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 autoantibodies could be a marker of disease recurrence, but do not necessarily predict a poor outcome.

P.038

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.

P.040

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

ravulizumab in the OLE (n=83) showed rapid improvement (least squares mean, 95%CI) in Myasthenia Gravis-Activity of Daily Living (MG-ADL) and Quantitative Myasthenia Gravis (QMG) total scores, which were maintained through 34 weeks (MG-ADL: -1.7, -2.7 to -0.8; QMG: -3.1, -4.2 to -1.9). Improvements achieved by ravulizumab-treated patients (n=78) in the RCP were sustained through 60 weeks (MG-ADL: -4.0, -4.8 to -3.1; QMG: -4.1, -5.4 to -2.9). Ravulizumab was well tolerated; no meningococcal infections were reported. Four deaths unrelated to study treatment occurred. Conclusions: Ravulizumab demonstrated sustained improvements in MG symptoms and was well tolerated for up to 60 weeks in adults with AChR Ab+ gMG.

P.041

Guillain Barre syndrome could be a rare presenting finding of nodal and paranodal autoantibodies in immune-mediated neuropathies (IMN): A clinical utility of Cell based Assay

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Background: Guillain Barre Syndrome (GBS) and chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) are the two most common forms of treatable IMNs. Antibodies targeting proteins at paranodal cell-adhesion molecules such as contactin-1 (CNTN1), neurofascin-155 (NF155), contactin-associated protein 1 (CASPR1), and nodal neurofascins-NF140 and NF186, have been discovered in CIDP patients. Methods: Between August 2021 and January 2023, at BC Neuroimmunology laboratory, Vancouver we screened a total of 214 sera of patients for detecting nodal and paranodal antibodies with a fixed CBA. These patient sera were assayed for the presence of NF140, NF155, NF186, CNTN1, plus Caspr1 antibodies. The final diagnosis and response to therapy of positive cases were evaluated by a questionnaire requested from their physicians. Results: 10 cases were positive for nodal/paranodal antibodies by CBA (mean age 52.4 ± 15.4 years). Two cases were NF155 Ab positive CIDP with good response to conventional therapies. Three cases were double positive for NF140 and 186 Abs, three were double positive for CNTN1 and CASPR1 Abs. Interestingly, two cases were triple positive with GBS presentation. Conclusions: We identified a subgroup of nine patients with CIDP nodal and paranodal antibodies. Among them, two cases had triple positive antibodies with GBS presentation and poor response to plasma exchange and IVIg.

P.042

Could Live Cell-Based Assay increase the acetylcholine receptor autoantibodies seropositivity in patients with clinical suspicion of myasthenia gravis?

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Background: AChR antibodies (Abs) in Myasthnia Gravis (MG) are detected in approximately 50% of ocular and 85% of generalized MG by the current gold standard radioimmunoprecipitation assay (RIPA). Recently, fixed and lived Cell-Based assays (L-CBA) are developed. We clinically validated our in-house L-CBA in detecting AChR Ab in clinically suspected MG patients. Methods: Between January 2020 and April 2022, we assayed 10167 sera for AChR Ab by RIPA. We also assayed 4349 of AChR Ab seronegative sera of the above suspected MG samples for anti-MuSK Ab by RIPA. Then 1228 sera of double seronegative and/or borderline AChR Ab was assessed by L-CBA for AChR Ab. For clinical validation, we obtained clinical information on 36 seropositive cases for AChR Ab by L-CBA. Results: We found additional eighty-four cases seropositive for AChR Ab by L-CBA. The clinical information was obtained for 36 cases and based on their final diagnosis, twenty had generalized MG, thirteen had ocular MG, 2 not yet diagnosed and 1 case was of not-MG. Conclusions: The L- CBA has demonstrated improved sensitivity and higher diagnostics performance than RIPA. The L-CBA allowed improved clinical diagnosis and increased seropositivity (by 7%) in clinically suspected MG patients who were earlier seronegative/borderline for AChR Ab by RIPA.

P.043

Long-term safety, tolerability, and efficacy of efgartigimod in patients with Generalized Myasthenia Gravis: concluding analyses from ADAPT+

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Background: Efgartigimod is a human IgG1 antibody Fc-fragment that reduces total and pathogenic IgG autoantibody levels through FcRn blockade. ADAPT was a phase 3 trial evaluating efgartigimod in patients with generalized myasthenia gravis (gMG). Patients who completed ADAPT could enroll in ADAPT+ (open-label extension). Methods: Efgartigimod (10 mg/kg intravenous) was administered in cycles of 4 weekly infusions, with subsequent cycles initiated based on clinical evaluation. ADAPT+ evaluated long-term safety and tolerability of efgartigimod in patients with gMG. Efficacy was assessed utilizing MG-ADL and QMG scores. Results: Of 167 patients from ADAPT, 151 (90%) entered ADAPT+, and 145 received ≥1 cycle as of January 2022. Over 217.55 patient-years of follow-up (mean duration per patient, 548 days), incidence of adverse events did not increase with subsequent cycles. AChR-Ab+ patients with ≥1 year of follow-up across ADAPT/ADAPT+ (n=95) received a median (range) 5.0 (0.4–7.6) cycles per year. All AChR-Ab+ patients (n=111) demonstrated consistent improvements (mean change [SE], week 3 of cycle 1) in MG-ADL (-5.0 [0.33]; up to 14 cycles) and QMG (-4.7 [0.41]; up to 7 cycles) scores during each cycle. Conclusions: These ADAPT+ analyses suggest long-term efgartigimod treatment is well tolerated and efficacious. Additional final data cut analyses will be presented at CNSF 2023.