migraines, but only 24% offered any support. Migraine had negative impacts on relationships.

Conclusion The Migraine Voice Survey highlights the true burden of migraine in Australia and limitations of current management options.

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GEMCITABINE-RELATED RADIATION RECALL AS A CAUSE OF FOCAL MYOSITIS AND MUSCLE NECROSIS

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Introduction Radiation recall is a phenomenon in which chemotherapy triggers an inflammatory response in tissue previously subjected to radiation therapy. A wide variety of agents have been implicated. Cutaneous tissue is most frequently affected but other tissue can be involved; myositis has been associated with administration of gemcitabine in particular. Incidence has been estimated at less than 6% and the pathophysiology is not understood. We present a case report from Gosford Hospital, with the additional feature of positive SRP antibodies.

Case A 74 year old female presented with a one day history of left hip pain and inability to weight bear. She had been diagnosed with metastatic squamous cell carcinoma of the lung five months earlier and underwent palliative radiotherapy to a left acetabular metastasis. 12 days prior to presentation she completed her second cycle of chemotherapy with carboplatin and gemcitabine. Pre- and post-contrast CT and MRI demonstrated necrosis in left sartorius, with foci of myositis in other muscles of the thigh, and surrounding soft tissue oedema. Symptoms improved after chemotherapy was ceased. Myositis antibody studies subsequently revealed low level positive Ku and SRP antibodies.

Conclusion Radiation recall should be considered in the differential diagnosis of myositis in oncology patients. The serum of our patient contained SRP antibodies, which are associated with immune mediated necrotising myopathy. A previous case study reported gemcitabine-induced radiation recall muscle necrosis associated with dermatomyositis. These findings hint that radiation recall myositis may occur in the setting of a predisposition to immune mediated myopathy.

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CORRELATING STRUCTURE AND FUNCTION TO BETTER IDENTIFY SURROGATE END POINTS FOR CLINICAL TRIAL DESIGN: A LONGITUDINAL CLINICAL AND IMAGING STUDY OF PRIMARY PROGRESSIVE APHASIA

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Introduction Measuring longitudinal change in white matter tracts offers a highly sensitive way of monitoring the course of a range of neurodegenerative conditions. However, it remains unclear how structural changes correlate with symptom progression. Clinically meaningful outcomes remain a key requirement in therapeutic trial design so

imaging biomarkers need to accurately predict these outcomes. Identifying surrogate clinical end points is of particular importance in neurodegenerative conditions where clinical change evolves slowly. To address this the current study aims to identify potential surrogate end points by assessing correlations between clinical and neuroimaging measures.

Methods 30 patients meeting consensus criteria for a diagnosis of primary progressive aphasia underwent longitudinal imaging and neuropsychological assessments at baseline and one year. A mixed effects model was designed to test for significant interactions over time between changes in neuropsychological performance and Fractional Anisotropy (FA) in key white matter tracts.

Results Declining single word comprehension correlated with reducing FA within bilateral inferior longitudinal fasciculus (ILF), bilateral superior longitudinal fasciculus (SLF) and the genu of the corpus callosum; declining naming ability correlated with reducing FA in the left ILF, right uncinate fasciculus and right SLF; declining word repetition correlated with reducing FA within the left ILF.

Conclusions Declining neuropsychological scores correlated with longitudinal decline in FA in a number of white matter tracts across an anatomically distributed language network. Correlations between function and structure provide evidence that monitoring structural white matter changes in the tracks identified may have value as a surrogate end point for future clinical trials.

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CHARACTERISING SLEEP AND FATIGUE IN PATIENTS WITH PRIMARY MITOCHONDRIAL DISEASE

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Introduction Fatigue is common in patients with primary mitochondrial disease (PMD). There has been little prospective research into sleep pathology in these patients and assessment of contributory factors to fatigue.

Methods Patients with PMD were prospectively assessed with overnight polysomnography in addition to measures of fatigue, muscle fatigability, disease severity, sleep propensity and depression.

Results 16 patients participated, 15 completing inpatient polysomnography. Obstructive sleep apnoea (OSA) was common (53%), affecting 5/9 females (56%) and 3/6 males (50%), although most cases were mild in severity. There was a trend to higher incidence of OSA in older patients but not other traditional risk factors, nor presence of myopathy. The Epworth Sleepiness Scale (ESS) was the best predictor of OSA, although not reaching significance.

