rs-fMRI. Rs-fMRI data was acquired from 16 TLE-HS and nine TLE-nonHS, along with 25 healthy controls (HC). RSNs were established using a data-driven independent component analysis approach, in order to determine significant connections between HC and patient groups ipsilateral and contralateral to the seizure focus. Results: When comparing TLE-HS to HC, FC changes were found for the dorsal-attentional (DAN), visual, fronto-parietal (FPN), sensorimotor and default-mode networks (DMN). Alterations in the DAN, DMN and FPN were found when comparing TLE-nonHS to HC. Conclusions: This study demonstrated widespread network reorganization across TLE subtypes. These FC patterns hold promise as a prognostic biomarker, and may be used to define subsequent function and dysfunction in this patient population.

P.009

Canadian Survey of the neurological care provided to women living with epilepsy: preliminary results

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Background: In Canada, approximately 300,000 women with epilepsy (WWE) are of childbearing potential. Given the unique aspects of providing care for WWE, our objective was to gather demographic and practice characteristics of health care professionals providing care for WWE to identify potential gaps. Methods: We developed a questionnaire to understand the demographic and practice characteristics of professionals providing care for WWE. We invited all French and English practitioners (physicians, physician assistants and nurse practitioners), recruited through the Canadian League Against Epilepsy (CLAE), Canadian Neurological

Sciences Federation (CNSF) and Canadian Epilepsy nursing Group (CENG), to complete our online questionnaire. Results: Preliminary data show 43% were between 32-40 years of age and 76% were medical doctors. Sixty-three percent had been in practice for less than 20 years; 81% considered themselves epilepsy specialists and 66% devoted their practice entirely to epilepsy patients and 78% practiced exclusively in academic centers. Conclusions: Our data shows providers involved in the care of women with epilepsy in Canada are predominantly academic experts in epilepsy. Potential gaps in care include mid-late career physicians, non-specialized health care practitioners, and community-based practices, as less likely to provide care for WWE. This snapshot may provide future insights to the unmet needs of WWE Canada.

P.010

Extreme delta brush in anti-NMDAR encephalitis correlates with poor functional outcome and death

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Background: The electroencephalography (EEG) pattern extreme delta brush (EDB) is felt to be highly specific for anti-N-methyl-D-aspartate receptor (NMDAR) encephalitis. This study aimed to characterize EEG findings in anti-NMDAR encephalitis patients looking for the proportion of abnormal EEGs, presence of EDB, and to relate EDB to clinical outcomes (Glasgow Outcomes Scale (GOS) at 6 months, need for ICU admission, and death). Methods: This retrospective single centre study included anti-NMDAR encephalitis patients who had ≥1 EEGs obtained from 2014-2021. EEGs were retrospectively analyzed by 2 reviewers. Clinical outcomes of interest were extracted through hospital and clinic chart review. Results: Twenty-one patients with anti-NMDAR encephalitis were included. Sixty-four EEGs were analyzed. Four EEGs (6.3%) were within normal limits. Focal or generalized slowing (without EDB) was seen on 44 EEGs (68.8%). EDB was seen on 16 EEGs (25.0%) in 9 of 21 patients. The presence of EDB was significantly associated with need for ICU admission (p=0.02), poorer outcome at 6 months as per the GOS (p=0.002), and with death (p=0.02) Conclusions: The presence of EDB on EEG in anti-NMDAR encephalitis patients is associated with increased need for ICU admission, risk of death, and worse functional outcomes at 6 months.

P.011

Piano Player Hand Sign: description of a novel clinical sign elicited by cortical electrical stimulation in epileptic patients

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Background: Cortical electrical stimulation (CES) may produce different motor responses according to the brain area stimulated. In this study, we describe a new motor response characterized by finger movements such as a person playing piano, which we named the Piano Player Hand (PPH) sign. Methods: We retrospectively reviewed the CES results of 252 patients with drug-resistant epilepsy who underwent SEEG between January 2005 and December 2019 at the Grenoble-Alpes University Hospital. The patients' characteristics, SEEG findings and CES parameters were extracted to find common clinical and anatomical features. Results: The PPH sign was identified 20 times from 12 patients, with stimulation of either the supplementary motor area (SMA), anterior cingulate gyrus (ACG), pre-SMA, middle frontal gyrus and anterior insula. It was obtained with high frequency stimulation, with intensity ranging from 0,7 to 3mA and mostly contralateral to the stimulation side (19/20). It was part of the ictal semiology of five patients. An afterdischarge was observed in five of the relevant CES. Conclusions: The PPH sign is a novel clinical sign, obtained mainly, but not exclusively, with CES of a small vicinity encompassing the SMA, pre-SMA and ACG. The PPH sign, when occurring ictally, may point to the premotor mesial frontal surface of the brain.

P.012

The new-onset refractory status epilepticus (NORSE/FIRES) family registry

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Background: New-onset refractory status epilepticus (NORSE) is a rare clinical presentation affecting previously healthy individuals. Febrile infection-related epilepsy syndrome (FIRES) is a subcategory of NORSE and applies when a preceding fever occurs. The NORSE/FIRES Family Registry has been developed to gain insight into risk factors and to assess the spectrum of clinical outcomes amongst survivors. Methods: Survivors, surrogates, and physicians can enter patient data into the REDCap-based registry: https://www.norseinstitute.org/norse-registry-2. Information collected includes medical history, clinical presentation, and quality of life, among others. Participants are invited to complete follow-up surveys for up to two years following presentation of seizures. Enrollment is ongoing in multiple languages. Results: 56 participants are enrolled from 12 countries (2-78 years, median: 12.5, IQR: 20.5, 31 survivors). At ≥6 months after onset, survivors experience a mean of ≥12 seizures per month and remain on a median of 4 (IQR: 3) anti-seizure medications. The median quality of life amongst all survivors was rated 4/10 (IQR: 3.5). Conclusions: Preliminary data suggests that survivors of NORSE/FIRES have a high seizure burden and poor quality of life. This international multi-lingual family registry will help develop hypotheses for future studies and provides an opportunity for families to contribute to the scientific understanding of this disease.

P.013

Stereo-encephalographic presurgical evaluation of temporal lobe epilepsy

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Background: Drug resistant epilepsy is present in nearly 30% of patients. Resection of the epileptogenic zone has been found to