MGFA 2020 Scientific Session POSTER PRESENTATIONS:

Overlap Paraneoplastic Syndrome: Thymoma Presenting as Myasthenia Gravis and Immune Mediated Myopathy - A Case Series. M Cuchanski, Fairport, NY
Tumors of the thymus gland, thymomas are known to alter the function of the immune system. The precise reason is unclear, but may be related to the belief that T-cell lymphocytes mature in the thymus. Perhaps abnormalities of the structure of the thymus, such as the development of tumor of the thymus may alter the normal maturation of T-cells. People in the MG community know that thymomas are associated with MG. This presentation shows that thymomas can disturb the immune system to produce other autoimmune disorders. They describe a person who developed an autoimmune disorder that involved entire muscle fibers, not just the region of the neuromuscular junction.

Follow-Up Care in Myasthenia Gravis During Covid-19: Comparison of Telemedicine and In-Person Encounters. C Farmakidis, Prairie Village, KS and University of Kansas, Kansas City KS
Telemedicine is an important element of outpatient care for a variety of patients, particularly people who live far from care givers and when isolation is needed as is true during the current pandemic. This study examined how effective caregiver-patient interactions were for people with MG when the assessments were done using videoconferencing, telephone or in-person. The remote interactions via telephone or videoconferencing appeared to be in equally effective to in-person interactions for people who were not severely compromised by MG. I am pleased that this group found that video conferencing and telephone exchanges are effective. I agree that people who have more severe disease or during challenging times benefit from in-person assessments. I am encouraged that the work being done by Dr. Guidon and her colleagues - described elsewhere in the discussion of this excellent meeting – will make future telemedicine assessments even more effective.

Validation of a Surface Plasmon Resonance Assay for the Diagnostic Detection and Characterization of Muscle-Specific Tyrosine Kinase (MuSK) Antibodies in Myasthenia Gravis Patients - E Gibbs Univ British Columbia, Vancouver, Canada
This was a technical study of a new way of assaying for antibodies to MuSK. The technique being described holds promise for improving the detection of MuSK antibodies.

Characteristics and Outcomes of Myasthenia Gravis Patients with Covid-19 – A Case Series - P Kwon, Brooklyn, NY
A point of clarification – the poster refers to both Covid-19 and SARS-CoV-2. SARS-CoV-2 is the specific strain of Coronavirus that causes the disease Covid-19. This study resports on 7 people with MG who developed Covid-19. Prior to the Coronavirus infection, none of the people were seriously ill. Five of 7 developed breathing difficulty that was severe enough to require hospitalization and two of the five hospitalized patients died. Two of the three surviving hospitalized patients received a dose of a monoclonal antibody against interleukin-6 (an inflammatory protein that causes tissue damage, inhibition of IL-6 is employed to treat rheumatoid arthritis, the agent was

tocilizumab) had marked improvement in their clinical course, ultimately discharged at their baseline neurologic status. The IL-6 inhibitor is different from the antibody cocktail used to treat President Trump. The number of cases reported is large for this type of report, but too small to draw firm conclusions. Combined with other reports it appears to me that people with MG are at risk to develop severe manifestation if they acquire Covid-19. Different treatment approaches that target the SARS-CoV-2 or that inhibit an exaggerated inflammatory response in Covid-19 need to be further studied.

Myasthenia Gravis Population at U Conn Health: A Single Center Profile Analysis Comparing Patient Epidemiology and IVIg Treatment Requirements. A Healy Farmington, CT, M Imperioli (Southbury, CT)
This study considers the use of IVIG treatment in patients with MG. They reported 17.7% required IVIG only for acute exacerbations, while 18.9% required maintnenance IVIG treatment. The patients with who developed MG after 65 years of age and who had AChR-MG were more likely to require maintenance IVIG.

A Novel Diagnostic Method for Myasthenia Gravis - G Kocak, Istanbul, Turkey
This was a very clever study. In the 1980's to early 2000's I participated in research led by Drs. Daroff, Dell"Osso and RJ Leigh that described the eye movement abnormalities in people with MG. While those studies defined eye movement abnormalities that are specific for MG, the technology employed to measure the eye movements could not be employed in a clinic setting. Thus, while people with MG had distinctive eye movement abnormalities, eye movement measurements could not be performed in a clinically practical manner. The importance of this study was to employ technology that could be used in a clinic setting. The researchers were made the research done in the over 30 years ago clinically relevant today.

