P.038

The incidence and characteristics of chronic pain and fatigue after 12 months later admitting with COVID-19; The Post-COVID 19 syndrome

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Background: This study aimed to evaluate chronic pain and fatigue in COVID-19 patients after 12 months of hospitalization. Methods: We studied the COVID-19 patients discharged from Hospital, between March 10 to April 20, 2020. Results: A total of 157 patients were included in this study. Forty-three patients (27.4%) complained of chronic fatigue and muscle weakness in the last six months. The visual analog fatigue scale (VAFS) score of 3.84 ± 1.48 was obtained. Forty patients (25.5%) were suspected of Chronic Fatigue Syndrome (CFS). Twenty-four patients (15.3%) had severe chronic pain or exacerbation of previous chronic pain, most of which were reported in the lower back (70.8%) and lower extremities (66.7%). Pain intensity had a mean score of 2.33 ± 0.87 and was mainly described as "muscle cramps," "persistent dull pain," and "boring and numbing." In women, chronic pain and fatigue, extended hospital stays, ICU admission, and depressed mood were common than in men. For these pain and fatigue, 37% used nonsteroidal anti-inflammatory drugs, and 16.3% used antidepressants. Only one person had applied for physiotherapy, and none of the patients had received psychotherapy Conclusions: Fatigue and chronic pain in patients recovering from COVID-19 are common complications, even after 12 months of illness.

Neuromuscular Disease and EMG

P.040

Value-based approach to the management of Inflammatory Neuropathies: Incorporating objective outcome measures in clinical care

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Background: Measuring outcomes that matter to patients is a key component of ensuring patient-centred care. In Chronic Inflammatory Neuropathies (CINs), where immunomodulatory treatments have risks and high costs, systematic evaluation of disease progression is needed to ensure patients are achieving outcomes that reflect their values and goals. The aim of this project is to evaluate the feasibility of objective outcome measure (OOM) use in the clinical setting. Methods: Prospective data was collected from 27 participants with CIDP or MMN. Participants completed and provided feedback on patient-reported outcome measures including quality of life, activity and participation, pain and fatigue, as well as grip strength, 9-hole peg test, 10 meter walk, muscle strength and sensation. Focus groups were conducted to collect qualitative data. Results: The majority of OOMs were considered relevant to 90% of participants. The top three ranked measures were muscle strength testing, daily activities questionnaire and quality of life questionnaire. 52% of participants identified balance and/or detailed gait assessment as an important factor that was not part of collected OOMs. Conclusions: OOMs allow for appropriate monitoring of patients and optimization of immunotherapy treatment. By tracking longitudinal results that matter to patients, patients can better participate in shared-decision making. Clinicians should adopt OOMs going forward.

P.041

Characteristics of carpal tunnel syndrome in wild-type transthyretin amyloidosis

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Background: Wild-type transthyretin amyloidosis (wtATTR) is an important cause of infiltrative cardiomyopathy in older adults. Carpal tunnel syndrome (CTS) is one of the most common extra-cardiac manifestations of wtATTR; however, the prevalence, severity, and risk of recurrence following carpal tunnel release (CTR) remain poorly understood. Methods: This retrospective cohort study reports findings from a single-centre experience of routine neurological screening of newly diagnosed wtATTR patients including nerve conduction studies. Consecutive wtATTR patients between 2014 and 2021 were included. Results: Seventy-nine wtATTR patients were included, 73 (92%) males, mean age of 79 years. Seventy-four (94%) had median neuropathy at the wrist (MNW), 50% having a prior diagnosis with the remaining 50% being diagnosed at screening. The majority with MNW were symptomatic (53, 67%) with moderate or severe disease (66, 84%) bilaterally (42, 53%) on electrophysiologic testing. Nineteen (24%) had recurrent CTS despite previous CTR. At the time of screening, 19 (24%) were prescribed wrist splinting and 36 (46%) were referred for CTR. Conclusions: Carpal tunnel syndrome is common in wtATTR. Most have bilateral disease with moderate to severe MNW at the time of wtATTR diagnosis. Recurrence of CTS after CTR is more common in wtATTR patients than in the general population.

P.042

A novel SOD1 mutation associated with rapidly evolving lower motor neuron syndrome and MR ventral nerve root enhancement

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Background: Mutations in the Cu/Zn superoxide dismutase 1 (SOD 1) gene are estimated to cause 20% of familial ALS and 1-2% of sporadic cases. Accurate gene variant classification of novel mutations in amyotrophic lateral sclerosis (ALS) has deepened our understanding of clinical phenotypes, provided pathologic insights, and is crucial to incorporating emerging

therapies. Methods: We describe a case of a 75-year-old female who presented with a rapidly progressive lower motor neuron syndrome leading to flaccid quadriparesis and complete loss of independence over a five month period. Results: Genetic testing demonstrated a heterozygous variant of uncertain significance in the SOD1 gene with a g > c point mutation at position 382 that has been described in one other patient in available literature. MR of the lumbar spine demonstrated abnormal smooth nerve root enhancement. Conclusions: This novel mutation in the SOD1 gene may be associated with a rapidly progressive phenotype of sporadic ALS. Ventral nerve root enhancement should not exclude a diagnosis of ALS especially in the absence of nodularity or nerve enlargement.

