Poster Presentations

Retrospective Longitudinal Assessment of MG-ADL Score with Treatment of Myasthenia Gravis
Matthew Varon (Kansas City, KS), Mazen Dimachkie (Kansas City, KS)

One of the big advances in the clinical evaluation of MG is the development and increasingly widespread use of clinical measures of patient strength, limitations in carrying out activities of daily life (ADL) and patient perspective on their satisfaction with life. The MG-ADL assesses the symptoms and the impact of MG on the ability of people with MG to perform ADL. This very measure is currently completed at a clinical visit. The purpose of this study was to determine the practicality and accuracy of determining MG-ADL scores from a person’s electronic medical record (EMR). These researchers found that it was possible to obtain an MG-ADL score from the information in the EMR and that the EMR-acquired MG-ADL scores agreed with the MG-ADL scores obtained at a clinical visit. To be able to deduce the MG-ADL score, information needs to entered in the EMR in a disciplined fashion so that the details needed to determine the MG-ADL are present. One importance of this study is that outcome measures such as the MG-ADL need to be studied over the duration of the study. Traditionally, clinical studies, such as clinical trials, have required participants to attend multiple evaluation sessions at study sites. Having to go to study sites can prevent some patients from participating in a clinical study. This study indicates that a carefully documented EMR can enable subjects to remain close to home provided their clinicians are able to carefully document the EMR so that outcome measures such as the MG-ADL can be ascertained to measure the outcomes of different interventions, enabling participants in clinical trials to have data collected without having to travel to a main study site.

Thymectomy in Seropositive Myasthenia Gravis in a Quaternary Neuromuscular Division Before and After Publication of the MGTX Study
Constantine Farmakidis (Kansas City, KS), Mazen Dimachkie (Kansas City, KS)

The MG thymectomy trial (MGTX) established that thymectomy improved the clinical state of people with AChR-MG. This study assessed whether the findings of the MGTX altered the number of thymectomies performed. They found that people with AChR-MG were 1.8 times more likely to have a thymectomy in the period since MGTX compared to before MGTX. This study indicates that the MGTX, by demonstrating the benefits of thymectomy, has encouraged thymectomy to be performed more often.

Exercise Provocation of Stimulated Single Fiber Electromyography, an Attempt to Increase the Diagnostic Yield in Myasthenia Gravis Patients
AyatAllah Farouk Hussein (Cairo, Egypt)

This was a clever study that utilizes exercise-induced worsening of NMJ function in MG to enhance the sensitivity of single fiber EMG (SFEMG) testing for detecting MG. The techniques described in this poster should be considered when SFEMG results are negative or questionable in a patient with clinically suspected MG. Recall that for people with clinical manifestations of MG who do not have
detectable levels of antibodies against MG-associated muscle proteins such as AChR and MuSK (sero-negative), EMG studies are important to confirm a diagnosis of MG.

**Co-Occurrence of Ocular Myasthenia Gravis and Chronic Inflammatory Demyelinating Polyradiculoneuropathy**

Anirudha Rathnam (Detroit, MI), Ritika Suri (Detroit, MI), Naganand Sripathi (Detroit, MI), Kavita Grover (Detroit, MI)

This was a case report of a middle-aged man who initially developed MG at age 43 and 26 years later he developed a second autoimmune condition, damaging motor nerve fibers (chronic immune demyelinating polyradiculopathy (CIDP)). This case emphasizes that people with MG may have a tendency to develop other auto-immune conditions such as thyroid disease, lupus or in the case of this person CIDP. Relevant to the next poster, one possible reason for people with MG to be at increased risk for autoimmune disease is that may have impairment in the immune “checkpoint” system that normally prevents the immune system from attacking the body.

**Lambert-Eaton Myasthenic Syndrome in the Setting of Immune Checkpoint Inhibitor Treatment of Small Cell Lung Cancer**

Nadim Jiwa (Boston, MA)

One way of treating advanced cancer is to “rev-up” the immune system by suppressing the “checkpoint” mechanism that normally acts to prevent the immune system from attacking the body. The objective of checkpoint inhibition is to enhance the immune system so that immune cells attack cancer cells (remember that cancer cells are derived from normal cells). A side effect of using checkpoint inhibitor therapy is that autoimmune disorders such as MG may emerge. Lambert-Eaton Myasthenic Syndrome (LEMS) is an autoimmune disorder that is caused by antibodies directed against the nerve terminal resulting in impaired release of ACh. This is a case report of a person who developed LEMS in association with checkpoint inhibition treatment used to treat lung cancer. It may seem odd to use a treatment that can induce autoimmune disorders, but the treatment is being used for people who are critically ill with advanced cancer. While the side effects of checkpoint regulator inhibition can be serious, advanced cancer is fatal.

