

ELECTROPHYSIOLOGICAL CHARACTERIZATION OF CERVICAL SPONDYLOTIC MYELOPATHY

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Introduction: Cervical spondylotic myelopathy (CSM) is defined as cervical cord damage due to spondylosis of the cervical spine. It has proven to be the most common cause of myelopathy for over 60 years. Electrodiagnostic tests are very useful in demonstrating the location and intensity of lesions, functional disability, and prognosis.

Objects: The authors will describe and demonstrate the various damages in terms of electrophysiological parameters.

Methods: Thirty patients of both sexes with magnetic resonance imaging (MRI) confirmed CSM were studied. Electromyography (EMG), somatosensory and motor evoked potentials (MEPS), nerve conduction studies, and F-waves were applied to them.

Results: MEPS proved to be the most affected. It is suggested that corticospinal tracts are affected early and in the most intense form. Electrophysiological parameters were affected in different forms: MEP amplitude was decreased in 69.5%, MEP amplitude ratio was decreased in 78%, and P40 wave latency was increased in 52.1% of patients. EMG was affected in all patients: 72.7% showed a diffuse neurogenic pattern, and the rest of the patients showed a radicular pattern. The authors also demonstrated moderate to severe abnormalities of motor and sensory nerve conduction of the median nerve in 52.1% of the patients. There was great correlation between electrophysiological parameter abnormalities and the intensity of MRI cord damage.

Conclusions: MEPs are most affected in CSM. There is involvement of the median nerves in an important group of CSM patients that is an overlapping abnormality. In addition, there is great correlation between abnormalities of electrophysiological parameters and MRI damage of the cord.

DECREMENTING RESPONSE IN REPETITIVE NERVE STIMULATION IN AMYOTROPHIC LATERAL SCLEROSIS

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Introduction: It has been long known that the decrementing response (decrement) in repetitive nerve stimulation is observed in amyotrophic lateral sclerosis (ALS), as well as in neuromuscular junction disorders such as myasthenia gravis (MG). However, the details of decrement in ALS have not been fully investigated.

Objectives: To clarify the frequency, distribution, and other features of decrement in ALS, in comparison to those of MG.

Methods: Subjects included 50 patients with ALS, 39 generalized MG (gMG) patients, and 20 ocular MG (oMG) patients. All MG patients were examined before treatment. The authors tested six muscles including upper trapezius, deltoid, and nasalis, and distal muscles. The decrement of the amplitude of the fourth response from the first one following 3Hz stimulation was measured, and a decrement exceeding 5% was interpreted to be abnormal.

Results: Significant decrement at least in one muscle was observed in 84% of ALS patients, and in 74% and 45% of gMG and oMG patients. Decrement was most frequently observed in proximal muscles in ALS, as well as in MG (deltoid 77% and 62%, and trapezius 68% and 51% for ALS and gMG, respectively). Decrement in nasalis was frequent in gMG (54%), but was rare in ALS (8%). In general, there was little difference between ALS and MG regarding the characteristics of decrementing response, such as the degree of U-shape (i.e., the recovery after the bottom of decrement, or the response to different stimulus frequencies).

Conclusions: Decrement is a more frequently observed finding in cases of ALS than in generalized MG. Its detection may be helpful in the diagnosis of ALS.

ASSESSING THE ACCURACY OF CLINICAL TESTS WITH ELECTRODIAGNOSTIC STUDY FINDINGS FOR IDENTIFYING CARPAL TUNNEL SYNDROME; SHOULD HAND SURGEONS RELY ON CLINICAL FINDINGS

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Introduction: The aim of the study was to investigate whether a combination of selected provocative maneuvers and sensory testing could improve the accuracy of a carpal tunnel syndrome (CTS) diagnosis, or if hand surgeons should rely on electrodiagnostic (EDX) study findings.

Method: Retrospective studies that examined 200 patients who were referred with suspected CTS and had undergone nerve conduction studies (NCS) were conducted. Responses to Tinel sign, Phalen's test, and carpal compressions test (CCT) and sensory testing over the median sensory nerve distribution were assessed for each patient. Complete EDX studies were performed and the data was collected.

Results: The sensitivity (2.33, 2.54; 3.64) of compound motor action potential, and sensory action potential proved to be better than the nerve conduction velocity (0.49; 0.36, 0.89); also, the positive likelihood ratio and negative likelihood ratio were calculated for each test. The inclusion of sensory testing did not improve the sensitivity and specificity for Tinel sign, Phalen's test, or CCT (sensitivity: 0.51, 0.64, 0.14; specificity: 0.61, 0.75, 0.96).

Conclusion: EDX findings prove to be the most sensitive test in the identification of CTS. In many situations when clinical impressions of CTS were made, such determinations could not be verified by electrophysiological findings. More importantly, these data indicate that the assessment of sensation of the median sensory nerve distribution does not improve the diagnostic accuracy of CTS. However, a positive Phalen's test is more likely to be associated with NCS changes that are consistent with CTS.

NERVE CONDUCTION STUDIES IN SPASTIC PARAPLEGIA OPTIC ATROPHY AND NEUROPATHY SYNDROME

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Introduction: Spastic paraplegia, optic atrophy and neuropathy syndrome (SPOAN) is an autosomal recessive disorder originally characterized in 70 individuals living in a geographic isolate in Northeast Brazil. It is characterized by progressive spastic paraplegia, congenital optic atrophy and early onset axonal polyneuropathy. Linkage studies have shown that its responsible gene is located at 11q12-13, in a 2.3 Mb region.

Objective: To evaluate nerve conduction in patients with SPOAN syndrome.

Methods: Twenty-three individuals (18 females) with confirmed SPOAN syndrome ranging in age from 4 to 58 years participated in this study. They were submitted to nerve conduction studies (NCSs) which included motor conduction studies of the right median, ulnar, tibial, and peroneal nerves; bilateral tibial H reflexes, and sensory NCSs studies of the right median, ulnar, radial, sural, and superficial peroneal nerves.

Results: The NCSs of all patients were clearly abnormal, and demonstrated a severe sensory motor axonal polyneuropathy. Sensory nerve potentials were consistently absent in the lower limbs, and were usually absent in median (21/23), ulnar (19/23), and radial (17/23) nerves. Motor NCSs revealed reduced amplitudes and borderline conduction velocities in the upper limbs, and usually absent potentials in peroneal (20/23) and tibial (19/23) nerves. Tibial H reflexes were usually absent (21/23), despite the spastic paraplegia.

Conclusion: Patients with SPOAN syndrome have a severe, early onset sensory motor axonal polyneuropathy. A normal NCS appears to effectively rule out this condition.

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EVALUATING SYMPATHETIC DYSFUNCTION AND NEUROPATHY IN PARKINSON'S DISEASE

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Introduction: Autonomic dysfunction in Parkinson's disease may be partly mediated by the peripheral nerves, but this component has not been well characterized. The incidence of neuropathy in these patients has not been thoroughly examined. While some studies of Parkinson's patients have used either autonomic testing or nerve conduction studies (NCSs), none have utilized both simultaneously.

Objectives: To determine whether there is a higher incidence of peripheral nerve dysfunction in Parkinson's disease and to investigate a potential correlation with a disturbance of the sympathetic autonomic system.

Methods: Ten patients with Parkinson's disease completed questionnaires for neuropathy and autonomic symptoms. Ankle reflex and vibration testing were performed with sympathetic skin responses (SSRs) and NCSs of the sural and peroneal nerves. R-R intervals were recorded at rest and during various maneuvers. Statistical methodology included correlation testing and chi-square analysis of measures of neuropathy and sympathetic dysfunction.

Results: Seven subjects (70%) had clinical signs of neuropathy. Of those, six had abnormal SSRs. Six subjects (60%) had abnormal NCSs, and of those, five had abnormal SSRs. In total, eight subjects (80%) had abnormal SSRs, and one subject (10%) had abnormal R-R intervals. Five subjects (50%) complained of autonomic symptoms; all had abnormal SSRs, and three had abnormal NCSs. Pearson correlation coefficients and chi-square analysis of measures for neuropathy and sympathetic dysfunction did not achieve statistical significance.

Conclusions: In this pilot study, there was an increased prevalence of abnormal SSRs and various measures of neuropathy. Further study is required

to establish a conclusive link between neuropathy and autonomic dysfunction in Parkinson's disease.

PROGRESSIVE ELECTROPHYSIOLOGIC WORSENING DESPITE CLINICAL IMPROVEMENT IN ACUTE INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY

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Introduction: In acute inflammatory demyelinating disease (AIDP), electrophysiological abnormalities and accompanying clinical indices have been assumed to reach their nadir by 3 to 4 weeks, followed by improvement.

Objectives: To understand the correlation between serial electrophysiological indices in nerve conduction studies (NCSs) of demyelination in AIDP and clinical status, particularly during recovery.

Methods: Retrospective analysis of AIDP patients with standardized serial clinical and NCS data from January of 2008 to January to 2010.

Results: The mean age (n=15) was 51.5, 80% were males, 87% had a preceding infection, 40% required intubation, and 100% received either intravenous immunoglobulin or plasma exchange. NCS and clinical examination on average were performed at day 7, 29, and 85. The average Guillain-Barre syndrome disability score was 3.7, 3.1, 1.9, respectively. Nerve conduction study results (% of normal for our laboratory) were: Median distal motor latency (DML) 126%, 127%, 203% (p = 0.03), median motor conduction velocity (CV) 97%, 84%, 83% (p=0.02), and median compound muscle action potential (CMAP) 68%, 78%, 88% (p= 0.08) respectively; Ulnar DML 125%, 150%, 174% (p = 0.11), ulnar motor CV 100%, 95%, 92% (p= 0.09), and ulnar CMAP 61%, 59%, 72% (p = 0.32), respectively; peroneal DML 110%, 130%, 160% (p = 0.01), peroneal motor CV was 94%, 90%, 86% (p =0.42), and peroneal CMAPs 44%, 33%, 36% (p = 0.18) respectively. Several examples were identified with dramatic electrophysiological decline despite clinical improvement.

Conclusions: Progressive electrophysiologic worsening beyond 4 weeks, despite clinical improvement in AIDP, is common. Evolutionary electrophysiological worsening should not be mistaken for recurrent disease or rebound deterioration.

ASSESSMENT OF H-REFLEX AND ANKLE REFLEX IN UNILATERAL S1 RADICULOPATHY

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Introduction: In addition to clinical examination, there are many tools used for the diagnosis of lumbosacral radiculopathy, including S1 roots lesion. Magnetic resonance imaging (MRI) and electrodiagnostic (EDX) studies are well known and are routinely used.

Objectives: To determine the value of ankle reflex in comparison to H-reflex in diagnosis of unilateral S1 radiculopathy.

Methods: The authors examined 76 patients with a confirmed diagnosis of unilateral S1 radiculopathy based on physical examination and MRI studies. Each was assessed using EDX studies that included nerve conduction studies, needle electromyography, and tibial H-reflex. Clinical examination with special attention to ankle reflex was also performed.

Results: There was moderate agreement between H reflex latency and ankle reflex (P value= 0.001). No correlation was found between H reflex latency or ankle reflex with severity of disk involvement in MRI.

Conclusion: Ankle reflex and tibial H-reflex are used in the assessment of S1 radiculopathy with different specificities and sensitivities. In this study, moderate agreement between ankle reflex and H-reflex was found in patients with evidence of ipsilateral S1 radiculopathy (KAPPA coefficient=0.45, $P=0.001$).

POSTERIOR INTEROSSEOUS NERVE CONDUCTION: A NOVEL TRANSCUTANEOUS TECHNIQUE FOR DIAGNOSIS OF PATHOLOGY

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Introduction: The posterior interosseous nerve (PIN) is a deep motor branch of the radial nerve. Clinical symptoms of posterior interosseous nerve syndrome (PINS) include finger extensor weakness as well as tenderness over the elbow lateral epicondyle and arcade of Frosche. Current diagnostic methods of PINS include electromyography, needle stimulation/recording of PIN conduction, and imaging with ultrasound or magnetic resonance imaging. Development of a more cost effective and less invasive diagnostic tool for PINS would be beneficial.

Objectives: This study evaluates the sensitivity and specificity of a transcutaneous, nerve conduction study technique for the diagnosis of PINS.

Methods: Twenty-two subjects with clinically suspected PINS were compared to 40 control subjects. Recording electrodes were placed over the brachioradialis (Br) and extensor carpi ulnaris (ECU) muscles with stimulation 5 cm above the lateral epicondyle for each upper limb. The Br-ECU latency was calculated for each limb, and the side to side latency difference was compared. The authors also compared the side to side latencies to the ECU as another option for identifying PIN impairment.

Results: The mean side to side Br-ECU latency difference was 0.15 ± 0.12 in the control subjects; the mean of PINS subjects was 0.52 ± 0.44 . The control group's ECU side to side difference mean was 0.016 ± 0.371 ; the PINS subject's mean was 0.571 ± 0.333 . The area under the receiver operating characteristic curve was 0.878 for the Br-ECU latency difference, and 0.859 for the ECU difference.

Conclusions: Transcutaneous nerve conduction detection using the technique described provides a non invasive means of evaluation of PINS with high sensitivity and specificity (>85%).

OPTIMAL RECORDING ELECTRODE SITES FOR RADIAL MOTOR NERVE CONDUCTION STUDIES WITH EXTENSOR INDICIS: CADAVERIC AND ELECTROPHYSIOLOGIC STUDIES

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Introduction: Active (E1) and reference (E2) recording electrodes for the extensor indicis proprius muscle (EIP) have not been subjected to scientific scrutiny. Additionally, a concentric needle electrode was used as the recording device.

Objectives: To demonstrate the optimal combination of surface E1 and E2 electrode placement for radial motor nerve conduction study (MNCS) with EIP, based on cadaveric and electrophysiologic studies.

Methods: Thirty-six upper limbs from 19 fresh frozen cadavers were dissected. The distance (MIDPOINT) from the tip of ulnar styloid process (USP) to the midpoint between the upper and lower margin of EIP was measured and the ratio of MIDPOINT over the forearm length was calculated. Radial MNCS with surface electrodes was performed in 158 arms of 79 normal volunteers (23 women, 56 men). A total of six combinations of three E1 sites (3.5 cm proximal to USP; distal one-fourth of forearm length; the most prominent point during active contraction) and two E2 sites (tip of ulnar styloid process; EIP tendon at wrist) were studied. Stimulation site was 8 cm proximal to each E1.

Results: Among combinations of E1 and E2, the optimal combination to obtain the greatest amplitude of radial compound muscle action potential is E1 of the distal 1/4 point of forearm length and E2 of EIP tendon point at the wrist.

Conclusions: The optimal E1 and E2 recording electrodes for radial MNCS with EIP is the distal one-fourth point of the forearm length and EIP tendon at the wrist, respectively.

SPECTRUM OF NEUROIMAGING FINDINGS IN L5 LUMBAR RADICULOPATHY

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Introduction: L5 radiculopathy has characteristic clinical and electrodiagnostic (EDX) features of weakness and denervation of hip abduction, ankle dorsiflexion, and inversion with pre ganglionic sensory loss resulting in preserved lower extremity sensory responses. It is unknown how often patients with this distinctive clinical EDX presentation have isolated L5 root compression on neuroimaging versus other single root compressions (i.e., L4 or S1) or more widespread lumbar foraminal or spinal stenosis.

Objectives: To review the neuroimaging findings in electromyography (EMG) confirmed L5 radiculopathy for: 1) the presence of severe or moderate to severe isolated L5, L4, or S1 root compression, 2) calculation of a combined L3-S1 foraminal, lateral recess, and 3) spinal stenosis severity score (SSS) ranging from 0 to 60.

Methods: Twenty-six consecutive patients with EMG confirmed L5 radiculopathy who had lumbar magnetic resonance imaging or computerized tomography myelogram within 3 months of the EDX examination were retrospectively analyzed by a blinded neuroradiologist.

Results: Only 9 patients (35%) had isolated or predominant L5 root compression, 14 (54%) had more diffuse neuroimaging abnormalities, and 3 (12%) had essentially normal neuroimaging. Compared to the 14 patients with diffuse disease, the nine patients with more focal L5 compression were significantly younger (mean age: 47 versus 61), and had a lower SSS (mean score: 13 versus 21). There was a strong positive correlation between age and the SSS.

Conclusions: Neuroimaging evidence of isolated L5 root compression in EMG confirmed L5 radiculopathy appears to decline with age due to the pres-

ence of diffuse multilevel neuroforaminal, lateral recess, and spinal stenosis. The authors did not find neuroimaging evidence of L5 radiculopathy due to isolated L4- or S1-root compression.

PROTEOMIC ANALYSIS OF AMYLOID-LIKE DEPOSITS IN WALDENSTROM'S IgM NEUROPATHY

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Introduction: Rarely, IgM neuropathy patients have nerve biopsies with protein deposits morphologically similar to amyloid but not staining with Congo red. The exact nature of these deposits is unknown. Laser microdissection tandem mass spectrometry (MS), a validated technique in the analysis of immunoglobulin derived deposits, allows for a simultaneous and unbiased search of multiple peptides from paraffin embedded tissue.

Objective: To describe the exact protein composition of amyloid-like deposits in Waldenstrom's neuropathy using a novel MS based proteomic analysis.

Methods: The authors performed amyloid stains, immunohistochemistry, electron microscopy, and laser microdissection/dual MS of proteinaceous deposits in sural nerves of two patients with neuropathy and IgM lambda Waldenstrom's macroglobulinemia.

Results: Sural nerve biopsies had similar histopathological, immunohistochemical, and ultrastructural features. There was severe nerve fiber loss, epineurial inflammation, and no lymphomatosis. Dense hyaline endoneurial deposits with amyloid-like fibrillar ultrastructure were Congo red negative and did not react to proteins universally associated with amyloid (i.e., serum amyloid protein (SAP) and apolipoprotein E (ApoE)). Amyloid controls were positive. In both cases, MS of microdissected tissue containing the deposits detected a peptide profile that was consistent with the whole IgM lambda immunoglobulin molecule based on the

detection of immunoglobulin mu heavy chain constant region, immunoglobulin lambda light chain constant region, and immunoglobulin J chain.

Conclusions: Endoneurial amyloid-like deposits in Waldenstrom's neuropathy are composed of all the peptides that comprise an IgM pentameric molecule and lack the amyloid associated proteins SAP and ApoE. Deposition of the whole circulating IgM molecule may be a mediator of nerve injury. Autoimmune mechanisms may be contributory.

COMBINED SENSORY INDEX UTILITY IN CARPAL TUNNEL SYNDROME

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Introduction: Combined sensory index (CSI), a summation of three latency differences between the median nerve and the ulnar and radial nerves (i.e., comparison tests of median and ulnar nerve at digit IV (split ring finger (SR)) of median/radial nerve at digit I (numb thumb (NT)) and of median and ulnar sensory latencies at palm (P8) is reported to improve the diagnostic sensitivity in the electrodiagnosis of carpal tunnel syndrome (CTS). However, performing all three tests might not be necessary.

Objectives: To investigate how often it is necessary to perform all full three components of the CSI to meet the diagnostic criteria of CTS.

Methods: The authors retrospectively analyzed electromyography reports of patients who were diagnosed with CTS. Patients with a previous carpal tunnel surgery or with evidence of peripheral neuropathy were excluded from the study.

Results: A total of 243 extremities of 145 patients with CTS were analyzed. The mean age was 48.4 ± 9.9 years. Conventional median sensory and motor distal latency studies were diagnostic in 83% of the extremities. Performing only one component of the CSI test increased the diagnostic yield by 15%. The three part CSI test was required to increase the diagnostic yield by 2%. Of the indi-

vidual component tests that included NT, SR and P8, NT was found to be the most sensitive.

Conclusions: Although there is no single test that best determines the diagnosis of CTS, the authors believe that performing only one component of the CSI is required in 15% of cases, while all three components are required in only 2% of cases.

DIFFERENTIATING SYMPTOMATIC FROM ASYMPTOMATIC ULNAR SLOWING ACROSS THE ELBOW

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Introduction: Ulnar neuropathy at the elbow (UNE) is the second most frequently observed focal peripheral neuropathy of the upper limb. A surgical intervention is often considered for the treatment of UNE. It is common to find asymptomatic ulnar slowing across the elbow in routine nerve conduction studies.

Objectives: To differentiate symptomatic and asymptomatic ulnar slowing across the elbow.

Methods: A retrospective chart review was done on patients with and without ulnar symptoms who underwent upper extremity nerve conduction studies. Patients with peripheral neuropathy or a history of previous surgery to the elbow were excluded from the study.

Results: A total of 169 extremities were included in the study. Eighty-five had symptomatic UNE, and 84 had ulnar slowing, but experienced no ulnar symptoms. There was no difference between the two groups with regards to age, gender, side of involvement, or percentage of ulnar nerve slowing across the elbow. The only significant correlation between symptomatic and asymptomatic ulnar nerve slowing across the elbow was found with a percent ulnar nerve amplitude drop across the elbow ($p=0.000$). A 17% amplitude drop was seen in the symptomatic group as compared to 1% in the asymptomatic group.

Conclusions: One important difference between symptomatic and asymptomatic ulnar nerve slowing across the elbow is that the existence of significant amplitude drop is more commonly seen in the symptomatic group. This should be considered in the diagnosis of UNE.

CONCURRENT NERVE ENTRAPMENT IN PATIENTS REFERRED FOR ELECTRODIAGNOSTIC EVALUATION OF CARPAL TUNNEL SYNDROME

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Introduction: When patients are referred for electrodiagnostic (EDX) evaluation of possible carpal tunnel syndrome (CTS), laboratories may be asked to test only the median nerve(s) in an effort to minimize cost. This practice based retrospective review will help determine whether such minimization represents good clinical practice.

Objective: To determine the incidence of diagnoses other than CTS in a population referred for CTS evaluation.

Methods: From July of 2001 through December of 2006, a specific laboratory received 449 upper limb referrals. Of these, 207 (46%) were for CTS evaluation: 90 unilateral and 117 bilateral. Thus, 324 limbs were tested for CTS as follows: median and ulnar motor and sensory studies, ulnar F wave, radial sensory (examiner's discretion), and monopolar electromyography of C5-T1 myotomes, including abductor pollicis brevis. One electromyographer performed all EDX examinations and diagnosed CTS as well as other nerve entrapments using standard criteria.

Results: CTS was present in 284 limbs (88%). Of the remaining 40, 19 (5.9%) showed another diagnosis, and 21 (6.1%) were normal. In the 284 limbs with CTS, it was a case of sole diagnosis in 178 (63%), but 106 (37%) had one or more other diagnoses. These other diagnoses included ulnar neuropathy at the elbow (84 limbs), ulnar neuropathy

at the wrist (n=6), peripheral neuropathy (n=19), cervical radiculopathy (n=15), brachial plexopathy (n=1), and radial neuropathy (n=2).

Conclusions: Among those referred for CTS evaluation, other diagnoses were found to occur at an incidence of 39% (125/324) overall, and 37% (106/284), even with CTS present. Complete upper limb examination is indicated in patients referred to the EDX laboratory for CTS.

ABSENCE OF NECK PAIN: CERVICAL RADICULOMYELOPATHY MISDIAGNOSED AS NEUROMUSCULAR DISEASE

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Introduction: Four cases that demonstrate an occasional difficulty to consider cervical myelopathy with radiculopathy as important differential diagnosis early in the course of evaluation of symmetric or asymmetric leg and arm weakness are presented. This is especially important when neck pain or incontinence are absent, if the examination is not unequivocally indicating myelopathy, and if other neuromuscular conditions are present, such as polyneuropathy or myopathy.

Case Study: The initial working diagnoses in these cases included peripheral neuropathy and superimposed myositis, possible statin myopathy, Vitamin B12 deficiency-related neuropathy with corticospinal tract involvement, and possible motor neuron disease. The patients presented 2 to 6 months after initial work up after developing significant difficulty walking. Magnetic resonance imaging (MRI) in all cases indicated severe cervical degenerative disc disease with spinal canal stenosis and with myelomalacia of different degrees. Surgery was performed in each case, but provided a lack of symptom improvement.

Conclusion: These cases highlight the importance for neuromuscular specialists, in particular, to con-

sider cervical radiculopathy with myelopathy early in the evaluation of a patient even if presentation is atypical. Early detection of cervical radiculomyelopathy may improve and/or halt the potential progression of neurologic dysfunction.

NEUROMUSCULAR ABNORMALITIES IN PATIENTS WITH GASTROPARESIS

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Objective: To determine whether or not patients with gastroparesis also had peripheral neuromuscular abnormalities.

Methods: This descriptive study assessed neuromuscular (NM) abnormalities seen in a cohort of 46 patients with symptoms of gastroparesis refractory to conventional medical and neurostimulator therapies. All patients (mean age 41 ± 25 years; diabetic gastroparesis 12%; post surgical gastroparesis 10%; and idiopathic gastroparesis 78%) underwent detailed NM examination. Of 46 patients, 23 (50%) were found to have abnormal neurological examinations and were further studied with electromyography (EMG) and nerve conduction studies NCSs; anti neuronal nuclear autoantibody types 1, 2, and 3; purkinje cell cytoplasmic autoantibodies type 1, 2; voltage gated potassium channels (VGKC); and genetic markers for thymidine phosphorylase.

