There was also decreased connectivity between the anterior cingulate and right lateral occipital cortex, and between the left anterior insula to the cerebellum and precuneus cortex. Conclusions: The process of effort discounting is correlated to functional connectivity changes involving the precuneus, anterior cingulate, and left anterior insula in healthy older adults.

P.002

Saccade parameters reveal cognitive impairment and differentially associate with cognitive domains across neurodegenerative diseases

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Background: Eye movements reveal neurodegenerative disease processes due to overlap between oculomotor circuitry and disease-affected areas. Characterizing oculomotor behaviour in context of cognitive function may enhance disease diagnosis and monitoring. We therefore aimed to quantify cognitive impairment in neurodegenerative disease using saccade behaviour and neuropsychology. Methods: The Ontario Neurodegenerative Disease Research Initiative recruited individuals with neurodegenerative disease: one of Alzheimer's disease, mild cognitive impairment, amyotrophic lateral sclerosis, frontotemporal dementia, Parkinson's disease, or cerebrovascular disease. Patients (n=450, age 40-87) and healthy controls (n=149, age 42-87) completed a randomly interleaved pro- and anti-saccade task (IPAST) while their eyes were tracked. We explored the relationships of saccade parameters (e.g. task errors, reaction times) to one another and to cognitive domain-specific neuropsychological test scores (e.g. executive function, memory). Results: Task performance worsened with cognitive impairment across multiple diseases. Subsets of saccade parameters were interrelated and also differentially related to neuropsychology-based cognitive domain scores (e.g. antisaccade errors and reaction time associated with executive function). Conclusions: IPAST detects global cognitive impairment across neurodegenerative diseases. Subsets of parameters associate with one another, suggesting disparate underlying circuitry, and with different cognitive domains. This may have implications for use of IPAST as a cognitive screening tool in neurodegenerative disease.

P.003

CJD in the modern era: The value of clinical features and diagnostic tests

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Background: The advent of real-time quaking-induced conversion (RT-QuIC) assays has transformed the diagnostic approach to sporadic Creutzfeldt-Jakob disease (CJD) facilitating earlier recognition of affected patients. Recognizing this, we evaluated the performance of clinical features and diagnostic tests for CJD in the modern era. Methods: Clinical data were extracted from the electronic medical records of 115 patients with probable or definite CJD assessed at Mayo Clinic from 2014-2021. Clinical features and diagnostic tests were evaluated at presentation, and associations with diagnosis and prognosis determined. Results: Mean age-at-symptom onset was 64.8 ±9.4 years; 68 patients were female (59%). The sensitivity of clinical markers (myoclonus) and tests historically considered in patients with suspected CJD was poor (stereotyped EEG abnormalities, 16%; CSF 14-3-3, 60%). Conversely, RT-QuIC (93%), t-tau >1149 pg/mL (88%), and characteristic signal abnormalities on MRI (77%) identified most patients. Multivariable linear regression confirmed shorter days-to-death in patients with myoclonus (125.9, $CI_{95\%}$ 23.3-15.5, p=0.026), visual/cerebellar signs (180.19, $CI_{95\%}$ 282.2-78.2, p<0.001), positive 14-3-3 (193, $CI_{95\%}$ 304.9-82.9; p<0.001), and elevated t-tau (9.0, $CI_{95\%}$ 1.0-18.0, for every 1000 pg/ml elevation; p=0.041). Conclusions: CSF RT-QuIC and elevated t-tau, and stereotyped MRI abnormalities were consistently detected in CJD patients. Myoclonus, EEG findings, and CSF protein 14-3-3 were less useful in the modern era.

P.004

Dissecting the neuropathological causes of rapidly progressive dementia

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Background: A clear understanding of the neuropathological causes of RPD is needed to inform the diagnosis and treatment of patients with rapidly progressive dementia (RPD). Methods: Patients with <4.0 years from symptom onset to death were identified within the Mayo Clinic Neurodegenerative Brain Bank (1998-2020). Relevant clinical details were extracted from available records. Neuropathological diagnoses were assigned following standard protocols. Results: 310/8586 (3.6%) cases met RPD criteria. Relative to typically progressive cases, prion disease most commonly presented as RPD (74%, 32/43), followed by progressive supranuclear palsy/corticobasal degeneration (PSP/

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