AAN Conference 2024 – Myasthenia Gravis-Focused Sessions

Session 1: NMJ Disorders

Abstract: NMJ Disorders: MG, Ocular, and MUSK Myasthenia

- BEAT-MG did not show steroid sparing effect; RINOMAX trial did due to early patient recruitment.
- MINT trial offers hope as another option for MG treatment.
- Ocular MG patients lack access to approved therapies due to exclusion from trials based on MGFA scores.
- MUSK MG patients respond well to B-cell depletion therapies like rituximab.
- Memory B-cells play a role in antibody production in MUSK MG.
- Complement inhibitors eculizumab and raviluzumab show improvement in MG-ADL scores.

Session 2: B Cells in gMG Pathogenesis

Abstract: The Role of B Cells in gMG Pathogenesis

- Discussion on CD20 vs. CD19 expression in treating AChR+ versus MuSK+ gMG.
- BEAT-MG study showed no significant differences in steroid sparing.
- RINOMAX study showed positive outcomes for AChR+ gMG patients.
- Comparative efficacy in AChR+ versus MuSK+ gMG discussed.

Session 3: Improvement with Rozanolixizumab

Abstract: Clinically Meaningful Improvement in Physical Fatigue and Muscle Weakness Fatigability with Rozanolixizumab

- Post-hoc analysis of MycarinG study showed significant improvements with rozanolixizumab.
- Significant percentage of patients achieved improvements in muscle weakness fatigability and physical fatigue.

Session 4: Cost-effectiveness of Efgartigimod

Abstract: Cost-effectiveness Analysis of Efgartigimod Versus IVIg for AChR-Ab+ gMG in Canada

- Efgartigimod showed cost savings and increased quality-adjusted life years compared to IVIg.

Session 5: Eculizumab Versus Rituximab

Abstract: Eculizumab Versus Rituximab for AChR+ gMG

- Real-world comparison showed eculizumab favored over rituximab for AChR+ gMG.
- Long-term remissions seen in patients responding to CD20 depletion.

Session 6: Comorbidity and Steroid Use

Abstract: Comorbidity Burden and Steroid Use in gMG

- Study analyzed comorbid conditions and steroid use patterns in gMG patients under Medicare.
- Significant comorbidity load observed, emphasizing cautious steroid use.

Session 7: Incidence and Prevalence of MG

Abstract: Incidence and Prevalence of Myasthenia Gravis

- Study estimated prevalence and incidence rates of MG in the US.
- Prevalence extrapolated to 116,255 individuals living with MG in the US.

Session 8: LUMINESCE Study on Satralizumab

Abstract: LUMINESCE Study on Satralizumab in gMG

- LUMINESCE study evaluating IL-6R antagonism with satralizumab in gMG.
- 186 participants enrolled, showing representative characteristics of gMG population.

Session 9: Early vs. Late Initiators of Eculizumab

Abstract: Early vs. Late Initiators of Eculizumab in gMG

- Early initiation within 2 years of diagnosis showed greater clinical benefit compared to late initiation.

Session 10: Rozanolixizumab Treatment Cycles

Abstract: Drivers of New Rozanolixizumab Treatment Cycles

- Initiation of new cycles driven by worsening MG-ADL and QMG scores.
- Physicians personalized treatment based on individual patient needs.

Session 11: Safety and Efficacy of Efgartigimod

Abstract: Long-term Safety and Efficacy of Efgartigimod in gMG

- Retrospective data analysis supports better outcomes for early initiators of eculizumab and ravulizumab.

Exploring Autoimmune Disease and Treatment Strategies in Myasthenia Gravis

Session 12: Immunopathogenesis in MG

Researchers conducted an exploratory proteomics analysis to study the differences in the immunopathogenesis of early and late-onset myasthenia gravis (MG) using the BeatMG study cohort. They identified distinct protein expressions in late-onset MG, suggesting a unique immunopathogenic mechanism.

- 80% of MG patients have autoantibodies against AChR
- Proteins highly expressed in late-onset MG: CXCL17, JCHAIN, CD83, TNFRSF11A
- LOMG potentially mediated by distinct immunopathogenic mechanism

Session 13: Zilucoplan Safety and Efficacy

An interim analysis of RAISE-XT demonstrated favorable long-term safety and sustained efficacy of zilucoplan up to 96 weeks in AChR Ab+ gMG patients. Notable reductions in MG-ADL scores and corticosteroid doses were observed.

- TEAEs occurred in 95.5% of patients
- Mean reduction in MG-ADL score: -6.33 (zilucoplan group)
- Reduction in corticosteroid dose: 61% of patients

Session 14: DNTH103 in Preclinical Model

DNTH103 showed promise in improving neurotransmission in a preclinical MG model, aiming to provide a safer and more convenient alternative to complement C5 therapies.

- DNTH103 selectively targets active C1s protein
- Potential for lower infection risk and improved dosing

Session 15: Shared Decision-making in gMG Care

Discussion on the importance of shared decision-making in the era of new targeted therapies for gMG, emphasizing the need for individualized treatment approaches.

- Exploration of evidence-based treatment decisions
- Highlighting the importance of optionality in gMG care

Additional Studies on Myasthenia Gravis

Study: Inclusion and Exclusion Criteria Impact

The study assessed the eligibility of AchR+ gMG patients in routine care for phase III clinical trials of complement and FCRN inhibitor therapies.

Key Findings:

- 89.5% of patients and 95% of clinic visits did not meet trial eligibility criteria.
- Main reasons for ineligibility: MG-ADL score >5 and recent IVIG use.
- Broader criteria could enhance patient eligibility and trial generalizability.

Study: Efgartigimod and Minimal Symptom Expression

The study focused on assessing the impact of achieving minimal symptom expression (MSE) in AchR+ gMG patients treated with efgartigimod.

Key Results:

- 44.6% of efgartigimod-treated participants achieved MSE.
- Participants with MSE showed substantial improvements in disease-specific measures and quality of life.
- Transitioning to efgartigimod resulted in sustained improvements.

Study: Transitioning to Ravulizumab from Eculizumab

The study evaluated the effectiveness and safety of transitioning gMG patients from eculizumab to ravulizumab.

Key Findings:

- 26% of patients transitioned from eculizumab to ravulizumab.
- Transition maintained stable MG-ADL scores and classifications.
- No new safety signals reported during the transition.

Study: Ravulizumab Wearing Off Phenomenon

The study investigated the "wearing off" phenomenon in gMG patients receiving ravulizumab.

Key Results:

- 33% of patients reported wearing off effect before the next infusion.
- Interventions included dosing adjustments and therapy switches.
- No specific factors identified for symptom fluctuation.