Results: EMG and NCS showed myopathic changes in 12 patients (50%) and polyneuropathy in 7 (30%). Plasma amino acids were present in 36%, striated muscle antibodies in 12%, and VGKC antibodies in 4%. P/Q calcium channel antibodies and anti acetylcholine receptor ganglionic antibodies were present in 4% patients.

Conclusions: Peripheral NM abnormalities were seen in half of all patients with refractory gastroparesis. The presence of peripheral nerve antibodies, indicated in a significant number of patients with abnormalities, suggests a generalized NM pathology of peripheral and gastrointestinal NM systems. NM

evaluation yields findings important in the evaluation of patients with refractory gastroparesis.

Earnest L. Murray, II, MD

Junior Member Recognition Award Recipient

MUSCLE INTRUSION INTO THE TUNNEL IN CARPAL TUNNEL SYNDROME

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Introduction: Carpal tunnel syndrome (CTS) is a common condition with an etiology that is unknown in most cases. One potential cause is muscle intrusion into the carpal tunnel, which may increase pressure within the tunnel and compress the median nerve.

Objectives: To determine if those with CTS have larger amounts of flexor digitorum and lumbrical muscle within the tunnel than those without CTS.

Methods: Poultry plant workers, which are considered to be a high risk population for development of CTS, were screened using a hand diagram, nerve conduction studies (NCS), and high resolution ultrasound. Those with normal NCSs and hand diagrams were categorized as “no CTS,” those with one abnormal finding were categorized as “possible CTS,” and those with abnormal diagrams in a classic CTS pattern plus median sensory onset latencies 0.5 ms or greater than ulnar latencies were classified as “definite CTS.”

Results: Screening was performed on 369 wrists. Of those, 3% met criteria for “definite CTS” while 26.8% met criteria for “possible CTS.” There was muscle in the tunnel in 100% of those with “definite CTS,” 92% of those with “possible CTS,” and 82% of those with “no CTS.” The amount of muscle in the tunnel with the wrist in neutral position was 8.1 mm² for “definite CTS” and 4.8mm² for “no CTS” (p = 0.01).

Conclusions: Muscle within the carpal tunnel was found more frequently and to a greater extent in those with “definite CTS” than those with “no CTS.” This suggests that muscle intrusion may play a role in idiopathic CTS, but prospective investigation is needed.

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ESTABLISHMENT OF NORMAL VALUES FOR THE ULNAR NERVE ACROSS THE ELBOW

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Introduction: Definitive diagnosis of ulnar neuropathy at the elbow is clinically and electrodiagnostically challenging. There are various electrodiagnostic (EDX) techniques used for diagnosis. It remains unclear whether one technique or a combination of techniques can be used to improve EDX sensitivity.

Objectives: To present normative, temperature controlled nerve conduction values for ulnar nerve motor studies to the first dorsal interosseous (FDI), abductor digiti mini (ADM), and mixed median ulnar nerve action potential (NAP) difference, for each subject.

Methods: Normal data were gathered at a hospital based university EDX laboratory for nerve conduction studies (NCS) of the three previously described techniques. Each of the NCSs was performed on 30 arms from 15 volunteers (age 26 to 59). None had symptoms of neuropathy or radiculopathy. Temperature was controlled at $\geq 33^{\circ}\text{C}$ for each stimulation site.

Results: Normative data is reported as mean \pm one standard deviation (mean \pm SD). The mean difference between the mixed median ulnar NAP study was found to be 0.3 ms \pm 0.2 ms. The mean conduction velocity across the elbow was 59.3 m/s \pm 6.5 m/s for the ulnar motor to FDI, and 61.3 m/s \pm 5.2 m/s for the ulnar motor to ADM.

Conclusions: The authors report normal values for the three temperature controlled tests administered on each arm of every subject. This normative data can be used for future reference in the diagnosis of ulnar neuropathy at the elbow.

Michael N. Horner, DO

Junior Member Recognition Award Recipient

COMPARISON OF THE MEDIAN NERVE CROSS SECTIONAL AREA, CARPAL TUNNEL CROSS SECTIONAL AREA, AND K-J INDEX BETWEEN NORMAL AND CARPAL TUNNEL SYNDROME SUBJECTS

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Objective: The ultrasonographic cross sectional area (CSA) of the median nerve is used to diagnose carpal tunnel syndrome (CTS). The ratio of the CSA of the median nerve to the carpal tunnel (K-J index) was obtained and compared between normal reference groups and subjects diagnosed with CTS using electromyography (EMG).

Materials and Methods: Fifty hands from 28 patients who were diagnosed with CTS using EMG were used to obtain the proximal and distal CSAs of the median and carpal tunnel with ultrasonography. The CSAs were compared to the normal reference group.

Results: The proximal median CSA, carpal tunnel CSA, proximal K-J index, distal median CSA, distal carpal tunnel CSA and distal K-J index of the patient group was $14.27 \pm 6.10\text{mm}^2$, $207.72 \pm 34.14\text{mm}^2$, 6.97 ± 2.98 , $11.20 \pm 2.83\text{mm}^2$, $154.18 \pm 22.02\text{mm}^2$, 7.42 ± 2.30 , respectively. Those of the normal reference group were $9.64 \pm 2.86\text{mm}^2$, $166.86 \pm 27.85\text{mm}^2$, 5.85 ± 1.66 , $9.22 \pm 2.83\text{mm}^2$, $130.20 \pm 21.27\text{mm}^2$ and 7.08 ± 1.71 , respectively. Thus, with exception of the distal K-J index, the remaining indexes and areas were increased in the patient group as compared the normal reference group ($P < 0.05$).

Conclusion: In ultrasonographic findings of CTS, an increase is seen not only in the median nerve CSA, but also in the carpal tunnel CSA. This implies that CTS occurs due to median nerve compression, secondary to flexor tenosynovitis, and that preventing edematous change of the flexor retinaculum is central in treating CTS.

SYMPTOMATIC SURGICAL BENEFIT IN CARPAL TUNNEL: ROLE OF MEDIAN SENSORY STUDIES

J. P. Ney, J. Jarvik (Seattle, WA)

Introduction: Carpal tunnel syndrome (CTS) is frequently treated with surgery and conservative measures. Median sensory and mixed nerve studies are among the most sensitive tests for diagnosing CTS.

Objectives: To retrospectively examine median sensory and mixed nerve studies in relation to surgical outcomes in a randomized clinical trial (RCT) and cohort study of mild to moderate CTS.

Methods: Subjects with reported median digital sensory nerve action potentials (SNAPs), or median palmar (8cm) studies were included. Latencies were converted to sensory and mixed nerve conduction velocities (SMNCVs). The adjusted 12 month carpal tunnel assessment questionnaire symptomatic (CTSAQS) score was the outcome of interest. The relationship between median SMNCVs and the adjusted 12 month CTSAQS were modeled with interaction of SMNCV and surgery/no surgery as a predictor in linear regression. Demographic, health related and psychosocial variables were added as covariates in separate regressions. In the RCT, intention to treat and as treated (AT) analyses were performed.

Results: SMNCVs (range 5 to 48 m/s) were calculated from 82 RCT subjects and 165 cohort subjects. For SMNCVs > 27 m/s, a consistent trend of improved (average 0.2 to 0.4 lower) adjusted CTSAQS scores were seen in all analyses for those receiving or randomized to surgery, relative to sub-

jects with lower SMNCVs. This was most prominent in the AT analysis of the RCT (interaction-p-value=0.009).

Conclusion: This study suggests that subjects with higher median SMNCVs have greater symptomatic benefit from surgical intervention for CTS than those with more electrodiagnostically severe disease. Future study using uniform electrodiagnostic techniques is warranted to confirm these findings.

John P. Ney, MD

Junior Member Recognition Award Recipient

DYNAMIC ULTRASOUND CHARACTERISTICS OF THE MEDIAN NERVE IN THE CARPAL TUNNEL

J.A. Strakowski, D.B. Guffrey (Columbus, OH)

Introduction: Previous median nerve studies using high frequency ultrasound (US) have included primarily static or low resistance dynamic measurements. Obtaining further information regarding the dynamic effects of grip resistance and wrist position could provide additional insight into occupational strains and other stresses on the median nerve at the carpal tunnel.

Objectives: To identify dynamic differences in the parameters of cross sectional area (CSA) and excursion of the median nerve in the carpal tunnel space, with changes in grip resistance and wrist position.

Methods: Eighty wrists of 40 electrodiagnostically confirmed normal volunteers were evaluated with high frequency US. Utilizing a standardized compression device, dynamic measurements of the median nerve were taken with the wrist in varied positions and flexion of the digits with varied resistance. The parameters of the median nerve measured included absolute longitudinal excursion, median to flexion tendon excursion ratio, relative excursion in short axis, and changes in CSAs.

Results: A decreased excursion of the median nerve as well as the median to tendon excursion ratio, in

all positions of the wrist, is seen with increased resistance. It is further accentuated with extreme wrist flexion and extension. Slightly smaller measured CSA is seen with increased grip resistance.

Conclusions: Increased grip force results in deformational changes and more limited dynamic excursion of the median nerve in the carpal tunnel space. This is further accentuated with grip position in extreme flexion and extension. Additional US studies could provide further insight into the relative effect of movements, grip, and hand and wrist position.

OXYNEUROGRAPHY IS A NOVEL APPLICATION OF THE NEAR INFRARED SPECTROSCOPY TECHNIQUE

J.F. Jabre, G.M. Squintani, D. De Grandis* (Boston, MA; Verona, Veneto, Italy*)*

Introduction: Near infrared spectroscopy (NIRS) is a non invasive technique that has been applied to the study of perfusion and oxygenation of skeletal muscle, tumor detection in breast tissue, and in functional studies of brain perfusion.

Objectives: To describe oxyneurography (ONG), a novel application of the NIRS technique in the measurement of the total oxygenation index (TOI) of human nerves.

Methods: The authors used ONG in the study of the ulnar, tibial and sural nerves in normal subjects, as well as in patients with ulnar neuropathy at the elbow. NIRS electrodes were applied directly over the nerves to measure their TOI, a ratio of the nerve's oxygenated hemoglobin content over its total hemoglobin content.

Results: The nerve TOI normal values were $70.65 + 4.23$, which were similar to TOI values obtained in normal muscle and other tissues. In patients with ulnar neuropathy at the elbow, there was a significant correlation between the drop of the ulnar nerve's TOI and its conduction velocity slowing at the elbow.

Conclusions: Routine nerve conduction studies assess a nerve's myelin sheath and axonal integrity, but leave the status of a nerve's vascular integrity unknown. Oxyneurography not only leads to the early detection of compromise in a nerve's perfusion and oxygenation status, but also proves to be effective in the assessment of pharmacological agents destined to prevent or reverse the effects of a nerve's vascular compromise which leads to neuropathy.

GUILLAIN-BARRE SYNDROME AFTER H1N1 VACCINATION IN THE UNITED STATES

H. Yacoub, A. Qureshi, H. Khan, N. Souayah (Newark, NJ; Minneapolis, MN*)*

Objective: To determine the reporting rate and characteristics of Guillain-Barre syndrome (GBS) after the administration of H1N1 vaccine in the United States in 2009.

Background: Although GBS can be associated with the influenza vaccine, it remains unknown whether GBS can be associated with H1N1 influenza. The issue has major public health importance due to the recommendations given by the Center for Disease Control's Advisory Committee on Immunization Practices in regards to the 2009 H1N1 vaccine administration (July 29, 2009).

Methods: The authors analyzed data from the Vaccine Adverse Event Reporting System, supplemented by data from the Center for Biologics and Research under the Freedom of Information Act.

Results: Twenty-seven cases (mean age 39.8 ± 21.7 years; 11 were men) of GBS were reported following the administration of the H1N1 influenza vaccination, with an estimated occurrence of 2.7 per 10 million vaccinations. All cases of GBS were reported within 6 weeks after vaccination; 26 GBS cases (85.2%) were reported in the first 2 weeks after vaccination, with maximum occurrence reported in the second week. No incidences of disability or death were reported in 23 patients who were hospitalized. In 2009, 57 patients were diagnosed with

GBS after vaccination with the seasonal influenza vaccine. There was an estimated occurrence of 7.3 per 10 million vaccinations. This time period of occurrence was similar to that reported for GBS cases after H1N1 influenza vaccination.

Conclusion: This is one of the first national reports documenting the occurrence of GBS after H1N1 influenza. These results warrant early recognition, active surveillance, and treatment of GBS after H1N1 influenza vaccination.

Hussam A. Yacoub, DO

Junior Member Recognition Award Recipient

ABDOMINAL WALL PAIN CAUSED BY CUTANEOUS NERVE ENTRAPMENT IN A PREGNANT WOMAN

A.M. Januario, J. Magalhães, O.G. Lins (Recife, Pernambuco, Brasil)

Introduction: Entrapment neuropathies of the anterior cutaneous branches of the intercostal nerves are uncommon. During pregnancy, only involvement of the lateral thoracic branches had been reported. Changes in the abdominal wall during pregnancy can anatomically explain the entrapment. Branches of the seventh and eighth intercostal nerves are more affected because they cross several possible anatomical sites of entrapment.

Case Report: The authors report the case of a pregnant woman who noted pain and dysesthesias in the lower chest and upper abdominal wall since her second trimester of pregnancy. Physical examination showed hypoesthesia at the area. Somatosensory evoked potential (SEP) by stimulation of the skin of the affected area was not obtained for the right side, and had reduced amplitude and increased latencies for the left side. SEPs by stimulation of the skin of the surrounding areas were normal. The patient was reviewed 2 years later. She reported improvement of the symptoms, but complained of dysesthesia on the right side, elicited by movements such as lifting the arms. Cutaneous SEP showed reduced amplitude and prolonged

latency in response to stimulation of the affected right side and was normal for the left side.

Conclusion: Hypoesthesia and/or dysesthesia in the abdominal wall during pregnancy raises the possibility of compression of cutaneous branch of intercostal nerve. Anatomical aspects explain the specific involvement of the branches of the seventh and eighth intercostal nerves.

COMPARISON OF QUANTITATIVE PSEUDOMOTOR AXON REFLEX TEST, UTAH EARLY NEUROPATHY SCALE AND SKIN BIOPSY IN HUMAN IMMUNODEFICIENCY VIRUS NEUROPATHY

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Introduction: The quantitative pseudomotor axon reflex test (QSART) and Utah early neuropathy scale (UENS), as well as small fiber specific measures were investigated for assessing axon reflex test (ART) associated toxic neuropathy (ATN) in human immunodeficiency virus infected persons.

Methods & Results: Twenty individuals with symptoms and signs of neuropathy (cases) and eighteen control subjects, all on ART, were evaluated with QSART and UENS. Ten randomly selected patients in each group had skin biopsy at the thigh and distal leg with intraepidermal nerve fiber density analysis (IENFD). Pain was assessed with use of a visual analog scale (VAS). Mean sweat volume (μL) was lower in cases than in controls at all testing sites except forearm ($p < 0.01$). UENS and VAS (mm) were higher in cases than controls ($p < 0.05$ for all). Lower sweat volume at all sites correlated with higher total UENS and pin subscore, and prior week VAS ($p < 0.05$ for all). Mean IENFD was similar at the thigh (20.7 ± 8.0 versus 19.8 ± 7.3 fibers/mm), and although lower at the ankle in cases compared to controls, the difference was not significant (12.7 ± 5.9 versus 15.3 ± 8.4 fibers/mm, $p = 0.4$). IENFD did not correlate with QSART or UENS at any site, but

the ankle IENFD did correlate with prior week VAS ($r = -.547$, $p = 0.02$). QSART was more likely to be abnormal in cases (7/10, using published adjusted normative volumes) than biopsy (3/10 cases identified using cutoff of 10 fibers/mm at distal site for IENFD), and was normal in the majority of control subjects (8/10).

Conclusion: QSART and UENS have not been previously studied in this population and appear promising in the detection of ATN.

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UTILITY OF SOMATOSENSORY EVOKED POTENTIALS FOR THE DIAGNOSIS OF CHRONIC INFLAMMATORY DEMYELINATING POLYRADICULONEUROPATHY

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Introduction: A pure sensory variant of the chronic inflammatory demyelinating polyradiculoneuropathy (CIDP), namely sensory CIDP, has been infrequently reported. Its diagnosis is problematic because demyelination may not be evident in the sensory nerve conduction study (SCS). In a preceding study, the authors documented that the tibial nerve somatosensory evoked potentials (SEPs) that can evaluate the proximal part of the sensory nerve, which is preferentially involved in CIDP, were useful in the diagnosis of typical CIDP. This may also hold true for the pure sensory form.

Objectives: To investigate the utility of tibial nerve SEPs for the diagnosis of sensory CIDP.

Methods: The study included four patients who presented with sensory symptoms and signs of insidious onset, depressed or lost tendon reflexes, but reported no weakness. Patients with sensory ganglionitis were not included. Two patients had anti myelin associated glycoprotein (MAG) antibody. Tibial nerve SEPs were segmentally evalu-

ated using N8-P15-N21 potentials. The sensitivity of conventional diagnostic criteria for CIDP was also tested.

Results: The criteria of the European Federation of Neurological Societies/Peripheral Nerve Society were satisfied in two MAG positive patients, mainly due to prolonged distal motor latencies. Two MAG negative patients were near normal in motor nerve conduction. SCS gave no definite evidence of demyelination for every patient. Tibial nerve SEPs of all four patients presented “proximal dominant findings”, which was defined in a previous study and was found to be sufficiently specific for CIDP.

Conclusions: Tibial nerve SEPs are useful in diagnosing sensory CIDP that is often difficult to diagnose by nerve conduction. Even MAG positive patients, who are typically believed to have definite distal predilection, presented a marked delay in the nerve roots.

NEUROMUSCULAR AND AUTONOMIC FEATURES OF VOLTAGE GATED POTASSIUM CHANNEL ANTIBODY

A. Pande, E. Murray, V. Vedanarayanan, A. Leis (Jackson, MS)

Introduction: The spectrum of neurological manifestations associated with voltage gated potassium channel (VGKC) autoimmunity is broad. In most cases, clinical features are attributed to brain dysfunction. Neuromuscular (NM) manifestations and autonomic nervous system (ANS) dysfunction are less common.

Objective: To describe five patients with VGKC antibodies in whom neuromuscular or ANS manifestations were the predominant features.

Methods: VGKC antibody testing was performed by Mayo Medical Laboratories in Rochester, Minnesota.

Case Reports: Case 1: A 17-year-old boy with recurrent painful muscle cramps, supraventricular tachycardia, and hyperhidrosis. The painful

cramps improved with intravenous steroids.

Case 2: A 69-year-old woman with progressive multifocal peripheral neuropathy and idiopathic gastroparesis requiring a gastric pacemaker.

Case 3: A 31-year-old woman with idiopathic gastroparesis (which required a gastric pacemaker), neurogenic bladder, and intermittent ptosis, double vision, and generalized weakness of undetermined etiology.

Case 4: A 23-year-old woman who satisfies original and modified El Escorial World Federation of Neurology criteria for the diagnosis of amyotrophic lateral sclerosis (ALS), except for autoimmunity.

Case 5: An 81-year-old man with a subacute progressive disorder suggesting ALS, including upper and lower motor neuron involvement and widespread denervation on electromyography. However, definitive diagnosis is confounded by underlying diabetes, advanced degenerative disc disease, and autoimmunity.

Conclusions: Patients with VGKC antibodies can present with NM or autonomic manifestations in the absence of brain dysfunction. Evaluation for VGKC antibodies should be considered in cases of idiopathic autonomic dysfunction and ALS.

CLINICAL SPECTRUM OF VOLTAGE GATED POTASSIUM CHANNEL ANTIBODY ASSOCIATED PERIPHERAL DISORDERS

C.N. Fournier, H.R. Jones, G. Abel, M. Rhoads, J. Srinivasan (Burlington, MA)

Introduction: Voltage gated potassium channel (VGKC) autoimmunity is associated with peripheral nerve system (PNS) involvement in 25% of cases. The authors present five patients with PNS disease and VGKC positivity.

Case Reports: Patient 1 is a 74-year-old male that has experienced weakness for over 3 years. He had a history of adenocarcinoma in the groin from an unknown primary cancer. Examination revealed distal greater than proximal weakness and reduced sensation in upper and lower extremities (LEs). Electromyography (EMG) revealed severe demy-

elinating polyneuropathy (PN). He declined treatment. Four years later, he died unexpectedly.

Patient 2 is a 45-year-old female with numbness in feet and bilateral foot drop that developed over 3 years. Examination demonstrated distal LE weakness and sensory loss. EMG demonstrated moderate length dependent axonal PN. The workup for malignancy was negative. She improved with intravenous immunoglobulin IVIg.

Patient 3 is 61-year-old female that presented with painful spasms, numbness, and gait unsteadiness. Paresthesias were present for 20 years, and painful spasms were present for 3 years. Examination revealed distal LE weakness and sensory loss, with fasciculations and myokymia. EMG demonstrated severe axonal PN; neuromyotonia was not seen. Work up for malignancy was negative. Isaac's syndrome was diagnosed. Transient improvement was seen with IVIg.

Patient 4 is an 87-year-old male with bulbar myasthenia gravis (MG). Patient 5 is a 66-year-old male with ocular MG. Acetylcholine receptor, striated muscle, and VGKC antibodies were positive. Chest computed tomography was negative. The patients were stabilized while managed with prednisone and mycophenolate.

Conclusion: Potentially treatable polyneuropathies that are associated with VGKC autoimmunity have a phenotypically varied presentation. VGKC antibodies may also be seen in seropositive MG.

ATYPICAL ELECTRODIAGNOSTIC FEATURES IN POEMS SYNDROME

A.S. Wu (Valhalla, NY)

Introduction: Electrodiagnostic (EDX) features of the polyneuropathy in polyneuropathy, organomegaly, endocrinopathy, M-protein, and skin changes (POEMS) syndrome have been characterized as mixed demyelinating and axonal. Due to similar EDX features in POEMS and chronic inflammatory demyelinating polyneuropathy (CIDP),

studies comparing EDX findings between the two have been previously conducted. The pattern of nerve conduction abnormalities has been found to differ between the two disorders. Therefore, recognition of typical EDX features in POEMS will be helpful for early diagnosis. However, this case reports a patient with diagnosed POEMS and Castleman disease that presented with EDX features that lacked typical patterns and remained indistinguishable from CIDP.

Case Report: Nerve conduction study of an Asian woman with a clinically diagnosed POEMS syndrome (polyneuropathy, IgA gammopathy, hypothyroidism, lymph nodes enlargement, skin changes) with Castleman disease revealed a primary demyelinating polyneuropathy with prolonged distal motor latencies in both upper extremities (UEs) and lower extremities (LEs), normal compound motor action potentials, slowing conduction velocities (30s in UE and 20s in LE), temporal dispersion, and significantly prolonged F waves. It did not show the typical patterns in POEMS, as reported in previous studies.

Conclusion: It is hoped awareness will be raised regarding the recognition of various EDX features of POEMS syndrome, which could be indistinct from CIDP. Caution should be taken when making a diagnosis of POEMS versus CIDP based on the typical EDX features in previously published studies. Diagnosis of POEMS syndrome should remain primarily clinical.

DIAGNOSIS OF HEREDITARY NEUROPATHY IN TYPE I DIABETES MELLITUS

A.S. Wu (Valhalla, NY)

Introduction: A 47-year-old man with type I diabetes mellitus (DM) since age 21 is now found to have a primary demyelinating hereditary neuropathy.

Case Report: A 47-year-old man with type I DM reported slowly progressive weakness, numbness, and tingling in his legs. He reported deformity in his feet and atrophic calf muscles while grow-

ing up. The man was never active in sports, and was unable to curl or spread his toes. Type 1 DM was diagnosed at age of 21 and was treated with insulin. He developed foot drop bilaterally when he was 30. Two years later, he had a severe left foot ulcer which resulted in the amputation of the first four toes. On his neurologic examination, there was distal muscle atrophy, weakness, sensory abnormality, and diminished reflexes. His nerve conduction study showed a unique primary demyelinating neuropathy with uniformly slow conduction velocities (10 to 30m/s) in the upper extremities (absent lower extremity responses). Because of conduction block in the forearm segment of the ulnar nerve, studies suggested possible superimposed chronic inflammatory demyelinating polyneuropathy. Further bloodwork and cerebrospinal fluid (CSF) study were performed (CSF protein 52). Genetic testing for demyelinating Charcot Marie Tooth revealed four DNA sequence alternations in four genes (EGR2, LITAF, PRX, SH3TC2).

Conclusion: This clinical picture best represents the diagnosis of a primary demyelinating hereditary neuropathy with a secondary diabetic neuropathy. Recognition and awareness is needed in regards to hereditary neuropathy in type I DM and conduction block in hereditary neuropathy.

