using open database containing muscle MRIs Mercuri scores from 950 individuals. Results: AI and machine learning were applied on 10 types of NMD muscle MRI Mercuri scores that represented muscle involvement based on the degree of fatty infiltration. Different models were generated, the one with highest accuracy was used. When tested on new patients, it achieved a 90% accuracy. Subsequently, was turned into a mobile application. Conclusions: Muscle MRI is a valuable tool to help in NMD diagnosis. Specific muscle involvement pattern can be predictive. Besides, AI facilitates the interpretation and comprehension of muscle imagining in NMD.

C.3

Childhood Absence Epilepsy: Prevalence of neuropsychiatric comorbidities and predictors of treatment resistance

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Background: Seizures in childhood absence epilepsy (CAE) are typically easily controlled with anti-seizure medications (ASMs). Factors predictive of treatment resistance remain unclear. Our objectives were to assess prevalence of neuropsychiatric problems and factors influencing treatment resistance in a cohort of CAE at a single centre. Methods: We retrospectively reviewed patients with CAE diagnosed between January 1999 and December 2016 with at least 1-year follow-up. Treatment resistance was defined as failure to respond to two appropriate ASMs. Exclusion criteria included eyelid myoclonia with absence, myoclonic absence, and generalized tonic-clonic (GTC) seizures prior to absence seizures. Results: The study population comprised 164 patients (65 males) with mean age at seizures onset of 6.1years. 21% had treatment-resistant seizures. The first ASM was Ethosuximide (63.4%), Valproic acid (23.2%), and Lamotrigine (6.7%). 32.9% of children had learning disabilities, 28.7% ADHD, and 12.8 % anxiety.

A stepwise binary logistic regression analysis identified GTC seizures, learning disability (LD) and ongoing general spike and wave on EEG as predictors of treatment resistance. At last follow-up (average of 5.4 years), 43.3% of children were seizure-free off ASMs. Conclusions: 21% of children with CAE had treatment-resistant seizures. LD, GTC and failure of normalization of the EEG were associated with treatment resistance.

C.4

Distinct BOLD signal variability changes in temporal and occipital cortices in pediatric epilepsy

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Background: Greater variability of neuronal signalling, measured as the standard deviation of the blood oxygen dependent signal ($BOLD_{SD}$), relates to information processing capacity. Resting-state functional magnetic resonance imaging was used to determine differences in $BOLD_{SD}$ between children with and without epilepsy. Methods: We studied 24 controls (mean age 8.52 ± 1.35 years) and 18 patients (mean age 11.5 ± 3.4 years) with medically refractory epilepsy that underwent imaging for preoperative planning.Standard preprocessing steps (FSL v6.0, FMRIB) were followed and AAL atlas was used. Whole-brain two sample t-tests were used for group comparisons and significance was set at $p < 0.05$ FDR-corrected. Results: Children with epilepsy showed significantly lower $BOLD_{SD}$ in left inferior and middle temporal gyri ($p < 0.001$), right caudate nucleus ($p < 0.01$), cuneus ($p < 0.001$), and fusiform gyrus ($p < 0.001$), and significantly increased $BOLD_{SD}$ bilaterally in inferior occipital gyri ($p < 0.0001$). There were no significant differences when comparing whole-brain $BOLD_{SD}$ values. Conclusions: Neuroplastic changes in epilepsy may depend on an optimal amount of internal neural variability driven by the identified key regions. Certain temporal and occipital regions may underlie neural processing differences in children with epilepsy. Further studies may correlate these findings with behavioral testing.

POSTER PRESENTATIONS

Adult Neurology (CNS/CSC)

Dementia and Cognitive Disorders

P.001

The neural correlates of effort-reward decision-making in older adults

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Background: Effort mobilization is important in older adults to stay healthy, notably for decision-making. The process of decreasing subjective value of a reward as required effort increases is called effort discounting. By identifying underlying neural correlates related to effort discounting, we can better understand factors affecting normal cognitive aging. Methods: We acquired resting-state functional magnetic resonance images from 19 cognitively normal older adults (10 males; 66±6 years). Participants completed a computerized cognitive task—called Effort Expenditure for Rewards Task—capturing the willingness to expend effort for rewards through binary choices between high-reward-high-effort or low-reward-low-effort option to obtain varying monetary rewards. We modelled subjective value to assess the k parameter, effort discounting. A functional connectivity analysis examined the involvement of regions associated to the salience network. Results: The seed-to-voxel analysis revealed increased connectivity within the precuneus cortex and to clusters in the right temporal and posterior cingulate gyri, with increased k-value or decreased willingness to expend effort.

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

HC Riek (Kingston)* *BC Coe* (Kingston) *DC Brien* (Kingston) *J Huang* (Kingston) *A Abrahao* (Toronto) *S Arnott* (Toronto) *D Beaton* (Toronto) *M Binns* (Toronto) *S Black* (Toronto) *M Borrie* (London) *L Casaubon* (Toronto) *D Dowlatshahi* (Ottawa) *E Finger* (London) *C Fischer* (Toronto) *A Frank* (Ottawa) *M Freedman* (Toronto) *D Grimes* (Ottawa) *A Hassan* (Thunder Bay) *M Jog* (London) *S Kumar* (Toronto) *D Kwan* (Kingston) *A Lang* (Toronto) *J Lawrence Dewar* (Thunder Bay) *B Levine* (Toronto) *W Lou* (Toronto) *J Mandzia* (London) *C Marras* (Toronto) *M Masellis* (Toronto) *P McLaughlin* (Kingston) *J Orange* (London) *S Pasternak* (London) *A Peltsch* (Kingston) *B Pollock* (Toronto) *T Rajji* (Toronto) *A Roberts* (London) *D Sahlas* (Hamilton) *G Saposnik* (Toronto) *D Seitz* (Kingston) *C Shoesmith* (London) *T Steeves* (Toronto) *S Strother* (Toronto) *S Sujanthan* (Toronto) *K Sunderland* (Toronto) *R Swartz* (Toronto) *B Tan* (Toronto) *D Tang-Wai* (Toronto) *C Tartaglia* (Toronto) *A Troyer* (Toronto) *J Turnbull* (Hamilton) *L Zinman* (Toronto) *ONDRI Investigators (), DP Munoz* (Kingston)

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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/