

inflammatory demyelinating polyneuropathy (CIDP). While reports suggest an acute onset is more likely than in antibody negative CIDP, little literature exists around the subsequent course of NF-155 positive cases that originally presented with an acute inflammatory demyelinating polyneuropathy (AIDP) phenotype. Methods: Two male patients, ages 51 and 59, presented with similar, <2 week histories of lower extremity weakness. Patients were diagnosed with AIDP and treated with IVIG. Following initial improvement, both patients relapsed. One patient was treated with IVIG and steroids with subsequent improvement; however, he was unable to be weaned from steroids without experiencing recurrence of symptoms. The other patient was not retreated. Testing for NF-155 IgG was sent. Results: The first patient ultimately required Rituximab for stable improvement, the other improved spontaneously. Both patients later had positive tests for NF-155 IgG4 antibodies. Conclusions: Both of our NF-155 positive cases had initial AIDP-like presentations, followed by a relapsing course and excellent eventual recovery. This result, along with limited other available cases, suggest that in patients with an AIDP-like presentation, NF-155 IgG4 auto-antibodies could be a marker of disease recurrence, but do not necessarily predict a poor outcome.

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Idiopathic inflammatory myopathies and malignancy screening: a survey of the current practices amongst Canadian neurologists and rheumatologists

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Background: There is a well-established association between idiopathic inflammatory myopathies (IIM) and malignancy. There are no evidence-based guidelines amongst neurologists and rheumatologists on the choice and timing of malignancy investigations. Our aim is to characterize the current gaps and uncertainties amongst neurologists and rheumatologists with malignancy screening in IIM patients. Methods: An online survey consisting of 18 multiple-choice questions related to IIM malignancy screening was distributed to adult neurologists and rheumatologists in Canada. Quantitative and descriptive analysis was performed. Results: The majority of respondents (96%, n=68) performed malignancy screening. There was variability in practice including delegation and choice of screening tests, influence of patient-specific factors, and time and length of repeat testing. Only 18% of respondents were confident in their malignancy screening practices. Between neurologists and rheumatologists, there were differences in the number of IIM patients seen, consideration of patient-specific factors and choice of screening investigations. Further details and data will be presented at the conference. Conclusions: There is a lack of consensus and confidence in the choice and timing of malignancy investigations in IIM, with neurologists and rheumatologists differing in their approaches. Further research is required to better understand the relationship between IIM and malignancy to create expert-led consensus guidelines.

P.039

Development of a checklist for treating adults with Myotonic Dystrophy Type 1: a neuromuscular disease network for Canada (NMD4C) Knowledge Translation Tool

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Background: The Neuromuscular Disease Network for Canada (NMD4C) aims to improve the care of Canadians with neuromuscular diseases. It has identified a need to support clinicians in implementing clinical guidelines with the use of checklists for initial evaluation and clinical follow-ups. The objective of the study was to develop a pragmatic management checklist to support clinical guidelines for diagnosis and follow-up of myotonic dystrophy type 1 (DM1). Methods: A practice-based DM1 checklist will be reviewed by a panel of 35 experts using an online survey. The survey has been drafted using the Appraisal of Guidelines Research and Evaluation tool for assessing Recommendation Excellence (AGREE-REX). The experts will rate: (1) the quality of each checklist recommendation, and (2) the applicability of each recommendation based on their clinical setting. Scores will be compiled and discussed among experts to achieve consensus. Results: The compiled checklist items were organized into three sections: (1) initial evaluation, (2) follow-up visit and (3) general treatment recommendations. Feedback from experts across Canada, results on feasibility, and a finalized checklist will be presented. Conclusions: The development of a feasible treatment checklist is a useful KT tool that DM1 experts across Canada could apply in their own clinical settings.

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Long-term efficacy and safety of ravulizumab in adults with anti-acetylcholine receptor antibody-positive generalized myasthenia gravis

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Background: The 26-week double-blind, randomized, placebo-controlled period (RCP) of the CHAMPION MG study (NCT03920293) demonstrated ravulizumab's efficacy and tolerability in anti-acetylcholine receptor antibody-positive (AChR Ab+) generalized myasthenia gravis (gMG). Methods: In the ongoing open-label extension (OLE), patients receive intravenous ravulizumab (blind loading dose in placebo-treated patients or bridging dose in ravulizumab-treated patients, then 3000–3600 mg according to body weight every 8 weeks) for ≤4 years. Data from RCP baseline up to Week 60 were analyzed. Results: Ravulizumab's long-term efficacy (n=161) and safety (n=169) were assessed. Patients who switched from placebo in the RCP to