11 QUANTITATIVE MUSCLE ULTRASOUND: A DIAGNOSTIC BIOMARKER IN AMYOTROPHIC LATERAL SCLEROSIS

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Objectives We prospectively assessed the diagnostic utility of quantitative muscle ultrasound to distinguish amyotrophic lateral sclerosis (ALS) patients from neuromuscular mimics. Additionally, we explored the association between ultrasound parameters and clinical markers of disease severity.
Methods 46 patients suspected of ALS were studied using ultrasound of 17 different muscle groups. Muscle fasciculations and quantitative muscle echo-intensity were recorded. Muscle specific echo-intensity reference values were generated from 39 healthy controls, with hyperechoic muscles defined as a Z score >1.5 SD. The diagnostic utility of these metrics and their association with disease severity was determined.
Results 34 patients met the diagnosis of ALS over the follow up period. 12 patients were diagnosed with other neuromuscular disorders. Longer ALS disease duration was associated with more hyperechoic muscles ($r=0.44$ $p=0.003$) and less fasciculating muscles ($r=0.35$ $p<0.001$). ALS subjects had significantly more fasciculating muscles when compared to mimic subjects (mean 7.9 ± 3.9 vs 1.9 ± 2.8 $p<0.01$). By contrast, the number of hyperechoic muscles was not significantly different between groups (mean 2.0 ± 0.3 vs 3.1 ± 0.9 $p=0.167$). In the diagnosis of ALS $\geq$ 5 fasciculating muscles was 82% sensitive and 87.5% specific (AUC 0.88) while a total fasciculation count of $\geq$7 was 82% sensitive and 75% specific (AUC = 0.86). The ratio of fasciculating muscles to hyperechoic muscles was calculated. A ratio of $\geq$ 1.5 was 87% sensitive and 89% specific (AUC 0.94) for ALS.
Conclusions The ratio of fasciculating muscles to hyperechoic muscles on ultrasound is highly accurate for the diagnosis of ALS.

12 THE IMPACT OF THE COVID-19 PANDEMIC IN PATIENTS WITH MYASTHENIA GRAVIS; AN AUSTRALIAN PERSPECTIVE

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Objectives Previous studies have demonstrated high morbidity and mortality in patients with myasthenia gravis (MG) who acquired COVID-19. We aim to provide an up to date perspective of the various impacts of COVID-19 in our population.
Methods A prospective observational cohort study was conducted to identify the impact of the COVID-19 pandemic in Australian patients with MG. We conducted an online survey through Myasthenia Alliance Australia from May 2022 to July 2022, focusing on the impact of the pandemic on MG disease control, treatment, quality of life and the mental health of patients.
Results Among the 229 patients who responded to the survey, 221 (96.5%) had received at least two doses of the COVID-19 vaccines, 65 (28.4%) had contracted COVID-19, with 7 patients (10.8%) requiring hospitalisation and one patient (1.5%) requiring ICU admission. A large proportion of patients responded that the pandemic had no impact on their MG disease control (123; 54%) or treatment (111; 48%). Most patients (128; 76.2%) felt the pandemic had at least a mild impact on their mood.
Conclusion Our study provides a snapshot of the types of impacts experienced by myasthenic patients during the pandemic. In addition to a hospitalisation rate of 10.8%, our cohort self-reported substantial psychological impacts, which was independent of acquiring COVID.

13 AUTOLOGOUS HAEMATOPOIETIC STEM CELL TRANSPLANTATION FOR NON-MS NEUROIMMUNOLOGICAL DISORDERS: A SINGLE CENTRE EXPERIENCE

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Objectives Autoimmunity affects the C/PNS in a range of heterogenous diseases. Whilst most patients respond to standard treatments, responses may be inadequate leading to significant and potentially permanent disability. Autologous haematopoietic stem cell transplantation (AHSCT) has been reported as a means of intensive immunomodulation. Here, we evaluate the safety and efficacy of AHSCT in patients with non-MS neuroimmunological disease.
Methods A Phase II clinical trial of AHSCT using a cyclophosphamide + ATG chemotherapeutic regimen for patients with treatment-refractory neuroimmunological disease commenced at St Vincent's Hospital in December 2010 (ACTRN12613000339752. Eligibility criteria for each disease are available via ANZCTR.
Results 12 patients underwent AHSCT between May 2011 and September 2021 for the following; Bechet's disease (n=1), CIDP (n=3), SLE/CNS vasculitis (n=4), opsoclonus myoclonus ataxia (n=1), stiff-person syndrome (n=2), neurosarcoidosis (n=1). Median follow-up is 39 months (19–67 months). Treatment related mortality was 0%. Adverse effects due to AHSCT were consistent with expected toxicities. A median of 7 (range = 5–11) prior disease-modifying therapies were trialled prior to AHSCT. Ten patients remained off immunotherapy following AHSCT. Quality of life metrics improved for the majority of patients transiently post-AHSCT but only remained below baseline in 5 of 12 patients at last follow up.
Conclusions Clinical evidence to support the use of AHSCT in treatment-refractory neuroimmunological disease is scant. This study represents a summary of the experience of the largest Australian autoimmune disease transplant unit, shedding light on which conditions and patient phenotype may be more likely to benefit from AHSCT.