D.3

Self-reported autonomic nervous system dysfunction among people with drug-resistant focal epilepsy

HG Gray (London) H Kreinter (London) M Elnazali (London) J Shoemaker (London) A Suller Marti (London)*

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Background: Autonomic nervous system (ANS) dysfunction in people with epilepsy (PwE) is a likely contributor to sudden unexpected death in epilepsy (SUDEP). However, the nature of autonomic dysfunction among PwE remains poorly understood. We aimed to delineate self-reported ANS functioning among people with drug-resistant epilepsy, a patient group at increased risk for SUDEP. Methods: People with focal drug-resistant epilepsy undergoing stereoelectroencephalography at the Epilepsy Monitoring Unit in London, Ontario completed the Composite Autonomic Symptom Score (COMPASS-31), a widely used questionnaire for ANS function. Results: The mean total COMPASS-31 score (N=34; 13 females) was 27.36 (SD=13.77). There was no significant correlation between total COMPASS-31 score and current age (mean=32.71 years, SD=10.58; r(32)= -0.04) or age of epilepsy onset (mean=17.31 years, SD=8.26; r(30)=0). Females scored higher than males (t(32)=3.41, p<.05), but scores did not differ between participants with an epileptogenic zone in the temporal lobe(s) (N=20) and participants with multi-focal, extra-temporal or unknown epileptogenic zones (t(32)=0.18). Participants prescribed 2-3 sodium channel blocking anti-seizure medications (cardiotoxic; N=17), scored worse than participants on 0-1 sodium channel blockers (N=17) (t(32)= -2.15, p<.05). Conclusions: Autonomic testing should be a standard component of clinical care for people with drug-resistant epilepsy, especially for females and for those on sodium channel blockers.

D.4

International consensus recommendations for the management of glucocorticoid complications in neuromuscular disease

CD Kassardjian (Toronto) C Bacher (Toronto) P Narayanaswami (Boston) R Laughlin (Rochester) M Bromberg (Salt Lake City) F Buttgereit (Berlin) D Dodig (Toronto) EP Flanagan (Rochester) C Gottlieb (Ottawa) JK Marshall (Hamilton) RG McCoy (Baltimore) S Mitchell (Toronto) PK Tosh (Rochester) P Sinha (Boston)*

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Background: Adverse effects and risks associated with glucocorticoid (GC) treatment are frequently encountered in immune-mediated neuromuscular disorders. However, significant variability exists in the management of these complications. Our aim was to establish international consensus guidance on the management of GC-related complications in neuromuscular disorders. Methods: An international task force of 15 experts was assembled to develop clinical recommendations for managing GC-related complications in neuromuscular patients. The RAND/UCLA Appropriateness Method (RAM) was employed to formulate consensus guidance statements. Initial statements were drafted following a comprehensive literature review and were refined based on anonymous expert feedback, with up to three rounds of email voting to achieve consensus. Results: Consensus was reached on statements addressing general patient care, monitoring during GC therapy, osteoporosis prevention, vaccinations, infection screening, and prophylaxis for *Pneumocystis jiroveci* pneumonia. A multidisciplinary approach to managing GC-related complications was highlighted as a key recommendation. Conclusions: This represents the first consensus guidance in the neurological literature on GC complications, and offer clinicians structured guidance on mitigating and managing common adverse effects associated with both short- and long-term GC use. They also provide a foundation for future debate, quality improvement, research work in this area.

D.5

Efgartigimod treatment in participants with anti-acetylcholine receptor seronegative generalized myasthenia Myasthenia Gravis clinical studies

O Blanchard (Montreal) S Steeland (Ghent) A Meisel (Berlin) V Bril (Toronto) M Jiang (Ghent) R Kerstens (Ghent) K Heerlein (Ghent) J Howard Jr (Chapel Hill) T Vu (Tampa) R Mantegazza (Milan) K Utsugisawa (Hanamaki)*

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Background: Antibodies directed against acetylcholine receptor (AChR) are absent in approximately 15% of patients with gMG. Approved treatment options represent an unmet need in the AChR-antibody (Ab)- gMG population. Efgartigimod is an immunoglobulin G1 (IgG1) antibody Fc fragment that selectively reduces IgG levels by blocking neonatal Fc receptor (FcRn)-mediated IgG recycling. Here, we describe efgartigimod efficacy in AChR-Ab- participants with gMG receiving either efgartigimod IV or subcutaneous (SC) efgartigimod PH20 (coformulated with recombinant human hyaluronidase PH20) across clinical studies. Methods: Post hoc analyses were conducted to examine efficacy and safety of efgartigimod IV and/or efgartigimod PH20 SC in AChR-Ab- participants in ADAPT/ADAPT+ and ADAPT-SC/ADAPT-SC+ trials. Results: Among pooled AChR-Ab- participants (n=56), mean (SE) MG-ADL total score improvement from baseline to Week 3 was -3.7 (Cycle 1: 0.44 [n=55]). Consistent MG-ADL improvements occurred with repeated cycles. Clinically meaningful improvements (CMI; ≥2-point MG-ADL decrease) occurred in 76.4% (n=42/55) of participants (Cycle 1, Week 3). In Cycle 1, 23.2% (n=13/56) of participants achieved minimal symptom expression (MG-ADL 0-1). Similar efficacy results occurred across all cycles. Overall safety profile was similar between AChR-Ab+ and AChR-Ab- participants. Conclusions: Both efgartigimod IV and efgartigimod PH20 SC were well tolerated and led to CMI in participants with AChR-Ab- gMG.