PERIPHERAL DEMYELINATING NEUROPATHY IN MULTIPLE SCLEROSIS

J.K. Baruah, G.R. Baruah (Milwaukee, WI)

Introduction: Peripheral neuropathy (PN) may be noted in some longstanding cases of multiple sclerosis (MS), irrespective of the given immunomodulation treatment. This report describes 12 patients with longstanding MS with PN, and their response to methylprednisolone (MP) intravenous immunoglobulin (IVIg).

Methods: Twelve patients (8 female; 4 male) with MS developed progressive weakness and were subsequently noted to have demyelinating PN on electrophysiologic evaluation. Prior to evaluation, each patient ambulated with use of a cane or walk-

er. As the weakness and numbness progressed, the tendon reflexes were reduced with decreased peripheral sensation. Each received 1000 mg of intravenous (IV) MP for 6 days, and intravenous immunoglobulin (IVIg) (0.4gm/kg body weight) for 5 days. Each patient then received maintenance IVIg infusions once a month for 6 to 8 months.

Results: The electromyography parameters were suggestive of demyelinating PN. Each patient responded well to treatment and in about 5 months, the patients returned to premorbid state. The spinal fluid studies revealed increased protein with normal cell count.

Conclusions: Demyelinating PN may appear in longstanding cases of MS. Aggressive treatment with MP and IVIg proved to be helpful. It is important to rule out other causes of PN such as B-12 deficiency, and vasculitis.

BOTULINUM TOXIN IN HEMIFACIAL SPASM

J.K. Baruah, G.R. Baruah (Milwaukee, WI)

Objective: To determine if there is modification in typical electrophysiologic (EP) findings in hemifacial spasm (HFS) with long term use of botulinum toxin treatment (BTX).

Methods: Eighteen patients (10 female; 8 male) were periodically treated initially at 4 to 6 month intervals with BTX for HFS. Each patient had facial nerve conduction, blink reflex (BR), and electromyography studies before and after in 6 to 8 weeks following BTX. The BTX was injected in frontalis, orb oculi, nasalis, zygomaticus, platysma, mentalis, and buccal pad of fat on HFS side.

Results: The patients studied tolerated BTX well and responded adequately. The facial nerve conduction parameters were normal in the orb oris/oculi. In 13 of 18 patients, R2 response of BR was absent (N=8) or reduced (N=5) in ipsilateral orb oris following supraorbital stimulation on HFS side after 3 to 4 years of BTX treatment.

Conclusions: The study concluded that in some patients with HFS, the BTX treatment had a tendency to improve EP parameters by modifying the excitability of nerve or suppressing the ephaptic discharge and thereby reducing the frequency of periodic BTS treatment.

ISOLATED MEDIAL ANTEBRACHIAL CUTANEOUS SENSORY NEUROPATHY STATUS POST BRACHIOPLASTY

W.S. Baek (Fontana, CA)

Introduction: A 54-year-old female underwent right arm brachioplasty 2 years ago, which was complicated by diffuse swelling, bruising, and pain.

Methods: Since undergoing a surgical revision 1 year following brachioplasty, she has had constant right medial forearm pain and dysesthesias starting proximal to the wrist, extending to medial upper arm distal to axilla. She denied any weakness. On examination there was mild weakness in the right deltoid, biceps, triceps, first dorsal interosseous (FDI), abductor digiti minimi, and abductor pollicis brevis.

Results: Nerve conduction studies revealed an absent right medial antebrachial cutaneous (MABC) response. The left MABC amplitude was 71uV. The right superficial radial sensory, median motor, and ulnar motor responses were all normal. Electromyography showed mild chronic neurogenic changes without denervation in the deltoid, biceps, triceps, flexor carpi radialis, extensor indicis proprius, and FDI. Magnetic resonance imaging showed C3-7 mild disc desiccation. Lidocaine patches and gabapentin improved her symptoms.

Conclusions: Isolated MABC neuropathy is rare. In cadaveric studies, 5 percent of all cases of standard brachioplasty resulted in injury to the MABC nerve. This penetrates the deep fascia 14 cm proximal to the medial epicondyle, lies within the plane of standard dissection, and is therefore at risk of injury. The asymmetric MABC responses were striking; com-

pletely absent on the affected side (right) and well within normal limits on the non affected side (left). An MABC neuropathy should be widely recognized as a complication of standard brachioplasty.

NITROUS OXIDE INDUCED SEVERE SENSORIMOTOR POLYNEUROPATHY WITHOUT PERNICIOUS OR MEGALOBLASTIC ANEMIA

W.S. Baek (Fontana, CA)

Introduction: Nitrous oxide (N₂O) was discovered in 1776 and was later used as anesthesia.

The neurological manifestations of N₂O are well described. Pernicious and megaloblastic anemia are common. The authors present a case of isolated N₂O induced neuropathy without anemia.

Case Report: A 20-year-old previously healthy male reported painful paresthesias in legs and foot drop for 6 months. He had distal arm painless paresthesias without weakness, which resolved 2 months ago. The patient failed the Marine Corps physical. Since the end of 2006, he inhaled 50, 9-inch balloons full of N₂O every Friday and Saturday. This practice ceased a month ago. Examination showed mild distal weakness in both legs with absent proprioception and vibration in feet and diffuse areflexia. Hemoglobin was 15 and motor conduction velocity was 92. Thyroid stimulating hormone, human immunodeficiency virus, rapid plasma reagin, and lead were all normal or negative. B12 levels were 139, methylmalonate levels were 0.60, and homocysteine levels were 36. Nerve conduction studies demonstrated absent sural sensory, peroneal and tibial motor responses with borderline superficial radial sensory amplitude and mild slowing. The ulnar motor response was unremarkable. Vitamin B12 injections and a methionine rich diet were initiated. He was lost to follow up.

Conclusions: Cobalamin is required for generating succinyl coenzyme A from methylmalonic acid and converting homocysteine to methionine. N₂O

deactivates cobalamin, thus decreasing methionine and s-adenosyl methionine levels, which are essential for cellular repair. Despite having completely absent sensorimotor responses in the leg, his complete blood count did not reveal any evidence of pernicious or megaloblastic anemia. The authors conclude that N2O can cause a severe isolated axonal sensorimotor polyneuropathy.

THE CLINICAL, ELECTROPHYSIOLOGICAL OF GUILLAIN-BARRE SYNDROME IN HONG KONG

C. Lee, T. Tsoi, C. Cheung, M. Au Yeung, R. Li, E. Yeung (Hong Kong)

Introduction: The clinical and electrophysiological features of Guillain-Barre Syndrome (GBS) are not well studied in Hong Kong.

Methods: A retrospective study of GBS patients was conducted from January of 2006 to December of 2009. The demographic data, clinical course, and electrophysiological findings were retrieved. The electrophysiological classification was defined according to the Ho and colleagues criteria.

Results: Thirty-one patients with GBS and its variants were identified. Twenty-four patients had acute inflammatory demyelinating polyradiculopathy. Six patients had a Miller-Fisher variant, confirmed by positive GQ1b antibody. One axonal variant was found. The mean age was 59 years (range 19 to 87). Twenty patients (64.5%) had preceding events of either enteritis or respiratory tract symptoms. Weakness was the most common symptom upon presentation (96.8%), followed by numbness (38.7%), facial weakness (22.6%), and ophthalmoplegia (22.6%). Cerebrospinal fluid protein was elevated in 15 of 21 patients (average 6.5 days from symptom onset). All patients underwent nerve conduction studies (average 7.3 days from symptom onset). Demyelinating neuropathy was demonstrated in 23 patients (74.2%). Prolonged or absent F wave response (67.7%) and increase in motor distal latency (61.3%) were the most common findings. Twenty-four patients re-

ceived intravenous immunoglobulin (IVIg) alone. Four patients received IVIg and plasma exchange due to disease progression. Two patients received no immunotherapy in view of mild impairment. The mean length of hospitalization was 42 days (range: 6 to 220). One patient died of respiratory complication. Eighteen patients (58.1%) had >1 score improvement in the modified Rankin scale upon discharge.

Conclusions: The pattern of GBS in Hong Kong is similar to that in Western countries, and has comparable outcomes.

CLINICAL COURSE OF PATIENTS WITH MYASTHENIA GRAVIS IN HONG KONG

E. Yeung, C. Lee, T. Tsoi, M. Au Yeung, R. Li, C. Cheung (Hong Kong)

Introduction: As a result of advanced treatment options, the prognosis for patients with myasthenia gravis (MG) has much improved in Western countries over past decades. However, there is a scarcity of data related to cases in Hong Kong.

Method: The authors conducted a retrospective review of patients with MG who experienced disease onset after 2000. The demographic data, clinical characteristics, and outcome information were obtained from written medical records and Clinical Management System (CMS).

Results: Eighty eight patients were included in the review. The mean age at onset was 54.2 years. Of those, 55 (63%) had pure ocular symptoms, 24 (27%) had generalized myasthenia at onset, 9 (10%) had ocular onset followed by secondary generalization. Thymic abnormality was diagnosed by computerized tomography in ten patients (nine thymoma, one hyperplasia). Thymectomy was performed in nine patients (five for thymoma, one for hyperplasia, three for normal thymus). Prednisolone and azathioprine were used in 38.6% and 28%, respectively, predominantly in generalized cases. Two refractory cases were treated with mycophenolate. Twenty-three patients experienced

significant deterioration in myasthenic symptoms requiring hospitalization, with a cumulative 671 days in the hospital. Fourteen patients developed myasthenic crisis, and a total of 21 courses of intravenous immunoglobulin (IVIg) was administered. Two patients did not respond to IVIg and required seven courses of plasmapheresis. All cause mortality was 9% (8/88) after an average follow-up of 5 years, with four deaths from each group. In the ocular group, none were disease related. Among the generalized group, two died of respiratory failure, and one from severe sepsis. The overall disease related mortality rate was 3.4%.

Conclusion: The outcome of local MG patients is comparable to that in Western countries.

GUILLAIN-BARRE SYNDROME AFTER GARDASIL VACCINATION IN THE UNITED STATES

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Introduction: Several reports have pointed to an unusually high number of neurological manifestations following Gardasil vaccination administration.

Objectives: The authors will determine the reporting rates and characteristics of Guillain-Barré syndrome (GBS) following the administration of Gardasil in the United States.

Methods: The authors used data derived from the Vaccine Adverse Event Reporting System. A neuromuscular specialist reviewed all physician reported possible events following vaccination to identify GBS cases after Gardasil vaccination between June of 2006 and September of 2009. Crude weekly reporting rate estimates were calculated and compared with GBS cases related to meningococcal and influenza vaccines in the general population.

Results: Sixty-nine cases of GBS were identified after Gardasil vaccination administration over a 3.3

year period. The onset occurred within 6 weeks after vaccination in 70% of patients in whom the date of vaccination was known. Hospitalization was reported in 42 (61%), and disability in 12 (17%) patients. GBS occurred in 47 (68%) cases after the administration of a single vaccine, and in 18 (26%) cases after Gardasil was administered, in combination with meningococcal vaccine. The estimated weekly reporting rate of post Gardasil GBS within the first 6 weeks (6.6 per 10,000,000) was higher than that of the general population (0.65 to 2.5 per 10,000,000), post meningococcal (3 per 10,000,000), and influenza vaccination (1.3 per 10,000,000).

Conclusions: These findings suggest a prominently high weekly reporting rate of GBS within 6 weeks following Gardasil vaccine administration. Due to possible underestimation incurred during passive surveillance ascertainment, a prospective active surveillance for accurate ascertainment and identification of high risk groups is warranted.

THE EXPANDING INDICATIONS FOR NERVE TRANSFERS

J.M. Brown (St. Louis, MO)

Introduction: Nerve transfers were originally used in cases of brachial plexus injury where, as a result of nerve root avulsions, no proximal axon donor was available to restore a particular function. Over the past decade, nerve transfers have proven to be an effective intervention in numerous cases of nerve injury, often producing results which are more rapid and robust than that accomplished through traditional grafting. Currently, nerve transfers are being applied in situations which previously would not have been considered indications for nerve reconstruction.

Case Reports: The authors present three cases in which nerve transfers were performed. First, a case of nerve transfers from the median nerve for the restoration of wrist and finger extension due to a radial nerve injury is presented. Second, a case of cervical spondylosis which resulted in a flaccid hand which

was treated with transfers from the proximal musculocutaneous and radial nerves to recover hand function is reviewed. Finally, a case of a chronic C7 ASIA A spinal cord injury in which ulnar intrinsic function was restored to a muscle group within the “intact” sublesional spinal cord segment.

Conclusion: Nerve transfers offer the potential for restoring function in a number of neurological conditions formerly considered cases for braces or assistive devices. If implemented effectively and in a timely fashion, recovery of effective hand function can be achieved.

NEURALGIC AMYOTROPHY: FIVE CASES WITH A DIVERSE PRESENTATION

R.R. Conway (Columbia, MO)

Introduction: Neuralgic amyotrophy is an uncommon condition characterized by the sudden onset of acute severe pain, most frequently in the shoulder girdle or other areas of the upper extremity followed within 2 weeks by weakness and sometimes sensory loss. It is idiopathic with a suspected autoimmune etiology. Some believe it primarily involves the brachial plexus, but as originally described by Parsonage and Turner and later by England and Sumner, the disorder may involve a variety of peripheral nerves.

Case Study: The author presents five cases with clinical presentations consistent with neuralgic amyotrophy with mononeuropathies documented by electrodiagnostic studies. Two patients had suprascapular neuropathy, two had anterior interosseous neuropathy, and the fifth patient had suprascapular and axillary neuropathies.

Conclusion: It is important to remember that although neuralgic amyotrophy may present as a brachial plexopathy, the presentation is often more diverse. Early recognition is important since recent studies have confirmed the benefit of early corticosteroids in the treatment of this condition.

SCIATIC NEUROPATHY AS A COMPLICATION OF ULTRASOUND GUIDED POPLITEAL BLOCK

R.R. Conway (Columbia, MO)

Introduction: Popliteal sciatic nerve block has become popular for anesthesia in foot and ankle surgeries. While it is difficult to find information on the incidence of sciatic neuropathy as a complication of these procedures, it is generally reported as rare, with one prospective study reporting an incidence of 0.5%. Recently, ultrasound (US) guidance of these procedures has been proposed to increase the efficacy of the injections and further decrease the rate of complications.

Case Study: A 67-year-old white female underwent an US guided popliteal sciatic nerve block for an open tibiotalar arthrodesis which was complicated by an incomplete sciatic neuropathy with severe axonotemesis. The findings are documented with electromyography and nerve conduction studies.

Conclusions: While US guided popliteal sciatic nerve block is considered to have few complications, such complications do exist and can be disabling.

PARANEOPLASTIC SENSORY NEUROPATHY AND GANGLIONIC ACETYLCHOLINE RECEPTOR ANTIBODY

U.K. Dhand, K. Sahaya, N.N. Singh (Columbia, MO)

Introduction: Sensory neuropathy or neuronopathy in patients with small cell lung cancer (SCLC) is among the classical paraneoplastic syndrome that is typically associated with anti-Hu antibody or CRMP-5 antibody. Association with ganglionic neuronal acetylcholine receptor (AChR) antibody has not been described.

Case Report: A 62-year-old woman developed numbness and paresthesia of hands, feet and tongue, followed by weight loss, nausea, and hyponatremia. Ten months later, she was diagnosed with SCLC and was treated with radiation, etoposide, and carbopla-

tin. She was evaluated in a neurology department for continued numbness and pain in her hands and feet, and a feeling of her tongue being numb and swollen. An examination revealed pain and touch sensory impairment over feet and fingers, altered feeling over the tongue (preserved taste sensation), and impaired vibration at the toes and malleoli. Electrodiagnostic study showed markedly reduced sensory nerve action potential amplitude of median, ulnar and sural nerves, normal motor nerve conduction, and mild distal chronic neurogenic changes. Routine blood counts and chemistry, serum B12, thyroid function, serum protein electrophoresis, antinuclear antibodies (including SS-A, SS-B) were normal. Paraneoplastic autoantibody profile was positive for ganglionic AChR antibody (0.06 nmol/L, normal <0.02). A follow up 4 months after cancer treatment showed no worsening of neuropathy nor any autonomic symptoms.

Conclusion: The patient presented had a non length dependent pattern of sensory neuropathy antedating the diagnosis of SCLC but consistent with paraneoplastic sensory neuronopathy, along with an unusual finding of positive ganglionic AChR antibody. The ganglionic AChR antibody is associated with autoimmune autonomic neuropathy, and has also been considered a putative onconeural antibody; however, its direct role in the pathogenesis of paraneoplastic sensory neuropathy is uncertain.

MONONEURITIS MULTIPLEX AS A RESULT OF ACUTE HEPATITIS B RELATED VASCULITIS

M.F. Hangan, J. Li (Valhalla, NY)

Introduction: Mononeuritis multiplex (MM) preceded by acute Hepatitis B virus (HBV) related seronegative polyarteritis nodosa (PAN) vasculitis is a rare clinical occurrence, with only a few reported cases.

PAN is a necrotizing vasculitis of small and medium arteries that can develop within 6 months of acute HBV infection. HBV-PAN patients may have negative anti neutrophilic cytoplasmic antibodies (ANCA). MM is a painful, asymmetric asynchronous, sensory motor peripheral axonal neuropathy involving isolated dam-

age to at least two separate nerves. The mechanism of axonal destruction is hypothesized as direct action of the HBV or vasculitis of the vasa nervorum triggered by deposits of immune complexes.

Case Report: A 62-year-old man who is right handed, presented with progressive pain and numbness in his hands and feet, followed by left foot drop and atrophy. Six weeks prior, he had acute HBV infection. Serology revealed positive HB surface and HBe antigen, also HB core antibodies and two million copies of HBV-DNA. Other hepatitis viruses were negative. He had nonreactive serum antibodies to ANCA. Nerve conduction study and needle electromyography revealed mononeuritis multiplex involving left sciatic, left radial and ulnar nerves with evidence of axonal loss. Sural nerve biopsy confirmed PAN. Polymerase chain reaction for HBV-DNA was negative following antiviral treatment. Plasmapheresis and pulse steroids improved the muscle strength and neuropathic pain.

Conclusion: MM caused by acute HBV-related PAN represents a diagnostic and therapeutic challenge as HBV-PAN patients have non reactive serum antibodies to ANCA. Immunosuppressants comprise the treatment choices.

Mihaela F. Hangan, MD

Junior Member Recognition Award Recipient

PROGNOSTIC DETERMINANTS IN DEMYELINATING NEUROPATHY WITH ANTI MYELIN ASSOCIATED GLYCOPROTEIN ANTIBODIES

G.D. Meekins, B.J. Distad, M.D. Weiss (Seattle, WA)

Introduction: Although demyelinating neuropathy with antibodies to myelin associated glycoprotein (MAG) is often considered to be a form of chronic inflammatory demyelinating polyradiculoneuropathy, it has a much less predictable response to treatment.

Objective: To determine whether the clinical and electrophysiological presentation of anti-MAG neu-

ropathy can determine prognosis in regards to treatment with rituximab and other immunomodulatory therapies, and to discover if the electrophysiological findings correlate with the clinical presentation.

Methods: Eleven patients were evaluated with comparisons made between clinical presentation, treatment response, and electrophysiological findings that included terminal latency index (TLI). Comparisons between the groups were made by unpaired student's t-test.

Results: Mean peroneal compound muscle action potential (CMAP) amplitude of treatment responders was significantly greater than that of non-responders ($1.3 \text{ mV} \pm 1.34$ vs $0.3 \text{ mV} \pm 0.05$, $p < 0.05$). In part, this is due to motor nerve inexcitability of the lower extremities (LEs) in the majority of non responders (peroneal or posterior tibial or both in 4/4 non responders versus 1/6 responders). The mean peroneal CMAP amplitude was greater in patients with a duration of illness less than 6 years, relative to those with a longer duration ($1.3 \text{ mV} \pm 1.4$ vs $0.1 \text{ mV} \pm 0.2$), though this was not particularly significant ($p = 0.069$). TLI and distal motor latency for all nerves tested failed to demonstrate any correlation with treatment response, duration of illness, or other parameters.

Conclusions: The evolution of the disease over long periods of time appears to be associated with inexcitable motor nerves in the LEs and a poorer response to therapy. Early diagnosis of this disorder is warranted, as delaying treatment may prevent a favorable response.

MOTOR UNIT NUMBER AND MOTOR UNIT ACTION POTENTIAL IN CARPAL TUNNEL SYNDROME

M. Sohn, S. Hwang, H. Shin (Daejeon, Korea)

Introduction: To evaluate the clinical significance of motor unit number estimation (MUNE) and quantitative analysis of motor unit action potential (MUAP) analysis in carpal tunnel syndrome (CTS) according to severity of nerve conduction studies

(NCSs), ultrasonographic (US) measurement and clinical symptoms.

Methods: Electrophysiologic studies and US studies were performed on 54 wrists of 31 patients diagnosed with CTS and compared to the findings on 40 wrists of 20 healthy controls. Median NCSs, amplitude and duration of MUAP, MUNE with incremental technique on the abductor pollicis brevis (APB) were measured. The cross sectional area (CSA) of the median nerve at the pisiform and distal radio ulnar joint level was determined by high resolution US.

Results: MUNE of the APB was significantly lower in the CTS group and had a negative correlation with electrophysiologic stage, amplitude and duration of MUAP, CSA at pisiform level, and Boston Carpal Tunnel Questionnaire (BCTQ). The amplitude and duration of MUAP were significantly higher in the CTS group, yet had a negative correlation with the electrophysiologic stage, however, no relationship with CSA and BCTQ was noted.

Conclusions: The authors suggest that MUNE serves as a good indicator of motor reserve and the severity of CTS. Therefore, MUNE could be helpful in the follow up of patients with CTS after treatment in clinical practice.

DIABETIC LUMBOSACRAL RADICULOPLEXUS NEUROPATHY: CASE REPORT OF NERVE BIOPSY AND POSTMORTEM FINDINGS

D. Younger, A.P. Hays (New York, NY)

Introduction: An autoimmune vascular ischemic etiopathogenesis has been suspected in diabetic lumbosacral radiculoplexus neuropathy (DLRPN). However, postmortem evaluation has been lacking. A case describing the clinicopathological features of a patient with DLRPN is presented.

Case Report: A 59-year-old man with diabetes mellitus was treated for 15 years with oral hypoglycemics. In January of 1995, he noted left thigh pain and par-

esthesia, followed by sensory changes and weakness in one leg, then in the other. In May of 1995, he was placed on insulin and had onset of weakness leading to quadriplegia. Examinations showed wasting and flail weakness in the legs, stocking and glove sensory loss, and areflexia. He was treated with intravenous corticosteroids, intravenous immune globulin, and cyclophosphamide for vasculitis. This was followed by acute renal failure, leucopenia, acute myocardial infarction, coma, and death.

Laboratory studies showed: erythrocyte sedimentation rate 60 mm/hour; glucose 250 mg/dl, cerebrospinal fluid protein 122 mg/d, with 1 white blood count/mm³. Electromyography and nerve conduction study showed severe mixed axonal and demyelinating polyneuropathy. Sural nerve biopsy showed vasculitis (see figures). General autopsy of systemic organs and brain failed to demonstrate vasculitis, however, specimens of femoral nerve, sciatic nerve, and lumbar plexus showed chronic inflammation in the epineurium, perineurium, and endoneurium (see figures).

Conclusion: The findings were consistent with a vascular ischemic etiopathogenesis of DLRPN related to microscopic vasculitis. Axonal degeneration was the preponderant finding on electrodiagnostic (EDX) studies, and was likely related to the primary ischemic event. The demyelinating changes on EDX studies were considered secondary to axonal degeneration.

MULTIFOCAL MOTOR NEUROPATHY: THE CLINICAL AND ELECTROPHYSIOLOGIC FEATURES

B. Kang, K. Kim, Y. Lim (Seoul, Korea)

Introduction: Multifocal motor neuropathy (MMN) is a very rare and chronic demyelinating neuropathy. Controversy surrounds the diagnosis, pathogenesis, and treatment of MMN.

Objectives: The authors will describe the clinical and electrophysiologic features and responses to therapy in five patients.

Methods: Five patients with MMN, who fulfilled the European Federation of Neurological Societies/Peripheral Nerve Society criteria, were reviewed. Clinical and laboratory findings, including electrophysiologic characteristics and anti-GM1 antibody, were analyzed.

Results: The age of onset ranged from 24 to 51 years. Disease duration lasted from 3 months to 7 years. The initial symptom was asymmetric hand weakness with progression to other distal limbs. Sensory loss was not observed. Four patients treated with intravenous immunoglobulin (IVIg) pulse therapy (3 to 5 days) demonstrated good response within 10 days. Three patients that were given IVIg monthly maintained the improvement of weakness. One patient, treated with IVIg bimonthly with a low dose azathioprine/prednisone combination therapy also had motor improvement. The average number of conduction blocks per patient was 2.8. All patients had conduction blocks (CBs) in upper limbs. Three patients had CBs in lower limbs. Other electrophysiological evidence of demyelinating features was not observed in motor nerve conduction studies. The IgM anti-GM1 antibody was positive in one patient (35.3 EU/ml).

