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STANDARD MEDIAN NERVE ULTRASOUND IN CARPAL TUNNEL SYNDROME: A RETROSPECTIVE REVIEW OF 1,021 CASES
Santoshi Billakota, Lisa Hobson-Webb (Durham, NC)

INTRODUCTION/OBJECTIVE: CTS is diagnosed with EDX studies. Investigations have examined US cross-sectional-area (CSA) and wrist to forearm ratio (WFR) cut-offs for screening EDX abnormalities in patients with suspected CTS. The objective of this study is to determine if these US parameters are effective in a real world population.

METHODS: This is a retrospective review of patients presenting to the Duke EMG Laboratory during 2013-2014 with a final diagnosis of CTS. US diagnosis of CTS was based upon median nerve cross-sectional area of >9mm2 and/or wrist-to-forearm ratio of >1.4. EDX studies were the gold standard for diagnosis.

RESULTS: A total of 1,021 patients and 2,042 extremities were studied; 1,905 extremities carried a final diagnosis of CTS. US was positive in 97.7% of EDX confirmed CTS.

SUMMARY/CONCLUSION: Median nerve US is nearly as sensitive as the gold standard for EDX testing for the diagnosis of CTS.

Santoshi Billakota, MD
Golseth Young Investigator Award Recipient

RESPONSE TO INTRAVENOUS IMMUNOGLOBULIN IN PATIENTS WITH CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY: THE INSIGHTS QUALITY IMPROVEMENT PROJECT
Todd Levine (Phoenix, AZ), Jonathan Katz (San Francisco, CA), Richard Barohn (Kansas City, KS), David Saperstein (Phoenix, AZ), Gil Wolfe (Buffalo, NY), Lara Katzin (Tampa, FL), Tahseen Mozaffar (Orange, CA), Mazen Dimachkie (Kansas City, KS), Gary Badger (Burlington, VT), Leslie Vaughan, Elissa Ritt, Michelle Greer (Temecula, CA)

INTRODUCTION: IVIg is prescribed for a wide variety of neuromuscular diseases. Despite established criteria for diagnosing diseases such as chronic inflammatory demyelinating polyneuropathy (CIDP), there is great variability in the types of patients who are prescribed IVIg.

OBJECTIVE: To utilize a quality-improvement project to determine the clinical, laboratory, and electrophysiologic criteria most predictive of a response to treatment in neuromuscular disease patients prescribed IVIg.

METHODS: The clinical, laboratory, and electrophysiologic data of 585 patients who were prescribed IVIg were reviewed by a panel of blinded, independent neuromuscular neurologists. Outcomes were determined based on quality-of-life measures, the Patient Global Impression of Change, and clinical documentation.

RESULTS: The largest group of patients were those diagnosed with neuropathy (n=166); 40% of these patients were judged appropriate candidates for IVIg. This mirrored the overall response rate of 48%. Meeting American Academy of Neurology (AAN) or European Federation of Neurological Societies (EFNS) criteria for a demyelinating neuropathy was a predictor of response (63% versus 33%). Among patients who met criteria for CIDP, the response rate was 58% and 63% for multifocal motor neuropathy (MMN). There was a 25% response rate for patients with axonal neuropathies. There was no association with age, distribution of symptoms, dose or brand of IVIg, or chronicity of disease.

SUMMARY/CONCLUSION: The overall response rate to IVIg in patients diagnosed with demyelinating neuropathies is 48%. Adhering to AAN or EFNS criteria improved this response rate to 58% for CIDP and 63% for MMN. This suggests more specific recognition of diseases can improve outcomes and limit over-utilization of IVIg.

Todd Levine, MD
Best Abstract Award Recipient
3 CHALLENGES IN THE DIAGNOSIS OF CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY: A REVIEW OF PATIENTS TREATED WITH INTRAVENOUS IMMUNOGLOBULIN FROM A SPECIALTY PHARMACY DATABASE
Kenneth Gorson (Brighton, MA), Jeffrey Allen (Chicago, IL), Deborah Gelinas (Research Triangle Park, NC), Daniel Serrano (Bronx, NY)

INTRODUCTION/OBJECTIVE: The aim of this study was to explore neurologists’ adherence to the European Federation of Neurological Societies (EFNS)/Peripheral Nerve Society (PNS) guidelines for the diagnosis of chronic inflammatory demyelinating polyneuropathy (CIDP) by reviewing data from a specialty pharmacy database.

METHODS: Clinical, electrophysiological, cerebrospinal fluid, MRI, and nerve biopsy data were reviewed for 65 consecutive patients treated with IVIg. Three neuromuscular neurologists classified cases according to EFNS/PNS criteria as (1) fulfilling CIDP criteria, (2) possibly CIDP (satisfied EFNS/PNS clinical but not electrophysiologic diagnostic criteria), (3) not CIDP (neither clinical nor electrophysiologic criteria met), or (4) insufficient information to make a diagnosis.

RESULTS: The patients were treated by 31 different community neurologists in 14 states. Nineteen providers referred only 1 patient and 1 provider referred 19 patients (none of whom met criteria for CIDP). Only 11% met criteria for CIDP, 1% met clinical but not electrophysiological criteria, 55% did not have CIDP, and 33% had insufficient information to make a diagnosis. The IVIg average induction dose was 1.26 g/kg (0.35-2.6) and the mean maintenance dose was 0.84 g/kg (0.2-2.0). These average IVIg doses were lower than those reported in controlled clinical trials.

SUMMARY/CONCLUSION: Misdiagnosis of CIDP by community neurologists was common. These findings suggest that widely accepted and readily available CIDP diagnostic guidelines are underutilized during routine clinical care. It was also observed that documentation in the clinical record is often insufficient to support diagnosis. These findings suggest a need for further education on diagnostic guidelines.

Jeffrey Allen, MD
Best Abstract Runner Up Award Recipient

4 THE SAFETY OF NERVE CONDUCTION STUDIES IN PATIENTS WITH CENTRAL LINES
Zachary London, Andrew Mundwiler, Hakan Oral, Gary Gallagher (Ann Arbor, MI)

INTRODUCTION: Central venous catheters breach the skin and extend towards the heart, reducing the normal electrical resistance provided by intact skin. Some authors recommend avoiding or modifying NCSs in patients with central venous catheters to avoid the theoretical risk of inducing cardiac arrhythmia.

OBJECTIVE: To determine if NCSs affect measures of cardiac conduction or cardiac rhythm in patients with central venous catheters.

METHODS: Under continuous 12-lead electrocardiogram (EKG) monitoring, subjects with and without central venous catheters underwent a series of NCSs. The protocol included a total of 16 NCSs, including every permutation of left and right, ulnar and spinal accessory motor nerves, short and long duration stimulations, and single and 2-Hz repetitive stimulations. EKG tracings were marked at the time of each stimulation. A board-certified cardiologist reviewed the EKG tracings for evidence of cardiac conduction abnormality or arrhythmia, defined as 3 or more consecutive ectopic beats or a triplet.

RESULTS: Ten control subjects and 6 subjects with central venous catheters underwent the NCS protocol. Single benign ectopic beats and atrial runs were found in both study subjects and control subjects. No serious arrhythmias or conduction abnormalities were noted in either group.

SUMMARY/CONCLUSION: NCSs of the upper extremities, including both proximal stimulations and repetitive stimulation, do not appear to confer increased risk of cardiac conduction abnormality or arrhythmia in patients with central venous catheters. The presence of a central venous catheter is not a contraindication to standard NCSs.

Gary Gallagher, MD
Best Abstract Runner Up Award Recipient
ULTRASOUND DIFFERENTIATION OF CHARCOT–MARIE–TOOTH TYPE 1 AND CHRONIC INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY
Mingsheng Liu, Liying Cui, Jingwen Niu (Beijing, China)

INTRODUCTION: Ultrasound (US) has been mainly applied for diagnosis of mononeuropathy. Some research has reported the features of peripheral nerve US in demyelinating neuropathies. However, most reports only scanned 1 or several sites along the nerve, which may not reflect the morphological features of demyelinating nerves which could be involved patchily.

OBJECTIVE: To prove whether nerve US can differentiate between Charcot–Marie–Tooth type 1 (CMT1) and chronic inflammatory demyelinating polyradiculoneuropathy (CIDP).

METHODS: Systematic US measurements of peripheral nerves were performed in 18 patients with CIDP, 13 patients with CMT1, and 16 healthy control subjects. Median and ulnar nerves were scanned from distal to proximal parts along the nerve. The cross-sectional areas (CSAs) were measured at 10 defined locations, respectively.

RESULTS: CSAs measured in all locations on median and ulnar nerve were significantly increased in CMT1 than in CIDP and healthy control subjects (p<0.01). CSAs measured in 8 locations of median nerve and 8 locations of ulnar nerve were significantly increased in CIDP than in healthy control subjects. Receiver operation characteristic (ROC) curve analysis revealed CSA measurements to be well suited for differentiation of CMT1 and CIDP, with area under curve of 8 locations of median nerve and 9 locations of ulnar nerve of more than 0.9. CSA may change abruptly along the nerve in CIDP but not in CMT1.

SUMMARY/CONCLUSION: Systematic US CSA can be used for differentiating CMT1 from CIDP.

Mingsheng Liu, MD, President’s Research Initiative Award Recipient

Mingsheng Liu, MD, Master in Neuroscience, Recipient of the 2016 IFCN North American Chapter Fellowship Award

PK/PD MODELING AND SIMULATION OF 3,4-DIAMINOPYRIDINE BASE IN PATIENTS WITH LAMBERT–EATON MYASTHENIA SYNDROME
Jeffrey Guptill (Durham, NC), Nilay Thakkar (Chapel Hill, NC), Charles Pelouquin (Gainesville, FL), Kathy Ales (Princeton, NJ), David Jacobus (Princeton, NJ), Laura Jacobus (Princeton, NJ), Michael Cohen-Wolkowiez (Durham, NC), Daniel Gonzalez (Chapel Hill, NC), DAPPER Study Group

INTRODUCTION: 3,4-diaminopyridine (3,4-DAP) is the drug of choice to treat Lambert–Eaton myasthenic syndrome (LEMS)-related weakness, but the optimal dose regimen is uncertain.

OBJECTIVE: To develop a population pharmacokinetic/pharmacodynamic (PK/PD) model of 3,4-DAP free base in LEMS and apply the final model to perform 3,4-DAP dosing simulations.

METHODS: A population PK/PD analysis was performed using concentrations of 3,4-DAP collected from the DAPPER study, a randomized double-blind, placebo-controlled, withdrawal study in LEMS patients. The triple timed up and go (3TUG) assessment was the primary PD measure. Clinically-relevant dosing strategies were simulated and the percentage of patients achieving an improved 3TUG response calculated.

RESULTS: A total of 1270 PK samples from 49 subjects were analyzed. 3TUG data for 32 randomized subjects and 1091 3TUG data points were used. The median (range) age and weight was 60 years (23-83) and 80 kg (46-117), respectively, and the total daily dose was 80 mg (30-100). A 2-compartment model described the PK data well. Important covariates in the final model were body weight and serum creatinine. A fractional inhibitory Emax model characterized the exposure-response relationship well: Effect (sec)=18.3*[1−(0.69*Cp)/(25.2+Cp)] with EC50 expressed in ng/mL. Simulated doses of 10-20 mg demonstrated that >90% of patients could achieve a ≥20% improvement in TUG time.

SUMMARY/CONCLUSIONS: PK/PD modeling confirmed a concentration-response relationship with 3,4-DAP in LEMS patients using the 3TUG assessment. The simulations support that LEMS patients can expect a modest improvement in 3TUG times at low 3,4-DAP doses, and up titration often improves the magnitude of improvement.

Jeffrey Guptill, MD, President’s Research Initiative Award Recipient
INNOVATIVE ASSESSMENT OF MUSCLE INJURY USING ELECTRICAL IMPEDANCE MYOGRAPHY
Benjamin Sanchez (Brookline, MA), Shama Iyer (Baltimore, MD), Jia Li (Boston, MA), Richard Lovering (Baltimore, MD), Seward Rutkove (Boston, MA)

INTRODUCTION: Duchenne muscular dystrophy is a genetic disorder characterized by muscle degeneration, weakness, increased susceptibility to injury, and ultimately premature death.

OBJECTIVE: To investigate the ability of noninvasive electrical impedance myography (EIM) as a novel tool to assess the physiopathology of muscular injury associated to disease.

METHODS: Injury was induced in the quadriceps of healthy wild type (WT, n=10) and dystrophic (mdx, n=10) mice from the C57BL/10ScSnJ strain (male, 2-month-old, body weight: 27.4±2.7 g and 27.8±2.1 g, respectively). Muscle injury resulted from 15 forced lengthening contractions through a 60 degree arc of motion. Standard functional outcomes and EIM were assessed pre-injury and 0 hours, 24 hours, and 48 hours post-injury. The opposite thigh served as a control and data were normalized to pre-injury. Statistical significance was assessed using Mann-Whitney and two-way ANOVA tests, two-tailed.

RESULTS: Compared to WT mice, mdx animals had larger loss in peak twitch, 66.6±5.5% and 37.3±6.5% (p<0.01), and peak tetanic torque, 68.4±4.5% and 22.1±5.0% (p<0.0001). Injured WT and mdx muscle showed an increase in characteristic frequency by 177% (p<0.0001) and 167% (p<0.0001), respectively; and a decrease in extracellular resistance by 46% (p<0.0001) and 45% (p<0.0001), respectively.

SUMMARY/CONCLUSION: This is the first use of EIM to assess in vivo injury. The changes in EIM parallel the outcomes from standard functional measurements. Results suggest that characteristic frequency does reflect membrane disruption between sarcolemma and t-tubule, and extracellular resistance is able to detect damage-induced muscle tear and interstitial edema.

Benjamin Sanchez, PhD, President’s Research Initiative Award Recipient

LONGITUDINAL EVALUATIONS OF UPPER EXTREMITY 3D REACHABLE WORKSPACE IN POMPE DISEASE BY KINECT SENSOR
Divya Reddy, Sarah Humbert (Loma Linda, CA), Evan de Bie, Alina Nicorici (Sacramento, CA), Jay Han (Irvine, CA)

INTRODUCTION: Pompe disease is a rare multisystem metabolic myopathy, inherited in an autosomal recessive manner, which is caused by a deficiency of glycogen-degrading lysosomal enzyme acid α-glucosidase (GAA). The pathological feature of Pompe disease is lysosomal accumulation of glycogen in muscle tissue throughout the body, with cardiac and skeletal muscle being the most commonly affected. Diagnosis can be challenging as patients show limb-girdle weakness similar to a myriad of other neuromuscular disorders. With the rising interest in the use of technology for reliable upper extremity functional assessment as a clinical endpoint, a depth-ranging sensor (Kinect) based upper extremity motion analysis system was applied to determine reachable workspace encountered in Pompe disease.

OBJECTIVE: To evaluate longitudinal changes in Kinect measured upper extremity reachable workspace relative surface area (RSA) in a cohort of patients diagnosed with Pompe disease.

METHODS: Ten patients diagnosed with late-onset Pompe disease (age range: 49-82 years) were monitored with Kinect reachable workspace RSA across visits spanning approximately 2.5 years. Changes in reachable workspace RSA are assessed using a linear mixed model.

RESULTS: Upper lateral quadrant RSA declined significantly per year by approximately 12.5% (F1,22 = 13.43, p=0.0014) versus the lower medial quadrant showing a slightly increased reachability of approximately 13.4% (F1,22 = 10.75, p=0.0034) and the upper medial quadrant RSA declined significantly in one year by approximately 15.0% (F1,22 = 9.69, p=0.0051) during this time period.

SUMMARY/CONCLUSION: Upper extremity reachable workspace is a novel marker capable of objectively quantifying declines in upper extremity functional ability in patients with Pompe disease. The reachable workspace is an intuitive functional measure that may be useful as a clinical endpoint for the efficacy evaluation of upper extremity targeted therapeutics in Pompe clinical trials or serve as a longitudinal clinical monitoring tool for patients already on treatment.

Divya Reddy, MD, President’s Research Initiative Award Recipient
USE OF KINECT SENSOR TO DETERMINE REACHABLE WORK SPACE AFTER BOTULINUM TOXIN INJECTION FOR UPPER LIMB SPASTICITY
Karina Del Rosario, Linda Johnson (Sacramento, CA), Lisa Williams, Evan de Bie, Brian Barry (Santa Cruz, CA), Jay Han (Orange, CA)

INTRODUCTION: Validated outcome measurement tools to quantitatively evaluate upper limb function after use of botulinum toxin (BTX) for spasticity management are limited. Use of the Modified Ashworth Scale (MAS) has been the mainstay outcome measure for determining reduction in spasticity after BTX injections. However, demonstrating improvement in upper limb function and ability to perform daily tasks has proven more difficult. The Kinect based motion system uses motion sensing technology to track change in functional reachability and range of motion over time. With the Kinect system, determining and monitoring the upper limb mobility impairment as measured by reachable workspace may be a valuable outcome measure for upper limb function in clinical trials.

OBJECTIVE: To evaluate changes in upper limb range of motion using the Kinect sensor to measure reachable workspace after botulinum toxin injection in patients with upper limb spasticity secondary to intracranial events.

METHODS: This was a prospective, open-label, single center study. After meeting inclusion and exclusion criteria, patients with cerebral insult resulting in shoulder adductor and/or elbow flexor spasticity with a MAS greater than 1+ were recruited. Primary outcomes measures utilizing the Kinect sensor were used and secondary outcome measures using the MAS, Functional Observational range of motion checklist, and passive/active range of motion were obtained.

RESULTS: Four patients were evaluated using primary and secondary outcome measures at baseline and 2, 4, 8 and 12 weeks after botulinum toxin injection. Functional range of motion as determined by the Kinect sensor improved at 2, 8 and 12 weeks in one patient, and 2, 4, and 8 weeks in a second patient.

SUMMARY/CONCLUSION: The Kinect based reachable workspace outcome measure may be a useful tool to quantitatively monitor efficacy of BTX for spasticity management.

Karina Del Rosario, MD, President’s Research Initiative Award Recipient

SIZE INDEX REVISITED: VERIFICATION BY INCREASED NUMBER OF SUBJECTS
Masahiro Sonoo, Go Ogawa (Tokyo, Japan), Erik Stalberg (Uppsala, Sweden)

INTRODUCTION: Size index (SI) is a motor unit potential (MUP) parameter in concentric needle EMG, which can sensitively discriminate between control and neurogenic MUPs. At the 2012 AANEM meeting, the authors proposed the revised SI (rSI) using the logarithmic scale for area/amplitude and investigated the utility of SI and rSI for both neurogenic and myogenic MUPs. One limitation of that study was the small number of patients: 4-6 for each group. In this study, the authors greatly increased the number of patients, and the age was matched between control subject and patient groups.

OBJECTIVE: To verify the utility of SI and rSI using the expanded data.

METHODS: The biceps brachii (BB) and tibialis anterior (TA) muscles were investigated. The study population was 1619 control MUPs (26 subjects), 340 neurogenic MUPs (10 subjects), and 498 myogenic MUPs (14 subjects) in the BB and 1245 control MUPs (23 subjects), 566 neurogenic MUPs (19 subjects), and 473 myogenic MUPs (13 subjects) in the TA. The coefficient for area/amplitude was variously changed for the SI and rSI. The sensitivity was judged using 1 percentile and 99 percentile of control MUPs.

RESULTS: The maximum sensitivity of the SI for neurogenic MUPs using an appropriate coefficient was 84% for the BB and 66% for the TA. Similar sensitivity was achieved using the rSI. That for the rSI for myogenic MUPs was 9% for the BB and 25% for the TA. These were higher than any other conventional parameters.

CONCLUSION: By choosing appropriate coefficients, the SI (for neurogenic MUPs) and the rSI (for both neurogenic and myogenic MUPs) will enhance the utility of MUP analyses.

Masahiro Sonoo, MD, PhD, President’s Research Initiative Award Recipient
11 ELECTRICAL IMPEDANCE MYOGRAPHY OF FACE AND NECK MUSCLES: ESTABLISHING NORMATIVE VALUES
Courtney McIlduff, Sung Yim, Adam Pacheck, Seward Rutkove (Boston, MA)

INTRODUCTION: Many neurological disorders translate into oropharyngeal dysfunction and facial weakness. However, there are few ways to objectively measure underlying changes in muscle. As a modality that has sensitively detected neuromuscular conditions using limb measurements, electrical impedance myography (EIM) could offer new, quantifiable information about clinical status when applied to facial and neck muscles.

OBJECTIVES: (1) To establish normative EIM values in selected facial and neck muscles of healthy adults and (2) to determine the reliability of EIM measurements in sternocleidomastoid and masseter muscles.

METHODS: EIM measurements were performed on the frontalis, temporalis, masseter, orbicularis oris, and sternocleidomastoid muscles of all participants. Average EIM phase (±SD) was determined in each muscle at 50 kHz. Reliability was assessed by calculating the intraclass correlation coefficient (ICC).

RESULTS: EIM measurements were performed in a total of 42 healthy men and women ranging in age 22-71 years. By muscle, the average EIM values were: frontalis 4.8° (SD 0.8), temporalis 6.1° (SD 2.3), masseter 6.3° (SD 1.9), orbicularis oris 7.5° (SD 1.2), and sternocleidomastoid 12.6° (SD 4.2) in healthy participants. The intra- and inter-rater ICCs were 0.91 and 0.93 for the sternocleidomastoid and 0.96 and 0.88 for the masseter.

SUMMARY/CONCLUSION: This study provides preliminary normative values for facial and neck muscles using EIM; further, the technology facilitated very reliable measurements. Additional work is needed to validate these findings in healthy and diseased populations. EIM could ultimately offer quantitative biomarker data of orofacial health for use in the clinic and therapeutic trials.

Courtney McIlduff, MD, President’s Research Initiative Award Recipient

12 AUTONOMIC DYSFUNCTION IN SENSORY NEURONOPATHIES: A QUANTITATIVE SUDOMOTOR AXON REFLEX EVALUATION
Alberto Martinez, Ingrid Faber, Maximiliano Carneiro, Carlos Martins Jr, Guilherme Gasque, Jose Domingues, Anamarli Nucci, Marcondes França Jr (Campinas, Sao Paulo, Brazil)

INTRODUCTION: Sensory neuronopathies (SNs) represent a subgroup of peripheral neuropathies caused by dorsal root ganglia (DRG) damage that leads to the classical features of sensory ataxia and asymmetrical sensory deficits. This peculiar presentation is opposed to the classical pattern seen in polyneuropathies (symmetric length-dependent damage). Although dysautonomic signs have been described in SNs, the full spectrum remains unclear. The Q-Sweat (quantitative sudomotor axon reflex test) (WR Medical Electronics Co., Maplewood, Minnesota) evaluates sudomotor function and enables detailed characterization of autonomic dysfunction, but it has not been fully explored in SNs.

OBJECTIVE: To describe the sweat patterns of SN patients through Q-Sweat evaluation.

METHODS: Patients with clinical and neurophysiological criteria for SNs were included and underwent Q-Sweat evaluation. The sweat volume was measured after the iontophoresis of a 10% acetylcholine solution in the 4 standard points on both sides of the body. The obtained volumes were considered abnormal if they did not reach the fifth percentile considering age, stature, and sex.

RESULTS: Twenty-five patients (13/25 male) with SNs were included. The most common etiology was idiopathic (16/25). Mean age and disease duration were 51.7±10.8 and 11.5±9.7 years, respectively. All patients had an abnormal result at least 1 point tested by Q-Sweat, with a mean of 4.08 abnormal points/patient. Asymmetrical findings were present in 52%, and in 68% there was not a length-dependent pattern.

SUMMARY/CONCLUSION: SNs exhibit frequent Q-Sweat abnormalities and a both-side test protocol is relevant to confirm asymmetry. These findings also corroborate small fiber damage and its role in the SN pathophysiology.

Alberto Martinez, MD, President’s Research Initiative Award Recipient

Alberto Martinez, MD, IFCN North American Chapter Fellowship Award Recipient
RELIABILITY OF BEDSIDE ULTRASOUND OF MUSCLE AND DIAPHRAGM THICKNESS IN CRITICALLY ILL CHILDREN
Kay Wei Ping Ng (Singapore, Singapore), Alexander Dietz, Ryan Johnson, Michael Shoykhet, Craig Zaidman (St. Louis, MO)

INTRODUCTION: Skeletal muscle atrophy occurs in adults in the ICU in proportion to the degree of illness. It is unknown if similar atrophy occurs in children.

OBJECTIVE: To determine if ultrasound is sufficiently reliable to measure muscle atrophy in children in the ICU.

METHODS: Muscle thickness (MT) was measured in the biceps brachii, quadriceps, tibialis anterior (TA), and right diaphragm using ultrasound in 25 sedated and intubated children, ages 1 week-17 years. An experienced sonographer trained novice sonographers on the first 10 patients. Each examiner performed 3 consecutive measurements. Calculations included inter-rater reliability between each examiner’s average measurements, intra-rater reliability between highest and lowest measurements, and limits of agreement as 1.96 SD of the mean difference.

RESULTS: MT (cm) was: quadriceps (2.2±0.8), biceps brachii (1.6±0.6), TA (1.2±0.5), and diaphragm (0.1±0.04). Inter- and intra-rater reliability was higher for limb muscles (intraclass correlation [ICC] >0.9) than diaphragm (ICC 0.7-0.8) for both experienced and novice examiners. Intra-rater mean difference plus or minus the limits of agreement were similar for novice and experienced examiners and were: quadriceps (0.03±0.24), biceps brachii (0.03±0.17), TA (−0.01±0.16), diaphragm (0±0.04). Inter-rater mean difference plus or minus the limits of agreement were similar to intra-rater values and were: quadriceps (0.09±0.18), biceps brachii (−0.04±0.22), TA (0.02±0.14), and diaphragm (−0.005±0.03).

SUMMARY/CONCLUSION: Ultrasound can be used to reliably measure muscle thickness in critically ill children, including infants. Repeated MT measures using ultrasound can detect a 8-15% change in MT for limb muscles, and a 22-36% change in diaphragmatic thickness in critically ill children.

Kay Wei Ping Ng, MBBS, MRCP, Mmed, President’s Research Initiative Award Recipient

SOLEUS-H REFLEX STUDY IN SPASTICITY, DYSTONIA, AND CONTRACTURE IN CHILDREN: IMPACT OF BOTULINUM TOXIN INJECTION
Debabrata Ghosh (Columbus, OH)

INTRODUCTION: Soleus H reflex may distinguish spasticity from dystonia or contracture. There is paucity of data regarding H reflex changes following botulinum toxin (BoNT) administration.

OBJECTIVE: To study soleus H reflex in children with spasticity, dystonia, and contracture and its changes following BoNT injection.

METHODS: Children younger than 21 with hypertonia involving soleus/gastrocnemius muscles receiving BoNT were included. Parameters included: H latency, M latency, Hmax-amplitude, Mmax-amplitude, and H/M-ratio before and 6-12 weeks after BoNT. Mean values were calculated and appropriate t-tests used to compare the means before and after injection.

RESULTS: Of the 13 patients (7 male, 6 female; age range: 12.34±6.92 years), diagnoses included: spasticity—5 (9 limbs), dystonia—5 (6 limbs), and contracture—3 (6 limbs). H reflex was normal in dystonia or contracture. The spastic group had a higher H/M-ratio versus non-spastic: 1.72±2.3 versus 0.32±0.29 (t=2.33, p=0.03). Hmax-amplitude trended higher in spasticity (4485.29±6124.23 μV versus 3932.36±2868.91 μV) but p>0.05. OnabotulinumtoxinA dose in the soleus was 32±9.02 U. Followup (n=9) after 2.9±0.88 months: all improved regarding ankle range of movement with goal attainment scale of 0 in 1, 1+ in 4, and 2+ in 4 patients. H/M-ratio and Hmax-amplitude showed a downward trend (H/M-ratio 1.89±2.52 to 0.75±1.09 in spasticity subgroup), though statistically insignificant. H latencies remained unchanged.

SUMMARY/CONCLUSION: Higher H/M-ratio in soleus H reflex can distinguish spasticity from dystonia and/or contracture. There is a trend towards higher Hmax-amplitude in spasticity, however latencies are unchanged. BoNT injection tends to lower H/M-ratio and Hmax-amplitude without altering H latency.

Debabrata Ghosh, MD, President’s Research Initiative Award Recipient

Kay Wei Ping Ng, MBBS, MRCP, Mmed, President’s Research Initiative Award Recipient

Kay Wei Ping Ng, MBBS, MRCP, Mmed, Resident and Fellow Member Award Recipient
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INTRAVENTOUS IMMUNOGLOBULIN TREATMENT-RELATED FLUCTUATIONS IN CHRONIC INFLAMMATORY DEMYLINATING POLYNEUROPATHY PATIENTS USING DAILY GRIP STRENGTH MEASUREMENTS (GRIPPER): STUDY DESIGN AND UPDATE
Timothy Walton (Lenexa, KS), Jeffrey Allen (Minneapolis, MN)

INTRODUCTION: The optimal treatment approach for chronic inflammatory demyelinating polyneuropathy (CIDP) patients on chronic IVIg therapy is unknown.

OBJECTIVE: To describe an investigator-initiated, multicenter study that prospectively evaluates IVIg-related fluctuations.

METHODS: The primary outcome measure is grip strength, performed daily for 6 months. Weekly home nursing visits capture the Rasch-built Overall Disability Score (R-ODS), Timed Up and Go Test (TUG), Overall Neuropathy Limitations Scale (ONLS), Modified Fatigue Severity Scale (mFSS), and Visual Analog Pain Severity Scale (VAS). The Quality of Life Short Form Physical Component Summary® is collected at baseline, week 12, and week 24. Serum IgG levels are collected at 3 time-points surrounding IVIg infusions (peak, trough, and mid-cycle). Total recruitment is anticipated to be 30 subjects from 7 different sites. Upon study completion, "wear-off" frequency will be analyzed by assessing the proportion of subjects with grip strength (GS) and R-ODS intracycle fluctuation and the proportion of cycles in which fluctuation occurs. To determine the extent of "wear-off," the difference between maximum and minimum GS, R-ODS, TUG, ONLS, and VAS scores will be analyzed.

RESULTS: Currently, 11 subjects from 3 sites have been enrolled. This interim study report will provide preliminary representative data, demonstrating IVIg "wear-off" effects on GS and other outcome measures.

SUMMARY/CONCLUSION: It is expected that the results of this study will help facilitate development of CIDP treatment optimization strategies. In addition, this information should be important in forming hypotheses to be tested in future studies (i.e., comparing different dosage intervals, optimal IVIg taper guidelines, or assessing the longterm outcome of short-term cycle-to-cycle clinical fluctuations).

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PARANEOPLASTIC MOTOR NEURON DISEASE AND CEREBELLAR ATAXIA ASSOCIATED WITH ANTI-HU ANTIBODIES
Julie Bucklan, Jinny Tavee (Cleveland, OH)

INTRODUCTION: Anti-Hu antibodies are associated with various paraneoplastic syndromes (PNS) including cerebellar degeneration and rarely, motor neuron disease (MND). We report a case of paraneoplastic anti-Hu syndrome with features of both cerebellar ataxia and MND.

OBJECTIVE: To describe a case of paraneoplastic cerebellar degeneration and MND.

CASE REPORT: A 75-year-old woman with diabetes, remote uterine cancer status-post hysterectomy, and a 55-pack-year tobacco history presented with a one-year history of progressive generalized weakness, slurred speech, and right hand clumsiness in the setting of a 30lb weight loss. No cognitive or behavioral changes were noted. Neurologic examination demonstrated dysarthria, bilateral upper extremity ataxia, and asymmetric hyperreflexia. In the lower extremities, diffuse muscle atrophy, fasciculations and severe weakness with flail feet were seen. Electromyography (EMG) revealed active and chronic neurogenic changes in cervical and lumbar segments along with chronic tongue denervation. Anti-Hu antibody titers were elevated in both the serum (1:61,440) and CSF (1:256). Extensive diagnostic testing, including MRI of the CNS axis, whole body PET scan, anti-GAD antibody, and ALS genetic testing, was otherwise negative. No improvement was seen after administration of IVIG, and the patient expired within two months of the initial visit. No autopsy was performed.

CONCLUSION: This is the first case report of motor neuron disease and cerebellar degeneration occurring simultaneously in the setting of high titer anti-Hu antibodies. While rare, a paraneoplastic etiology should be considered in MND patients with rapidly progressive symptoms, multiple risk factors for malignancy and atypical neurologic findings as demonstrated in our case.
ABNORMAL NEURONAL MIGRATION IDENTIFIED BY BRAIN PATHOLOGY IN CONGENITAL MUSCULAR DYSTROPHY 1A
Himali Jayakody (Saint Petersburg, FL), Katherine Mathews, Steven Moore (Iowa City, IA)

INTRODUCTION/OBJECTIVE: Congenital muscular dystrophy type 1A (MDC1A), or merosin-deficient congenital muscular dystrophy, is caused by recessive mutations in LAMA2 and loss of laminin alpha 2 chain expression. Individuals with MDC1A typically have white matter signal changes on MRI; rarely, structural brain abnormalities are seen. Described here is the second published case of autopsy neuropathology in MDC1A.

CASE REPORT: A 17-year-old male who died of an esophageal hemorrhage was born at term and hypotonic at birth. He had normal cognitive milestones, sat independently, but never walked. Nocturnal bilevel positive airway pressure (BPAP) was started at age 4. LAMA2 sequencing identified 2 frameshift mutations: c.2049_2050delAG, p.R683Sfs*21 and c.8669dupT, p.L2890Ffs*16. MRI of the brain at age 2 showed abnormal white matter T2 signal, but was structurally normal. Postmortem evaluation revealed a diffuse, bilateral cobblestone appearance of the cerebral cortex including many sites of fusion between adjacent gyri. There were multifocal disruptions of the glia limitans, associated with mildly abnormal cortical lamination. White matter was normal. Subcortical nodular heterotopia and sites of failed cerebellar granule cell migration were seen in the cerebellum. Cobblestone pathology is more commonly associated with dystroglycanopathies; a shared mechanism is likely as alpha-dystroglycan binds to merosin at the glia limitans in the developing brain.

CONCLUSIONS: Together with the single previously published autopsy in MDC1A, this case suggests that cobblestone neuropathology can be present despite a structurally normal brain MRI. Thus, it is more common than imaging studies demonstrate, and this pathology could account for epilepsy seen in some MDC1A patients.

MYOSITIS AND MYASTHENIA IN CANCER PATIENT RECEIVING NOVEL CHEMOTHERAPY
Narges Moghimi, Sudhakar Tummala (Houston, TX)

INTRODUCTION: Myasthenia gravis (MG) can present with a predominant myositis clinical picture. This case highlights the importance of timely diagnosis of MG in cancer patients receiving novel chemotherapy.

CASE REPORT: A 66-year-old male with clear cell renal carcinoma treated with chemotherapies was recently started on AMP-514 (anti-programmed death-ligand 1 [PD-L1] immune checkpoint inhibitor) and anti-PD-1. After 2 cycles, the patient developed acute bilateral proximal muscle weakness with elevated creatinine kinase. Further evaluation revealed eye ptosis, limited extraocular movements, dyspnea, dysphagia, neck flexor weakness, and fatigable weakness in upper and lower proximal muscles. Clinical findings were concerning for neuromuscular junction disorder. Needle EMG findings showed myopathy with muscle membrane instability with no compelling findings for pre- or post-neuromuscular junction dysfunction on repetitive NCSs. Anti-acetylcholine receptor (AChR) and anti-striated muscle antibodies were positive at 2 separate 15 day time points prior to IVlg. Patient condition improved following total plasma exchange and initiation of IVlg.

RESULTS: Blocking PD-1 interaction with its ligands with immunotherapy alters immune checkpoints and enhances immune responses. This can lead to a wide variety autoimmune side effects, including myositis. It remains unclear if MG is uncovered or actually triggered by up regulation of immune responses with novel antibody immunochemotherapies. It also remains unclear if use of combination immunotherapy has further enhanced this complication. Presence of AChR and anti-SM antibodies can help identify underlying myasthenia in cancer patients who have developed myositis.

SUMMARY/CONCLUSION: This case highlights MG and myositis as an immune-mediated side effect related to new immune checkpoint inhibitor combination chemotherapy.

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NOVEL MUTATION IN SYNE1 CAUSING A MUSCULAR DYSTROPHY PHENOTYPE
Jennifer Tay, Maureen Galindo, Katalin Scherer (Tucson, AZ)

INTRODUCTION: Increased usage of next generation sequencing is revealing patients with novel mutations in known and unknown genes. Phenotype–genotype correlations are important to understand the significance of these mutations.

OBJECTIVE: To provide phenotypic description of a novel mutation in SYNE1 (Emery–Dreifuss muscular dystrophy type 4).

CASE REPORT: A 14-year-old boy presented with proximal lower extremity weakness. He is a product of a full-term, uncomplicated pregnancy, walking at approximately 12-13 months. Progressive proximal lower limb weakness began at the age of 3. At 8 years old, his workup revealed a normal creatine phosphokinase level. Muscle biopsy showed mild hypertrophy, fatty infiltration, and atrophy and was suggestive of a dystrophy. Needle EMG was myopathic without spontaneous activity. Weakness plateaued at age 14, and the patient is ambulatory with a waddling gait. A next generation sequencing panel of 30+ genes revealed a novel variant in SYNE1 (c.16360-9G>T heterozygous mutation in intron IVS85). Family history is negative; the parents are undergoing testing for the same variant at this time and will be reported on the final poster.

SUMMARY/CONCLUSION: The SYNE1 mutation, which is associated with Emery–Dreifuss muscular dystrophy, presents with a variable phenotype clinically similar to limb-girdle muscular dystrophy (pelvifemoral type).

EPIDEMIOLOGICAL DATA OF A REFERENCE CENTER FOR NEUROMUSCULAR DISORDERS IN THE SOUTH OF BRAZIL
Juliana Varela, Bianca Madeira, Pedro Schestatsky, Pablo Winckler (Porto Alegre/RS, Rio Grande do Sul, Brazil)

INTRODUCTION: The Neuromuscular Disorders Clinics is part of the Neurology Division of Hospital de Clinicas de Porto Alegre, a community-based university hospital located in Porto Alegre, Brazil. It is one of the main reference centers for neuromuscular diseases in the state of Rio Grande do Sul.

OBJECTIVE: To present the epidemiological data of a reference center for neuromuscular disorders of a community based hospital in the south of Brazil.

METHODS: Using clinical, neurophysiologic, and laboratory data, patients were divided in the following topographic groups of disorders: nerve, muscle junction, anterior horn cell, and muscle.

RESULTS: There were a total of 791 registered patients; 419 patients (53%) were classified as nerve disorders, 99 (12.5%) as anterior horn cell disorders, 168 (21.2%) as muscle disorders, and 95 (12%) as muscle junction disorders. Patients that were still in the investigating process (n=10; 1.3%) were classified as having an undefined diagnosis.

SUMMARY/CONCLUSION: The nerve disorders group was the largest and had 37 patients with hereditary neuropathy, but also patients with different complex diseases as well as common disorders such as diabetes polyneuropathy. For muscle disorders, it was a heterogeneous group including rare genetic dystrophies but also common diseases like Duchenne/Becker muscular dystrophies. In the muscle junction group, the most common disorder was myasthenia gravis (n=90), and in the anterior horn cell group it was amyotrophic lateral sclerosis (n=78). This is the first epidemiologic work of a reference center for neuromuscular disorders in Brazil. The authors consider it important to know and understand the local epidemiology and reality of their community.
CASE REPORT: A 47-year-old man was referred to the EDX clinic for diagnostic evaluation of numbness, tingling, and pain in both hands. During his history, the patient reported a prior EDX study in 2013 that diagnosed him with bilateral CTS. He reported symptoms of numbness of both hands without any trigger and right wrist pain and paresthesias injected recently with a steroid injection with some relief. Physical examination revealed (1) normal strength, sensation, and reflexes in both upper extremities, (2) positive Tinel, Phalen, reverse Phalen, and carpal compression tests at both wrists, and (3) positive Tinel test at both elbows. An EDX consultation was followed by bilateral upper extremity NCSs and neuromuscular ultrasound (NMUS) evaluation. The NCSs revealed prolonged bilateral median sensory and motor conductions across the wrists consistent with mild bilateral median neuropathy at the wrists. NMUS evaluation revealed a trifid median nerve at the distal portion of the left carpal tunnel, with a combined cross-sectional area (CSA) of 0.14 cm². Left forearm median nerve CSA was 0.05 cm². Right median nerve CSA at the wrist was 0.12 cm² and at the forearm was 0.06 cm². No other sonographic abnormalities or anomalies were noted in the right median nerve.

SUMMARY/CONCLUSION: An extremely rare anomaly of a trifid median nerve was noted on NMUS examination of a patient with electrodiagnostically-confirmed bilateral CTS. Awareness of this rare anomaly is important as the patient was preparing to undergo surgical decompression of bilateral carpal tunnels.
CLINICAL UTILITY OF UTILIZING THE SHORT HEAD OF THE BICEPS FEMORIS AND THE TIBIALIS POSTERIOR IN TRIGGERED NEEDLE ELECTROMYOGRAPHY FOR L5 PEDICLE SCREW PLACEMENT IN AN INTRAOPERATIVE SETTING
Kimberley Butler (Lebanon, CT), Patrick Doherty (New London, CT)

INTRODUCTION: Standard and recommended muscle selection for triggered needle EMG for L5 pedicle screw placement during intraoperative monitoring is fairly limited. An investigation was conducted into the clinical utility of utilizing both the short head of the biceps femoris (SHBF) and the tibialis posterior to evaluate stimulation thresholds of pedicle screws placed at the L5 level.

OBJECTIVE: To evaluate the clinical utility of a more sophisticated array of muscles during triggered EMG for L5 level pedicle screw placement.

METHODS: A total of 65 waveforms were evaluated and divided into the following categories: firing of the tibialis posterior only (category A), firing of the tibialis posterior and the tibialis anterior simultaneously (category B), firing of the tibialis posterior, tibialis anterior, and the SHBF (category C), and firing of multiple (>3) muscles including the tibialis posterior and the SHBF (category D). Threshold values in mA was then correlated to compound muscle action potential (CMAP) findings.

RESULTS: CMAPs recorded from the tibialis posterior only (category A) represented 1.3% of all waveforms evaluated. CMAPs recorded simultaneously from the tibialis posterior and the tibialis anterior (category B) were noted on 14% of all waveforms evaluated. CMAPs recorded from the combination of the tibialis posterior, tibialis anterior, and the SHBF (category C) represented 9%. CMAPs elicited from multiple muscles (category D) represented 18%.

SUMMARY/CONCLUSION: Utilization of recording from the tibialis posterior in correlation to other L5-innervated muscles (e.g., the tibialis anterior) during triggered EMG for L5 pedicle screw placement may increase sensitivity of stimulation thresholds specific to the L5 level.

Ana Calzada-Reyes (Habvana, Cuba)

INTRODUCTION: Diabetes and alcoholism are the most common etiologies of peripheral neuropathy in developed countries. The present study takes into consideration the duration of the disease, the metabolic control over it, and the presence (or not) of clinical symptoms of peripheral neuropathy in insulin-dependent diabetic patients to evaluate in which circumstances peripheral NCSs are really advisable.

OBJECTIVE: To establish the usefulness of NCSs in the evaluation of diabetic neuropathy (DN) according to the evolution time and the metabolic control.

METHODS: NCSs were performed in 82 type I diabetic patients. Median, ulnar, peroneal, and sural nerves were studied. NCS parameters were related to clinical evidence of DN, the disease duration, and the values of glycosylated hemoglobin. A linear model of variance analysis (ANOVA) with 2 factors (disease duration and the metabolic control over it) was performed to detect significant differences in the electrophysiological parameters.

RESULTS: Of the NCSs performed for this study, 76.8% were abnormal. Sensory NCSs were the most affected. The ulnar sensory nerve had the highest percentage of damage. Reduce amplitudes of responses or absences of potentials were the main abnormalities in sensory nerves. There was a high clinical–electrophysiological concordance. The highest incidence of abnormalities corresponded to patients with longer disease duration, especially those who did not have longterm appropriate metabolic control of their glycemic blood levels. The 2 factor ANOVA showed dependence of the NCS sensory parameters with the metabolic control and of the motor parameters with the disease duration.

SUMMARY/CONCLUSION: NCSs provide useful information in diabetic patients even in cases without clinical evident DN.

Kimberley Butler, CNCT, Technologist Member Award Recipient
Ana Calzada-Reyes, MD, MSc, AANEM Foundation for Research and Education Award Recipient
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DECREMENTAL RESPONSES OF REPETITIVE NERVE STIMULATION IN ALS
Bei Cao, Qian-Qian Wei, Ruwei Ou, Yongping Chen, Jing Yang, Yaqian Xu, Yan Zeng, Hui-fang Shang (Chengdu, Sichuan, China)

INTRODUCTION: The decremental responses following repetitive nerve stimulation (RNS) has been reported in amyotrophic lateral sclerosis (ALS). This difference on the decrement in RNS between ALS, myasthenia gravies (MG), other mimic of motor neuron disease (MND) and healthy controls (HCs) was controversial.

OBJECTIVE: To investigate these features of the decrements in ALS, by comparing with MG, HC, and mimic of MND.

METHODS: We included in 63 ALS, 45 MG, 20 HCs and 29 patients with mimic of MND. All participants were performed standardized nerve conduction studies (NCS) and RNS. The disease progression of ALS was calculated by the formula, which was (48-the ALSFRS-R score at the enrollment)/disease duration (from symptom onset to the enrollment time).

RESULTS: Significant decrements (>10%) in at least one muscle were observed in 29.3% of the ALS patients and 86.7% of MG. Compared with the other groups, the ALS group had lower amplitude of compound muscle action potential (CMAP) on median nerve, ulnar nerve and tibia nerve. Additionally, the CMAP amplitude of median nerve, ulnar nerve and tibia nerve was related to the CMAP change on RNS in APB, ADM and TA, respectively. However, this relation was not observed in MG, HCs and mimic of MND.

CONCLUSIONS: The decrement following RNS was not rare in ALS, though, the positive rate of RNS was higher in MG than ALS. Our finding may suggest that the mechanism of the decrement following RNS may be not only associated with "prejunctional" factor but also neuromuscular junction.

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CARPAL TUNNEL SYNDROME: EVALUATION WITH A VARIETY OF NERVE CONDUCTION STUDY TECHNIQUES
Lidia Charroo Ruiz (Habvana, China)

INTRODUCTION: The combination of NCS techniques provides a more comprehensive assessment of the median nerve to help diagnose CTS.

OBJECTIVE: To describe the NCS findings in patients with CTS evaluated in the author's laboratory and to identify the best NCS techniques that detected the abnormalities present in the sample group.

METHODS: A sample of 165 patients with suspected CTS was studied with conventional NCSs as well as transcarpal NCSs, the combined sensory index (CSI), and the ring finger technique. Seventy-nine patients were then studied 3 months after surgery.

RESULTS: Conventional NCSs revealed abnormalities in 62.3% of the patient group. Transcarpal NCSs revealed that the palm-transcarpal latency difference was the variable that detected the greater number of abnormalities (83.12%) in the distal to median nerve segment. The ring finger and CSI techniques had detection rates of 73% and 67%, respectively. An analysis of receiver operating characteristic (ROC) curves for the NCS techniques showed that the variables that better identified CTS in the sample group were the palm–transcarpal latency difference, the transcarpal conduction velocity, and the wrist–index finger velocity. The postoperative study showed improvement in the distal motor latency of the median nerve.

SUMMARY/CONCLUSION: These findings support the high value of the various NCS techniques in diagnosis of CTS, and it may be possible to conclude that the conventional NCS may detect unambiguous severe CTS, while the transcarpal NCS, the ring finger technique, and the CSI are able to detect abnormalities at earlier stages.

Lidia Charroo Ruiz, MD, Master in Neuroscience, IFCN North American Chapter Fellowship Award Recipient

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ELECTRODIAGNOSTIC TESTS DURING INTRAOPERATIVE NEUROPHYSIOLOGICAL MONITORING: EXPERIENCES IN DETECTION OF ALARM SIGNALS AND SIGNALS OF GOOD PROGNOSIS
Lidia Charroo Ruíz (Habvana, China)

INTRODUCTION: Intraoperative neurophysiological monitoring (IONM) is important to minimize neurological morbidity from operative manipulation.

OBJECTIVE: To contribute to the preservation of the nervous system and to provide a prognosis after surgery through the use of IONM.

METHODS: In this prospective study the protocol was adjusted according to type and extent of the nervous system injury. IONM was performed in 89 patients (adults and children) with injury of the cervical and lumbosacral column. EDX tests—needle EMG, motor evoked potentials, somatosensory evoked potentials, F waves, and mapping—were performed. With the Neurosign 100 and Digitimer D185 stimulators, it was possible to evaluate neural pathway and direct electrical stimulation of motor nerve and nerve roots.

RESULTS: The clinical and IONM findings showed that only 2 patients had neurological deficits after surgery, which corresponded to patients who showed changes, “alarm signals,” (e.g., increases in threshold, decreases in amplitude, and neurotonic discharges). There was also improvement in electrophysiological responses (amplitude and waveform) which was interpreted as a signal of a good prognosis. In general, some authors refer to the “warning criteria”; however, this study provides evidence of signs of good prognosis, which are supported by results obtained in EDX evaluations postsurgery. An index of alarm signals and good prognosis signals has been calculated.

SUMMARY/CONCLUSION: IONM is helpful in the assessment of signs of a good prognosis and is effective during surgery in adults and children, with IONM possibly contributing to the preservation of function of the nerve roots and spinal pathways using a protocol with different EDX tests.

Lidia Charroo Ruíz, MD, Master in Neuroscience, IFCN North American Chapter Fellowship Award Recipient

ELECTROMAGNETIC INTERFERENCE FROM NERVE CONDUCTION STUDIES ON A CONTINUOUS GLUCOSE MONITOR: A CASE REPORT
Brian Chau, Sarah Humbert, Derek Rodeback (Loma Linda, CA)

INTRODUCTION: Electromagnetic interference (EMI) affecting implanted medical devices is a possible risk during NCSs. Continuous glucose monitors (CGMs) such as the Dexcom® system (Dexcom, Inc., San Diego, CA) provide viewing on mobile devices with real-time monitoring, auditory alerts, and data trends through subcutaneous electrical signals.

OBJECTIVE: To report a unique case of NCS and CGM signal impairment correlation.

CASE REPORT: Two resident physicians were performing practice lower limb NCSs on each other during their EDX training rotation. One resident had recently started wearing a Dexcom G5 sensor on the right lower abdominal quadrant. Sensory, motor, and mixed studies were performed on the right lower limb of this resident. Upon conclusion of the study, it was noted the CGM receiver stopped recording glucose levels at approximately the same time as the NCSs commenced. No audible warning alarms were noted from the receiver during the study.

SUMMARY/CONCLUSION: EMI through volume conduction has been proposed as a source of concern during NCSs with cardiac devices, but no literature exists on NCS EMI on CGMs. As these devices become more widely used, patients may rely solely on them and a gap in monitoring may result in rare, but possible, complications. Currently, no precautions regarding CGMs and EDX studies are published. Clinicians may consider screening diabetics for these devices prior to EDX examinations. Further studies help establish more than a limited correlation, identify the vicinity of stimulation locations leading to possible interference, and improve patient education.

Brian Chau, MD, Resident and Fellow Member Award Recipient
INTRAOPERATIVE ELECTROPHYSIOLOGY EXAMINATION OF MEDIAN NERVE SHOWS THE SHORT TERM OUTCOME OF CARPAL TUNNEL RELEASE
Jie Chen, Lei Xu (Shanghai, China), Dong Tian (Shanghai, China)

INTRODUCTION: An intraoperative electrophysiology examination is used to observe the surgical effect of neurolysis. But few studies have discussed which components of the examination are more credible.

OBJECTIVE: To explore a reliable predicator of surgical outcomes of carpal tunnel release (CTR) by exploring the changes of pre-and intraoperative median nerve–abductor pollicis brevis compound muscle action potentials (CMAPs).

METHODS: Fifteen patients with CTS were involved in the study. An electrophysiology examination was carried out to record CMAPs of the abductor pollicis brevis muscle at the following moments: before CTR, right after brachial plexus block, immediately after CTR and tourniquet release, 1 minute, 3 minutes, 5 minutes, and 7 minutes after tourniquet release. Statistical analysis compared these parameters with preoperative values.

RESULTS: There were statistically significant changes in both amplitude and latency of the CMAP within 5 minutes after tourniquet release (p<0.05), while not much difference was seen after 5 minutes. Both the latency and amplitude of CMAP greatly improved (p<0.05). Moreover, the improvement of amplitude had more statistical significance (p<0.01).

SUMMARY/CONCLUSION: CMAP amplitude is a better predictor than CMAP latency to evaluate the effectiveness of median nerve decompression. It is more reliable to carry out the electrophysiology examination at least 5 minutes after tourniquet release.

UTILITY OF NEEDLE ELECTROMYOGRAPHY FOR THE DIFFERENTIATION BETWEEN GUILLAIN–BARRÉ SYNDROME AND CONVERSION DISORDER
Takashi Chiba, Keiichi Hokkoku, Junpei Yamamoto, Midori Kuwabara, Yasuomi Kawamura (Tokyo, Japan), Tatsuya Ito (Itabashi-ku, Tokyo), Yuki Hatanaka (Tokyo, Japan), Masahiro Sonoo (Itabashi Ku, Japan)

INTRODUCTION: Conversion disorder (CD), having nonorganic weakness, is an important differential diagnosis of Guillain–Barré syndrome (GBS) because both disorders may present with acute limb weakness. Although GBS patients show typical findings, such as depressed deep tendon reflexes and abnormalities in NCSs, these may not be apparent at the earliest stages. A recruitment pattern seen with concentric needle EMG can differentiate central weakness from neurogenic weakness, and therefore it may be useful for the evaluation of such patients, although there have been few reports from this standpoint.

