tremor. The tapping test may not reliably distinguish between PD tremor and functional tremor.

MS / Neuroinflammatory Disease

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Utility of amyotrophic lateral sclerosis functional rating scale (ALSFRS) bulbar subscores for predicting need for gastrostomy tube

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Background: We evaluated the utility of the Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS) in predicting risk of gastrostomy tube (G-tube) insertion in patients with ALS. Methods: We conducted a retrospective study using the Pooled Resource Open-Access ALS Clinical Trials Database. People with ALS, at least two ALSFRS scores, and baseline swallowing subscore >1 were included. G-tube outcome was defined as reaching a swallowing subscore ≤1. Predictors were ALSFRS bulbar subscores (swallowing, speech, salivation). Survival analyses estimated median time to outcome and cumulative probability of outcome within 91 days. Individuals were censored at last ALSFRS score. Results: We included 6,943 participants. Median [95% CI] time to G-tube insertion was 245 [228, 285], 562 [547, 621], and 1,268 [980, 1,926] for baseline swallowing subscores of 2, 3, and 4, respectively. Probability of G-tube insertion was associated with baseline swallowing, speech, and salivation subscores (log-rank test $p < 0.0001$). For patients who transitioned to a swallowing subscore of 2 or 3, 18.1% [95% CI 16.1, 20.3] and 1.9% [95% CI 1.3, 2.7] required G-tube insertion within 91 days of score transition. Conclusions: ALSFRS bulbar subscores may identify patients at risk of G-tube insertion. Probability of G-tube insertion within 91 days is low if swallowing subscore ≥3.

Neuromuscular Disease and EMG

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Concomitant corticosteroid use in ravulizumab-treated adults with anti-AChR antibody-positive gMG: results from the CHAMPION MG open-label extension

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Background: Treatment of generalized myasthenia gravis (gMG) with reduced steroid dosages may minimize steroid-associated AEs. Corticosteroid dosage changes were not permitted during the 26-week, CHAMPION MG study of ravulizumab in adults with anti-acetylcholine receptor antibody-positive (AChRAb+) gMG. Participants who completed the study could receive ravulizumab in the open-label extension (OLE; NCT03920293); corticosteroid adjustments were permitted. Methods: Patients could receive intravenous ravulizumab (blind induction or bridging dose at Week 26 [OLE start] for those previously receiving placebo or ravulizumab, respectively, then 3000–3600 mg at Week 28 and every 8 weeks thereafter) for ≤4 years. Results: Among 161 patients (78 ravulizumab, 83 placebo) who entered the OLE and received ravulizumab for ≤164 weeks, 113 received oral or enteral corticosteroids during the OLE; the proportion treated with >10 mg/day corticosteroids decreased from 58% (n=66) at first OLE dose to 37% (n=42) (35 [31%] received ≤5 mg/day and 71 [63%] received ≤10 mg/day) at last reported dose. Fourteen patients (12%) discontinued corticosteroids. The mean (SD) corticosteroid dosage/patient decreased from 17.5 (11.9) mg/day at first OLE dose to 11.7 (10.9) mg/day at last assessment. Conclusions: Ravulizumab decreased corticosteroid use in patients with AChRAb+ gMG, suggesting a steroid-sparing role for ravulizumab.

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Long-term safety and efficacy of zilucoplan in myasthenia gravis: additional interim analyses of RAISE-XT

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Background: Zilucoplan, a macrocyclic peptide complement component 5 inhibitor, sustained efficacy for up to 60 weeks of treatment, with a favourable safety profile in patients with acetylcholine receptor autoantibody-positive generalised myasthenia gravis in an interim analysis of RAISE-XT (NCT04225871). We evaluate the safety and efficacy of zilucoplan up to 96 weeks. Methods: RAISE-XT, a Phase 3, multicentre, open-label extension study, included patients who participated in the double-blind Phase 2 (NCT03315130) and Phase 3 (NCT04115293) zilucoplan studies. Patients self-administered daily subcutaneous zilucoplan 0.3mg/kg injections. Primary outcome was incidence of treatment-emergent adverse events (TEAEs). Secondary outcomes included change from baseline in Myasthenia Gravis Activities of Daily Living (MG-ADL) score. Results: At data cut-off (11 May 2023), median (range) exposure to zilucoplan was 1.8 (0.11–5.1) years (N=200). TEAEs occurred in 191 (95.5%) patients; the most common TEAE was COVID-19 (n=64; 32.0%). At Week 96, mean (standard error) change in MG-ADL score from double-blind study baseline was –6.33 (0.49) and –7.83 (0.60) for patients who received zilucoplan 0.3mg/kg and placebo in the double-blind studies, respectively. Conclusions: Zilucoplan demonstrated a favourable long-term safety profile. Efficacy was sustained for 96 weeks in