Conclusions: MMN differs from idiopathic CIDP in clinical characteristics and in electrophysiological findings. Although the clinical and electrophysiological features of this study did not differ from other previous reports, anti-GM1 antibody was not frequently observed in the patients studied. Monthly IVIg therapy paired with combination immunosuppressants maintained improvements, suggesting major humeral immune mediated pathogenesis of MMN with very low effects of anti-GM1 antibody.

CLINICAL FEATURES OF 45 PATIENTS WITH TRUE NEUROGENIC THORACIC OUTLET SYNDROME

B.E. Tsao, R.W. Shields, Jr (Loma Linda, CA, Cleveland, OH*)*

Introduction: True neurogenic thoracic outlet syndrome (N-TOS) is a rare disorder affecting the lower trunk of the brachial plexus.

Objectives: To compare the clinical characteristics of 35 patients with electrodiagnostic evidence of true N-TOS who underwent surgical treatment to 10 patients who were managed conservatively.

Methods: A retrospective record review was conducted.

Results: In both groups, patients were predominantly female, were of similar age ranges, and experienced right sided involvement unrelated to handedness. Weakness, pain, and sensory loss were present in nearly all patients at diagnosis after an average duration of 55 months. Antecedent trauma was described in nearly 25% of participants. All patients had severe muscle atrophy and weakness of the median (T1>C8), more than ulnar or radial innervated muscles (C8>T1). A cervical rib or elongated transverse process was present in 83%, and was bilateral in 66%. Thirty-five underwent surgical resection of a congenital band or first thoracic rib. A band was found in nearly all patients, regardless of the presence of a cervical rib or elongated transverse process. Surgical outcome included improved pain (48%), improved pain and strength (21%), improved strength alone (10%), and no change in symptoms (21%). Patients who underwent surgery were more likely to experience improvement in strength and pain (80% surgical versus 33% non surgical).

Conclusions: True N-TOS is a rare disorder causing chronic progressive sensory and motor symptoms and signs in the C8/T1 myotomes and dermatomes. Surgical treatment, regardless of the anatomic abnormality noted prior to surgery, is effective in the majority of patients.

POST TRAUMATIC SUPERIOR GLUTEAL NEUROPATHY IN A BALLET DANCER

P. S. Hsu (Springfield, MA)

Introduction: The authors present a patient with a superior gluteal neuropathy after an assault with minor trauma. He complained of unsteadiness only while standing on his right leg and attempting to spin.

Case Report: A 64-year-old male ballet instructor presented with complaints of unsteadiness occurring immediately after an assault in which he was struck on the left side of his face and fell onto his right side. He suffered from soreness over the lateral aspect of his right hip which had resolved after a period of weeks. After the assault, he also experienced persistent instability in standing and spinning on his right leg.

Although unaware of obvious weakness in his leg or changes in his gait, he had clinical weakness of right hip abduction and internal rotation. He did not have Trendelenburg's sign or gait abnormalities. Electromyography performed over a year after the injury revealed chronic denervative changes limited to the gluteus medius and tensor fasciae latae, muscles innervated by the superior gluteal nerve.

Conclusions: This patient had a painless superior gluteal neuropathy presenting without gait abnormalities, but with the inability to spin on one leg following minimal trauma. No pelvic abnormalities were apparent on imaging. This case of a superior gluteal neuropathy is unique due to the subtlety of the patient's symptoms. It supports the need to consider an injury to the superior gluteal nerve in patients presenting in a similar manner, particularly in highly trained dancers.

THE INCIDENCE AND TIMING OF TRIGGERS AMONG 281 NEURALGIC AMYOTROPHY PATIENTS

M.A. Ferrante, A.J. Wilbourn (Memphis, T; Cleveland, OH*)*

Introduction: According to the literature, approximately 50% of neuralgic amyotrophy (NA), or Parsonage-Turner syndrome patients have an identifiable trigger. Most commonly, it is a flu like illness, although the incidence and distribution of triggers and the time between the trigger and NA onset are unclear. This information is important to consulting neurologists, especially when litigation is involved.

Objective: To determine the incidence, distribution, and timing of triggers associated with NA.

Methods: Over a 12 year period, the authors prospectively elicited a trigger history among NA patients studied in a given electromyography laboratory.

Results: Of 268 bouts of NA, a trigger preceded 196 (73%): operative or medical procedure (57%), recent illness (48%), excessive or unaccustomed activity (34%), closed trauma (19%), childbirth (13%), dental procedure (11%), vaccination (10%), and open trauma (4%). The time interval between the identified trigger and the onset of NA ranged from 1 to 28 days: 1 week (67%), 2 weeks (18%), 3 weeks (9%), and 4 weeks (6%).

Conclusions: In this series, the trigger incidence was 73%. The most common trigger was a recent operation or medical procedure, and the trigger occurred within 4 weeks of the NA bout, with roughly two-thirds occurring within the first week. When NA is triggered by an operative or medical procedure and is not recognized, the trigger may be mistakenly considered the cause of the neurologic abnormalities. To avoid misdirected lawsuits, it is important for consulting neurologists to recognize NA and to be familiar with its associated triggers.

RACEMOSE SPINAL NEUROCYSTICERCOSIS

H. Muqtadar, E. Vernier, I. Goldstein, N. Souayah (Newark, NJ)

Introduction: Spinal cord involvement is seen in 1 to 3 percent of neurocysticercosis cases. The racemose form of spinal neurocysticercosis is quite rare. The author presents a patient with racemose type of spinal neurocysticercosis causing progressive motor and sensory deficit.

Case Report: A 49-year-old Mexican man presented with 2 months of bilateral arm and leg numbness and weakness. Motor examination showed mild proximal and distal weakness in both upper extremities and mild left iliopsoas weak-

ness. Sensation was decreased to all modalities in both arms and legs. Deep tendon reflexes were brisk and Babinski sign was present on both sides. Magnetic resonance imaging of the cervical spine showed multiseptated cystic lesions in the upper cervical spinal canal that are compatible with racemose form of neurocysticercosis and diffuse meningeal enhancement. The patient underwent a suboccipital craniotomy and decompressive C1 laminectomy with partial evacuation of the cysts. After receiving 2 weeks of oral steroids combined with oral albendazole, he showed minimal clinical improvement.

Conclusion: The racemose neurocysticercosis constitutes a hydroptic change that leads to large or even giant vesicles usually devoid of a scolex. It is typically located in the basal cisterns and perisylvian cistern. Intra abdominal and intra thoracic pressure variation caused cysticerci larvae diffusion into the intradural-extramedullary spinal space by a retrograde larvae dissemination through the epidural venous plexus. Racemose spinal neurocysticercosis is more difficult to treat and is associated with significant neurological morbidity, as compared to a solitary cysticercosis granuloma.

Hurmina Muqtadar, MD

Junior Member Recognition Award Recipient

TURNS TO AMPLITUDE ANALYSIS IN RECURRENT LARYNGEAL MONONEUROPATHY

M.C. Munin, M. McCarty Statham, C.A. Rosen, S.D. Nandedkar** (Pittsburgh, PA; Cincinnati, OH*; Middleton, WI**)*

Introduction: A recent article from the Neurolaryngology Study Group called for the validation of quantitative laryngeal electromyography (LEMG) to better standardize methodology across medical centers and to improve diagnostic and prognostic accuracy.

Objectives: To develop normative data in controls for turns to amplitude analysis of the thyroarytenoid (TA) muscle and to compare results to

those of patients with subacute recurrent laryngeal nerve mononeuropathy.

Methods: LEMG of the TA muscle was performed in 21 controls and 16 patients with subacute recurrent laryngeal nerve mononeuropathy. Quantified turns and mean amplitude/turn were measured for greater than 10 epochs per individual. A linear scale cloud was constructed based on 90% of the normal data points falling within the cloud.

Results: In the control subjects, a normal cloud for the TA was delineated with a mean amplitude of 334 μ V and 450 mean turns per second. Analysis from patients showed a mean amplitude of 299 μ V and 290 mean turns per second. Seven patients with neuropathy all had data points with fewer than 400 turns per second, yet few patients had data points outside the normal cloud. Mean turns were statistically different from controls ($p=0.002$), but the mean amplitude was not statistically different.

Conclusion: This study is one of the first to present quantitative turns to amplitude analysis of the TA muscle from LEMG in controls and patients with recurrent laryngeal mononeuropathy. In patients with unilateral vocal fold paralysis, the authors found a significantly decreased number of turns during a range of phonatory efforts compared to controls.

MITOCHONDRIAL NEUROGASTROINTESTINAL ENCEPHALOMYOPATHY LIKE PHENOTYPE DUE TO POLG1 MUTATIONS

E.L. Dimberg, L.C. Wong, M. Milone** (Jacksonville, FL; Houston, TX *; Rochester, MN**)*

Introduction: POLG1 is a nuclear gene that encodes for the catalytic subunit of the only mitochondrial DNA polymerase gamma essential for mtDNA replication. POLG1 mutations cause a wide variety of phenotypes. The authors report a patient with a POLG1 related disorder resembling mitochondrial neurogastrointestinal encephalomyopathy (MNGIE).

Case Report: A 50-year-old man presented with cognitive difficulties, progressive ptosis, lower extremity sensory loss, proximal weakness, persistent diarrhea, vomiting, and severe weight loss. On examination, he had tangential speech and word finding difficulty. He had severe nonfluctuating ptosis, ophthalmoparesis, bifacial and limb weakness and distal sensory loss. The patient was cachectic with diffuse muscle atrophy. Resting serum lactate was 2.7 mmol/L (nl <1.7). Serum creatine kinase was normal. Computerized tomography of the abdomen and pelvis, esophagogastroduodenoscopy, and colonoscopy were unremarkable. Electromyography demonstrated a mild proximal myopathy and length dependent sensorimotor peripheral neuropathy. Magnetic resonance imaging of the brain showed diffuse cerebral and cerebellar vermian atrophy, but no leukoencephalopathy. Biceps brachii muscle biopsy revealed a few ragged red and ragged blue fibers and several cytochrome c oxidase negative fibers, suggestive of a mitochondrial myopathy. Sequencing of the thymidine phosphorylase (TYMP) gene detected no deleterious mutations, and the plasma thymidine level was normal. Sequencing analysis of the POLG1 gene detected two heterozygous mutations, p. W74BS and p. R953C.

Conclusion: This case underscores the need for sequencing POLG1 in patients with MNGIE like phenotype without TYMP mutations and the overlapping of clinical phenotypes in mitochondrial disorders with different molecular defects.

MITOCHONDRIAL DISORDER DUE TO NOVEL POLG1 SPLICE SITE MUTATION

M. Milone, T. Liewluck, J. Wang, J.A. Leavitt, L. Wong* (Rochester, MN; Houston, TX*)*

Introduction: POLG1 is a nuclear gene encoding for the catalytic subunit of the only mitochondrial DNA (mtDNA) polymerase gamma, which is essential for mtDNA replication and repair. Mutations in POLG1 result in a wide spectrum of phenotypes.

Case Reports: Patient 1 (Pt1) is a 65-year-old man with dyschromatopsia since childhood, ptosis, ophthalmoparesis, generalized weakness, cataract, and optic atrophy. Patient 2 (Pt2) is a 63-year-old man with a similar phenotype, but with no optic atrophy. He also has levodopa responsive Parkinsonism and hypoacusia. Electromyography showed myopathic and neurogenic motor unit potentials in Pt1 and myopathic changes in Pt2. Resting lactate was normal in both patients; creatine kinase was increased 1.8 fold in Pt2. Brain magnetic resonance imaging (MRI) showed cortical chronic infarcts in Pt1, and diffuse cerebral atrophy in Pt2, who had normal MRI spectroscopy. Muscle biopsy revealed scattered ragged red fibers and numerous ragged blue and cytochrome c-oxidase negative fibers in both patients. Pt1 had multiple muscle mtDNA deletions and no mutations in the OPA1 gene. A novel mutation, c.3104+3A>T, affecting the splicing was found in compound heterozygous with p.F749S in Pt1 and with p.G848S in Pt2. There is ongoing analysis of transcripts arising from this novel mutation.

Conclusions: Clinical features of POLG1 mutations are heterogeneous. Although the optic atrophy would favor an OPA1-related autosomal dominant disorder, Pt1 has a recessive POLG1-disorder. Therefore, molecular analysis is imperative in determining the correct diagnosis for proper patient care and genetic counseling.

IMPORTANCE OF ELECTROMYOGRAPHIC ASSESSMENT OF PARASPINAL MUSCLES IN MYOPATHY: EVALUATION OF 78 PATIENTS

G. Dahani; M. Al-Hakami, M.A. Lakhair, A. Dahani*M. Khan** (Khamis Mushayt, Aseer, Saudi Arabia; Jamshoro, Sindh, Pakistan*, Karachi, Sindh, Pakistan**)*

Introduction: Paraspinal muscles are supplied by posterior ramus, while limb and abdominal muscles are supplied by anterior ramus. Voluntary contraction of paraspinal muscles is difficult, making the assessment of motor unit potentials difficult. For this reason, it is not being used routinely in myo-

pathies. However, in some situations like vacuolar myopathies, its value is essential.

Objective: To evaluate the importance of electromyography of paraspinal muscles in myopathy.

Methods: This is a prospective interventional study. Consent was taken per protocol. Patients referred for myopathy were included, while those with diabetes mellitus and an age over 50 were excluded from the study. Nerve conduction studies and needle examination were (concentric needle) performed according to American Association of Neuromuscular and Electrodiagnostic Medicine criteria for myopathy. Cervical paraspinal muscles were assessed and patients were encouraged to tighten neck muscles for motor unit assessment. The findings were confirmed by a qualified neurophysiologist. SPSS, version 16, was used for data interpretation and the results were expressed in percentages.

Results: Fifty-one females and 27 males were included in the study. Inflammatory myopathic findings were noted in 60 subjects (76.92%), and non-inflammatory in 18 (23.08%). In the inflammatory myopathy group, 13 (21.66%) showed fibrillation and positive sharp waves in paraspinal muscles, but had negative results in limbs. Among those with inflammatory myopathy, 9 (15%) and 3 (16.6%) in non-inflammatory showed myopathic motor units in paraspinal muscles but not in limb muscles

Conclusion: Paraspinal muscle electromyographic assessment is statistically helpful for further evaluation of myopathic disorders, particularly if limb muscle electrography does not show fibrillation potentials, positive sharp waves, and myopathic motor units, or has inconclusive findings. Further muscle biopsy is needed to correlate paraspinal muscle predilection or its early involvement in the disease process.

A REPORT OF HYDROXYCHLOROQUINE TOXICITY AND STEROID MYOPATHY MIMICKING SCIATICA

M.N. Diaz, P.S. Bajaj (Maywood, IL)

Introduction: A 67-year-old female patient took steroids for 2 decades for suspected dermatomyositis due to an initial presentation of rash and proximal muscle weakness, despite concurrent electrodiagnostic findings that were more consistent with steroid myopathy. When her symptoms worsened, steroids were increased and hydroxychloroquine was added for a presumed connective tissue disorder based on nonspecific autoimmune serology and biopsy showing type II muscle atrophy. She subsequently suffered multi system disorders including retinopathy, which have been recently attributed to hydroxychloroquine toxicity.

Case Report: This patient presented for evaluation of presumed sciatica, though lumbosacral magnetic resonance imaging was normal. Her symptoms included cramping in the left buttock and leg that worsened with exercise. Except for unobtainable left sural nerve study (consistent with her history of post chemotherapy peripheral neuropathy), distal nerve conduction studies were normal. Physical and electromyographic needle exams revealed bilateral pseudo hypertrophy in calves, decreased insertional activity in gluteus maximus, and tibialis anterior and medial gastrocnemius with diminished motor unit firing throughout the left lower extremity. There was no evidence of lumbosacral radiculopathy.

Conclusion: Findings were consistent with end stage muscle disease where muscle has been replaced by fat or fibrosis. A chart review revealed how the side effects of hydroxychloroquine had been interpreted as progressive autoimmune disease and thus warranted increased steroid use, exacerbating the patient's underlying myopathy. Interestingly, her worst pain corresponded to areas with the least amount of insertional activity (i.e., where muscle was preferentially damaged). This

is a case of severe myopathy mimicking sciatica. It also raises awareness of how the steroid hydroxychloroquine treatment cycle can cause significant morbidity when myopathy is misdiagnosed.

DUCHENNE MUSCULAR DYSTROPHY: PROLONGATION OF SURVIVAL BY NONINVASIVE RESPIRATORY INTERVENTIONS

B. Saulat, J. Bach, N. Souayah (Newark, NJ)

Introduction: The average age for survival of Duchene muscular dystrophy (DMD) patients without noninvasive ventilation (NIV) is between 16 and 19 years.

Objectives: To report survival outcomes of ventilator dependent DMD patients who use a NIV and mechanically assisted coughing (MAC) oximetry protocol.

Methods: A retrospective study of 128 patients who used NIV was conducted. End tidal carbon dioxide, oximetry, vital capacity (VC), maximum insufflation capacity, and CPFs were monitored. Nocturnal NIV was initiated for symptomatic hypoventilation. Survival was quantitated by the duration of need for continuous NIV support.

Results: The patients were given access to MAC/oximetry for a mean of 10.0 ± 6.1 years. With advancing disease, 98 full time users were 30.3 ± 6.1 years of age in December of 2009. Fifty-two patients were still alive. Seven of these were NIV dependent for 20 years with no ventilator free breathing ability, and 22 patients had VC of less than 250 ml. Twenty-three of the 98 required continuous long term NIV without hospitalization. There were a total of 46 deaths: 21 died of cardiac related issues, 16 of respiratory causes, and 9 of other etiology. Ten patients were lost to follow up. Twenty-two unweanable intubated patients were extubated to NIV and MAC. Eight of nine deaths of TIV users were tube related.

Conclusions: Continuous NIV with MAC/oximetry as needed can prolong life and eliminate a need for DMD patients to resort to tracheotomy for

DMD. Unweanable patients can be decanulated and extubated without resort to tracheotomy.

Bilal Saulat, MD,MPH

Junior Member Recognition Award Recipient

ELECTROMYOGRAPHY FINDINGS IN DIFFERENT MUSCLES IN INCLUSION BODY MYOSITIS: THE UTILITY OF THE FLEXOR DIGITORUM PROFUNDUS MUSCLE

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Introduction: Sporadic inclusion body myositis (IBM) is a chronic myopathy. However, the co-existence of neurogenic process has been argued based on the presence of so-called neurogenic changes in needle electromyography (EMG), such as high amplitude or long duration motor unit potentials (MUPs). For this reason, IBM patients may be misdiagnosed with amyotrophic lateral sclerosis (ALS). In IBM, forearm flexor muscles, specifically the flexor digitorum profundus (FDP) muscle, is severely affected. The authors have experienced that EMG in FDP is unambiguously myopathic, and thus is helpful to the diagnosis. However, previous studies have not investigated the comparison of EMG findings between different muscles.

Objectives: To assess the utility of EMG in FDP for the diagnosis of IBM by comparing EMG features from several muscles.

Methods: The authors retrospectively reviewed EMG records of 16 biopsy confirmed IBM patients. EMG findings were scored qualitatively regarding spontaneous and voluntary activities. In a limited number of subjects, quantitative MUP analysis of EMG was performed to confirm the validity of qualitative scoring.

Results: Findings in FDP patients (n=14), biceps brachii (BB) patients (n=13), and rectus femoris (RF) patients (n=8) were investigated. Low amplitude MUPs were observed in 57%, 0%, and 0% of

patients for FDP, BB, and RF, respectively, whereas high amplitude MUPs were observed in 21%, 62%, and 75% of patients, respectively. Quantitative analysis corresponded with the qualitative impression.

Conclusions: If the FDP is closely examined, myopathic features would become evident and misdiagnosis would be avoided. Furthermore, typical myopathic EMG findings in this most affected muscle suggest that IBM is a genuine myopathy.

CREATINE DEFICIENCY SYNDROMES: POTENTIALLY TREATABLE ENCEPHALOMYOPATHIES

A. Verma (Miami, FL)

Introduction: Creatine is synthesized in the liver in a two step enzymatic process. First, it is carried via blood to target organs, then transported using creatine transporter-1 into the tissues. Creatine deficiency syndromes related to each of the two enzymatic steps and creatine transporter-1 have been recently reported in patients with developmental encephalopathy.

Objective: To describe two cases of arginine guanine amidinotransferase (AGAT) deficiency and to review creatine deficiency syndromes.

Methods: Two siblings (ages 26 and 24 years) from a consanguineous parentage with AGAT deficiency with features of brain and muscle involvement were studied. Clinical, biochemical, electrophysiological, muscle pathology, brain magnetic resonance (MR) spectroscopy, and molecular genetic tests were performed. Similar cases in literature were reviewed.

Results: Both patients had severe cognitive and speech impairment and moderate proximal muscle weakness (Medical Research Council grade 4/5). Biopsy from left quadriceps muscle showed fiber size variation, occasional degenerating fibers, and slight atrophy of both type 1 and 2 fibers. Investigations revealed undetectable guanidinoacetate in plasma (normal 0.4 – 3.7 μ M) and urine (normal 80 – 656 μ M), the findings characteristic of AGAT de-

iciency. Brain MR spectroscopy showed about half of normal creatine levels. Molecular analysis of AGAT gene from peripheral lymphocytes revealed homozygous R169X nonsense mutation in both cases. Initiation of an oral creatine supplement (400mg/kg wt) resulted in dramatic improvement in muscle strength and led to some cognitive improvement.

Conclusions: Early detection of AGAT deficiency and creatine supplementation can potentially prevent encephalomyopathy. Creatine deficiency should be excluded in all patients presenting with developmental delay, with or without myopathy.

DERMATOMYOSITIS WITH FOCAL SCLERODERMA LIKE SKIN ATROPHY

Y. Chai, R. Khan, T.E. Bertorini (Memphis, TN)

Introduction: Dermatomyositis (DM) manifests with proximal muscle weakness and a characteristic skin rash. Scleroderma-like disorders comprise a wide spectrum of heterogeneous diseases characterized by sclerosis of the dermis, subcutis, and sometimes the underlying soft tissues and bone. The hallmark of this group of diseases is skin thickening, as is present in systemic sclerosis, and some are associated with autoantibodies and/or autoimmune conditions. Previous reports have described a possible association between DM and scleroderma, which is considered a DM scleroderma overlap syndrome. However, there have been no previous reports showing scleroderma like focal skin atrophy in DM.

Case Report: A patient with DM, confirmed by muscle biopsy, developed unusual skin lesions characterized by the atrophy and hardening of focal areas over lateral upper arms, posterior shoulders, and left temporal areas. Focal scleroderma was highly suspected. The extensive blood work up for scleroderma and other connective tissue diseases, including serum C-reactive protein and antibodies for fluorescent antinucleus, ribonucleoprotein, Smith, double strand DNA, centromere, and Scl-70 were within normal limits. A skin biopsy showed no evidence of scleroderma or other connective tis-

sue findings except for minimal inflammation. The scleroderma like skin changes and muscle weakness improved after immunotherapy.

Conclusion: Patients with DM may present with multiple skin abnormalities that include focal scleroderma like skin atrophy and therefore, should also be categorized as a scleroderma like disorder.

HEREDITARY INCLUSION BODY MYOPATHY ASSOCIATED WITH CARDIOMYOPATHY

Y. Chai, T.E. Bertorini, F.A. McGrew (Memphis, TN)

Introduction: Hereditary inclusion body myopathy (HIBM) type is an autosomal recessive disorder characterized by a preferential involvement of the distal muscles of the lower extremities, especially the anterior compartment of the legs, with relative preservation of the quadriceps. It is often referred to as quadriceps sparing myopathy. Previous reports in HIBM described an exclusive involvement of skeletal muscles. There have been no previous reports showing the involvement of other organs such as the heart in HIBM.

Case Report: The authors report two siblings who presented with typical HIBM with diagnosis confirmed by genetic testing and muscle biopsy. Both developed exertional dyspnea 20 to 26 years after disease onset. Cardiac studies indicated a dilated cardiomyopathy.

Conclusion: This is the first report suggesting a possible association between HIBM and cardiomyopathy. It also suggests that HIBM may involve the myocardium. Although it is possible the cardiomyopathy may have other etiologies, it is important for clinicians to be aware of a possible association for proper management.

TRENDS IN OUTCOMES AND HOSPITALIZATION CHARGES IN INFLAMMATORY MYOPATHIES

Z.A. Al-Qudah, L. Mehyar, H.R. Khan*, S. Islam*, N. Souayah (Newark, NJ; Miami, Florida*)

Introduction: New therapeutic strategies have been introduced in the past 10 years to manage inflammatory myopathies.

Objective: To assess the impact of new therapeutic strategies on outcome and cost of hospitalization among patients with inflammatory myopathies.