OBJECTIVE: To investigate the role of recruitment pattern observed during needle EMG in discriminating between CD and GBS patients.

METHODS: Clinical and electrophysiological records were retrospectively reviewed for 3 GBS and 3 CD patients who were admitted to the authors’ department due to acutely-progressing limb weakness for whom needle EMG was conducted during the acute phase.

RESULTS: Initial NCSs were normal for all CD patients. Even for GBS patients, however, NCSs conducted 0-5 days from the clinical onset showed no or minimal abnormalities. Needle EMG was performed for 1-2 muscles presenting with moderate-to-severe weakness. A reduced recruitment pattern suggesting neurogenic change, few motor units firing at a frequency of 20 Hz or higher, was observed in all GBS patients. In contrast, poor activation suggesting central weakness was observed in all CD patients.

CONCLUSION: Recruitment patterns on needle EMG can contribute to the differentiation between CD and GBS at an early phase.
AN ATYPICAL CASE OF UPPER ARM WEAKNESS-MAN-IN-THE-BARREL SYNDROME: A CASE REPORT

Christopher Constantino, Teresita Joy Evangelista (Manila, Philippines)

INTRODUCTION: Brachial diplegia with intact leg and facial motor function describes someone inside a barrel, hence the term man-in-the-barrel-syndrome (MIBS). Its causes include cerebral ischemia, cervical myelopathy, and other etiologies. Prognosis is poor.

CASE REPORT: A 50-year-old man suffered transient loss of consciousness and a forehead laceration after a 9-foot fall. He had no deficits and was ambulatory. Three years later, he noted gradual, progressive weakness of his right hand. Nerve conduction velocity (NCV) tests showed reduced motor amplitudes of both median nerves and normal F wave latencies except on the right median nerve. Needle EMG showed increased insertional activities, spontaneous activities, few firing motor unit action potentials, and moderate-to-severe reduced recruitment, suggesting cervical radiculopathy with consequent axonal changes. MRI revealed focal cord signals at C5, diffuse disc bulge, and cord abutment at C4-5 and C5-6. Weakness progressed, eventually involving both upper extremities. One year after, he underwent anterior cervical corpectomy of C5 and fusion of C4-6, with iliac crest bone graft. Postoperatively, both arms had no motor function, but his legs, face, sensation, and bowel and bladder function were unaffected. He was lost to follow-up and was seen after 3 years with absent motor function and severe atrophy of his arms and shoulder girdle. Motor function of the legs and face, all sensory modalities, and bowel and bladder function were normal. He also noted loss of erection. Repeat EMG-NCV studies showed findings of bilateral cervical radiculopathy. Repeat MRI revealed fusion of C4-6, absence of disc bulge and cord abutment, and two small symmetrical hyperintense foci centrally located in both sides of C5.

SUMMARY/CONCLUSION: MIBS is characterized by brachial diplegia with intact leg and facial motor function, caused by several known etiologies. Prognosis is poor. This patient is a rare case of MIBS due to the unusual delayed onset and gradual progression of symptoms following trauma and the presence of erectile dysfunction.

Christopher Constantino, MD, AANEM Foundation for research and Education Award Recipient

A STUDY OF DYNAMIC F WAVES IN JUVENILE SPINAL MUSCULAR ATROPHY OF DISTAL UPPER EXTREMITY (HIRAYAMA DISEASE)

Chaojun Zheng (Shanghai, China), Lauren Del Prato (Syracuse, NY), Xiang Jin (Shanghai, China), Robert Weber (Syracuse, New York), Jianyuan Jiang (Shanghai, China), Yu Zhu (Syracuse, NY)

INTRODUCTION: Hirayama disease (HD), a focal motor neuron disease characterized by asymmetric weakness of the upper extremities, primarily affects young males of Asian descent. While the etiology of HD is unknown, current theories include compression of the cervical spinal cord from forward shifting of the dura during neck flexion. F wave studies performed in neck flexion may provide insight into the pathophysiology of this disease and have implications for clinical management.

OBJECTIVE: To analyze changes in upper limb F waves during neck flexion in patients with HD compared to patients with ALS and healthy control subjects.

METHODS: Forty-one healthy subjects and 38 HD and 24 ALS patients were included. Bilateral F waves were consecutively recorded 20 times in the neck standard position and after neck flexion for 30 minutes. The persistence, minimal latencies, chronodispersion, F/M ratio, amplitude of F waves, and repeater F waves were compared between the 2 neck positions.

RESULTS: During neck flexion, repeater F waves occurred in more HD patients, and both ulnar and median repeater F waves increased significantly, along with higher F/M ratios on the more symptomatic side (p<0.05). No differences in F wave measures were observed in control subjects and ALS patients during neck flexion (p>0.05).

SUMMARY/CONCLUSION: HD and ALS both present with focal muscle atrophy without sensory involvement. Statistically significant changes in F waves during neck flexion compared to the neck standard position only occurred in the HD group, which supports the theory that HD is likely a position-related dysfunction rather than an intrinsic spinal cord disease. Counseling patients with HD should include avoidance of prolonged neck flexion.
SELECTIVE AND BILATERAL INVOLVEMENT OF THE PERONEUS LONGUS MUSCLE IN LUMBAR SPINAL STENOSIS
Jorge Díaz-Ruiz, Alvaro Rodríguez-Lazaro, Edicson Ruiz-Ospina (Bogota, Colombia)

INTRODUCTION: Lumbar spinal stenosis usually affects multiple nerve roots and several muscles innervated by a specific root both in the lower limbs and in the paraspinal region.

CASE REPORT: A 77-year-old man, with no significant past medical history, presented with a chief complaint of low back pain that radiated to the thighs and calves, muscle weakness, and gait disturbance for 1 month. Physical examination revealed absent bilateral ankle jerk reflexes and weakness of plantar flexors (4/5) and foot evertor muscles (2/5). MRI showed lumbar spinal stenosis with cauda equina compression at L4-5 levels. Needle EMG demonstrated signs of denervation in the bilateral gluteus maximus, medial gastrocnemius, and peroneus longus muscles. There was no further evidence of any other muscles innervated by the L5 root or paraspinal muscles being affected. The patient was diagnosed with lumbar spinal stenosis, with bilateral involvement of the L5-S1 nerve roots, with only electromyographic evidence of the L5 level being affected and only clinical evidence of abnormality of the peroneus longus muscle.

SUMMARY: In lumbar spinal stenosis the muscle involvement can be selective and/or asymmetrical despite an MRI showing severe and diffuse involvement of the lumbosacral roots.

NEUROPATHY BY LEPROSY IN PATIENTS FROM A ENDEMIC AREA IN CUBA: ELECTROPHYSIOLOGICAL FINDINGS
Rivero Fernando Raúl (Bayamo, Cuba)

INTRODUCTION: In Cuba a National Program for Control of Leprosy has been implemented since 1962; however, there are regions with high endemicity.

OBJECTIVE: Patients with leprosy from an endemic area of the eastern region in Cuba were studied to characterize the damages appearing in peripheral nerves of these patients throughout their illness.

METHODS: NCSs of median, ulnar, peroneal, and sural nerves were conducted.

RESULTS: Patients were older than 18 years; the predominant clinical forms were Lepromatous and Dimorphous with 59% and 22%, respectively. Abnormal studies were found in 95.45%; 88.1% of them were sensorimotor dysfunctions. Demyelinating abnormalities were the main finding when the nature of the damages was taken into consideration. No patients showed pure motor lesions in their NCSs. Sensory fibers of the nerves displayed a high percentage of damages with 75%, 79.5%, and 86.4%, respectively, meanwhile in the motor ones only the peroneal nerve showed a similar index of dysfunctions with 77.3%. The NCS findings in the ulnar nerve and thickening of ulnar nerve enlargement were statistically related. Discriminant analysis of the NCS results showed a differentiation according to the degree of peripheral nerve damage: the group of patients with severe clinical signs and symptoms of leprosy neuropathy were significantly different from those with mild and moderate signs and symptoms. There were no statistical differences between these last 2 groups.

SUMMARY/CONCLUSION: A correlation was found between the severity of the signs and symptoms of leprosy multiple mononeuropathy and NCS findings.

Rivero Fernando Raúl, MD, IFCN North American Chapter Fellowship Award Recipient

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- Sensory Ganglionopathy
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TOXIC EFFECTS OF CHRONIC MERCURY EXPOSURE ON THE PERIPHERAL NERVOUS SYSTEM IN ELECTROCHEMICAL WORKERS
Yodeisy Ferrer González, Rodisnel del Toro Ramírez (La Habana, Cuba)

INTRODUCTION: Mercury is a heavy metal of known toxicity. It accumulates in the nervous system, thyroid, breasts, myocardium, liver, kidneys, and eyes, and may be associated with dysfunction of those organs. The clinical impact of smaller mercury exposures remains controversial.

OBJECTIVE: To evaluate the toxic effects of mercury on the PNS by using NCSs in workers from an electrochemical factory chronically exposed to mercury.

METHODS: Workers from an electrochemical factory (n=42) and healthy non-factory employee control subjects (n=20) participated in the study. Neurophysiological examinations included motor and sensory NCSs of both upper and lower limbs. To determine mercury exposure, urine samples were collected and mercury levels were assessed.

RESULTS: All the workers had normal urine levels, despite the time of exposition. Workers exposed to mercury had median, sural, and superficial peroneal sensitive action potentials (SAPs) with amplitude and area significantly slower than healthy control subjects. Superficial peroneal and median SAPs amplitude and area had a significant negative correlation with time of exposition to mercury.

SUMMARY/CONCLUSION: NCSs can be useful for evaluating the toxic effects of chronic exposure to mercury.

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RETROSPECTIVE REVIEW OF ELECTRODIAGNOSTIC CHARACTERISTICS OF CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY CASES WITH AND WITHOUT DIABETES
Krishna Pokala, Alexander Gevorgyan, Said Beydoun (Los Angeles, CA)

INTRODUCTION: Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) is reported to have different electrodiagnostic findings when it is associated with Diabetes Mellitus (DM) but this has not been rigorously tested.

OBJECTIVES: To examine the distinguishing electrodiagnostic characteristics between CIDP patients with DM and CIDP patients without DM.

METHODS: Medical records of 54 CIDP patients seen at the neuromuscular division of Keck Medical Center and LAC+USC Hospital at USC (Los Angeles, CA) from 2005-2015 were retrospectively reviewed. Patients were identified and categorized and divided into two groups: CIDP patients without DM (CIDP-DM; n=47) and CIPD patients with DM (CIDP+DM; n=7). Analysis of electrodiagnostic values in these two groups was performed according to strict EFNS/PNS electrodiagnostic criteria for CIDP.

RESULTS: 1. CIDP+DM cohort demonstrated lower percentage (71.4% versus 82.9%) of “Definite CIDP” according to strict EFNS/PNS criteria. 2. CIDP+ DM cohort demonstrated more axonal loss on sensory nerve conductions manifested by lower amplitudes and a higher percentage of “No Response”. 3. CIDP+DM cohort demonstrates lower percentage of length-dependent demyelinating features (28.5% versus 61.7%)

CONCLUSION: The CIDP+DM cohort has some characteristic nerve conduction study findings when compared to the CIDP-DM cohort, including lower percentages of “Definite CIDP,” more axonal features on the sensory nerve conduction studies, and fewer length-dependent demyelinating changes. This is somewhat unexpected as DM has a known association with length-dependent changes seen on nerve conduction studies.
CONVENTIONAL NERVE CONDUCTION STUDIES IN INDIVIDUALS WITH PREDIABETES
Javier Gutierrez, Ana Conesa, Rafael Perez-Lalana, Yaima Fabregas-Deulofeo, Reinel Alvarez-Plasencia, Manuel Licea-Puig (La Habana, Cuba)

INTRODUCTION: Previous studies have demonstrated that individuals with prediabetes can suffer several types of a peripheral neuropathy. However, the specific features of this neuropathy are not fully described. NCSs can help to quantify and characterize this disorder.

OBJECTIVE: To evaluate NCS results in individuals with prediabetes.

METHODS: Conventional NCSs were performed in 26 individuals with a laboratory diagnosis of prediabetes (fasting glucose: 5.6-6.9 mmol/l or 2-hour glucose: 7.8-11 mmol/l). Motor NCSs and F waves were performed on ulnar, peroneal, and tibialis posterior nerves. Sensory NCSs were performed on ulnar, peroneal, and sural nerves. NCS results were compared to the authors' normal reference values database.

RESULTS: Completely normal results were reported in 10 subjects (38.4%). Signs of peripheral demyelination (either sensory or motor) were identified in 10 subjects (38.4%), while signs of axonal damage (either sensory or motor) were recorded in 6 individuals (23%). Motor NCSs were abnormal in 9 subjects (34%) while sensory NCSs were abnormal in 8 subjects (30.7), however only 3 patients had mixed sensory and motor disorders. Abnormal F waves were found in 10 subjects (38.4%). NCS abnormalities were rated as mild in 15 (57.6%) and severe in only 1 subject.

SUMMARY/CONCLUSION: These results confirm that approximately half of patients with prediabetes may have mild but definite peripheral neuropathy. Surprisingly, demyelination was slightly more common than axonal damage, while sensory and motor disorders showed similar incidence. These findings suggest that the peripheral neuropathy associated with prediabetes could have different profiles of presentation.

Joel Gutierrez, MD, PhD, IFCN North American Chapter Fellowship Award Recipient

ELECTROPHYSIOLOGIC CHANGES IN HUMAN MUSCLE FOLLOWING A TRAUMATIC TEAR: A CASE STUDY
Sandra Hearn, Hassen Berri, James Richardson (Ann Arbor, MI)

INTRODUCTION: The electrophysiologic response and associated EDX findings in human muscle after a traumatic tear remain poorly defined. Denervation potentials have been reported following semimembranosus tears in humans, and a rat model has shown a chronological trajectory of denervation followed by reinnervation, in a proximal-to-distal fashion.

OBJECTIVE: To characterize electrophysiological changes following a human gastrocnemius muscle tear over 15 weeks.

CASE REPORT: A 30-year-old man sustained an MRI-confirmed grade 3 tear of the medial gastrocnemius at the musculotendinous junction. He reported no neurologic symptoms. Five weeks post-injury, he had symmetric Achilles reflexes and robust, symmetric tibial motor NCS amplitudes. Needle EMG of the affected and contralateral (control) gastrocnemii was performed and recorded at 5 weeks post-injury. Five areas in the affected muscle and 2 areas in the control muscle were studied. At 15 weeks post-injury, needle EMG of the affected muscle was repeated. Recordings were then de-identified and analyzed. At 5 weeks post-injury, the affected gastrocnemius displayed increased insertional activity as compared with the control gastrocnemius. By 15 weeks post-injury, insertional activity had diminished to approach the level observed in the control sample in all but 1 area of the affected muscle. However, motor unit action potentials showed chronic neurogenic morphological changes not previously observed at the 5 week post-injury time point.

SUMMARY/CONCLUSION: This case study illustrates electrophysiological findings of increased insertional activity followed by reinnervation changes in a human muscle after local trauma. EDX physicians unaware of this phenomenon are at risk for making erroneous interpretations and inaccurate diagnoses.
L3 AND L4 INNERVATION INVESTIGATED FROM PATIENTS WITH SINGLE-ROOT RADICULOPATHY
Mana Higashihara, Yuki Hatanaka, Go Ogawa (Tokyo, Japan), Toshio Fukutake (Kamogawa, Japan), Shigeo Murayama, Masahiro Sonoo (Tokyo, Japan)

INTRODUCTION: It has been generally supposed that the rectus femoris (RF) and the vastus medialis (VM) muscles are mainly innervated by L3 and L4 roots equally, and the iliopsoas mainly by L2-3.

OBJECTIVE: To determine myotomal innervation of the femoral muscles based on the clinical and needle EMG findings of patients with single-root lesions.

METHODS: Clinical and needle EMG records of patients with L3 or L4 single-root radiculopathy confirmed by lumbar MRI were retrospectively reviewed.

RESULTS: Enrolled were 4 patients with L4 radiculopathy and a patient with L3 radiculopathy. Neurologically, weakness of the iliopsoas was observed in 2 patients with an L4 lesion and the quadriceps was weak in a patient with an L3 lesion. All of the patients with an L4 lesion showed hypesthesia or pain in the medial aspect of lower leg. A patient with an L3 lesion presented with hypesthesia in the medial part of the distal thigh and knee. During needle EMG, profuse denervation potentials were observed in the RF in all patients with L4 lesions, while the VM muscle showed no evidence of denervation. Needle EMG of the iliopsoas was conducted in 2 patients with an L4 lesion and profuse denervation was present for both patients. The VM muscle showed chronic denervation with denervation potentials at rest in a patient with an L3 lesion, while there was no evidence of denervation in the RF muscle in this patient.

SUMMARY/CONCLUSION: L4 innervation of the RF and iliopsoas muscles, and L3 innervation of the VM muscle, were suggested from this study. Such information for myotomal innervation will improve localization for electrodiagnosis.

COMPARISON OF MUSCLE ULTRASOUND FINDINGS BETWEEN CHRONIC INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY AND AMYOTROPHIC LATERAL SCLEROSIS
Keiichi Hokkoku, Masahiro Sonoo, Takashi Chiba, Chizuko Oishi, Hiroshi Tsukamoto, Yuki Hatanaka (Tokyo, Japan)

INTRODUCTION: It has been documented that denervation causes increased echo intensity (EI) and decreased muscle thickness (MT) in muscle ultrasound (MUS). Demyelinating neuropathies, including chronic inflammatory demyelinating polyradiculoneuropathy (CIDP), would not show denervation unless secondary axonal degeneration occurs. Hence, MUS may help discriminate CIDP from ALS causing axonopathy.

OBJECTIVE: To compare MUS findings between CIDP and ALS using quantitative assessments.

METHODS: Subjects consisted of 13 CIDP patients and 17 ALS patients, along with 60 normal control (NC) subjects. MUS was conducted in the abductor pollicis brevis (APB), abductor digiti minimi (ADM), and first dorsal interosseous (FDI) muscles in each patient. EI and MT of the examined muscles were quantitatively measured. The raw value was converted into the Z-score calculated from the corresponding sex and age group, and the data for 3 muscles were pooled together.

RESULTS: The number of examined muscles were 36, 51, and 180 for the CIDP, ALS, and NC groups, respectively. The EI and MT of the CIDP patients were not significantly different from those of the NC group. The ALS group showed significantly higher EI and lower MT compared to the CIDP group and the NC group (p<0.001 for all combinations).

SUMMARY/CONCLUSION: These results suggest that CIDP patients do not show MUS changes due to the absence of denervation. These MUS features may be useful in discriminating between CIDP and ALS.
OXYNEUROGRAPHY STUDY OF TOURNIQUET-INDUCED CHANGES IN NERVE TISSUE OXYGENATION
Joe Jabre (Los Angeles, CA), Srinivas Pyati, Karthik Raghunathan (Durham, NC)

INTRODUCTION: Tourniquet application, routinely used to reduce perioperative bleeding, induces endoneurial ischemia. This may in turn promote chronic postsurgical pain.

OBJECTIVE: To test the hypothesis that tourniquet-induced endoneurial ischemia can be assessed via measurement of nerve tissue oxygenation using the oxyneurography (ONG) technique (Jabre and colleagues 2012), a ratio of oxygenated over total hemoglobin concentration in nerve tissue acquired via near infrared spectroscopy (NIRS).

METHODS: Ten healthy volunteers were recruited. The Fore-Sight Oximeter was used to measure the ONG in the ulnar and common peroneal nerves at baseline. Sensory nerve conduction threshold (sNCT) measurements in areas known to be innervated by these nerves were also recorded. Changes were then assessed in ONG and sNCT after tourniquet inflation (to pressures exceeding systolic blood pressures), after 3 and 5 minutes, and then again after tourniquet deflation.

RESULTS: Following tourniquet inflation in both the upper and lower extremities, a significant drop in the ONG index at 3 and at 5 minutes was observed when compared to baseline values. After tourniquet deflation, there was significant overshoot in the ONG (above the baseline value) with subsequent variation in time to return to baseline. In contrast to changes in the ONG, changes in sNCT were highly variable.

SUMMARY/CONCLUSION: Monitoring nerve tissue oxygenation intraoperatively using the ONG technique can be a valuable addition to limb surgical procedures where a tourniquet is used so as to alert the surgeon to endoneurial ischemia that may result in chronic postsurgical pain.

Carpal Tunnel Syndrome Electrophysiological Parameters Acquired Using the E-NORMS Technique
Joe Jabre (Los Angeles, CA), Joao Kouyoumdjian, Vanessa Fernanda Moreira Ferreira (Sao Jose Do Rio Preto, Sao Paulo, Brazil)

INTRODUCTION: The e-norms technique (Jabre and colleagues 2015) relies on identifying normal studies from a mixed normal and abnormal laboratory cohort.

OBJECTIVE: To use the e-norms technique to derive reference values for 3 median nerve absolute latencies.

METHODS: From October 2011 to February 2016, 443 female CTS patients (mean age: 52 years) and 285 female patients (mean age: 42 years) with normal NCSs were selected. CTS EDX studies were based on: (1) median sensory peak (MSP) latency to the middle finger, 14 cm (≥3.70 ms), (2) median mixed palm peak (MPP) latency, palm to the wrist, 8 cm (≥2.30 ms), and (3) median motor distal latency (MMDL), wrist to the abductor pollicis brevis, 8 cm (≥4.30 ms). If these 3 absolute latencies were normal, comparative latencies—sensory median to the ulnar ring finger, sensory median to the radial thumb, and/or mixed palmar median to ulnar—were studied.

RESULTS: The e-norms derived practical limits were MSP ≥3.70 ms, MPP ≥2.00 ms, and MMDL ≥4.00 ms. Sensitivity, specificity, positive predictive value, and negative predictive value were, respectively, 67.2%, 99.8%, 99.7%, and 70.7% for MSP; 90.7%, 92.8%, 92.0%, and 91.7% for MPP; and 61.9%, 99.6%, 99.5%, and 64.4% for MMDL.

SUMMARY/CONCLUSION: The e-norms technique proved useful in identifying normal values for NCs. The new reference values obtained using this method did not change sensitivity/specificity for MSL, increased sensitivity (52.3% to 90.4%) with mild loss of specificity (100% to 92.8%) for MPP, and increased sensitivity (50% to 61.9%) with mild loss of specificity (100% to 99.6%) for MMDL.
GUILLAIN–BARRÉ LIKE SYNDROME: A MANIFESTATION OF NELARABINE THERAPY
Bushra Javed (White Plains, NY), Prachi Kale, Anila Thomas, Brij Singh Ahluwalia (Valhalla, NY)

INTRODUCTION: Nelarabine is an FDA-approved antineoplastic purine analog used for the treatment of refractory or relapsed T-cell lymphoblastic leukemia or lymphoma. The side effects include peripheral neuropathy, myelopathy, dysautonomia, somnolence, and seizures usually within 10-12 days of therapy.

OBJECTIVE: To present a case of nelarabine-induced neurotoxicity which appears clinically and electrophysiologically similar to Guillain–Barré syndrome (GBS).

CASE REPORT: A 54-year-old man with a history of T-cell acute lymphoblastic lymphoma presented with a relapse of lymphoma. He was treated with nelarabine, cytarabine, mercaptopurine, and cyclophosphamide and then developed sudden onset of ascending weakness of the bilateral lower extremities, mild upper extremity weakness, and areflexia at 9 days post nelarabine therapy. Spinal imaging did not show evidence of cord compression or signal change. Cerebrospinal fluid results showed an elevated protein of 70 mg/dl and white blood cell count of 0 cells/mm³. NCSs of the lower extremities showed conduction block, prolonged distal motor latency, and absent F waves while reduced recruitment of normal appearing motor units was found on needle EMG, suggestive of an acute demyelinating polyneuropathy. He was treated with IVIg followed by plasma exchange with minimal improvement in symptoms.

CONCLUSION: In this patient with a clinical and electrophysiologic presentation similar to GBS after nelarabine therapy, treatment for an immune-mediated process was initiated with minimal improvement. Whether nelarabine toxicity is an aberrant immune response against myelin or chemotoxicity remains unknown. Awareness and early identification of this medication side effect could be utilized in screening and preventative strategies during treatment.

SIMULTANEOUS SMALL AND LARGE FIBER NEUROPATHY IN GUILLAIN–BARRÉ SYNDROME
William Jens, Aiesha Ahmed (Hershey, PA)

INTRODUCTION: Guillain–Barré syndrome (GBS) is an acute autoimmune reaction that attacks the peripheral nervous system and can have a variety of presentations depending on the localization of damage. Small fiber damage as well autonomic system dysfunction can result in dysautonomia. Typically, demyelination is noted on EDX testing but an axonal pattern can also be seen. Small fiber involvement can only be seen on autonomic testing, which is rarely performed.

OBJECTIVE: To report a case of simultaneous presentation of small as well as large fiber neuropathies with GBS.

CASE REPORT: A 20-year-old woman presented with ascending sensory loss and burning dysesthesias with classic preceding diarrheal illness 3 weeks prior to onset. She also reported subjective weakness and palpitations. Physical examination found no objective weakness but hyporeflexia with liable heart rate. GBS was confirmed with primarily demyelinating sensorimotor polyneuropathy on EDX studies. IVIg was given and she had improvement with sensory findings but issues with dysautonomia remained at discharge. These symptoms continued through followup, so autonomic testing was obtained. Sudomotor testing was positive, consistent with involvement of small nerve fiber, and heart rate testing was also abnormal, showing autonomic dysfunction.

SUMMARY/CONCLUSION: Small fiber neuropathies can be seen as a rare GBS subset. Also, autonomic instability can be a driver for mortality which is not usually explored during the standard GBS workup. Both of these are easily tested on autonomic testing and should be considered in the evaluation for appropriate patients.

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IS SURAL-SPARING PATTERN SEEN IN THE VERY EARLY STAGES OF GUILLAIN–BARRÉ SYNDROME?
Jingxia Dang, Jiaoting Jin, Fangfang Hu, Xing Qin (Xian, Shaanxi, China)

INTRODUCTION: Many studies have shown the sural-sparing pattern to be a useful EDX feature in evaluation of suspected Guillain–Barré syndrome (GBS). It is seen in axonal and demyelinating forms of GBS. However, it is unknown if this phenomenon occurs in the very early stages.

OBJECTIVE: To identify if the sural-sparing pattern is present in the very early stages of GBS.

METHODS: Standard EDX studies were performed in 51 clinically-defined GBS patients with a history of less than 4 days, between 4-10 days, and more than 10 days. Those with GBS variants and Miller–Fisher syndrome were excluded. The sural-sparing pattern was the focus of the studies and it was defined as a “spared” normal sural sensory nerve action potential (SNAP) with abnormal median or ulnar SNAPs (measured antidromically). Results were compared to age- and height-matched normal subjects.

RESULTS: Eleven patients (7 male, 4 female; mean age: 48.0±11.2 years; age range: 25-61 years) had a history of less than 4 days. The sural-sparing pattern was present in 9 (81.8%), which included 3 with acute inflammatory demyelinating polyneuropathy, 6 with acute motor axonal neuropathy/acute motor-sensory axonal neuropathy, and 2 unclassified cases. In those with a 4-10 day history (13/20, 65%) and more than 10 days (12/20, 60%), the sural-sparing pattern occurs but decreases over time.

SUMMARY/CONCLUSION: The sural-sparing pattern can occur in the very early stages of GBS. It reflects a pathological process in axonal and demyelinating conditions. When patients are suspected with GBS in the very early stages clinically, the sural-sparing pattern is crucial for diagnosis.

INTERACTION OF CATHODAL AND ANODAL STIMULATIONS IN NERVE CONDUCTION STUDIES
Takamichi Kanbayashi, Takaharu Yamauchi, Yosuke Miyaji, Masahiro Sonoo (Itabashi-ku, Japan)

INTRODUCTION: At the 2015 AANEM annual meeting, the authors suggested that the F-wave latency may be substantially shortened due to the anodal stimulation that occurs more than 20 mm proximal to the anode. This pitfall may be more problematic if the cathode is shifted away from the nerve trunk. During investigating this effect, the authors discovered an unexpected interaction between cathodal and anodal stimulations.

OBJECTIVE: To investigate the interaction of cathodal and anodal stimulations.

METHODS: Subjects were 10 healthy volunteers. Their ulnar nerves were stimulated at the wrist using the cathode-distal setting. The antidromic mixed nerve action potential (MNAP) recorded at the elbow was used as an indicator to infer the antidromic impulse. First, both cathode and anode were placed right on the nerve (0 mm shift), and the stimulus current values necessary to achieve the threshold and half size of the maximal MNAP for anodal stimulation were measured. Next, the cathode was shifted 4 mm and 8 mm away from, or perpendicularly, to the nerve, and the similar experiments were repeated.

RESULTS: The thresholds were 15.9±4.3, 13.2±3.2, 11.3±2.6, and 8.0±3.0 mA for 0, 4, and 8 mm and perpendicular settings, respectively. The stimulus current values necessary to achieve the threshold and half size of the maximal MNAP for anodal stimulation were measured. Next, the cathode was shifted 4 mm and 8 mm away from, or perpendicularly, to the nerve, and the similar experiments were repeated.

RESULTS: The thresholds were 15.9±4.3, 13.2±3.2, 11.3±2.6, and 8.0±3.0 mA for 0, 4, and 8 mm and perpendicular settings, respectively. The stimulus current value to achieve half size was 33.2±15.0, 23.9±6.6, 20.6±4.7, and 18.2±5.0 mA, respectively.

SUMMARY/CONCLUSION: The anodal stimulation became more likely to occur when the cathode was shifted away from the nerve. Hyperpolarization generated around the cathode must suppress the anodal stimulation.
INTRODUCTION: There is dearth of electrophysiological studies on pediatric ulnar neuropathy (PUN). OBJECTIVE: To analyze patterns of nerve injury in PUN. METHODS: A retrospective analysis was conducted on 49 children with PUN.

RESULTS: The mean age was 14 years; two-thirds were male. Half had traumatic etiologies. Sensory loss in the little finger was the prevailing complaint (89%). Predominant localization was at the elbow (55%), followed by the forearm (23%) and humerus (14%). In PUN localized at the elbow, slowing was seen in about half of the cases, while conduction block was seen in a quarter. In all cases, a diminished ulnar sensory nerve action potential (SNAP) was the most common abnormality (71%) with a median axon loss estimate (MAXE) of 62%. The dorsal ulnar cutaneous (DUC) SNAP was reduced in 55% with a MAXE of 43%. Abductor digiti minimi (ADM) and first dorsal interosseous (FDI) compound muscle action potentials (CMAPs) were reduced half of the time, with a MAXE of 30% and 28%, respectively. There was high correlation between ulnar SNAP MAXE and ADM MAXE (r=0.76, p<0.0001), FDI MAXE (r=0.81, p<0.0001) and DUC SNAP MAXE (r=0.60, p=0.0048). Neurogenic changes were seen in the ADM, FDI, flexor carpi ulnaris, and flexor digitorum profundus in 79%, 77%, 25%, and 35%, respectively. In proximal localizations, intrinsic hand muscles were more affected than proximal ulnar muscles in 52% of cases. Pathophysiology was demyelinating in 27%, axonal in 59%, and mixed in 14%.

SUMMARY/CONCLUSION: There is frequent axonal and fascicular injury in PUN. In proximal axonal lesions, sensory and motor fibers to distal muscles are predominantly affected, whereas in demyelinating lesions, slowing occurs twice as commonly as conduction block.

INTRODUCTION: In cervical spondylotic myelopathy (CSM), level with structural stenosis seems not correspond to neurological level of deficit. However, it has not been clearly demonstrated.

OBJECTIVE: To demonstrate agreement or discrepancy between structural stenosis level and abnormal neurological level in patients with CSM. Structural stenosis level was determined by MRI, and myotome with neurological deficit was determined by needle EMG.

METHODS: The results of needle EMG and MRIs in 17 patients with CSM were reviewed to show objectively the relationship between structural level of stenosis and neurological level with lower motor neuron involvement. Structural level was defined by spinal canal stenosis with definite cord compression. Neurological level was determined by myotomes with abnormal spontaneous activity.

RESULTS: In all but 1 patient, myotomes with abnormal spontaneous activity on needle EMG were lower by 1, 2, 3, or 4 levels (S1) than stenotic canal shown in MRI.

SUMMARY/CONCLUSION: Motor neuron involvement in CSM is not concordant with the structural lesion.
A CASE OF INFLAMMATORY NEUROPATHY ASSOCIATED WITH ANTI-MYELIN-ASSOCIATED GLYCOPROTEIN ANTIBODY AND SERIAL NEUROSONOGRAPHIC FINDINGS

Sang Beom Kim (Seoul, South Korea), Dong Suk Shim (Incheon, South Korea), Yang-Ki Minn, Byung-Ok Choi (Seoul, South Korea)

CASE REPORT: A 63-year-old man presented with 3 days of weakness of both hands. Neurological examination showed mild motor weakness of both arms and left leg (impossible heel gait and hopping). He complained of no sensory loss, but deep tendon reflexes were hypoactive in the upper limbs. NCSs revealed sensorimotor demyelinating polyneuropathy with conduction block (CB) over the left median, ulnar, and peroneal nerves. Cerebrospinal fluid showed slightly increased protein. After a diagnosis of Guillain–Barré syndrome, IVIg was started. Serum tests of anti-ganglioside antibody and anti-MAG antibody were only positive for anti-MAG. Serum and urine protein electrophoresis with immunofixation showed no monoclonal gammopathy. Neurosonography exhibited relative swelling of the bilateral proximal median nerves, right whole ulnar nerve, right sciatic nerve, and left tibial nerve at the calf, and bilateral peroneal nerves at the fibular heads. After IVIg, motor power was restored progressively but not completely. In followup NCSs, compound muscle action potentials were increased and CBs disappeared. Serial nerve ultrasound (US) showed mild decrements of cross-sectional areas of formerly enlarged nerves.

SUMMARY/CONCLUSION: A neuropathy with anti-MAG antibodies is an acquired demyelinating polyneuropathy with slowly progressive primary sensory impairment associated with gait ataxia and postural tremor at the upper limbs. The neurophysiological findings indicate a symmetric demyelinating damage with uniform slowing of nerve conduction velocity and prolonged distal motor latency, without CB. But in this case, the patient had no ataxia and NCSs showed multifocal CB. US in a patient with anti-MAG neuropathy often reveals nerve regional enlargement in and out of entrapment sites. Further prospective studies may clarify the evolution of US changes according to history, neurophysiology, and therapeutic response.

FIVE YEAR SERIAL NEUROSONOGRAPHIC FINDINGS IN PATIENTS WITH AXONAL CHARCOT–MARIE–TOOTH DISEASE

Sang Beom Kim (Seoul, South Korea), Dong Suk Shim (Incheon, South Korea), Yang-Ki Minn (Seoul, South Korea), Byung-Ok Choi (Seoul, South Korea)

INTRODUCTION: Ultrasonography has been used widely for identifying pathology of peripheral nerve.

OBJECTIVE: In this study, ultrasonographic findings in the median and ulnar nerves in patients with the axonal type of Charcot–Marie–Tooth (CMT) type 2A disease were compared annually for 5 years.

METHODS: Median and ulnar nerve ultrasound studies were performed in 19 patients with CMT2A, and 5 patients underwent nerve cross-sectional area (CSA) study annually or biennially for 5 years.

RESULTS: Median and ulnar nerve CSAs were similar or slightly increased in the followed CMT2A patients, compared with the first study. But, in clinically deteriorated patients with worsening difficulties of hand grip or gait nerve CSAs were moderately enlarged.

SUMMARY/CONCLUSION: In demyelinating polyneuropathies, which are hereditary or acquired, enlarged nerve CSA is well known. But change of nerve CSA in axonal neuropathy is controversial. In this study, moderate increase of nerve CSA in some CMT2A patients was found. Further studies involving more CMT2A patients are needed to elucidate the natural history and neurosonographic features of hereditary axonal neuropathies.
STIMULATION ACROSS THE TRANSVERSE CARPAL LIGAMENT IN CARPAL TUNNEL SYNDROME  
Stephen Kishner, Chadwick Murphy, Margaret Maxi, Malia Cali, Donald Mercante (New Orleans, LA)

INTRODUCTION: CTS is the most common entrapment neuropathy and is typically caused by compression of the median nerve under the transverse carpal ligament (TCL). Routine CTS studies do not typically stimulate distal to the TCL.

OBJECTIVE: To determine if stimulating distal to the TCL will provide useful clinical information.

METHODS: In this prospective study, 105 consecutive patients referred to the EMG laboratory to rule out CTS were chosen. They were asked if they had classical symptoms of CTS and for how long. Severity of CTS on NCSs was graded in a similar nature to Bland’s classification. After routine NCSs were completed, an additional median motor stimulation was performed distal to the TCL and its latency and amplitude were recorded.

RESULTS: Wrist-to-palmar latency difference was found to have a positive correlation with symptom duration (p=0.0230). Wrist-to-palmar amplitude difference did not have any correlation with symptom duration. Both the wrist-to-palmar latency and amplitude were shown to have a positive correlation with symptom presence (p=0.0357, p=0.0031). The strongest correlation made in this study was wrist-to-palmar latency difference to severity grade (p<0.001). No correlation was found between wrist-to-palmar amplitude difference and severity grade.

SUMMARY/CONCLUSION: The wrist-to-palmar motor latency difference across the TCL correlates with symptom duration and symptom presence, as well as severity grade. The wrist-to-palmar motor amplitude difference across the TCL correlates with symptom presence, but not with severity grade or symptom duration.

CHARACTERISTICS AND SIGNIFICANCE OF DOUBLETS ON NEEDLE ELECTROMYOGRAPHY  
Christopher Lamb, Devon Rubin (Jacksonville, FL)

INTRODUCTION: Voluntary doublets, or double discharges, are an electrophysiological phenomenon sometimes associated with metabolic derangements or neuromuscular conditions. The association of doublets with other neuromuscular diagnoses has not been fully described.

OBJECTIVE: To define the frequency, characteristics, and significance of doublets on needle EMG and their relationship to neuromuscular disease or metabolic abnormalities.

METHODS: A total of 232 consecutive patients examined by a single examiner referred for routine needle EMG were prospectively studied. The frequency of doublets in individual patients, specific muscles, neuromuscular conditions, electrolyte levels, and doublet characteristics were evaluated. Twenty-five subjects with doublets were matched to 25 non-doublet subjects by age, diagnosis, and gender.

RESULTS: Of 232 patients, 25 (10.7%) exhibited doublets for a total of 35 unique doublets analyzed. Mean patient age was 59 years (52% male). Of 1303 (2.5%) muscles studied, 32 exhibited doublets. Lower extremity and paraspinal groups represented 91% of muscles with doublets. Five patients had doublets in more than 1 muscle. Doublet frequency grouped by needle EMG diagnoses was: myopathy (4/12, 33.3%), ALS (4/13, 30.8%), axonal polyneuropathy (7/29, 24.1%), radiculopathy (5/38, 13.2%), and no disease (7/109, 6.4%). Doublets were seen in 2/3 patients with peroneal neuropathies and 1 patient with an obturator neuropathy, but 0/41 patients with median, ulnar, or other upper limb mononeuropathies. Differences in serum electrolyte concentrations between doublet subjects and matched subjects were not statistically significant.

CONCLUSION: Doublets occur in approximately 10% of patients, are more common in lower extremity and paraspinal muscles, and do not seem to be clinically correlated to a specific metabolic pathology or neuromuscular diagnosis.
A PRESENTATION OF AMYOTROPHIC LATERAL SCLEROSIS AND PARAMYOTONIA CONGENITA
Peggy Lazerow, Brett Morrison, Vinay Chaudhry (Baltimore, MD), Payam Mohassel (Bethesda, MD)

INTRODUCTION: Paramyotonia congenital (PC) is an autosomal dominant nondystrophic myotonia caused by a missense mutation in the SCN4A gene. ALS is a motor neuron disease that can be sporadic or congenital. Some genes implicated in the pathogenesis of ALS include SOD1 and c9ORF72.

OBJECTIVE: To report a patient with genetically-confirmed ALS and PC.

CASE REPORT: A 61-year-old woman had been followed for cold- and exercise-induced muscle cramps since age 29. She had clinical and electrophysiological myotonia, and an episode of cold-induced paralysis requiring intubation. Exercise and cold provocation produced a significant drop in compound muscle action potential (CMAP) amplitudes. She now presented with a 1-year history of progressive weakness and shortness of breath. Family history was significant for 4 generations with muscle cramps or respiratory distress. Examination showed asymmetric weakness, atrophy, percussion myotonia, and incongruously brisk reflexes. Vital capacity was 1.15 L (40% predicted).

RESULTS: Muscle biopsy demonstrated esterase positive angular and polygonal atrophic fibers. NCSs showed reduced motor amplitudes. Repetitive stimulation showed a decremental response in several muscles. Needle EMG demonstrated abnormal spontaneous activity with reduced recruitment, large and long motor units, and myotonic discharges. Genetic testing was positive for previously reported pathogenic SOD1 and SCN4A mutations. She died from respiratory failure within 6 months.

SUMMARY/CONCLUSION: This patient had typical symptoms of PC for many years before being diagnosed with ALS, both present in family history and confirmed with genetic testing. This study underscores the importance of detailed clinical evaluation and genotype–phenotype correlation, especially when 1 condition does not fully explain a patient’s symptoms.

EFFECT OF CONVERGENCE RTMS AND SCALP ACUPUNCTURE ON NEUROMOTOR FUNCTION IN ISCHEMIC STROKE RAT MODEL
Sam-Gyu Lee, Eun-Jong KIM, Ara Jo (Gwangju Metropolitan city, South Korea)

OBJECTIVE: To investigate the mechanism and the therapeutic effect of the combined stimulation intervention of repetitive transcranial magnetic stimulation (rTMS) and scalp acupuncture stimulation (SAS) in the cerebral ischemic stroke (MCAo) rat model.

METHODS: Seven male Sprague Dawley® rats were used and grouped as follows: A—normal; B—MCAo; C—MCAo with rTMS; D—MCAo with SAS; and E—MCAo with rTMS+SAS. The rTMS was delivered at M1. SAS was performed at GV21 and GB6. Intervention was performed from day 3 after stroke induction. The neurological deficit score (NDS) was measured before and after stroke induction. Grip strength meter (GSM) and ladder rung walking test (LRWT) were performed for neuromotor function measure at days 1, 7, and 14 after stroke. Hematoxylin and eosin (H&E) stain and immunohistochemistry, quantitative reverse transcription polymerase chain reaction (qRT-PCR), and western blot were performed.

RESULTS: The NDS of all rats was 0±0 before stroke induction and 3.85±0.59 after a day of stroke induction. Grasping power of GSM increased and the error ratio of LRWT decreased more in group E than each single stimulation group on day 7 post-stroke. The qRT-PCR revealed no significant differences in all groups. Brain-derived neurotrophic factor (BDNF) and neurotrophin-3 in immunohistochemistry and western blot study were higher in all 3 stimulation groups than in the control and MCAo groups, but N-methyl-D-aspartate receptor 1 (NMDAR1) and microtubule-associated protein 2 (MAP2) showed no significant differences in all groups.

SUMMARY/CONCLUSION: Single intervention and convergence stimulation intervention would be effective, but convergence intervention of rTMS and SAS revealed no more additive or synergistic effect to any single intervention.
THE EFFECT OF SCALP ACUPUNCTURE AND REPETITIVE TRANSCRANIAL MAGNETIC STIMULATION ON NEUROMOTOR FUNCTION IN PHOTOTHROMBOTIC STROKE RAT MODEL
Sam-Gyu Lee, Eun-Jong Kim, Ganbold Selenge (Gwangju Metropolitan City, South Korea)

OBJECTIVE: To investigate the effects of scalp acupuncture stimulation (SAS) and repetitive transcranial magnetic stimulation (rTMS) intervention on neuromotor function in the phototrombotic cerebral infarction (PCI) rat model.

METHODS: PCI was induced on M1 of the right frontal lobe of 60 male Sprague Dawley® rats. SAS was performed at the GV21 and GB6 acupoints of ipsilesional M1. Low-frequency rTMS was delivered to contralesional M1. The rats were sorted into groups: A—normal; B—PCI; C—PCI with SAS; and D—PCI with rTMS. A regional cerebral blood flow (rCBF) with a quantitative reverse transcription polymerase chain reaction (qRT-PCR) and an MRI of the brain were performed.

RESULTS: Limb withdrawal test showed that left forelimb function improved significantly more in groups C and D than in group B at week 4 post PCI induction. Histopathological study of ipsilesional M1 showed that the cellularity was more dense and eosinophilic cells were decreased more in groups C and D than in group B. Western blot revealed that brain-derived neurotrophic factor (BDNF) and microtubule-associated protein 2 (MAP2) were more increased in groups C and D than in group B at week 2, more increased in group C than in group B at week 5. Immunohistochemistry expressed more BDNF and MAP2 in groups C and D than in group B at week 5. qRT-PCR revealed that gene expression for BDNF and MAP2 increased more in groups C and D than in group B. MRI showed the focal infarction of right M1 after PCI induction, and the infarction volume progressively decreased in groups C and D than in group B from week 5 to week 8.

CONCLUSION: rTMS and SAS work effectively for individual intervention of neuromotor function of the PCI rat model. rTMS was more effective for neurogenesis.

ELECTRODIAGNOSTIC AND Q-SWEAT ABNORMALITIES IN PATIENTS WITH NGLY1 CONGENITAL DISORDER OF DE-GLYCOSYLATION DISORDER
Tanya Lehky, Christina Lam, Carlos Ferreira, Donna Krasnewich, Lynne Wolfe, William Gahl (Bethesda, MD), Camilo Toro (Rockville, MD)

INTRODUCTION: Congenital disorders of glycosylation (CDG) are a group of genetic disorders marked by errors in the glycosylation of proteins and other molecules. In 2012, the first congenital disorder of de-glycosylation (NGLY1-CDDG) was described. NGLY1 encodes for N-glycanase-1, an enzyme involved in the cytosolic degradation of glycoproteins. The clinical manifestations are a triad of hepatic disease, complex choreoathetotic movements and ophthalmological findings, plus a manifold of accompanying abnormalities including seizures, global developmental delay, constipation, and auditory and peripheral neuropathies (PN).

OBJECTIVE: In this study, patients with NGLY1-CDDG underwent multimodality testing including EDX and Q-Sweat (WR Medical Electronics Co., Maplewood, Minnesota), a commercial quantitative sweat measurement system.

METHODS: Subjects with NGLY1-CDDG were enrolled in the National Human Genome Research Institute study 14-HG-0071 “Clinical and Basic Investigations into Known and Suspected Congenital Disorders of Glycosylation” (NCT02089789), and 76-HG-0238 “Diagnosis and Treatment of Patients with Inborn Errors of Metabolism and Other Genetic Disorders” (NCT00369421). EDX studies included NCSs, needle EMG, and Q-Sweat.

RESULTS: Thirteen subjects (age range: 2-21 years, mean: 9.4 years) were evaluated. All subjects had abnormal NCSs; 8 had sensorimotor PN with demyelinative features, 2 had severe axonal PN, 2 had only mild slowing of conduction velocity, and 1 had a sensory neuropathy. Needle EMG showed chronic plus or minus active neurogenic changes in 11 subjects. Ten subjects had abnormal Q-Sweat responses, either in both upper extremities and lower extremities (4/13) or lower extremities alone (6/13).

SUMMARY/CONCLUSION: Patients with NGLY1-CDDG frequently present with sensorimotor PN and length-dependent autonomic small fiber dysfunction, marked by abnormal Q-Sweat results.
ELECTRODIAGNOSTIC STUDIES IN MOEBIUS SYNDROME AND OTHER CONGENITAL FACIAL WEAKNESS DISORDERS
Tanya Lehky, Reversa Mills, Carol Van Ryzin (Bethesda, MD), Andrea Gropman (Washington, DC), Hyun Cho (Bethesda, MD), Ethylin Wang Jabs (NYC, NY), Elizabeth Engle (Boston, MA), Camilo Toro, Irini manoli (Bethesda, MD)

INTRODUCTION: Moebius syndrome is defined as a congenital, non-progressive facial weakness with limited abduction of 1 or both eyes and normal vertical gaze. It is part of a spectrum of congenital cranial dysinnervation disorders affecting the facial (CN VII) and other cranial nerves (CNs). Facial weakness can also be a manifestation of congenital myopathies.

OBJECTIVE: To explore the utility of EDX findings in the differential diagnosis of individuals with congenital facial weakness.

METHODS: Subjects participated in “Study on Moebius Syndrome and Other Congenital Facial Weakness Disorders” (NCT02055248). EDX studies included the blink reflex and facial and peripheral nerve sensory and motor NCSs. Needle EMG was performed on several subjects.

RESULTS: Thirty-five subjects (age range: 10-68 years) were evaluated. Twelve Moebius and 11 hereditary congenital facial paresis (HCFP) patients had findings limited to low CN VII responses except for 1 Moebius patient with a motor neuropathy. Four patients with TUBB3 mutations had a sensorimotor polyneuropathy with demyelinating features. Seven patients had a primary muscle disease: Carey–Fineman–Ziter syndrome (3 patients), syndromic facial palsy with arthrogryposis and ophthalmoplegia (2 patients), myotonic dystrophy phenotype (1 patient), and 1 patient had an undetermined myopathy. One patient had hemifacial myohyperplasia without a CN VII neuropathy. Three Moebius patients exhibited mirror hand movements.

SUMMARY/CONCLUSION: Congenital facial weakness can present as isolated CN VII involvement (HCFP), affect multiple cranial nerves (Moebius), or be part of a more complex neuropathic or myopathic disorder. EDX testing of this patient population can assist characterizing the underlying pathogenesis.

AN ANALYSIS OF ELECTROPHYSIOLOGICAL FINDINGS IN 33,363 PATIENTS REFERRED TO THE ELECTROMYOGRAPHY LABORATORY IN THE FOSHAN HOSPITAL OF TRADITIONAL CHINESE MEDICINE
Ming Li, Yu Zhu (Foshan, China)

INTRODUCTION: Electrophysiological studies enhance knowledge of the types of neuromuscular pathology encountered in a busy hospital and can help improve preparedness and prevention strategies to deal with them.

OBJECTIVE: To investigate the types of neuromuscular pathology encountered in the Hospital of Traditional Chinese Medicine with a large orthopedic center at Foshan City, a heavy labor industrial city in southern China.

METHODS: A review of 33,363 records of patients referred for needle EMG studies over 7 years, between 2009 and 2015, was carried out to determine the type of pathologies found in these patients.

RESULTS: Studies were abnormal in 87.7% of the cases. The EDX diagnoses included: lumbosacral radiculopathy (16.7%), median neuropathy at the wrist (13.9%), traumatic multi-nerve injury (11.5%), polyneuropathies (8.7%), facial neuropathy (8.2%), ulnar neuropathy at the elbow (4.3%), cervical radiculopathy (3.7%), traumatic radial nerve injury (2.5%), traumatic peroneal nerve injury (2.3%), traumatic brachial plexus injury (2.3%), peroneal neuropathy (2.1%), motor neuron disease (1.5%), sciatic neuropathy (1.3%), and muscle disorders (0.9%). Overall, trauma accounted for 27% of the referrals while non-trauma accounted for 83%.

SUMMARY/CONCLUSION: The large percentage of patients with peripheral nerve pathology diagnosed through electrophysiological studies emphasizes the importance of establishing a qualified EMG laboratory. Determining the types of neuromuscular pathology encountered in a hospital setting can help hospital administration better understand the needs of departmental structure and staffing to better serve their patient population.

Ming Li, MD, 2016 IFCN North American Chapter Fellowship Award Recipient
Dystonia in Machado–Joseph Disease Is Associated with Brainstem Hyperexcitability
Alberto Martinez, Guilherme Gasque, Carlos Martins Jr, Ingrid Faber, Jose Domingues (Sao Paulo, Brazil), Marcelo Nunes (Sao Paulo, Brazil), Anamarli Nucci, Marcondes França Jr (Campinas, Sao Paulo, Brazil)

Introduction: Machado–Joseph disease (MJD/SCA3) is the most common autosomal dominant ataxia worldwide. MJD/SCA3-related dystonia is present in 5-33% of the patients and has unclear pathophysiology. The blink reflex recovery curve (BRRC) is a neurophysiological test considered a marker of brainstem excitability. As neuronal hyperexcitability has been suggested to underlie MJD/SCA3-related dystonia, the authors decided to evaluate BRRC in these subjects.

Objective: To evaluate whether patients with MJD/SCA3 and dystonia have brainstem hyperexcitability.

Methods: Patients with MJD/SCA3 and healthy subjects were invited to participate. They underwent a standard BRRC protocol with 3 stimuli pairs in the supraorbital fissure and crescent between stimuli intervals (200, 300, 500, 1000, and 3000 ms). The orbicular oculi muscle was the recording site and the area under the R2 curve for each stimuli pair for each interval was obtained. The mean value for the BRRC was compared between groups through linear regression with age as a covariate. P values <0.05 were considered significant.

Results: The subjects were divided into 3 groups: MJD/SCA3–dystonic (A: n=10), MJD/SCA3–non-dystonic (B: n=9), and control subjects (C: n=10). Mean age/disease duration were 36.8±11/8.2, 49.7±15/11.4 and 32.2±9.64/-years, respectively. Mean BRRC areas were 0.72, 0.57, and 0.48, respectively. Group A had a significantly higher result than B (p=0.048) and C (p=0.012), however groups B and C presented no significant difference (p=0.536).

Summary/Conclusion: These results indicate that brainstem hyperexcitability is present in MJD/SCA3-related dystonia and BRCC is a useful tool to identify those patients at risk of developing dystonia.