Fatigue was common, with 81% of patients having significant fatigue on the Fatigue Severity score and 69% on the Fatigue Impact Scale. The two scores correlated well ($r = 0.85$, $p = 0.01$).

The mean 5 times sit to stand time of 16 seconds was significantly higher than community normals ($p = 0.04$), demonstrating muscle fatigability, not isolated to patients with myopathy.

Conclusion OSA is common in patients with PMD, although mostly mild in severity. ESS likely remains a relevant tool in screening patients when considering overnight polysomnography to investigate fatigue in these patients. OSA does not explain the high prevalence of fatigue, with other factors including depressive symptoms, important considerations.

126 INCIDENCE OF DEMENTIA IN ELDERLY PATIENTS WITH EPILEPSY

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Introduction The incidence of epilepsy is higher in patients with underlying dementia.¹ The goal of the present study is to look at the incidence of dementia in patients presented to our epilepsy clinic and analyse electroencephalogram (EEG), imaging findings and response to antiepileptic drug (AED) in these individuals.

Methods A retrospective study was performed on patients presented to Nepean Hospital epilepsy clinic from 2015 to 2017. Multiple clinical parameters were obtained from electronic medical records.

Results A total of 258 patients presented to the clinic, of which 38 patients were above the age of 65 years. 11 patients were excluded due to insufficient information or patients without any history of seizures. Out of the remaining 27 patients studied, nine patients (33%) had dementia including five patients (19%) with Alzheimer's dementia. Sixteen patients (59%) experienced complex partial seizures. Brain MRI was performed in twenty one patients (78%). Sixteen patients (59%) had MRI-identified structural lesions including prior stroke or intracerebral haemorrhage. EEGs were performed in twenty patients (74%). Eight patients (40%) had abnormal EEG with one patient (5%) having epileptiform discharges, three patients (15%) having focal slowing and four patients (20%) having generalised slowing. Overall, nineteen patients (70%) were on AEDs with good control and four patients (15%) required more than one AED to achieve seizure control.

Conclusion The study showed that there is higher incidence of dementia in patients with epilepsy compared with general population.² Most patients experience complex partial seizures and can be adequately controlled on single AED.

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128 LATE-ONSET POMPE DISEASE (LOPD) PRESENTING WITH FULMINANT HYPERCAPNIC RESPIRATORY FAILURE

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Introduction We report a case of LOPD with acute-on-chronic respiratory failure.

Case A 57 year-old retired farmer presented with obtundation requiring intubation. He reported a 4 month history of hypophonia, intermittent diplopia, lethargy and orthopnea.

Initial arterial blood gas measurement displayed acute-on-chronic hypercapnic respiratory failure (pH 7.19, pO₂ 98 mmHg, pCO₂ 112 mmHg, HCO₃ 43 mmol/L). Muscle biopsy was suggestive of LOPD with myofibres demonstrating acid phosphatase and periodic acid-schiff positive vacuoles. Diagnosis was confirmed with low α -glucosidase activity on dried blood spot (0.4 μ mol/h/L) and elevated urinary tetrasaccharide level (5 mmol/mol creatinine). Mutation analysis of the GAA gene demonstrated two known pathogenic mutations (c.-32-13T>G and c.1075+1G>T). With improved ventilation, he was able to be extubated. The only respiratory support on discharge was overnight bilevel positive airway pressure ventilation.

Conclusion LOPD is a rare autosomal recessive metabolic disorder caused by a deficiency in acid α -glucosidase. This leads to intra-lysosomal accumulation of glycogen in tissues. Particularly in the late form, there is significant phenotypic variability.¹ Diagnosis remains challenging. Cases have been reported with a range of initial symptoms including stroke,² syncope³ and chronic respiratory failure.⁴ Acute on chronic respiratory failure at presentation is rare.

Enzyme replacement therapy has been shown to improve both morbidity and mortality in LOPD.⁵ Earlier treatment is associated with better outcomes.⁶ Prompt recognition of cases is paramount. Unexplained acute-on-chronic respiratory failure should raise the possibility of this condition. In such cases, management of ventilation is vital.

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