Methods: Using a retrospective analysis of cross sectional survey, the authors determined the rates of occurrence, in-hospital outcomes, and mean hospital charges for pediatric patients hospitalized with inflammatory myopathies in 1997 using the Nationwide Inpatient Survey. These outcomes were compared with those observed among patients hospitalized in 2007.

Results: When comparing data from 2007 with data from 1997, more admissions were found for inflammatory myopathies patients (2617 versus 2856). The average patient's age was significantly higher in 1997 as compared to those in the 2007 group (51.25 ± 22.6 versus 48.17 ± 22.25 , $p < 0.0001$). Gender distribution was similar in both groups with noted female predominance. The length of hospitalization (in days) was significantly longer in the 2007 group (9.79 ± 2.88 versus 8 ± 10.4 , $p < 0.001$). Hospital mortality was determined to be significantly higher in 1997 (86 (3.3%) versus 43 (1.5%), $p < 0.0001$). The average cost of hospitalization was significantly higher in the 2007 group ($\$35,068 \pm \$33,198$ versus $\$13,751 \pm \$13,194$, $p < 0.0001$), even when the 1997 average cost of hospitalization was adjusted to allow for inflation ($\$19,251$).

Conclusions: With the improvement of therapeutic strategies for the management of inflammatory myopathies between 1997 and 2007, a significant increase in the average cost and length of hospital-

ization was found with a significant reduction of mortality in 2007 as compared to that in 1997.

AUTOSOMAL RECESSIVE BETHLEM MYOPATHY

A. Arshad, G.R. Hayat, S.J. Iyadurai (St.Louis, MO)

Introduction: Bethlem myopathy is a collagen VI disorder characterized by early onset, slowly progressive weakness, and joint contractures. It follows a relatively milder course compared to Ulrich congenital muscular dystrophy. Previously, the autosomal recessive inheritance pattern for Bethlem myopathy was considered to be uncommon, but recent literature suggests that Bethlem and Ullrich myopathies share the spectrum of collagen VI disorders. The authors propose probable autosomal recessive Bethlem myopathy in a child.

Case report: A 9-year-old girl presented with a 2 year history of progressive generalized weakness with difficulty rising from a chair. Bilateral finger contractures and decreased wrist motion was noted by the parents. Family history for myopathy was negative. Physical examination showed generalized weakness, proximal more than distal, and flexion contractures in bilateral hands. Electrophysiological study showed right median sensory neuropathy and a few myopathic units in proximal muscles. Creatine phosphokinase and electrocardiogram were normal. Mitochondrial deoxyribonucleic acid analysis was normal. Genetic testing for Lamin A/C and emerin were negative. Muscle biopsy showed non-specific changes.

Conclusion: The reported patient shows a possible occurrence of autosomal recessive Bethlem myopathy. This finding indicates that both Bethlem and Ullrich myopathy may present with autosomal recessive and dominant mutations. This finding has implications for genetic counseling as well as genotype and phenotype correlations in collagen VI disorders. Both of these disorders likely represent a spectrum of the same underlying genetic disorder with differing severities.

DIFFERENCES IN OCULAR MYASTHENIA GRAVIS IN ALABAMA BETWEEN RACES

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Introduction: In a 2009 report on 235 patients with myasthenia gravis (MG) in Alabama, differences were noted between whites and African American (AAs). In the AA group, the incidences of MuSK-ab MG and seronegative ocular MG were found to be higher.

Objectives: To determine whether there is a difference in the incidence of ocular MG between whites and AAs in Alabama based on an extensive survey performed at the University of Alabama Birmingham's Medical Center.

Ocular MG was defined as patients with ocular findings that persisted for at least 2 years, and were the only clinical manifestation at the last examination. The authors surveyed available charts of MG patients from three sources that included a neuromuscular (NM) disease clinic, the neuroophthalmology clinic (from 2003 to current), and all historical charts from the NM disease clinic (between 1990 and 2002).

Results: Sixty-nine ocular MG patients were identified (AAs=21, white= 48). AChR antibody was positive in 57% of all ocular MG cases. AChR antibody was positive in 71% of 48 cases in white patients, and in 24% of 21 cases in AA patients.

Conclusion: A difference exists in AChR antibody rates between white patients and AA patients in cases of ocular MG. This difference is statistically significant (P value < 0.0004).

CONCENTRIC NEEDLE JITTER IN MYASTHENIA GRAVIS

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Introduction and Objectives: To estimate the jitter parameters in myasthenia gravis (MG) in stimulated frontalis (F) and extensor digitorum communis (EDC) muscles using concentric needle electrodes (CNE).

Methods: Fifteen MG patients, 10 male and 5 female (age 43.7 ± 15 years) were studied. MG Foundation of America Clinical Classification: I (n=4), IIb (n=4), IIIa (n=2), IIIb (n=2), IVa (n=1), IVb (n=1) and V (n=1). Jitter was expressed as the mean consecutive difference (MCD) in 515 and 494 analyzed potentials for F and EDC, respectively. Filter settings 1 to 10 kHz. Upper limit of normal was set to 99% of control values.

Results: Mean MCD in F was $62.3 \mu\text{s}$ (12.4 to 124), and abnormal in 80% ($> 24.2 \mu\text{s}$). Mean MCD in EDC was $45.8 \mu\text{s}$ (12.1 to 103), and abnormal in 73.3% ($> 24.8 \mu\text{s}$). Mean percentage of number of outliers ($> 33.9 \mu\text{s}$) were 65% (F) and 42.5% (EDC), and abnormal in 80% for both muscles. Some jitter parameters were abnormal in 14 (93.3%) patients in combined studies. Acetylcholine receptor antibody (AChRab) was abnormal in 12 (80%), $9.31 \pm 5.92 \text{ nmol/L}$ (1.53 to 19.5). Muscle specific kinase antibodies were negative in all three AChRab negative patients. These were measured together with the jitter studies.

Conclusions: Stimulated jitter recordings measured from CNE can be used for MG diagnosis with a high sensitivity. Extensive normative studies are lacking and therefore, borderline findings should be evaluated with great caution. Study supported by Foundation for Research Support of the State of São Paulo (FAPESP)

ANTI MUSK MYASTHENIA GRAVIS RESPONSIVE TO RITUXIMAB

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Introduction: Myasthenia gravis (MG) is an autoimmune neuromuscular disease that is commonly associated with anti acetylcholine receptor antibodies. Few cases, mostly those with bulbar involvement,

are associated with anti MUSK antibodies. Epidemiologic studies reveal a severe course and poor response to therapy of anti MUSK MG. Rarely, anti MUSK MG is associated with thymus disease.

Case Report: A 43-year-old right handed man presented with a 6 month course of diplopia, variable ptosis, and severe dysarthria. He gradually developed axial weakness and shortness of breath. High titers of anti MUSK antibodies were detected. Anti-acetylcholine receptor and anti striated muscle antibodies were absent. Repetitive nerve stimulation (RNS) of the facial nerve at 3 Hz revealed significant decrement. Needle electromyography revealed short duration polyphasic potentials in the face, deltoid, and biceps. After his computerized tomography of the chest identified a thymic mass, he underwent thymectomy. Pathological examination revealed thymic hyperplasia and no malignant cells. He showed no clinical response after plasma exchange, anti cholinesterase therapy, and steroids. The treatment with intravenous immunoglobulin, in combination with immunosuppressors, did not offer benefit. As a last resort, infusions with rituximab were initiated and significantly improved his muscle strength, diplopia, and dysarthria.

Conclusion: Anti MUSK MG is a rare autoimmune condition which represents a diagnostic and therapeutic challenge. Rituximab could be used to treat otherwise poorly responsive cases. Further investigation is needed for a better understanding of the phenomena of thymus disease with anti-MUSK MG and its clinical response to immunomodulatory therapy.

A CONCOMITANT PRESENTATION OF ANDERSEN-TAWIL SYNDROME AND MYASTHENIA GRAVIS IN A PATIENT WITH GENERALIZED WEAKNESS

J.J. Luo (Philadelphia, PA)

Introduction: Andersen-Tawil syndrome (ATS) is a rare channelopathy caused by potassium channel (Kir2.1) mutation. ATS is characterized by periodic paralysis, cardiac arrhythmias, and dys-

morphic features. The periodic paralysis may be hypo, hyper, or normokalemic. Myasthenia gravis (MG) is an autoimmune mediated disorder of neuromuscular junction. Anti acetylcholine receptor antibodies (AAR) have been considered pathognomonic and pathogenetic for MG.

Case Report: A 31-year-old woman with a history of morbid obesity and periodic weakness was admitted and intubated in August of 2008 due to hemodynamic instability and cardiogenic shock. Electrocardiography revealed a prolonged QT-interval, ST-elevation in the inferior and anterolateral leads (1 mm), and subsequently, 3rd degree AV block. Laboratory results upon admission discovered hypokalemia (K=2.5 mEq/l), and acute renal insufficiency (creatinine=1.7 mg %), but was unremarkable in all other tests. She was stabilized. A pacemaker was implanted. Neurological examination showed she had dysarthria, bilateral ptosis, horizontal diplopia, and a lax jaw with poor dentition. She was unable to completely close her eyes and hold up her neck. Muscle strength was 2/5 in the lower extremities, and 3/5 in the upper extremities. Tendon reflexes were normal. No muscle tenderness or pathologic reflexes were seen. Clinical findings were consistent with those of ATS. Follow up neurophysiologic studies showed abnormal decremental responses on repetitive nerve stimulation. High titers of AAR were identified. She was treated with oral potassium supplementation and an anti acetylcholinesterase inhibitor (pyridostigmine). Her symptoms were improved and remained in a relatively stable condition.

Conclusion: Concomitant presentation of both ATS and MG in a patient is rarely found in the literature and represents a diagnostic and therapeutic challenge.

SEROPOSITIVE MYASTHENIA GRAVIS ASSOCIATED WITH SMALL CELL LUNG CARCINOMA

M. Ohira, D. Jeong, S.J. Oh (Birmingham, AL)

Introduction: Lambert-Eaton myasthenic syndrome (LEMS) is a well known classical para-

neoplastic syndrome of small cell lung carcinoma (SCLC). An association between myasthenia gravis (MG) and SCLC is rarely known.

Case Report: A 65-year-old man noticed weakness in leg muscles. The weakness worsened and spread to his upper arms. Shortness of breath also developed. A neurologic examination showed bilateral ptosis, marked weakness in proximal muscles, moderate weakness in distal muscles, diminished reflexes, and normal sensory function. Due to marked decremental response in the repetitive nerve stimulation test, the diagnosis of myasthenia gravis crisis was made. Single fiber electromyography in the extensor digitorum communis muscle showed a markedly increased mean value of mean consecutive difference (174 μ s), and frequent blocking in seven single fiber potential pairs (SFPPs). AChR-ab tests were positive. He was treated with plasmapheresis, prednisone, and pyridostigmin. One edrophonium test showed a definite positive response. Chest x-ray showed a nodule in the right upper lobe. A computerized tomography showed a lobulated parenchymal mass in the right hilar area, and a transthoracic needle biopsy confirmed the diagnosis of SCLC. Thymoma was not found. The patient was treated with chemotherapy for SCLC. One month after chemotherapy, the patient developed septicemia and respiratory failure which led to adult respiratory distress syndrome. The patient died 2 months after the onset of weakness.

Conclusion: The patient presented had seropositive MG and SCLC. Only three cases showing the combined features of MG and SCC have been reported. These cases were all seronegative. Thus, this patient is the first with seropositive MG and SCLC.

QUANTITATIVE ANALYSIS OF SURFACE ELECTROMYOGRAPHY FOR THE EVALUATION OF LOWER MOTOR NEURON INVOLVEMENT IN SPINAL AND BULBAR MUSCULAR ATROPHY

M. Higashihara, M. Sonoo, T. Yamamoto, Y. Nagashima, H. Uesugi**, Y. Terao, Y. Ugawa***, E.*

*Stålberg****, S. Tsuji (Bunkyo-ku, Tokyo, Japan; Itabashi-ku, Tokyo, Japan*; Sapporo, Hokkaido, Japan**; Fukushima, Fukushima, Japan***; Uppsala, Uppsala, Sweden****)*

Introduction: Spinal and bulbar muscular atrophy (SBMA) is a motor neuron disease for which some promising clinical trials have been carried out in recent years. A reliable electrophysiological marker for disease progression is of increasing importance. In a preceding study, the authors developed a new quantitative analysis of surface electromyography (SEMG), the clustering index (CI) method, for the tibialis anterior muscle. It can differentiate neurogenic from myopathic changes. This technique was applied to the evaluation of lower motor neuron (LMN) involvements in SBMA.

Objectives: To apply the CI method to the abductor digiti minimi (ADM) muscle and to show its utility in SBMA.

Methods: The subjects included 20 genetically confirmed SBMA patients, and 35 normal controls. The recording electrode was placed over the abductor digiti minimi (ADM) belly with a reference electrode 2 cm proximally, in order to reduce contamination of far field potentials from other muscles. The CI is used to evaluate the way in which the SEMG signal is clustered into individual large motor unit potentials. More than 20 epochs of one second length at various contraction levels were collected from each subject. Their CI values were plotted against the total area of SEMG. The authors constructed a normal cloud and calculated the Z-score for each subject. The compound muscle action potential (CMAP) amplitude of the ADM muscle was also evaluated.

Results: Z-scores were abnormal for all SBMA patients (range: 2.67 to 8.80), whereas the CMAP amplitude was decreased in 65% of patients.

Conclusions: The CI method achieved acceptable sensitivity in detecting LMN abnormality in

SBMA, and is promising as a simple and noninvasive electrophysiological marker.

Mana Higashihara, MD

Golseth Young Investigator Award Recipient

RISK OF HEMATOMA FOLLOWING NEEDLE ELECTROMYOGRAPHY OF THE PARASPINAL MUSCLES

J. Gertken, A.J. Boon, E.J. Sorenson, C.H. Hunt, J.M. Morris (Rochester, MN)

Introduction: In one retrospective review, hematoma formation in the paraspinal muscles following needle electromyography (EMG) was reported to be as high as 11% in non-anticoagulated patients. Based on this finding, needle EMG of the paraspinal muscles is often avoided in anticoagulated patients.

Objectives: To establish the incidence of hematoma, detected by magnetic resonance imaging (MRI) following paraspinal EMG.

Methods: The study population, identified via a computer generated search engine, was comprised of patients who all underwent paraspinal EMG and subsequent concordant level spine MRI within 1 week of EMG over a 1 year period. EMGs were performed at a tertiary care academic medical center by staff and resident neurologists and physiatrists. Two neuroradiologists independently reviewed the spine MRIs of these patients. A hematoma was defined as any acute or subacute blood products at the approximate level of the previous paraspinal muscle EMG.

Results: A total of 4860 charts were identified and reviewed. Of those, 373 charts met the inclusion criteria. No paraspinal hematomas were observed.

Conclusions: To the authors' knowledge, there has been only one previous study investigating the incidence of hematoma formation following paraspinal EMG. In contrast to those results, in which 5 hematomas were identified in 17 patients, the authors

report no MRI evident paraspinal hematomas in the 373 MRIs reviewed. These results should help further the development of evidence based guidelines for EMG procedures in high risk muscles and in patients with an increased bleeding risk. These results support the notion that paraspinal EMG is a relatively safe procedure with a minimal risk of bleeding.

Jonathan Thomas Gertken, MD

Best Abstract Award Recipient

EFFECT OF LIDOCAINE IONTOPHORESIS ON PAIN DURING NEEDLE ELECTROMYOGRAPHY

T. Annaswamy, A. Morchower (Dallas, TX)

Introduction: Acute pain associated with needle electromyography (EMG) can cause premature termination or may interfere with data collection or interpretation. No prior studies have evaluated the effect of lidocaine iontophoresis on pain reduction during EMG.

Objectives: The authors will determine if lidocaine iontophoresis applied prior to EMG examination mitigates pain.

Methods: This was a prospective, randomized, placebo controlled, double blinded, study. Local institutional review board approval was garnered and informed consent processes were followed. Subjects were recruited from patients scheduled for EMG evaluation of bilateral upper extremities in a hospital based clinic. Subjects were randomized to lidocaine (4%) or placebo (normal saline) groups. A research pharmacist provided the blinded medication with a total of 40 mA.min of iontophoresis administered over the left opponens pollicis (OP). The right OP was untreated. Monopolar needle EMG of bilateral OP was then performed and was immediately followed by subjects rating their pain on a 10 cm visual analog scale (VAS).

Results: Fourteen subjects participated in the study. Paired t-tests were performed to evaluate group

differences. Untreated side VAS (6.61 ± 1.96) was significantly higher than the treated side (4.63 ± 2.90 ; $p < 0.05$). However, there were no significant differences between the untreated and treated sides in either treatment group (lidocaine group: 7.29 ± 1.56 versus 5.63 ± 3.12 ; $p = 0.19$ and placebo group: 5.93 ± 2.19 versus 3.63 ± 2.48 ; $p = 0.1$), indicating a placebo effect. Seventy-two percent of subjects indicated a preference for iontophoresis.

Conclusions: The administration of iontophoresis prior to needle EMG significantly reduces pain associated with the test. However, lidocaine iontophoresis was not significantly different than saline iontophoresis, indicating a placebo effect. Further studies are needed to determine if lidocaine iontophoresis is beneficial for use in the clinical setting. Study supported by UT Southwestern Medical Center Department of PM&R

Thiru Annaswamy, MD, MA
Best Abstract Runner-Up Award Recipient

TONGUE ULTRASOUND IN OCULOPHARYNGEAL MUSCULAR DYSTROPHY

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(Albuquerque, NM)

Introduction: Reports suggest that magnetic resonance imaging or computed tomography are sensitive in detecting subclinical fat infiltration of the tongue in patients with late oculopharyngeal muscular dystrophy (OPMD), and specific when compared to myasthenia gravis (MG). Tongue ultrasound (US) could provide a safe, inexpensive, and quick alternative to neuroimaging in OPMD.

Objectives: To determine the sensitivity, specificity, and reliability of US in detecting tongue fat infiltration in patients with OPMD and bulbar MG.

Methods: The tongues of 10 consecutive patients with genetically confirmed OPMD and eight consecutive patients with bulbar MG were evaluated with US in two planes. A radiologist and a neuromuscular neurologist, both with experience in

skeletal muscle US and blinded to the patient's diagnosis, assessed the images for fat infiltration defined as visually determined, clear cut, high muscle echogenicity. Sensitivity, specificity, and inter observer agreement were determined.

Results: The mean age (61.6 ± 4.9 versus 60.5 ± 14.1 years, $p = 0.839$) and time of disease (7.3 ± 3.9 versus 5 ± 4.8 years, $p = 0.274$) at US were no different between OPMD and MG groups. The proportion of females was higher in the OPMD group ($8/10$ versus $4/8$, $p = 0.025$). For ultrasound readers one and two, the sensitivity was 60% ($6/10$) and 40% ($4/10$), and the specificity was 50% ($4/8$) and 50% ($4/8$), respectively. There was slight interobserver agreement ($k = 0.12$).

Conclusion: In contrast to reports using delayed neuroimaging, non quantitative tongue US performed relatively early in the disease may not be diagnostically informative when considering OPMD versus bulbar MG diagnoses. Quantitative US with enhanced spatial resolution should be considered for future studies.

CLINICAL FEATURES IN MUSK ANTIBODY POSITIVE AND NEGATIVE MYASTHENIA GRAVIS

K. Kim, Y. Lim, B. Kang (Seoul, Republic of Korea)

Introduction: The anti muscle specific tyrosine kinase (MuSK) antibodies (Ab) are specific for AChR Ab-negative myasthenia gravis (MG).

Objectives: To determine the clinical characteristics of MuSK Ab-positive and negative MG patients who lack AChR Ab.

Methods: The authors assayed MuSK Ab in 22 seronegative MG patients and compared the features of patients with MuSK Ab to those without MuSK Ab.

Results: Among 22 patients, eleven (one man, 10 women) had MuSK Ab. The mean onset age was 36.9 years (range: 22 to 52 years) in the MuSK

Ab-positive group. The age of onset in the MuSK Ab-negative group ranged from 21 to 55 years ($F=0.4652$). Hyperthyroidism was more prominent in MuSK Ab-negative patients (4 of 11 patients, 36%). In the MuSK Ab-positive group, weakness initially involved the extraocular and bulbar muscles in five (45%) patients. In the MuSK Ab-negative group, initial weakness involved ocular muscles in seven (64%), and the bulbar muscles in three (27%). Repetitive nerve stimulation test (RNST) was normal in five MuSK Ab-negative patients and in four MuSK Ab-positive patients. Five MuSK Ab-positive patients showed positive RNST at proximal muscles of deltoid or trapezius. Patients moderately responded to immunosuppressants, regardless of the presence of MuSK Ab. Two MuSK Ab-negative patients were completely treated after thymectomy for thymoma without persistent immunosuppressant or anticholinesterase.

Conclusions: In a group of generalized female MG patients without AChR-Ab and hyperthyroidism,, the probability of MuSK Ab positive was high, despite the RNST and presentation of ocular symptoms. The responses of anticholinesterase and immunomodulatory agents in MuSK Ab-positive and negative groups were similar and were considered to be generally successful.

AMYOTROPHIC LATERAL SCLEROSIS IN PATIENTS WITH A HISTORY OF POLIO-MYELITIS

Sadeghian, R. Alkawadri, S.P. Nations, J.R. Trivedi (Dallas, TX)

Introduction: The authors report two patients with a history of poliomyelitis who developed amyotrophic lateral sclerosis (ALS) later in life.

Case Report: Patient 1: A 59-year-old man with history of arrested hydrocephalus since childhood had paralytic poliomyelitis at age eight with bulbar, respiratory, and leg weakness. He recovered and was able to walk independently with use of a special boot. At age 45, he developed left leg pain and new weakness and was diagnosed with post polio-

myelitis syndrome. At age 56, he developed cognitive dysfunction and abnormal behavior. One year later, he developed rapidly progressive arm and leg weakness. Electromyography (EMG) showed active denervation with minimal reinnervation. A muscle biopsy showed active neurogenic atrophy. He died at age 60. Autopsy revealed Bunina bodies as well as ubiquitin and TDP-43 positive inclusions in surviving neurons.

Patient 2: A 59-year-old man had a history of poliomyelitis resulting in paraparesis at age two. Following recovery, he was left with mild residual weakness in the legs. At age 51, he developed left arm weakness, which progressed to involvement of the right arm. At age 54, he developed neck extensor and bulbar weakness. He became wheelchair dependent and used a bi level positive airway pressure device at age 59. An EMG showed active denervation in three myotomes. Genetic test for Kennedy disease was negative.

Conclusion: ALS rarely occurs in patients with a history of poliomyelitis, and the course appears to be similar to patients with no history of poliomyelitis. The occurrence of rapidly progressive weakness in muscles that were clinically spared during poliomyelitis favors ALS.

AN ADAPTIVE SAFETY AND DOSE FINDING STUDY OF FAMPRIDINE-SR IN EARLY AMYTROPHIC LATERAL SCLEROSIS

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Introduction: Respiratory insufficiency becomes evident earlier during sleep in patients with amyotrophic lateral sclerosis (ALS). Non invasive ventilation (NIV) is associated with increased survival, but dominant bulbar symptoms cause difficulties adapting to it. Thus, it is important to investigate alternatives to delay NIV. A statistically significant improvement with 3, 4 DAP in ALS was previously shown in the literature. Fampridine SR is a sustained release 4-aminopyridine.

Objective: The authors' primary objective is to determine whether Fampridine SR is safe and well tolerated in patients with ALS. A secondary objective is to explore a dose to response relationship.

Methods: A single center study will identify the maximum tolerated dose of Fampridine SR by an adaptive continual reassessment method. The secondary objective will evaluate a percentage change in sleep disordered breathing parameters. Peripheral arterial tonometry algorithm will help to identify surges of sympathetic activation and 'autonomic arousals' associated with nocturnal respiratory events in a home setting.

Results: The cumulative effect of 4-AP is an increased duration of the presynaptic action potential by blocking potassium channels, prolonging depolarization, and enhancing ACh release with improved neuromuscular (NM) transmission. Decreased transmitter release and/or conduction failure in motor terminals might play a role in the human motor unit (MU) failure. Electromyography decrement during repetitive MU activation in motor neuron disease has been observed where 4-AP can increase force production.

Conclusions: As 4-AP improves NM transmission and muscle contractility, the authors hypothesized that it would improve SDB and delay NIV. An evaluation of the safety and dose finding of Fampridine-SR as a symptomatic treatment in ALS is recommended. These results can serve as a guideline in the creation of future efficacy trials.

ELECTROMYOGRAPHY AND MAGNETIC RESONANCE IMAGING IN THE EVALUATION OF LOWER BACK PAIN

S. Im, J. Moon (Jeju, South Korea, Seoul, South Korea*)*

Objectives: To assess the findings and correlation of electrophysiologic study, electromyography (EMG) and imaging modality, and magnetic resonance imaging (MRI) in patients with lower back pain.