Alberto Martinez, MD, IFCN North American Chapter Fellowship Award Recipient

Does Electrophysiology and Treatment Response Differ in Idiopathic versus Diabetic Chronic Inflammatory Demyelinating Polyneuropathy?
Anza Memon, Sarah Madani, Bashiruddin K. Ahmad, Lonni Schultz, Kavita Grover, Ximena Arcila-Londono, Naganand Sripathi (Detroit, MI)

Introduction: Sensory electrophysiology and terminal latency index (TLI) differences have been described in various chronic inflammatory demyelinating polyneuropathy (CIDP) subgroups.

Objective: To evaluate sensory electrophysiology, TLI, and treatment response in idiopathic and diabetic CIDP.

Methods: In a retrospective review of 147 patients with CIDP who underwent EDX evaluation (January 2000-December 2015), 89 fulfilled electrophysiological criteria described by the ad hoc subcommittee of the American Academy of Neurology and Albers and colleagues. Excluded were patients (31) with acute inflammatory demyelinating neuropathy, hereditary sensorimotor neuropathy, vasculitis, and polyneuropathy with paraproteinemia. The remaining 58 patients were divided into idiopathic (40) and diabetic (18) groups. These groups were compared for age, sex, history of cancer, cerebrospinal fluid (CSF) protein, response to treatment, sensory response abnormalities, and TLI measurements using chi-square tests for binary and categorical variables and t-tests for continuous measures. All testing was at the alpha=0.05 level.

Results: Group differences for age, sex, history of cancer, CSF protein, and treatment response were not significant. Comparing TLI values in measurable responses, the difference between the 2 groups for tibial TLI was significant (p=0.012), with the idiopathic group having a lower mean as compared to the diabetic. TLI values differences for median, ulnar, and peroneal nerves were not significant. The difference in abnormal rates of sensory responses was significant for the sural nerve with the idiopathic group having a lower rate compared to the diabetic (80% versus 100%, p<0.05). No differences were noted for the ulnar, median, and radial nerves.

Conclusion: Tibial TLI and sural sensory responses have some value in differentiating the 2 groups. Larger prospective studies are needed to confirm these findings.

Anza Memon, MD, Resident and Fellow Member Award Recipient
THE IMPORTANCE OF ELECTRODIAGNOSTIC STUDIES IN EARLY RECOGNITION AND EVALUATION OF PATIENTS WITH RADICULOPATHIES
Anza Memon (Novi, MI), Sarita Maturu, Lonni Schultz, Arun Chandok (Detroit, MI)

INTRODUCTION: EDX testing is an extension of a neurological evaluation. Diagnosis of radiculopathy is based on clinical history, examination, imaging studies, and EDX testing.

OBJECTIVE: To attempt early recognition of radiculopathies on EDX testing and to compare EDX findings with spinal neuroimaging (CT/MRI).

METHODS: This was a retrospective study on patients with cervical and lumbosacral polyradiculopathies who underwent EDX evaluation within the last year. Patient demographics, reason for EDX evaluation, grading, radiological correlation recommendation, and review of pre- and post-EDX spinal imaging (CT/MRI) were recorded.

RESULTS: Patients were divided into cervical (n=76) and lumbosacral (n=106) groups. Radiculopathy was the most common reason to obtain EDX evaluation in the cervical (41.5%) and lumbosacral groups (63%). The second most common reason was CTS (36%) and peripheral neuropathy (35%) in the cervical and lumbosacral groups, respectively. A total of 51% of patients in the cervical and 36% of patients in the lumbosacral groups were graded mild. No grading was used in 35% of patients in the cervical and 48% of patients in the lumbosacral groups. Radiological correlation was recommended in 22% (41% obtained spinal imaging) in the cervical and 13% (21% obtained spinal imaging) in the lumbosacral groups. Finally, 69% of the cervical and 64% of the lumbosacral groups were found to have a radiological correlation on spinal imaging.

CONCLUSION: Comparison of EDX findings with an MRI in the evaluation of radiculopathies is complimentary. EDX examination helps find the functional abnormalities, whereas neuroimaging studies detect structural abnormalities. It is reasonable to add EDX testing whenever there is uncertainty or discrepancy between the spinal neuroimaging and clinical presentation.

Anza Memon, MD, Resident and Fellow Member Award Recipient

SPINAL ACCESSORY NEUROPATHY: ETIOLOGY, ELECTROPHYSIOLOGY, AND RECOVERY
Anza Memon, Bashiruddin K. Ahmad (Detroit, MI)

INTRODUCTION: Isolated spinal accessory neuropathy (SAN) is rare and associated with significant morbidity. Electrophysiological studies help in recognition of injury but not in predicting outcome.

OBJECTIVE: To identify etiologies, electrophysiology, and prognostication in patients with SAN.

METHODS: A retrospective review was conducted of patients with SAN over 10 years (2000-2016). Patient age, gender, side of injury, etiology, and clinical features were recorded. Initial and repeat electrophysiological studies, followup visits, and recovery data were collected.

RESULTS: A total of 31 patients (mean age: 51 years; 16 women, 15 men) with SAN (19 right, 11 left, and 1 bilateral) were divided into 5 groups: postsurgical (20), stretch injuries (4), idiopathic (4), intracranial (2), and radiotherapy induced (1). Pain, winging, and joint restriction were common in the postsurgical group. Trapezius wasting was more common in idiopathic SAN. Sternocleidomastoid (SCM) wasting was equally common in idiopathic and intracranial groups. Compound muscle action potential (CMAP) amplitude and reduced trapezius recruitment did not correlate with prognosis. CMAP amplitude improvement on repeat study did correlate with functional recovery. An abnormal SCM needle examination was found in 8/22 patients (8 reduced recruitment, 6 fibrillations), 6 due to extracranial pathology. Five of the patients had additional mononeuropathies (1 vagus, 1 hypoglossal, 2 suprascapular, 1 long thoracic). An initial MRI of the brain was normal in the patient with vagal involvement; subsequent MRI showed lymphoma.

SUMMARY/CONCLUSION: Initial CMAP amplitude and reduced recruitment do not correlate with SAN outcome. CMAP amplitude improvement on followup study correlates better with functional recovery. Outcome in postsurgical cases was poor. SCM and additional muscles should be sampled to exclude coexistent mononeuropathies. Polyneuritis cranialis patients warrant followup neuroimaging.

Anza Memon, MD, Resident and Fellow Member Award Recipient
A PITFALL OF THE ANTIDROMIC SENSORY NERVE CONDUCTION STUDY OF THE MEDIAN NERVE: PARTIAL STIMULATION AT THE PALM
Chizuko Oishi, Yosuke Miyaji, Takamichi Kanbayashi, Atsuro Chiba, Masahiro Sonoo (Tokyo, Japan)

INTRODUCTION: The antidromic sensory NCS following wrist and palm stimulations, recording from the index finger, is 1 of the tests to evaluate CTS, and it has an advantage that it can detect sensory conduction block (SCB). The authors have found a previously-undescribed pitfall for this test: partial stimulation at the palm. Two branches from the first and the second webspace (i.e., radial and ulnar branches) contribute to the sensory nerve action potential (SNAP). The radial branch is buried in the thenar eminence and is rather difficult to stimulate, and therefore partial stimulation of the ulnar branch may easily occur.

OBJECTIVE: To describe a pitfall of partial stimulation in antidromic sensory NCSs of the median nerve.

METHODS: Subjects were 6 healthy volunteers. The active recording electrode was placed at the proximal interphalangeal joint of the index finger, and the stimulation was given at 7 cm (palm) and 14 cm (wrist) proximally. In the palm stimulation, separate stimulation of each branch was attempted.

RESULTS: Selective stimulation of each branch was possible for all subjects. Isolated supramaximal stimulation of the ulnar branch was achieved at 8-15 mA, and spread to the radial branch did not occur up to 40 mA in 3 cases, which suggests that fallacious partial stimulation may easily occur. The palm-to-wrist amplitude ratio of the SNAP was 108±15% for partial stimulation and 77±6% following the stimulation of both branches.

CONCLUSION: Partial stimulation is a pitfall of antidromic sensory NCSs that may interfere with the evaluation of SCB.

END-PLATE SPIKES OR FUSIMOTOR SPIKES?
Juhani Partanen (Helsinki, Finland)

INTRODUCTION: End-plate spikes (EPSs) are often observed in needle EMG of relaxed muscles. They have a negative onset or brief positive onset. They are seen in “active spots” of a muscle, often with miniature end-plate potentials (MEPPs). They are supposed to be elicited by irritation of a motor nerve twig by the EMG needle and recorded postsynaptically by the same electrode.

OBJECTIVE: Ectopic motor nerve action potentials should spread to the whole motor unit. The aim of this study is to point out that the prevailing hypothesis is incorrect.

METHODS: EPSs were studied using 2-3 EMG needle electrodes in the given muscle in order to see spreading or propagation of these potentials. Also, activation methods were used, especially passive stretch of the muscle.

RESULTS: Using multiple needle electrodes longitudinally to the muscle fibers, it was observed that most EPSs were local with negative onset. Rarely, an EPS was propagating for a short or long distance. Sometimes a synchronous EPS was observed in another electrode transversal to the muscle fibers. Passive stretch of the muscle activated EPSs in the active spot.

SUMMARY/CONCLUSION: It is concluded that EPSs represent action potentials of intrafusal muscle fibers. Nuclear bag fibers have local junction potentials, which do not propagate and these potentials are represented as negative-onset non-propagating potentials. Potentials propagating for a short distance represent action potentials of nuclear chain fibers. EPSs propagating like MUPs represent beta motor unit potentials. Passive stretch of a muscle activates II-afferents causing spinal reflex activation of gamma and beta units, observed as EPSs.
QUANTITATIVE ANALYSIS OF THE SOUND WAVES PRODUCED BY ABNORMAL SPONTANEOUS POTENTIALS: DIFFERENCES BETWEEN MYOTONIC AND COMPLEX REPETITIVE DISCHARGES
Alexandre Recchia (Sao Paulo, Brazil)

INTRODUCTION: Morphology, stability, and firing pattern (FP) are some of the characteristics that allow the identification of myotonic discharges (MDs) and complex repetitive discharges (CRDs). However, what makes them more easily recognizable are the peculiar sounds produced by their respective FP. Therefore, a careful hearing of these potentials becomes more important than their viewing.

OBJECTIVE: To quantify the differences between sound waves produced by MDs and CRDs through analysis of their frequencies and correlate them with the pitch of the sound.

METHODS: Ten MDs and twelve CRDs were collected from distinct patients using concentric needle electrodes. After collected, 2 different small portions of each discharge of 2 seconds duration were isolated (20 MD/24 CRD samples). CRD samples were selected randomly, whereas MD samples, due to the irregular FP, were isolated in the beginning of each discharge and after an alteration in the firing rate. Once isolated, the sounds produced were recorded and submitted to a Fast Fourier Transform spectrum analyzer to make frequency domain measurements.

RESULTS: Mean frequencies and standard deviation in the CRD and MD groups were 823.08±99.13 Hz/886.02±76.71 Hz and 1198.9±69.76 Hz/1560.02±127.28 Hz, respectively. MD samples presented significant quantitative differences in the frequencies due to the characteristic irregular FP, unlike the CRD group that presented slight differences (regular FP). MD sound waves produced high audible frequencies and pitch when compared with CRD.

SUMMARY/CONCLUSION: The quantitative analysis of the sound waves produced by their respective FP allows an accurate differentiation of these 2 types of discharges, leading to an understanding of the human pattern of auditory recognition.

Alexandre Recchia, MD, IFCN North American Chapter Fellowship Award Recipient

ELECTROMYOGRAPHIC ASSESSMENT OF THE TEMPORALIS MUSCLE PRIOR TO A LENGTHENING MYOPLASTY IN PATIENTS WITH MOEBIUS SYNDROME
Francis Renault, Cyril Gitiaux, Bernard Sergent (Paris, France), Valérie Charpillet (Neuilly sur Seine, France), Marie-Paule Vazquez (Paris, France)

INTRODUCTION/OBJECTIVE: Temporalis muscle lengthening myoplasty improves tightening of the lips and rehabilitates smiling function for patients with congenital facial palsies. Because Moebius syndrome is heterogeneous, a careful evaluation is mandatory before deciding to perform myoplasty. This series shows the role of needle EMG for investigating temporalis muscle and trigeminal nerve motor functions.

METHODS: A retrospective study of 18 patients with no upward movements of the labial commissure was conducted. Needle EMG was used to study the temporalis muscle bilaterally. Analysis focused on the recruitment pattern of voluntary contraction and electrical silence or activities at rest. Traces were classified as normal, neurogenic, or low-amplitude. Functional outcomes of myoplasty were evaluated by measuring commissural upward movement (mm) and qualified as: high (≥10); medium (>5); or little (≤5).

RESULTS: Surgery was cancelled for 5 patients with abnormal needle EMG signs including neurogenic (3) or low-amplitude (2) traces. Myoplasty was performed in 7 patients (age range: 8-17 years), unilaterally (3) or bilaterally (4). Preoperative needle EMG was normal (3) or showed neurogenic (2) or low-amplitude (2) moderate changes. Followup period after surgery was 2-12 years. Functional outcomes were high (5), medium (1), or little (1).

SUMMARY/CONCLUSION: Needle EMG study of the temporalis can detect muscle denervation or atrophy, or dyspraxia, and guide the decision to encourage or discourage performing myoplasty, or enhance the rehabilitation program and make the patient aware of a possibly modest outcome.
SEROPositive myasthenia gravis with superimposed cerebellar degeneration: A case report
Dennys Reyes (Sunrise, FL), Efrain Salgado (Weston, FL)

Introduction: The presence of myasthenia gravis (MG) and cerebellar degeneration (CDG) has been reported in very rare cases of paraneoplastic syndrome. There is no available report of positive acetylcholine receptor antibodies (AChR-abs) in a patient with cerebellar degeneration neither prior reports of coexistent MG and CDG without evidence of a paraneoplastic process.

Case Report: A 33-year-old female with a past medical history of hydrocephalus after shunting (1992) presented with symptoms of MG (2000) after experiencing generalized weakness, diplopia, and right eye ptosis followed by CDG (2003) with progressive walking difficulties, ataxia, and dysarthria. She received a thymectomy. Neurological examination revealed ataxic dysarthria, hyporeflexia, no motor deficit, diminished vibratory sensation of the left hallux, dysmetric finger-to-nose and heel-to-chin, ataxic gait, and positive Romberg test. She was on daily prednisone and Mestinon®. A needle EMG study revealed normal repetitive nerve stimulation. Median nerve somatosensory evoked potentials (SEPs) were normal but tibial nerve SEPs showed very mild symmetrical cervical potential prolongation. Vitamin E, thyroid-stimulating hormone, and HbA1c were normal. Extensive paraneoplastic workup showed negative anti-Yo, anti-MaTa, anti-CV2, anti-Ri, anti-CAR, anti-GAD, anti-Hu, anti-Zic4, anti-NR1, and anti-alpha 3AChR abs. AChR-abs including the binding, blocking, and modulating were all positive. MRI of the brain with and without contrast demonstrated severe cerebellar atrophy consistent with congenital spinocerebellar ataxia, thinning of the corpus callosum, and a possible tectal cyst or glioma. The patient refused immunosuppressant therapy given her plans to become pregnant.

Conclusion: The autoimmune mechanism in MG could possibly result in cerebellar degeneration without paraneoplastic evidence.

DO patients with CARP AL TUNNEL SYNDROME NEED A NEEDLE ELECTROMYOGRAPHY EXAMINATION?
Mark Ross, Leslie Zuniga, Charles Gervais, Benn Smith (Scottsdale, AZ)

Introduction: There are different approaches to the EDX evaluation of patients suspected of having CTS. Some physicians believe that if a patient has clinical symptoms of CTS and NCSs provide evidence of median neuropathy localized to the wrist, then needle EMG examination (NEE) is not necessary. Some have advocated that ultrasound evaluation of the median nerve at the wrist should be the initial diagnostic test for patients suspected of having CTS. The most common EDX practice is to use both NCSs and the NEE for evaluation of suspected CTS.

Objective: To determine the number of patients who have EDX evidence of CTS on NCSs who also have concurrent NEE evidence of cervical radiculopathy (CR).

Methods: The Mayo Clinic Arizona EMG Laboratory database was searched over a 1-year period (2015-2016) for diagnostic codes indicating CTS alone and CTS plus CR.

Results: Of 667 patients with CTS, 595 patients had CTS alone (89%) and 72 had CTS plus CR (11%).

Summary/Conclusion: This series demonstrated that roughly 11% of patients seen with CTS have an associated CR. The EDX confirmation of CR can only be diagnosed by performing the NEE. If such patients had only NCS or ultrasound evaluation, the diagnosis of CTS could be established but the diagnosis of CR would be missed. The possibility of diagnoses of concurrent CR and CTS provides a strong argument in favor of performing both NCSs and the NEE in patients with suspected CTS.
PATIENT PREPAREDNESS FOR NEEDLE ELECTROMYOGRAPHY TESTING BY REFERRING PHYSICIANS, NEUROPHYSIOLOGY STAFF, AND AANEM EDUCATIONAL VIDEO

Julia Whitlock, Devon Rubin (Jacksonville, FL)

INTRODUCTION: Referring physicians provide patients with varying information about the needle EMG procedure. Appropriate pre-procedure education of patients may improve patient tolerance.

OBJECTIVE: To determine the extent of patient needle EMG preparation by the ordering physician, neurophysiology staff, and the AANEM patient education video and correlate that with pain.

METHODS: Patients undergoing needle EMG during January-March 2016 were voluntarily surveyed about patient preparation by (1) the ordering physician, (2) neurophysiology staff, and (3) the AANEM video (shown prior to beginning the study in 1 group). Surveys included patient assessment of preparedness and pain. The frequency and degree of preparedness and pain among the groups was compared.

RESULTS: Of 142 patients surveyed, 57% were ordered by a neurologist. According to the survey, 42% of all patients (31% neurologist-referred; 59% non-neurologist referred) indicated that the needle EMG was NOT explained by their ordering physician. When explained, the explanation was “mostly” or “extremely” accurate in 38% based on the referring physician’s explanation, 86% by neurophysiology staff explanation, and 74% by AANEM video. Additionally, 41% of patients who received an accurate explanation from their physician and 83% of those who received information from the neurophysiology staff still indicated interest in watching a video. There was no significant difference in the degree of pain between patients who did and did not receive explanations from physicians or in patients who did and did not view the educational video.

SUMMARY/CONCLUSION: Referring physicians frequently do not explain needle EMG to patients. Proper explanation by the ordering physician, neurophysiology staff, or AANEM educational video may improve patient satisfaction with the study.

CLINICAL AND RADIOGRAPHIC CORRELATES IN AN INTERESTING CASE OF OCULOPHARYNGEAL MUSCULAR DYSTROPHY

George Jakubek (Jacksonville, FL), Jonathan Smith (Bethesda, MD), Mark Landau (Laurel, MD)

INTRODUCTION: Oculopharyngeal muscular dystrophy (OPMD) is an autosomal dominant or recessive muscular dystrophy affecting 1/100,000 individuals worldwide with a late onset. Although this disease is well described in the literature, there is scant documentation of prominent atrophy of proximal lower extremity muscles.

OBJECTIVE: To highlight an interesting case of OPMD and correlate proximal lower extremity weakness with radiographic findings.

CASE REPORT: A 75-year-old man presented 17 years after onset of extraocular muscle weakness and dysphagia and was diagnosed with OPMD. Eight years later, he reported difficulty ascending stairs and non-radiating lower back pain over a 6-month period. Workup for back pain with plain films revealed only an incidental pancreatic cyst; he was sent for a lumbar spine MRI and referred back to neurology. On examination, the patient had bilateral ptosis, restricted extraocular movement, and hypophonic voice. Marked atrophy was noted in proximal lower extremities, however no fatigable weakness and MRC 4/5 muscle strength in the proximal upper and lower extremities. Reflexes were brisk at the patella but otherwise normal. Needle EMG revealed increased insertional activity with myopathic units in the left tensor fascia lata. Although MRI was initially non-diagnostic, CT of the pancreatic cyst noted atrophy of bilateral psoas muscles and extensive fatty replacement of the gluteus and iliacus muscles.

CONCLUSION: OPMD usually manifests with weakness of the extraocular and pharyngeal muscle groups, but, as time goes on, proximal muscle groups can be involved in the upper and lower extremities. This case highlights the important clinical and radiographic findings of OPMD.
AUTONOMIC NERVOUS SYSTEM CHANGES ASSOCIATED WITH RHEUMATOID ARTHRITIS: A CLINICAL AND ELECTROPHYSIOLOGICAL STUDY
Hussein Sultan (Alexandria, Egypt)

INTRODUCTION: The authors have noticed some autonomic manifestation in patients with rheumatoid arthritis (RA).

OBJECTIVE: To evaluate clinically and electrophysiologically the autonomic nervous system changes associated with RA.

METHODS: The present study included 25 patients with RA (22 women, 88%) and 30 apparently healthy control subjects (27 women, 90%). A thorough clinical examination was carried out. Disease activity and functional disability were assessed. Tests for assessment of autonomic functions included active and passive orthostatic stress tests and the sympathetic skin response (SSR). The presence of abnormality in 2 or more tests was a clue for the presence of autonomic neuropathy (AN). Sural sensory NCSs and posterior tibial motor NCSs were performed.

RESULTS: There was a statistically significant decrease in standing systolic and diastolic blood pressure components of the active orthostatic stress test and SSR amplitude as well as a statistically significant prolongation of SSR latency of RA patients when compared to control subjects. Three patients (12%) had clinical symptoms suggestive of AN, which increased to 14 patients (56%) when orthostatic stress tests and the SSR were utilized. There were no statistically significant differences between patients with different disease activity scores with variables grades of RA activity and SSR latency and amplitude. There were no statistically significant differences between patients with different Stanford Health Assessment Questionnaire Disability Index grades of RA functional disability and SSR latency and amplitude.

SUMMARY/CONCLUSION: AN is a common extra-articular manifestation of RA affecting sympathetic and parasympathetic fibers.

A TRANSFORMED POEMS SYNDROME CASE FROM CIDP
Erkan Tokgöz (Elazığ, Turkey), Tayfun Kaşıkçı, Semai Bek, Oğuzhan Öz, İltel Uysal, Zeki Odabaşı (Ankara, Turkey)

INTRODUCTION: The POEMS syndrome was first reported in 1938 and it is characterized by the presence of (P)polyneuropathy, (O) organomegaly, (E) endocrinopathy, (M) M-protein, and (S) skin changes. The characteristic feature of POEMS syndrome is peripheral polyneuropathy that patients are often misdiagnosed as chronic inflammatory demyelinating polyneuropathy (CIDP) or unknown monoclonal gamopati. In this report, we present a patient who were followed for 6 years with a diagnosis of CIDP and developed POEMS syndrome with other systemic findings during the clinical course of her CIDP.

CASE REPORT: 43-year-old female patient had been followed up with CIDP since 2004. She was admitted in 2010 with the presentation of bilateral sudden visual loss, diarrhea, and prolonged menstrual bleeding. She had cachexia, hirsutism, hypermenore, “white nail” sign and distal hypoesthesia in extremities with moderate motor deficits, absent deep tendon reflexes, bilateral papill edema, retinal hemorrhages in and could not count fingers a meter away. Acetazolamide was started and visual acuity improved significantly in following days. Pericardial effusion, ascites, hepatosplenomegaly, myoma uteri and sclerotic bone lesions were detected. Aspiration and biopsy of bone marrow was found to be compatible with plasma cell dyscrasias and chemotherapy was planned by oncology clinic.

SUMMARY: Patient should have the following: the peripheral neuropathy; osteosclerotic myeloma, a clonal plasma cell dyscrasia and at least one sclerotic bone lesion or Castleman disease; and at least one of the other features for diagnosis of POEMS syndrome.

POEMS syndrome is a very rarely seen clinical syndrome. Our patient was followed with the diagnosis of CIDP for years. In additon our case transformed to POEMS syndrome with systemic sign and symptoms 6 years after first diagnose. POEMS syndrome was diagnosed with a multidicipliner aproach and due to several signs. Because of all these reasons we found worth to present our case.
CLINICAL CHARACTERISTICS OF PATIENTS WITH AMYOTROPHIC LATERAL SCLEROSIS BASED ON FASCICULATION DETECTION USING MUSCLE ULTRASOUND
Jun Tsugawa, Sho Takeshita, Toshiyasu Ogata, Jiro Fukae, Yoshio Tsuboi (Fukuoka, Japan)

INTRODUCTION: Fasciculations are a characteristic finding among patients with ALS. Muscle ultrasound (US) is a practical and efficient tool to detect such potentials. Generally, fasciculations have been demonstrated in some muscles among patients with ALS during the early stages; however, some patients do not exhibit any detectable fasciculations.

OBJECTIVE: To study the association between the clinical type of ALS and fasciculation grade to examine whether these patients with ALS differ in clinical presentation.

METHODS: The fasciculation grade was measured in 9 muscles using muscle US from 28 ALS patients. The fasciculation scale was categorized into 4 grades based on a previously published report. The patients were divided into 2 groups according to the fasciculation detected: grades 2-3 in 2 or more muscles (fasciculation ≥2 group) or fewer than 2 muscles (fasciculation <2 group). The differences between the 2 groups were then compared.

RESULTS: Disease duration was significantly shorter in the fasciculation ≥2 group than the fasciculation <2 group (8.0 versus 42.0 months, p<0.001). The fasciculation ≥2 group was significantly older than the fasciculation <2 group (70.0 versus 61.5 years, p=0.010).

SUMMARY/CONCLUSION: These findings show that it is challenging to detect fasciculations using muscle US for the diagnosis of ALS among young cases who have a prolonged disease duration. Although muscle US appears to be a useful tool in the diagnosis of ALS, the physician should be sensitive to hard-to-detect fasciculations in young patients with ALS who have a slow disease progression or prolonged disease duration.

USEFULNESS OF COMBINED SOMATOSENSORY AND MOTOR EVOKED POTENTIALS AND NEEDLE ELECTROMYOGRAPHY MONITORING IN INTRAMEDULLARY SPINAL CORD TUMOR SURGERIES
Minjung Youn, Seong Rae Jo, Dong Gun Kim, Kyung Seok Park (Seongnam, South Korea)

INTRODUCTION: During surgery for intramedullary spinal cord tumors (IMSCTs), the risk of critical injury to motor and sensory pathways is higher due to direct tract manipulation. Among the intraoperative neuromonitoring (IONM) modalities, needle EMG can provide immediate results without averaging during critical phases of the procedure.

OBJECTIVE: To evaluate whether the combined use of needle EMG with somatosensory evoked potential (SSEP)/motor evoked potential (MEP) monitoring had any advantages over SSEP/MEP monitoring alone.

METHODS: The clinical and neurophysiological data of 25 operations in 21 patients (median age: 41.05 years, age range: 26-83 years; 12 men, 9 women) with IMSCTs were reviewed. SSEPs, MEPs, and continuous free-running needle EMG were monitored during the surgeries.

RESULTS: Combined needle EMG/SSEP/MEP recording was successfully performed in 14/25 operations. Combined SSEP/MEP recording was possible in 11 operations. New postoperative neurological deficits occurred in 15 operations: 5 had combined sensory and motor deficits, 9 had new sensory deficits, and 1 had increased motor weakness. Combined SSEP/MEP monitoring had a sensitivity of 67%, a specificity of 60%, a positive predictive value (PPV) of 67%, and a negative predictive value (NPV) of 40%. Combined needle EMG/SSEP/MEP had a sensitivity of 89% and a PPV of 62%.

SUMMARY/CONCLUSION: Combined needle EMG/SSEP/MEP monitoring provided higher sensitivity than Combined SSEP/MEP monitoring techniques during IMSCT surgery. Combined monitoring with SSEP/MEP and needle EMG can be more helpful for predicting neurological deficit in this surgery.
PROGNOSTIC VALUE OF INTRA- AND EXTRAOPERATIVE LATERAL SPREAD RESPONSES IN MICROVASCULAR DECOMPRESSION SURGERIES OF HEMIFACIAL SPASM
Minjung Youn (Seoul, South Korea), Seong Rae Jo, Dong Gun Kim, Kyung Seok Park (Seongnam, South Korea)

INTRODUCTION: The lateral spread response (LSR) is observed in patients with hemifacial spasm (HFS) by electrically stimulating 1 branch of the facial nerve by needle EMG which activates facial muscles innervated by other branches of the facial nerve.

OBJECTIVE: To evaluate whether extraoperative LSR monitoring has any advantages over intraoperative LSR monitoring.

METHODS: A retrospective review was conducted between January and December 2012 at Seoul National University Bundang Hospital of 25 patients (5 men, 20 women) who underwent continuous intraoperative monitoring during microvascular decompression (MVD). LSR and continuous needle EMG were monitored in the frontalis, orbicularis oculi, and mentalis muscles. Each patient was evaluated before and after surgery, on discharge, and at 3 months after surgery.

RESULTS: On admission and discharge, extraoperative LSR recording was possible in 23 patients. Intraoperative LSRs disappeared during surgery for 17 patients. LSRs were absent for 5 patients. Extraoperative LSRs disappeared after surgery for 10 patients. LSRs were absent for 7 patients. In 6 patients, LSRs were present before and after MVD. For 5 patients, intraoperative LSRs disappeared during surgery but extraoperative LSRs persisted. Statistically, extraoperative disappearance of LSRs was correlated with HFS relief at 4 days after surgery and at 3-month followup (p=0.049 and 0.044).

SUMMARY/CONCLUSION: In this study, extraoperative LSR monitoring was more predictive of surgical outcome compared with intraoperative LSR at 3-month followup. But further research on a larger group for a longer followup period is needed to clarify the prognostic values of intraoperative and extraoperative LSR monitoring.

CONSTITUENT MUSCLE CONTRIBUTION TO THENAR COMPOUND MUSCLE ACTION POTENTIAL
Qing Yue (Bloomsburg, PA), Tyson Hale, Aaron Knecht (Danville, PA), Jessica DeLaurentis, Anna Morusiewicz, Kristiana Barbarevech (Bloomsburg, PA)

INTRODUCTION: Within the thenar eminence, the median nerve innervates 3 muscles: abductor pollicis brevis (APB), flexor pollicis brevis (FPB), and opponens pollicis (OP). In theory, these muscles should all contribute to the thenar compound muscle action potential (CMAP).

OBJECTIVE: To evaluate subcomponents of the thenar CMAP from the APB, FPB, and OP.

METHODS: Surface and monopolar EMG (moEMG) needle recordings were obtained from normal human subjects at or near the 3 recording sites: proximal (position 1), middle (position 2), and distal (position 3) aspects of the thenar eminence during activation of median nerve or muscle fibers.

RESULTS: Compared to a “peak–trough” waveform pattern recorded from positions 1 and 2 by surface electrodes, position 3 yielded a “trough–peak” CMAP pattern. The moEMG needle recordings in the vicinity of position 3 revealed propagating sources that primarily derived from FPB based on the recordings of muscle fiber action potentials. Recordings in the vicinity of position 1 by moEMG needle detected a possible OP potential, which did not appear to have a major influence on the APB-derived thenar CMAP peak.

SUMMARY/CONCLUSION: Specific thenar CMAP components of APB, FPB, and possibly OP origins were identified. The moEMG needle study suggested that the thenar CMAP peak of the surface recording was mainly determined by the FPB at position 3 and by the APB at position 1.
**COMPUND MUSCLE ACTION POTENTIAL AMPLITUDE CAN BE ALTERED AT THRESHOLD BY MINOR CHANGE OF THENAR SHAPE**
Qing Yue, Jessica DeLaurentis, Kristiana Barbarevech, Anna Morusiewicz (Bloomsburg, PA)

INTRODUCTION: It had been documented that the configuration of the compound muscle action potential (CMAP) has been influenced by a number of variables including thumb positioning, suggesting that the profile/shape of the muscle may influence the quadrupoles being registered as it travels from motor point to tendon. However, thumb movement may decouple the surface recording electrode and the underneath muscle group from their original match, which, by itself, leads to a recording difference.

OBJECTIVE: To study whether thenar shape change without muscle/electrode decoupling alters thenar CMAP configuration.

METHODS: Surface CMAP recordings were obtained from 13 normal subjects at 3 recording sites: proximal (position 1), middle (position 2) and distal (position 3) aspects of the thenar eminence. A reference electrode was placed on the hypothenar eminence. Thenar shape change was achieved by applying pressure at the first metacarpophalangeal joint and tendon of the abductor pollicis longus at the wrist by the examiner. Care was taken not to stretch thenar muscles.

RESULTS: The majority (11/13, 85%) of the subjects showed drastic decreased CMAP amplitudes after the induced thenar shape change when the median nerve was stimulated at a level slightly above threshold. Similar changes were noted in all 3 recording sites. At the level of supramaximal stimulus, however, no such a change was observed.

SUMMARY/CONCLUSION: When a fraction of muscle fibers are being activated, their net summinating quadrupoles can be altered by changing the spatial configuration of these fibers.

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**VALIDATION OF THE E-NORMS METHOD TO DERIVE REFERENCE VALUES OF THE FLEXOR CARPI RADIALIS H-REFLEX LATENCY**
Christina Zaccarini (Syracuse, NY), Chao Zheng (Shanghai, China), Joe Jabre (Los Angeles, CA), Jianyuan Jiang (Shanghai, China), Robert Weber (Syracuse, NY), Yu Zhu (Syracuse, NY)

INTRODUCTION: Extrapolated norms or e-norms (Jabre and colleagues 2015) is a method used to derive reference values from laboratory populations when such data is unavailable or difficult to acquire. The method ranks a variable's data points in ascending order, calculates their first-order difference, and plots it against rank, resulting in an inverted S-curve. The plateau component of this curve is used to derive a variable's descriptive statistics.

OBJECTIVE: To validate the E-norms method by applying it to a new measure, the flexor carpi radialis H reflex latency (FCR-HL) derived from recordings in patients with known C6 and C7 radiculopathies.

METHODS: The authors’ previously published FCR-HL data from these patients was sent in raw format to an independent researcher blinded to the results. The data was analyzed using E-norms to derive the mean FCR-HL recorded from the patients’ affected and contralateral, unaffected sides. E-norms reference values were then compared to the published values in the literature.

RESULTS: E-norms derived FCR-HL means of 16.22±1.03 ms (unaffected sides), 17.20±0.28 ms (C6 radiculopathy), and 18.36±0.97 ms (C7 radiculopathy) compared favorably to published means of 16.68±1.7 ms (unaffected sides) (p=0.063), 16.91±1.8 ms (C6 radiculopathy) (p=0.50), and 18.40±1.8 ms (C7 radiculopathy) (p=0.93). E-norms derived FCR-HL was prolonged in C7 compared to C6 radiculopathies (p<0.001) and unaffected sides (p<0.001), consistent with previously published results.

SUMMARY/CONCLUSION: FCR-HL reference values obtained using the E-norms method compared well to previously published values in the literature. This validates the E-norms method and supports the previous finding of prolonged FCR-HL as a specific indicator of C7 radiculopathy.

Christina Zaccarini, MD, Resident and Fellow Member Award Recipient
INTRODUCTION: Diabetic peripheral neuropathy (DPN) affects 50% of patients with diabetes mellitus (DM). DPN has different forms; the most common would be the symmetrical, distal onset, slow progressing sensory polyneuropathy. Another less common form of DPN, known as diabetic proximal radiculoplexus neuropathy or diabetic amyotrophy, is found in approximately 1% of DM patients in usually middle-to-old age groups. The most studied type is characterized by acute-to-subacute onset of asymmetric pain in the proximal lower extremity that progresses to weakness and atrophy. However, the involvement of the upper extremity is rarely recognized.

CASE REPORT: A 59-year-old female, recently diagnosed to have non–insulin-dependent DM, presented with progressive weakness of bilateral shoulder muscles, associated with pain on movement of shoulders, with atrophy of the muscles of the hands, as well as the bilateral supraspinatus muscles, and sensory deficits on radial, median, and ulnar nerve distributions of both upper extremities. Needle EMG/NCSs showed bilateral C6, C7, C8, and T1 radiculopathy, incomplete with more chronic than acute denervation changes, demyelination changes, affecting sensory more than motor roots to its peripheral nerves.

SUMMARY/CONCLUSION: The less common variants of DPNs are asymmetric proximal, which include lumbosacral, thoracic, and cervical radiculoplexus neuropathy, which could be present even in newly diagnosed cases. When presented with DM patients with pain, weakness, and atrophy of a unilateral upper extremity, these should be considered in the diagnoses. Proper diagnostic workup of this condition could prevent unnecessary treatment when it is mistaken as another disease entity.

Jhoana Marie Zambrano, MD, IFCN North American Chapter Fellowship Award Recipient

INTRODUCTION: Ultrasound-guided needle EMG is becoming a popular option to help localize nerves and muscle in patients with altered anatomy.

OBJECTIVE: To describe a case of traumatic left upper limb polyneuropathy with pre- and postoperative ultrasound-guided EDX studies.

CASE REPORT: A 37-year-old patient presented with traumatic upper limb polyneuropathy after a fall. Initial EDX studies revealed severe left median neuropathy distal to innervation of the flexor digitorum superficialis, proximal to innervation of the flexor pollicis longus; severe left ulnar neuropathy distal to innervation of the flexor digitorum profundus, proximal to innervation of the abductor digiti minimi; and severe left radial neuropathy distal to innervation of the extensor carpi radialis and proximal to innervation of the extensor carpi radialis longus/brevis. He underwent several operations. Repeat EDX studies with ultrasound guidance 4 years later revealed persistent left median neuropathy distal to the flexor carpi radialis and proximal to the abductor pollicis brevis; improved left ulnar neuropathy distal to innervation of the flexor carpi ulnaris, proximal to innervation of the first dorsal interosseous; and persistent sensory radial neuropathy. Ultrasound guidance ensured proper EMG needle localization and revealed distorted anatomy/bony landmarks and evidence of atrophy in affected muscles. Patient will undergo tendon transfers: pronator trees to extensor carpi radialis brevis, flexor carpi radialis to extensor digitorum communis, and palmaris longus to extensor pollicis longus.

SUMMARY/CONCLUSION: Final conclusions will be determined after tendon transfers and ultrasound-guided needle EMG. Postoperative sonographic images and EDX findings will be compared to preoperative studies. Affect of altered anatomy, strength impairments, and positional changes will be assessed. Technical aspects of performing ultrasound-guided needle EMG will be assessed as will risks/benefits of the technique.
EFFICACY OF MULTILEVEL BOTULINUM TOXIN A TREATMENT OF HEMIPLEGIC AND DIPLEGIC SPASTIC CEREBRAL PALSY: A CLINICAL AND NEUROPHYSIOLOGICAL STUDY
Nayera Saber, Dalia El Mikawy (Cairo, Egypt)

INTRODUCTION: Spasticity in cerebral palsy (CP) places obstacles in the path to achieving rehabilitation goals.

OBJECTIVE: To detect favorable outcomes of multilevel botulinum toxin type A (BTX-A) injection in spastic CP children at a single session from clinical and electrophysiological aspects and to localize dependable factors for those outcomes.

METHODS: Twenty-two spastic CP patients (12 male, 10 female) with lower limb spasticity were enrolled. All patients were assessed by the modified Ashworth scale (MAS), timed 10 meter walk (TMW), pain scale, and knee and ankle range of motion (ROM). Also assessed were F waves, H reflexes with F/M and H/M amplitudes, and gastrocnemius (Gas) surface EMG (SEMG) for interference pattern (IP) recording. Single multilevel BTX-A injection of Gas, soleus (Sol), hamstring (Ham), and tibialis posterior muscles was performed in a single session. Reassessment was at 1 and 6 months.

RESULTS: Fourteen patients were diplegic and 8 were hemiplegic. There was significant increase in knee and ankle ROM (p<0.01) at 1 and 6 months post injection. The MAS, TMW, pain scale, and F/M and H/M amplitude were significantly reduced with significant improvement of IP 1 month after injection which persisted until the end of the study. A significant negative correlation existed between the MAS of Gas, Sol and Ham at 6 months and baseline ROM of the ankle and knee (p<0.05). ROM of the knee and amplitude of H/M and F/M were dependable factors for improved TMW after 6 months (TMW2) as a secondary outcome at the study end point.

SUMMARY/CONCLUSION: Multilevel BTX-A injection reduces spasticity and improves the short- and longterm outcomes.

BRACHIAL PLEXOPATHY AND HORNER'S SYNDROME AS A RARE COMPLICATION OF KELOID FORMATION
Kateryna Kurako, Ramon Lugo (Weston, FL)

INTRODUCTION: Keloids are an excessive tissue response to injury caused by fibroblast proliferation and collagen overproduction. While they can sometimes cause direct functional impairment, they are not known to proliferate deep enough to cause functional nerve compression.

CASE REPORT: A 58-year-old woman with a past medical history of breast cancer with a left modified radical mastectomy and left supraclavicular lymph node biopsy consulted for a left upper extremity weakness and numbness and a left “lazy eye.” The patient also complained of a large mass compressing her left neck and supraclavicular area which she stated began after her lymph node biopsy 1 year prior and continued to grow in size. Physical examination demonstrated mitotic pupils and ptosis on the left, along with shoulder shrug weakness on the left. MRI of the brachial plexus showed a left supraclavicular mass which infiltrated the scalene muscles medially and involved the brachial plexus more in the upper trunks of C5 and C6. Alongside this, there was also asymmetric edema of the right shoulder muscles which reflected denervation edema.

RESULTS: This case demonstrates the potential neurological impairments that large keloids can cause if not immediately managed appropriately.

SUMMARY/CONCLUSION: It is important to take into consideration the location, size, and growth rate of keloids to guide appropriate treatment.
NOVEL PMP22 MUTATION CAUSING DEJERINE–SOTTAS DISEASE, CLINICALLY INDISTINGUISHABLE FROM CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY
Katherine Bonsell, Maureen Kelly Galindo, Monica Chacon, Dinesh Talwar, Katalin Scherer (Tucson, AZ)

INTRODUCTION: Health insurance companies routinely deny genetic testing, despite it often being more cost effective than the wrong diagnosis.

OBJECTIVE: To demonstrate how delay in diagnosis and wasteful healthcare utilization occurs when insurance companies deny genetic testing.

CASE REPORT: An 8-year-old boy was repeatedly evaluated for neuropathy. First year development was normal. He pulled-to-stand at 1, then walked on his knees until 2.5. At age 3, he was described as clumsy, unable to run or jump, with diffuse distal greater than proximal weakness, hypotonia, unobtainable deep tendon reflexes (DTRs). He had normal creatine phosphokinase and elevated cerebrospinal fluid (CSF) (62 mg/dl). Needle EMG showed absent sensory nerve action potentials and severely slow, low amplitude motor responses with conduction blocks. MRI demonstrated thickening and diffuse enhancement of lumbar nerve roots. Genetic testing was denied, thus monthly IVIg was given for 2 years. PMP22 duplication/deletion testing approved at age 7 was normal. Clinical examination at age 8 showed profound distal greater than proximal weakness, inability to jump/run, unobtainable DTRs, and absent vibration sense in the feet. CSF again showed elevated protein (76 mg/dl). Motor nerve velocities were severely slowed with conduction blocks and temporal dispersion. Needle EMG demonstrated active reinnervation. Next generation sequencing revealed a novel heterozygous PMP22 mutation at codon 94 resulting in a frameshift and truncated protein. Unaffected parents tested negative.

SUMMARY/CONCLUSION: Dejerine–Sottas disease is clinically indistinguishable from chronic inflammatory demyelinating polyneuropathy, when the onset of symptoms is not preceded by normal walking due to developmental age. Children presenting with a demyelinating neuropathy should first undergo genetic testing to avoid unnecessary and costly diagnostic testing and treatment.

USE OF A CHECKLIST TO IMPROVE GUIDELINE COMPLIANCE AND QUALITY OF CARE IN AMYOTROPHIC LATERAL SCLEROSIS PATIENTS
Jason Fleming, Debra O’Reilly, Vera Fridman, Steven Ringel, Stacy Dixon, Dianna Quan (Aurora, CO)

INTRODUCTION: When patients are diagnosed with ALS, the neuromuscular physician’s role shifts to symptom management, optimization of function, moral and educational support, and anticipation and treatment of related comorbidities. Specialty society or academy guidelines help physicians decide upon needed interventions, but guidelines are lengthy and sometimes difficult to implement in a clinic visit. A checklist based on the American Diabetes Association disease management guidelines was shown to improve specific diabetes monitoring. Checklists in surgical and ICU settings have resulted in decreased morbidity and mortality. A checklist based upon guidelines for care of ALS patients could be incorporated into the electronic medical record and used at followup visits to ensure comprehensive, evidence-based care to improve quality of life and morbidity.

OBJECTIVE: To develop a guideline-based, user friendly checklist to deliver optimal care for patients with ALS.

METHODS: The most recent American Academy of Neurology guidelines for care of patients with ALS were used to create a comprehensive list of symptoms and laboratory data to review at each followup clinic visit. The list was refined through review with all neuromuscular providers at the authors’ center and made available as a “dot phrase” in their institution’s electronic medical records system.

RESULTS: Data on compliance with guideline recommendations are being collected from before and after implementation of this checklist.

SUMMARY/CONCLUSION: Checklists have reduced morbidity and mortality in numerous care settings. Their power can be harnessed in neuromuscular clinics to enhance compliance with guidelines and improve quality of care.
TWO TELEHEALTH MODELS FOR VIRTUAL INTERDISCIPLINARY AMYOTROPHIC LATERAL SCLEROSIS CARE: PARALLEL VERSUS TANDEM APPROACH
Richard Strozewski (Cleveland, OH), Ileana Howard (Seattle, WA)

INTRODUCTION: Multidisciplinary care delivered by specialized ALS clinics is associated with better patient outcomes and is the standard of care defined by clinical practice guidelines published by the American Academy of Neurology (Miller and colleagues 2009) and the Veterans Health Administration (VHA Handbook 1101.07). However, access to an experienced care team can be burdensome or impossible for patients who are affected by the disease, particularly for those living in rural areas. Telehealth technologies such as Clinical Video Telehealth (CVT) provide an opportunity for specialized teams to provide services to individuals who are unable to travel or access care at the tertiary medical centers. Previous studies suggest high patient and caregiver satisfaction with Tele-ALS care (Vitacca 2010). Despite these promising findings, no consensus exists on the optimal strategy for the incorporation of Telehealth into specialized ALS care.

OBJECTIVE: To describe 2 current models of Tele-ALS interdisciplinary care currently implemented in the Veterans Health Administration: late-stage implementation (tandem approach) and early-stage implementation (parallel approach). For each model, a pathway for implementation and how quality outcomes are monitored will be presented.

RESULTS: The authors will present two practical models for incorporation of Telehealth into specialized ALS care which provide models for complete, virtual, ALS interdisciplinary care.

SUMMARY/CONCLUSION: Virtual ALS interdisciplinary care is feasible and provides efficient, cost-effective, and patient-centered care. A multicenter, outcome data collection cohort study is in progress.

COMPLICATION RISK OF ELECTRODIAGNOSTIC STUDIES IN AN ADULT AMBULATORY CARE SETTING
Lisa Williams (Santa Cruz, CA), Min Kim (Redwood City, CA), Ninad Karandikar (Cupertino, CA)

INTRODUCTION: The complication rate of EDX studies is thought to be rare. There have been several isolated reports of infections; however, no large scale studies to date have looked at complication rates of needle EMG and NCSs.

OBJECTIVE: To estimate the risk of complications from needle EMG.

METHODS: After Institutional Review Board approval, all patients who had a needle EMG/NCS from December 2005 to December 2014 were identified retrospectively using Current Procedural Terminology (CPT) codes. The Veterans Affairs hospital was utilized given the ubiquitous and long standing use of an Electronic Medical Record for data query as well as the contained nature of the patient population. Complications were identified by using International Classification of Diseases (ICD)-9 codes within 90 days of the needle EMG. A total of 202 individual ICD-9 codes were searched, including infection, hematoma, and pacemaker complications. When complications were found medical records were reviewed by an independent physician to determine if the complication was causally related to the needle EMG.

RESULTS: Infection complications were found in 68/3993 patients (1.7% [95% CI 1.3-2.2%]) in the study. Independent physician chart review revealed these infections were not associated with needle EMG. Pacemaker complications were found in 0 patients (0% [95% CI 0-0.1%]), compartment syndrome was found in 0 patients (0% [95% CI 0-0.1%]), and hemorrhage was found in 0 patients (0% [95% CI 0-0.1%]).

SUMMARY/CONCLUSION: This is the first large scale retrospective chart review study looking at complications of needle EMG/NCSs in a contained population. There is a low risk of complications from needle EMG procedures, including infection, hemorrhage, compartment syndrome, and pacemaker malfunction.

Lisa Williams, MD, Resident and Fellow Member Award Recipient
INTRODUCTION: A high rate of needle EMG referrals to the authors’ center from primary care providers (PCPs) for suspected CTS did not result in an invasive treatment or diagnosis of median neuropathy.

OBJECTIVE: To (1) increase the rate of bracing prior to needle EMG, (2) increase the rate of injection or surgical treatment after needle EMG, and (3) reduce the number of referrals for isolated wrist pain without median neuropathy.

METHODS: Baseline data prior to intervention were ascertained retrospectively. An educational memo to PCPs was communicated via recurrent email and discussed in care team huddles. Patients were prospectively ascertained and scheduled (n=26) by a single nurse providing education on CTS treatment options. Treatment outcomes were reassessed via chart review at 6 months.

RESULTS: Referrals for isolated wrist pain declined from 15% to 4% (p=0.22). Significantly more patients were treated with bracing prior to needle EMG (from 68% to 92%; p<0.02). Nurse-to-patient education at the time of scheduling did not lead to cancelation of needle EMG or trial of bracing before needle EMG. The invasive treatment rate did not significantly increase (from 65% to 73%; p=0.40).

SUMMARY/CONCLUSION: Primary care team education may improve the value and utilization of needle EMG for patients with suspected CTS. Patients who elect conservative treatment for CTS likely still value needle EMG findings. Nurse-to-patient education at the time of scheduling needle EMG appears to have little impact on utilization and outcomes.

INTRODUCTION: Post-stroke, spastic elbow flexor muscles must be treated to improve function.

OBJECTIVE: To assess changes in elbow flexor spasticity after phenol injections to the musculocutaneous nerve (MCN), and to identify postinjection complications.

METHODS: For patients who have elbow flexor spasticity due to CNS disorders, changes in spasticity with phenol injections to the MCN were retrospectively assessed. For a subgroup of post-stroke patients, elbow flexor spasticity was assessed with a Modified Ashworth Scale (MAS) from 0 (no spasticity) to 4 (severe spasticity) pre- and post-injections, and analyzed with a t-test.

RESULTS: For 196 MCN injections with phenol 5% in sterile water, 106 injections were for stroke patients (48 women, age range: 68.0±13.7 years; 58 men, age range: 63.6±12.9 years). The data (mean±SD) showed: 0.54±0.16 mA to stimulate the MCN when phenol was injected; 1.3±0.56 sites injected; 0.67±0.53 ml phenol injected; and MAS pre-injection was 3.2±0.3 and post-injection was 0.9±0.7. The MAS post-injection was significantly less (p<0.05) than pre-injection with a t-statistic of 28.1. Of 196 MCNs injected with phenol, 1 patient had transient numbness in the distribution of the lateral antebrachial cutaneous nerve.

SUMMARY/CONCLUSION: Phenol injections to the MCN are an effective and safe approach to decrease elbow flexor spasticity for post-stroke patients.

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A PILOT TRIAL TO ASSESS THE FEASIBILITY AND EFFICACY OF SUBCUTANEOUS IMMUNOGLOBULIN IN PATIENTS WITH MYASTHENIA GRAVIS EXACERBATION: AN UPDATE
Grayson Beecher, Dustin Anderson, Ashley Mallon, Derrick Blackmore, Zaeeem Siddiqi (Edmonton, Canada)

INTRODUCTION: Subcutaneous immunoglobulin (SCIg) is an efficacious and safe alternative to IVIg in the treatment of immune-mediated neuropathies; however, its use in myasthenia gravis (MG) has yet to be formally studied. Presented here are the updated results of a phase III clinical trial assessing the use of Hizentra® (20% SCIg) in MG exacerbation.

OBJECTIVE: To assess the safety, feasibility, and efficacy of Hizentra® in patients with MG exacerbation.

METHODS: Thirty patients with MG (Myasthenia Gravis Foundation of America Class II and III) will be enrolled in a prospective, open-label, single blind study to receive SCIg at 2g/kg in a flexible dosing regimen over 4 weeks. The primary outcome measure is the change in quantitative MG (QMG) score from baseline to end of study (6 weeks). Standardized assessment tools are used weekly to evaluate secondary efficacy outcomes, incidence of side effects, serum IgG levels, and patient compliance.

RESULTS: Seventeen patients have been enrolled; thirteen have completed the study. All patients required 1 training session for self-infusion. There were no major adverse reactions to SCIg, including hemolysis. Mild infusion site reactions, flu-like symptoms, and headache have been reported, none causing discontinuation of SCIg. QMG scores showed clinically significant mean reduction of 5.08 points at end of study. Additional efficacy assessment tools, including manual muscle testing and MG composite, yielded similar results. Serum IgG levels gradually increased by a mean of 97%. Standardized patient surveys indicate reasonable satisfaction with SCIg use.

SUMMARY/CONCLUSION: Ongoing analysis suggests that SCIg may be safe, effective, and well tolerated in patients with MG exacerbation.

