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

MS / Neuroinflammatory Disease

P.052

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

P.053

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.

P.054

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

patients who had previously received zilucoplan and who switched from placebo.

P.055

Corticosteroid dose tapering in patients with generalised myasthenia gravis on zilucoplan: Interim analysis of RAISE-XT

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Background: In the Phase 3 RAISE study (NCT04115293), zilucoplan significantly improved MG-specific outcomes in patients with acetylcholine receptor autoantibody-positive generalised MG. After the first 12 weeks of the open-label extension study, RAISE-XT (NCT04225871), corticosteroid dose could be changed per the investigator's discretion. We evaluate changes in corticosteroid dose during treatment with zilucoplan in RAISE-XT. Methods: In RAISE-XT, adults who completed the Phase 2 or RAISE studies (N=200) self-administered daily subcutaneous zilucoplan 0.3mg/kg, either continuing with zilucoplan or switching from placebo. Primary outcome was incidence of treatment-emergent adverse events (TEAEs). We assessed (*post-hoc*) the proportion of patients who discontinued/reduced or increased corticosteroid dose relative to double-blind baseline up to Week 60. Results: At Week 60, 30% (n=18/60) and 22% (n=12/54) of patients receiving corticosteroids in the zilucoplan and placebo-switch groups, respectively, reduced/discontinued corticosteroids (mean dose reductions: 14mg and 16mg; mean [SD] CFB in MG-ADL scores: -5.00 [3.96] and -5.67 [6.89]. 12% (n=7/60) and 7% (n=4/54) of patients in the zilucoplan and placebo-switch groups, respectively, increased corticosteroid dose (~12mg mean increase in both groups). TEAEs occurred in 188 (94.0%) patients (data cut-off: 08 September 2022). Conclusions: While receiving zilucoplan, discontinuation or dose reduction in concomitant corticosteroids was possible with maintained efficacy.

P.056

Real-world reduction in oral corticosteroid utilization following efgartigimod initiation in patients with generalized myasthenia gravis

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Background: Reducing oral corticosteroids (OCS) use can alleviate the risk of many adverse events related to long-term OCS use. Here, we evaluate real-world utilization of OCS among patients with generalized myasthenia gravis (gMG) over the first 6 months following efgartigimod initiation. Methods: Patients with gMG using OCS who initiated efgartigimod treatment were identified retrospectively from an open US medical and pharmacy claims database (IQVIA Longitudinal Access and Adjudication Data [LAAD], April 2016-April 2023). Average daily dose (ADD) of OCS was analyzed during the 3-month period preceding efgartigimod initiation, and at 3 and 6 months post-efgartigimod initiation. Results: Of 231 patients assessed, 17 (7.4%), 109 (47.1%), and 105 (45.5%) had baseline OCS ADD of 0–5 mg, 5–20 mg, or >20 mg, respectively. At 3 and 6 months post-efgartigimod, 82 (35%) and 99 (43%) patients, respectively, reduced ADD by ≥5 mg. Proportion of patients with ADD of 0–5 mg increased >3-fold (7% baseline vs. 26% 6 months post-efgartigimod) and proportion of patients with ADD of >20 mg decreased by 35% (45% baseline vs. 29% 6 months post-efgartigimod) following efgartigimod initiation. Conclusions: Approximately 43% of patients were able to decrease steroid use or achieved steroid-free status within 6 months of efgartigimod treatment initiation.

P.057

Achievement of minimal symptom expression in acetylcholine receptor antibody-positive participants treated with efgartigimod in ADAPT/ADAPT+

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Background: A key efficacy indicator in generalized myasthenia gravis (gMG) treatment is improvement in MG-ADL score. Minimal symptom expression (MSE, MG-ADL total score of 0 or 1) is explored as a novel proposed treatment target in gMG in the phase 3 study of intravenous efgartigimod, ADAPT, and its open-label extension, ADAPT+. Methods: Post hoc analyses of acetylcholine receptor antibody positive participants in ADAPT (n=129) and ADAPT+ (n=111) were performed. Results: In ADAPT, 44.6% receiving efgartigimod achieved MSE vs 10.9% of participants given placebo. Despite less frequent assessment during ADAPT+, 40.5% of participants achieved MSE. Eighty-one percent of participants treated with efgartigimod who achieved MSE in ADAPT also achieved MSE during ADAPT+; 23% who had not achieved MSE in ADAPT did in ADAPT+. Achieving MSE was associated with substantial improvements in QMG, MGC, MG-QoL15r, and EQ-5D-5L mean scores of 11.4, 16.0, 12.4, and 0.3 points, respectively, from baseline to best score (across all visits). These drastic improvements resulted in quality of life (QoL) comparable to that of healthy populations. MSE achievement also resulted in sustained improvements in these disease-specific and QoL measures. Conclusions: Participants who achieved MSE showed substantial and consistent improvements across multiple disease measures and experienced QoL comparable to that of healthy populations.