Methods: The authors reviewed the case records of all patients with lower back pain that were referred to a particular EMG laboratory over an 24 month period. Only patients that had undergone a spine MRI performed within a prescribed 6 month period were included. The electrodiagnostic evaluation included nerve conduction studies and needle EMG. One group included only patients with needle examination abnormalities consistent with radiculopathy, ongoing denervation in two or more muscles of the same myotome, innervated by different peripheral nerves, and with no abnormalities in adjacent myotomes. MRI studies were evaluated based on neuroradiologist reports and by the assessing specialist. The authors compared the results of EMG and MRI findings read by two neuroradiologists.

Results: Overall, 78.3% of patients had an EMG abnormality, and 64.6% of patients had an MRI abnormality. EMG abnormalities were found in 84.4% of patients with abnormal MRI findings. MRI abnormalities were found in 69.7% of patients with abnormal EMG findings. EMG and MRI findings agreed in 66.2% of cases, but disagreed in 33.8%.

Conclusions: EMG and MRI findings will show discrepancy in a significant minority (33.8%) of patients with lower back pain. Therefore, the profound understanding of the advantages and limitations of each study is necessary in the interpretation of study results and in the determination of treatment modalities.

ATYPICAL ELECTRODIAGNOSTIC FINDINGS IN SCAPULAR WINGING SECONDARY TO MULTIPLE NEUROPATHIES: A CASE SERIES

M. Kapadia, O.A. Olufade, G. Siu, T. Vachranukunkiet, C.R. Sridhara (Philadelphia, PA)

Introduction: Scapular winging is easily diagnosed by visible inspection. Causes of scapular winging usually involve one nerve (long thoracic, spinal accessory, or dorsal scapular nerve), resulting in either lateral or medial scapular winging. The authors present a series of four patients who

presented for electrodiagnostic (EDX) evaluation with scapular winging.

Case Report: One patient presented with right shoulder pain after a flu like illness. EDX studies revealed an axon loss neuropathy involving the right upper and middle spinal accessory nerve branches and dorsal scapular nerve. The second patient presented with right arm weakness after a fall with a traumatic brain injury and was found to have isolated neurapraxic block with preservation of axons involving the upper and lower spinal accessory nerve branches and long thoracic nerve. The third patient presented with right arm weakness after thyroidectomy and was found to have axon loss neuropathy involving the upper and middle spinal accessory nerve branches and long thoracic nerve. The fourth patient presented with right shoulder weakness after a fall and was found to have axon loss neuropathy involving all three spinal accessory nerve branches, the long thoracic nerve, and a C5 radiculopathy.

Unilateral scapular winging with similar clinical presentation can be the result of various neuropathic etiologies. Physical examination findings in such patients were inconclusive in determining scapular winging etiologies. EDX findings in these four cases revealed multiple neuropathies involving two or three nerves that cause winging of the scapula. Etiologies included trauma, infection, and cancer related surgeries.

Conclusions: Although uncommon, this case series provides examples of multi nerve involvement in scapular winging and the importance of EDX studies in evaluating the involvement of the specific nerves.

ELECTRODIAGNOSTIC SERVICES IN THE UNITED STATES: ANALYSIS OF CORTICAL MAGNETIC STIMULATION DATA

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Objective: The authors will identify patterns of electrodiagnostic (EDX) service provisions in the United States.

Methods: EDX related claims were identified using current procedural terminology codes for clinical nerve conduction studies and electromyography by reviewing outpatient claim data from 2006. The information was comprised of Medicare claims filed in 2006.

Results: There were 241,295 EDX consultations that represented a random sample of 25% of the total EDX consultations in the United States during the given year. Neurologists accounted for the highest percentage of physician providers (52.6%), physiatrists accounted for 19.7%, and other types of physicians provided 21.8%. EDX encounters by non physician providers accounted for 2.6% of all studies. Claims were billed under Independent Diagnostic and Treatment Facilities (IDTFs) for 2.5% of these encounters. Differences in the extent of testing were noted with physiatrists and neurologists concordant in the extent of testing. Non physicians performed fewer tests per encounter, but had more reliance on nerve conduction testing and had less reliance on electromyography. The IDTFs employed the greatest proportion (10%) of studies, consisting of more than six individual tests.

Conclusions: This study provides a large scale perspective on the provision of EDX services in the United States for Medicare beneficiaries. Differences in the types of testing and the extent of testing by the provider type should be examined in future studies as they relate to differences in diagnostic outcomes and clinical trajectories.

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USEFUL PARAMETER FOR PREDICTION OF TREATMENT RESPONSE IN CARPAL TUNNEL SYNDROME

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Objectives: To verify the feasibility of initial parameters of ultrasound (US) or electromyography for response prediction after surgical release or steroid injection in carpal tunnel syndrome (CTS).

Methods: The authors recruited individuals with clinical, electrodiagnostic, and US evidence of CTS. Evaluations include the Boston self assessment questionnaire, median nerve motor and sensory conduction studies, and US at baseline, 3 weeks and 3 months after steroid injection or surgical release. The median nerve area of maximal swelling point (MS), 2 cm proximal from MS (2MS) and 12 cm proximal from MS (12MS) were measured by US and its ratio was calculated. The correlation between symptom score improvement after treatment and baseline parameters of nerve conduction study or US were estimated.

Results: Ten individuals (mean age 57.1 years, Boston symptom score 26) with 16 affected wrists were treated by steroid injection, and ten individuals (mean age 57.2 years, Boston symptom score 33) with 20 affected wrists were treated by surgical release. Boston symptom score were significantly improved 3 weeks and 3 months after treatment in both groups. After 3 months, in the surgical release group, the initial ratio of MS / 12 MS had a positive correlation with an improvement of the Boston symptom score ($p < 0.05$). In contrast, the steroid injection group showed positive correlation in the initial ratio of 2 MS / 12 MS after 3 months ($p < 0.01$). No other initial parameters of study show significant correlation with symptom score improvement.

Conclusions: The initial median nerve swelling ratio may be a useful predictor of response following steroid injection or surgical release.

ARESEARCH ON THE PULMONARY REHABILITATIVE MANAGEMENT IN PATIENTS WITH NEUROMUSCULAR DISEASE

D. Kim, S. Kang, W. Choi (Seoul, Seoul, Korea)

Introduction: Proper pulmonary rehabilitation is a sole method in prolonging the lifespan of ad-

vanced neuromuscular disease (NMD) patients. The authors aimed to review the actual application state of pulmonary rehabilitation.

Objectives: To investigate the real condition of pulmonary rehabilitation for patients with advanced NMDs on mechanical ventilation in Korea.

Methods: In order to estimate the current state of pulmonary rehabilitative management, chart review and pulmonary function evaluation were conducted in a total of 327 NMD patients who had used a mechanical home ventilator in specific conditions from March of 2001 to December of 2009.

Results: The 327 patients included 114 with Duchenne muscular dystrophy, 83 with other types of myopathy, 107 with amyotrophic lateral sclerosis, and 23 with spinal muscular atrophy. Twenty-one were previously intubated, and 18 patients had undergone tracheostomy and were switched into volume limited noninvasive ventilation (NIV). At the time of hospital discharge, 279 patients had applied NIV successfully. Twenty-four other patients who once used breathing apparatus products inappropriately were switched into volume limited NIV. However, 20 patients who had successfully applied NIV initially underwent tracheostomy later due to the exacerbation of underlying disease.

Conclusions: Although adequate pulmonary rehabilitation has been established as a cornerstone treatment of advanced NMD patients, proper management is not universal in a true clinical setting. If more clinicians take an active approach in treating pulmonary problems, more NMD patients could improve the quality and prolong their lives through proper pulmonary rehabilitation. Such steps include regular pulmonary function checkups and ventilatory state monitoring, as well as early NIV application.

TRENDS IN OUTCOMES AND HOSPITALIZATION CHARGES IN PEDIATRIC GUIL-LAIN-BARRE SYNDROME

K. Farhad, H.R. Khan, S. Islam*, N. Souayah (Newark, NJ; Miami, FL*)*

Introduction: New therapeutic strategies have been introduced in the past 10 years to manage Guillain-Barre Syndrome (GBS) among pediatric patients.

Objective: To assess the impact of new therapeutic strategies on the outcome and cost of hospitalization among pediatric patients with GBS.

Methods: Using a retrospective analysis of cross sectional survey, the authors determined the rates of occurrence, in-hospital outcomes, and mean hospital charges for pediatric patients hospitalized with GBS in 1997 using the Kids' Inpatient Database. These outcomes were compared to homologous data from 2006.

Results: When comparing data from 2006 with data from 1997, it was determined that there were more admissions for GBS patients (873 versus 759). The average patient's age in years was significantly higher in 2006 as compared to those in the 1997 group (10.9 ± 6 versus 9.2 ± 5.7 , $p < 0.0001$). However, gender distribution was similar in both groups and had slight male predominance. The length of hospitalization (in days) was significantly higher in the 1997 group (8 ± 6.3 versus 7.1 ± 5 , $p = 0.001$). No in-hospital mortality was reported in the studied periods. The average cost of hospitalization was significantly higher in the 2006 group ($\$42,000 \pm \$27,000$ versus $\$18,000 \pm \$12,000$, $p < 0.0001$) even when the 1997 average hospitalization cost was adjusted to account for inflation ($\$42,000 \pm \$27,000$ versus $\$22,500 \pm \$15,000$).

Conclusions: With the improvement of therapeutic strategies for the management of pediatric GBS between 1997 and 2006 there was a dramatic increase in the average cost of hospitalization with a modest reduction in length of stay in 2006, as compared to 1997.

Khorso Farhad, MD

Junior Member Recognition Award Recipient

TRENDS IN OUTCOMES AND HOSPITALIZATION CHARGES IN NEONATAL BRACHIAL PLEXUS PALSY

K. Farhad, H.R. Khan, S. Islam*, N. Souayah (Newark, NJ; Miami, FL*)*

Introduction: In the past 10 years, new therapeutic strategies have been introduced to manage neonatal brachial plexus palsy (BPP).

Objective: To assess the impact of new therapeutic strategies on the outcome and cost of hospitalization among patients with BPP.

Methods: Using a retrospective analysis of cross sectional survey, the authors determined the rates of occurrence, in-hospital outcomes, and mean hospital charges for BPP patients hospitalized in 1997 using the Kids' Inpatient Database. These outcomes were compared with homologous data from 2006.

Results: When comparing data from 1997 with data from 2006, more admissions for BPP patients requiring mechanical ventilation were found in the latter group (157 versus 82). The average patient's age in days was higher in 1997 when compared to those in the 2006 group (179 ± 124 versus 129 ± 117 , $p = 0.0024$). However, gender distribution was similar in both groups, with noted male predominance. The length of hospitalization (in days) was higher in the 1997 group (3.3 ± 2.1 versus 2.5 ± 1.9 , $p = 0.0032$). Routine discharge was reported with most patients in both groups, and no in-hospital mortality was reported in the studied periods. The average cost of hospitalization was significantly higher in the 2006 group ($\$28,200 \pm \$21,600$ versus $\$16,400 \pm \$13,800$, $p < 0.0001$) even when the 1997 average hospitalization cost was adjusted to account for inflation ($\$28,200 \pm \$21,600$ versus $\$19,000 \pm \$12,700$).

Conclusions: Without significant changes in hospitalization outcome or length of stay, a dramatic increase in average hospitalization charges for pa-

tients with BPP was observed in 2006 as compared to 1997.

Khorso Farhad, MD
Junior Member Recognition Award Recipient

TRENDS IN OUTCOMES AND HOSPITALIZATION CHARGES IN PEDIATRIC MYASTHENIA GRAVIS

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Introduction: In the past decade, new treatments for pediatric myasthenia gravis (MG) patients have been introduced that are expected to improve outcomes associated with hospitalization.

Objective: To assess the impact of new therapeutic strategies on outcome and cost of hospitalization among pediatric MG patients in United States.

Methods: The authors determined the rates of occurrence, in-hospital outcomes, and mean hospital charges for pediatric patients hospitalized with MG in 1997 using the Kids' Inpatient Database (KID). These outcomes were compared with those observed among patients hospitalized in 2006.

Results: There were 307 and 351 admissions for MG in 1997 and 2006, respectively. The proportion of female patients was higher in both time periods. The average age (in years \pm standard deviation) was 12.2 ± 5.5 in 1997, and 12 ± 5 in 2006 ($p=0.626$). The length of hospitalization was significantly higher in 1997 compared to 2006 (4.8 ± 3.4 versus 4.15 ± 3 $p=0.009$). In 1997 and 2006, routine discharge occurred in 295 (96.3%) and 312 (96.4%) patients, respectively. Transfer to short term hospital, home health care, and skilled nursing facilities occurred in 12 (4%) and 11 patients (3.2%), respectively. In-hospital deaths were not reported in either time period. There was a significant increase in mean hospital charges in 2006 compared with 1997 (\$31,810 versus \$19,244), even when the inflation rate was included (\$31,810 versus \$27,518).

Conclusions: Despite improvement in therapeutic strategies and the significant reduction in length of hospitalization of pediatric MG patients from 1997 to 2006, there was a dramatic increase in hospitalization charges without change in clinical outcomes.

Pavel P. Tishuk, MD
Junior Member Recognition Award Recipient

DIRECTIONAL TUNING OF EARLY SHOULDER MUSCLE ACTIVITY

L. Boissé (Kingston, Ontario, Canada)

Introduction: Previous work in the authors' laboratory revealed early electromyography (EMG) activity in shoulder girdle muscles (posterior deltoid, anterior deltoid, supraspinatus, and pectoralis major) of subjects performing a reaching task. This phasic visual response occurred 80 to 100 ms after the presentation of a visual target. This activity preceded the onset of muscle activity associated with volitional arm motion by approximately 85 ms and was time locked to a presentation of a visual stimulus. The results suggest a previously undescribed direct link between the visual system and the musculature of shoulder girdle muscles, thus linking the visual response with motor initiation.

Objectives: To determine whether early EMG activity is tuned to the direction as well as the onset of the visual stimulus.

Methods: Continuous intramuscular EMG of shoulder muscles, as well as elbow and shoulder joint kinematics, was recorded while subjects performed a reaching task with a rotational component.

Results: The posterior deltoid muscle was the only tested muscle to show a significant directional component to the early EMG activity.

Conclusions: While all tested shoulder muscles displayed time locked early EMG activity during a reaching task, only the posterior deltoid displayed direction locked tuning. These results suggest a

previously undescribed direct link between the visual system and the musculature of shoulder girdle muscles, thus linking the visual response with motor initiation. This may have significant implications for hand and eye coordination, as the results suggest that the direction of a target primes relevant muscles before they are called into action to reach for an object.

Lysa Boisse, MD, MSc

Junior Member Recognition Award Recipient

NEUROREHABILITATION CHALLENGES FOLLOWING TIBIAL TO PERONEAL NERVE TRANSFER

S. Macaluso, T.A. Miller, D.C. Ross (London, Ontario, Canada)

Introduction: Nerve transfer is a surgical option that can be employed following nerve injury when an undesirably long nerve graft would be required. This procedure relies on the patient's ability to adapt the activation of a particular nerve to perform a novel motor function. This incorporates the remarkable neuroplasticity of the brain. The authors present a case of lower limb nerve transfer in which the rehabilitation that followed was less than intuitive for the patient, and provided interesting challenges.

Case Report: A 17-year-old male sustained a traumatic dislocation of his right knee with immediate inability to dorsiflex and evert the foot. No functional recovery was seen over the course of 5 months. Electromyography (EMG) showed signs of deinnervation in the tibialis anterior (TA) and peroneous longus (PL), with no evidence of nerve recovery. Surgical nerve transfer was performed using sural nerve grafts to connect two branches supplying the lateral gastrocnemius with the TA and PL. Functional recovery of eversion was evident within 8 months, but with no dorsiflexion. EMG of the TA revealed only a single volitional motor unit (MU) with dorsiflexion, but many MUs present with plantar flexion.

Conclusion: This case highlights neurorehabilitation challenges often experienced following nerve transfer. Recovery of function requires more than peripheral nerve re growth, and involves reorganization of the brain to appropriately activate the desired muscle. The role of EMG biofeedback and functional magnetic resonance imaging in rehabilitation to assist in the process of neuroplasticity is still uncertain. The appropriate timing of these interventions requires discussion.

UNITED STATES PHARMACEUTICAL EXPENDITURES AND TRENDS FOR PERIPHERAL NEUROPATHIC PAIN FROM 1997 TO 2007

J.P. Ney, J. Watanabe (Seattle, Washington)

Introduction: Pain associated with neuropathy is managed with medications including analgesics, anticonvulsants, and antidepressants. As awareness of neuropathic pain and available treatments expanded over the past decade, so have outpatient pharmaceutical expenses for these conditions.

Objective: To estimate trends in pharmaceutical expenses for peripheral neuropathic pain.

Methods: The authors conducted a secondary analysis of data from the 1997 to 2007 Medical Expenditures Panel Survey (MEPS), a nationally representative survey of the civilian United States non-institutionalized population. Survey respondents were identified with prescription drug subclasses commonly prescribed for pain with associated ICD-9 codes for peripheral neuropathic pain conditions. Annual costs (adjusted for medical inflation to 2007 dollars) were tallied using person level weights. Standard errors were adjusted for variance of sample strata and primary sampling unit.

Results: The estimated number of prescription drug users for peripheral neuropathic pain conditions (in the millions) rose steadily from 2.24 (95% CI 1.88 to 2.59) in 1997 to 3.38 (95% CI 2.92 to 3.85) in 2007. Total estimated expenditures for prescription pharmaceuticals for peripheral neuro-

pathic pain increased three-fold from 1997 to 2007 and from \$363 million (95% CI 2.45 to 4.81 million) to \$1.12 billion (95% CI \$837 million to \$1.40 billion). In particular, total costs for GABA-ergic anticonvulsants (gabapentin, pregabalin) for these conditions rose 5200% from \$11.6 million (95% CI \$1.9 to 21.2 million) in 1997 to \$606 million (95% CI \$392 to 819 million) in 2007.

Conclusions: The authors estimate that the total expenditures for prescription drug treatment of peripheral neuropathic pain have risen dramatically from 1997 to 2007. Additionally, individual expenditures have increased out of proportion to the number of new users.

John P. Ney, MD

Junior Member Recognition Award Recipient

PAINFUL MUSCLE SPASM AND MUSCLE SOMATOSENSORY EVOKED POTENTIALS: CHANGES DURING TREATMENT OF LOW BACK PAIN

J.S. Dhaliwal, Y. Zhu, Y. Guan, R. Weber (Syracuse, NY)

Introduction: The role of muscle pain and muscle spasm in low back pain syndrome has been a subject of controversy due to the inadequate measurement techniques that correlate to patient symptoms. Somatosensory evoked potential (SEP) after magnetic stimulation of muscle (mSEP) has provided a new method for examining muscle afferent activity.

Objectives: To define the correlation between the changes in painful muscle spasm and mSEP during treatment.

Methods: A prospective study of 27 patients (16 male, 11 female, aged 21 to 62 years) with diagnoses of lumbar sprain (n=8), myofascial pain (n=9), disc bulging (n=6), and lumbosacral radiculopathy (n=4). The low back pain visual analogue scale (VAS) evaluation, the assessment of the degree of a palpable muscle spasm ranging from no spasm (n=0) to marked spasm (n=4), and mSEP were

completed before and 1 month post non surgical treatment, which included physical therapy, manipulation, and nerve root blocks. The first cortical response of mSEP (P30/N40) was measured following magnetic stimulation of paraspinal muscles on each side.

Results: The patients demonstrated a significant decrease in pain in accordance with the VAS evaluation (from 6.7 ± 0.8 to 2.8 ± 1.2). The amount of palpable muscle spasm also decreased (from 3.5 ± 0.3 to 1.2 ± 0.4). There was an increase in the amplitude of P30/N40 ($0.7 \pm 0.5 \mu\text{v}$ to $1.6 \pm 0.7 \mu\text{v}$) after treatment. There was a significant correlation between the changes in the pain VAS, muscle spasm, and the changes in mSEP.

Conclusions: The data provides evidence of muscle pain, muscle spasm, and muscle afferent activity as mechanisms of low back pain.

CONTRIBUTION OF ELECTRODIAGNOSTIC STUDIES IN LUMBAR SPINAL STENOSIS

J.L. Cosgrove, S.L. Chase, G.K. Cosgrove (Mars, PA)

Objectives: To determine the relationship between electrodiagnostic (EDX) findings and the response to epidural steroid injections (ESI) in subjects with symptomatic lumbar spinal stenosis (LSS). Efficacy of treatment was determined by changes in pre and post injection results on the Swiss Spinal Stenosis Questionnaire (SSSQ) and the Six Minute Walk Test (SMWT).

Methods: Fifteen subjects presenting with painful ambulation and LSS confirmed by magnetic resonance imaging, underwent EDX testing and completed the SSSQ and SMWT. EDX was performed using previously published methods. Results were classified as A (axial muscles only), R (radicular pattern), P (peripheral neuropathy), or N (normal). All subjects received between one and three ESIs, according to routine clinical protocol. Approximately 6 weeks after completion, the SSSQ and SMWT were repeated.

Results: Significant improvement was noted after ESI on both the SSSQ, with a mean score change of 6.7 ($p = .033$), and SMWT, with an average increase of 51 meters ($p = .023$). There was a trend toward smaller changes in walking distances for subjects with any abnormal EDX classification ($p = 0.11$) when compared to those with normal EDX results. However, the differences were not significant. Similar trends were seen in the subgroups with only axial involvement ($p = 0.27$), and peripheral neuropathy ($p = 0.23$).

Conclusions: In subjects with LSS, treatment with ESI resulted in significant improvement in both measured walking ability and self reported functional status. The trend of greater improvement in walking distance between subjects with normal versus abnormal EDX findings did not reach statistical significance.

IS UPPER EXTREMITY PAIN IN WRITER'S CRAMP AN ASSOCIATED REPETITIVE STRAIN INJURY?

A. Recchia (São Paulo, Brasil)

Introduction: Writer's cramp (WC) is a task specific dystonia characterized by abnormal movements or posturing of the upper limb due to inappropriate muscle contractions that interfere with writing movement.

Objectives: To determine whether symptoms inherent to writer's cramp are associated with a repetitive strain injury (RSI).

Methods: Thirty-four patients with WC were evaluated. The patients were classified as being of a simple type, having difficulty only when writing ($n = 14$), or a dystonic type, where symptoms are present when performing writing and other activities ($n = 20$). In five patients, the authors observed the presence of an associated tremor (type A/task specific tremor and type B/position-specific tremor). Each patient had their diagnostics of WC confirmed by electromyography. All patients presented with some degree of painful movements, usually when over gripping a

pen or experiencing uncomfortable tightness in the upper extremity (UE) while attempting to overcome the dystonic movements.

Results: Of the 34 patients studied, 14 had tenosynovitis, 3 bursitis, 2 carpal tunnel syndromes, and 1 had cervical radiculopathy. The remaining fourteen patients showed no associated pathologies, despite the presence of some degree of pain. Specific treatment has been adopted, focusing on WC and the underlying conditions. There was an improvement of pain symptoms in most treated cases, although a marked improvement in performance during the writing was not observed.

Conclusions: The incidence of RSI in WC seems to be much higher than it is recognized to be. Patients with WC presenting with UE pain should be investigated in search of associated conditions to benefit from pain reduction and improve performance when writing.

HETERONIMOUS PERONEUS LONGUS H REFLEX IN THE EVALUATION OF L5 RADICULOPATHIES

A. Recchia (São Paulo, Brasil)

Introduction: The H reflex (HR) is a monosynaptic spinal reflex that assesses both motor and sensory fibers and can be used to assess the integrity of proximal nerve pathways (PNP).

Objectives: To perform H wave study bilaterally, to assess L5 root in control subjects, and compare to patients with an L5 radicular compression.

Methods: The control group consisted of 50 healthy adults ranging in age from 20 to 59 years with no history or signs of neurological disease. The group of patients consisted of 17 subjects ranging in age from 36 to 62 years with a diagnosis of L5 radiculopathy. Lumbosacral nuclear magnetic resonance performed in all patients revealed herniated discs with moderate to severe degrees of foraminal compression. To elicit the HR, tibial nerve was stimulated at the knee with bipolar surface electrodes.

Results: The H wave's mean latency was 28.5 +/-1.7 msec (mean+/-SD) in the control group, and 32.3 +/-1.9 ms (mean+/-SD) in the patient group. HR was absent on the affected side in ten patients. In the remaining 7 patients, when comparing latencies and amplitudes of the both sides, there was an asymmetry greater than 2 ms and a reduction of up to 55% lower in the affected side, respectively.

Conclusions: Needle examination proves to be the better neurophysiological test to assess the integrity of PNP, however, it does not allow for the evaluation of preganglionic sensory fibers which constitutes an advantage when using HR. The heteronomous HR is easy to perform and provides an alternative and complementary technique to assess the integrity of proximal segments. It may also emphasize the findings obtained in a needle examination.