Grayson Beecher, MD, Resident and Fellow Member Award Recipient

ONABOTULINUM TOXIN IN THE TREATMENT OF CARPAL TUNNEL SYNDROME
Jonathan Benfield (San Antonio, TX), Benjamin Sucher, Kinal Bhatt, Ralph Bennett (Phoenix, AZ), Anthony Lee (Scottsdale, Arizona)

INTRODUCTION: CTS is caused by compression of the median nerve within the carpal tunnel. Neuromuscular ultrasound (NMUS) can identify thenar muscle contraction compressing the median nerve in CTS patients. There is no available treatment to decrease thenar muscle activity as a mechanism to relieve median nerve compression.

OBJECTIVE: To evaluate whether CTS patients with NMUS-confirmed nerve compression by thenar muscles could benefit from onabotulinumtoxinA (onabot) injection to reduce muscle size and activity.

METHODS: A prospective, randomized, double-blind, placebo-controlled study of 10 patients with mild-to-moderate bilateral CTS, confirmed by NCS and NMUS, was conducted. Patients were divided into control and treatment groups of 5 subjects each. Non-dominant hands were injected under US guidance with 40 units of onabot or 0.4 cc normal saline (NS), with 2 equally-divided doses (20 units onabot or NS each) into the abductor pollicis brevis and opponens pollicis muscles. Participants were evaluated with NMUS, NCSs, the Levine grading scale, and Jamar® (Lafayette Instrument®, Lafayette, IN) dynamometer at baseline and at 6, 12, and 18 weeks.

RESULTS: Three subjects injected with onabot demonstrated decreases in median distal motor latencies, 2 had decreases in median distal sensory latencies, and some had decreases in cross-sectional area on NMUS. One onabot subject did show improvement or progression in median distal latencies while the non-injected hand had increasing median distal latencies.

SUMMARY/CONCLUSION: Onabot injections for CTS may be a viable nonsurgical treatment alternative in some patients. Further studies with a greater number of subjects are required to confirm efficacy and determine the optimum dose of onabot.
FACTORS LINKED TO MORTALITY IN AMYOTROPHIC LATERAL SCLEROSIS PATIENTS DURING 1998-2014: A NEW YORK STATEWIDE PLANNING AND RESEARCH COOPERATION SYSTEM DATABASE REPORT
Mohammad Chaudhary, Abu Nasar (Newark, NJ), Francisco Gomez (West Orange, NJ), Nizar Souayah (Newark, NJ)

INTRODUCTION: Some reports suggest diabetes and prostate cancer may slow the progression of ALS.

OBJECTIVES: To analyze the relationship between various demographic and comorbid conditions and mortality in ALS patients.

METHODS: The authors compared factors affecting mortality in patients diagnosed with ALS during 1998-2014 utilizing the New York Statewide Planning and Research Cooperation System (SPARCS) database. Logistic regression analysis was performed to identify independent risk factors for mortality.

RESULTS: A total of 2841 patients were analyzed (median age: 65±13.33 years, 60% male). Overall mortality rate for this period was 20%. ALS patients with hypertension demonstrated an overall reduced mortality rate (OR 0.70, CI 95% 0.57-0.86, p=0.001). Advanced age was associated with a higher mortality rate: 61-80 years (OR 1.72, CI 95% 1.39-2.14, p<0.001) and above 80 (OR 3.17, CI 95% 2.28-4.40, p<0.001). Other factors associated with higher mortality rates were respiratory complications including mechanical ventilation (OR 2.08, CI 95% 1.60-2.70, p<0.001) as well as the use of noninvasive ventilation (OR 2.77, CI 95% 2.15-3.56, p<0.001). Diabetes and cancer did not affect mortality in this study (OR 1.03, CI 95% 0.78-1.36, p=0.831; OR 1.03, CI 95% 0.39-2.75, p=0.947, respectively).

CONCLUSION: Advanced age and respiratory complications were associated with higher mortality, whereas hypertension was associated with lower mortality. Diabetes and cancer had no statistically significant effect on mortality. Work is in progress to determine if any comorbid condition can delay disease onset and slow progression of ALS.

COMPARING OUTCOMES AND CHARGES OF MYASTHENIA GRAVIS PATIENTS TREATED WITH INTRAVENOUS IMMUNOGLOBULIN VERSUS PLASMA EXCHANGE: A 1998-2014 NEW YORK STATEWIDE PLANNING AND RESEARCH COOPERATION SYSTEM DATABASE REPORT
Mohammad Chaudhary, Abu Nasar (Newark, NJ), Francisco Gomez (West Orange, NJ), Nizar Souayah (Newark, NJ)

INTRODUCTION: IVIg and plasma exchange (PE) are accepted treatments in the management of myasthenia gravis (MG).

OBJECTIVE: To compare outcomes and charges for MG inpatients managed with IVIg versus PE.

METHODS: Using the New York Statewide Planning and Research Cooperation System (SPARCS) inpatient database, MG patients above age 18 were determined per International Classification of Diseases (ICD) codes (358.0, 358.00, and 358.01). Outcomes and mean hospital charges for MG patients from 1998-2014 were determined utilizing χ² and the Mann-Whitney U test.

RESULTS: Of the 3168 patients (median age: 58 years, 36% male) analyzed, 1506 received IVIg only and 1662 received PE only. Patients in the IVIg group were more likely to be discharged to home versus transferred to care facilities (75% versus 64% and 23% versus 33%, respectively, p<0.001). The IVIg arm showed shorter lengths of stay (median: 5 (4-7) versus 10 (6-15) days) and lower mortality (2% versus 3%) (p<0.001). Incidence of respiratory failure, mechanical ventilation, tracheostomy, and pneumonia was significantly lower in the IVIg group (10% versus 20%, p<0.001; 7% versus 18%, p<0.001; 1% versus 4%, p<0.001; respectively) as well as readmissions (58% versus 80%, p<0.001). Average hospitalization charges were also significantly lower in the IVIg group ($67,400 versus $77,900, p<0.001).

CONCLUSION: Hospitalization charges, length of hospitalization, and incidence of respiratory complications were significantly lower in patients treated with IVIg compared to PE. Work is in progress to determine the outcome of mechanically ventilated patients and whether MG patients with respiratory failure are preferentially treated with PE.
INTRODUCTION: Guillain–Barré syndrome (GBS) is usually treated with IVIg and plasma exchange (PE).

OBJECTIVE: To compare outcomes and costs between PE and IVIg in GBS.

METHODS: Adults admitted for GBS during 2009-2013 were analyzed utilizing the New York Statewide Planning and Research Cooperation System (SPARCS) database. Patients were divided into PE and IVIg groups and outcomes and costs analyzed with the Chi-square and Wilcoxon rank-sum tests.

RESULTS: Of 1027 patients (median age: 56 years, 55% male) analyzed, 822 received IVIg and 205 PE. The IVIg group fared better overall versus the PE group, with more home discharges (35% versus 18%) and decreased mortality (1% versus 2%) (improved discharge status, p=0.001). The IVIg group also had a decreased length of stay (median of 7 days versus 14, p=0.017) and fewer reported nosocomial infections (19% versus 26%, p=0.027). The IVIg group presented fewer instances of endotracheal intubation and spent less time on mechanical ventilation with 2% requiring <96 hours of mechanical ventilation versus 5% for the PE arm (p=0.034) as well as 8% versus 18% requiring >96 hours of mechanical ventilation, respectively (p=0.001). There was no significant difference in readmissions or multiple readmissions between both groups (p=0.838). Furthermore, there was a significant decrease in average hospitalization costs and charges over 5 years with an average of $115,950 for IVIg patients versus $158,000 for those receiving PE (p=0.001).

SUMMARY/CONCLUSION: IVIg was superior to PE in all tested metrics outcomes and costs, except readmissions which showed no statistical difference.

INTRODUCTION: IVIg is increasingly used as an alternative treatment to steroids for management of chronic inflammatory demyelinating polyneuropathy (CIDP).

OBJECTIVE: To compare outcomes and costs between CIDP treatments: steroids or other treatments versus IVIg.

METHODS: The New York Statewide Planning and Research Cooperation System (SPARCS) inpatient database was used to analyze CIDP patients between 2009-2013 for outcomes with treatment by IVIg, steroids, or other treatments. The categorical Chi-square test and two-sample t-test or Wilcoxon rank-sum tests were used.

RESULTS: Of the 1070 patients analyzed, those who received IVIg were significantly older than those who received steroids (median age: 66 versus 52 years, p<0.001). No significant difference in length of stay between IVIg and steroid groups (p=0.261), nor in average charges $74,600–62,800 (p=0.702), was observed between the 3 groups. A significantly higher association for the steroids group with diabetes was observed versus IVIg or other modalities (47% versus 25% and 28%, p=0.011). Home discharge rates were significantly higher for IVIg patients (52%) compared to patients receiving steroids (p<0.002). Multiple readmissions were significantly higher in the steroid group compared to the IVIg group (p<0.001).

SUMMARY/CONCLUSION: No significant difference was found in charges or hospitalization length between CIDP patients treated with IVIg versus steroids. There was found a significantly higher discharge to hospice and skilled nursing facility/short-term facility as well as more prevalence of diabetes in CIDP patients treated with steroids compared to those treated with IVIg. This may impact the cost of CIDP patients treated with steroids when the outpatient cost is taken into consideration.

Jesyre Veitia, Francisco Gomez (West Orange, NJ), Nizar Souayah (Newark, NJ)

INTRODUCTION: There are reports of Guillain–Barré syndrome (GBS) after influenza vaccination.

OBJECTIVE: To investigate the correlation between GBS and influenza vaccination in adults.

METHODS: Data from the Vaccine Adverse Event Reporting System (VAERS) database for 1991-2015 was utilized. The initial 6 weeks post-vaccination was defined as the risk period for possible cause and effect between vaccination and GBS; the subsequent 6 weeks were defined as the control period. Case-centered and self-controlled case analyses were utilized.

RESULTS: A total of 1174 GBS cases were reported after influenza vaccination (mean age: 56±16.21 years, 54.94% male). The reporting rate of post-influenza vaccination GBS was 0.55 per million, similar to that in the general population. Of these cases, 83.98% were reported within the first 6 weeks after vaccination, with 54.5% of these occurring within the first 2 weeks. Eighteen GBS cases fulfilled Brighton criteria levels 1, 2, or 3, with 17 (90%) of these cases reported within the risk period and 61.11% occurring within the first 2 weeks. Self-controlled case analysis demonstrated that most (82.58%) GBS cases were reported within the risk period regardless of Brighton criteria. Similar results were obtained with case-centered analysis.

SUMMARY/CONCLUSION: No significant increase was observed in the reporting rate for post-vaccination GBS. However, there was an unbalanced distribution of the GBS reporting rate, skewed towards the first 6 weeks post-vaccination, suggesting some cases could be associated with vaccination. These results warrant the use of active surveillance as well as continuous and careful analysis of post-vaccination GBS cases.


Francisco Gomez (West Orange, NJ), Mohammad El-Ghanem, Abu Nasar (Newark, NJ), Nizar Souayah (Westfield, NJ)

INTRODUCTION: Guillain–Barré syndrome (GBS) after influenza vaccination has been reported and the authors have attempted to find an association.

OBJECTIVE: To investigate the correlation between GBS and influenza vaccination in adults, analyzing both the Vaccine Adverse Event Reporting System (VAERS) and Vaccine Safety Datalink (VSD) databases.

METHODS: The VAERS and VSD 1991-2000 data was analyzed. Self-controlled case series and case-centered study designs were used. The initial 6 weeks post-vaccination was defined as the risk period of possible cause and effect between vaccination and GBS.

RESULTS: A total of 69 VSD and 62 VAERS GBS cases after vaccination were analyzed. The incidence of GBS after vaccination in the VSD population was 2/100,000, similar to that in the general population. There was no significant difference between the risk and control periods in the VSD database (10% versus 8.5%, p<0.771). However, in VAERS most cases were reported within the risk period. (96.5% versus 3.5%, p<0.001).

SUMMARY/CONCLUSION: There was a significant difference in GBS incidence distribution between VSD and VAERS. No significant increase was found in the incidence of GBS after influenza vaccination in the risk period compared to the control period or the general population in the VSD database; however, most VAERS cases of GBS were reported within the risk period. Case ascertainment differed between databases, no case in VAERS fulfilled the Brighton level 1 criteria. This discrepancy may be explained by several factors: VSD constitutes active surveillance whereas VAERS is passive surveillance; all VSD, but not VAERS, patients belong to 1 healthcare system; and acute events closer to vaccination are reported more than events occurring late after vaccination.

Francisco Gomez, MD, Resident and Fellow Member Award Recipient
GUILLAIN–BARRÉ SYNDROME AFTER INFLUENZA VACCINATION 1999–2015: ANALYZING THE VACCINE ADVERSE EVENT REPORTING SYSTEM

Jesyree Veitia (West Orange, NJ), Francisco Gomez (West Orange, NJ), Nizar Souayah (Westfield, NJ)

OBJECTIVE: To investigate the correlation between Guillain–Barré syndrome (GBS) and influenza vaccination in adults.

METHODS: Data from the Vaccine Adverse Event Reporting System (VAERS) database for 1999-2015 was utilized. The initial 6 weeks post-vaccination was defined as the risk period for possible cause and effect between vaccination and GBS; the subsequent 6 weeks were defined as the control period. Case-centered and self-controlled case analyses were utilized.

RESULTS: A total of 841 GBS cases were reported after influenza vaccination (mean age: 55.73±16.56 years, 55.18% male) for an incidence rate of 0.765 per million, similar to that in general population. Of these cases, 82% were reported within the first 6 weeks after vaccination, with 64.4% of these occurring within the first 2 weeks. Eighteen GBS cases fulfilled Brighton criteria levels 1, 2, or 3; 16 (90%) of these were reported in the risk period, with 11 (61)% of these cases occurring within the first 2 weeks. Self-controlled case analysis demonstrated that more than 85% of GBS cases were reported within the risk period whether or not the case fulfills Brighton criteria. Similar results were obtained with case-centered analysis.

SUMMARY/CONCLUSION: An increase in incidence of GBS post-influenza vaccination was not observed. There is an unbalanced distribution of incidence within the first 6 weeks, suggesting some cases could be associated with vaccination. Most GBS cases were reported within the 6-week risk period, regardless of Brighton level. These results warrant the use of active surveillance as well as continuous and careful analysis of post-vaccination GBS cases.

EFFECT OF ADD ON YOGA INTERVENTION ON PULMONARY FUNCTION IN CHILDREN WITH DUCHENNE MUSCULAR DYSTROPHY

Pradnya Dhargave, Atchayaram Nalini, Ragupathy Sendhil Kumar, Tiruchur Raju, Meghana Adoor, Raghuram Nagarathna, Kandhavelu Thennarasu, Talakad Sathyaprabha (Bangalore, India)

INTRODUCTION: Respiratory complications in Duchenne muscular dystrophy (DMD) is the major cause of morbidity and mortality. Therapies to maintain respiratory functions in these children are essential to delay ventilator dependency and to increase life expectancy. Physiotherapy is used as regular care in the management of respiratory functions in DMD. Yoga is used in the rehabilitation of asthma, Guillain–Barré syndrome, epilepsy, arthritis, musculoskeletal pains, etc. The added effect of yoga on pulmonary functions in DMD children has not been studied.

OBJECTIVE: To find the added effect of yoga practices on pulmonary functions in children with DMD.

METHODS: A total of 124 children with DMD (age range: 5–10 years) were randomly allocated to 2 groups. Group 1 consisted of home-based physiotherapy. Group 2 consisted of home-based yoga and physiotherapy. Pulmonary function tests—forced vital capacity (FVC), peak expiratory flow rate (PEFR), tidal volume (VT), maximal voluntary ventilation (MVV), minute ventilation test (MVT)—were performed every 3 months for a period of 1 year. RmANOVA and the least significant difference test was performed to compare the effect of treatments.

RESULTS: FVC improved in group 1 from 0.9±?? to 1.0±0.3 and in group 2 from 0.83±0.3 to 1.0±0.3 (F=14.165, p<0.001). PEFR improved in group 1 from 100.6±41.7 to 126.8±44.2 and in group 2 from 103.4±41.2 to 116.6±48.4 (F=4.818, p<0.001). MVV improved in group 1 from 28.1±9.7 to 34.6±11.0 and in group 2 from 28.7±9.4 to 32.6±10.7 (F=5.569, p<0.001).

SUMMARY/CONCLUSION: Adding yoga practices along with regular physiotherapy care in children with DMD was beneficial in the younger age group.
102 A CASE OF GAUCHER DISEASE WITH DOUBLE MUTATIONS OF N370S AND L444P RECEIVING ENZYME REPLACEMENT TREATMENT FOR THREE YEARS
Sleiman El Jamal, Jin Jun Luo (Philadelphia, PA)

INTRODUCTION: Gaucher disease (GD) is an inherited metabolic disorder due to β-glucosidase deficiency. There are 3 types of GD. Type I (94% of GD) is characterized by progressive lysosomal storage of glucocerebrosidase in macrophages predominantly in bones, bone marrow, liver, and spleen. Types II (1%, infantile form) and III (5%, adult form) have neurological involvement.

OBJECTIVE: To present an adult case of GD with double mutations receiving enzyme replacement therapy (ERT) for 3 years.

CASE REPORT: A Hispanic-American man was diagnosed with GD at age 10. At age 51, he presented for persistent severe headache and generalized body aches. His past medical history includes porencephaly, diabetes, traumatic brain injury, schizoaffective disorder, alcohol and illicit drug abuse, and splenectomy for hepatosplenomegaly with thrombocytopenia. His family history is notable for the death of his brother and cousin from GD. Genetic testing confirmed GD with double mutations (N370S, +/-, common in type I; and L4444P, +/-, types II and III). Laboratory studies disclosed abnormal β-glucosidase (0.10 [5.5-9.7] nmol/hr/ml; patient's value [normal reference]), angiotensin converting enzyme (337 [51-119] IU/L); chitotriosidase (25,765 [8-65] nmol/hr/ml); tartrate resistant acid phosphatase (35 [4.8-8.2] IU/L); IgA (1093 [81-463] mg/dL), free light chain (101.1 [3.3-19.4] mg/L) and kappa to lambda ratio (4.53 [0.26-1.65]). Electrophysiologic study disclosed pure motor neuropathy with significantly distal active denervation. IV infusion of Cerezyme® (imiglucerase, GenZyme Therapeutics, Cambridge, MA) significantly improved his symptoms.

RESULTS: A repeated electrophysiologic study 2 years after ERT initiation suggested improvement. Detailed data will be presented.

SUMMARY/CONCLUSION: This case supports the observation that longterm ERT is well tolerated and improves GD symptoms.

Sleiman El Jamal, MD, Resident and Fellow Member Award Recipient

103 MAINTENANCE THERAPY WITH SUBCUTANEOUS IMMUNOGLOBULIN IN PATIENTS WITH CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY: A LONGTERM FOLLOWUP
Brittany Harvey, Natalie Tucker, Adam Hart, Lara Katzin, Jerrica Farias, Nichole Jones, Samuel Dang, Clifton Gooch, Tuan Vu (Tampa, FL)

INTRODUCTION: Chronic inflammatory demyelinating polyneuropathy (CIDP) is an autoimmune neuropathy responsive to immunoglobulin (Ig). Following a study comparing subcutaneous Ig (SC Ig) to IV Ig treatment in terms of efficacy and quality of life (QOL, as a reflection of safety and tolerability) in patients with CIDP, 3 patients (to date) who had completed the trial chose to continue with SC Ig.

OBJECTIVE: To conduct a longterm followup on patients who continued on SC Ig after the trial to assess whether the benefits seen during the trial are durable.

METHODS: Using a phone survey, patients' QOL was assessed using the Rasch-built Overall Disability (R-ODS) and CIP-PRO20 scales. Patients were also surveyed regarding whether they had remained on SC Ig, whether they had relapses (requiring rescue or change in therapy), and their impression of SC Ig treatment.

RESULTS: One patient underwent autologous hematopoietic stem cell transplantation 7 months after completing the study and is no longer on treatment. She felt the effect was durable and had no relapse while on SC Ig. The other 2 patients remained on SC Ig 12 and 5 months after the study. The subjects' R-ODS and CIP-PRO20 scores remained unchanged since the time they completed the study. Neither had relapses, and both prefer SC Ig because of lack of side effects and stable control of symptoms.

SUMMARY/CONCLUSION: The preliminary data shows that SC Ig is well tolerated, and the improvement in QOL is persistent. In addition, efficacy appears durable, as none of the patients had worsening of symptoms or relapse while on SC Ig.
IMPACT OF AN AGGRESSIVE LIFESTYLE PROGRAM ON DIABETIC NEUROPATHY AS EVALUATED BY NERVE CONDUCTION STUDIES
James Horsley (Clay City, Kentucky)

INTRODUCTION: Therapeutic impact of lifestyle and drug/supplements on diabetic polyneuropathy have been documented. Only rarely has this been studied with lifestyle alone using nerve conduction studies as the outcome measure.

OBJECTIVE: Compare nerve conduction studies early, then later, in a lifestyle change program.

METHOD: Among many patients in the lifestyle program for 18 days for various diagnoses, ten with diabetic polyneuropathy went through nerve conduction studies on 15 nerves in the first few days of their stay. After one to two weeks of serum glucose monitoring during the aggressive diet, exercise and other lifestyle changes the nerve conduction studies were repeated.

RESULTS: Of the 15 nerves presented here, 7 motor and 8 sensory, 14 had improvement in the time of conduction in the terminal segment (the distal latency) and over half became normal. These normalized distal latencies had decreased (improved) an average of 25.2% from the first electrodiagnostic evaluation. The one exception to improvement of distal latency in the 15 nerves was in the only patient who had significant worsening of his glycemic control before the second evaluation compared to the glycemic control before the first.

The nerve conduction velocity was obtained in 6 of the 7 motor nerves. It improved in the 3 nerves of the 3 patients with the best glycemic control before the second electrodiagnostic evaluation. The other 3 nerves in which nerve conduction velocity changes were obtained all had negative changes, i.e. worse results, and these were all in patients with less than ideal glycemic control before the second electrodiagnostic evaluation.

Amplitude changes in the motor and sensory nerve responses between the two nerve conduction studies were very much less consistent than the above-mentioned results.

CONCLUSIONS: Substantial improvement in some aspects of nerve conduction in diabetic polyneuropathy can be achieved in one to two weeks of a lifestyle program. The therapeutic effect occurs primarily in the very distal nerve segment.

ANIMAL ASSISTED INTERVENTION AS A FORM OF QUALITY IMPROVEMENT IN A MULTIDISCIPLINARY AMYOTROPHIC LATERAL SCLEROSIS CLINIC
Kathleen Pearson (Richmond, VA), Scott Vota (Glen Allen, VA)

INTRODUCTION: Animal assisted interventions (AAIs), including animal assisted therapy, activities, and service animal activities, are well-known to provide benefits to patients’ well-being, both in inpatient and outpatient settings. Psychosocial benefits of AAIs for patients, families, and staff have been demonstrated in prior studies involving palliative care settings. There are limited studies regarding the impact of AAIs in the ALS patient population. Animal assisted therapy may be a complement to existing therapies available in these multispecialty clinics and may further improve quality of life for both patients and staff in this setting.

OBJECTIVE: To determine benefits to patients, family members, and staff of the addition of animal assisted therapy to existing therapies within a multidisciplinary ALS clinic.

METHODS: Over a 3-month period 10 canine therapy teams interacted with a total of 32 patients, family members, and staff at the Virginia Commonwealth University multidisciplinary ALS clinic. Surveys were completed by participants following the visits regarding overall satisfaction, mood, and quality of care following the visit and collected anonymously. Scores were reported with a 5-point rating scale.

RESULTS: Survey responses were overwhelmingly positive. Average scores were above 4 in all domains, including enjoyment and benefit of visit, improved mood, and perception of care in the clinic. No participants reported worsening of mood, lack of benefit from the visit, or worsened perception of quality of care.

SUMMARY/CONCLUSION: Animal assisted therapy is a useful adjunctive therapy in a multidisciplinary ALS clinic, improving mood and patient, family, and medical staff satisfaction.
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EXAMINING INPATIENT OUTCOMES FOR
CHRONIC INFLAMMATORY DEMYELINATING
POLYNEUROPATHY POPULATIONS
ADMINISTERED INTRAVENOUS
IMMUNOGLOBULIN AND PLASMA EXCHANGE
Tamara Opila (Sharon Hill, PA), Nizar Souayah (Newark, NJ)

INTRODUCTION: IVlg and plasma exchange (PE) are among the most common treatments for chronic inflammatory demyelinating polyneuropathy (CIDP). Long-term outcomes may include partial to complete remission, or progressive disability.

OBJECTIVE: To assess inpatient outcomes for CIDP populations treated with IVlg and PE, when stratifying by age and zip code classification of median income.

METHODS: A retrospective analysis using the Nationwide Inpatient Sample (NIS) was conducted for patients with a primary diagnosis of CIDP. Patient demographics were assessed; length of stay (LOS), total charges, and disposition status were also tabulated from 2003-2012.

RESULTS: Among 7,972 CIDP patients, the average age was 60.2±14.7 years; rates of mechanical ventilation were consistent among age groups. Despite the documented increased prevalence among men, nearly equal gender proportions were observed. Total charges and LOS averaged $46,971 and 4.5 days, respectively. IVlg was the primary treatment for 74.3% of patients, while 24.3% received PE and 1.4% were administered IV steroids. A significant difference in therapy cost was detected by longitudinal analysis; IVlg total average charges ranged from $20,238 to $72,641 and PE from $9,406 to $89,901 (p<0.001). The 3.5 day average LOS for IVlg patients was significantly shorter than PE (6.7; p<0.0001); IVlg patients also demonstrated a significantly greater rate for routine discharges (p<0.0001).

Patients in the lowest median income quartile were also more likely to be discharged to skilled nursing facilities and home health care compared to their higher income counterparts (p<0.0001).

SUMMARY/CONCLUSION: CIDP is heterogeneous in presentation and may require complementary treatments to achieve remission. Advanced age, comorbidities, and socioeconomic status may contribute to patient outcomes.

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EXAMINING ACUTE CARE PATIENT OUTCOMES
FOR POLYMYOSITIS AND DERMATOMYOSITIS
IMMUNOGLOBULIN AND INTRAVENOUS STEROID
TREATMENT GROUPS, USING THE NATIONWIDE
INPATIENT SAMPLE
Nizar Souayah (Newark, NJ), Tamara Opila (Sharon Hill, PA)

INTRODUCTION: IV steroids and IVlg are among the gold standard treatments for polymyositis and dermatomyositis; this population is at-risk for sequelae including generalized muscle weakness, infection, and arrhythmia.

OBJECTIVE: To assess inpatient outcomes for polymyositis and dermatomyositis cases treated with IV steroids and IVlg, when stratifying by age and median income quartiles.

METHODS: A retrospective analysis (2003-2012) of the Nationwide Inpatient Sample (NIS) was conducted by extracting aggregate patient data using primary procedure codes for these neuromuscular disorders. Case demographics and disposition status, mortality rates, length of stay (LOS), and total charges were tabulated.

RESULTS: Average ages for polymyositis and dermatomyositis patients were 50.9±14.9 and 28.0±22.4 years, respectively. Racial distributions varied greatly among the 2 disorders (p<0.001), with twice the number of African-American polymyositis cases observed. The average LOS for patients receiving IV steroids was 4.5 days, versus 3.1 for IVlg (p<0.001). A longitudinal analysis compared the cost of both therapies; IVlg total charges ranged from $20,201 to $79,523 and IV steroids from $11,996 to $69,766 (p<0.001). Total charges were significantly different when comparing age groups (p<0.001). Patients 41-57 years old displayed the highest average total charges ($57,051) and the only mortalities, while those older than 57 demonstrated a significantly greater discharge rate to longterm care and home health care (p<0.0001). Hospital charges and LOS were not associated with patient income level.

SUMMARY/CONCLUSION: IVlg and IV steroids are effective treatments for this population, however IVlg is more widely utilized in the acute care setting by 10:1. Indications such as refractory disease may factor into therapy selection.
INVESTIGATION OF AUTONOMIC FINDINGS BEFORE AND AFTER BOTULINUM TOXIN INJECTION
Gazi Yozgatlı (Eskisehir, Odunpazari), Yasar Kutukcu, Umit Ulas, Hakan Akgun, Erkan Tokgoz, Semir Mazman, Oguzhan Oz, Uzeyir Erdem, Zeki Odabasi (Eskisehir, Turkey)

INTRODUCTION: Botulinum toxin (BoNT), known as the most powerful neurotoxin, has been used for many years especially effectively in the treatment of dystonia in neurology clinics.

OBJECTIVE: Normally, BoNT shows local effect on applied areas involving the muscles and glands. However, according to recent research, it has been illustrated that BoNT causes a distant effect in a way of hematogenic or retrograde axonal transport along with its local effect. Since it is well-known that the autonomic nervous system (ANS) is affected in patients with botulism, the goal of this study is to investigate the distant effect of BoNT injection on autonomic functions.

METHODS: In order to assess autonomic functions, pupillometry and vasomotor reactivity were measured by transcranial doppler tests used in patients receiving BoNT injections. Measurements before injection were compared with measurements 7-10 days after injection when maximum effect starts to be seen.

RESULTS: There were no significant differences in the results of both tests. Nevertheless, it was found that middle cerebral artery average flow velocity, a sub-parameter in this study, was significantly lower in the group applied with a low dose of BoNT on facial areas.

SUMMARY/CONCLUSION: There are 3 possible explanations for this decrease: it may be caused by the effects of local BoNT, the distant effect on the ANS without a dose-dependent BoNT, and more likely by technical differences. Thus, BoNT, which has been used for many years, does not have serious complications and side effects on autonomic functions.

TAKOTSUBO CARDIOMYOPATHY IN AMYOTROPHIC LATERAL SCLEROSIS
Kyomin Choi, Seok-Jin Choi, Yoon-Ho Hong, Jun-Soon Kim, Seol-Hee Baek, So Hyun Ahn, Chan Soon Park, Sung-Min Kim, Je-Young Shin, Jung-Joon Sung (Seoul, South Korea)

INTRODUCTION: Respiratory insufficiency is the primary cause of death in advanced stages of ALS. Meanwhile, cardiomyopathy has rarely been reported in ALS patients. Takotsubo cardiomyopathy (TSC) represents an acute heart failure syndrome characterized by “reversible” left ventricular dysfunction. Several case reports involving occurrence of TSC in ALS patients have been reported in the literature.

OBJECTIVE: To investigate the clinical presentation and prognosis of TSC in patients with ALS.

METHODS: Detailed clinical, laboratory, and cardiovascular data from 75 ALS patients who underwent echocardiographic evaluation for various reasons at a single referral center between January 2006 and December 2015 were reviewed.

RESULTS: Nine of 75 patients (12%) were diagnosed with TSC (mean age: 61.3 years, age range: 55-71 years) and the median disease duration of ALS was 51.5 months (range: 18-134 months). All patients were at advanced stages of ALS with various degrees of chronic respiratory compromise and 5 were bedridden. Acute dyspnea was an invariable presentation in all patients, and chest discomfort mimicking acute coronary syndrome was present in 2 patients. Six patients underwent significant hypotension. Three patients showed altered mentality and 2 of them suffered cardiopulmonary arrest.

SUMMARY/CONCLUSION: TSC is not an uncommon condition in ALS. It presents invariably as acute dyspnea, with or without chest discomfort, in advanced stages of ALS, and two-thirds of patients may have serious in-hospital complications. This study suggests that TSC might be under-recognized in ALS patients and highlights the need for proper cardiac evaluation and management for this condition.
ACUTE ONSET CHRONIC INFLAMMATORY Demyelinating Polyneuropathy Following Anthrax Vaccination
Kyomin Choi, Seok-Jin Choi, Min Kyung Kang, Jun-Soon Kim, Seol-Hee Baek, So Hyun Ahn, Yoon-Ho Hong, Sung-Min Kim, Je-Young Shin, Jung-Joon Sung (Seoul, South Korea)

INTRODUCTION: Chronic inflammatory demyelinating polyneuropathy (CIDP) typically presents as either a relapsing or progressive neuropathy with proximal and distal weakness which develops over at least 8 weeks. However, acute onset CIDP (A-CIDP) is an increasingly recognized type of CIDP. It is crucial to distinguish A-CIDP from Guillain–Barré syndrome with treatment-related fluctuations (GBS-TRF) because of the differences in the therapeutic strategies and prognosis.

OBJECTIVE: To report a rare case of CIDP following anthrax vaccination and emphasize the importance of differentiating A-CIDP from GBS-TRF.

CASE REPORT: A 33-year-old man with hypothyroidism and dyslipidemia presented with myalgia and tingling sensation in 4 extremities of acute onset after anthrax booster vaccination. Neurological examination showed hypoesthesia in the medial side of the bilateral forearm and palms without motor weakness, and deep tendon reflexes were preserved. NCSs revealed demyelinating sensorimotor polyneuropathy. The patient was treated with IVIg 400 mg/kg for 5 days, and sensory symptoms almost disappeared. However, bilateral arm weakness developed a week after discharge (MRC grade 4). Furthermore, NCSs showed aggravated results compared to the previous study performed at 2 weeks prior. Oral prednisolone 60 mg daily was prescribed and the patient had considerable improvement. Nonetheless, he complained of slowly progressive weakness, once the steroid was discontinued 4 months from onset. The patient was diagnosed with A-CIDP rather than GBS-TRF.

SUMMARY/CONCLUSION: CIDP could be one of the important side effects of anthrax vaccination. The diagnosis of A-CIDP should be carefully considered, if the clinical deterioration occurs after 8 weeks from onset.

EARLY DETECTION OF FASCICULATIONS IN AMYOTROPHIC LATERAL SCLEROSIS BY ULTRASONOGRAPHY
Hazem Elkarabaty (Alex, Egypt), Franz Glocker (Freiburg, Germany)

INTRODUCTION: ALS is a fatal progressive neurodegenerative disorder. The combination of overactive tendon jerks with weak wasted fasciculating muscles was taken as sign of ALS. Many physicians report that the first symptom of ALS was fasciculations that may have anteceded weakness by several years. Fasciculations are a frequent ultrasonographic (US) sign in neuromuscular diseases. It has been proved that US is more sensitive than electromyographic examination in visualizing fasciculations in patients with lower motor neuron disorders. Additionally, it is more reliable than the clinical examination.

OBJECTIVE: To detect and quantify US recorded fasciculations of the tongue and extremity muscles in patients with definite and suspected ALS according to El Escorial criteria.

METHODS: Forty patients (20 with a definite diagnosis of ALS and 20 with a suspected diagnosis) were studied along with 30 control subjects. A transverse (M-mode) US examination (5-10 MHz) of the tongue, extremity muscles, and paravertebral muscles was conducted using an ATL Ultramark HDI apparatus (Philips Healthcare) along with NCSs and needle EMG of the selected muscles.

RESULTS: US is an easy way to detect deep fasciculations in the tongue and extremity muscles. The US results showed a significant increase of the number and mean of duration of fasciculations after 6 months.

SUMMARY/CONCLUSION: US is an easy method to detect deep fasciculations in patients with a suspected diagnosis of ALS that could not be detected clinically or by needle EMG. An increase in the number and mean of duration of fasciculations in followup US for patients with a suspected diagnosis of ALS will help to confirm the diagnosis.

Hazem El-Karabaty, MD, Recipient of the 2016 IFCN North American Chapter Fellowship Award
BRUGADA SYNDROME AND OTHER CARDIAC ISSUES IN AMYOTROPHIC LATERAL SCLEROSIS
Rohit Gummi, Raghav Govindarajan (Columbia, MO)

BACKGROUND: Primary cardiac involvement is uncommon in ALS. Reported here are 3 cases where patients developed cardiac disease including the first case of Brugada syndrome reported in ALS.

CASE REPORTS: (1) Brugada syndrome: A 45-year-old woman presented with right upper limb weakness and fasciculations and was diagnosed with ALS. She was wheelchair bound and had developed bulbar symptoms when she presented with sudden onset shortness of breath. CT of the chest did not show pneumonia; CT angiogram of chest did not show pulmonary embolus. Electrocardiogram (EKG) showed pronounced elevation of the J point, a coved-type ST segment, and an inverted T wave in V1 and V2 diagnostic of Brugada syndrome. Extensive workup for reversible causes was nondiagnostic. (2) Dilated cardiomyopathy: A 70-year-old man presented with lower limb weakness and was wheelchair bound before he was diagnosed with ALS. Three months later he presented with sudden onset shortness of breath. CT of the chest showed pulmonary edema; CT angiogram did not show pulmonary embolus. An echocardiogram revealed an ejection fraction of 20% with dilated left ventricular cavity. Extensive workup for reversible causes was nondiagnostic. (3) Atrial fibrillation: A 62-year-old woman presented with slurred speech and difficulty in swallowing and was diagnosed with ALS. Two months after diagnosis she presented with sudden onset shortness of breath. CT/angiogram of the chest was normal. EKG revealed atrial fibrillation with rapid ventricular rate. She was treated with IV sotalol and placed on digoxin treatment.

CONCLUSION: While pulmonary issues are a common cause of dyspnea in ALS, cardiac involvement can be a potentially life threatening presentation.

Rohit Gummi, BS, Resident and Fellow Member Award Recipient

LUCK OF THE DRAW: A CASE OF CO-MORBID AMYOTROPHIC LATERAL SCLEROSIS AND MULTIPLE SCLEROSIS
Nasheed Jamal, Jonathan Morrill, Matthew Harmelink, Patricia Pittman (Los Angeles, CA), Shrikant Mishra (North Hills, CA)

INTRODUCTION: A neurodegenerative disorder affecting upper and lower motor neurons, ALS has an incidence rate of 1.8/100,000 in North America. Multiple sclerosis (MS), an autoimmune disease involving the CNS, has an annual incidence of 7.6-20.6/100,000 in Canada and the United States. Their concurrent presence in 1 patient is exceptional.

OBJECTIVE: To report a case of a patient initially diagnosed with relapsing-remitting MS and later confirmed to have co-morbid ALS.

CASE REPORT: A 61-year-old right-handed man presented at age 50 years with episodic “clumsy feet,” “funny” feeling in his legs, and difficulty ambulating. Neurological examination revealed ataxia. Initial MRI of the brain and lumbar puncture were consistent with MS. After starting Avonex®, he experienced several exacerbations with worsened ataxia and gait impairment that resolved with IV steroids. Four years later, he developed increased left lower extremity tone and right upper extremity weakness. Avonex® was switched to Rebif®. Needle EMG/NCSs in 2010 were normal. Repeat studies in February 2013 showed neurogenic changes in all C5-T1 muscles tested. Examination demonstrated decreased strength in all extremities. Needle EMG/NCSs in April 2013 revealed abnormal spontaneous activity in bilateral upper extremities (BUE) and lumbosacral paraspinal muscles. He later reported significant BUE weakness that has since progressed. In September 2014, tongue fasciculations were noted. That Thanksgiving, he began having difficulty feeding and toileting and began feeling painless muscle twitches. In September 2015, needle EMG/NCSs showed evidence of motor axonopathy involving the lumbar, cervical, and bulbar myotomes consistent with motor neuron disease in this clinical setting.

CONCLUSION: This case illustrates that 2 uncommon neurological diseases, MS and ALS, may manifest contemporaneously.
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OPTIMIZING ELECTRICAL IMPEDANCE MYOGRAPHY OF THE TONGUE IN AMYOTROPHIC LATERAL SCLEROSIS
Courtney McIlduff, Sung Yim, Adam Pacheck, Seward Rutkove (Boston, MA)

INTRODUCTION: Electrical impedance myography (EIM) can quantify muscle health at a range of frequencies, including that most commonly employed: 50 kHz. However, disease-related changes in EIM data suggest the distinction between normal and patient EIM values could be more apparent at frequencies above 50 kHz.

OBJECTIVE: To investigate at what other selected frequencies tongue EIM might differentiate healthy individuals and ALS patients, remain reliable, and correlate with a standard metric of bulbar function.

METHODS: Tongue EIM phase data from 30 volunteers and 11 ALS patients were analyzed at 6 discrete frequencies from 50 to 500 kHz.

RESULTS: Of the frequencies assessed, EIM demonstrated maximal separation and reliability at 100 kHz, where phase value was also significantly correlated with the ALS Functional Rating Scale, Revised Bulbar Sub-score.

SUMMARY/CONCLUSION: One-hundred kHz could serve as an optimal frequency at which to measure EIM phase values of the tongue in ALS. Additional work is needed to further characterize the role of tongue EIM as a biomarker of bulbar dysfunction in ALS and other neurological disorders.

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DIAGNOSTIC DILEMMA WITH AMYOTROPHIC LATERAL SCLEROSIS
Syed Shah, Raghav Govindarajan (Columbia, MO)

BACKGROUND: ALS is a devastating disease with no known cure. Reported here are 4 cases in which clinical mimickers delayed diagnosis and thus quality of care.

CASE REPORTS: (1) Alcoholic neuropathy: A 70-year-old male developed lower limb weakness and numbness/tingling. He was diagnosed with alcoholic neuropathy on a limited EDX examination which showed small sural responses. He was treated with Vitamin B1 but in a year weakness had spread to the upper limbs. He had seen 3 physicians before his ALS diagnosis. (2) Hyperparathyroidism: A 65-year-old female developed right upper limb weakness and fasciculations. She had hypercalcemia and was diagnosed with primary hyperparathyroidism. The symptoms were attributed to this and underwent parathyroidectomy. Despite surgery the weakness progressed to other limbs along with bulbar symptoms. Four physicians and 1.5 years later ALS was diagnosed. (3) Autoimmune neuropathy/oligoclonal bands: A 46-year-old female presented with right upper limb weakness and fasciculations. Workup showed 10 oligoclonal bands in cerebrospinal fluid. She was treated with high-dose steroids and immunosuppressive therapy for a year by 2 physicians, with weakness spreading to lower limbs before her ALS diagnosis. (4) Lumbosacral radiculopathy: A 75-year-old male presented with right foot drop. MRI of the lumbar spine showed compression, and he underwent surgery. His weakness spread to other leg, and he had revision surgery. He had esophageal stretching for dysphagia. Three years and 10 physicians later he was diagnosed with ALS.

CONCLUSION: ALS remains a clinical diagnosis performed by an expert neuromuscular physician. Diagnostic biomarkers are urgently needed to reduce lag so that patients can be directed to multidisciplinary clinics sooner.

Syed Shah, MBBS, MD, Resident and Fellow Member Award Recipient
A CASE OF SCHWARTZ–JAMPEL SYNDROME: A RARE MYOTONIC-LIKE DISORDER
Ashish Khanna, Mohammad Aalai (Brooklyn, NY), Joan Hou, Kirill Alekseyev, Armando Ianicello, Todd Lefkowitz, Adrian Cristian (Brooklyn, NY)

INTRODUCTION: Schwartz–Jampel syndrome (SJS), or chondrodystrophic myotonia, is a rare congenital myotonic-like disorder with a prevalence of less than 1:1,000,000. The gene affected is known to encode for perlecan, which is responsible for heparin sulfate proteoglycan that results in abnormal myelination.

OBJECTIVE: To investigate the natural history and needle EMG/NCS features of SJS type 1.

CASE REPORT: A 33-year-old female was diagnosed with SJS at around age 3 with the help of needle EMG/NCSs. She had classic features including blepharophimosis, short stature, puckered lips, and skeletal abnormalities. She had multiple orthopedic and ophthalmologic procedures at a young age, including cervical spinal surgery for repair of a congenitally short neck. She has symptoms of myotonia in all 4 extremities, which is controlled with medication. The patient presented to the authors' Acute Inpatient Rehabilitation Facility after an ICU stay including tracheostomy with mechanical ventilation for respiratory failure. Physical examination revealed distal atrophy with preserved proximal musculature, genu varum, but intact strength within range of all limbs. Needle EMG findings included fasciculations and complex repetitive discharges as well as myotonic discharges. The NCSs revealed a mild sensory median nerve entrapment at the wrist, which those with SJS are predisposed to have.

SUMMARY/CONCLUSION: SJS, also known as chondrodystrophic myotonia, is a congenital myopathy that has been reported in consanguineous parents. This case describes a patient with known type 1 disease that had multiple classic physical examination and EDX features of this extremely rare disorder.

DIVERSE ETIOLOGIES OF HYPERCKEMIA IN CHILDREN
Fouad Alghamdi, Partha Ghosh (Boston, MA)

INTRODUCTION: Creatine kinase (CK) is a sensitive marker of muscle diseases.

OBJECTIVE: To identify various etiologies of hyperCKemia in children.

METHODS: A retrospective chart review of children with elevated CK evaluated in the authors' Neurology clinic over a 10-year period was conducted. Elevated CK was defined as values greater than 150 U/L (4-150) or greater than 175 U/L (4-175) based on laboratory control subjects.

RESULTS: Of the 1948 children with hyperCKemia identified, 468 (24%) had various neuromuscular disorders: genetic myopathies (muscular dystrophies, congenital myopathies) either confirmed by genetic testing or suspected based on clinical features, needle EMG, or muscle biopsy—243 (52%); inflammatory myopathies (dermatomyositis, polymyositis, and associated with other connective tissue disorders)—21 (4.5%); metabolic myopathies (glycogen/lipid metabolism disorders, mitochondrial myopathies, unspecified rhabdomyolysis)—115 (24.5%); muscle channelopathies (periodic paralysis, myotonic disorders)—24 (5%); infectious/drug induced myositis—12 (2.5%); and neurogenic causes (spinal muscular atrophy, neuropathy)—53 (11.5%). These patients presented with 1 or more symptoms suggestive of muscle disorders (weakness, myalgia, cramps/stiffness, rhabdomyolysis). In addition, a significant proportion (1480, 76%) had miscellaneous causes (genetic/metabolic disorders, neurodevelopmental disorders, brain malformations/epilepsy, developmental delays/hyponatremia, movement disorders, CNS infections, hypoxic/ischemic events, arthrogryposis) where symptoms for muscle disorders were nonspecific.

CONCLUSION: CK is an inexpensive screening tool to diagnose muscle disorders. HyperCKemia along with symptoms suggestive of muscle disorders increases the final diagnostic yield of myopathies. Neurogenic disorders particularly spinal muscular atrophy can have high CK and mimic myopathies which should always be considered in the differential diagnosis.

Fouad Alghamdi, MBBS, Resident and Fellow Member Award Recipient
SPECTRUM OF NON-DYSTROPHIC PEDIATRIC MUSCLE CHANNELOPATHIES
Fouad Alghamdi, Partha Ghosh (Boston, MA)

INTRODUCTION: The skeletal muscle channelopathies are a heterogeneous group of disorders whose clinical manifestations range from flaccid paralysis to myotonia.

OBJECTIVE: To describe the spectrum of non-dystrophic muscle channelopathies in children.

METHODS: A retrospective review of children with channelopathies diagnosed in a pediatric neuromuscular clinic was conducted. The diagnosis was established by clinical features, needle EMG findings, and/or genetic testing.

RESULTS: Thirty-one patients were identified in this cohort. Fifteen children had periodic paralysis (PP). Twelve of them had hypokalemic PP (CACNA1S-5, unknown genetic cause [3], Andersen–Tawil syndrome, KCNJ-3, not confirmed [1]) and electrocardiogram abnormalities were detected in 7; preventive treatment included acetazolamide (7) and spironolactone (1). Three children had hyperkalemic PP (SCN4A-3) and clinical myotonia detected in 1; preventive treatment included acetazolamide (1). Sixteen children had non-dystrophic myotonia. Six of them had a chloride channelopathy (CLCN1, Thomsen disease [5]), Becker disease [1]), 5 had clinical myotonia, and all had electrophysiological myotonia. Ten of them had a sodium channelopathy (SCN4A, autosomal dominant myotonia congenita [8], paramyotonia [2]), 8 had clinical myotonia, and 7 had electrophysiological myotonia (needle EMG not performed [3]). One presented with neonatal hypotonia and feeding and respiratory difficulties. Out of 16 children with non-dystrophic myotonia, 11 received treatment (mexiletine [10], acetazolamide [1]).

CONCLUSION: Non-dystrophic muscle channelopathies are a rare but distinct group of neuromuscular disorders in children with diverse clinical manifestations (PP, proximal weakness, myotonia, muscle hypertrophy, and neonatal feeding and respiratory difficulties). Cardiac arrhythmias can be life-threatening in hypokalemic PP. Acetazolamide and mexiletine are mainstays of therapy in PP and myotonia, respectively.

Fouad Alghamdi, MBBS, Resident and Fellow Member Award Recipient

UNIQUE MUSCLE PATHOLOGICAL FINDINGS IN A PATIENT WITH VERY-LONG-CHAIN ACYL-COA DEHYDROGENASE DEFICIENCY
Nigel Arruda, Amarilis Sanchez-Valle (Tampa, Fl), Jonathan Vu (Providence, RI), Lara Katzin, Clifton Gooch, Tuan Vu (Tampa, FL)

INTRODUCTION: Very-long-chain acyl-CoA dehydrogenase (VLCAD) is an enzyme involved in the oxidation of fatty acids, a crucial energy source for muscle when glycolysis cannot meet metabolic demands. VLCAD deficiency leads to cardiomyopathy, myopathy, recurrent rhabdomyolysis, and myoglobinuria.

OBJECTIVE/RESULTS: A 29-year-old woman with VLCAD deficiency had occasional, brief episodes of diffuse muscle pain and weakness since childhood. At 18, she had an acute episode of severe weakness and elevated creatine kinase (CK) (77,000 IU/L). She was diagnosed with polymyositis and treated with azathioprine and prednisone. Her symptoms continued, and at age 20 she was switched to mycophenolic acid and Solu-Medrol®. At 22, after prolonged walking, she had an acute exacerbation of severe weakness with inability to ambulate, nausea, vomiting, acute renal failure (requiring hemodialysis), and CK elevation to 33,000 IU/L. Needle EMG showed myopathic changes and spontaneous activity, consistent with an inflammatory myopathy. A muscle biopsy showed necrotizing myopathy with multifocal myofiber sarcoplasmic calcification without evidence of metabolic or mitochondrial myopathies. An acylcarnitine panel showed VLCAD deficiency. With dantrolene and medium chain triglyceride (MCT) oil, she did not experience further acute exacerbations but continued to have intermittent and mild muscle weakness and pain with normal-to-mildly elevated CK.

DISCUSSION: The sarcolemmal calcification is an unusual, previously unreported, feature of VLCAD deficiency. The sarcolemmal calcification is thought to be due to tissue damage or necrosis from ischemia.

SUMMARY/CONCLUSION: Sarcolemmal calcification can be a feature of VLCAD deficiency, and dantrolene may offer protection from attacks of rhabdomyolysis and weakness.
UTILITY OF ULTRASOUND IN MUSCLE BIOPSY
Santoshi Billakota, Lisa Hobson-Webb (Durham, NC)

INTRODUCTION: Core or needle muscle biopsies are generally performed on previously selected muscle for diagnoses of various muscle pathologies. At the authors’ institution, biopsies are performed by trained neuromuscular specialists on muscle selected utilizing needle EMG findings without ultrasound (US) guidance.

OBJECTIVE: To determine whether US guidance of needle EMG selected muscle will increase the diagnostic yield of the biopsy.

METHODS: Two trained physicians performed biopsies based upon prior clinical and EDX examination results. Forty consenting patients underwent core muscle biopsy and were randomized for additional US guidance. Demographic information and results of prior needle EMG testing was recorded. Amount of tissue in each biopsy was recorded along with the final pathologic diagnosis.

RESULTS: From the 40 patients studied, 16 of the muscle biopsies were performed with US guidance. Out of these, 50% had a definitive diagnosis, 38% had findings without a definitive diagnosis, 1 patient was intolerant, and 1 of patient had an inadequate biopsy sample. Out of the non-US guidance group, 58% had a definitive diagnosis, 33% had no definitive diagnosis, 1 patient was intolerant, and 1 patient had an inadequate biopsy sample. There were no significant differences found between the 2 groups and no complications occurred.

CONCLUSION: Both US and non-US guidance groups performed equivocally when biopsy was performed on muscles previously selected by needle EMG. Guiding more specific site selection within the muscle did not improve outcomes. Further studies are proposed with omission of needle EMG muscle selection to determine if US alone offers a noninvasive, inexpensive alternative.

NEEDLE ELECTROMYOGRAPHY IDENTIFICATION OF CHRONIC MUSCLE SPASM
Roger Coletti (Lewes, DE)

INTRODUCTION: Multiple treatments for muscle spasm are utilized without regard to the physiology of the muscle. Muscles in chronic spasm can persist for decades and require specialized treatment.

OBJECTIVE: The unique physiology of chronic muscle spasm has not been well described. Systematic differentiation is needed to distinguish between temporary and chronic muscle spasm.

METHODS: Patient selection required a pain syndrome of greater than 1 month. Physical examination of reported sites of muscular pain were evaluated by compression to identify sites of non-compressibility and discomfort. Needle EMG evaluation consisted of identification of spontaneous electrical activity (SEA) also know as “end-plate noise.” Treatment of only those sites with significant SEA was performed with onabotulinumtoxinA or phenoxybenzamine/lidocaine until no residual SEA was present.

RESULTS: More than 100 patients were evaluated and treated. Selective chemodenervation based upon needle EMG identified SEA activity resulted in longterm relief, regardless of the duration of the prior pain syndrome, with a single treatment. Muscle spasms that did not exhibit SEA were treated with conventional therapies with reported good outcomes.

SUMMARY/CONCLUSION: SEA represents the needle EMG physiologic identifier and likely etiology of chronic muscle spasm. Hypothetically, longterm neurotransmitter blockade with these agents relieving chronic spasm allows normalization of spasm-induced restricted circulation, facilitating membrane stabilization and preventing recurrence of SEA-induced chronic muscle spasm.