Determination of Complement Activation in Myasthenia Gravis: A Pilot Study - I Lee Univ of Alabama in Birmingham, AL
In AChR-MG, damage of the neuromuscular junction is mediated by activation of the inflammatory chemical system called complement. Inactivation of complement by treatments such as eculizumab is useful to treat AChR-MG. This study examined whether measuring the small peptide fragments produced when complement is activated and broken down by the body. The idea was worthy, but the findings so far do not indicate that employed techniques are able to reliably detect fragments of recently activated complement in people with active MG.

Rapidly reversible tongue atrophy in Seronegative Myasthenia Gravis following treatment - J Massey Duke Univ., Durham. NC
The tongue is a very unusual skeletal muscle that moves itself and does not move a bone. Tongue atrophy can occur in MG, but is more often associated with motor neuron disease (amyotrophic lateral sclerosis, ALS) than with MG. This is a report of 61 year old man with seronegative MG who had prominent atrophy of the tongue. He responded to MG treatment prednisone-like agents and PLEX with improvement in muscle strength and restoration of tongue mass.

Familial Musk Myasthenia Gravis - J Massey & EW Massey Duke Univ., Durham. NC
This is the first report that the authors are aware of in which a brother and sister both manifest MuSK-MG. It is not clear if the presence of MuSK-MG in the siblings was a rare but random event or if the siblings had a genetic predisposition toward MuSK-MG.

Variability of Complement Levels in a Spectrum of Myasthenia Gravis Patients
D Post Stanford, CA
The idea of this study was similar to the above poster by Dr. Lee, to determine if measuring amounts of components of complement can be a useful biomarker for MG. As was true in Dr. Lee's report, the investigators reported that the measured levels of complement components were highly variable. Consequently, as measured in this and Dr. Lee's study, complement would not be a useful biomarker for MG.

Acetylcholine Receptor (AChR) Antibody Overshoot Following Plasmapheresis Associated with Clinical Deterioration of Myasthenia Gravis - D Richards Cleveland Clinic Foundation Cleveland, OH and collaborators in Los Angeles, CA
Plasmapheresis (PLEX) is effective treatment for generalized MG. There is a theoretical risk that PLEX will also remove factors that may be suppressing antibody production. Thus there is a risk or increased antibody production leading to increased antibody levels following PLEX. The authors report 5 people with AChR-MG who showed increased AChR antibody levels within 1 to 6 months following PLEX. Four patients had worsening of their MG to point of myasthenic crisis or impending crisis. The authors feel it important to utilize immunosuppression in concurrence with PLEX to prevent a rebound in AChR antibody levels due to PLEX.

Real-World Experience of Eculizumab for Myasthenia Gravis - J Suh Boston, MA
This is a report of the effectiveness of Eculizumab in clinical practice, not in a research protocol. They found that about 2/3rds of patients improved with Eculizumab. Patients no longer needed IVIG or PLEX. Reduction of prednisone and other oral immunosuppresants was modest. The types of side effects were similar to those noted in clinical trials. (please also refer to the Data Blitz presentation of Dr. Muley which reported that the complication rates for Eculizumab in clinical practice was similar to what was reported in clinical trials).

MuSK Mutation in an Anti-Acetylcholine Receptor and Musk Antibody Negative Patient. N Suresh Riverview, FL and Univ of South FL
This is a report of an unusual adult who manifest progressive limb weakness without antibodies to AChR or MuSK. The person had a unique mutation that altered MuSK. EMG findings were not those of a person with MG, but he did improve with mestinon and prednisone. More studes will be needed to understand how the MuSK mutation led to the person's weakness and why he responded to mestinon or prednisone.

Performance of Different Criteria for Refractory Myasthenia Gravis - C Tran Scarborough, Canada and Univ. Toronto

This study compared different criteria used to determine when an individual had refractory MG. This group favored the criteria from the International Consensus Guideline.

A Case of Musk Myasthenia Gravis Presenting with Isolated Respiratory Insufficiency. C Tsai, Chapel Hill, NC and Univ North Carolina
This is a report of an unusual clinical presentation of a young adult woman with MuSK-MG. Throat/mouth weakness are breathing muscle weakness can occur in MuSK-MG, but this case was unusual in that the initial manifestation was progressive breathing muscle weakness.

Efficacy of Eculizumab in Myasthenia-Gravis-Foundation-of-America (MGFA) Grade-V Myasthenia Gravis. U Usman New Haven, CT and others in AZ, and OH
The MGFA grading system measures the clinical severity of MG. Grade V is a person in crisis with severe MG. Eculizumab has not been studied in people with MG crisis. They report on 3 AChR-MG patients who were hospitalized in MG crisis and treated with eculizumab. Each person improved clinically. This study suggests that eculizumab can be a clinically useful treatment for people with the severest forms of MG.