POPLITEUS MUSCLE HEMORRHAGE: A RARE CAUSE OF PROXIMAL TIBIAL NEUROPATHY

E.L. Dimberg, D.I. Rubin, C.J. Ortiguera, K.D. Kennelly (Jacksonville, FL)

Introduction: Popliteus muscle strain or rupture is a rare cause of a proximal tibial neuropathy. Patients typically present to orthopedists with complete or nearly complete recovery, which is usually achieved with conservative management. The authors present a patient who developed a tibial neuropathy due to a traumatic popliteus muscle strain and hemorrhage whose clinical presentation was initially masked by a chronic L5 radiculopathy.

Case Report: A 54-year-old man with a longstanding history of back pain and a chronic L5 radiculopathy developed severe right leg pain and numbness, pain, and weakness in the right foot after jumping onto a dock. Examination demonstrated severe weakness in right toe flexion, foot dorsiflexion, eversion, inversion, and plantar flexion, but proximal leg strength was normal. Sensation was reduced in his first and second toes and plantar foot, and his Achilles reflex was absent. Electrodiagnostic (EDX) studies demonstrated a proximal right tib-

ial neuropathy with a superimposed chronic right L5 radiculopathy. Magnetic resonance imaging (MRI) of the leg showed edema and hemorrhage within the popliteus muscle displacing the tibial nerve. He was treated with conservative measures. After 1 year, only a partial recovery was achieved.

Conclusions: Popliteus muscle rupture or strain should be considered in the differential diagnosis of tibial neuropathy. Although often presenting to orthopedists or sports medicine physicians, neurologists and physiatrists should be aware of this entity. EDX and MRI are essential for confirmation of the diagnosis. Although previous studies suggest good recovery, the patient studied did not fully recover. Surgical decompression may be considered in future cases.

RELIEF OF PAIN SYMPTOMS IN CHRONIC TRIGEMINAL NEURALGIA WITH REPETITIVE TRANSCRANIAL MAGNETIC STIMULATION

L. Gómez, E. Padilla, E. Infante (Havana, Cuba)

Introduction: Repetitive transcranial magnetic stimulation (rTMS) has been proposed as a potential treatment for neuropathic pain.

Objective: To evaluate the effect of 20 Hz rTMS on painful symptoms in a group of 10 patients suffering from therapy resistant chronic trigeminal neuralgia.

Method: Patients received one daily session of 15 trains of 20 Hz rTMS. Each one was 10 seconds in duration with 10 seconds of intertrain interval over 15 days, except weekends. A visual analogue scale (VAS) and the Leeds Assessment of Neuropathic Symptoms and Signs (LANSS) scale were used for the assessment of pain before and after the fifth, tenth, and the last rTMS session.

Result: There were noted significant differences in LANSS scale (Wilcoxon, $p=0,043$) after five rTMS sessions, and in VAS after completing ten sessions (Wilcoxon, $p=0,044$); but the best punctuation was observed after 15 sessions (VAS initial: 8,2; final 1,6; LANSS initial: 21; final:4).

Conclusion: rTMS seems to provide short term pain relief in patients with therapy resistant chronic trigeminal neuralgia.

EFFICACY OF LIDOCAINE TRIGGER POINT INJECTIONS IN CHRONIC MUSCULOSKELETAL PAIN

M.F. Hangan, N. Dhadwal, F.M. Dyro, J. Li (Valhalla, NY)

Introduction: Managing chronic musculoskeletal pain (CMP) can be challenging. Lidocaine trigger point injections (LTPI) is a safe procedure, has minimal discomfort, with rare, localized side effects. Whether it is used as a sole treatment or as an adjunct to other non invasive or pharmacologic therapies, it can reduce CMP symptoms.

Objective: To ascertain the clinical efficacy and safety of long term LTPI in alleviating CMP.

Methods: The authors conducted a retrospective chart review of patients treated between 2001 and 2009 with 1% LTPI for CMP (pain over 3 months). Age, gender, location of and interval between injections, total number of visits, and cointerventions were analyzed.

Results: The authors identified 37 patients (10 males, 27 females). Mean age was 52.14 years (27 to 74). The mean number of visits was 53.5 (9 to 178) with mean length of treatment 5.1 years (1 to 9). Frequency of LTPI was 1 to 3 weeks (n=5), 4 to 6 weeks (n=27), and over 6 weeks (n=5). Location of LTPI was neck or shoulder (n=33), thoracic or lumbar region (n=18), gluteii region (n=13), legs (n=11), and arms (n=3). CMP was secondary to spine diseases (n=23), fibromyalgia (n=19), post traumatic (n=10), shoulder disease (n=9), headache (n=5), and temporo mandibular joint disease (n=3). Patients experienced minimal to complete relief after LTPI with transient soreness at the injection site, but no other side effects were reported. Co interventions included surgery (n=19), opiates (n=18), antidepressants (n=13), GABA analogues (n=11), muscle relaxants (n=11), steroids (n=8),

non steroid anti inflammatories (n=7), and botulinum toxin (n=5).

Conclusion: The authors noticed meaningful improvement in alleviating CMP with long term 1% LTPI used as an adjunct rather than a standalone treatment. Further research will establish its value within the multidisciplinary approach to CMP management.

Mihaela F. Hangan, MD

Junior Member Recognition Award Recipient

PAIN IN PARKINSON'S DISEASE

J.K. Baruah, G.R. Baruah (Milwaukee, WI)

Introduction: Patients with Parkinson's disease (PD) suffer from various non motor disorders that include pain. Pain significantly interferes with PD patients in activities of daily living and complicates the conventional management. This observational study outlines the type and nature of painful conditions and management thereof.

Methods: PD patients with pain (29 male; 20 female) were evaluated. Each underwent complete neurological examination and brief pain inventory. Appropriate painful conditions were identified and treated with medications, physical therapy, and simple procedural interventions whenever indicated.

Results: The PD patients with pain were grouped in this study as follows: aggravated spine disorder (radiculopathy), myofascial pain/cramp, dystonic pain (focal or axial), headache (cervicogenic) including drug induced, peripheral neuropathies (peripheral and focal), central pain, abdominal pain (constipation/bowel obstruction), and shoulder hand syndrome. In five patients, pain preceded the diagnosis of PD.

Conclusions: Management of pain is an important component in the treatment of PD. It is important to realize that the patient may initially present with painful conditions to a family practitioner or internist prior to the time the diagnosis of PD is made.

The treatment of painful conditions with medicine and simple procedural interventions is often helpful. Dystonic pain tends to respond to PD drugs or botulinum toxin. If the neurologist is proactive in treating pain in PD patients, this will help in their mobility and will eventually improve their quality of life and slow the progression of disease.

CHRONIC AUTOIMMUNE AUTONOMIC NEUROPATHY RESPONDING TO PREDNISONE

M. Stanton (Rochester, NY)

Introduction: Chronic autonomic neuropathy may be immune mediated or may be part of a neurodegenerative process. Chronic immune autonomic neuropathy is rare, and its treatment is not well defined.

Case report: A 35-year-old man presented with a 3 year history of heat intolerance associated with loss of sweating that spread over his body. He denied numbness, weight loss, and other autonomic symptoms. His neurologic examination was normal, including pupillary responses and orthostatic blood pressure measurement. Routine nerve conduction studies were normal (sural amplitude 20 mcV and medial plantar amplitude 17 mcV). Skin sympathetic potentials were abnormal and offered no response in the feet, and 50 mcV response in the right hand to startle. Heart rate variability to deep breathing was normal (variability of 20 bpm). Skin biopsy showed normal epidermal and eccrine sweat gland innervation and morphology. Nicotinic acetylcholine receptor antibody titer was normal. Treatment with pyridostigmine resulted in modest benefit, and had no side effects. He was started on 60 mg of prednisone daily for a month, which was decreased to 30 mg every other day by his 3rd month. He reported a marked benefit in terms of sweating and heat tolerance. Seven months after starting treatment with prednisone, skin sympathetic responses were repeated and his responses to startle were normal in the foot (650 mcV). Marked improvement was noted in the right hand (700 mcV).

Conclusions: Anhydrosis can be a limited form of autoimmune autonomic neuropathy. Prednisone can be effective and should be given consideration for treating autoimmune autonomic neuropathy.

NEUROMUSCULAR SAFETY OF SALINE TRANSVENOUS LIMB PERFUSION IN MUSCULAR DYSTROPHY

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Introduction: High pressure retrograde transvenous limb perfusion has been successfully used to deliver plasmid deoxyribonucleic acid and adeno-associated virus minidystrophin transgenes into skeletal muscle in experimental animals. Translating this promising technique to humans with muscular dystrophy (MD) requires addressing multiple safety and logistical aspects including analgesia, vascular access and competence, limb compartment pressure, and integrity of nerve and muscle function.

Objectives: To determine the safety profile of a dose escalation study of retrograde transvenous single limb perfusion with 0.9% saline in adults with Becker and limb girdle MDs.

Methods: An 18 or 20 g intravenous catheter was inserted into the distal lesser saphenous vein. A single cuff tourniquet was placed just above the knee at 310 mm Hg. Infusion of normal saline was carried out. Infusion volume was escalated from 5 to 20% of limb volume. Pre and post perfusion studies of the peroneal and tibial motor nerves and the sural sensory nerve were performed in triplicate within 3 days of perfusion.

Results: No significant change was noted in distal motor latency, compound muscle action potential amplitude, F-wave latency, and conduction velocity of the perfused limb. No change was noted in post perfusion serum creatine kinase or potassium levels or in 6 minute walk distance. Anterior and lateral compartment pressures transiently reached 100 mm Hg in the subject receiving 20% of limb volume.

Conclusions: High pressure retrograde transvenous limb perfusion with saline up to 20% of limb volume is safe and feasible. These studies will serve as a basis for future gene therapy clinical trials.

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CLINICAL IMPLICATION OF COUGH ASSISTING DEVICE SUBSTITUTING GLOTTIC FUNCTION

W. Choi, S. Kang, J. Park, D. Kim (Seoul, Korea)

Introduction: Neuromuscular disease patients who have weak respiratory muscles with bulbar muscle weakness and/or tracheostomy cannot obtain sufficient intrathoracic pressure to produce enough cough flow due to an inability to hold precough volume. These patients have difficulty initiating the compressive phase, so cough flow cannot be sufficiently increased in spite of using the several cough assisted methods. Mechanical insufflation-exsufflation (MI-E) can be effective in clearing airway secretion. However, many patients hesitate in purchasing MI-E due to its high cost. Therefore, the authors have developed a cough assisting device that substitutes glottic function.

Objective: The aim of this study is to estimate the efficiency of the device in patients who have weak respiratory muscles with bulbar muscle weakness and/or tracheostomy.

Methods: Forty-eight patients with bulbar muscle weakness and/or tracheostomy, as well as respiratory muscle weakness, were recruited. The forced vital capacity (FVC), unassisted peak cough flow (UPCF), maximal insufflation capacity (MIC), and assisted peak cough flow (APCF) were measured via tracheostomy or oronasal interface. MIC and APCF were measured using the cough assisting device substituting glottic function.

Results: In all 48 subjects, MICs (1443.3 ± 582.1 ml) measured with the device were significantly

higher than FVCs (776.1 ± 492.2 ml, $p < 0.01$). APCFs (147.5 ± 104.3 L/min) measured using the device were also significantly higher than UPCFs (54.9 ± 42.2 L/min, $p < 0.01$).

Conclusions: The cough assisting device is effective in helping to increase cough flow and assist in air stacking exercises by substituting the function of the glottis in patients who have glottis dysfunction or tracheostomy tube.

CHANGES OF CLINICAL SCALES AND ULTRASONOGRAPHIC FINDINGS AFTER ENDOSCOPIC CARPAL TUNNEL RELEASE SURGERY

D. Park (Kangdonggu, Seoul, South Korea)

Objectives: To evaluate the changes of clinical scales and ultrasonographic findings after endoscopic carpal tunnel release surgery.

Methods: The authors evaluated 31 hands with carpal tunnel syndrome using the Boston carpal tunnel questionnaire (BCTQ; symptom and function), Simovic clinical scale and the historical objective (Hi-Ob) scale preoperatively, and 1, 12, and 24 weeks postoperatively. The flattening ratio (FR) and the cross sectional area (CSA) of the median nerve within the carpal tunnel were measured using ultrasound (US). Paired t-tests were used to compare the changes of clinical scales and US findings.

Results: BCTQ (symptom), Simovic clinical scale, and the Hi-Ob scale showed a significant improvement at 1, 12 and 24 weeks after surgery ($p < 0.05$). But the BCTQ (function) only showed a significant improvement at 12 and 24 weeks after surgery. On US, the CSA of the median nerve within the carpal tunnel decreased significantly at 12 and 24 weeks after surgery ($p < 0.05$). The FR of the median nerve showed no significant change.

Conclusions: This study monitored the changes of clinical scales and US findings 24 weeks after surgery. Postoperatively, the clinical symptoms improved after 1 week, but function improved after

12 weeks. A significant decrease in the CSA of the median nerve showed after 12 weeks, but the FR of the median nerve did not show a significant change.

A LONG TERM FOLLOW UP STUDY AFTER SURGICAL TREATMENT OF CARPAL TUNNEL SYNDROME

D. Park (Kangdonggu, Seoul, South Korea)

Objectives: To evaluate a long term prognosis of carpal tunnel syndrome (CTS) following endoscopic carpal tunnel release.

Methods: Eighty-six hands in 65 patients that underwent endoscopic carpal tunnel release 4 to 7 years previously were enrolled in the study. CTS was diagnosed by electrophysiologic study and clinical symptoms. Electrophysiologic severity was determined by Stenvens' criteria. The authors conducted phone surveys in regards to the clinical improvement (6 point ordinal transition scale by Geritsen) and experience recurrent symptoms. Then, the medical records of respondents were reviewed on the basis of their electrophysiologic severity.

Results: Of all respondents, ten (11.62%) hands had mild CTS, 32 (37.20%) hands had moderate CTS, and 44 (51.16%) hands had severe CTS. There were 10 hands (31.25%) with recurrence of symptoms in moderate CTS, seven hands (15.9%) in severe CTS, and zero hands in mild CTS. In mild CTS, five (50.0%) hands had "complete recovery", four (40.0%) hands had "much improvement" and one (10.0%) hand had "improvement". In moderate CTS, 17 (53.12%) hands had "complete recovery", 11 (34.37%) hands had "much improvement", and 4 (12.5%) hands had "improvement". In severe CTS, 28 (63.63%) hands had "complete recovery", 6 (13.63%) hands had "much improvement", 7 (15.9%) hands had "improvement", and 2 (4.54%) hands had no change.

Conclusions: In the long term follow up study of patients who underwent endoscopic carpal tunnel release, 19.76% of the patients experienced recurrence of symptoms in moderate and severe cases of CTS, but most patients (97.67%) showed improvement.

COMPARISON OF SONOGRAPHY AND ELECTROPHYSIOLOGIC STUDY IN CARPAL TUNNEL SYNDROME

D. Park (Kangdonggu, Seoul, South Korea)

Objectives: To compare the diagnostic usefulness of ultrasonography (US) and electrophysiologic study in carpal tunnel syndrome (CTS) patients.

Methods: The study included 166 hands. The diagnosis of CTS was based on clinical signs and symptoms. All patients had sonographic measurement of the cross sectional area (CSA) of the median nerve and measurement of the difference of wrist palm median sensory latency (DMSL). Normal values were obtained from 40 controls. The sensitivity and specificity of US and electrophysiologic study were studied by means of a receiver operating characteristic (ROC) curve. The kappa coefficient was used to evaluate the relationship between US, electrophysiologic study, and clinical diagnosis.

Results: The CSA and the DMSL were significantly increased in the patient group. The best cutoff value of CSA was 9.5 mm², which had a sensitivity of 87.5%, and a specificity of 92.3%. The DMSL showed a sensitivity of 93.0% and a specificity of 92.3%. The kappa coefficient for DMSL versus clinical evaluation was 0.78, and for CSA, this coefficient was 0.74. Bifurcation of the nerve was present in eight hands, persistent median artery within the tunnel was seen in two hands, and a cyst within the tunnel was found in one hand. Additionally, tenosynovitis was observed in one hand.

Conclusions: In patients with a clinical diagnosis of CTS, electrophysiologic studies showed higher sensitivity than US. However, some anatomical abnormalities were detected by US.

SPREAD TO THE DORSAL ULNAR CUTANEOUS BRANCH: A PITFALL FOR THE ANTI-DROMIC SENSORY NERVE CONDUCTION STUDY OF THE ULNAR NERVE

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Introduction: The dorsal cutaneous branch of the ulnar nerve, or dorsal ulnar cutaneous branch (DUC), is a forearm branch of the ulnar nerve, whose conduction study is used for the differential diagnosis of ulnar neuropathy. The authors found a previously undescribed pitfall for the routine antidromic sensory nerve conduction study (SCS) of the ulnar nerve, which is the spread of the wrist stimulation to the DUC, which may interfere with the examination results.

Objective: To investigate the frequency and the influence of this pitfall on studies of the ulnar nerve.

Methods: The authors stimulated the ulnar nerve at the wrist for ten control subjects, then recorded the antidromic sensory nerve action potential (SNAP) over the proximal interphalangeal joint of the little finger (routine antidromic SCS). Concurrently, the DUC response over the dorsum of the hand was recorded to check the occurrence of the spread. The stimulus intensity and the stimulating position were changed, and the effects were investigated.

Results: In each subject, the spread occurred when the stimulus intensity was increased and led to an increase in the amplitude of the routine SNAP. However, the degree of increase varied among subjects, as 13 to 72% of the genuine response was from the superficial branch. A careless localization of the stimulating point may cause the spread from a weak intensity, which would not be recognized without monitoring the DUC response. In a patient with ulnar neuropathy at the wrist (UNW), the spread caused an erroneous normal onset latency of the routine SNAP.

Conclusions: This pitfall may interfere with the normal values of the ulnar SNAP amplitude and with the diagnosis of UNW.

THE APPROPRIATE MANEUVER OF REPETITIVE NERVE STIMULATION FOR THE TRAPEZIUS MUSCLE

G. Ogawa, M. Sonoo*, Y. Hatanaka* (Tokorozawa, Saitama, Japan; Itabashi, Tokyo, Japan*)

Introduction: The trapezius muscle is a commonly used muscle for repetitive nerve stimulation (RNS) tests. Most textbooks recommend maneuvers that provide downward pressure or have the subject hold a chair in a sitting position. However, significant pseudofacilitation, typically at the second wave, is often difficult to avoid by these maneuvers. The authors have obtained good results by passively elevating the subject's shoulder in the supine position, instead of forcefully pressing down.

Objectives: To compare several maneuvers for RNS in the trapezius muscle with specific regards to pseudofacilitation, and to validate the utility of the method described.

Methods: The group of subjects included 11 healthy volunteers. The authors compared three maneuvers in the sitting position: 1) no restriction, 2) to ask the subject to grip on to the chair, and 3) to give a firm downward pressure to the shoulder, and two maneuvers in the supine position: 4) to firmly hold down the shoulder, and 5) to elevate and hold the shoulder (the authors' method).

Results: The average increment of the second wave in amplitude as compared to the first one was 5.5, 4.8, 4.2, 7.9, and 2.3% respectively, for each maneuver. It was significantly lower in the authors' method than in any other ones. The persistence of voluntary activity was generally prominent for maneuvers in the sitting position, especially for the second method.

Conclusions: The pseudofacilitation in the trapezius muscle is caused by the irresistible force to shorten the muscle. Thus, a maneuver to shorten the muscle from the first, by elevating and holding the shoulder, would reduce such an artifact.

CONCURRENT BRACHIAL PLEXOPATHY IN TRAUMATIC CERVICAL CORD INJURY PATIENTS

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Introduction: Brachial plexopathy is often missed initially due to the fact neurological deficit is masked by other injuries in spinal cord trauma patients.

Objectives: To investigate the clinical and electrodiagnostic (EDX) findings of concurrent brachial plexopathy in traumatic cervical cord injury patients.

Methods: The authors retrospectively reviewed the medical records of 18 patients (12 men, 6 women; age 44.9 ± 13.7 years) who were diagnosed with brachial plexopathy on an EDX study. The patients were among 448 that were admitted to a specific rehabilitation center from 1995 to 2009 with traumatic cervical cord injuries.

Results: The most common neurological level of injury was C4 (27%). On the American Spinal Injury Association impairment scale (AIS), AIS D was most the common. The most common cause of trauma was attributed to car accidents (44%). The combined injuries around the shoulder joint associated with trauma included clavicle fracture, humerus fracture, clavicle scapular injury, scapular fracture, and acromioclavicular joint injury. The severity of brachial plexopathy was incomplete in 94%, and complete in 6%. The most common location of brachial plexopathy was at the root level. The most common presenting symptom among patients was motor weakness. The duration between a traumatic incident and the diagnosis of brachial plexopathy was 64.5 ± 51.9 days. Follow up EDX studies were performed in only four cases. All had electrodiagnostically improved.

Conclusions: The diagnosis of brachial plexopathy was commonly delayed in patients with traumatic cervical cord injuries. The most common level of injury was the root of brachial plexus and/or the cervical cord. This may be attributed to the traction or fracture related injury.

SUPINE ANTERIOR TECHNIQUE OF OBTAINING TIBIAL H REFLEX

A. Shukla, K. Butler (Willimantic, CT)

Introduction: The most common technique used to obtain tibial H reflex studies is with the patient in a prone position with recording electrodes placed over the medial margin of the gastrocnemius or over the soleus with stimulation of the tibial nerve in the mid-popliteal region. The authors describe a technique where the patient is positioned supine and recording electrodes are placed over the anterior portion of the soleus as it crosses over the belly of the flexor digitorum longus (FDL).

Methods: The patient is placed in a supine position. The active recording electrode is placed on most anterior position of the soleus midshaft of the medial portion of the tibia where the soleus crosses over the belly of the FDL. The reference electrode is placed distally on the Achilles tendon. Stimulation is performed using standard techniques (cathode proximal, long stimulation duration, and sub-maximal stimulation technique).

Results: When performed in this manner, all recorded H reflexes and M waves demonstrated an initial negative take off from baseline. H reflex latencies never varied more than 1.5 ms from those obtained in the more traditional manner.

Conclusion: Performing H-reflex studies using this method has advantages over more traditional methods. Placing the patient in a supine position offers ergonomic relief to the patient and to the examiner. Responses which routinely demonstrate good negative take off help reduce measuring error. Negativity generally correlates to techniques which are considered superior due to the proximity to the generator source. This, combined with the placement of the active recording electrode, may also suggest contribution from the FDL.

LOW CONTRAST VISUAL EVOKED POTENTIALS AID IN EARLY DETECTION OF OPTIC DEMYELINATION

F. Bumanlag, J. Luo (Philadelphia, PA)

Introduction: Visual evoked potential (VEP) is a very useful tool in evaluating the anterior visual

pathways. Pattern reversal visual evoked potentials (PRVEPs), which can reveal clinically silent optic demyelination, have a well documented role in the diagnosis of multiple sclerosis (MS). However, the sensitivity in detecting the optic abnormality as a visual function surrogate remains unsatisfied.

Objectives: To study whether low contrast VEPs can increase the sensitivity in identifying early optic demyelination.

Methods: Normal volunteers without neurological disorders and subjects who fulfilled the revised McDonald criteria for MS were recruited. Conventional PRVEPs using a video display were performed monocularly utilizing 28 foot checkerboard stimuli. The illuminate of the contrast stimuli was set up at 90%, 70%, 50%, 30%, and 10% via a computer controlled program provided by the manufacturer. Reversal rate was 1.1 Hz. Two trials were obtained from each eye. Resultant responses of N75, P100, and N145 were recorded from LO-Fpz, MO-Fpz, RO-Fpz and Fpz-A1 derivations. Filters were set at LFF: 0.5 Hz, HFF: 100 Hz. Peak latency, interpeak latency, and interlateral latency were obtained.

Results: Seven normal controls (age: 27.6 ± 9.3 , range: 18 to 45 years old) and six subjects with MS (39.5 ± 14.3 , 26 to 60) were studied. N75 and P100 latencies were significantly prolonged in the MS subjects as compared to normal controls ($95.82 \pm 6.22/77.46 \pm 2.01$ ms, $p < 0.01$; $112.82 \pm 9.51/85.71 \pm 2.57$, $p < 0.01$; and $132.83 \pm 9.14/114.07 \pm 3.06$, $p < 0.05$; $141.73 \pm 10.26/117.86 \pm 2.6$, $p < 0.02$ at the 50% and 30% contrast, respectively).

Conclusions: The authors' preliminary data demonstrated that low contrast VEPs may increase sensitivity in the early detection of optic demyelination.

CRITERION VALIDITY AND FACTORIAL ANALYSIS OF THE BOSTON CARPAL TUNNEL QUESTIONNAIRE IN RELATION TO NERVE CONDUCTION STUDIES

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Introduction: The Boston Carpal Tunnel Questionnaire (BCTQ) is a well known instrument that assesses two separate domains, function impairment and symptom severity, of patients with carpal tunnel syndrome (CTS).