CLINICAL RELEVANCE: Needle EMG of muscle in apparent spasm provides the ability to differentiate and treat selectively muscle that will return to normal function with conventional treatment and muscle that is in a state of chronic spasm requiring selective interventions.
CLINICAL AND PATHOLOGICAL HETEROGENEITY IN A FAMILY WITH NOVEL ACTA1 MUTATION
Priya Dhawan, Eric Sorenson, Teerin Liewluck, Milone Margherita (Rochester, MN)

INTRODUCTION: Mutations in skeletal muscle α-actin gene (ACTA1) cause congenital myopathies, manifesting with a spectrum of clinical severity and pathological findings, including nemaline bodies, actin aggregates, cores, zebra bodies, and fiber type disproportion.

OBJECTIVE: To describe a novel ACTA1 mutation and associated myopathy.

CASE/RESULTS: The proband is a 68-year-old male with foot drop since childhood. In his 50s his leg weakness worsened; he then developed arm weakness, dyspnea, and dysphagia. Examination showed facioscapuloperoneal weakness. Needle EMG revealed myopathic and neurogenic findings with minimal fibrillations. Triceps and gluteal biopsies demonstrated amorphous material, surrounded by a zone lacking oxidative enzyme reactivity, and type 1 fiber predominance. Whole exome sequencing detected novel ACTA1 (c.757G>C; p.G253R) and 2 TTN variants. The proband's son is a 34-year-old with normal motor development and reduced arm bulk. He developed arm weakness in his 30s. Examination showed arm weakness, pes cavus, and finger hyperextensibility. The proband's daughter is a 31-year-old with normal motor development and is a lifelong slow runner, who in her 20s developed leg weakness. Examination showed mild distal weakness, pes cavus, and ankle contractures. Needle EMG revealed myopathic units in weak muscles. Triceps biopsy (daughter) showed nemaline bodies, core-like structures, and type 1 fiber predominance. Sanger sequencing confirmed the ACTA1 and 1 TNN variant in both.

SUMMARY/CONCLUSION: The novel ACTA1 variant is predicted as deleterious, segregating with myopathy in an autosomal dominant pattern. The daughter's nemaline myopathy supports ACTA1 pathogenicity and underscores pathological heterogeneity of actinopathies. The possibility that TTN variants contributed to the proband's more severe phenotype cannot be excluded.

IS CORTICOSTEROID TREATMENT ASSOCIATED WITH SLEEP-RELATED BREATHING DISORDERS IN DUCHENNE MUSCULAR DYSTROPHY? A PRELIMINARY STUDY
Elias Karroum, Manisha Malik, Han Phan (Atlanta, GA)

INTRODUCTION: Duchenne muscular dystrophy (DMD) is an X-linked neuromuscular disorder due to mutations in the dystrophin gene. This results in progressive muscle degeneration leading to severe ambulation restriction, cardiopulmonary insufficiency, and death. Corticosteroids remain the only recommended therapy with established results, but they have many adverse effects, including weight gain, cataracts, and glucose intolerance. Weight gain in DMD patients, along with their weakened upper airway and respiratory muscles, can result in sleep disordered breathing.

OBJECTIVE: To study the impact of corticosteroids treatment on sleep-disordered breathing in DMD patients.

METHODS: Forty-four DMD patients (age range: 14.3±3.5 years; BMI z-score=0.1±2.0; 23% ambulatory) were retrospectively studied based on treatment (n=18) or not (n=26) with corticosteroids. Polysomnographic data were collected on total sleep time, sleep efficiency, arousals/hour of sleep, percentages of rapid eye movement (REM) and supine sleep, and respiratory measures including: apneas–hypopneas/hour of sleep (overall/during supine sleep/during REM sleep/strictly obstructive/strictly central), oxygen desaturations/hour of sleep, hypoxic burden (% SpO2<90%), nadir SpO2, and hypercapnic burden (% PCO2>50 mmHg).

RESULTS: Patients on corticosteroids were more ambulatory (44% versus 8%, p=0.008). They had similar obstructive sleep apnea (50% versus 65%, p=0.3) and hypoventilation (6% versus 8%, p=1.0) prevalences compared to patients not on corticosteroids. They also had a lower hypoxic burden (0.01±0.03% versus 0.3±0.9%, p=0.04) and tendency for a lower hypercapnic burden (3.7±11.5% versus 6.8±19.2%, p=0.06) and percentage of supine sleep (52±39% versus 70±38%, p=0.07).

SUMMARY/CONCLUSION: These preliminary data do not suggest a clinically significant association between corticosteroids treatment and sleep disordered breathing in DMD patients.

Elias Karroum, MD, PhD, Resident and Fellow Member Award Recipient
A CASE OF MYOSITIS ASSOCIATED WITH CHRONIC LYME DISEASE
Serge Khelemsky, Shan (Sarah) Chen (New Brunswick, NJ), James Van Gurp (New Brunswick, NJ)

INTRODUCTION: Inflammatory myositis associated with Lyme disease is rare and typically presents as a focal myositis. Diffuse myositis with a normal creatine phosphokinase (CPK) level is even more rare and is reported here.

OBJECTIVE: To illustrate the importance of including myositis within the differential diagnosis in scenarios involving myalgias in patients with chronic Lyme disease.

CASE REPORT: A 62-year-old woman with chronic Lyme disease presented with a new onset severe low back pain radiating down both legs over the course of 2 weeks, along with progressive gait dysfunction over several years to the point of being wheelchair bound. She was previously treated for Lyme disease with multiple antibiotic regimens. On examination, both lower extremities had symmetric proximal weakness (1/5 on MRC scale) and hyporeflexia, reduced distal vibration sense, and severe pain with movement. CPK was normal. Cerebrospinal fluid showed mild pleocytosis, elevated protein, and negative Lyme titers by PCR. MRI of the right thigh showed diffuse muscle edema suggesting inflammatory myopathy. Needle EMG showed diffuse irritable myopathy, lumbosacral polyradiculopathy, and sensorimotor axonal polyneuropathy. Left quadriceps muscle biopsy showed interstitial focal chronic lymphoplasmacytic inflammation, consistent with Borrelia burgdorferi infection. She was given a short course of IV steroids, which helped relieve her pain without significant improvement in strength.

SUMMARY/CONCLUSION: Lyme myositis, while rare, can present with debilitating clinical patterns including profound myalgias and progressive weakness. CPK can be normal if the inflammation is mostly interstitial. Needle EMG, muscle specific MRI, and muscle biopsy can help to establish the diagnosis and guide appropriate therapies.

WHAT CAUSED THE FOOT DROP? ELECTRODIAGNOSIS IN CONJUNCTION WITH ULTRASOUND SPARED A PATIENT UNNECESSARY NEUROSURGERY
Rupali Kumar, Neal Varghis (Redwood City, CA), Roger Klima (Palo Alto, CA)

INTRODUCTION: Foot drop can be caused by lesions at multiple different levels, from brain or spinal cord injury to motor neuron disease to radiculopathy to peripheral mononeuropathy to myopathy.

OBJECTIVE: To describe a case in which a patient with multiple potential etiologies for foot drop was diagnosed by EDX testing and imaging together, and thus spared unnecessary neurosurgery.

CASE REPORT: A 56-year-old male with facioscapulohumeral muscular dystrophy (FSHMD) presented with chronic low back pain and new right foot drop, with tibialis anterior (TA) weakness on examination. Lumbar spine MRI showed moderate-to-severe right L4-5 neuroforaminal stenosis, concerning for radiculopathy. The neurosurgery department scheduled the patient for L4-5 laminectomy/foraminotomy/discectomy. Prior to surgery, needle EMG was performed, revealing acute denervation, polyphasic motor units, and reduced recruitment in the right TA. Decreased right TA insertional activity indicated a partial loss of muscle and replacement with either adipose tissue (classically seen in FSHMD) or fibrosis (seen in denervation from radiculopathy). The quality of needle penetration was soft, consistent with adipose tissue. Ultrasound demonstrated muscle atrophy and areas of increased echogenicity consistent with fat. Fatty atrophy was redemonstrated on MRI. These findings suggested that FSHMD was likely the major contributor to the right foot drop, and not radiculopathy.

SUMMARY/CONCLUSION: A careful diagnostic approach is required when multiple potential etiologies coexist in 1 patient. While needle EMG alone could not have excluded radiculopathy, in concert with diagnostic ultrasound and MRI, the authors were able to determine the correct diagnosis. As a result, the patient was spared an inappropriate and potentially morbid neurosurgical procedure.
SEVERE AXIAL WEAKNESS IN ANOCTAMINOPATHY
Hani Kushlaf (Cincinnati, OH)

INTRODUCTION: Anoctaminopathy has a variable phenotype that includes limb-girdle muscular dystrophy type 2L (LGMD 2L), distal myopathy, and exercise intolerance with rhabdomyolysis. Axial weakness in anoctaminopathy has not been reported.

OBJECTIVE: To report the clinical history, examination, and diagnostic testing in a patient with severe axial weakness caused by anoctaminopathy.

CASE REPORT: A 53-year-old female presented with leg weakness. Symptoms began with thoracolumbar scoliosis in her early 20s. This led to discovering hyperCKemia that ranged 1000-3495 mg/dl. In her early 30s, she noted progressive difficulty walking up inclines. A maternal male second cousin has Duchenne muscular dystrophy. Examination revealed significant paraspinal muscle atrophy, thoracolumbar dextroscoliosis, mild difficulty standing up without arm support, moderate-to-severe difficulty sitting up from laying down position without support, asymmetric right greater than left iliopsoas weakness, and mild weakness of the bilateral gluteus maximus. Electrodiagnosis revealed myopathic motor unit potentials with increased recruitment in arm and leg muscles. Increased insertional activity and fibrillations were noted in the right medial gastrocnemius and thoracic and lumbar paraspinal muscles. MRI of the lumbosacral spine showed significant paraspinal muscle atrophy, thoracolumbar dextroscoliosis, mild difficulty standing up without arm support, moderate-to-severe difficulty sitting up from laying down position without support, asymmetric right greater than left iliopsoas weakness, and mild weakness of the bilateral gluteus maximus. Electrodiagnosis revealed myopathic motor unit potentials with increased recruitment in arm and leg muscles. Increased insertional activity and fibrillations were noted in the right medial gastrocnemius and thoracic and lumbar paraspinal muscles. MRI of the lumbosacral spine showed significant paraspinal muscle atrophy. Echocardiogram was normal except for left ventricular grade 1 diastolic dysfunction. Electrocardiogram, lung function test, antinuclear antibody, and erythrocyte sedimentation rate were normal or negative. A right deltoid biopsy in her early 30s showed “inflammation.” Facioscapulohumeral muscular dystrophy 1 genetic testing was negative. Gene panel testing for LGMD revealed 2 heterozygous ANO5 mutations (c.14-1 G>A in IVS1 and c.989dupT in exon 10).

SUMMARY/CONCLUSION: Severe axial myopathy represents an additional phenotype of Anoctamin 5 mutations. Anoctaminopathy should be considered in any patient with adult onset axial myopathy.

POSTERIOR REVERSIBLE ENCEPHALOPATHY LEADING TO THE DIAGNOSIS OF McARDLE DISEASE
Bing Liao, Edrich Rodrigues, Sean Pittock, Margherita Milone (Rochester, MN)

INTRODUCTION: McArdle disease (glycogen storage disease V) is an autosomal recessive disease caused by a deficiency of the muscle isoform glycogen phosphorylase. This enzyme catalyzes the first step in glycogen catabolism. McArdle disease manifests with exercise-induced myalgia, muscle stiffness, and cramps. Myoglobinuria occurs in a large proportion of patients and can cause acute renal failure. There is a phenotypic heterogeneity and the diagnosis can be delayed to adulthood.

OBJECTIVE: To describe an adult patient who presented with seizures in the setting of posterior reversible encephalopathy (PRES), which led to the diagnosis of McArdle disease.

CASE REPORT: A 36-year-old man with history of fatigue since childhood started experiencing exercise-related myalgia and muscle “spasms” in his late teens. At age 35, following intense physical activity, he developed severe muscle pain and stiffness and persistent urine discoloration. A few days later, he had 3 generalized tonic–clonic seizures which led to intubation. He was found to be in acute renal failure secondary to rhabdomyolysis. Serum creatinine kinase (CK) level was >200,000 U/L. MRI of the brain revealed bilateral subcortical T2 hyperintensity in the occipital, parietal, and posterior frontal lobes consistent with PRES. Histochemical studies of a quadriceps biopsy showed absence of myophosphorylase reactivity, indicative of McArdle disease. After hemodialysis for 2 weeks, his renal function improved. Two months later, the brain MRI abnormalities resolved; CK values remained elevated at 1400-3700 U/L.

SUMMARY/CONCLUSION: This case highlights the importance of early recognition of McArdle disease to try to prevent catastrophic outcomes.
MOTOR NEUROPATHY IN PATIENTS WITH INCLUSION BODY MYOSITIS WITH RESPONSE TO INTRAVENOUS IMMUNOGLOBULIN: TWO CASES AND A REVIEW OF THE LITERATURE

Erin Manning, Dale Lange (New York, NY)

CASE REPORTS: Patient 1 is an 85-year-old woman with 5 years of progressing muscle weakness and falls. Her weakness was asymmetric and most prominent in finger flexion. Her needle EMG showed polyphasic and short duration motor units consistent with a myopathic process. Patient 2 is a 54-year-old man with 19 years of slowly progressive muscle weakness. His weakness was asymmetric and diffuse but most prominent in finger flexion and quadriceps muscles. His needle EMG showed polyphasic and low amplitude motor units with complex repetitive discharges consistent with a myopathic process. Both patients had muscles biopsies with rimmed vacuoles and endomysial infiltrate consistent with inclusion body myositis (IBM) as well as motor nerve biopsies with loss of myelinated fibers and regenerative fibers consistent with motor neuropathy. Both patients had a response to IVIg with a mild increase in strength by examination and patient-reported improvement in strength and endurance with less falls. A literature review for other patients with both IBM and a motor neuropathy did not yield any results, although there are patients with IBM and a sensory neuropathy.

SUMMARY/CONCLUSION: This is the first report of patients with IBM and a motor neuropathy with a response to IVIg.

LYMPHEDEMA COMPLICATING THE TREATMENT OF MULTIFOCAL ACQUIRED DEMYELINATING SENSORY AND MOTOR NEUROPATHY: A CASE REPORT

Erin Manning (New York, NY)

CASE REPORT: A 63-year-old woman presented with a chief complaint of progressive weakness and numbness and pain in the left arm. She had a history of left breast cancer treated 14 years ago with lumpectomy and full axillary node dissection followed by radiation therapy and chemotherapy. She developed lymphedema in 2006 that was well controlled with compression sleeves. She developed progressive weakness, pain, and numbness 5 months prior to presentation. Her examination showed distal left arm weakness and atrophy, hyperesthesia in the ring and little fingers and half of the middle finger, and edema in left forearm and hand. Her needle EMG/NCSs of the arms showed evidence of demyelination in the median and ulnar nerves of the left arm. MRI of the left brachial plexus showed fascicular enlargement of the C8-T1 nerve roots and the medial cord. She was diagnosed with the multifocal acquired demyelinating sensory and motor neuropathy (MADSAM) variant of chronic inflammatory demyelinating polyneuropathy (CIDP). She started IVIg with stabilization of her weakness. The edema in the left arm and especially the hand worsened. Her occupational therapy had to put on hold due to the need to complete dedicated lymphedema therapy for 2 weeks with improvement in the edema in the hand. However, her left hand weakness worsened and later stabilized with an increase in her IVIg dosing.

SUMMARY/CONCLUSION: This is the first case report of a patient with MADSAM and lymphedema.
ROD MYOPATHY DUE TO COINCIDENCE OF TWO HETEROZYGOUS VARIANTS IN RYR1 GENE: A NEW PATHOLOGIC FEATURE AND DETECTION OF A NOVEL VARIANT

Jafar Kafaie (Saint Louis, MO), Miguel Guzman (Saint Louis, MO), Saeed Hamidi (Thornhill, Canada), Michael Marsh (Saint Louis, MO)

INTRODUCTION: Mutations in the skeletal muscle ryanodine receptor (RYR1) are a known cause of congenital myopathies and are associated with malignant hyperthermia and central core and minicore myopathy with external ophthalmoplegia.

OBJECTIVE: To report a novel pathologic variant in the RYR1 gene.

CASE REPORT: A 2-year-old girl presented with congenital axial and limb-girdle weakness and hypotonia. Examination showed gross motor delay with normal sensations and mild bilateral facial weakness with no ophthalmoplegia. She was areflexic. She had history of malignant hyperthermia in her paternal great uncle. MRI of the brain and spine were normal. She had normal creatine phosphokinase, aldolase, ammonia, and acetylcarnitine. Needle EMG showed fibrillations and positive sharp waves with normal NCSs. Muscle biopsy showed myopathic changes including increased endomysial connective tissue, few scattered atrophic muscle fibers, increased fiber size variability, and central nuclei. Myonecrosis, regenerating fibers, vacuoles, or inflammatory cells were not identified. Electron microscopy (EM) revealed rods. She was heterozygous for c.11590+1 G>T: IVS82+1, a new pathogenic variant in the RYR1 gene. She was also heterozygous for p.V4849I: c14545 1 G>A, a known pathologic variant in the RYR1 gene. Her father and mother are carriers of c.11590+1 G>T and p.V4849I variants, respectively, without any history of myopathy, weakness, or gross motor delay.

SUMMARY/CONCLUSION: Pathological variants in the RYR1 gene are typically associated with core myopathies which present in infancy or early childhood. Rod myopathy with ophthalmoplegia and severe weakness has been reported in RYR1 mutation, but to the best of the authors’ knowledge this is the first case of congenital myopathy with rod-like structures in EM imaging with no ophthalmoplegia and mild limb-girdle weakness.

STATIN-ASSOCIATED AUTOIMMUNE NECROTIZING MYOSITIS WITH ANTI-HMGCR AUTOANTIBODIES AND HELIOTROPE RASH RESPONSIVE TO PLASMA EXCHANGE

Prachi Parikh, Jinny Tavee, Yuebing Li (Cleveland, OH)

INTRODUCTION: Necrotizing autoimmune myositis (NAM) is a rare complication of statin use that is associated with anti-3-hydroxy-3-methylglutaryl-coenzyme A reductase (anti-HMGCR) antibodies and requires aggressive immune-modulating treatment.

OBJECTIVE: To report a case of statin-associated NAM mimicking dermatomyositis with creatinine kinase (CK) levels >25,000 U/l and a dramatic response to plasma exchange (PE).

CASE REPORT: A 47-year-old man with hyperlipidemia presented with a 4-week history of progressive speech and swallowing difficulties, rash, and generalized weakness. The patient had been on atorvastatin 20 mg for 1 year prior to onset. Neurologic examination demonstrated nasal speech, dysarthria, and diffuse weakness most prominent in the facial muscles, neck flexors, and distal extremities with bilateral wrist and foot drop. A heliotrope rash with additional skin lesions throughout the neck, trunk, and shoulders was also noted. HMGCR antibody levels in blood were greater than 200 units (normal <20) with a peak CK of 37,527 U/l. Myositis panel and anti-NXP2 antibodies were negative. Needle EMG was consistent with generalized necrotizing myopathy. Muscle biopsy revealed mild inflammatory changes without perifascicular atrophy. Despite 5 days of IV methylprednisolone and 2 gm/kg IVIg, HMGCR antibodies and CK levels remained elevated with continued clinical decline. Subsequent treatment with PE led to significant improvement in muscle strength, dysarthria, and dysphagia. The patient experienced further recovery with initiation of mycophenolate mofetil, oral corticosteroids, and continued PE.

SUMMARY/CONCLUSION: Elevated CK levels (>25,000 U/l), dermatomyositis-like skin rash, and a robust response to PE have not been previously reported in statin-associated NAM. In severe cases refractory to IVIg, PE may be helpful.
PREVALENCE OF POMPE DISEASE IN PATIENTS WITH MYOPATHY IN A TERTIARY CARE HOSPITAL IN NORTHEASTERN BRAZIL
Cleonisio Rodrigues, Matheus Barros, Pedro Braga Neto (Fortaleza, Brazil)

INTRODUCTION: Late-onset Pompe disease (LOPD) is a rare, but potentially treatable, inherited autosomal recessive metabolic myopathy caused by deficiency of acid alpha-glucosidase (GAA). Its prevalence in tertiary neurologic centers in Brazil is unknown.

OBJECTIVE: To evaluate the prevalence of LOPD diagnosis in adult patients with myopathies attended at the Ambulatory of Neuromuscular Diseases of the Hospital Geral de Fortaleza, Ceará, Brazil, from March 2012 to March 2015.

METHODS: A total of 52 patients with an initial clinical and electrophysiological diagnosis of myopathy were prospectively evaluated; 27 of which who had a myotonic dystrophy phenotype were excluded. Then, the remaining 25 patients underwent a dried blood spot test and subsequently a GAA gene mutation screening, if their GAA enzyme dosage was low.

RESULTS: The most common clinical presentations of the 25 studied patients overall were proximal and/or limb-girdle weakness. Two patients (8% of sample) were found to have low GAA enzyme dosage and pathological heterozygous mutations: intron 1 of the GAA gene (c.-32-13T>G) and a deletion encompassing exon 18. They were siblings and shared the same mutations and neuromuscular findings: limb-girdle and abdominal weakness and a typical pattern in electroneuromyography. The diagnosis of the others 23 patients after appropriate investigation were: limb-girdle muscular dystrophies (12; 48%), facioscapulohumeral dystrophy (4; 16%), inflammatory myopathies (3;12%), endocrine myopathies (2; 8%), mitochondrial myopathy (n=1; 4%), and oculopharyngeal dystrophy (n=1; 4%).

SUMMARY/CONCLUSION: LOPD diagnosis screening should be strongly considered in individuals with initial clinical and electrophysiological results suggestive of myopathy with no myotonic dystrophy phenotype. The most common alternative diagnosis of this subgroup is limb-girdle muscular dystrophy.

NEUROLEPTIC MALIGNANT SYNDROME IN STIFF PERSON SYNDROME
Amit Sachdev (Novi, MI), Sara Moussa (East Lansing, MI)

INTRODUCTION: Stiff person syndrome (SPS, also known as Moersch–Woltmann syndrome) is a rare disorder of diffuse muscle rigidity and spasticity increased startle response. An autoimmune etiology to SPS is recognized. Immune modulatory and symptomatic therapy is utilized. Baclofen, including intrathecal delivery, has been attempted. Six cases have been described since 1998 of neuroleptic malignant syndrome (NMS) associated with baclofen withdrawal.

OBJECTIVE: To report the first case of NMS associated with abrupt accidental cessation of intrathecal baclofen in a patient with SPS and to review special management considerations and challenges in recognizing the appropriate disease process.

CASE REPORT: A 64-year-old male with antibody associated SPS presented with sudden onset of tremor and muscle spasm. He had a previous intrathecal pump implanted. Two days prior he reports interrogation of the pump. He developed hyperthermia. A failed baclofen pump was identified, attributed to a defective battery. Oral replacement of baclofen and IV dantrolene were administered. Additional immune therapy was not added.

SUMMARY/CONCLUSION: The autoimmune component of SPS brought into question the possibility of disease flare up. However the patient's symptoms of hyperthermia and autonomic instability, not typical of SPS, made the diagnosis of NMS secondary to baclofen withdrawal more likely. It was important to include NMS in the differential for this patient as it was essential when considering treatment.
TWO CASES OF HYPOTHYROID MYOPATHY WITH IMPROVEMENT AFTER THYROID REPLACEMENT THERAPY
Radhika Sampat, Rocio Garcia-Santibanez, Jonathan Glass
(Atlanta, GA)

INTRODUCTION: Hypothyroid myopathy may present as a polymyositis-like syndrome. Clinical and laboratory improvement occurs with thyroid replacement therapy.

OBJECTIVE: To report 2 cases of hypothyroid myopathy and their respective workup, treatment, and outcomes.

CASE REPORTS: (1) A 27-year-old woman presented with a 6-month history of progressive weakness despite immunosuppressive therapy for presumed polymyositis. Her examination showed bilateral proximal upper and lower extremity weakness and impaired pulmonary function tests. Her creatine kinase (CK) was 6760 U/L, thyroid-stimulating hormone (TSH) 55 uIU/ml, thyroglobulin antibody 4.7 IU/ml, and thyroid peroxidase antibody >1000 IU/ml. Needle EMG demonstrated a proximal irritable myopathy. Left quadriceps muscle biopsy showed marked type-II atrophy, necrosis, and regenerating fibers without inflammation. After stopping the immunosuppressants and 6 months of thyroid replacement, her pulmonary function tests and strength normalized with the exception of mild proximal upper extremity weakness.
(2) A 61-year-old woman presented with 2 months of progressive cold intolerance, severe proximal upper and lower extremity weakness, and dysphagia resulting in respiratory failure and intubation. Her CK was 800 U/L, TSH 32 uIU/ml, and thyroglobulin antibodies 264 IU/ml. Needle EMG and biopsy were similar to above, but with primary inflammation. After 6 weeks of thyroid replacement (with normalization of TSH) and steroids, she was no longer ventilator dependent but strength had not improved. Long term follow up is still needed.

SUMMARY/CONCLUSION: Hypothyroid myopathy is an uncommon but disabling complication of untreated hypothyroidism. It may present as a polymyositis-like proximal irritable myopathy and improves with thyroid replacement therapy. Serological recovery usually precedes clinical recovery, which may take an average of 12 weeks.

Radhika Sampat, DO, Resident and Fellow Member Award Recipient

CHRONIC ENDURANCE EXERCISE TRAINING IN A MOUSE MODEL OF MYOTONIC DYSTROPHY TYPE 1
Lydia Sharp, Thomas Cooper (Houston, TX)

INTRODUCTION: Progressive muscle loss is a major feature of myotonic dystrophy, which negatively impacts a patient's quality of life. Pathogenesis stems from the RNA from the expanded allele (CUGexp RNA) disrupting the activities of splicing regulatory proteins. The molecular mechanisms of muscle wasting are unknown, and no therapies are available to combat muscle loss. Several small studies have evaluated the effects of exercise on muscle function in myotonic dystrophy. Results have been varied, and range from no effect to improvement in muscle strength. However, there is insufficient data available to make definitive recommendations to patients, and concern remains that some forms of exercise could lead to increased muscle damage and muscle loss.

OBJECTIVE: To evaluate the effects of chronic endurance exercise on skeletal muscle in a mouse model of myotonic dystrophy.

METHODS: Two groups of HSA-LR mice (a well-characterized model of myotonic dystrophy) and 2 groups of wild type mice were subjected to 2 months of treadmill running or were kept sedentary. The groups were compared using body composition, grip strength, exhaustive treadmill running, histological analysis of skeletal muscle, quantification of CUGexp RNA levels, quantification of splicing regulator proteins, and analysis of alternative splicing events altered in myotonic dystrophy.

RESULTS: Exercise in the HSA-LR mice reduced body fat percentage, increased bone density, increased endurance, and led to a significant reversal in several alternative splicing events.

SUMMARY/CONCLUSION: The results demonstrate that chronic endurance exercise is associated with improvements in muscle function and molecular abnormalities in a mouse model of myotonic dystrophy.

Radhika Sampat, DO, Resident and Fellow Member Award Recipient
FEMORAL NERVE AND LUMBAR PLEXUS INJURY AFTER LATERAL RETROPERITONEAL TRANSPSOAS APPROACH: ELECTRODIAGNOSTIC PROGNOSTIC INDICATORS
Naomi Abel, Andrew Vivas, Juan Uribe (Tampa, FL)

INTRODUCTION: Injury to the lumbosacral plexus and nerves is a well described complication after lateral retroperitoneal transpsoas approach to the spine. The prognosis for functional recovery after plexopathy or neuropathy is unknown.

OBJECTIVE: (1) To review cases of lateral retroperitoneal transpsoas lumbar interbody fusion (LLIF) at the University of South Florida and define the potential degrees of nerve injury which occur intraoperatively, and (2) to suggest intervals for EDX examination and prognostic indicators for recovery.

METHODS: Patients undergoing 1 level LLIF between January 2011 and August 2013 were identified. Postoperative MRI and EDX and physical examinations were performed to assess clinical and electrophysiologic recovery of function.

RESULTS: Of the 114 patients who underwent 1 level LLIF, 4 patients sustained severe femoral or femoral/obturator neuropathy, defined as loss of sensation and strength less than MRC grade 3/5 in corresponding peripheral nerve distributions. Three of these patients demonstrated spontaneous activity in the distribution of the femoral or femoral/obturator nerves at baseline, consistent with axonotmesis. All patients improved to at least MRC grade 4: 2 by 3 months, 1 by 9 months, and 1 by 12 months.

SUMMARY/CONCLUSION: Traction injuries to the lumbosacral plexus are an infrequent but potentially devastating complication following LLIF and result in variable degrees of nerve injury along the length of nerve. While reinnervation by collateral sprouting has limitations, novel axonal regrowth may continue for 12 months or more. With no clinical recovery, EDX studies performed at 6 weeks, 3 months, 6 months, and 8 months provide information to localize and delineate the extent of injury and provide prognostic indicators for recovery.

A CASE OF PHARYNGEAL–CERVICAL–BRACHIAL WEAKNESS MIMICKING MYASTHENIA GRAVIS CRISIS
Valeria Alvarez, Hardaman Cristian, Ariel Bustos, Debora Nadur, Micke Espinosa, Gonzalo Etchepareborda, Dario Rueda, Miguel Angel Chaves, Alejandra Pernazza, Maria Conti, Ricardo Reisin, Federico Micheli (Buenos Aires, Argentina)

INTRODUCTION: Pharyngeal–cervical–brachial weakness (PCB) is a rare variant of Guillain–Barré syndrome. Myasthenia gravis (MG) occasionally debuts as a crisis. Even though MG is more frequent, PCB is a differential diagnosis to keep in mind.

OBJECTIVE: To describe a challenging case of PCB.

CASE REPORT: A 27-year-old woman presented a week previous to consultation with rapidly progressive bilateral ptosis, diplopia, weakness in all 4-limbs, dysphonia, dysarthria, dyspnea, and dysphagia. At examination bilateral ophthalmoplegia, facial paresis, and predominant upper limb muscle weakness were evident. Needle EMG, rapid and slow repetitive nerve stimulation, F responses, and sensory and motor NCSs were normal. The patient rapidly worsened and invasive ventilation was required. Despite plasma exchange, IVIg, and corticosteroid therapy, she had to be ventilated for 50 days. Three months after onset she is still recovering. Cranial nerves, respiratory, and bulbar evaluation are normal, but proximal-muscle strength had a slower improvement. Corticosteroid therapy was gradually stopped. Autoantibodies against acetylcholine-receptor and muscle-specific kinase, antinuclear antibody, rheumatoid factor, serologies for b-viral-hepatitis, cytomegalovirus, Epstein–Barr virus, botulinum toxins, thallium serum level, cerebrospinal fluid analysis, and repeat NCS and single fiber studies were all normal. However, GD1a/aGM1/GM2 gangliosides antibodies were positive.

SUMMARY/CONCLUSION: PCB is characterized by acute progressive oropharyngeal, neck, and shoulder weakness, facial palsy with preserved tendon reflexes in lower limbs, and absence of sensory disturbances. Clinically, this case resembles Ropper’s description adding a lower limb paresis. As prognosis and treatment are quite different, PCB should be considered as a differential diagnosis of MG.

Valeria Alvarez, MD, AANEM Foundation for Research and Education Award Recipient
RACE DOES NOT AFFECT ULTRASOUND CROSS-SECTIONAL AREA OF UPPER EXTREMITY NERVES
Nathan Anderson, LeLand Finley, Bryce Betteridge, Weston Pratt, Eneko Larumbe, Jongyeol Kim (Lubbock, TX)

INTRODUCTION: Age, gender, and body mass index (BMI) are known to affect cross-sectional area (CSA) of peripheral nerve on ultrasonography. However, the effect of race on the CSA of nerves is not well defined.

OBJECTIVE: To investigate the effect of race on nerve size.

METHODS: Normal CSA of median and ulnar nerves of 120 healthy volunteers from 3 different ethnicity groups (Caucasian: 20 M, 20 F; African-American: 20 M, 20 F; Mexican-American: 20 M, 20 F), aged between 18 and 30 years, were compared. Measurements were made at 5 standard points along each nerve in the upper limb. Anthropometric data—age, height, weight, fat mass, muscle mass, total body water, bone mass, basal metabolic rate, metabolic age, visceral fat rating, body mass index (BMI), and degree of obesity (%)—were recorded.

RESULTS: There was no statistically significant difference in normal values of CSA of median and ulnar nerves at 5 spots between races after adjusting for other confounders. Females showed smaller CSA at each point (p<0.05), but the difference between genders was insignificant when controlled for fat free mass index (FFMI). Contrary to previous studies, BMI did not show any significant correlation with sex.

SUMMARY/CONCLUSION: For ultrasonography of major upper extremity nerves, race-specific reference values will not be needed but normative data of CSAs should be obtained according to anthropometric parameters including FFMI and gender. FFMI has the potential to be a powerful indicator for explaining differences in nerve CSA.

MILLER–FISHER SYNDROME: ELECTRODIAGNOSTIC FEATURES OF SIX CASES
Vitor Caldas, Julieanne Cordenonssi, Alberto Mello, Carlos Heise (Brazil, São Paulo)

INTRODUCTION: Miller–Fisher syndrome (MFS) is considered a variant of Guillain–Barré syndrome (GBS), characterized by the classic triad of ataxia, areflexia, and ophthalmoplegia.

OBJECTIVE: To evaluate the electrophysiological findings of patients with MFS.

METHODS: Six cases of MFS, seen within 3 weeks from onset, were retrospectively analyzed. Four patients presented with complete MFS, 2 patients presented with ophthalmoplegia without ataxia, and 2 showed bilateral facial palsy. In all patients, cerebrospinal fluid showed protein-cytological dissociation and MRI of the brain was unspecific. Antiganglioside antibodies were not available. All patients underwent motor NCSs and F wave studies of median, ulnar, tibial, and peroneal nerves; sensory NCSs of median, ulnar, radial, superficial peroneal, and sural nerves; and H reflex studies of tibial nerves. In selected cases, facial motor NCSs, repetitive stimulation tests, blink reflex, and needle EMG were performed.

RESULTS: EDX abnormalities were found in 67% of patients. Sensory NCSs showed non–length-dependent sensory axonal neuropathy with absent tibial H reflexes in 3 patients. One patient had only minor sensory abnormalities. Motor NCSs disclosed facial neuropathy in 2 patients and were otherwise normal in all limb nerves. Blink reflex was abnormal in 1 patient. F waves, needle EMG, and repetitive stimulation tests were normal in all patients.

SUMMARY/CONCLUSION: Most patients with MFS showed a sensory axonal neuropathy or a sensory neuronopathy. Facial neuropathy can be seen in some patients. The pattern of abnormalities is distinct from the usual features seen in GBS.

Vitor Caldas, MD, AANEM Foundation for Research and Education Award Recipient
TORSIONAL ANATOMY OF THE CARPAL TUNNEL: A REFERENCE FOR HEMISPASTIC INJECTION
Faye Chiou-Tan, Joslyn John (Houston, TX), Lawrence Robinson (Toronto, Canada)

INTRODUCTION: The hemispastic position of the upper limb (UL) after stroke can be difficult to inject for spasticity and CTS. This is the author's fourth publication documenting anatomic changes in the UL in the internally rotated, pronated, and flexed (hemispastic) position compared to anatomic neutral.

OBJECTIVE: To provide musculoskeletal ultrasound (MSKUS) anatomy of the carpal tunnel in the hemispastic compared to anatomic position.

METHODS: The MSKUS probe was centered over the carpal tunnel at the level of the proximal and distal flexor retinaculum in a healthy 51-year-old female. A pair of MSKUS images was recorded for each site: (1) supinated neutral and (2) in a hemispastic position with clenched fist. Video recordings were made to track the movement of the muscles and nerves during internal rotation.

RESULTS: The median nerve at the proximal flexor retinaculum measured 0.08 cm² in cross sectional area (CSA) in the anatomic position (normal). CSA increased to 0.11 cm² in the pronated clenched fist hemispastic position (borderline abnormal). NCSs were normal. The lumbricals were not visible with the hand supinated and open, but they appeared in the distal carpal tunnel region in the hemispastic fist clenched position.

SUMMARY/CONCLUSION: Precaution should be taken in diagnosing CTS in non-anatomic positions using CSA values on MSKUS. It is unclear if the increase in CSA was due to an inability to position the MSKUS probe perpendicular in the hemispastic position or if the nerve CSA increases with wrist flexion. The images show that the lumbricals may contribute to crowding of the distal carpal tunnel.

ACUTE INTERMITTENT PORPHYRIA: RETROSPECTIVE ANALYSIS OF NEUROPHYSIOLOGIC PATTERN IN SEVEN PATIENTS
Julianne Cordenonssi, Vitor Marques Caldas, Marcelo Megale, Carlos Heise (Sao Paulo, Brazil)

INTRODUCTION: Acute intermittent porphyria (AIP), a result of porphobilinogen deaminase deficiency, is the main porphyria associated with neuropathy.

OBJECTIVE: To evaluate the neurophysiologic pattern of cases with biochemical confirmation of AIP.

METHODS: Neurophysiological data from patients with confirmed biochemical diagnosis of AIP was retrospectively evaluated. All patients underwent motor NCSs of median, ulnar, peroneal, tibial, axillary, and femoral nerves; sensory NCSs of median, ulnar, and sural nerves; and needle EMG at rest and during contraction of various muscles. Four patients underwent autonomic studies, including sympathetic skin response and assessment of heart rate variability.

RESULTS: Motor NCSs showed axonal motor impairment in 6 patients: all of them showed reduced amplitudes of compound muscle action potential (CMAP) in axillary and femoral nerves and only 3 patients had reduced amplitudes of CMAP in distal nerves. The sensory NCSs showed reduced sensory nerve action potential amplitudes in only 2 patients. Regarding needle EMG, rest muscle abnormalities were observed in 6 patients, while only 2 showed signs of chronic reinnervation during voluntary contraction. The autonomic study was performed in 4 patients and was abnormal in 3.

SUMMARY/CONCLUSION: AIP is a predominantly acute proximal axonal motor polyneuropathy. Autonomic assessment can be very useful in cases of AIP. Julianne Cordenonssi, MD, AANEM Foundation for Research and Education Award Recipient
ANTI-SOX1 ANTIBODIES-ASSOCIATED NEUROPATHY MIMICKING GUILLAIN–BARRÉ SYNDROME IN A PATIENT WITH A TONGUE SQUAMOUS CELL CARCINOMA
Astrid Corlobe (Nimes, France)

INTRODUCTION: Paraneoplastic neuropathies are classically subacute sensory neuronopathies associated with anti-Hu antibodies. Anti-SOX1 antibodies have been reported in Lambert–Eaton myasthenic syndrome linked to small cell lung cancer. However, antibodies against SOX1 have also been found in a small number of patients with paraneoplastic axonal or mixed axonal-demyelinating neuropathy, and in a few patients with neuropathy of unknown origin. The associated tumor, when present, was exclusively lung cancer.

OBJECTIVE: To report a patient with tongue cancer who presented an acute demyelinating polyneuropathy associated with anti-SOX1 antibodies.

CASE REPORT: A 59-year-old man with a recent medical history of tongue squamous cell carcinoma presented with acute respiratory failure leading to tracheostomy. Five days later he complained of painful paresthesia, first in the left arm, then in the feet. He rapidly evolved toward tetraparesis. NCSs revealed a heterogeneous demyelinating sensorimotor polyneuropathy with multiple conduction blocks and temporal dispersion; needle EMG showed only signs of active denervation in left tibialis anterior muscle. Cerebrospinal fluid analysis was unremarkable: normal protein level, normal cell counts, and no oligoclonal bands. Anti-SOX1 antibodies were positive in blood, but other antineural antibodies and antiganglioside antibodies were negative. He underwent 2 plasma exchanges. Unfortunately, he presented a hypoxic cardiac arrest, was resuscitated, but which nevertheless lead to death a few days later.

SUMMARY/CONCLUSION: This is the first case reported of anti-SOX1 antibodies-related acute demyelinating neuropathy associated with tongue cancer, underscoring the wideness of the paraneoplastic neuropathy spectrum.

PEDIATRIC SACRAL PLEXOPATHY SECONDARY TO A VASCULAR LESION: A CASE REPORT
Lisa Williams (Santa Cruz, CA), Karina Del Rosario, Craig McDonald (Sacramento, CA), Divya Reddy (West Sacramento, CA)

INTRODUCTION: Sacral plexopathy is most commonly associated with trauma; however, other causes such as vascular malformations should be considered. Vascular lesions include both hemangiomas and vascular malformations. While vascular hemangiomas comprise tumors, vascular malformations result from altered vascular embryogenesis during fetal life. There are rare reported cases of vascular malformations resulting in sciatic neuropathy; however, there are no reported cases of vascular lesions causing sacral plexopathy in a pediatric population.

OBJECTIVE: To describe a rare pediatric case of sacral plexopathy secondary to vascular etiology.

CASE REPORT: A 21-month-old male presented with asymmetric gluteal musculature and difficulty progressing to walking since the age of 12 months. Physical examination findings are significant for right gluteus maximus and gluteus medius atrophy. He has weakness with hip extension and slight Trendelenburg gait. The skin overlying the right buttock has discoloration consistent with a vascular malformation. He has normal tone and reflexes and no obvious pain or discomfort or sensory deficits. He has ongoing difficulty with walking and frequent falls. EDX studies revealed a right sacral plexus injury proximal to the take off to the inferior gluteal nerve specifically affecting the peroneal division of the sciatic nerve and the inferior gluteal nerve at or distal to the greater sciatic foramen with evidence of reinnervation/denervation. He had an MRI with short tau inversion recovery (STIR) images without abnormalities.

SUMMARY/CONCLUSION: Vascular lesions should be considered in the workup of sacral plexopathy in the pediatric population. The current case describes a rare cause of sacral plexopathy secondary to a vascular lesion.

Lisa Williams, MD, Resident and Fellow Member Award Recipient
PATHOGENESIS OF ACUTE INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY: A CRUCIAL ROLE OF ALPHA-M INTEGRIN

Chaoling Dong, Rebecca Beacham, Steven Palladino, Eric Scott Helton, Eroboghene Ubogu (Birmingham, AL)

INTRODUCTION: The mechanisms of pathogenic leukocyte trafficking across the blood–nerve barrier (BNB) in acute inflammatory demyelinating polyradiculoneuropathy (AIDP) are incompletely elucidated. Prior work using untreated AIDP patient mononuclear leukocytes in a flow-dependent in vitro human BNB model demonstrated a crucial role for alpha-M integrin (CD11b) in pathogenic trafficking.

OBJECTIVE: To determine whether CD11b+ leukocytes are present in AIDP nerve biopsies and ascertain functional relevance in inflammatory demyelination using a representative animal model.

METHODS: Indirect immunohistochemistry was performed on sural nerve biopsies from 3 patients with AIDP and 3 age- and sex-matched normal control subjects. The relevance of CD11b in vivo was determined by evaluating cohorts of 8-12 week old female SJL mice with bovine peripheral nerve myelin-induced experimental autoimmune neuritis treated with a function-neutralizing monoclonal antibody compared to vehicle-treated and isotype antibody-treated mice. Neuromuscular severity score (NMSS) evaluation, motor nerve electrophysiology, and histopathological quantification of inflammatory cells and area of demyelination in sciatic nerves were performed. The Mann-Whitney U or student’s t tests were used to determine statistical significance.

RESULTS: CD11b antibody inhibition a week following disease onset reduced NMSS to near normal levels. This was associated with statistically significant increased dorsal caudal tail nerve (DCTN) amplitudes, as well as increased DCTN and sciatic nerve conduction velocities and reduced DCTN and sciatic nerve total waveform durations compared to control subjects. Statistically significant reductions in the numbers of F4/80+ monocytes/macrophages, CD3+ T-cells, and total CD11b+ mononuclear leukocytes, as well as percentage of demyelinated area per section, were also observed.

SUMMARY/CONCLUSION: This study supports a crucial pathogenic role of CD11b in AIDP.

A CASE REPORT OF EXTENSIVE LEPTOMENINGEAL SARCOIDOSIS

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INTRODUCTION: Sarcoidosis is a systemic granulomatous disease with an unknown etiology. The most common manifestations of sarcoidosis involve the lungs. An estimated 5% of patients will exhibit a variety of neurological symptoms often as the only clinical indication of the disease.

CASE REPORT: A 58-year-old woman presented to the emergency department with a 4-month history of increasing difficulty walking compounded by episodes of sudden lightheadedness, loss of balance, and orthostatic hypotension. Careful neurological examination noted unsteady gait with asymmetric lower extremity weakness and hyperreflexia. Contrast enhanced MRI of the neuraxis showed diffuse nodular leptomeningeal enhancement from the posterior fossa to the cauda equina. Lumbar puncture showed increased protein with elevated white blood cells, IgG synthesis rate, Q albumin ratio, and cerebrospinal fluid angiotensin-converting enzyme level. Biopsy of an inguinal lymph node revealed non-necrotizing granulomatous inflammation consistent with sarcoidosis. The patient improved on IV methylprednisolone therapy.

CONCLUSIONS: This is a case of neurosarcoidosis without signs or symptoms of systemic sarcoidosis with extensive leptomeningeal enhancement and lymph node biopsy consistent with sarcoidosis.
UTILITY OF ULTRASOUND IN THE MANAGEMENT OF ULNAR NEUROPATHY ACROSS THE ELBOW
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INTRODUCTION: Ulnar neuropathy at the elbow (UNE) is the second most common entrapment neuropathy of the upper extremity. Traditionally, UNE has been diagnosed by examination and EDX testing. Ultrasound may have advantages over EDX testing, as it allows for anatomic evaluation and provocative maneuvers and is better tolerated.

OBJECTIVE: To assess the utility of ultrasound in the management of UNE.

METHODS: Patients scheduled for release surgery were recruited. Patients with diabetes, ulnar nerve surgery, or subluxation were excluded. EDX testing was conducted. Patients were splinted for 6 weeks. Ultrasound was performed 4 weeks preoperatively and 6 weeks postoperatively. McGowan scores were recorded preoperatively and 3 months postoperatively.

RESULTS: Of the 11 patients screened, 2 were excluded, 4 did not complete their postoperative ultrasound, and 5 completed the study. Of those 5 cases, preoperative EDX testing was positive for UNE in 3, negative in 1, and inconclusive in 1. Average preoperative ulnar nerve CSA was 19.2 mm2. Average postoperative CSA was 15.2 mm2. All were improved (range: 2-6 mm2). In 4/5 patients, improvements of 4-6 mm2 at 6 weeks post-surgery corresponded with an increase of 1 grade in McGowan score at 3 months. In 1 patient, an improvement of only 2 mm2 at 6 weeks post-surgery corresponded with no change in McGowan score at 3 months, but improvement of 1 grade by 6 months.

SUMMARY/CONCLUSION: Ultrasound correlated with the clinical diagnosis of UNE more frequently than EDX testing, corresponded with clinical improvements in McGowan score, and may be a useful adjunct in the management of UNE, particularly when EDX testing is inconclusive.

A CASE OF CAUDA EQUINA SYNDROME AFTER INTRATHECAL CYTARABINE
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INTRODUCTION: Cauda equina syndrome (CES) is an uncommon but feared complication of intrathecal cytarabine.

OBJECTIVE: To report a case of CES after intrathecal cytarabine with slow improvement after discontinuation of the drug.

CASE REPORT: A 30-year-old woman with T-cell acute lymphocytic leukemia presented with bilateral lower extremity weakness. She initially had lower extremity radicular pain that developed 4 weeks after starting intrathecal cytarabine for malignant cells in her cerebrospinal fluid (CSF). She received cytarabine for 7 months followed by maintenance intrathecal methotrexate. After 9 months, she developed right leg weakness and numbness followed by left leg weakness, as well as urinary and fecal incontinence. Her MRC rating scale of iliopsoas and hip abductors/adductors on the right was 1/5 and on the left was 4/5. Quadriceps and hamstrings were 2/5 on the right and 4+/5 on the left. Distal lower extremity strength (gastrocnemius, tibialis anterior, and extensor hallucis longus) was 0/5 on the right and 1/5 on the left. Pinprick was decreased in L5-S2 distribution bilaterally with lower extremity areflexia. Electrophysiologic studies showed a polyradiculoneuropathy affecting L2-S2 on the left and L3-S2 on the right. Lumbosacral MRI showed cauda equina nerve root enhancement. Three consecutive CSF samples were negative for malignant cells. Therefore, the CES was attributed to the intrathecal cytarabine. Seven months after the last dose of cytarabine her motor strength moderately improved.

SUMMARY/CONCLUSION: CES secondary to intrathecal cytarabine was initially reported in 1% of patients. Recent reports have shown that up to 21-25% of patients develop CES, partial or complete, with incontinence, lower extremity paresis, and/or radicular pain.
LENGTH-DEPENDENT SENSORIMOTOR PERIPHERAL NEUROPATHY OFTEN RESULTS IN VENTRAL ABDOMINAL SENSORY LOSS
Charles Gervais, Mark Ross, Brent Goodman, A. Arturo Leis, Benn Smith (Scottsdale, AZ)

INTRODUCTION: Length-dependent sensorimotor peripheral neuropathy (LDSMPN) affects the body's longest nerve fibers, namely those innervating structures in the feet and hands. What is less appreciated, and essentially absent from the current literature, is that length-dependent involvement of sensory fibers in LDSMPN in thoracic segments gives rise to ventral abdominal sensory loss.

OBJECTIVE: To describe clinical, examination, and electrophysiological findings in cohorts of consecutive patients with LDSMPN with and without ventral abdominal sensory loss.

METHODS: Consecutive patients seen for LDSMPN (n=30) were evaluated for the presence or absence of ventral abdominal sensory loss. Demographic variables, symptoms, and quantitative neurologic findings using the Neuropathy Impairment Score (NIS) were examined using descriptive statistics. Final diagnostic categories were noted.

RESULTS: Ventral abdominal sensory loss (which was asymptomatic in all patients tested) was documented in 20/30 LDSMPN patients (66.7%); mean age 64.1 years (age range: 33-81 years), M:F gender ratio 19:11, mean NIS 21.4 (range: 0-77). Needle EMG/NCS abnormalities were found in 25/30 patients, with 5/30 having other objective evidence of peripheral neuropathy. LDSMPN patients without ventral abdominal sensory loss (n=10) had a mean age of 61.2 years (age range: 45-73 years), M:F gender ratio of 7:3 (range: 0-54).

SUMMARY/CONCLUSIONS: (1) Ventral abdominal sensory loss appears to be common in patients with LDSMPN of a variety of causes. (2) In addition to axons innervating distal limb territories, distal sensory fibers from the thoracic region represent another category of length-dependent involvement in LDSMPN. (3) The clinical examination of LDSMPN patients should include the ventral abdomen.

Charles Gervais, MD, Resident and Fellow Member Award Recipient

PARSONAGE–TURNER SYNDROME IN ACUTE IMMEDIATE POST-TRANSPLANT CASE
Hunaid Hasan, Rebecca Harrison, Sudhakar Tummala (Houston, TX)

INTRODUCTION: Parsonage–Turner syndrome is a rare acute immediate complication of autologous stem cell transplantation (ASCT) in multiple myeloma attributed to graft-versus-host disease, chemotherapy, and immunosuppression.

OBJECTIVE: To understand the acuity of onset of symptoms and its mechanism as a function of prognosis.

CASE REPORT: A 59-year-old right-handed male with kappa light chain multiple myeloma was admitted for ASCT with conditioning melphalan, revlimid, and natural killer (NK)-cell regimen. At baseline, he was athletically active. On admission day 2, immediately after chemotherapy induction, there was acute onset of right shoulder pain followed by worsening weakness. On day 9, neurological examination demonstrated (2-3/5) weakness of shoulder-girdle muscles and of right deltoid, supraspinatus, and infraspinatus pronation and supination as well as numbness of lateral right upper extremity. Cervical/right shoulder MRI demonstrated no nerve root compromise and denervation edema of the rotator cuff, respectively. Needle EMG demonstrated absent/half-amplitude median sensory responses at the right thumb and right index finger, respectively, with abnormal spontaneous activity of the right deltoid, infraspinatus, supraspinatus, and serratus anterior. There was moderate axonal right brachial plexitis of C5/6 and less of C7 motor/sensory roots. Lumbar puncture was unremarkable. Prednisone and IVIg were given with acupuncture, pain management, and physical therapy.

SUMMARY/CONCLUSION: Studies have reported onset of peripheral nerve disease from 5 days to 4 months after ASCT. Acute onset is associated with poorer functional recovery compared to chronic onset. Although the mechanism has been proposed to be secondary to immune dysfunction, variations exist that are not understood which predict functional outcome. Understanding these will facilitate management to improve functional outcome.
THE ROLE OF ELECTRODIAGNOSIS IN CONSERVATIVE VERSUS SURGICAL MANAGEMENT OF BRACHIAL PLEXUS INJURIES
David Impastato, Aaron Bunnell, Katherine Impastato, Jila Dabestani, Jason Ko (Seattle, WA)

INTRODUCTION: Brachial plexus injuries occur in less than 5% of trauma cases but severely limit function and quality of life. Currently, treatment options consist of conservative management, nerve transfer, or tendon transfer surgeries. EDX studies are often used to plan surgical management, but minimal evidence exists to predict which injuries will respond best to conservative management versus nerve transfer.

OBJECTIVE: To determine if recruitment or compound muscle action potentials (CMAPs) could help determine whether brachial plexus injuries would best be managed with conservative measures or nerve transfer surgery.

METHODS: This retrospective study included patients with brachial plexus injuries affecting muscles innervated by the axillary and suprascapular nerves who received care at Harborview Medical Center from January 2005 until December 2015. Patients were excluded if they had concomitant peripheral nerve injury, cerebrovascular accident, spinal cord injury, amputation, or vascular injury of the involved limb.