P.043

Plasmapheresis for treatment of light chain amyloidosis related myopathy

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Background: Light chain (AL) amyloidosis is a plasma cell disorder characterized by abnormal fibrillary light chain deposition causing cardiac, renal, hepatic, gastrointestinal and peripheral nervous system dysfunction. Muscle disease occurs in 1.5% of individuals causing progressive proximal weakness thus far considered untreatable. Methods: We reviewed two cases of AL amyloidosis associated myopathy at our institution who had robust response to plasmapheresis. Both were at stringent clinical response following CyBorME therapy during peak severity of their myopathy. Results: In case 1, a 70-year-old male with recently diagnosed kappa light chain multiple myeloma and cardiac/renal amyloidosis developed severe subacute proximal weakness preventing ambulation. CK was normal and electromyography was consistent with irritable myopathy. Deltoid biopsy showed perimysial and endomysial amyloidosis. A trial of plasmapheresis in a tapering schedule resulted in robust recovery of strength. In case 2, a 67-year-old female with recently diagnosed kappa light chain multiple myeloma with amyloidosis on fat pad aspirate developed severe subacute proximal weakness requiring prolonged hospital admission. CK was normal and electromyography demonstrated non-irritable myopathy. Bicep biopsy showed perivascular amyloidosis. A trial of plasmapheresis in a tapering schedule resulted in robust recovery of strength. Conclusions: Plasmapheresis is a novel and potentially effective treatment for patients with AL amyloidosis associated myopathy.

P.044

GMPPB mutation causes a muscular dystrophy-myasthenic spectrum

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Background: Mutations in GDP-Mannose Pyrophosphorylase B (GMPPB) cause a spectrum of disease ranging from muscular dystrophy to congenital myasthenic syndrome (CMS). Recognition of neuromuscular junction dysfunction has important treatment implications. Methods: We describe a person with GMPPB mutation causing an overlapping limb girdle muscular dystrophy - myasthenic syndrome with robust response to acetylcholinesterase inhibitors. We review the literature on the muscular dystrophy - CMS and explore the phenotypic features that aid in recognizing neuromuscular junction dysfunction. Results: A 35-year-old woman presented with a 10-year history of debilitating myalgias, symmetrical limb girdle and neck weakness, and chronic CK elevation. Electromyography showed a non-irritable myopathy. Biopsies were consistent with muscular dystrophy. Whole genome sequencing revealed two heterozygous pathogenic mutations in the GMPPB gene, giving a diagnosis of genetically confirmed limb girdle muscular dystrophy. Subsequently, repetitive nerve stimulation revealed decrement in the trapezius muscle suggestive of an overlap myasthenic syndrome. She was started on pyridostigmine resulting in recovery of full motor power with significant functional improvement. Conclusions: Identification and treatment of neuromuscular junction dysfunction caused by GMPPB mutations can significantly improve motor power and function. Early onset of progressive fatigable proximal weakness, spared ocular and facial muscles, and pyridostigmine responsiveness are important features of GMPPB-related CMS.

P.045

Biopsies of the transverse carpal ligament and tenosynovium for tissue confirmation of transthyretin amyloidosis

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Background: Transthyretin Amyloidosis (ATTR) is a common cause of both cardiomyopathy and carpal tunnel syndrome, with many patients needing carpal tunnel release (CTR). Although tafamidis is now an approved treatment of ATTR cardiomyopathy, insurers in most provinces require biopsy confirmation of amyloidosis. Endomyocardial biopsy is often the chosen approach due to optimal sensitivity, albeit with risk of serious adverse events such as stroke, cardiac tamponade, and arrhythmias. CTR may present an ideal opportunity for obtaining amyloidosis biopsy confirmation. Methods: ATTR patients undergoing CTR had biopsy of their transverse carpal ligament (TCL) and/or flexor tenosynovium to assess the sensitivity of both sites for biopsy confirmation of amyloidosis. Results: Twelve patients consecutively underwent biopsies during CTR, with 4 (33%) having bilateral CTR and biopsy. Among 16 TCL biopsies and 14 tenosynovium biopsies, 100% demonstrated amyloid deposition. Another patient had isolated tenosynovium biopsy without CTR and also demonstrated amyloidosis. There were no serious adverse events, and 1/13 (8%) had wound dehiscence requiring repeat suturing. Conclusions: Biopsy of the TCL and/or tenosynovium during CTR is safe, cost-effective, and sensitive, and may represent an alternative to endomyocardial biopsy in patients requiring tissue confirmation for tafamidis approval. ATTR patients eligible for tafamidis may benefit from early neurology assessment.