**Congenital Myasthenic Syndromes: A Clinical, Electrophysiological, and Genetic Review**

Joshua Kaltman (Decatur, GA)

This study discussed congenital disorders of neuromuscular transmission, aka congenital MG (CMG) and congenital myasthenic syndromes. The subjects were children seen at a regional children’s hospital from 2013 to 2019. They reported on thirteen children with a mean age of about 7 ½ years of age. The manifestations of the CMG varied among the subjects varied according to the causative mutation that could impair release of ACh, ACh processing in the synaptic cleft, or the interaction of ACh with the muscle (for example too few or dysfunctional AChRs). The encouraging point of this study was that knowing what the NMJ dysfunction was enabled clinicians to use agents that would effectively address the underlying disorder.

**Natural Course and Treatment of ACHR-MG Converted to MuSK-MG or DP-MG in Children: 2 Case Studies and Literature Review**

Yaru Lu (Guangzhou, China), Hao Ran (Guangzhou, China), Qian Ma (Guangzhou, China), Xiaoxi Liu, Guangzhou, China, Weibin Liu (Guangzhou, China)

This poster addressed two unusual cases of childhood MG in which children who initially had AChR-MG, later developed antibodies to MuSK.
Hand Myasthenia in a Patient with Positive Acetylcholine Receptor Antibodies
Ricardo Maselli (Davis, CA)

This is a description of an unusual case of MG in which the patient later developed weakness in one hand with EMG findings indicating that the EMG supported a diagnosis of hand weakness due to MG. Note that weakness in MG is often asymmetric, with some muscles on one side of the body being weaker than the same muscles on the other side. This case was unusual in the extreme asymmetry.

LRP4 and Agrin Antibodies in Myasthenia Gravis (MG): Augusta University (AU) Experience
Michael Rivner (Augusta, GA), Brandy Quarles (Augusta, GA), Jin-Xiu Pan (Cleveland, OH), Zheng Yu (Cleveland, OH), Kristy Bouchard (Augusta, GA), Lin Mei (Cleveland, OH)

This study reported on 61 people with clinical MG who had antibodies to LRP4, agrin or both. These patients had symptoms of MG and abnormal EMG studies. Patients were re-tested for antibodies against AChR and MuSK. Fifteen had AChR antibodies, 7 had MuSK antibodies, 6 subjects (9.8%) had LRP4 antibodies and 5 (9.6%) had agrin antibodies. Four (7.5%) had antibodies to both LRP4 and agrin. No patient had AChR and MuSK antibodies. One person had LRP4 and AChR antibodies, 1 person had agrin and AChR antibodies and 1 person had LRP4 and MuSK antibodies. No person had antibodies to both agrin and MuSK. Among this group of people with an initial diagnosis of seronegative MG, I was impressed by the following points: 1) More than 1/3 of people had antibodies to AChR or MuSK on retesting, so were not really seronegative; 2) 11% had antibodies to LRP4, agrin or both; 3) so overall almost half of the people had antibodies to AChR, MuSK, LRP4 or agrin; 4) people did not have both AChR and MuSK antibodies, but people could have both LRP4 and agrin, LRP4 and AChR, LRP4 and MuSK and agrin and MuSK. This group of people with MG had clinically moderate or severe MG. It is presently unknown if these patients will respond to newer treatments such as rituximab or eculizumab. The more that seronegative people are studied and understood the better treatment for seronegative-MG will become.

Evaluation of Medications Implicated in Promoting Myasthenia Gravis (MG) Exacerbation Fact or Fiction
George Small (Pittsburgh, PA), Mohammad Ali (Wexford, PA), Carol Schramke (Pittsburgh, USA)

The Myasthenia Gravis Foundation of America (MGFA) lists multiple medications as relatively contraindicated (RCM) in MG patients (see MGFA website). The listing of medications is based upon the nature of the medication (does it have the potential to impair neuromuscular function) and have there been clinical cases where a medication caused clinical worsening of MG. However, since MG is a snowflake disease, two patients may react differently to the same medication. This study reviewed the medical records of 100 people with MG over a 3 year period. They looked for the presence of MG exacerbations in people who did and did not take a RCM. 67 patients took a RCM and 37% had an exacerbation. In contrast, 46% of people who did not take a RCM had an exacerbation. Looking at specific classes of medications such as beta-blockers and antibiotics on the RCM list, they did not find a higher rate of exacerbation among people who took a RCM. Of note, beta-blockers were placed on the RCM list at a time when this class of medications was just starting to be used. Beta-blockers have evolved with many new preparations in use that have largely replaced early preparations. Today, beta-blockers are commonly used medications. This study emphasizes that the MGFA list is medications that are RELATIVELY contraindicated, not banned for use in people with MG. The decision to use a medication on the RCM list needs to be made between the patient and clinician based upon the individual characteristics of the patient and whether the medication in question is the best medication to use.