RESULTS: Statistical significance was not achieved for any recruitment or CMAP value in predicting which surgical or nonsurgical patients would achieve greater than 3/5 strength at least 6 months after injury. It does appear that patients with no, discrete, or markedly reduced recruitment tend to have worse outcomes where patients with reduced recruitment tend to improve with surgery.

SUMMARY/CONCLUSION: Multicenter studies should be performed to attempt to obtain statistical significance as it is still unclear which patients would perform best with nerve transfers. Patients with reduced recruitment have the best chance of improving to greater than 3/5 strength with surgery, although it is possible for patients with worse recruitment to achieve functional strength with surgical intervention.

PERIPHERAL NERVE ULTRASOUND FINDINGS IN A CHILD WITH SEVERE DEMYELINATING HEREDITARY NEUROPATHY
Jennifer Jaskiewicz, Tracy Levy, Mark Landau, David Dennison, Jonathan Smith (Bethesda, MD)

INTRODUCTION: Diagnosis of hereditary neuropathy in young children is complicated by the need for sedation during prolonged, invasive, or painful procedures such as electrodiagnosis, lumbar puncture, or MRI. Ultrasound (US) provides a relatively quick and well-tolerated means of assessing nerves in such patients.

OBJECTIVE: To discuss the role of US in the diagnosis of severe hereditary neuropathy in pediatric populations.

CASE REPORT: Presented here is a case of suspected Dejerine–Sottas syndrome and the diagnostic workup thereof. A 28-month-old girl with no family history of neuropathy presented with lifelong gross motor delay. Examination revealed occipital plagiocephaly, café au lait spots, diffuse hypotonia, lower-extremity predominant weakness, absent reflexes, and a wide-based, waddling gait. Cerebrospinal fluid showed mildly elevated protein of 54 mg/dL. NCSs disclosed severe diffuse, symmetric slowing with velocities of 9.9-15.9 m/s. MRI demonstrated enhancement of spinal and cranial nerve roots. US demonstrated diffusely increased cross-sectional area compared to published values for age (z scores: 1.7-12.6); lower extremity nerves were larger. Whole exome sequencing is pending.

SUMMARY/CONCLUSION: Motor delay, hypotonia, weakness, and areflexia raised concern for severe hereditary neuropathy. Electrodiagnosis and US suggested demyelination. Dejerine–Sottas was suspected. Data on imaging in pediatric patients with this condition is extremely limited, and to the authors’ knowledge there are no reports of US findings. US has several advantages in this setting, allowing quick, painless assessment of multiple nerves. Further study of US in pediatric hereditary neuropathies might allow targeted genetic testing even in children with limited or no ability to tolerate NCSs.

Jennifer Jaskiewicz, DO, Resident and Fellow Member Award Recipient
CLINICAL AND LABORATORY FEATURES OF SMALL FIBER NEUROPATHIES WITH IMMUNOGLOBULIN M VERSUS TRISULFATED HEPARIN DISACCHARIDE
Jafar Kafaie, Minsoo Kim (Saint Louis, MO)

INTRODUCTION: Serum IgM binding to IdoA2S-GlcNS-6S, a trisulfated heparin disaccharide (TS-HDS), is associated with small fiber neuropathies (SFNs).

OBJECTIVE: To describe characteristics of SFNs associated with IgM versus TS-HDS.

CASE REPORTS: (1) A 16-year-old male presented with paresthesia in the lower extremity and progressed to generalized pain. His IgM versus TS-HDS titers were 16,000. Intraepidermal nerve fiber density (IENFD) on left thigh was 6.44/mm and calf was 6.91/mm. Sweat gland nerve fiber density (SGNFD) was normal. (2) A 16-year-old male presented with bilateral lower extremity pain which progressed to generalized intermittent pain, dry skin, dry eye/mouth, and urinary urgency. IgM versus TS-HDS titers were 23,000. He had an elevated creatine kinase (CK) at 967 IU/L and aldolase at 19.3 U/L. IENFD on right thigh was 5.79/mm and calf was 4.62/mm. SGNFD was normal. (3) A 15-year-old female presented with sharp abdominal pain 1 month after contracting infectious mononucleosis. This progressed to numbness/tingling on bilateral distal extremities and postural orthostatic tachycardia syndrome. IgM versus TS-HDS titers were 18,000. IEFND on left thigh was 12.74/mm and calf was 8.87/mm. SGNFD was normal. (4) A 16-year-old female presented with a jabbing spasm and stiffness in her legs, dry eyes, and orthostatic dizziness. IgM versus TS-HDS titers were 26,000. IEFND on left thigh was 13.31/mm and calf was 8.76/mm. SGNFD was normal.

SUMMARY/CONCLUSION: All patients presented with acute onset of progressive painful neuropathy confirmed as SFN with abnormally low IENFD. Noticeable characteristics include non–length-dependency (C1), high CK (C2), onset in 1 extremity (C1 and C4), 50% male (C1 and C2), and 1 preceding event (C3). Despite autonomic dysfunction in the majority (C2, C3, and C4), SGNFD was normal in all. IgM versus TS-HDS may enhance diagnostic yield for acute onset of SFN.

A PERIPHERAL NERVE SHEATH TUMOR PRESENTING AS CALF AND FOOT SENSORY SYMPTOMS
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INTRODUCTION: Peripheral nerve sheath tumors are rare in patients without a history of neurofibromatosis.

CASE REPORT: A 48-year-old right-handed woman with prior hypertension, asthma, pre-diabetes, and Vitamin D deficiency presented with several months’ paroxysmal right medial calf pain radiating to the plantar surface of the foot and 1 months’ paresthesia. On examination, sensation to pinprick and temperature was reduced over the right plantar foot and sensation to light touch was reduced over the dorsum and plantar surface of the right foot. Proprioception and pallesthesia were intact without allodynia, muscle weakness, or atrophy. A vague, deep mass was palpable over the right medial calf just distal to the knee with Tinel sign. Bilateral peroneal and tibial motor and bilateral sural and superficial peroneal sensory NCSs were normal, but the right tibial H reflex was absent. Needle EMG was normal. MRI revealed a globular, heterogeneous mass with peripheral enhancement in the posterior right calf in the region of the tibial neurovascular bundle. Ultrasound-guided core needle biopsy followed by complete excision of the mass revealed a Schwannoma attached to the tibial nerve.

SUMMARY: Schwannomas may present as palpable lesions in patients without a history of neurofibromatosis as an isolated mononeuropathy. Inspection and palpation of the affected limb may help determine diagnostic imaging.

Nidhi Kapoor, MBBS, Resident and Fellow Member Award Recipient
RETROSPECTIVE REVIEW OF CLINICAL PRESENTATION IN PATIENTS WITH MULTIFOCAL MOTOR NEUROPATHY
Sanaz Karimi, Said Beydoun, David Girard (Los Angeles, CA)

INTRODUCTION: Multifocal motor neuropathy (MMN) is a demyelinating neuropathy with good response to immunotherapy.

OBJECTIVE: A retrospective chart review of patients with MMN was performed to look for the clinical presentation, severity of symptoms, progression, and response to treatment.

METHODS: A retrospective chart review of 18 patients with a diagnosis of MMN who were seen at the Neuromuscular Division of Keck Medical Center/University of Southern California during last 8 years (2007-2015) was performed. The European Federation of Neurological Societies 2010 guidelines criteria for diagnosis of MMN was used.

RESULTS: Among the 18 MMN patients (11 male, 7 female; M/F=1.5), the median age of symptom onset in females and males was 35 years (female age range: 28-46 years, male age range: 33-66 years). Median duration between symptom onset to diagnosis was 5.5 years (range: 6 months-17 years). Six (33%) were GM1 antibody positive. Sixteen presented with asymmetric upper extremity weakness. The most common weakness was grip weakness followed by wrist drop and foot drop. Fifteen (83%) had conduction block. Eleven (61%) had definite MMN and 3 (16%) had probable MMN. Three (16%) were positive for monoclonal gammopathy. Seventeen (94%) had good response to IVIg treatment based on the Overall Disability Sum Score (ODSS). The quantitative treatment response and disease severity and progression will be assessed (ODSS, dynamometer).

SUMMARY/CONCLUSION: Early diagnosis and treatment in patients with MMN is consistent with good treatment outcome.

BRACHIAL PLEXOPATHY AS A PRESENTATION OF SJÖGREN'S SYNDROME
Gupreet Khakh, Raghav Govindarajan (Columbia, MO)

INTRODUCTION: Sjögren's syndrome can have a wide variety of neurological (both CNS and PNS) manifestations. Reported here is an uncommon case of brachial plexopathy/plexitis as a presentation of Sjögren's syndrome.

CASE REPORT: A 22-year-old male woke up with severe shoulder pain which later became a boring severe arm pain. He was diagnosed with shoulder sprain and put in a sling. The pain persisted and he was prescribed multiple medications including opioids. About 2 weeks later he developed hand and arm weakness. He was then referred to the authors. On examination he had patchy involvement of weakness affecting multiple nerves but no obvious scapular winging. There was no clear sensory gradient. Needle EMG performed a month from onset showed ongoing denervation in multiple muscles with patchy distribution affecting multiple nerves including median, ulnar, radial, and axillary, but curiously long thoracic and suprascapular nerves were spared. MRI of the brachial plexus showed patchy hyperintense T2 signal. Given the unusual presentation, further workup was performed which showed antinuclear antibodies at 1:1200 and a positive SSA antibody. Lip biopsy showed chronic inflammation of the minor salivary glands confirming the diagnosis of Sjögren's syndrome. He is currently in 6 weekly IVIg for his weakness, in addition to Plaquenil®.

SUMMARY/CONCLUSION: Sjögren's syndrome is a differential for a variety of neurological diseases. It should also be kept as a differential for idiopathic brachial plexitis.

Gupreet Khakh, MD, Resident and Fellow Member Award Recipient
NEUROLOGIC INVOLVEMENT IN V122I FAMILIAL AMYLOIDOSIS

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INTRODUCTION: Late-onset cardiomyopathy with heart failure is the phenotype of familial amyloidosis, occurring in African Americans in the United States at a rate 4 times that of whites after the age of 60; 3.9% of African Americans are heterozygous for the amyloidogenic allele, where isoleucine substitutes for valine at position 122 (Val 122-ile) of the serum carrier protein transthyretin (TTR). Reported here are the neurologic changes seen in addition to the cardiomyopathy.

OBJECTIVE: To describe a new phenotype in V122i familial amyloid cardiomyopathy and neuropathy.

CASE SERIES: Seven patients with the ATTR V122i gene mutation had amyloid staining on myocardial biopsies and 1 patient had amyloid staining on a flexor retinaculum biopsy. All had symptoms of peripheral neuropathy. Causes of a peripheral neuropathy other than amyloid were excluded. All 7 patients had axonal predominant peripheral neuropathy, confirmed by needle EMG. One patient had a superimposed painful lumbar radiculopathy, without corresponding structural MRI abnormalities. One patient had multiple entrapment neuropathies and a small fiber neuropathy (reduced intraepidermal nerve fiber density) and dysautonomia with reduced sweat gland nerve fiber density.

SUMMARY/CONCLUSION: Of these 8 patients with the ATTR V122i mutation, 7 have amyloid cardiomyopathy and a peripheral neuropathy possibly due to amyloid deposition in the nerve. One patient had carpal tunnel and a peripheral neuropathy without a cardiomyopathy. One had symptoms of radiculopathy, confirmed by needle EMG, possibly due to dural amyloid infiltration. Prospective studies of this phenotype are warranted to confirm these findings and to determine whether the peripheral neuropathy occurs earlier than the cardiomyopathy. This disease may be underestimated.

ELECTROPHYSIOLOGICAL SUBTYPES AND PROGNOSIS OF GUILLAIN–BARRÉ SYNDROME IN SOUTH KOREA

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INTRODUCTION: Guillain–Barré syndrome (GBS) is a potentially life-threatening disease characterized by acute onset, progressive flaccid paralysis with sensory or autonomic symptoms. GBS is usually associated with a good prognosis, but some patients show severe motor weakness or respiratory distress which can lead to a poor functional outcome.

OBJECTIVE: To describe the electrophysiological subtypes and to search for prognostic factors of GBS in South Korea.

METHODS: Clinical and laboratory data were collected from 85 GBS cases between 2004-2015 and those factors which can predict functional outcome were analyzed. Electrophysiological subtypes were classified as normal, demyelinating, axonal, equivocal, and inexcitable. The severity of motor weakness was evaluated by the sum of MRC grades of all 4 limbs, and Hughes function grade score (HFGS) was used to assess the prognosis at 12 months.

RESULTS: The most common electrophysiological subtype was demyelinating (n=50), followed by normal (n=14), equivocal (n=14), and axonal (n=6). The axonal group showed more severe motor weakness at nadir than the demyelinating group (MRC sum score: 33.7 versus 45.6, p=0.008) and higher HFGS at discharge (3.2 versus 2.3, p=0.002). When the good outcome (HFGS: 0-1) group was compared at 12 months with the poor outcome (HFGS: 2-6) group at same period, the former group showed younger age, less severe motor weakness at nadir, and higher distal compound muscle action potential amplitude of ulnar and peroneal motor NCSs than the latter group (for all parameters, p<0.05).

SUMMARY/CONCLUSION: Older age, severe motor weakness at nadir, and axonal type in the NCS were demonstrated as poor prognostic factors of GBS in South Korea.
HUMAN T-LYMPHOTROPHIC VIRUS TYPE 1 PRESENTING AS A MOTOR NEUROPATHY
Vanessa Tiongson (Elmhurst, NY), Nina Kim, Perrin Pleninger (New York, NY)

INTRODUCTION: Human T-lymphotropic virus type 1 (HTLV-1) is a retrovirus known to neurologically manifest as a myelopathy called tropical spastic paraparesis. It is important to recognize that HTLV-1 can present with other neurological symptoms including motor neuropathy.

OBJECTIVE: To report 2 cases of HTLV-1 presenting as a predominant motor neuropathy.

CASE REPORTS: (1) A 49-year-old woman presented with 7 years of progressive weakness and muscle fasciculations in her extremities. Her examination revealed atrophy of bilateral forearms, hands, thighs, and calves; fasciculations in both arms; distal weakness in all extremities; and hyperreflexia. NCSs and needle EMG showed evidence of axonal motor neuropathy. MRI of the cervical and thoracic spine was unremarkable. MRI of the brain showed nonspecific T2 abnormalities. Laboratory workup was unremarkable except for a reactive HTLV-1. (2) A 53-year-old female requested a second opinion for 3 years of bilateral hand and foot weakness. Examination was notable for bilateral hand contractures and atrophy and distal greater than proximal arm weakness. MRI of the entire neural axis was unremarkable. NCSs and needle EMG showed evidence of an axonal motor neuropathy. The patient was initially told she had ALS. Laboratory workup revealed positive HTLV-1.

SUMMARY/CONCLUSION: Motor neuropathy is included within a spectrum of neurological presentations associated with HTLV-1 infection and can be mistaken for ALS. It is important to consider HTLV-1 infection when evaluating patients with motor neuron disorders as this diagnosis may alter prognosis and management. Additional research of this population may lead to advances in treatment of HTLV-1 disorders and other motor neuropathies.

Vanessa Tiongson, MD, Resident and Fellow Member Award Recipient
Nina Kim, MD, Resident and Fellow Member Award Recipient

PLEXOPATHY DUE TO MICROWAVE TREATMENT OF AXILLARY HYPERHIDROSIS
Stephen Knox, Roderick Ballesteros (Sacramento, CA)

INTRODUCTION: Brachial plexopathy is uncommonly seen. Direct trauma, localized malignancy, inflammation, and physical compression are common etiologies. Remote plexus injury can occur as a consequence of radiation and, rarely, electrical injury. Plexus injury is confirmed by the combination of the clinical and electrophysiological presentation, with low amplitude (postganglionic). Hyperhidrosis can be a debilitating condition hindering patient social, personal, and professional interactions. Treatment options have been limited to topical applications, botulinum toxin injections, and invasive surgical interventions. Recently approved is an apparatus that delivers localized microwave energy to thermally necrose the axillary sweat glands. Complications are rare and are limited to transient patches of altered sensation in the treatment limbs.

OBJECTIVE: To describe a case of a plexopathy following microwave thermolysis of axillary sweat glands to treat hyperhidrosis.

CASE REPORT: The electrophysiological presentation and longterm outcomes of a patient who developed a plexopathy as a consequence of the procedure described are reviewed. Plexopathy with predominant involvement of the median nerve was identified and confirmed electrophysiologically. Symptoms were present bilaterally following the thermolysis, with persistent unilateral sensory loss of the middle finger 2 years later.

SUMMARY/CONCLUSION: Hyperhidrosis can be a debilitating social condition. Newer modalities of treatment are available with trailing complications. This case extends the etiological causes of plexopathy and recognizes its overall good prognosis.
PERIPHERAL NERVE INJURIES: A RETROSPECTIVE SURVEY OF 1124 CASES
Joao Kouyoumdjian, Carla Graca, Vanessa Ferreira (Brazil, Sao Paulo)

INTRODUCTION: Peripheral nerve injuries (PNIs) constitute a serious risk of severe motor disabilities.

OBJECTIVE: To retrospectively analyze 26 years of PNIs based on EDX consultations.

METHODS: Between 1989 and 2014, 1124 consecutive patients experienced 1418 PNIs and were referred for an EDX examination. Symptomatic PNIs were located in the lower and upper limbs and facial nerves. Cases with iatrogenic lesions and spinal/root lesions were excluded. Brachial plexus (BP) injuries with root avulsion were included in the BP group. Injury categories included vehicular accidents, penetrating traumas, falls, gunshot wounds, car accidents involving pedestrians, sports, and miscellaneous.

RESULTS: Patients’ mean age was 34.2 years and most were male (76.7%). PNIs were isolated in 80.9%; combined lesions most commonly involved the ulnar and median nerves. Upper limb PNIs occurred in 72.6%. The ulnar nerve was injured most often, either singly or in combination. Vehicular accidents were the most common cause of injury (46.4%), affecting the BP or radial, fibular, and sciatic nerves. BP lesions occurred in 46.1% of motorcycle accidents. Penetrating trauma (23.9%) commonly affected the ulnar and median nerves. Falls and gunshot wounds frequently affected the ulnar, radial, and median nerves. Sports injuries, mostly due to soccer, affected mainly the fibular nerve. BP injuries were much more common in motorcycle accidents than in car accidents (46.1% versus 17.1%) and root avulsions were more frequent.

CONCLUSIONS: Most PNIs were caused by vehicular accidents and penetrating trauma, and occurred in young men. Overall, ulnar nerve, BP, and median nerve PNIs were the most prevalent.

Joao Kouyoumdjian, MD, PhD, IFCN North American Chapter Fellowship Award Recipient

VOLTAGE-GATED POTASSIUM CHANNEL AUTOANTIBODY IN A PATIENT WITH SMALL FIBER NEUROPATHY AND PARKINSONISM
Ikjae Lee, Hani Kushlaf (Cincinnati, OH)

INTRODUCTION: Voltage-gated potassium channel (VGKC) autoantibody has been associated with acquired neuromyotonia, Morvan syndrome, epilepsy, limbic encephalitis, and dysautonomia. There are no prior reports of coexisting small fiber neuropathy and Parkinsonism in patients with VGKC autoantibody.

OBJECTIVE: To report the clinical presentation, results of diagnostic testing, and course of a patient with small fiber neuropathy and positive VGKC antibody who subsequently developed Parkinsonism.

CASE REPORT: A 63-year-old man developed constant paresthesia and burning pain in both feet followed by finger tingling in 3 months. Neurological examination revealed decreased pinprick sensation in the toes. Needle EMG of the left arm and leg showed mild left CTS without large fiber peripheral neuropathy or lumbosacral radiculopathy. Sweat gland and epidermal nerve fiber densities were decreased in skin biopsy from the left calf but not from the left thigh. Rheumatoid factor, antinuclear antibody, double stranded DNA, SSA, SSB, endomysial IgA, gliadin antibodies, rapid plasma reagin, fasting blood glucose, HbA1c, hepatitis C antibody, vitamin B12, erythrocyte sedimentation rate, thyroid-stimulating hormone, T4, serum protein electrophoresis, Lyme Ab IgG and IgM, and HIV testing were all normal or negative. VGKC autoantibody titer was 4.11 (normal <0.02 nmol/L). He was treated with IVlg without improvement. He developed resting tremor in the left arm 2 years after the onset of neuropathic symptoms. Detailed examination confirmed Parkinsonism with asymmetric resting tremor, rigidity, and bradykinesia. His Parkinsonism symptoms improved significantly with levodopa treatment.

CONCLUSION: Coexisting small fiber neuropathy and Parkinsonism with VGKC autoantibodies has not been reported. The relatively short period of symptom development suggests an association with VGKC autoimmunity. Further research is required to prove this association.

Ikjae Lee, MD, Resident and Fellow Member Award Recipient
INTRANEURAL PERINEURIOMA: THE VALUE OF MAGNETIC RESONANCE NEUROGRAPHY

Luciana León Cejas, Lita Vargas, Daniela Binaghi, Cintia Marchesoni, Ana Pardal, Mariano Socolovsky, Alberto Dubrovsky, Soledad Monges, Analia Taratuto, Ricardo Reisin (Buenos Aires, Argentina)

INTRODUCTION: Intraneural perineurioma (IP) is a benign, yet under-recognized, hypertrophic peripheral nerve tumor. It occurs in the young and is characterized by a mild and progressive motor and sensitive compromise. Diagnosis requires a high clinical suspicion combined with electrophysiology and magnetic resonance (MR) neurography.

METHODS: To review the authors’ experience in patients with IP and analyze retrospectively their clinical aspects, electrophysiological studies, neurography images, and biopsy results.

RESULTS: Six patients (5 male, mean age: 19 years) were identified between 2011-2015. Five presented with unilateral steppage gait and only 1 with unilateral gastrocnemius wasting. All patients had mild sensory loss, and none of them had pain. Physical and neurophysiological examinations showed involvement of the sciatic nerve extending into the peroneal (5 patients) and posterior tibial nerves (1 patient). MR neurography identified the characteristic pattern of perineurioma: nerve enlargement, extensive affection of the nerve, and iso-intensity of the tumor in T1, hypo-intensity in T2, with hyper-intensity in T2 with fat suppression sequences. In all patients a nerve biopsy confirmed the diagnosis of IP. After a mean followup of 3 years, 4 patients remained stable and required a tendon transfer to minimize a neurological deficit (steppage gait) in 2 cases.

CONCLUSIONS: IP is an underdiagnosed focal neuropathy. MR neurography allows the identification of this tumor, and therefore it is a key tool in the diagnosis of patients with slowly progressive focal neuropathies. MR neurography should be used to diagnose this entity when suspected.

Luciana León Cejas, MD, Recipient of the 2016 IFCN North American Chapter Fellowship Award

PERIPHERAL NERVE MAGNETIC RESONANCE IMAGING AND SERIAL NERVE CONDUCTION STUDY DATA IN A PATIENT WITH THE MADSAM VARIANT OF CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY

Dora Leung (New York, NY)

INTRODUCTION: The multifocal acquired demyelinating sensory and motor neuropathy (MADSAM) variant of chronic inflammatory demyelinating polyneuropathy (CIDP) presents with symptoms suggestive of mononeuropathy multiplex, with sensory and motor involvement.

OBJECTIVE: To present abnormal peripheral nerve MRI findings, as well as serial EDX data, after treatment in a patient with MADSAM.

CASE REPORT: A 36-year-old man presented with a 1-year history of numbness and paresthesia in the right leg, without leg or back pain. Muscle cramps and weakness developed in the right leg with foot drop gait 6 months prior to presentation. During neurologic evaluation, the patient had distal right leg weakness in both a fibular and tibial nerve distribution, with decreased sensation. His strength and sensation were normal in the left leg and both arms, with diffuse hyporeflexia. Shortly after presentation, he developed new sensory symptoms in the left leg and right arm, but no motor deficits. He was diagnosed with the MADSAM variant of CIDP, and his symptoms improved with monthly IVIg treatment. MRI of the lumbosacral and brachial plexuses showed diffuse, markedly enlarged and thickened nerves. Blood tests for an autoimmune/inflammatory process were all normal or negative. Cerebrospinal fluid analysis was normal. EDX studies showed demyelinating features with conduction block, even in clinically-affected limbs, with evidence of denervation. With monthly IVIg treatments, serial NCSs over the next 18 months showed improvement with a decrease in demyelinating features and resolution of conduction block, corresponding with clinical improvement.

SUMMARY/CONCLUSION: This patient with the MADSAM variant of CIDP showed classic features of thickened nerves on imaging studies, and demyelinating features with conduction block on EDX studies, which improved with treatment.
NOVEL MPZ MUTATION IN THREE GENERATIONS WITH DOMINANTLY-INHERITED, DISTAL DEMYELINATING POLYNEUROPATHY

David Lorance, Kelly Mandigo, Michael Hehir (Burlington, VT)

INTRODUCTION: Charcot–Marie–Tooth (CMT) disease is classified by clinical severity, demyelinating versus axonal electrophysiology, inheritance pattern, and genetic mutation. High-throughput, next-generation sequencing (NGS) expanded the number of mutations known to cause CMT and increased identification of sequence variants of undetermined significance. Distinguishing among causative and incidental novel sequence variants is imperative to guide clinical decisions in patients with possible inherited neuropathies. Clinical phenotype description in families with variants of undetermined significance will lead to understanding the clinical significance of these mutations.

OBJECTIVE: To define the clinical phenotype in 3 consecutive generations with demyelinating CMT that possess a novel base-pair substitution.

METHODS: The proband, age 7, underwent NGS through MNG Laboratories (Atlanta, Georgia). A novel sequence variant was identified and compared to the exome aggregation consortium (ExAC) data set to verify absence in control subjects. Four family members from 3 generations, who possess the clinical phenotype of dominantly-inherited demyelinating CMT, were interviewed, examined, and studied with EDX testing. Single gene analysis was performed on a remaining 3 patients.

RESULTS: NGS identified a novel base-pair substitution of MPZ (c.314C>T), resulting in a missense variant (p.Pro105Leu). All 3 generations have symmetric, distal weakness of the feet and hands, calf cramping, high foot arches, curled toes, diminished deep tendon reflexes, and distal sensory loss to pinprick, temperature, and vibration. EDX testing was consistent with distal demyelination (prolonged distal motor latencies) with secondary axon loss.

SUMMARY/CONCLUSION: The novel MPZ base-pair substitution in this family is associated with dominantly-inherited, distal demyelinating, inherited neuropathy with extremity cramping. This mutation should be reclassified as pathogenic.

David Lorance, MD, Resident and Fellow Member Award Recipient

IATROGENIC SUPERFICIAL PERONEAL SENSORY MONONEUROPATHY DUE TO ELECTIVE ANTERIOR COMPARTMENT FASCIOTOMY RELEASE IN A 20-YEAR-OLD RUNNER

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INTRODUCTION: Chronic exertional compartment syndrome (CECS) typically presents as anterolateral leg pain with exercise, and it is most common among young adult runners. An elective fasciotomy release is the treatment of choice. The anterior compartment is most commonly involved. Iatrogenic superficial peroneal nerve injury is a rare surgical complication, but it can occur when the fasciotomy is performed too distally.

OBJECTIVE: To describe a case of a superficial peroneal sensory mononeuropathy after elective fasciotomy release.

CASE REPORT: A 20-year-old female runner with a history of bilateral CECS presented with pain and numbness over the dorsum of the left foot after an elective left anterior compartment fasciotomy release. On examination, she had sensory loss with monofilament testing in the left superficial peroneal nerve distribution. Her motor examination was intact. NCSs revealed normal tibial and peroneal motor responses and a normal sural sensory response. The left superficial peroneal sensory response was unobtainable, while the right side was normal. Needle EMG revealed no abnormalities. Specifically, the left peroneus longus did not show any abnormal spontaneous activity or motor unit morphology changes. She was diagnosed with an isolated axonal superficial peroneal sensory mononeuropathy. The prognosis for recovery is guarded with an unobtainable sensory response 14 months postoperatively.

SUMMARY/CONCLUSION: A study of 118 patients who underwent elective percutaneous anterior compartment fasciotomy release reported a 2% rate of iatrogenic superficial peroneal nerve injury. CECS is a common diagnosis seen in sports medicine clinics. Patients should be made aware of the potential for iatrogenic nerve injury from elective fasciotomy release when discussing treatment options.
FASCICULATION POTENTIALS AND DECREMENTAL RESPONSES IN AMYOTROPHIC LATERAL SCLEROSIS

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INTRODUCTION: Many textbooks or reviews mention a positive correlation between fasciculation potentials (FPs) in needle EMG and decremental responses following repetitive nerve stimulation (RNS) in ALS patients, although this is based on only 1 previous study. Some authors have argued that FPs are more frequent in less-affected muscles, whereas decremental responses have been reported to be greater in more severely-affected muscles. These contradictory features raise doubt over the above correlation.

OBJECTIVE: To investigate the correlation between FPs and decremental responses in ALS patients.

METHODS: Enrolled subjects were 30 ALS patients for whom both needle EMG and RNS were conducted on the same trapezius muscle from February 2013 to October 2014. FPs were identified and counted by inspection of the raw needle EMG signal of around 3 minutes length, and the firing rate of FPs per minute (FR) was calculated. The decremental percentage (Decr%) was defined as the decrease of the peak-to-peak amplitude of the compound muscle action potential (CMAP) from the first to the fourth response. Pearson's correlation coefficient was calculated for 2/3 parameters: the FR, Decr%, and the amplitude of the initial CMAP (CMAPamp).

RESULTS: There was no correlation between the FR and Decr% (r=0.03) or between the FR and CMAPamp (r=0.04). A significant negative correlation was observed between the CMAPamp and Decr% (r=−0.56, p<0.005).

SUMMARY/CONCLUSION: FPs are not correlated with the decremental response or CMAPamp, both of which reflect disease severity.

CASTLEMAN’S DISEASE NEUROPATHIES

Elie Naddaf, Angela Dispenzieri, Jay Mandrekar, Michelle Mauermann (Rochester, MN)

INTRODUCTION: Castleman’s disease (CD) is a rare lymphoproliferative disorder with a broad spectrum of clinical manifestations. It can be associated with POEMS (polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes) syndrome.

OBJECTIVE: To define the peripheral neuropathy (PN) phenotypes seen with CD.

METHODS: A retrospective chart review for patients with biopsy-proven CD between January 2003 and December 2014 was conducted. Patients with PN were divided into 2 groups: CD without POEMS (CD-PN) and CD with POEMS (CD-POEMS). POEMS patients were used as control subjects. Clinical, electrophysiologic, and laboratory characteristics were collected.

RESULTS: There were 7 patients with CD-PN, 20 with CD-POEMS, and 122 with POEMS. CD-PN patients had the mildest neuropathy characterized by predominant sensory symptoms and mild distal sensory deficits (median Neuropathy Impairment Score [NIS] of 7 points). Although both CD-POEMS and POEMS patients had a severe sensorimotor neuropathy, CD-POEMS patients were less affected (median NIS of 33 and 66 points, respectively). The degree of severity was also reflected on EDX testing where CD-PN patients demonstrated a mild degree of axonal loss, followed by CD-POEMS and then POEMS. Demyelinating features, defined by the European Federation of Neurological Societies/Peripheral Nerve Society criteria, were present in 43% of CD-PN, 78% of CD-POEMS, and 86% of POEMS.

CONCLUSION: There is a spectrum of demyelinating PN phenotypes associated with CD. CD-PN is sensory predominant and is the mildest phenotype, whereas CD-POEMS is a more severe, sensory and motor neuropathy. Compared to the POEMS cohort, CD-POEMS neuropathy has a similar but less severe phenotype. Whether these patients respond differently to treatment deserves further study.
SPINAL ACCESSORY NERVE: CLINICAL CORRELATIONS AND NEW DISCOVERIES
Subhadra Nori (Roslyn Heights, NY)

INTRODUCTION: Nearly 20 years ago, the author and colleagues published a Muscle & Nerve paper titled “Utilization of Intraoperative Electroneurography to Understand the Innervation of the Trapezius Muscle.”

OBJECTIVE: To determine whether there has been any useful novel information since.

METHODS: A literature search was performed using the key words spinal accessory nerve (SAN), neck dissections, and trapezius muscle and resulted in a total of 539 articles found with 15 remaining after exclusion criteria were applied. Of those, 7 were anatomic cadaveric dissections, 6 were live neck dissections, and 2 were retrospective literature reviews.

RESULTS: The literature review further confirms the previous findings that the SAN is the main motor supply to the trapezius muscle. Although several motor contributions from the cervical plexus—mainly C2/3/4—exist, these contributions are not consistent or significant enough to substitute for SAN function. Complex anastomoses do exist between the cervical nerves and lower cranial nerves. Novel descriptions and further details of branching patterns of the SAN have been discussed by a few authors. It is the opinion of the majority of authors that a serious effort should be made to identify all these branches and different patterns of innervation. Preservation and identification of these branches may result in better postoperative functional outcomes.

SUMMARY/CONCLUSION: This review confirms the author and colleagues’ previous finding that the SAN is the main nerve supply to the trapezius muscle. Although there are cervical plexus contributions, they are not consistent or significant. In order to achieve better functional outcomes, and to avoid the disabling shoulder syndrome, all nerves should be preserved.

DIABETIC TRUNCAL POLYNEUROPATHY: CLINICAL AND ELECTROPHYSIOLOGICAL ABNORMALITIES
Chilvana Patel, Suryanarayan Vishnubhakat (Greatneck, NY)

INTRODUCTION: Diabetic truncal polyneuropathy (DTPN) is a well-recognized form of diabetic neuropathy, though it is often misdiagnosed and leads to unnecessary investigations. While clinical presentations have been described, electrophysiological abnormalities have not been described in detail.

OBJECTIVE: To present 9 cases of DTPN with clinical and electrophysiological abnormalities over a period of 12 years.

METHODS: This study was a retrospective analysis of clinical and EDX studies in 9 patients with DTPN.

RESULTS: All 9 patients (6 male, 3 female; age range: 48-71 years) had type 2 diabetes mellitus (DM). Three patients were on insulin, and 6 patients were treated with oral hypoglycemic agents. Eight of 9 patients had DM for 2-10 years, and 1 patient was discovered to be diabetic at presentation. All patients reported truncal paresthesias in a unilateral thoracic dermatomal distribution (mostly in 2 or 3 segments). Six of 9 patients had abdominal muscle weakness and 1 had proximal limb muscle weakness. NCSs revealed sensory motor polyneuropathy in all. Needle EMG revealed abnormalities in limb muscles in 7 patients, acute/chronic neuropathic changes in paraspinal muscles in 6, and denervation in abdominal muscles in 5.

SUMMARY/CONCLUSION: DTPN presents with abdomino-thoracic painful paresthesias in usually 2-3 thoracic dermatomes with abdominal muscle weakness in two-thirds of patients. All patients have distal sensory motor peripheral neuropathy. Diabetic patients with truncal paresthesias with or without abdominal muscles weakness should raise suspicion of DTPN, and careful electrophysiology studies will confirm the diagnosis in majority of the patients, resulting in proper treatment and avoidance of unnecessary investigations.
NEUROPATHY ASSOCIATED WITH RECURRENT INTERMITTENT HYPOXIA
Hebatallah Rashed, Adel Marei, Nagia Fahmy (Cairo, Egypt)

INTRODUCTION: Chronic hypoxia is a risk factor for polyneuropathy. Yet, the impact of recurrent hypoxia (as in sleep apnea disorders) on nerve function has not been well established.

OBJECTIVE: To study (1) the risk of occurrence of neuropathy and its pattern in a group of patients with sleep apnea and (2) its correlation with the degree of oxygen desaturation.

METHODS: Thirteen patients diagnosed clinically and by polysomnogram (PSG) with sleep apnea (apnea hypopnea index >5) were included. Patients with symptoms of neuropathy, concomitant illnesses that may predispose to neuropathy (i.e., diabetes, renal or liver dysfunction, or chronic obstructive pulmonary disease), heavy smokers, patients over 50, and patients with family history of neuropathy were excluded. Neurological examination was performed with no signs of neuropathy. Patients then underwent NCSs. After that, patients were categorized into 2 groups: group 1 (apnea+neuropathy) and group 2 (apnea only).

RESULTS: It was found that 6/13 patients (46.15%) had electrophysiological abnormalities consistent with neuropathy. Among them, 1 patient had purely motor neuropathy, 2 patients (33.3%) had purely sensory polyneuropathy, and 3 patients (50%) had mixed sensorimotor polyneuropathy. Regarding the pathology, 4 patients (66.6%) had axonal neuropathy, and 2 patients (33.3%) had mixed axonal-demyelinating neuropathy, and this was statistically significant. Patients in group 1 showed statistically significant oxygen desaturation compared to patients in group 2.

SUMMARY/CONCLUSION: Recurrent intermittent hypoxia could be an independent risk factor for axonal damage of peripheral nerves. Also, severity of neuropathy correlates with the degree of oxygen desaturation. Thus, continuous positive airway pressure (CPAP) treatment could offer a possible treatment for neuropathy in those patients.

Hebatallah Rashed, PhD, IFCN North American Chapter Fellowship Award Recipient

ULNAR NEUROPATHY AT THE WRIST ASSOCIATED WITH DUPUYTREN'S CONTRACTURE: MAGNETIC RESONANCE IMAGING AND ELECTRODIAGNOSTIC FEATURES
Brion Reichler, Darryl Sneag (New York, NY)

INTRODUCTION: Ulnar neuropathy at the wrist (UNW) is rare by comparison with that at the elbow. The most common causes include external pressure and compression by a mass. Since Dupuytren's contracture (DC) is often associated with finger flexion deformities, weakness due to focal motor neuropathy could easily be overlooked. There is only 1 case report in the literature of DC compressing the ulnar nerve in Guyon's canal.

OBJECTIVE: To report a case of UNW isolated to the deep motor branch, in association with DC and no other discernible cause.

CASE REPORT: A man with no history of trauma, arthropathy, or activities involving repetitive extrinsic compression presented with chronic atrophy and weakness of the right hand, without sensory complaints. Examination revealed diffuse atrophy and weakness limited to ulnar intrinsic hand muscles.

RESULTS: The right ulnar motor amplitude at the abductor digiti minimi (ADM) was 12% of the contralateral, with prolonged distal latency but normal proximal conduction. Ulnar sensory responses were normal. Needle EMG showed evidence of active denervation and reinnervation limited to the ADM, first dorsal interosseous, and ulnar lumbricals. MRI showed thickening of the palmar fascia, with scarring and remodeling of the perineural fat within the distal portion of Guyon's canal, surrounding the ulnar nerve. It also showed denervation atrophy of the hypothenar muscles, fourth lumbrical, and medial interossei.

SUMMARY/CONCLUSION: DC is a rare cause of UNW. In view of the finger flexion deformities that often accompany it, associated ulnar neuropathy may actually be underrecognized and needs to be considered and ruled out.
MASSIVE FIBROLIPOMATOUS HAMARTOMA CAUSING RECURRENT MEDIAN NEUROPATHY
Kaye Sedarsky, Scott Wallace, Patrick Malafarpace, Robert Shih, Leon Nesti, Walter Faillace, Mark Landau, Jonathan Smith (Bethesda, MD)

INTRODUCTION: Fibrolipomatous hamartoma (FLH), a benign, rare tumor characterized by proliferation of fibroadipose tissue within the epineurium and perineurium, is in the differential of peripheral nerve tumors. Characteristic imaging findings may facilitate diagnosis of FLH and allow conservative management, as operative treatment often results in neurological deficits.

OBJECTIVE: To discuss electrodiagnosis, imaging findings, and treatment options in a rare median nerve tumor.

METHODS: Presented here is a case of late failure of carpal tunnel release due to progressive enlargement of a median nerve tumor with initial imaging and biopsy concerning for neurofibroma, subsequently treated with complete excision with biopsy confirming FLH.

CASE REPORT: A 24-year-old man presented with recurrence of disabling wrist pain, paresthesia, and an enlarging wrist mass 1 year after successful carpal tunnel release. Pre-release MRI and biopsy were consistent with neurofibroma. There were no stigmata or family history of neurofibromatosis. Examination revealed intact strength, decreased sensation in the thumb and index and middle fingers, and no macrodactyly. Electrodiagnosis disclosed median mononeuropathy at the wrist. Ultrasound demonstrated an enlarged nerve with large, hypoechoic fascicles and prominent hyperechoic perineurium. MRI showed a fusiform, coaxial cable-like mass. Concern for malignant transformation prompted excisional biopsy; pathology confirmed FLH. Sensory deficits worsened, but pain resolved and thenar motor function was preserved.

SUMMARY/CONCLUSION: The differential for intraneural mass lesions includes benign and potentially malignant lesions. Aggressive surgical treatment of FLH is usually unnecessary, but ambiguous data and refractory pain may necessitate resection. Imaging may clarify the differential. Relatively preserved function postoperatively suggests that variations in anatomy may allow better surgical outcome in some patients.

Kaye Sedarsky, MD, Resident and Fellow Member Award Recipient

STATIC AND DYNAMIC SONOGRAPHIC EVALUATION OF POSTERIOR INTEROSSEOUS NERVE INJURIES
Elena Shanina, Ruby Patton, Robert Smith (Galveston, TX)

INTRODUCTION: The posterior interosseous nerve (PIN) is vulnerable to proximal radius fractures and dislocations. EDX studies can diagnose posterior interosseous neuropathy, but they seldom give etiological data. Sonographic evaluation not only provides nerve/surrounding tissue morphology, but it also enables dynamic assessment of anatomical topography and nerve compression during flexion/extension and supination/pronation.

OBJECTIVE: To explore the diagnostic role of neuromuscular ultrasound in evaluating PIN injury.

METHODS: In a series of 6 PIN traumatic injuries, all patients underwent EDX testing and sonographic evaluations. Peripheral nerve ultrasound employed an 18 MHz transducer, with video-loop recording documenting dynamic elbow flexion/extension and pronation/supination tests.

RESULTS: All patients had EDX evidence of posterior interosseous neuropathy. One patient had double crash syndrome, including humeral shaft and radial head fractures resulting in radial nerve palsies at both the spiral groove and PIN, respectively. Sonographic evaluations showed enlargement of the PIN to 5.14±1.07 mm² (30 normal control subjects: 1.60±0.51 mm²) and defined sites of short or extended foci of enlargement. Hardware was visualized by ultrasound: screws could be counted and positions of their tips in relation to surrounding tissues could be easily identified. Dynamic testing did not document change in PIN cross-sectional area (p=0.588), but such testing revealed dynamic impingement of the PIN by a screw in 1 case.

SUMMARY/CONCLUSION: Sonographic evaluation provides significantly enhanced detail regarding location and etiology of PIN injury, allowing examiners to differentiate traumatic and iatrogenic causes, and guide appropriate surgical exploration.
TWO CASES OF UNILATERAL FLAIL ARMS: AN ARGUMENT FOR NON-DIABETIC CERVICAL RADICULOPLEXUS NEUROPATHY
Kong Truong, Theresa LaBarte, Bryan Tsao (Loma Linda, CA)

INTRODUCTION: The association between nondiabetic cervical radiculoplexus neuropathy and Parsonage-Turner syndrome is uncertain.

OBJECTIVE: To report 2 cases of unilateral pan-brachial plexopathy whose clinical and EDX features are more convincing for nondiabetic cervical radiculoplexus neuropathy.

CASE REPORTS: Patient 1: A 68-year-old male presented with abrupt onset of left upper limb pain followed weeks later by weakness. His examination disclosed left upper limb weakness, atrophy, reduced sensation, and reduced reflexes. Sixteen years after his initial onset he had another acute bout of paroxysmal pain and weakness. Needle EMG/NCSs 3 years after his last bout demonstrated a left pan-brachial plexopathy with widespread chronic motor axon loss. Patient 2: A 69-year-old female with breast cancer and radiation therapy 16 years prior presented with acute right hand pain for 7 months. This was followed by progressive right upper limb weakness and numbness over a 1-year period. Her examination disclosed diffuse right upper limb weakness, atrophy, reduced sensation, and reduced reflexes. Needle EMG/NCSs 4 years after onset showed a right pan-brachial plexopathy with widespread motor axon loss and no myokymic potentials. In both patients, needle EMG sampling of paraspinal muscles and the contralateral limb was normal. Extensive tests for autoimmune disorders, vasculitis, and diabetes, as well as MRI of the brachial plexus, were normal.

SUMMARY/CONCLUSION: The widespread nature of both patients’ plexopathies, the recurrent bout in patient 1, and the prolonged course of pain and weakness in patient 2 are highly atypical for Parsonage-Turner syndrome. It is proposed that they fit within the spectrum of nondiabetic cervical radiculoplexus neuropathy.

THE PATHOGENIC ROLE OF FIBRONECTIN CONNECTING SEGMENT-1 IN CHRONIC INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY
Kelsey Greathouse, Steven Palladino, Rebecca Beacham, Eric Scott Helton, Chaoling Dong, Eroboghene Ubogu (Birmingham, AL)

INTRODUCTION: The mechanisms of leukocyte trafficking across the blood-nerve barrier (BNB) in chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) are incompletely understood. Fibronectin connecting segment-1 (FNCS1) is an alternatively spliced fibronectin variant that serves as a counterligand for leukocyte α4 integrin implicated in pathogenic neuroinflammation.

OBJECTIVE: To determine the role of FNCS1 in CIDP patient leukocyte trafficking across the BNB in vitro and in chronic demyelinating neuritis in vivo using a spontaneous murine CIDP model.

METHODS: Seven untreated CIDP patients’ leukocytes were infused into a flow-dependent in vitro BNB model with 25 µM FNCS1 peptide (contains the critical binding sequence for α-4 integrin) with appropriate control subjects including an irrelevant (FNCS1C) peptide. Real-time video microscopy was performed to quantify trafficking and an adhesion/migration index (AMI) was calculated. 24-week old female B7-2-deficient non-obese diabetic mice were treated with 2 mg/kg FNCS1 or FNCS1C peptide for 5 days. Neuromuscular Severity Score (NMSS) evaluation, dorsal caudal tail nerve (DCTN) and sciatic motor nerve electrophysiology, and sciatic nerve inflammatory cell counts and morphometric assessment of demyelination were performed. The Mann Whitney U or student’s t-test determined statistical significance.

RESULTS: FNCS1 peptide maximally inhibited CIDP leukocyte trafficking in vitro, with a mean AMI of 0.77 compared to an AMI of 2.09 for FNCS1 peptide. FNCS1 peptide treatment resulted in statistically significant differences in mean NMSS, motor conduction velocities, and distal waveform durations in the DCTN and sciatic nerve as well as CD45+ leukocytes and percent demyelinated area per section compared to control subjects.

SUMMARY/CONCLUSION: These results imply an important role for FNCS1-α4 integrin-mediated leukocyte trafficking in CIDP.
DIAGNOSTIC VALUE OF MAGNETIC RESONANCE NEUROGRAPHY IN PATIENTS WITH SUSPECTED NEUROGENIC THORACIC OUTLET SYNDROME

Lita Vargas, Ricardo Claudio Reisin, Ana Pardal, Cintia Marchesoni, Luciana Leon Cejas, Daniela Binaghi (Buenos Aires, Argentina)

INTRODUCTION: The neurogenic thoracic outlet syndrome (nTOS) is caused by compression of the lower trunk of the brachial plexus. It is characterized by pain, paresthesias, weakness, and atrophy in the C8-T1 distribution.

OBJECTIVE: To analyze clinical, imaging, and electrophysiological findings in a series of patients with nTOS.

METHODS: Medical records of patients with clinical suspicion of nTOS who had both needle EMG and magnetic resonance neurography (MRN) studies from 2010 to 2014 were evaluated retrospectively.

RESULTS: Four of 14 patients fulfilled the clinical, needle EMG, and MRN criteria (3 women, mean age: 29.5 years, age range: 14-41 years). Needle EMG showed reduced sensory amplitude of the ulnar nerve with preserved median sensory responses, decreased motor amplitude of median or ulnar nerves, and acute and chronic denervation in C8-T1 myotomes. MRN revealed in all patients increased signal on T2-weighted sequence of the lower trunk, with presented contrast enhancement in 2. Patients 1 and 2 showed a rudimentary cervical rib and a fibrous band; patient 3, a rudimentary cervical rib and a transverse mega-process; and patient 4, a scalenus minimus muscle. The mean delay until diagnosis was 2 years (range: 6 months to 3 years). In the remaining group (10/14 patients), plexitis (3), infiltration by cancer (2), schwannoma (2), post-surgical fibrosis (1), multifocal motor neuropathy (1), and CIDP (1) were detected.

SUMMARY/CONCLUSION: Clinical and electrophysiological studies are fundamental tools to diagnose nTOS. MRN has become the imaging technique of choice to rule out mimicking conditions.

Lita Vargas, MD, IFCN North American Chapter Fellowship Award Recipient

THE IMPORTANCE OF GENETIC TESTING IN FAMILIAL AMYLOID POLYNEUROPATHY CAUSED BY TRANSTHYRETIN GENE MUTATIONS

Rocio Vazquez do Campo, Elizabeth Mauricio (Jacksonville, FL)

INTRODUCTION: Transthyretin familial amyloid polyneuropathies (TTR-FAPs) present with a length-dependent axonal peripheral neuropathy and various degrees of autonomic dysfunction. Sporadic cases with no prior family history have been reported. Mutations in the TTR gene, a precursor of amyloid, lead to accumulation of amyloid in various organs causing tissue damage and dysfunction. Due to its focal and irregular distribution, amyloid deposits are not identified in tissue biopsies in 30% of cases.

OBJECTIVE: To recognize the limitations of tissue biopsy and highlight the importance of genetic testing in FAP.

CASE REPORT: A 62-year-old man presented with an 8-month history of progressive distal weakness and painful paresthesias in all extremities preceded by 2 years of constipation and orthostatism. His father had died prematurely from unknown cardiac disease. On examination, he had distal weakness and sensory loss to all modalities in a high stocking-and-glove distribution; his reflexes were absent. Needle EMG revealed a severe length-dependent, primarily axonal, sensorimotor peripheral neuropathy. Sural nerve biopsy demonstrated decreased density of myelinated fibers and axonal degeneration without inflammation. Both sural nerve biopsy and fat aspirate did not demonstrate amyloid deposits on congo red preparations. Despite these findings, amyloid genetic studies were pursued and a mutation in the TTR gene was identified.

SUMMARY/CONCLUSION: TTR-FAP must be considered in patients with progressive axonal polyneuropathy and autonomic features. The absence of amyloid deposits on biopsy or family history does not exclude the diagnosis. Genetic testing to identify TTR gene mutations is often necessary. Establishing the correct diagnosis is crucial, as promising targeted therapies are currently under investigation.
VEUMURAFENIB-ASSOCIATED NERVE LARGE ARTERIOLE VASCULITIS
Amy Visser, Wei Wang, Gita Thanarajasingam, Judith Kaur, Shreyasee Amin, Christopher Klein, Joseph Matsumoto (Rochester, MN)

INTRODUCTION: Vemurafenib is a small molecule serine/threonine protein kinase with potent inhibition of mutated BRAF used in the treatment of metastatic melanoma.

OBJECTIVE: To report a patient with metastatic melanoma who developed nerve large arteriole vasculitis during treatment with vemurafenib.

CASE REPORT: A 67-year-old woman presented with 1 year of progressive dysesthesia, numbness, and weakness involving her distal lower and upper limbs. She was prescribed vemurafenib for treatment of metastatic melanoma 9 months prior to symptom onset with induction of an oncologic remission. EDX testing was notable for a severe, primarily axonal, sensorimotor polyradiculoneuropathy. A right sural nerve biopsy showed a decreased density of myelinated fibers in a multifocal pattern with an increased rate of axonal degeneration, segmental demyelination, and myelin remodeling. Small, moderate, and large collections of polyclonal perivascular inflammatory cells were present in the epineurium with associated destruction of small, medium, and large arteriole vessel walls. These areas also showed hemosiderin deposition and neovascularization. Shortly prior to these findings, vemurafenib was discontinued after discovery of new metastatic lesions and treatment with pembrolizumab was initiated. Marked symptomatic improvement was noted with close neurologic followup over the ensuing 6 months.

SUMMARY/CONCLUSION: Although treatment with vemurafenib has been linked to the development of panniculitis with or without vasculitis, facial palsy, as well as sarcoidosis, this is the first case of nerve large arteriole vasculitis associated with vemurafenib use. The mitogen-activated protein kinase (MAPK) pathway seems to play a critical role; however, the pathophysiology of this process remains unclear.

Amy Visser, MD, Resident and Fellow Member Award Recipient

OCULOMOTOR AND MASTICATOR MUSCLE ENHANCEMENT IN A CASE OF MULTIPLE CRANIAL NERVE PALSIES AND INTERNUCLEAR OPHTHALMOPLEGIA ASSOCIATED WITH HERPES SIMPLEX VIRUS INFECTION
Crystal Yeo, Ericka Simpson, Milvia Pleitez, Robert Smith (Houston, TX)

INTRODUCTION: Acutely or subacutely denervated skeletal muscle has high signal on T2-weighted MRI and enhances with gadolinium contrast, whereas chronically denervated muscle shows atrophy and fatty infiltration with high signal on T1-weighted MRI. Similar muscle pathology and MRI changes can be found with inflammatory, infectious, or neoplastic processes. However, denervation should be considered when the clinical context is appropriate.

OBJECTIVE: To report for the first time oculomotor and masseter muscle enhancement in the context of multiple cranial nerve palsies, gaze center involvement, and herpes simplex virus (HSV) infection.

CASE REPORT: A young, previously healthy patient developed multiple cranial nerve palsies and internuclear ophthalmoplegia over 1 year. Her MRIs showed oculomotor and masticator muscle enhancement. Workup was negative except for the presence of HSV IgM.

SUMMARY/CONCLUSIONS: In cranial nerve investigations where needle EMG and NCSs are difficult to perform, MRI can be used as an ancillary test to support denervation. Further studies to investigate MRI findings during reinnervation would be desirable for diagnostic and prognostic purposes. HSV IgM is found in acute HSV infection or chronic infection with reactivation. HSV can be latent in many cranial ganglia but has not been previously reported to occur with multiple cranial neuropathies. Patients with subacute presentation of multiple cranial neuropathies should be evaluated for HSV infection and reactivation.
NERVE ULTRASOUND IDENTIFIES ABNORMALITIES IN CHRONIC INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY
Dongqing Zhu (Shanghai, China), Yu Zhu (Syracuse, NY), DONG TIAN (Shanghai, Shanghai), Kai Qiao, xiangjun chen (Shanghai, China), Robert Weber (Syracuse, NY)

INTRODUCTION: Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is an autoimmune disease characterized by recurrent demyelination and remyelination with resultant thickening of the peripheral nerves. The diagnosis is based mainly on the clinical presentation and electrophysiological detection of demyelination.

OBJECTIVE: To explore the new research area of detection of nerve abnormalities in CIDP by sonography.

METHODS: Three patients with CIDP underwent NCSs as well as ultrasonographic examination of the median, ulnar, sciatic, fibular, posterior, and tibial nerves and the brachial plexuses bilaterally, using a standardized protocol. The cross-sectional area (CSA) of each nerve was calculated and the segment nerve thickening as well as nerve vascularization was assessed.

RESULTS: On sonography, multiple nerves in 3 patients were found to be diffusely hypertrophic and hypoechoic. CSAs of the brachial plexuses and the median, sciatic and femoral nerves were increased compared to normal values. The brachial plexuses findings were confirmed by MRI. All 3 patients were evaluated with NCSs, including short segment studies to localize motor conduction block. It was also identified that multifocal enlargement of nerve trunks precisely correlated with neurophysiological conduction block. Increased vascularization was seen in 2 patients in 1 nerve and in 1 patient in multiple nerves.

SUMMARY/CONCLUSION: The findings of diffusely hypertrophic and hypoechoic nerve (proximal and distal) segment enlargement at the site of conduction block as well as increased nerve vascularization may be tools to better diagnosis CIDP.

Dongqing Zhu, MD, IFCN North American Chapter Fellowship Award Recipient

TELEPHONIC SINGLE BREATH COUNT TEST ADMINISTERED BY NURSES IN DIAGNOSING MYASTHENIA EXACERBATION
Syeda Alqadri (Columbia, MO), Raghav Govindarajan (Columbia, MO)

BACKGROUND: Delay in diagnosing myasthenia gravis (MG) exacerbation can result in significant morbidity and rarely mortality. Since there are no definite tests for MG exacerbation, the symptoms can be nonspecific and confusing. It is not uncommon for patients to call about their MG exacerbation and for nurses to answer them. This study assessed the role of the single breath count test (SBCT) administered by nurses over the phone in triaging MG exacerbation.

METHODS: This prospective, non-blinded pilot study included all consecutive patients who called the clinic with suspected symptoms of exacerbation from January to December 2015. The diagnosis of MG exacerbation was at the discretion of treating neuromuscular physician. Nurses were trained in administering the SBCT and received continuing nursing education in MG prior to the study. In suspected MG exacerbation (<25 SBCT for this study), the SBCT was administered over the phone.

RESULTS: The study included 25 patients (10 male, 15 female, age range: 16-75 years; 12 seropositive); 5 were on IVIg maintenance therapy, 1 on IVIg and steroids, and the rest on steroids. Of the 45 individuals who called, 20 had an SBCT less than 25 and did had a true exacerbation, 8 had an SBCT less than 25 and did not have an exacerbation, 5 had an SBCT greater than 25 and were deemed to not have an exacerbation but were diagnosed as having one. The positive predictive value of telephonic SBCT was 71%.

CONCLUSION: The SBCT is a good telephonic quantitative biomarker which can identify MG exacerbation and can be administered even by allied healthcare professionals.

Syeda Alqadri, MD, Resident and Fellow Member Award Recipient
MYASTHENIA GRAVIS IN GRAFT-VERSUS-HOST DISEASE
Jose Avila, Matthew Burford, Robert Bucelli (St. Louis, MO)

INTRODUCTION: Neuromuscular complications may occur in 8.1% of patients after allogeneic hematopoietic stem cell transplant (HSCT), usually in the setting of graft-versus-host disease (GVHD). Myasthenia gravis (MG) is the least common of these complications, with an overall frequency of <1%.

OBJECTIVE: To describe 2 patients with MG as a manifestation of chronic GVHD.

CASE REPORTS: Case 1 was a 32-year-old man with 4 weeks of oculobulbar and proximal arm weakness. He had a history of acute lymphocytic leukemia and received HSCT 81 months prior to presentation. His course was complicated by refractory GVHD, treated with prednisone, mycophenolate, and tacrolimus. Case 2 was a 61-year-old man with 6 months of ocular and neck extensor weakness. He had a history of acute myeloid leukemia and underwent HSCT 71 months prior to presentation. He developed GVHD and was on chronic prednisone. He had previously received tacrolimus, mycophenolate, and cyclosporine. Both patients progressed to respiratory failure, requiring mechanical ventilation. Both had acetylcholine receptor antibodies and no evidence of thymoma. Treatment consisted of plasma exchange (PE) and rituximab in case 1. Case 2 was initially managed with a higher dose of prednisone but later required PE and mycophenolate due to adverse effects of corticosteroids.

SUMMARY/CONCLUSION: MG is a rare, late manifestation of chronic GVHD. It should be considered in patients with HSCT presenting with oculobulbar and ventilatory deficits. Treatment is challenging as patients are already on immunosuppressive drugs for GVHD and may have developed adverse effects due to longterm use, which limits therapeutic options.

Jose David Avila, MD, Resident and Fellow Member Award Recipient

PEDIATRIC LAMBERT–EATON MYASTHENIC SYNDROME: A CASE REPORT
Carolina Vivar, Jose Avila (St. Louis, MO), Hoda Abdel-Hamid (Pittsburgh, PA)

INTRODUCTION: Lambert–Eaton myasthenic syndrome (LEMS) is an autoimmune presynaptic neuromuscular junction disorder associated with voltage-gated calcium channel antibodies (VGCC abs). Pediatric LEMS is rare and less likely to be paraneoplastic compared to adult patients.

CASE REPORT: A 16-year-old girl presented with 1 year of progressive proximal leg weakness. MRI of the brain showed multiple subcortical and periventricular white matter hyperintensities without enhancement or restricted diffusion. Extensive testing, including creatine kinase and cerebrospinal fluid analysis, was unremarkable. She was ultimately diagnosed with multiple sclerosis and received IV methylprednisolone with significant improvement. Symptoms relapsed, requiring repeated corticosteroid infusions. MRI of the brain remained unchanged. EDX studies, performed 3 months later, showed small amplitude compound muscle action potentials (CMAPs) and normal sensory nerve action potentials. Brief exercise produced a >100% increase in CMAP amplitude, and 30-Hz repetitive nerve stimulation demonstrated an incremental response, consistent with LEMS. VGCC abs were negative. There was no evidence of malignancy on CT of the chest, abdomen, and pelvis on multiple occasions. The patient was treated with prednisone. IVIg and azathioprine were added later. Nine years after presentation, she has mild proximal weakness with intermittent exacerbations.

SUMMARY/CONCLUSION: LEMS is a treatable condition that should be considered in pediatric patients presenting with proximal limb weakness. Although non-paraneoplastic LEMS is more common in children, malignancy should be excluded. The significance of the white matter changes in this patient is unclear, but may represent a central demyelinating disorder.

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BEYOND THE ANTIBODIES: SERA METABOLOMIC BIOMARKER SIGNATURES DISCRIMINATE MYASTHENIC AND HEALTHY COHORTS
Derrick Blackmore, Nan Wang, Liang Li, Zaeem Siddiqi (Edmonton, Canada)

INTRODUCTION: Few biomarker studies have assessed the serum metabolome in patients with myasthenia gravis (MG). Recent advancements in metabolomic profiling with high coverage may facilitate description of an enhanced MG biomarker signature.

OBJECTIVE: To construct and compare the serum metabolomic profiles of MG patients and healthy individuals using a novel chemical isotope labeling liquid chromatography-mass spectrometry (CIL LC-MS) technique.

METHODS: CIL LC-MS uses different labeling reagents to target chemical group-based submetabolomes to provide in-depth metabolomic analysis. 12C-dansylation labeling of individual samples and 13C-dansylation labeling of pooled samples from 49 patients with seropositive MG and 50 age/gender matched healthy control subjects was undertaken. The amine/phenol submetabolome changes among the labeled samples were quantified based on subsequent analysis of the 13C- and 12C-labeled mixture by LC-MS.

RESULTS: On average, 4084±149 (n=49) and 3972±492 (n=50) metabolites were detected in sera from MG samples and control subjects, respectively—a total of 5711 metabolites in all samples. Orthogonal partial least squares discriminant analysis showed a clear separation of 2 groups (R2=0.98, Q2=0.80). The receiver operating characteristic (ROC) curve using 7 metabolites produced an area under the curve (AUC) value of 0.859 (0.806-0.920, 95% CI) with 91% specificity and 70% sensitivity.

CONCLUSIONS: High-coverage metabolomic profiling reveals that serum metabolomes of MG patients differ considerably from healthy control subjects, substantiating the probability of finding metabolic biomarkers specific to MG.

PURE OCULAR WEAKNESS AS THE SOLE CLINICAL PRESENTATION OF LAMBERT–EATON MYASTHENIC SYNDROME
Alaa Bouzhar, Raghav Govindarajan (Columbia, MO)

BACKGROUND: Pure ocular weakness as the sole clinical presentation of Lambert–Eaton myasthenic syndrome (LEMS) is uncommon. The case described here started with a presentation of ptosis misdiagnosed as ocular myasthenia which was later diagnosed as LEMS.

CASE REPORT: A 59-year-old woman presented to the ophthalmology clinic with a 4-month history of daily headaches, double vision, and droopy eyes. Ophthalmological examination showed bilateral right worse than left ptosis, dysconjugate gaze, and complaints of double vision with images side-by-side on lateral gaze bilaterally. Ice pack test showed significant improvement in right ptosis. Acetylcholine binding/blocking/modulating receptor antibodies were negative. She was diagnosed with seronegative ocular myasthenia and referred for further evaluation. Motor/sensory examination was normal. Reflexes were 2+ and symmetric. Left ulnar motor NCS showed a compound muscle action potential (CMAP) of 4.5 mV (normal ≥5 mV). Slow repetitive nerve stimulation of the left ulnar nerve showed 20% decrement in the CMAP amplitude. Given her history of hyponatremia (sodium between 125-130 mmol/l thought secondary to selective serotonin reuptake inhibitors) and 30 pack/year smoking, short exercise testing (10 second) was initiated, following which her left ulnar CMAP increased to 10 mV (more than 100% increment). CT of the chest showed precarinal/left lower pretracheal mass with central necrosis, and biopsy confirmed small cell carcinoma. P/Q antibody was 3.62 nmol/l (normal ≤0.02, Mayo Clinic). Hyponatremia was due to paraneoplastic SIADH.

CONCLUSION: Presence of unexplained hyponatremia (or other paraneoplastic syndromes) along with history of smoking should alert to the possibility of LEMS, even in those patients who have been labelled as “seronegative ocular myasthenia.”

Alaa Bouzhar, Technologist Member Award Recipient
INTRODUCTION: Delay in diagnosing myasthenia gravis (MG) exacerbation can result in significant morbidity and rarely mortality. Since there are no definite diagnostic tests for MG exacerbation, easy to administer, quantitative clinical biomarkers—eye closure strength (ECS), neck flexion strength (NFS), and single breath count test (SBCT)—were assessed.

METHODS: This prospective, non-blinded pilot study included all consecutive patients who presented with symptoms of MG exacerbation from January to December 2015. The diagnosis of MG exacerbation was at the discretion of treating neuromuscular physician. ECS was graded on a scale: grade 4: eyelashes dug in, grade 3: eyelids can be opened but with some resistance, grade 2: eyelids can easily be opened, and grade 1: cannot completely close the eye. NFS was assessed in the supine position and graded according to the MRC scale. The SBCT was performed as per previously defined protocols.

RESULTS: Of the 20 patients (8 male, 12 female, age range: 16-70 years; 8 seropositive) included, 8 were on IVIg maintenance therapy, 1 on IVIg and steroids, and the rest on steroids. ECS grade 2 or less, NFS grade 3 or less, and SBCT less than 25 had significant association with MG exacerbation (p<0.05). There was strong correlation between the 3 parameters (r=0.55, p<0.05) at these values. In 3 cases, an ECS of 2 was seen with normal NFS/SBCT values, and 3 days later the patients returned with worsening shortness of breath and their SBCT was below 20 and NFS was 2.

CONCLUSION: Quantitative clinical biomarkers are good predictors of MG exacerbation, and ECS may be more sensitive.

INTRODUCTION: Single fiber EMG (SFEMG) is the most sensitive method to detect and diagnose neuromuscular junction (NMJ) diseases such as Lambert–Eaton myasthenic syndrome (LEMS) and myasthenia gravis (MG). Concentric and single fiber needle electrodes yield comparable jitter results in normal healthy and MG patients.

OBJECTIVE: To analyze jitter by concentric needle electrode (CNE) SFEMG in LEMS patients.

METHODS: Fifteen subjects diagnosed with LEMS underwent CNE SFEMG in the extensor digitorum communis (EDC) and tibialis anterior (TA). Twelve healthy control subjects were also enrolled for examination of the EDC and 8 for the TA. Electrophysiological examinations—including nerve conduction velocity, needle EMG, and repetitive nerve stimulation (RNS) with low (1-5 Hz) and high (50 Hz) frequency—were performed in all patients to exclude other neuromuscular diseases.

RESULTS: Ten males and 5 females were diagnosed with LEMS via a compound muscle action potential increase of 100% during 50-Hz RNS. All patients showed a markedly increased jitter in the EDC (88.8±25.6 µs) and TA (92.2±30.2 µs) compared to healthy control subjects (28.0±4.4 µs, 30.5±9.3 µs, respectively).

SUMMARY/CONCLUSION: These results suggest that CNE SFEMG can uncover abnormality of NMJ transmission.

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EPITOPE SPREADING: A POSSIBLE MECHANISM OF CLINICAL DETERIORATION IN GUILLAIN–BARRÉ SYNDROME
Ghazala Hayat, Jafar Kafaie, Elizabeth Strong (St. Louis, MO)

INTRODUCTION: Autoimmune diseases can be triggered by a variety of factors; epitope spreading can contribute to their development and also can lead to diverse presentation. Many infectious agents, including viruses and bacteria, can trigger autoimmune processes in the CNS and PNS. Epitope spreading can explain the diversification of the epitopes recognized by humoral or cell mediated immune systems leading to distinct concomitant clinical presentations.

OBJECTIVE: To report a case of pre- and postsynaptic neuromuscular junction disorder followed by axonal Guillain–Barré syndrome (GBS) associated with Campylobacter jejuni infection.

CASE REPORT: A 58-year-old woman presented with a 1-week history of vomiting and abdominal pain, followed by difficulty walking. Examination showed generalized areflexia with mild 4/5 (MRC) proximal weakness in lower extremities. Laboratory evaluation showed albuminocytologic dissociation and positive C. jejuni titers. Electrophysiological studies were consistent with axonal GBS. She had fluctuating confusion. IVIg therapy was instituted; weakness progressed to involve upper extremities, primarily proximally. Evaluation revealed positive acetylcholine receptor binding antibodies, calcium channel antibodies, and neuronal voltage-gated potassium channel antibodies. Plasma exchange (PE) improved the motor strength and mental status.

SUMMARY/CONCLUSION: Intra- and intermolecular epitope spreading of B- and T-cells has been speculated in various autoimmune diseases. To the best of the authors’ knowledge this is the first case of axonal GBS associated with C. jejuni infection progressing to involve pre- and postsynaptic junctions as well as nodes of Ranvier. Physicians should consider this phenomenon in deteriorating cases of autoimmune disorders, such as GBS. Early PE may be beneficial in these cases.

CONGENITAL MYASTHENIC SYNDROME MISDIAGNOSED AS SERONEGATIVE MYASTHENIA GRAVIS FOR A LIFETIME: A REPORT OF A NOVEL MUTATION IN CHRNE
Darine Kassar, Mohamed Teleb (El Paso, TX), Stanley Iyadurai (Columbus, OH)

INTRODUCTION: Congenital myasthenic syndrome (CMS) is a rare disorder. History, clinical evaluation, and absence of detectable antibodies should prompt the search for it.

OBJECTIVE: To report a case of CMS mistaken for myasthenia gravis for over 35 years.

CASE REPORT: A 35-year-old man with a prior diagnosis of seronegative myasthenia gravis (SNMG) (diagnosed at age 18) was seen in the clinic with complaints of ptosis, diplopia, weakness, and poor exercise tolerance. Ptosis and fatigue with physical activity had been noted in early childhood and a diagnosis of SNMG was made, in the setting of negative acetylcholine receptor (AChR) and muscle-specific kinase antibodies. Prior treatments included thymectomy, IVIg, plasma exchange, prednisone, and pyridostigmine, with favorable outcome only with pyridostigmine. No similar family history. Examination revealed fatigable ptosis, limitation in extraocular movements, and proximal weakness in limb-girdle muscles. 3-Hz repetitive nerve stimulation showed significant decrement. CMS was suspected and gene testing revealed 2 heterozygous frameshift mutations (c.130; 1 bp duplication of G; codon 44 and c.1229; 1 bp duplication of C; codon 434) in the epsilon subunit of the AChR (CHRNE) gene. While the former mutation has been reported previously, the latter has not. Symptoms improved with albuterol therapy.

SUMMARY/CONCLUSION: To the authors’ knowledge, this is the first study to report a novel frameshift mutation in the CHRNE locus at the nucleotide location 1229. Both the mutations found here lead to premature truncation of the CHRNE protein leading to a CMS phenotype. Finally, this study lends additional support to the usefulness of albuterol treatment in CHRNE CMS.
MYASTHENIA GRAVIS AN UNCOMMON STROKE MIMICKER
Victoria Levasseur, Raghav Govindarajan (Columbia, MO)

BACKGROUND: It’s not uncommon for myasthenia gravis (MG) to present with episodic or even with first time symptoms to the ER, thus mimicking stroke. The cases here were worked up as stroke in the ER/hospital and were later diagnosed as MG.

CASE REPORTS: (1) A 35-year-old morbidly obese woman presented with slurred speech/dysphagia. She had a normal MRI of the brain/CT angiogram and was discharged on aspirin. Two weeks later she had similar symptoms, and aspirin was changed to clopidogrel. A month later she had similar symptoms and was put on Coumadin®. Clinic workup showed elevated acetylcholine receptor antibodies (AChR-abs) with decrement on repetitive stimulation. Coumadin was stopped, and she is now on IVIg. (2) Following a concert, a 40-year-old singer presented with slurred speech. She had a normal MRI of the brain/CT angiogram and was discharged on aspirin and statin. Workup showed tongue atrophy, elevated muscle-specific receptor tyrosine kinase (MuSK) antibody, and decrement on repetitive stimulation. She is on IVIg after stopping aspirin. (3) A 75-year-old man with hypertension presented with slurred speech. He had a normal CT angiogram with an MRI showing moderate-to-severe white matter disease. He was put on aspirin and clopidogrel. With similar symptoms a month later, Coumadin and statins were added. Workup showed elevated AChR-abs and decrement on repetitive stimulation. Coumadin was stopped, and he is now on prednisone.

CONCLUSION: Acute or first time bulbar/ocular symptoms of MG can be an uncommon stroke mimicker. A high degree of clinical suspicion, especially in a busy ER, is needed to diagnose MG and avoid potentially harmful therapy.

Victoria Levasseur, BS, Resident and Fellow Member Award Recipient

CONCURRENT CHRONIC INFLAMMATORY Demyelinating Neuropathy and Myasthenia Gravis with Thymoma
Jin Li (Scarsdale, NY)

INTRODUCTION: Chronic inflammatory demyelinating neuropathy (CIDP) and myasthenia gravis (MG) are distinct and uncommon immune-mediated neuromuscular diseases. The concurrence of these 2 distinct autoimmune diseases is extremely rare.

OBJECTIVE: To report a rare case of concurrent CIDP and MG with thymoma.

CASE REPORT: A 65-year-old woman had a 1-year history of progressive CIDP that was refractory to IVIg and plasma exchange (PE), and she was only stabilized with cyclophosphamide. She was able to ambulate with a walker, but utilized a wheelchair most of the time. She presented to Westchester Medical Center with chest pain and shortness of breath. She was found to have an 11-cm stage I thymoma. Serum acetylcholine receptor antibodies were positive. After thymectomy, her MG was treated with steroid, IVIg, and PE. Her hospital course was prolonged and complicated with respiratory failure, thrombocytopenia second to IVIg, herpes encephalitis, and refractory seizures.

SUMMARY/CONCLUSION: The concurrence of CIDP and MG imposes clinical challenges for effective treatments and complicates the clinical course with life-threatening comorbidities.
ASSOCIATION OF MYASTHENIA GRAVIS AUTOANTIBODIES TO CLINICAL FEATURES AND MYASTENIC CRISIS
Yasir Malik, Abubaker Almadani, Javeed Dar, Suhail Alrukun (Dubai, UAB)

OBJECTIVE: To collect demographic details of the MG patients and to assess correlation of their clinical features with auto-antibodies.

BACKGROUND: Myasthenia gravis is a rare disorder of neuromuscular junction with annual incidence of approximately 10 to 20 new cases per million. It is mediated by auto-antibodies against the acetylcholine receptor (AChR-Ab), muscle specific tyrosine kinase (MuSK-Ab) or other striated muscle antibodies.

METHODS: We reviewed record of 143 patients of Myasthenia gravis, who visited Rashid Hospital, Dubai from 2000 to 2010. It was a retrospective observational study in which patient's record and > 5 years follow up was reviewed. It was a retrospective observational study in which patient's record and > 5 years follow up was reviewed

RESULTS: Total number of 143 MG patients were reviewed. Motor complaints were almost equal in both groups, bulbar features predominated in seropositive group 63% vs 25% (p=0.016) and none of the seronegative patients had respiratory complaint. Patients without autoantibodies had milder degree of disease (Osserman’s classes 1 and 2) whereas patients with autoantibodies had comparatively severer disease (predominantly Osserman’s class 3). Similar trends were shown on MGFA severity scale.

CONCLUSIONS: Myasthenia gravis is a rare autoimmune postsynaptic neuromuscular disorder mediated by auto-antibodies. Patients with antibodies display severer form of disease in comparison to seronegative patients. Seronegative MG mainly include ocular myasthenia, they have less incidence of autoimmune co-morbidities and experience myasthenia crisis very rarely.

TAKOTSUBO CARDIOMYOPATHY WITH MYASTHENIA GRAVIS EXACERBATION
Shail Thanki, Raghav Govindarajan (Columbia, MO)

INTRODUCTION: Myasthenia gravis (MG) is an autoimmune disorder with weakness and fatigability of skeletal muscles. The cardiovascular system is typically spared. The following 2 cases of MG developed Takotsubo cardiomyopathy during myasthenic exacerbation.

CASE REPORTS: (1) Bulbar myasthenia with Takotsubo cardiomyopathy: A 35-year-old woman with MG presented with worsening dysphagia and slurred speech following respiratory infection. During hospitalization she complained of chest pain and shortness of breath which was thought to be secondary to anxiety. Workup showed mildly elevated troponins. An electrocardiogram (EKG) showed ST segment elevation in V2-V3. An echocardiogram showed mild apical left ventricular hypokinesis. Cardiac catheterization was normal. The patient was shifted to the ICU and started on an angiotensin-converting-enzyme inhibitor. She was successfully treated with IVIg and discharged home with normal troponins and EKG. An echo performed a month later was normal. (2) Generalized myasthenia with Takotsubo cardiomyopathy: A 65-year-old woman with generalized MG presented with shortness of breath, fatigue, and dysphagia. She was admitted to the ICU and underwent plasma exchange (PE). A day later she developed chest pain and complained of worsening shortness of breath. Her pulmonary function tests were getting better so the symptoms were attributed to anxiety, but troponin was mildly elevated. An EKG showed ST segment elevation in V2-V3. An echo showed mild apical left ventricular hypokinesis. Cardiac catheterization was normal. An echo performed 2 months later was normal.

CONCLUSION: Takotsubo cardiomyopathy can be an uncommon presentation during myasthenic exacerbation. Careful cardiac monitoring (and not just pulmonary functions) during an exacerbation is needed so as to prevent fluid overload and blood pressure fluctuations from treatment with IVIg/PE.

Shail Thanki, BS, Resident and Fellow Member Award Recipient
USE OF RITUXIMAB IN MYASTHENIA GRAVIS: A CASE REPORT
Juliana Varela, Bianca Madeira, Pedro Schestatsky, Pablo Winckler (Porto Alegre, Brazil)

OBJECTIVE: To present the case of a patient with myasthenia gravis (MG) using rituximab.

CASE REPORT: A 26-year-old female patient diagnosed with MG since the age of 10 had frequent relapses and hospital admissions, obesity, depression, diabetes, and hypertension due to longterm corticosteroid use. She had positive serology for acetylcholine receptor antibody, and although she did not have a thymoma a thymectomy was performed but without clinical improvement. During followup, different immunomodulators were tried (azathioprine, mycophenolate, and methotrexate), but none of these medications were either successful or tolerated. After monthly applications of immunoglobulin were started, she achieved a better disease control, but still had relapses with respiratory failure. In June 2015 the use of rituximab was initiated with a dose of 875 mg (375 mg/m2). She was re-evaluated monthly and showed a marked clinical improvement by 4 months.

SUMMARY/CONCLUSION: The use of rituximab in MG has been suggested as an alternative to refractory and severe cases. It has been related to mild infusion adverse effects but no serious adverse reactions, and there is no ideal dose established. This was the first patient in the authors’ center with MG to use rituximab. She had a refractory disease and many attempts to control her disease were made without success, so it was decided to start rituximab. There was no serious infusion or late adverse effects. In the followup evaluations she showed a marked improvement in the myasthenia control and in the depressive symptoms as well.

INSIGHTS: ANALYSIS OF INTRAVENOUS IMMUNOGLOBULIN RESPONSIVENESS IN PATIENTS WITH MYASTHENIA GRAVIS
Todd Levine (Phoenix, AZ), Leslie Vaughan (Temecula, CA), Gary Badger (Burlington, VT), Gil Wolfe (Buffalo, NY), Lara Katzin (Tampa, FL), David Saperstein (Phoenix, AZ), Tahseen Mozaffar (Orange, CA), Richard Barohn (Kansas City, KS), Jonathan Katz (San Francisco, CA), Mazen Dimachkie (Kansas City, KS), Michelle Greer (Temecula, CA), Elissa Ritt (Temecula, NM)

INTRODUCTION: IVIg is commonly used off-label for treatment of myasthenia gravis (MG). Due to a lack of controlled trials, there is tremendous variability in the types of patients who are prescribed IVIg for MG in clinical practice.

OBJECTIVE: Through INSIGHTS, a quality-improvement project, the clinical, laboratory, and electrophysiologic criteria of neuromuscular patients prescribed IVIg were examined.

METHODS: The clinical, laboratory, and electrophysiologic data of 585 neuromuscular patients who were prescribed IVIg were reviewed by a panel of independent, expert neuromuscular neurologists. Outcomes were determined based on quality-of-life measures, the Patient Global Impression of Change, and clinical documentation.

RESULTS: A total of 89 MG patients received IVIg; 44 of these were naïve to IVIg, and 68% of these patients had a positive response. Forty-five patients had received IVIg previously, and 73% of these had a positive outcome. Fifty-nine patients (66%) were receiving ongoing maintenance therapy, while 30 patients (34%) received treatment only for an exacerbation. Positive response was most strongly associated with the reviewers’ determination the patient was appropriate for IVIg based on progressive generalized weakness (92% versus 50%, p=0.06). Patient’s age, sex, distribution of weakness, disease time course, and antibody status had no significant association with response.

SUMMARY/CONCLUSION: The overall response rate of patients with MG to IVIg was 70%. This suggests larger, controlled trials of IVIg in MG should be performed with the potential to receive an indication for therapy both for exacerbations and maintenance.
JITTER AND BLOCKING PRE- AND POST-TREATMENT IN PEDIATRIC GENERALIZED MYASTHENIA GRAVIS
Sumit Verma (Atlanta, GA), Marie Collop (Mt. Berry, GA), Jenny Lin (Atlanta, GA)

INTRODUCTION: Stimulated jitter analysis (stim-JA) of the orbicularis oculi using concentric needle electrode examination is a sensitive and well-tolerated technique to diagnose myasthenia gravis (MG) in children. However, serial stim-JA studies to evaluate MG disease course is not performed in children.

OBJECTIVE: To measure jitter and blocking pre- and post-treatment in pediatric MG.

METHODS: Of the 5 girls (age range: 4-16 years) with seropositive generalized MG followed over 2014-2016, 3 were newly diagnosed and the other 2 were referred to the MG clinic with poorly controlled symptoms. Serial stim-JA studies and clinical examination were performed. Subjects were treated (pyridostigmine 5/5, corticosteroids 5/5, azathioprine 1/5, IVIg 3/5, thymectomy 1/5, plasma exchange 1/5) with the goal of achieving complete remission.

RESULTS: Twelve stim-JA studies were performed without complications or need for sedation. Two-hundred-seven apparent single fiber action potentials (ASFAPs) were recorded. The mean interval between 2 stim-JA studies was 13.6±5.3 months. Jitter measurement (mean consecutive difference: pre-treatment 82±37 µs, post-treatment 34±7 µs; p≤0.03) and percent ASFAPs with blocking (pre-treatment 55±33%, post-treatment 0%; p≤0.01) showed statistically significant improvement. Followup examination recorded improvement in ptosis (80%), ophthalmoplegia (100%), facial (70%), bulbar/respiratory (100%) and proximal muscle strength (75%).

SUMMARY/CONCLUSION: Improvement in jitter and blocking correlated with clinical improvement in treated cases of pediatric MG. The study was limited by the fact that the EDX physician was not blinded and there was a relative small study number. Stim-JA can be used as an objective measure of clinical improvement in pediatric MG.

A CASE OF OCCULT PAPILLARY THYROID CARCINOMA IN MYASTHENIA GRAVIS
Vimala Vajjala, Mansoureh Mamarabadi, Shan (Sarah) Chen, Tomer Davidov (New Brunswick, NJ)

INTRODUCTION: Myasthenia gravis (MG) is an autoimmune disorder affecting the neuromuscular junction characterized by fatigable weakness. It can be associated with thymoma, thyroid disorders such as Graves disease and Hashimoto thyroiditis, whereas extrathymic malignancies especially thyroid cancers are rare. Reported here is a case of MG with papillary thyroid carcinoma without coexistent thymoma.

OBJECTIVE: To present a case of MG associated with occult thyroid cancer and to acknowledge the importance of screening for thyroid cancer in refractory cases.

CASE REPORT: A 27-year-old woman presented with recurrent fatigue, dysarthria, diplopia, ptosis, and dyspnea. Repetitive nerve stimulation showed decrement, and acetylcholine receptors antibodies were elevated. Treatment with pyridostigmine and high-dose prednisone resulted in frequent exacerbations needing IVIg infusions. A CT of the neck revealed a possible ectopic thymoma, and ultrasound of the thyroid showed a large heterogeneous mass lateral to left lobe. A total thyroidectomy was performed and pathology showed multifocal papillary carcinoma with metastases to the lateral neck/jugular lymph nodes. Post procedure, her MG symptoms are controlled with pyridostigmine and low-dose steroids with no exacerbations.

SUMMARY: Thyroid carcinoma is rare in patients with MG but more than in the general population, papillary type being most common. Often, they have early extrathyroidal or lymph nodal metastases and a tendency to be at higher stage with small tumor size. Patients with refractory MG and frequent crises should be screened for thyroid cancer with a neck CT, thyroid ultrasound, or other modalities. Tumor treatment may improve the MG therapy as seen in this patient.
MYASTHENIA GRAVIS ASSOCIATED WITH SMALL-CELL LUNG CANCER: INSIGHT INTO INITIATION OF PARANEOPLASTIC AUTOIMMUNITY?
Anastasia Zekeridou, Guy Griesmann, Vanda Lennon
(Rochester, MN)

INTRODUCTION: Myasthenia gravis (MG) often associates with thymic epithelial neoplasia. Paraneoplastic MG sometimes associates with other cancers including primary lung carcinomas, both small-cell and non–small-cell types. Approximately 50% of small-cell lung carcinomas (SCLCs) express nicotinic acetylcholine receptors (AChRs) of neuronal or muscle type. The mechanism initiating AChR autoimmunity is not understood.

METHODS: The Mayo Clinic Neuroimmunology Laboratory database was retrospectively reviewed to identify patients with serologically- and clinically-confirmed MG in the context of SCLC diagnosis. AChR expression (both mRNA and protein) was investigated in a cancer cell line established from a patient with limited SCLC and MG.

RESULTS: Five patients were identified with clinically- and serologically-confirmed MG and SCLC. The cell line established from 1 of those patients was confirmed to express muscle (α1) AChR by using α1-subunit-selective monoclonal IgGs to immunoprecipitate 125I-α-bungarotoxin-complexed proteins from detergent-solubilized membranes. Reverse transcription polymerase chain reaction identified 8 informative mRNA transcripts: 2 encoded full-length native AChR α1 subunit (1 containing the exon P3A); the other 6 mRNA sequences, products of both in-frame and out-of-frame alternative splicing, encoded potential foreign epitopes.

SUMMARY/CONCLUSION: The mutated mRNA transcripts identified would encode truncated polypeptide products predicted to be immunologically foreign (i.e., "non-self"). If released from necrotic SCLCs and presented by major histocompatibility complex class II molecules on antigen-presenting-cells, these peptides would have potential to activate helper T-cells. This in turn could stimulate AChR autoantibody production by activating self-reactive B-cells recognizing linked AChR self-epitopes on longer peptide fragments, thus initiating clinical MG.
This activity has been planned and implemented in accordance with the accreditation requirements and policies of the Accreditation Council for Continuing Medical Education (ACCME). The AANEM is accredited by the ACCME to provide continuing medical education for physicians.

It is the policy of the AANEM to ensure balance, independence, objectivity and scientific rigor in all of its educational activities. All participating speakers, planning committee members, and authors are required to disclose to the program audience any financial relationships with proprietary entities, producing, marketing, re-selling, or distributing health care goods or services consumed by, or used on, patients that is related to the subject matter of this program. Spouse/partner relationships with commercial interests are also disclosed. Disclosure information was reviewed in advance in order to manage and resolve any possible conflicts of interest. All conflicts of interest have been resolved in accordance with ACCME Standards for Commercial Support℠.

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AN EXTENDED VIEW OF TREATMENT PATTERNS IN CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY

Jeffrey Allen (Minneapolis, MN), Jeffrey Guptill (Durham, NC), M. Chris Runken (Research Triangle Park, NC), Michael Eddy, Orsolya Lunacsek (Palm Harbor, FL)

INTRODUCTION: Real-world treatment patterns in chronic inflammatory demyelinating polyneuropathy (CIDP) are largely unknown.

OBJECTIVE: To gain a better understanding of CIDP treatment patterns.

METHODS: Treatment-naïve patients were identified from the PharMetrics-Plus database with ≥2 CIDP diagnoses ≥90 days apart or ≥1 CIDP diagnosis and a CIDP-related treatment between January 1, 2010-June 30, 2012. Patients were required to have continuous eligibility for medical and pharmacy benefits at least 1 year pre- and 2 years post-initial diagnosis. Patients who received IVIg for reasons other than CIDP were excluded. Steroid, immunotherapy (IM), and IVIg therapy exposure were evaluated during the 2-year followup.

RESULTS: A total of 1233 patients met inclusion criteria (mean age: 54.6 years, mean Charlson comorbidity index: 1.6, 53.4% male). During the study, 48% of patients received steroid therapy only, 13% IVIg only, 2% IM only, 18% IVIg and/or steroid and/or IM, and 5% IM and steroid; 15% received no treatment. Steroid-treated patients, alone or in combination, were younger than patients without steroid use (52.5 versus 58.3 years), while patients receiving no treatment were oldest (58.9 years). Overall, patients were treated within 4.9 months of diagnosis. Patients receiving IVIg and/or steroid and/or IM were treated earliest (1.4 months) compared to IM and steroid (3.4 months), IM only (4.4 months), and steroid only (7.1 months).

SUMMARY/CONCLUSION: In this analysis of treatment-naïve patients with a new International Classification of Diseases (ICD)-9 CIDP diagnosis code, steroids were the most often prescribed therapy, either alone or in combination. IVIg was utilized in only 30% of the population and usually initiated within 2 months of diagnosis.

CME certification was excluded for this abstract and cannot be claimed as part of participation for the poster sessions of the 2016 Annual Meeting.
ETEPLIRSEN, A PHOSPHORODIAMIDATE MORPHOLINO OLIGOMER FOR DUCHENNE MUSCULAR DYSTROPHY: CLINICAL UPDATE AND LONGITUDINAL COMPARISON TO EXTERNAL CONTROL SUBJECTS ON SIX-MINUTE WALK TEST
J R Mendell (Columbus, OH), N Goemans (Leuven, N/A), L Rodino-Klapac, Z Sahenk, LP Lowes (Columbus, OH), L Alfano (Columbus, OH), K Berry (Columbus, OH), P Duda, C Donoghue, F Schnell, J Dworzak, B Wentworth (Cambridge, MA), E Mercuri (Rome, Italy), DMD Italian Network

INTRODUCTION: Duchenne muscular dystrophy (DMD), a rare degenerative X-linked genetic disease, results in progressive muscle loss and premature death, occurring in about 1:3500-5000 males worldwide. DMD is primarily caused by frameshift-causing whole-exon mRNA deletions that prevent production of dystrophin protein.

OBJECTIVE: Eteplirsen, a phosphorodiamidate morpholino oligomer (PMO), is designed to induce production of internally-shortened dystrophin in patients amenable to exon 51-skipping.

METHODS: An analysis of 6-minute walk test (6MWT) performance over 4 years compared boys treated with 30/50 mg/kg eteplirsen weekly IV (n=12) versus a cohort of comparable external control subjects (EC, n=13) defined based on age, corticosteroid use, and genotype.

RESULTS: At year 4, a statistically significant treatment benefit of 162 meters on the 6MWT was observed in eteplirsen-treated patients compared with EC (p=0.0005). Two (17%) eteplirsen patients lost ambulation by year 1 with no additional losses observed, compared with 10 (85%) EC patients by Kaplan-Meier estimate at year 4 (log-rank p=0.011).

SUMMARY/CONCLUSION: After 4 years of eteplirsen treatment, DMD patients had a mean 6MWT that was 162 meters longer (p=0.0005) than the comparable external cohort.

CME certification was excluded for this abstract and cannot be claimed as part of participation for the poster sessions of the 2016 Annual Meeting.

ABOBOTULINUMTOXINA INJECTION PATTERNS IN PATIENTS WITH CERVICAL DYSTONIA FROM THE ANCHOR-CD REGISTRY STUDY
Cynthia Comella (Chicago, IL), Daniel Truong (Fountain Valley, CA), Alberto Espay (Cincinnati, OH), Daniel Snyder, Dominic Marchese (Basking Ridge, NJ), Richard Trosch (Farmington Hills, MI)

INTRODUCTION: ANCHOR-CD is a 1-year, noninterventional study that includes 350 patients with idiopathic cervical dystonia (CD) treated with abobotulinumtoxinA (Dysport®) according to routine practice.

OBJECTIVE: To assess usage patterns of abobotulinumtoxinA from the ANCHOR-CD registry of patients with CD.

METHOD: Botulinum neurotoxin (BoNT-A)-naive and non-naive patients ≥18 years of age with a diagnosis of idiopathic CD were eligible. The muscles injected, dose per muscle, number of sites into the muscle, and use of guidance technique were at the investigator’s discretion. Patients continued in the study for ≤4 treatment cycles.

RESULTS: A total of 347/350 patients from 41 sites completed the study (75% female, mean age: 59.0±13.6 years); 73% were non-naïve to BoNT-A therapy. The most common types of CD treated were mixed postures (68%), and the most frequent single predominant posture was torticollis (25.6%). Median abobotulinumtoxinA dose was 500 U (range: 100-2000 U) over 4 cycles. Median time between retreatment intervals was 14 weeks, with 25% having been re-injected >14 weeks. During each cycle, 4-5 muscles across 11-12 injection points were injected. Almost half of the investigators used needle EMG for injection guidance. The most frequently injected muscles included splenius capitis levator scapulae, trapezius, and sternocleidomastoid. The most common adverse events (AEs) included dysphagia and muscle weakness. Five patients discontinued due to AEs.

CONCLUSION: This study demonstrates the usual clinical practice of BoNT-A injections across multiple sites and demonstrates a consistent approach to abobotulinumtoxinA injection patterns and doses. Use of needle EMG guidance was variable, with less than half of the injections using this technique.

CME certification was excluded for this abstract and cannot be claimed as part of participation for the poster sessions of the 2016 Annual Meeting.
PHASE THREE TRIAL TO EVALUATE ABOBOTULINUMTOXINA (DYSPORT®) INJECTIONS IN CHILDREN WITH UPPER LIMB SPASTICITY DUE TO CEREBRAL PALSY

Mauricio Delgado (Dallas, TX), Ann Tilton (New Orleans, LA), Jorge Carranza (Celaya, Guanajuato), Marcin Bonikowski (Zagorze, Poland), Nigar Dursun (Kocaeli, Turkey), France Catus (Les Ulis Courtaboeuf, France), Philippe Picaut (Les Ulis Courtaboeuf, France)

INTRODUCTION: Although the use of botulinum toxin-A (BoNT-A) to treat upper limb spasticity (ULS) in cerebral palsy (CP) is long established, most clinical trials in CP have been small, of limited duration, and without adequate control.

OBJECTIVE: To assess the efficacy and safety of abobotulinumtoxinA (ABO) at doses of 8 and 16 U/kg versus ABO 2 U/kg (control group) for the treatment of pediatric ULS due to CP (4 treatment cycles over 1 year).

METHODS: Approximately 210 children (2-17 years old) with ULS due to CP (Gross Motor Function Classification System levels I-IV) will be recruited by specialist centers. Patients will have a Modified Ashworth Scale score ≥2 in the primary targeted muscle group (PTMG; elbow or wrist flexors). Patients will be randomized (1:1:1) to injections of ABO 2, 8, or 16 U/kg in cycle 1, using a prespecified injection protocol under electrical stimulation/ultrasound guidance. Patients will perform home exercises (under trained carer supervision) consistent with the chosen treatment goals throughout the study. Control patients (ABO 2 U/kg in cycle 1) will be re-randomized to either ABO 8 or 16 U/kg in subsequent cycles; the double-blind will be maintained. Eligibility to move to the next cycle will be assessed at week 16 of each cycle. The primary endpoint is change from baseline to week 6 of cycle 1 in muscle tone.

RESULTS: Patient recruitment is ongoing.

SUMMARY/CONCLUSION: This will be one of the largest and most robust studies to simultaneously evaluate the effects of BoNT-A on muscle tone, spasticity, and function in pediatric ULS due to CP.

CME certification was excluded for this abstract and cannot be claimed as part of participation for the poster sessions of the 2016 Annual Meeting.

EFFICACY AND SAFETY OF A 2 ML DILUTION OF ABOBOTULINUMTOXINA COMPARED WITH PLACEBO IN ADULT PATIENTS WITH CERVICAL DYSTONIA

Mark Lew (Los Angeles, CA), Daniel Snyder (Basking Ridge, NJ)

INTRODUCTION: Cervical dystonia (CD) is characterized by involuntary cervical muscle contractions leading to sustained and painful head and neck postures. AbobotulinumtoxinA is effectively administered in solutions, up to 1 mL, to affected muscles. Off-label 2 mL dilutions have been reported in the United States, and trials supporting this dilution would support dosing flexibility.

OBJECTIVE: To determine efficacy and safety of a 500 unit, 2 mL dilution of abobotulinumtoxinA (Dysport®) versus placebo in CD patients.

METHODS: In this 12-week, double-blind study, patients were randomized (2:1) to abobotulinumtoxinA or placebo. Toxin-naïve abobotulinumtoxinA patients received 500 units/2 mL in ≥2 affected neck muscles. AbobotulinumtoxinA patients who had previously received botulinum treatment (non-naïve) received 250-500 units/2 mL (2.5:1 abobotulinumtoxinA: previous onabotulinumtoxinA [Botox®] dose) into muscles injected during prior treatments. The primary endpoint was change from baseline to week 4 (W4) in Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS) total score.

RESULTS: A total of 129 patients (abobotulinumtoxinA, n=84; placebo, n=45) completed the W4 primary endpoint evaluation. Versus placebo, abobotulinumtoxinA patients experienced significantly greater changes from baseline in TWSTRS score at W4 (−2.5 versus −10.8, p<0.001; based upon the modified intent to treat population). Adverse events (AEs) occurred in 41% and 22% of abobotulinumtoxinA and placebo patients, respectively. Dysphagia was reported in 9% of treated patients. Other AEs in treated patients were muscle weakness, neck pain, and headache.

SUMMARY/CONCLUSION: This study indicates a 2 mL dilution of abobotulinumtoxinA was significantly more effective than placebo. No unexpected AEs were observed relative to previous studies that used the 1 mL dilution volume.

CME certification was excluded for this abstract and cannot be claimed as part of participation for the poster sessions of the 2016 Annual Meeting.
EFFICACY AND SAFETY OF ABOBOTULINUMTOXINA (DYSPORT®) IN ADULT HEMIPARETIC PATIENTS WITH UPPER LIMB SPASTICITY PREVIOUSLY TREATED WITH BOTULINUM TOXINS
Christina Marciniak (Chicago, IL), Allison Brashear (Winston Salem, NC), Bruce Rubin (Doral, FL), Peter Hedera (Nashville, TN), Stuart Isaacson (Boca Raton, FL), Philippe Picaut (Les Ulis, France), Jean-Michel Gracies (Creteil, France)

INTRODUCTION: Patients with upper limb spasticity (ULS) often require repeat botulinum neurotoxin (BoNT) injections and may have to switch BoNT products during their longerterm management.

OBJECTIVE: To evaluate abobotulinumtoxinA (Dysport®) treatment in adult hemiparetic patients who had previously received other BoNT products for ULS.

METHODS: Of patients enrolled in a double-blind, placebo-controlled study, 105/243 had previously been treated with another BoNT product for ULS. Patients were randomized (1:1:1) to a single injection of abobotulinumtoxinA 500 or 1000 U, or placebo.

RESULTS: Of this patient subgroup (mean age: 52±14 years, 62% male) the ULS etiology was stroke (87%, mean 6 years post-event) or traumatic brain injury (13%, mean 7 years post-event). Most (88%) had previously received onabotulinumtoxinA (mean/max dose 292/800 U), and 16% had previously received incobotulinumtoxinA (mean/max dose 312/1000 U). Of these, 69 were randomized to 500 (n=37) or 1000 U (n=32) of abobotulinumtoxinA. At 4 weeks post-injection, 78% demonstrated ≥1 point improvement in Modified Ashworth Scale scores (versus 25% for placebo), and 80% showed overall clinical improvement (mean improvement of ≥1 grade in physician’s global assessment). The Tardieu scale “angle of catch” improved in finger, elbow, and wrist flexors by 18–46 degrees (versus –1–11 degrees [placebo]) and spasticity angle improved by 15–31 degrees (versus –3–11 degrees [placebo]), resulting in a gain in extension (active range of motion) from 12-19 degrees (versus –1–4 degrees [placebo]). No unexpected safety events were observed.

SUMMARY/CONCLUSION: In this subpopulation of previously-treated hemiparetic adults, abobotulinumtoxinA (500/1000 U) injections improved muscle tone, spasticity, active movement for overall clinical improvement.

CME certification was excluded for this abstract and cannot be claimed as part of participation for the poster sessions of the 2016 Annual Meeting.

ABOBOTULINUMTOXINA (DYSPORT®): DOSES USED TO TREAT UPPER LIMB MUSCLES OF ADULTS WITH SPASTICITY PARTICIPATING IN A PHASE III RANDOMIZED, DOUBLE-BLIND PLACEBO-CONTROLLED STUDY
Peter McAllister (Stamford, CT), Heather Walker (Chapel Hill, NC), Christina Marciniak (Chicago, IL), Steven Edgley (Salt Lake City, UT), Fatuma Gul (Dallas, TX), Bruce Rubin (Doral, FL), David Simpson (New York, NY), Philippe Picaut (Les Ulis, France)

INTRODUCTION: Primary efficacy results of a large Phase III study showed that treatment with abobotulinumtoxinA at doses of 500 or 1000 U injected into upper limb muscles provided tone reduction and clinical benefit in patients with hemiparesis due to stroke or traumatic brain injury.

OBJECTIVE: To report mean doses of abobotulinumtoxinA administered to upper limb muscles.

METHODS: Patients (n=243) received abobotulinumtoxinA 500 or 1000 U or placebo by intramuscular injection into their primary targeted muscle group (PTMG, selected from extrinsic finger flexors, wrist flexors, and elbow flexors) and at least 2 other upper limb muscles, including shoulder muscles.

RESULTS: Mean doses (500 and 1000 U groups, respectively) were as follows: Finger muscles—flexor digitorum profundus: 93.5±17.0 and 195.5±25.9 U; flexor digitorum superficialis: 95.4±14.3 and 196.8±28.4 U; thumb muscles: 76.9±26.8 and 157.0±53.3 U. Wrist muscles—flexor carpi radialis: 92.2±18.1 and 178.1±45.5 U; flexor carpi ulnaris: 89.9±25.7 and 171.2±45.2 U. Elbow muscles—brachioradialis: 88.3±28.5 and 172.1±44.8 U; brachialis: 148.5±60.2 and 321.4±103.2 U; “other” (biceps brachii, pronator teres): 108.6±49.5 and 216.5±92.2 U. Average dosing in shoulder muscles (triceps brachii, pectoralis major, subscapularis, and latissimus dorsi) was 122.2±44.1 and 300.0±129.1 U.

SUMMARY/CONCLUSION: In these hemiparetic patients, mean doses administered were 76.9–196.8 U in the finger muscles, 89.9–178.1 U in the wrist flexors, 88.3–321.4 U for the elbow muscles, and 122.2–300.0 U in the shoulder muscles. Total dose administered (in the PTMG and at least 2 upper limb muscles) was 500 or 1000 U, which was previously shown to improve muscle tone in this patient population. The most common treatment-related adverse event was mild muscle weakness.

CME certification was excluded for this abstract and cannot be claimed as part of participation for the poster sessions of the 2016 Annual Meeting.
SAFETY AND TOLERABILITY OF ABOBOTULINUMTOXINA (DYSPORT®) IN CHILDREN (2-17 YEARS) WITH LOWER LIMB SPASTICITY DUE TO CEREBRAL PALSY: A POOLED ANALYSIS OF EIGHT CLINICAL TRIALS

Ann Tilton (New Orleans, LA), Dennis Matthews (Aurora, CO), Mark Gormley (St Paul, MN), Adnan Mahmood (West Sussex, England), Philippe Picaut (Les Ulis, France), Daniel Snyder (Basking Ridge, NJ), Mauricio Delgado-Ayala (Dallas, TX)

INTRODUCTION: Current American Academy of Neurology (AAN) guidelines consider botulinum toxin-A as a "generally safe" treatment for localized/segmental spasticity in children with cerebral palsy (CP).

OBJECTIVE: To systematically evaluate the safety of abobotulinumtoxinA (Dysport®) treatment in children with lower limb spasticity due to CP.

METHODS: Presented here are pooled adverse event (AE) data from 3 double-blind, randomized, placebo-controlled trials (single injections) and 5 open-label studies of repeated cycles of abobotulinumtoxinA treatment conducted in children with paretic lower limb CP (2-17 years). All subjects received abobotulinumtoxinA injections (up to 30 U/kg and ≤1000 U) into distal (gastrocnemius with/without soleus) and proximal (hamstring, adductors) muscles.

RESULTS: In the single-cycle studies, 57.9% of abobotulinumtoxinA-treated subjects (n=280) reported ≥1 AE versus 47.8% of placebo-treated subjects (n=136). The most common AEs (both groups) were related to childhood infections. Treatment related AEs (TRAEs) were more common with abobotulinumtoxinA versus placebo (11.8% versus 5.9%); the most common was pain in extremity (2.1% of abobotulinumtoxinA-treated subjects). Fewer serious AEs (SAEs) were reported in abobotulinumtoxinA-treated subjects (1.8%) versus placebo (4.4%). There were no treatment-related SAEs. In the open-label studies, where subjects (n=476) received up to 7 injections and were followed for up to 28 months, TRAEs were reported in 21% of treated subjects; the most common being pain in the extremity and muscular weakness. Two SAEs (ataxia due to walking on tiptoes and head injury resulting from a fall) were reported.

SUMMARY/CONCLUSION: These data from >700 children demonstrate a low incidence of TRAEs when abobotulinumtoxinA is injected into the distal and/or proximal leg muscles of children with CP.

CME certification was excluded for this abstract and cannot be claimed as part of participation for the poster sessions of the 2016 Annual Meeting.

COMPARISON OF METHODOLOGY, PATIENT CHARACTERISTICS, AND TREATMENT RESULTS FROM ANCHOR-CD AND OTHER REGISTRY STUDIES OF BOTULINUM TOXIN TYPE A IN CERVICAL DYSTONIA

Cynthia Comella (Chicago, IL), Alberto Espay (Cincinnati, OH), Daniel Synder, Dominic Marchese (Basking Ridge, NJ), Daniel Truong (Fountain Valley, CA)

INTRODUCTION: ANCHOR-CD is a real world, prospective, open-label registry study designed to collect patient responses and health economics data in patients with cervical dystonia (CD) who were treated with abobotulinumtoxinA in the United States.

OBJECTIVE: To compare methodology, patient characteristics, and treatment results from 3 botulinum neurotoxin (BoNT-A) registry studies.

METHOD: The authors examined ANCHOR-CD and 2 other open-label registries: CD-PROBE (onabotulinumtoxinA) and XCiDaBLE (incobotulinumtoxinA). Each evaluated similar efficacy assessments (e.g., Toronto Western Spasmodic Torticollis Rating Scale [TWSTRS], patient and clinical global impression of change [PGIC and CGIC], and CD impact profile [CDIP-58]).

RESULTS: The mean age (59.0±3.6 years), percent of females (75%), and mean age of onset (49±15.6 years) were comparable among registries. More patients in ANCHOR-CD (73%) and XCiDaBLE (77%) had previously received BoNT treatment versus CD-PROBE (36.5%). Change from baseline in TWSTRS total scores were similar in ANCHOR-CD (−12.1, n=304, week 4 of cycle 1) and CD-PROBE (−11.8, n=479, visit 3, >4-6 weeks after second injection). TWSTRS was not assessed in XCiDaBLE. Much improved or very much improved ratings on CGIC were similar in ANCHOR-CD (62.7%; n=316, week 4/cycle 1) and CD-PROBE (61.4%; n=479, visit 2, >4-6 weeks after first injection). Changes in CDIP-58 subscale scores were similar between ANCHOR-CD (cycle 1/day 1-cyle 3/day 1) and XCiDaBLE (CD patients; baseline-cycle 1/week 4), but larger in CD-PROBE (baseline-final visit, >4-6 weeks after cycle 3), possibly due to a higher proportion of BoNT-naive patients.

CONCLUSION: Patient characteristics and response patterns were generally similar across BoNT-A registries, supporting their individual effectiveness in CD.

CME certification was excluded for this abstract and cannot be claimed as part of participation for the poster sessions of the 2016 Annual Meeting.
Abstract Guide

DISCLOSURE APPENDIX (Non-CME Abstracts)

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It is the policy of the AANEM to ensure balance, independence, objectivity and scientific rigor in all of its educational activities. All participating speakers, planning committee members, and authors are required to disclose to the program audience any financial relationships with proprietary entities, producing, marketing, re-selling, or distributing health care goods or services consumed by, or used on, patients that is related to the subject matter of this program. Spouse/partner relationships with commercial interests are also disclosed. Disclosure information was reviewed in advance in order to manage and resolve any possible conflicts of interest. All conflicts of interest have been resolved in accordance with ACCME Standards for Commercial Support℠.

Non-CME Poster Disclosure Information

Continuing medical education (CME) certification was excluded for abstracts number 208-217 and cannot be claimed as part of participation for the poster sessions of the 2016 Annual Meeting. These abstracts will be available to view in a separate, non-CME area of the poster hall. Specific disclosure information for abstract authors of non-CME eligible posters, who had relevant financial relationships to disclose is listed below. All other authors of non-CME eligible abstracts, who are not included below, had no relevant financial relationships to disclose.

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INTRODUCTION: The usual approach of neuromuscular neurologists treating patients with Myasthenia are quite myopic dominated by the spree of looking for monistic somatic aspects of the disease only, with psychological aspects heaping up in the blind spots. Psychological factors such as depression, anxiety, or stress can trigger myasthenia, influence under-diagnosis or over-diagnosis by causing physical fatigue, or influence the overall quality of life. Available data in this field is scattered, not ample, or satisfying.

OBJECTIVE: To study the relatively left-out psychological aspects of Myasthenia gravis.

METHODS: A meta-analysis of Pubmed and Ovid articles published in original or translated English between 1980 and 2016 from five continents sourcing from Australia, Brazil, China, England, Germany, Japan, Serbia, and USA, was done.

RESULTS: Over 2600 patients were studied, highest being from Europe, especially Germany. Important revelations were: mood disorders, present in about a third of the patients, were higher in the late-onset disease forms. Emotional stress was a trigger of myasthenia aggravation especially with older age of onset, lower educational level, more severe and longer duration disease. Depression was a powerful quality of life affecting factor second only to decreased mobility, influenced by the doses of oral corticosteroid. Plasma exchange treatment failed to improve mood in spite of uplifted muscle power and ambulation, suggesting that mood disorders were not solely due to decreased somatic abilities.

SUMMARY/CONCLUSION: More holistic research and management strategies, considering the mind as well, and not just the body only, may take Myasthenia management to higher notches with better outcomes and patient satisfaction.
A PILOT TRIAL OF SUBCUTANEOUS IMMUNOGLOBULIN IN PATIENTS WITH MYASTHENIA GRAVIS EXACERBATION: AN UPDATE ON SAFETY, FEASIBILITY, AND PHARMACOKINETICS
G Beecher, D Anderson, D Blackmore, A Mallon, Z Siddiqi (Edmonton, Alberta)

INTRODUCTION: Subcutaneous immunoglobulin (SCIg) treatment in myasthenia gravis (MG) exacerbation has yet to be formally studied. Presented here are the updated safety, feasibility, and pharmacokinetic results from an ongoing phase III clinical trial assessing the use of Hizentra® (20% SCIg) in MG exacerbation.

OBJECTIVE: To assess the safety, feasibility, and pharmacokinetics of Hizentra® in patients with MG exacerbation.

METHODS: Thirty patients with MG (MGFA Class II and III) will be enrolled in a prospective, open-label, single blind study to receive SCIg at 2g/kg in a flexible dosing regimen over 4 weeks. The primary outcome measure is the change in quantitative MG (QMG) score from baseline to end of study (6 weeks; data presented separately). Patient compliance and incidence of side effects is monitored throughout the study. Laboratory safety parameters, including blood cell counts and differential, creatinine, liver function tests, markers of hemolysis, and IgG levels, are assessed weekly. Standardized assessment of patient satisfaction is performed at end of study.

RESULTS: Twenty-one patients have been enrolled; seventeen have completed the study. All patients required 1 training session for self-infusion. Pump rather than manual infusions resulted in better compliance. There were no major adverse reactions to SCIg, including hemolysis and impaired renal function. Mild infusion site reactions, headache, and flu-like symptoms have been reported, none causing discontinuation of SCIg. Serum IgG levels gradually increased by a mean of 97% by end of study. Standardized patient surveys indicate reasonable satisfaction with SCIg.

SUMMARY/CONCLUSION: Ongoing analysis suggests that SCIg is safe and well tolerated in patients with MG exacerbation.

ALN-CC5, AN INVESTIGATIONAL RNAI THERAPEUTIC FOR THE TREATMENT OF MYASTHENIA GRAVIS: INTERIM PHASE 1 DATA IN HEALTHY VOLUNTEERS AND EFFICACY IN PRE-CLINICAL ANIMAL MODELS OF MG.
A Borodovsky, N Kawahata, H Mclean, A Partisano, J Kim, N Najafian (Cambridge, MA), Linda Kusner, Henry Kaminski (Washington, DC), Jorg Taubel, (London) Jim Bush (Leeds)

INTRODUCTION: Uncontrolled complement activation plays a pivotal role in pathology of several neurological disorders including Myasthenia Gravis (MG) and Neuromyelitis Optica. ALN-CC5 is a subcutaneous (SC) investigational RNAi therapeutic designed to potently silence C5 mRNA.

METHODS: A placebo-controlled, double-blind phase 1 study of SC treatment with ALN-CC5 in NHVs has completed dosing. Primary endpoints are safety and tolerability. Secondary endpoints are pharmacokinetics, serum C5 and complement activity. Pre-clinically, C5 silencing was evaluated in Lewis rat EAMG models. Disease activity measures, MAC, AChR and complement activity levels were studied.

RESULTS: 44 NHVs were randomized (1:3) to placebo or single and multiple ascending SC doses of ALN-CC5. ALN-CC5 was generally well tolerated with multi-dosing. The PD effect was highly potent and durable with 97% knockdown (KD) of serum C5 at day 98 and 94% KD at day 180 after a single 600 mg dose. In a PTMG model, rats pre-treated with a SC C5 siRNA showed improvement in disease score, reduction in MAC deposition and preservation of AChR. At study end, 8/9 controls had severe disease, vs 1/9 treated animals with mild disease. Disease modifying effect was observed at 40% complement activity suppression. Evaluation of therapeutic treatment in active EAMG is ongoing.

SUMMARY/CONCLUSION: ALN-CC5 was generally well tolerated in NHVs to date. The PD effect was highly durable, supporting once monthly and possibly quarterly SC dosing. C5 silencing in EAMG models abrogated complement-mediated damage at the NMJ and blocked development of disease symptoms. These initial results support further investigational evaluation of ALN-CC5 in the treatment of MG.
AUTOANTIBODIES AS PREDICTOR OF MYASTHENIC CRISIS AND CLINICAL PARAMETERS
Y Malik, A Almadani, J Dar (Dubai)

INTRODUCTION: MG is a rare disorder of NMJ with annual incidence of approximately 10-20/million. It is mediated by auto-antibodies against the acetylcholine receptor (AChR-Ab), muscle specific tyrosine kinase (MuSK-Ab) or other striated muscle antibodies.

OBJECTIVE: To assess correlation of auto-antibodies with Myasthenic crisis and other clinical parameters.

METHODS: We reviewed records and > 5 years follow up of 143 patients, who visited Rashid Hospital from 2000-2010. It was a retrospective observational study. We encountered demographic details, clinical features with an intentional concentration on myasthenia crisis.

RESULTS: Amongst 143 patients Females predominated (61.8%) over males (38.2%) with a M:F ratio of 1:1.6. Clinically 16.7% patients had ocular and 83.3% had general myasthenia. 75.5% patients carried AchR-Ab, 7% had MuSK-Ab and 17.6% were seronegative. Seronegative MG was meaningfully associated with earlier onset (p=0.07). Bulbar features predominated in seropositive group (p=0.016). In terms of crisis 67% of AchR-Ab patients, all MuSK-Ab and 33% of seronegative patients underwent crisis. Hence, crisis occurred predominantly in seropositive MG(P<0.05). Higher the Osserman’s class at presentation severer was the crisis (p=0.006). Crisis frequency was two-fold in MuSK-Ab patients than others and they obviated relatively severer crisis, as well. One of our patients expired during crisis, so our mortality rate is 1.6 %.

SUMMARY/CONCLUSION: Patients with antibodies display severer form of disease. Seronegative MG has less incidence of autoimmune co-morbidities and experience myasthenia crisis very rarely. Seropositive MG patients had about 30% more tendency to develop crisis. Poor prognosis is predicted by infection during crisis, respiratory involvement, prolonged ventilator dependency and elderly age.

B CELL TOLERANCE DEFECTS AND ABNORMAL B CELL REPERTOIRE FORMATION IN MYASTHENIA GRAVIS
P Stathopoulos, J V Heiden, A Kumar, S Kleinstein, K O'Connor, R Nowak (New Haven, CT) R Barohn, M Dimachkie (Kansas City, KS)

INTRODUCTION: Myasthenia gravis (MG) is an autoimmune condition in which neurotransmission is impaired by pathogenic autoantibodies targeting acetylcholine receptors or muscle-specific kinase. The mechanisms underlying autoantibody production are not well understood.

OBJECTIVE: We investigated whether the autoimmune mechanisms contributing to MG include compromised B-cell tolerance and distorted B-cell repertoires, both of which have not been explored in MG.

METHODS: Validated assays to assess B-cell tolerance checkpoint fidelity were performed. The frequency, phenotype and repertoire of B-cell subsets were determined using flow cytometry, Sanger and next-generation B-cell sequencing.

RESULTS: Abnormally high frequencies of self-reactive naive B-cells accumulate in MG, implicating a breach in tolerance. Consistent with these tolerance defects was a distorted naïve B-cell repertoire revealed by sequencing. Circulating, antibody-producing plasmablasts were present in MG subjects. B-cell sequencing identified abnormal clonal expansions of both memory B-cells and plasmablasts that may directly contribute to the production of circulating pathogenic autoantibodies.

CONCLUSION: First we demonstrated that the naïve B-cell repertoire in MG is abnormally formed as a consequence of B-cell tolerance checkpoint defects. This represents a fundamental component of autoimmunity contributing to the initiation of this disease, which develops prior to antigen exposure. Secondly, autoantibodies that affect disease may be produced by the high frequency circulating peripheral blood plasmablasts, which are more easily targetable by therapeutics than long-lived autoantibody-producing plasma cells residing in the bone marrow or the thymus. These newly described mechanistic components of MG autoimmunity are of particular importance when considering the durability of MG treatment modalities.
B10 DEFICIENCY IN MYASTHENIA GRAVIS IS NOT ASSOCIATED WITH DEFECTIVE T CELL SUPPRESSION

J Yi, M Russo, J Massey, V Juel, L Hobson-Webb, K Gable, A Dawson, K Balderson, J Guptil (Durham, NC)

INTRODUCTION: B10 cells are interleukin-10 (IL-10) producing B cells that strongly inhibit B- and T-cell inflammatory responses. IL-21 and IL-35 are cytokines proposed to enhance the production of B10 cells.

OBJECTIVE: To test the role of IL-21 and IL-35 in promoting B10 cell generation in MG patients. Additionally, we examined whether B10 cells have defective capacity to suppress T cell activation.

METHODS: To proliferate B10 cells in-vitro, isolated B cells from acetylcholine receptor autoantibody-positive MG patients were stimulated with CpG or LPS and rCD40L in the presence and absence of rIL-21 and rIL-35. Cells were re-stimulated with PMA/ION, and analyzed by polychromatic flow cytometry. To assess B10 function, B10 cells were cultured in a 1:1 ratio with enriched CD4 T-cells in a single well or in a transwell plate that prevents cell-to-cell contact. CD4 T-cells were stained with a violet proliferation dye and the number of divisions after CD3/CD28 stimulation determined the magnitude of B10 cell suppression.

RESULTS: The addition of rIL-21 or rIL-35 did not increase the frequency of B10 cells as previously described in murine studies. B10 cells from MG patients were as effective in suppressing CD4 T-cell proliferation as B10 cells from controls. The transwell experiments suggest T-cell suppression is mediated primarily through soluble IL-10 and not by cell-to-cell contact.

SUMMARY/CONCLUSION: Although B10 cell frequencies are lower in MG patients, their ability to suppress T-cells is not affected. B10 cells are of interest as a potential therapeutic source to inhibit auto-reactive responses and restore immune homeostasis.

BEYOND THE ANTIBODIES: SERA METABOLOMOMIC BIOMARKER SIGNATURES DISCRIMINATE MYASTHENIC AND HEALTHY COHORTS

D Blackmore, N Wang, L Li, Z Siddiqi (Edmonton, Alberta)

INTRODUCTION: Few biomarker studies have assessed the serum metabolome in patients with myasthenia gravis (MG). Recent advancements in metabolomic profiling with high coverage may facilitate description of an enhanced MG biomarker signature.

OBJECTIVE: To construct and compare the serum metabolomic profiles of MG patients and healthy individuals using a novel chemical isotope labeling liquid chromatography mass spectrometry (CIL LC-MS) technique.

METHODS: CIL LC-MS uses different labeling reagents to target chemical-group-based submetabolomes to provide in-depth metabolomic analysis. 12C-dansylation labeling of individual samples and 13C-dansylation labeling of pooled samples from 49 patients with seropositive MG and 50 age/gender matched healthy controls was undertaken. The amine/phenol submetabolome changes among the labeled samples were quantified based on subsequent analysis of the 13C-/12C-labeled mixture by LC-MS.

RESULTS: On average, 4084 ± 149 (n = 49) and 3972 ± 492 (n = 50) metabolites were detected in sera from MG samples and controls, respectively - a total of 5711 metabolites in all samples. Orthogonal partial least squares discriminant analysis showed a clear separation of two groups (R2 = 0.98, Q2 = 0.80). The Receiver Operating Characteristic (ROC) curve using 7 metabolites produced an Area Under the Curve (AUC) value of 0.859 (0.806–0.920 at the 95% confidence interval) with 91% specificity and 70% sensitivity.

CONCLUSIONS: High-coverage metabolomic profiling reveals that the serum metabolome of MG patients differs considerably from healthy controls substantiating the probability of finding metabolic biomarkers specific to MG.
BLINDED RETROSPECTIVE ANALYSIS OF RITUXIMAB TREATMENT IN ANTI-MUSK MYASTHENIA GRAVIS

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INTRODUCTION: Anti-muscle specific kinase (MuSK) antibody myasthenia gravis (MG) often has a more severe phenotype with earlier involvement of bulbar and respiratory musculature than other forms of MG. Although plasma exchange and high dose prednisone (1 – 1.5mg/kg daily) are effective, many patients relapse with prednisone taper, exposing them to adverse events and monetary costs. Rituximab, an anti-CD-20 monoclonal antibody, is emerging as an effective treatment for MuSK MG based on evidence from case series and clinical experience. Rarity of MuSK MG is a barrier to developing randomized prospective trials; innovative trial design is needed. We performed a “blinded, retrospective review” of rituximab in MuSK MG that included a randomization step to mimic prospective trial enrollment.

OBJECTIVE: Rigorously evaluate rituximab as treatment for MuSK MG.

METHODS: Multi-center, blinded, retrospective, cohort study of MuSK MG patients treated and not treated with rituximab. Patients included for analysis if 5-member neuromuscular expert panel agreed it would have been reasonable to enroll them in a trial of rituximab vs. placebo. Panel decision based on review of the first year of available clinical data. Primary outcome measure is attainment of an MGFA PIS status of minimal manifestations, pharmacological remission, or stable remission. Secondary outcomes include dose of immune treatments and number of hospital admissions.

RESULTS: Over 100 patients have been identified for expert panel review. Data review is ongoing.

SUMMARY/CONCLUSION: This study will provide the largest comparative data set of MuSK MG patients treated with rituximab vs. those treated with other immune based therapies. Final study results will be presented.

CHARACTERIZATION OF SERONEGATIVE MYASTHENIA GRAVIS PATIENTS’ NEED FOR AGGRESSIVE THERAPEUTIC INTERVENTION AND THEIR RESULTING ADVERSE EVENTS

G Small (Pittsburgh, PA), S Rana (Sewickley, PA)

INTRODUCTION: Serological confirmation for myasthenia gravis (MG) is lacking in many patients with fluctuating weakness in whom electrophysiological evidence supports the diagnosis of MG, and in whom pyridostigmine and immunosuppressant therapies improve muscle strength. Discovery of additional serological markers such as low-density lipoprotein receptor related protein-4 (LRP4) antibodies, and others, in seronegative populations may allow for more confidence in a diagnosis of MG, and more targeted therapies.

OBJECTIVE: We retrospectively reviewed our MG patients’ database to characterize clinically our seronegative population. Our MG clinic serves 250 patients in an area of several thousand square miles in the eastern Ohio, Southwestern Pennsylvania, and West Virginia areas.

METHODS: 60 patients tested negative for acetylcholine receptor (ACHR) binding, blocking, modulating and muscle specific kinase (MuSK) antibodies, and 16 of these 60 tested positive either by repetitive stimulation (RNS) or single fiber electromyography (SFEMG) for a post-junctional neuromuscular junction disorder. No other neurological explanation for these patients’ symptoms was identified. 11 of the 16 patients suffer a generalized MG syndrome severe enough to warrant repeated plasma exchanges (PE) or intravenous immunoglobulin (IVIG) therapies necessitating multiple hospitalizations, and in many cases, life-threatening treatment complications of thrombosis, bleeding, and sepsis. 9 of these 11 patients have been hospitalized 2 or more times in the last 4 years, resulting in millions of dollars in health care costs.

RESULTS:

SUMMARY/CONCLUSION: Although the authors are confident in their therapeutic approach to these patients, MG serological verification may allow more specific treatment protocols, fewer medical adverse events, and less suffering.
CLINICODEMOGRAPHIC PREDICTORS OF IVIG MAINTENANCE THERAPY IN MYASTHENICS
S Johnson, Raghav Govindarajan (Columbia, MO)

BACKGROUND: The standard therapy for myasthenia gravis includes steroids and immunosuppressants. IVIg has been used for the treatment of exacerbations although multiple studies have demonstrated its efficacy as maintenance therapy either in isolation or in combination with steroids. The objective of this study is to identify clinicodemographic predictors of switching from oral steroids to IVIg maintenance therapy.

METHODS: This is a retrospective chart review of all newly diagnosed myasthenics from 2014-2016 who were transitioned from oral steroids to IVIg maintenance. Descriptive statistics was used to collect clinical and demographic data. Paired student t-tests were used to identify predictors of steroid discontinuation or inadequate dosing. A p value \( \leq 0.05 \) was considered statistically significant.

RESULTS: 14 patients (8 females, 6 males) age range 16-70 (mean=36), MGFA grade 2-3 were included. 10 patients were on daily prednisone, 4 were on weekend dosing. Patients were on steroids for range 1-3 months (median=2 months), average dose 30mg daily (median=20). In 8 patients steroids were completely discontinued and placed on IVIg and in 6 IVIg was added to steroids (due to inadequate dosing). Psychosis resulted in discontinuation of steroids (p<0.05) within 6 weeks of starting treatment in 4 patients and inadequate dosing in 6. Weight gain and body image disturbance was seen in 4 patients (all females) resulting in complete discontinuation steroids in all.

CONCLUSION: Psychosis and weight gain (causing body image disturbance) are common reasons for steroid discontinuation or inadequate dosing and predict switch to IVIg maintenance therapy.

COMORBID AUTOIMMUNE DISEASES IN PATIENTS WITH MYASTHENIA GRAVIS
J Farias, B Harvey, I Katzin, A Hart, C Jones, N Tucker, S Dang, C Gooch, T Vu (Tampa, FL)

INTRODUCTION: Myasthenia Gravis (MG) is an autoimmune condition affecting neuromuscular transmission, resulting in ocular, bulbar, and limb weakness.

OBJECTIVE: The objective of this study was to determine the prevalence of comorbid autoimmune conditions in patients with MG.

METHODS: We reviewed medical records of patients with the diagnosis of MG referred to or seen in our academic tertiary care center from January 1, 2013 through May 1, 2016 to identify other autoimmune disorders. De-identified data was collected, coded, and analyzed using SPSS 23. Chi-Square testing was used to analyze for correlations between discrete variables.

RESULTS: Of the 286 charts reviewed, 262 (91.6%) patients had a confirmed diagnosis of MG. Of these patients, 63 (24%) had another autoimmune condition, and 13 (5%) had two or more coexisting autoimmune diseases. These comorbidities included asthma (n=19; 7.3%), systemic lupus erythematosus (n=9; 3.4%), rheumatoid arthritis (n=7; 2.7%), Sjogren’s disease (n=7; 2.7%), chronic inflammatory demyelinating polyneuropathy (n=6; 2.3%) and psoriasis/psoriatic arthritis (n=6; 2.3%). Acetylcholine receptor (AchR) antibody negative patients were significantly (\( \chi^2=0.002 \)) more likely than AchR antibody positive patients to have a secondary autoimmune condition, particularly lupus (\( \chi^2=0.036 \)). There was no correlation (\( \chi^2=1.40 \)) between distribution of MG weakness and presence of another autoimmune condition.

SUMMARY/CONCLUSION: In our study, 29% of myasthenic patients had one or more autoimmune comorbidities. We propose that MG patients should be screened for other autoimmune conditions. However, additional analysis is needed to determine whether treatment of MG impacts autoimmune comorbidities.
CONCENTRIC‐NEEDLE SINGLE‐FIBER ELECTROMYOGRAPHY IN PATIENTS WITH CONGENITAL MYOPATHIES AND SECONDARY NEUROMUSCULAR JUNCTION DEFECTS
JD Domingues, (Jundiai, Sao Paulo) T Rosa, I Faber, CR Martins, Jr, M Martins, M Carneiro, A Martinez, A Nucci, M Franca, Jr. (Campinas, Sao Paulo)

INTRODUCTION: Congenital myopathies (CM) may have clinical fatigability by dysfunction of the neuromuscular junction (NMJ). The evaluation of this dysfunction may have therapeutic implications.
Objective: To describe the single fiber electromyography (SFEMG) in 3 patients with CM and the relevant therapeutic results.

METHODS: Patients underwent SFEMG with stimulated technique in the frontalis muscle (FR); minimum of 90 records, at different points. Data was compared to the reference values by Stalberg, Kouyoumodjian 2016. Jitter analysis from voluntary technique in extensor digitorum communis was excluded because technical artifacts (myogenic early recruitment). Case # 1, girl, 9 years, with centronuclear myopathy (CNM), periods of intense fatigue and apparent aggravation of myopathy. Case # 2: Man, 37, with (CNM). Case # 3: woman, 37 years, congenital fiber-type disproportion.

RESULTS: All patients had evidence of dysfunction of NMJ by increased jitter and / or transmission block (TB). The average Mean Consecutive Difference (MCD) jitter values in FR were 50.9 uS, 38 μs and 161 μs, respectively to # 1,2,3. The percentage of the TB was 16.2 (# 1), 6.3 (# 2) and 23.3 (# 3). Pyridostigmine was started to # 1, # 2, # 3 and # 3 subsequently to salbutamol with partial improvement of fatigue.

SUMMARY/CONCLUSION: Patients with CM may have fatigability caused by secondary NMJ defects, detectable by SFEMG. Relevant drugs can reduce these symptoms, and open a therapeutic window with partial improvement of the motor performance.

DOES CHANGE IN NEUROMUSCULAR JITTER PREDICT OR CORRELATE WITH CLINICAL CHANGE IN MG?
D Sanders, J Massey (Durham, NC)

INTRODUCTION: Neuromuscular jitter has been recommended as a biomarker for monitoring disease severity in myasthenia gravis (MG).

OBJECTIVE: To determine how well various jitter parameters reflect change in MG severity.

METHODS: We reviewed data from MG patients with at least two jitter measurements in the extensor digitorum (ED) (n=227) or frontalis (n=51) muscle. Jitter was measured with SFEMG electrodes during voluntary activation. Results for each test were reported as: mean jitter (calculated as Mean value of Consecutive Differences - MCD) of all measured action potential (AP) pairs; percentage of AP pairs with blocks (% blocks); and percentage of AP pairs with normal jitter (% normals). Change in disease severity was rated by the treating physician. Positive and negative predictive values (PPV & NPV) for improvement were calculated for change in mean MCD, % blocks and % normals between studies. Receiver operator characteristic curves were calculated to determine the optimum discriminant value predicting improvement.

RESULTS: A 14% decrease or 9 μsec fall in mean MCD in the ED had a 90% PPV and an 80% NPV for improvement. Jitter change in the frontalis was slightly less predictive of improvement. Change in mean MCD was a slightly better predictor of improvement than % normals or % blocks.

SUMMARY/CONCLUSION: Jitter is a sensitive measure of severity in MG and has a potential role as a biomarker in clinical trials and in the clinic. Absolute or percentage change in the mean jitter in the ED seems to be the best jitter parameter to follow.
ECULIZUMAB SAFETY AND EFFICACY IN REFRACTORY MYASTHENIA GRAVIS: A PHASE 3 RANDOMIZED, DOUBLE-BLEND, PLACEBO-CONTROLLED, MULTI-CENTER STUDY

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INTRODUCTION: Eculizumab, a humanized monoclonal antibody that inhibits terminal complement, may protect the neuromuscular junction from the destructive effects of antibody-mediated complement activation.

METHODS: AChR+ patients ≥18 years of age with generalized MG (Myasthenia Gravis Foundation of America Class II-IV), an MG-ADL screening/baseline score of ≥6 and inadequate response to immunosuppressive therapies were evaluated and randomized 1:1 to receive intravenous eculizumab infusion for 26 weeks (900 mg/week for 4 weeks, followed by 1200 mg/every 2 weeks) or placebo. Rescue therapy was permitted with physician discretion. The primary efficacy endpoint was change from baseline to week 26 in MG-ADL total score, as analyzed by worst rank. Pre-specified analyses are presented. Safety was also evaluated.

RESULTS: 63 placebo and 62 eculizumab patients were randomized. The primary endpoint did not achieve statistical significance (p=0.0698). Key prespecified secondary endpoints: change from baseline at week 26 in MG-ADL total score, as analyzed by worst rank. Pre-specified analyses are presented. Safety was also evaluated.

RESULTS: 63 placebo and 62 eculizumab patients were randomized. The primary endpoint did not achieve statistical significance (p=0.0698). Key prespecified secondary endpoints: change from baseline at week 26, as analyzed by worst rank in QMG score and responder analyses for the MG-ADL and QMG achieved p-values < 0.05 with the differences favoring patients treated with eculizumab over placebo. 3 of 4 MG-ADL and 4 of 4 QMG prospectively-defined sensitivity analyses, achieved p-values < 0.05 favoring patients treated with eculizumab over placebo. The most common adverse events across both groups were headache (17.6%), upper respiratory tract infection (17.6%) and nasopharyngitis (15.2%).

DISCUSSION: Although the primary endpoint narrowly missed statistical significance, the totality of data supports clinically meaningful improvements for patients treated with eculizumab compared to placebo. Eculizumab was well-tolerated.

This study (NCT01997229) was sponsored by Alexion Pharmaceuticals (New Haven, CT, USA).

FEAR OF FALLING IN THE MYASTHENIA GRAVIS POPULATION

J Naumes (Portland, OR), C Hafer-Macko (Baltimore, MD)

INTRODUCTION: To our knowledge, there are only six case reports on elderly people with myasthenia gravis (MG) who had recurrent falls. No published articles examine fall prevalence in the MG population across the lifespan. However, clinical experience and MG support group discussions indicate that fear of falling is a limiting factor for many.

OBJECTIVE: To determine the prevalence of falls in patients with MG and which activities are most associated with fear of falling.

METHODS: The International Falls Efficacy (IFE) questionnaire was administered to patients during MG clinic follow-up. MG severity was characterized by the MG Quality of Life 15, MG Activities of Daily Living, and Quantitative MG instruments. Social engagement and fatigue were also assessed. After IRB approval, researchers will distribute an anonymous, web-based survey via the MG Foundation of America (MGFA) website and online MG support groups on Facebook to examine the broader MG population.

RESULTS: Participants ranged from 25 to 73 years, QMG score of 1 to 27, and MG-QOL 15 score of 6 to 57. Preliminary data demonstrated a concern with fear of falling in the MG population. Thus far, all participants (n=10) reported fall concern, with 10% low, 60% moderate, and 30% high fall concern according to the IFE ratings. Even participants with well-controlled MG symptoms, classified as in pharmacological remission, still demonstrated low to moderate fear of falling.

SUMMARY/CONCLUSION: Fear of falling is prevalent in the MG population. Additional research is required to understand how fear of falling impacts engagement, psychosocial health, and best strategies to counter this fear.
IMPACT OF EXERCISE ON FUNCTION IN THE MYASTHENIA GRAVIS POPULATION
C Hafer-Macko, R Macko (Baltimore, MD), J Naumes (Portland, OR)

INTRODUCTION: Few studies have been conducted on the safety and effectiveness of exercise for individuals with myasthenia gravis (MG). Many healthcare practitioners state that exercise is contraindicated for the MG population. However, without exercise, physiological capabilities diminish and fatigue increases, further limiting ability to engage in meaningful occupations.

OBJECTIVE: To determine if a three-month exercise program will improve function and fitness in people with stable MG. Aims were to determine whether exercise is safe/feasible and improves functional performance, fitness, and energy cost of walking.

METHODS: Individuals (n=9; mean age 63) with stable, mild to moderate MG engaged in one hour of exercise three times a week for three months. Exercise consisted of aerobic walking, strength training, and breathing exercises. MG activities of daily living (MG ADL), Quantitative MG (QMG), MG quality of life (MG QOL-15), vital capacity, self-selected walking speed, 1-repetition maximum (1-RM), 6-minute walk, and Timed get Up and Go (TUG) instruments assessed functional performance.

RESULTS: Data demonstrates that MG is characterized by low fitness reserve, slow gait, and limited endurance while walking. Significant changes were TUG (-14%, p < 0.01), 1-RM leg press (15%, p < 0.02), peak walking speed (37%, p = 0.1), and peak ventilatory exchange (19%, p < 0.02). Exercise did not have an impact on MG ADL, QMG, and MG QOL-15.

SUMMARY/CONCLUSION: Engagement in exercise can be safe for the MG population if done purposefully and under the supervision of a medical professional. Improved strength, walking function, exercise capacity, and efficiency is possible and can greatly improve functional status.

INSIGHTS: ANALYSIS OF IVIG RESPONSIVENESS IN PATIENTS WITH MYASTHENIA GRAVIS
T Levine, D Saperstein (Phoenix, AZ), L Vaughan, M Greer, E Ritt (Temecula, CA), G Wolfe (Buffalo, NY), L Katzin (Tampa, FL), T Mozaffar (Orange, CA), R Barohn, M Dimachkie (Kansas City, KS), J Katz (San Francisco, CA)

INTRODUCTION: IVIG is commonly used as an off-label treatment of exacerbations and for maintenance therapy in myasthenia gravis. Due to a lack of controlled trials, there is tremendous variability in the type of patients who are prescribed IVIG for myasthenia gravis in clinical practice.

OBJECTIVE: As part of INSIGHTS, a quality-improvement project, we have examined the clinical, laboratory, and electrophysiologic criteria of neuromuscular patients prescribed IVIG across the US.

METHODS: We collected clinical, laboratory, and electrophysiologic data on 585 neuromuscular patients who were prescribed IVIG across the country. A panel of independent, expert neuromuscular neurologists reviewed the information. Positive outcomes were independently determined based on Quality-of-Life measures, Patient Global Impression of Change, and clinical documentation.

RESULTS: We present data from 89 MG patients who received IVIG. 44 of these patients were naïve to IVIG and 68% of these patients had a positive response. Of the 45 patients who had received IVIG previously, 73% had a positive outcome. 59 patients (66%) were receiving ongoing maintenance therapy, while 30 patients (34%) received treatment only for an exacerbation. Positive response was most strongly associated with the reviewers’ determination the patient was appropriate for IVIG (92% vs 50%, p=.06) based on progressive, generalized weakness. Other clinical features had no significant association with response.

SUMMARY/CONCLUSION: Overall response rate of patients with MG to IVIG was 70%. This suggests larger, controlled trials of IVIG in MG should be performed with the potential for an indication for both for exacerbations and maintenance. Updated data will be presented at the time of the meeting.
INTRODUCTION: The rarity of myasthenia gravis (MG) and the long time it takes for many immunosuppressive medications to work makes randomized controlled trials (RCTs) logistically difficult. Many current MG treatments are based on uncontrolled studies, which have a high risk of bias. Even good RCTs have limited generalizability and do not address the clinical heterogeneity of MG.

OBJECTIVE: To develop formal consensus-based guidance for the management of MG.

METHODS: In October 2013, the Myasthenia Gravis Foundation of America appointed a Task Force to develop treatment guidance for MG. A panel of 15 international experts was convened. As the first step, definitions were developed for: goals of treatment, minimal manifestations, remission, ocular MG, impending crisis, and refractory MG. An in-person meeting of the panel determined 7 treatment topics to be addressed. The RAND/UCLA appropriateness methodology was used to develop consensus guidance statements. Initial statements were developed for each topic from the literature. Up to three rounds of anonymous voting with modifications of the guidance statements based on panel input were used to attain consensus.

RESULTS: Guidance statements were developed for: symptomatic and immunosuppressive treatments, intravenous immunoglobulin and plasma exchange, management of impending and manifest myasthenic crisis, thymectomy, juvenile MG, MG associated with antibodies to muscle specific tyrosine kinase and MG in pregnancy.

SUMMARY/CONCLUSION: Given the insufficiency of studies to guide treatment in MG, this formal consensus of international MG experts is presented as a supplement to evidence-based guidelines, to assist clinicians caring for MG patients worldwide.

LRP4 ANTIBODIES IN MYASTHENIA GRAVIS
R Roda (Bethesda, MD), D Drachman, A Hoke (Baltimore, MD)

INTRODUCTION: Myasthenia gravis is an autoimmune condition caused by the presence of antibodies against components of the neuromuscular junction. Approximately 90% of patients have antibodies against the acetylcholine receptor (ACHR) or muscle specific kinase (MuSK). Recently antibodies against Lrp4, an essential component in the development and maintenance of the neuromuscular junction, have been detected in patients with myasthenia, suggesting that it could be a new antigenic target.

OBJECTIVE: We investigated the feasibility of developing an ELISA assay to detect Lrp4 antibodies, and tested samples from patients with myasthenia gravis that had previously tested negative to antibodies against the acetylcholine receptor.

METHODS: We tested a group of normal controls as well as patients with myasthenia that had tested negative to antibodies against the acetylcholine receptor.

RESULTS: Preliminary data analysis indicates that 8.2% of AChR negative myasthenia gravis patients tested positive for the presence of Lrp4 antibodies.

SUMMARY/CONCLUSION: Antibodies against Lrp4 can be detected using the ELISA we have developed.
MICRO-RNA AND M-RNA PROFILES ASSOCIATED WITH ECTOPIC GERMINAL CENTER FORMATION IN THYMUS SAMPLES OF PATIENTS WITH MYASTHENIA GRAVIS

M Sengupta, BD Wing, N Lee, L Kusner, H Kaminski (Washington, DC), G Cutter (Birmingham, AL)

INTRODUCTION: A characteristic pathology of early onset MG is thymic hyperplasia with ectopic germinal centers (GC). However, mechanisms that trigger and maintain thymic hyperplasia are poorly characterized. Dysregulation of micro-RNAs (miRNA) has been identified in the pathology of several autoimmune diseases.

OBJECTIVE: In this study we accessed the miRNA and mRNA profile of MG thymus.

METHODS: Thymus samples of patients were assessed by histology and grouped based on appearance of GC. MiRNA and mRNA were evaluated using GeneChip® miRNA 4.0 Array and GeneChip® Human Transcriptome Array 2.0, respectively. Partek Genomic Suite 6.6 and Transcript Analysis Console 2.0 programs were used for further analysis.

RESULTS: Thirty-four mature miRNA and forty eight annotated mRNA transcripts were identified that were differentially expressed between the two groups with greater than 1.5 fold difference in expression (ANOVA p<0.05). The cellular and molecular functions of the mRNAs involve cell death and cell survival, cellular proliferation, cytokine signaling and extra cellular matrix reorganization. The miRNAs identified are involved in cancer pathways. We identified 7 mi-riRNA and mRNA pairs that are reciprocally regulated. Regulator of G protein Signaling 13 (RGS13), which is known to be involved in GC regulation was identified to be associated with specimens having increased GC and was paired with downregulation of miR-452-5p and miR-139-3p.

SUMMARY/CONCLUSION: Our study shows that there is a distinct mRNA and miRNA expression pattern in the thymus with ectopic GC with expression of GC specific protein such as RGS13. Also maintenance of autoimmunity is supported by regulatory pathways known to be involved in anti-apoptotic pathway.

MYASTHENIA GRAVIS AN UNCOMMON STROKE MIMICKER IN THE EMERGENCY ROOM

V Levasseur, R Govindarajan (Columbia, MO)

BACKGROUND: It's not uncommon for MG to present with episodic or even with new onset symptoms to ER thus mimicking stroke. Herein we report 4 cases that were worked up as stroke in ER/hospital later on to be diagnosed with MG.

CASE REPORTS: 35 year old woman presented to ER with slurred speech/dysphagia. She had normal MRI brain/CT angiograms and was discharged on aspirin. Two weeks later she had similar symptoms and aspirin changed to clopidogrel. Month later she had similar symptoms and was on Coumadin. Clinic work up showed elevated Ach receptor antibody with decrement on repetitive stimulation. Coumadin was stopped.

40 year old singer presented with slurred speech following concert to the ER. She had normal MRI brain/CT angiogram and was discharged on aspirin and statin. Clinic work up showed tongue atrophy, elevated Musk antibody and decrement on repetitive stimulation.

50 year old woman presented with sudden onset right eye ptosis and binocular double vision with mild right facial droop to ER. She had normal MRI brain/CT angiogram. Clinic work up showed fatigable ptosis and positive Cogan sign. She is on pyridostigmine.

75 year old man with hypertension presented with slurred speech. He had normal CT angiogram/MRI brain. He was on aspirin and clopidogrel. When he had similar symptoms a month later Coumadin and statin were added. Clinic work up showed elevated Ach receptor antibodies and decrement on repetitive stimulation. Coumadin was stopped.

CONCLUSION: Acute or new onset time bulbar/ocular symptoms of MG can be an uncommon stroke mimicker.
PSYCHOMETRIC EVALUATION OF MG MANUAL MUSCLE TESTING (MG-MMT), QUANTITATIVE MYASTHENIA GRAVIS (QMG) AND MG COMPOSITE (MGC) IN PATIENTS WITH MUSK-ANTIBODY POSITIVE MG USING RASCH ANALYSIS.  
R Sadjadi (Boston, MA), D Sanders, R Mahmood, J Guptil (Durham, NC), T Burns (Charlottesville, VA)

INTRODUCTION: Rasch analysis compares psychometric properties of individual test items and assigns category responses in a scoring system to determine which items are the most informative.

Objective: To determine whether outcome measures currently validated for acetylcholine receptor antibody (AChR) MG are appropriate for MuSK MG patients.

METHODS: We performed Rasch analysis on 321 MG-MMT, 157 QMG and 92 MGC scores using data from the Duke MG Registry database. Winsteps software was used to explore data for targeting, stability, fit statistics, dimensionality and category response functioning and made comparisons with previously presented measures for AChR MG patients.

RESULTS: There was a significant floor effect comparing relative distribution of item and person location estimates, mostly secondary to a large paucisymptomatic or asymptomatic patient population. Lower extremity items were more sensitive to changes in more disabled patients while oculobulbar items were more appropriate for less severely affected patients. Item redundancy was noted between left and right sided items in the MG-MMT. Category response thresholds were reasonably organized and ordered with minor exceptions in the MG-MMT.

SUMMARY/CONCLUSION: All three disease severity outcomes had relatively reliable psychometrics. QMG seemed to have a better sensitivity to change in patients with less severe disease and therefore was psychometrically more stable than MGC and MG-MMT in this patient population. This is a work in progress and additional data from a more symptomatic patient population would help better understand the psychometric properties of these tests.

QUANTITATIVE CLINICAL ASSESSMENT OF MYASTHENIA GRAVIS EXACERBATION
S Alqadri, R Govindarajan (Columbia, MO)

INTRODUCTION: Delay in diagnosing myasthenia gravis (MG) exacerbation can result in significant morbidity and rarely mortality. While there are no definite diagnostic tests for MG exacerbation, we assessed 3 clinical tests: eye closure strength (ECS), neck flexion strength (NFS) and single breath count test (SBCT) as an easy to administer, quantitative clinical biomarkers.

METHODS: This is a prospective, non-blinded, pilot study of all consecutive patients who presented with symptoms of MG exacerbation from January to December 2015. The diagnosis of MG exacerbation was at the discretion of treating neuromuscular physician. ECS was graded: grade 4- eyelashes dug in, grade 3- eyelids can be opened but with some resistance, grade 2-eye lids can easily be opened, grade 1-cannot completely close the eye. NFS was assessed in supine position and graded according to MRC. SBCT was done as per previously defined protocols.

RESULTS: 20 patients (8 males, 12 females, age: 16-70 years, 8 seropositive) included. 8 were on IVIg maintenance therapy, one on IVIg and steroids and rest were on steroids. ECS grade 2 or less, NFS 3 or less and SBCT<25 had significant association with myasthenia exacerbation (p<0.05). There was strong correlation between the three clinically assessed parameters (r=0.55, p<0.05) at above values. In three cases ECS 2 was seen with normal NFS/ SBCT. 3 days later patients came back with worsening shortness of breath when the SBCT was below 20 and NFS was 2.

CONCLUSION: Quantitative clinical biomarkers are good predictors of MG exacerbation and eye closure strength may be more sensitive.
SERUM PROTEOMIC PROFILING OF MYASTHENIA GRAVIS FOR THE IDENTIFICATION OF CLINICALLY POTENTIAL BIOMARKERS
F Hussain, RS Piragasam, D Blackmore, R Fahlman, Z Siddiqi (Edmonton, Alberta)

INTRODUCTION: Myasthenia gravis (MG) is an antibody-mediated autoimmune disorder that affects the neuromuscular junction. Variable clinical course and similarities in signs and symptoms with other neuromuscular disorders, post challenges in the diagnosis. Furthermore, no biologic marker is currently available to predict the course of the disease.

OBJECTIVE: This study is aimed to establish serum proteomic profiles to identify and validate potential biomarker(s) for MG.

METHODS: For the initial phase of identification, an unbiased shotgun proteomic approach has been adopted using high-resolution quadrupole-Orbitrap mass spectrometry (MS) coupled with nano- Liquid chromatography (nLC). Three different groups of serum samples are being analyzed: MG, control group and a reference disease (Rheumatoid Arthritis).

RESULTS: Twelve samples (four from each subject group) have been analyzed in the initial pilot study. Collectively 28 proteins exhibit significant variations among the different groups (confidence interval 95%).

SUMMARY/CONCLUSION: These are being used to establish the outline of a predictive model. The proteomic profiling of serum may be an effective screening tool for discovery of a novel MG biomarker.

SLEEP DISORDERS IN PATIENTS WITH MYASTHENIA GRAVIS
J Farias, B Harvey, L Katzin, A, Hart, C Jones, N Tucker, S Dang, C Gooch, T Vu (Tampa, FL)

INTRODUCTION: Myasthenia Gravis (MG) is an autoimmune disorder associated with antibodies to the acetylcholine receptor (AchR). Patients with MG may have respiratory and oropharyngeal muscle weakness and fatigue, which may predispose them to sleep disorders.

OBJECTIVE: The objective of this study was to determine the prevalence of comorbid sleep disorders in patients with MG.

Methods: We reviewed medical records of patients labeled as having MG who were seen in our academic neuromuscular clinic from January 1, 2013 through May 1, 2016 to determine the incidence of sleep comorbidities. De-identified data was collected, coded, and analyzed using SPSS 23. Chi-Square testing was used to analyze for correlations between discrete variables.

RESULTS: Of 286 charts reviewed, 262 (91.6%) patients had an established diagnosis of MG. Of these patients, 55 (21.0%) had a diagnosed sleep disorder. Eight patients (3.1%) had two or more sleep disorders. Sleep disorders in patients with MG included obstructive sleep apnea (n=47; 17.9%), restless leg syndrome (n=7; 2.7%), insomnia (n=6; 2.3%), and narcolepsy (n=4; 1.5%). AchR-negative patients were significantly ($\chi^2 = 0.039$) more likely than AchR-positive patients to have a comorbid sleep condition.

SUMMARY/CONCLUSION: A high percentage of patients with MG had comorbid sleep conditions. Providers should screen for concurrent sleep disorders, particularly sleep apnea, which may contribute to fatigue in myasthenic patients. A longitudinal study is needed to determine whether treatment of sleep disorders can reduce the incidence of MG exacerbations.
SURVEY OF LONG TERM OUTCOMES IN JUVENILE MYASTHENIA GRAVIS
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INTRODUCTION: Juvenile onset autoimmune myasthenia gravis (JMG) shares a common mechanism to that of adult onset myasthenia, but little is known about the long-term outcome of children with JMG, and much is extrapolated from experience in adults.

OBJECTIVE: We hypothesize that subjects with JMG, despite manifesting disease across a wide range of expression, nonetheless achieve a good functional status.

METHODS: Target a population of children with JMG, age 21 or less at time of initial visit, seen in a major academic referral neuromuscular practice between 1987 and review of existing medical records with follow up contact of subjects or their families.

RESULTS: 67 subjects were identified, of which 25 were located and agreed to participate. Of these, mean age was 17.3 years with 11.2 years follow up. 67% were acetylcholine receptor positive. 75% had chiefly ocular manifestations. At follow up, 64% were still taking medication and 80% were still seeing a neurologist. 40% underwent thymectomy. 20% were hospitalized for myasthenic weakness at some time in the course of their disease. Overall, all subjects noted improvement over their course, but most manifested some residual symptoms with persistent fatigable eyelid ptosis in 48%. This group was highly functional, with 96% either full time students or fully employed. All but 2 were completely independent in their activities of daily living. Characteristics of the surveyed group suggest there may be ascertainment bias toward more symptomatic long-term course.

SUMMARY/CONCLUSION: The vast majority of patients with JMG are functioning well today appropriate to age. Subgroup analysis may be valuable to prognosis.

SURVIVIN EXPRESSION IN B CELLS OF EAMG RAT MODEL IS SUPPRESSED WITH SURVIVIN PEPTIDE VACCINE
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INTRODUCTION: T cell subsets and B cells contribute to the pathogenesis of myasthenia gravis (MG). The mechanisms that underlie the development and maintenance of autoimmunity in myasthenia gravis are poorly understood.

OBJECTIVE: We assessed the role of survivin, a member of the inhibitor of apoptosis protein family, in the survival of B cells in experimental autoimmune MG animal model.

METHODS: Lewis female rats were treated with survivin peptide one injection per week for three weeks. Control group received PBS. The following week, animals were induced with acetylcholine receptor (AChR). Animals were assessed for clinical score, weight, and grip during the course of the experiment. Survivin antibodies and AChR-specific antibody IgG isotypes were determined from sera. Spleenic population was assessed for survivin-expressing B cells and B cells that recognized AChR by Flow cytometry.

RESULTS: The survivin-vaccine treated animals demonstrated improved clinical score assessment, survivin antibodies, a reduction in acetylcholine receptor specific autoantibodies, and associated with marked reduction of survivin-expressing circulating CD3-/CD45RA+ cells. In the control animals, a portion of survivin-expressing cells specifically bound a peptide derived from the alpha subunit of acetylcholine receptor indicating that they recognize the AChR.

SUMMARY/CONCLUSION: Utilizing animal models of MG, we have shown the efficacy of a survivin vaccination strategy. Survivin expression may be part of a mechanism that inhibits the apoptosis of autoreactive B cells in myasthenia gravis and other autoimmune disorders.
TH17 POLARIZING ENVIRONMENT IN MYASTHENIA GRAVIS
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INTRODUCTION: Myasthenia gravis (MG) immunopathogenesis involves autoantibody production and increases in soluble interleukin-17 (IL-17) and T follicular helper (Tfh) cell frequencies. However, their interconnection has not been elucidated.

OBJECTIVE: To determine whether CD4+ T (Th) cells in MG patients are more prone to differentiating into IL-17 producing Th17 cells. Additionally, we examined the effect of this pro-inflammatory environment on Tfh cells.

METHODS: T-cells from five acetylcholine receptor autoantibody positive MG patients and eight healthy subjects were polarized for Th17 differentiation. A Th17 environment was produced using the T-cell activators αCD3/αCD28; recombinant cytokines IL-6, IL-1β, TGF-β1, and IL-23, and an antibody cocktail against IL-4 and IFN-γ. After one week of culture, T cells were re-stimulated with PMA/ION and the frequency of CXCR5+ Tfh cells and IL-17 producing T-cells were assessed by polychromatic flow cytometry.

RESULTS: Th17 polarization resulted in similar increases in IL-17+ CD4 T-cell frequencies in MG patients and controls. Interestingly, we observed increased frequencies of CD8 T-cells capable of producing IL-17 in MG patients. We also observed an increase in the frequency of CXCR5+ Tfh cells and IL-17 producing T-cells were assessed by polychromatic flow cytometry.

SUMMARY/CONCLUSION: Although we did not conclusively show that Th cells are predisposed to differentiate into Th17 cells, the detection of IL-17 producing CD8 T-cells is a novel finding that implies they may be involved in MG pathology. Collectively, the proinflammatory Th17 environment in MG may be a common theme that elevates IL-17 levels and differentiation of Tfh cells to promote the generation of autoantibody producing B cells.

THE PREDICTIVE VALUE OF THE TITERS OF ANTI-ACETYLCHOLINE RECEPTOR ANTIBODIES TO THYMUS PATHOLOGY AND TO THE CLINICAL OUTCOME IN PATIENTS UNDERWENT THYMECTOMY
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INTRODUCTION: To analyze the predictive value of anti-acetylcholine receptor antibodies (anti-AChRAb) for a predictive factor of thymus pathology, and for clinical outcome in myasthenia gravis (MG) underwent thymectomy.

OBJECTIVE: To evaluate if high anti-AChRAb levels might predict a more malignancy grade of thymic neoplasm and associate with a non remissionable clinical course in MG.

METHODS: The clinical data of 80 patients with MG undergoing thymectomy from January 2012 to December 2015 were retrospectively reviewed. Anti-AChRAb assayed pre and after thymectomy. The following factors were analyzed in relation to the clinical issues: Osserman-stage, Myasthenia Gravis Foundation of America (MGFA) stage. Quantitative Myasthenia Gravis Score (QMGS), frequency of recurrence, World Health Organization (WHO) histologic classification and Masaoka staging system.

RESULTS: Anti-AChRAb were detected in 90.6% of patients. Thymic lymphoid follicular hyperplasia (LFH) was present in 26.6%, thymoma in 59.3%, cyst of thymus in 6.3%, undegraded thymus in 7.8%. LFH was associated with a lower anti-AChRAb compared with thymoma(10.04±6.06 vs 15.26±5.70, Radio Immunoprecipitation Assay). Non remission within one year followed operation tend to have a higher anti-AChRAb compared with total remission group (16.39±4.02 vs 14.26±3.11). Multivariate logistic regression analysis showed that only Osserman-stage (IIB- OR = 3.49, 95% CI = 1.09-15.12 P = 0.041) were associated with statistically significant risk of recurrence on multivariate analysis.

SUMMARY/CONCLUSION: The highest chance of recurrence of MG post operation was observed in patients with IIB stage. Compared with LFH, while higher anti-AChRAb levels is associated with thymoma, High anti-AChRAb levels with non remissionable clinical course indicating a more aggressive chemical therapy among thymic MG.