Objective: To assess BCTQ structure and its relation to nerve conduction studies (NCS).

Methods: The assessment of 403 consecutive patients with clinical and/or electrophysiological definitions of CTS was performed using the BCTQ. The structure of the questionnaire was assessed by means of factor analysis. In addition, the factors obtained were compared with NCS and a neurophysiological grading scale for CTS.

Results: Factor analysis showed that three factors represented 60% of the variance of BCTQ. Factor 1 relates to the function domain questions and to the weakness and difficulty of grasping the questions of symptom domain. Factor 2 relates to numbness and tingling questions and to the pain awakening question of the symptom domain. Factor 3 relates to pain questions of the symptom domain. Factor 2 was found to have a stronger correlation with latencies and latency differences of NCS and with the neurophysiological scale than the other two factors.

Conclusions: The BCTQ assesses function and symptoms of patients with CTS by means of questions related to numbness and tingling, pain, and functional status. Among the questions, those related to numbness and tingling better correlate with electrophysiologic abnormalities distinctive of CTS and to the neurophysiologic severity of the disease.

INTRODUCTION: DIABETIC PERIPHERAL POLYNEUROPATHY IS POSTULATED TO BEGIN IN THE EARLY STAGES OF IMPAIRED GLUCOSE TOLERANCE

G-Y Park (Bucheon-si, Gyeonggi-do, Republic of Korea)

Objective: To determine if there were significant nerve conduction study (NCS) parameter dif-

ferences of the medial dorsal cutaneous (MDC), dorsal sural (DS) and medial plantar (MP) nerves among the following three groups: 1) diabetes mellitus (DM), 2) impaired glucose tolerance (IGT), and 3) healthy normal groups (HN).

Methods: Standard NCS of the sural, superficial peroneal, MDC, DS and MP were performed in all three groups. In the DM group, only those with normal sural and superficial peroneal sensory nerve action potential (SNAP) amplitudes were enrolled in this study.

Results: A total of 150 cases were enrolled (50 in each group). The mean \pm standard deviation of the MDC, DS, and MP SNAP amplitudes (uV) for the DM/ IGT, HN groups were 7.5 ± 3.9 , 7.8 ± 3.5 , 9.2 ± 5.3 / 8.4 ± 3.3 , 8.0 ± 2.9 , 13.9 ± 7.8 / 10.1 ± 3.3 , 11.2 ± 4.6 , 13.9 ± 7.2 , respectively. Statistical results showed that there were significant differences between the HN and DM group in SNAP amplitudes of these nerves. Results showed that there were significant differences between the HN and IGT group in the MDC and DS SNAP amplitudes.

Conclusions: NCSs of the three distal sensory nerves of the feet can well reflect the NCS changes in DM patients, who otherwise show normal NCS parameters of the superficial peroneal and sural nerves. The DS and MDC reflect the early changes of peripheral polyneuropathy in IGT patients. NCS of these nerves should help detect the early changes of peripheral polyneuropathy.

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CHANGE OF DERMATOMAL SOMATOSENSORY EVOKED POTENTIALS BY LOWER STIMULATION INTENSITY IN CARPAL TUNNEL SYNDROME

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Introduction: For the diagnosis of sensory cervical radiculopathy, the presence of carpal tunnel syndrome (CTS) can affect the result of dermatomal so-

matosensory evoked potentials (DSEP). However, the effect is not yet understood, especially when using lower than conventional stimulation intensity.

Objective: To investigate the change of cervical DSEP according to the severity of CTS and the stimulation intensity.

Method: Twenty-eight normal control hands and 29 CTS hands were enrolled. The DSEP study was performed with a stimulation intensity of 1.5x and 2.5x sensory threshold (ST) to normal controls and CTS patients. Stimulation sites were C6, C7, and C8 dermatomal area. The data of CTS hands were divided into mild, moderate, and severe groups. The authors analyzed the DSEP data in comparison with each dermatomal area, stimulation intensity, subjects, and DSEP values.

Results: In moderate CTS, the latency of C6 and C7 DSEPs during 1.5 x sensory threshold stimulation and C7 DSEP during 2.5 x ST were significantly delayed when compared with the values of normal subjects, but not with the mild CTS group. There was significant correlation between latency of C7 DSEP and sensory nerve conduction velocity during 2.5 x sensory threshold stimulation.

Conclusion: The authors surmise that these data can be helpful in the diagnosis of cervical sensory radiculopathy in CTS patients.

NORMAL DATA OF CERVICAL DERMATOMAL SOMATOSENSORY EVOKED POTENTIALS USING LOWER INTENSITY STIMULATION

J. Seo, M. Seo, M. Ko, S. Park (Jeonju, Jeollabuk-do, Republic of Korea)

Introduction: Dermatomal somatosensory evoked potential (DSEP) studies can be used in the electrophysiologic diagnosis of cervical radiculopathy. However, the sensitivity and the specificity of such studies remain a topic of debate. Using lower intensity stimulation in DSEP studies can be helpful in achieving increased diagnostic sensitivity.

Objectives: To establish normal reference data for the DSEP using stimulation intensity that is lower than the conventional one.

Methods: Fifty subjects without a history of neck pain or cervical spine surgery were enrolled in the study (25 adults older than 48, 25 adults younger than 32). The DSEP study was performed with stimulation intensity of 1.0×, 1.5×, 2.5× ST (sensory threshold) on right arms for C5, C6, C7, and C8 dermatomes.

Results: The mean latencies of DSEP stimulating C5, C6, C7, and C8 dermatomes with 1.5×ST intensity were 17.4 ± 1.6 ms, 22.1 ± 2.0 ms, 22.8 ± 1.4 ms and 22.7 ± 1.7 ms. The mean amplitude (N1P1) of DSEP stimulating C5, C6, C7 and C8 dermatomes with 1.5×ST intensity were 0.9 ± 0.4 μ V, 0.9 ± 0.5 μ V, 1.0 ± 0.6 μ V and 1.1 ± 0.7 μ V. The C5, C6, C7 and C8 DSEP were evoked in 84%, 98%, 100% and 96% of cases with the 2.5×ST compared with 64%, 56%, 60%, 62%, with the 1.5×ST. When one DSEP was not evoked, the DSEP of the opposite side was evoked in only four subjects.

Conclusion: This study provides normal values of DSEP with lower than conventionally used stimulation intensities. The absence of the response was bilateral in most cases.

DETAILED ORIGIN OF FAR FIELD POTENTIALS IN THE ULNAR CMAP STUDIED USING VOLUNTARY CONTRACTION

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Introduction: Recent studies have suggested that the interosseous muscles significantly contribute to the compound muscle action potential (CMAP) recorded over the abductor digiti minimi (ADM) muscle following ulnar nerve stimulation through the mechanism of the far-field potentials (FFPs). However, its details are not known yet.

Objectives: To investigate which muscle generates the FFPs in the ulnar CMAP by identifying individual motor unit potentials (MUPs) of each muscle during weak voluntary contraction.

Methods: Seven healthy volunteers were studied. Recording electrodes were placed over motor points of ulnar innervated muscles and at a common proximal reference over the forearm. The subjects were asked to perform a weak movement corresponding to the action of each muscle. Single MUPs ascribed to that muscle were identified. The authors summated MUPs from individual muscles and reconstructed CMAPs including FFPs, then compared them with the actual CMAPs.

Results: FFPs in the actual CMAPs were composed of negative (N1), positive (P1), and negative (N2) triphasic waves, N2 being often bilobed. FFPs in the reconstructed CMAPs coincided well with the actual ones. This enabled the authors to infer the origin of each FFP component. N1, P1, and early P2 components were generated primarily by ulnar and palmar interosseous muscles (the second and third palmar interossei), N1 also contributed by dorsal interossei. The ADM muscle also generated positive and negative biphasic FFPs, the latter generating the late N2 component.

Conclusions: The mechanism of FFP generation should be revisited. For example, early onset of the N1 FFP component, concurrent with the CMAP onset, cannot be explained by the theory of action potential termination.

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COOLING DIMINISHES VIBRATION INDUCED REDUCTION OF SENSORY NERVE ACTION POTENTIAL

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Introduction: It has been demonstrated that vibration reduces digital sensory nerve action potential (SNAP) amplitude. Steady firing in A- β fibers by activation of vibration receptors reduces the responsiveness of sensory nerve axons to the testing electrical stimulation. The effect of cooling on vibration induced reduction of SNAP has not been studied.

Objective: To examine the effects of cooling on vibration induced masking of SNAP.

Methods: The antidromic SNAP in the long finger was measured in four adults. Vibration stimulation was applied at 60 Hz and 2 mm amplitude on the palm near the third digit metacarpophalangeal joint. The SNAP was recorded prior to and during vibration at both 32°C and 22°C of the finger temperature.

Results: At 32°C, the average SNAP amplitude was 46.0 μ V at pre vibration and 26.5 μ V during vibration, a decrease of 43%. At 22°C, the average SNAP amplitude was 65.3 μ V at pre-vibration (increased by 42% compared to that at 32°C), and 54.5 μ V during vibration, a decrease of 17%. The onset latency of SNAP increased with cooling by 26%. Within each temperature group, there was no notable change in latency during conditioning vibration.

Conclusions: The effects of conditioning vibration on the reduction of SNAP are significantly less with cooling. The responsiveness of skin mechanoreceptors is reduced with a magnitude which is enough to overcome the adverse effect of enhanced vibration masking of SNAP by cooling, due to the increased refractoriness in the digital sensory nerve axons. The function of skin mechanoreceptors may be more significantly affected than that of large sensory nerve axons in lower temperature.

NEUROPHYSIOLOGICAL FINDINGS heralding NEOPLASTIC BRACHIAL PLEXOPATHY

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(Columbus, Ohio)*

Introduction: Invasion of the brachial plexus is a well known complication of lung cancer. When assessing the brachial plexus, sensory nerve conduction studies (NCSs) are helpful in localizing nerve injury distal to the dorsal root ganglia, and can identify milder injury when motor axon loss is not apparent. The authors report the subtle electromyography (EMG) findings in two cases of neoplastic brachial plexus invasion.

Case Reports: Case one involves a 59-year-old man with a history of tobacco abuse who presented with medial hand numbness and shoulder pain. Examination demonstrated weakness in C8/T1 innervated muscles with diminished sensation of the medial hand. EMG study was notable for reduced right ulnar sensory amplitude and active and chronic denervation in C8/T1 innervated muscles on the needle electrode examination (NEE). Case two involves a 49-year-old man with a history of tobacco abuse who presented with medial left forearm numbness, and pain in the shoulder and neck. Examination demonstrated normal motor strength, diminished medial forearm sensation, and a left Horner's. An EMG study showed reduced left medial antebrachial cutaneous sensory amplitude with active and chronic denervation in the abductor pollicis brevis on the NEE. In both cases, the electrodiagnostic findings were suggestive of lower brachial plexopathy. These findings prompted orders for a computerized tomography of the chest, despite negative chest x-rays. Eventually, testing led to a diagnosis of lung cancer.

Conclusion: These cases highlight the importance of sensory NCSs in the evaluation of brachial plexopathy. Subtle abnormalities in sensory amplitudes in high risk patients can indicate an ominous underlying process.

*William D. Arnold, MD
Junior Member Recognition Award Recipient*

DYNAMIC ELECTROMYOGRAPHY OF SPASTIC UPPER EXTREMITY IN PATIENTS WITH STROKE

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Introduction: The measurement of spasticity in stroke patients has primarily been performed with specific clinical scales. Recently, electrophysiologic analysis of the resistance of a limb to a movement has also been frequently used.

Objectives: To evaluate the specific movement pattern of the spastic hemiplegic upper extremity after stroke using dynamic electromyography (EMG).

Methods: Nine patients with spastic hemiplegia after stroke and eight control subjects were recruited. Participants were evaluated with dynamic EMG based on tasks involving individual grasp of hand and shoulder flexion. The muscular activities of upper trapezius, anterior deltoid, biceps brachii, triceps brachii, flexor digitorum superficialis, and extensor digitorum communis were investigated. Participants' hand and arm functions were evaluated with Fugl-Meyer motor assessment (FMA) before testing. Parameters evaluated by dynamic EMG were co-contraction ratio (CCR), initial contraction time (ICT) of muscle, maximal voluntary contraction force, root mean square of each muscle. The authors compared the difference of CCR and ICT of each muscle between patients and controls.

Results: Initial contraction times of flexor digitorum superficialis and extensor digitorum communis muscle between the two groups differed ($p < 0.05$). The muscle co-contraction ratio was higher in the spastic hemiplegia patients. A low FMA score correlated with increased proximal muscle tone of hemiplegic upper limbs.

Conclusions: Increased proximal tone is related to poor functional outcome of the hand. These findings are important clues in the management of patients who suffer from spasticity. Muscles tested in the present study might be selected as the points of

botulinum toxin injection or motor block for functional recovery.

NEEDLE ELECTROMYOGRAPHY STUDY IN PARKINSON'S DISEASE PATIENTS WITH DROPPED HEAD SYNDROME

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Introduction: Dropped head syndrome (DHS) in Parkinson's disease (PD) patients has been attributed to neck dystonia or unbalanced muscle rigidity in most cases, and neck extensor myopathy in rare cases. However, in Japan, needle electromyography (EMG) has rarely been performed in such patients.

Objectives: To evaluate EMG findings in PD patients with DHS and to explore whether there is evidence of myopathy.

Methods: The authors prospectively performed needle EMG in PD patients with DHS who visited a selected hospital since July of 2009. EMG was performed in the cervical paraspinal and deltoid muscles. Cervical magnetic resonance imaging (MRI) was also performed. The effects of prednisolone for the treatment of DHS were evaluated.

Results: Seven consecutive PD patients (2 men and 5 women) showed evidence of DHS. EMG revealed myopathic changes in all seven patients. EMG abnormalities were localized to the cervical paraspinal muscles. Presence of spontaneous activity in these muscles indicated that the myopathic changes were active. MRI revealed abnormal intensity in the splenius capitis muscles in three out of seven patients. Four patients were given 20mg of prednisolone per day. Three patients responded well to the therapy.

Conclusions: Findings suggest that neck extensor myopathy is not uncommon in PD patients with DHS. Needle EMG should be performed to explore the neck extensor myopathy in these patients, as there are some whose myopathy is responsive to steroid therapy.

SPECTRUM OF NEUROIMAGING FINDINGS IN C8 CERVICAL RADICULOPATHY

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Introduction: C8 radiculopathy is a distinctive syndrome (radicular pain, hand weakness, and medial hand/forearm sensory disturbance) with characteristic electrodiagnostic (EDX) findings (denervation of C8-innervated muscles with preserved sensory responses). Interestingly, C8 root impingement by C7/T1 lesions on neuroimaging studies is not often observed. The authors hypothesized that either a pre-fixed brachial plexus (C7 root compression at the C6/7 level) or upper cervical spinal cord compression resulting in lower cervical cord vascular compromise might explain some C8 radiculopathies without C8 root compression.

Objectives: To determine the spectrum of clinically relevant neuroimaging abnormalities in patients with EMG confirmed C8 radiculopathy.

Methods: Thirty-two patients with EMG confirmed C8 radiculopathy who underwent cervical magnetic resonance imaging or a computerized tomography within 3 months of EDX examination were retrospectively analyzed. The images were reviewed for the presence of: 1) severe or moderate to severe C8, C7, or T1 root compression ipsilateral to the symptoms, and 2) spinal cord compression with cord deformation or signal due to C3/4 - C6/7 disc herniations.

Results: Seven patients (22%) had C8-root compression at C7/T1. Of those without C8-root compression, five (16%) had C7-root compression at C6/7, one (3%) had T1-root compression at T1/T2, seven (22%) had cervical cord compression, four (12.5%) had low cervical intramedullary cord lesions (syrinx 2, mass 2), and eight (25%) had mild, or non specific findings.

Conclusions: C8 radiculopathy without C8 root compression on neuroimaging may be due to C7 root compression in the setting of a “pre-fixed”

brachial plexus, upper cervical spinal cord compression with vascular compromise of the distal cervical spinal cord (“myelopathic hand”), or lower cervical intramedullary cord lesions.

SUPRASCAPULAR MONONEUROPATHIES: ETIOLOGICAL CONSIDERATIONS AND DIAGNOSIS

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Objective: To identify underlying etiologies and electrodiagnostic (EDX) characteristics in patients with suprascapular mononeuropathies.

Methods: The charts of patients diagnosed with suprascapular mononeuropathy (SSMN) in the last 10 years, were reviewed. The underlying causes, if known, detailed the results of EDX testing that included nerve conduction studies (NCSs) and electromyography (EMG).

Results: Forty three patients (17 females, 26 males) with EDX evidence of SSMN (26 on left) were included in the study. Clinically diagnosed causes included trauma (n=27), postoperative (n=4), brachial plexitis/neuritis (n=9), spinoglenoid mass/cyst (n=2), and unknown (n=1). A total of 48 studies were performed (four had repeat studies). NCSs to infraspinatii (ISN) were done in 28, supraspinatii (SSN) in two, and both were done in eight studies. NCSs showed absent response (n=4), reduced amplitude (> 50% of the contralateral side) (n=11), prolonged distal latency (n=7), both amplitude and latency changes (n=9), and normal (n=7). Needle testing was performed in all studies, with both muscles sampled in 45. Of the 45, neurogenic changes were seen in both muscles for 28, ISN alone in 11, SSN alone in 1 and none in 5 studies. Isolated SSMN was noted in 62.5% of studies. Associated neuropathies included axillary (n=12), proximal median (n=2), and one each of phrenic, long thoracic, musculocutaneous, lateral antebrachial, and radial. Ten had brachial plexopathy electrodiagnostically, and three had cervical radiculopathy.

Conclusions: Trauma (71%) and brachial neuritis (20%) were the predominant causes of suprascapular mononeuropathies. Both NCSs and needle EMG should be performed in suspected SSMN cases, since 11% and 18% of the studies tested normal for one examination but had significant findings on the other.

UTILITY OF THE TIBIAL MOTOR NERVE CONDUCTION STUDY IN ROUTINE ELECTRODIAGNOSTIC TESTING

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(Washington, DC)*

Introduction: In the recognition that isolated tibial mononeuropathies are relatively uncommon and tibial motor nerve conduction studies (NCS) have technical limitations, the utility of this study is questioned in terms of routine electrodiagnostic (EDX) testing of the lower extremities.

Objectives: To examine completed EDX studies and determine if the tibial motor NCSs contributed to diagnoses when other NCSs were normal.

Methods: The authors retrospectively examined the most recent EDX studies completed at a specific institution until 50 patients were found that met inclusive criteria. In all, a common peroneal motor NCS, superficial peroneal, and sural sensory NCS were completed and interpreted as normal. The result of the tibial motor NCS was analyzed. The percentage of normal tibial motor NCS was calculated. The abnormal studies were further evaluated with respect to the clinical presentation.

Results: A total of 333 cases were reviewed in order to obtain 50 which met inclusion criteria. In 47 cases, the tibial-AH motor NCS was normal. In the other three, the nerve conduction velocity (NCV) was slightly below the lower limit of normal (42 m/s) but otherwise had normal compound muscle action potential amplitude and distal motor latency. The NCV of those three cases ranged between 37 and 40 m/s. The electromyographer deemed the findings insignificant and unrelated to the clinical presentation.

Conclusion: Limited added value exists when executing a tibial motor NCS in routine EDX studies after obtaining normal sensory studies and a common peroneal motor NCS.

A FREE WEB BASED TUTORIAL AND REFERENCE REGARDING ELECTROMYOGRAPHY IN THE UPPER EXTREMITY

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Introduction: Designed for the purposes of physician education, a free, web based tutorial and reference was created. It describes and illustrates techniques in electromyography (EMG) specific to the upper extremity (UE).

Objectives: The tutorial was created to provide a thorough, accurate, easy to understand, and reliable educational resource on the topic of EMG in the UE. It was designed for use by practicing physicians and residents, and could be accessed anywhere in the world, free of charge.

Methods: A searchable, internet based tutorial was created using the framework available at Frontal-Cortex.com, a free website dedicated to neurology education. The tutorial incorporates pages on 34 muscles in the UE, as well as the cervical paraspinous muscles at three levels. For each muscle, the tutorial describes the anatomy, and includes attachments, actions, and innervations. At least one video is provided that details each muscle, demonstrating needle placement and activation. Interactive anatomical illustrations are provided for most muscles.

Results: The tutorial has received over 90,000 hits since its inception in April of 2009. User demand for individual muscles varies. The abductor pollicis brevis page has been requested 1,732 times. Feedback from users has been positive. It is usable on desktop, laptop, and palmtop devices with internet capability.

Conclusions: The EMG tutorial provides a free, easily accessible educational resource for physicians. The multimedia content should facilitate mul-

multiple methods of learning and retention. This internet based resource should be of value to residents, and to those in locations with few economic resources.

Study supported by FrontalCortex.com

DOES STRETCHING THE SKIN BEFORE NEEDLE INSERTION REDUCE THE PAIN OF ELECTROMYOGRAPHY?

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Objective: To study whether stretching the skin before needle insertion reduces the pain of electromyography (EMG).

Background: EMG is a valuable technique for recording motor unit action potentials from skeletal muscles. Pain during the performance of needle EMG is an important clinical problem. It distresses the patient and can interfere with diagnostic accuracy. Anecdotal observation by experienced electromyographers suggests that needle EMG causes less discomfort if the skin is stretched prior to needle insertion.

Design/Methods: Retrospective chart reviews were performed on 10 charts from the Drexel University College of Medicine EMG laboratory. The proposal was examined by the institutional review board. The pain scale was recorded for each patient (0 to 10, with 0 being no pain and 10 being the worst pain) with and without stretching the skin while inserting the needle. These results were compared. All studies were performed by the same electromyographer. Statistical analysis was completed to determine if there was a difference between the pain scores reported by patients with and without stretching the skin.

Results: The mean pain score reported by patients with the skin stretched before needle insertion was 1.7, while the same patients reported a pain score of 4.6 when the needle was inserted without stretching the skin. This value reached statistical significance (p value: 0.013).

Conclusions: The authors concluded that stretching the skin prior to needle insertion may significantly decrease the pain of needle EMG.

SUPERNORMAL ROUTINE MEDIAN SENSORY NERVE CONDUCTION VELOCITIES PREDICTS AGAINST THE PRESENCE OF CARPAL TUNNEL SYNDROME

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Introduction: Carpal tunnel syndrome (CTS), the most common entrapment neuropathy in clinical practice, is frequently encountered in the electrodiagnostic (EDX) laboratory. In cases of mild CTS, a variety of comparison studies add sensitivity to the diagnostic protocol when routine nerve conduction studies are normal. The necessity of performing these additional studies, however, is not always clear in patients with "supernormal" median sensory nerve conduction velocities (NCV), which is defined as greater than 60 m/s.

Objectives: To determine whether a supernormal median sensory NCV obviates the need for additional comparison studies.

Methods: A retrospective review of 1206 EDX studies in patients referred for evaluation of CTS was conducted. All studies included routine testing of the median sensory NCV and palmar mixed nerve comparison studies. Patients were stratified by median sensory NCVs, beginning at NCV below 50 m/s and in 1 m/s increments up to 70 m/s. For each 1 m/s increment in NCV, the authors determined the percentage of patients with an abnormal palmar mixed nerve comparison study.

Results: The palmar mixed nerve comparison study was abnormal in only 11 of 237 patients (4.6%) with supernormal sensory NCVs. None of the 71 subjects with a median sensory NCV greater than 66 m/s had an abnormal comparison study.

Conclusions: A supernormal median sensory NCV strongly predicts against abnormalities of palmar mixed nerve comparison studies. The re-

sults of this study suggest that comparison studies may be avoidable in patients with sensory NCVs greater than 66 m/s.

INPUT OUTPUT CURVE OF CUTANEOUS SILENT PERIOD AND THE NUMBER OF TRACES AVERAGED

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Introduction: In the clinical setting, there has been an increased use of cutaneous silent period (CSP). However, the number of traces averaged varies from only a few to 200. To ensure patient cooperation and efficiency, the fewest necessary number of traces should be identified.

Objectives: To determine the minimum number of traces that preserves an input output relationship between stimulus intensity and CSP duration.

Methods: CSP was evoked from the index finger and recorded in the abductor pollicis brevis muscle in 16 healthy subjects (age 40 ± 11 years, 7 men) at stimulus intensities of 2.5, 5, 10, and 20 times individual perception threshold (xPT, 1.4 ± 0.4 mA). In total, 60 traces were recorded per intensity in each subject. Individual traces were rectified and consecutively averaged in blocks of 5, from N=5 to N=60. CSP duration was calculated over the segment where electromyography was below 80% of pre stimulus baseline. Data were pooled across subjects in order to fit and compare the input output curves between different blocks.

Results: CSP duration increased with stimulus intensity but did not significantly differ between the blocks (i.e., 2.5xPT: mean CSP 16 ms for N=60 versus 10 ms for N=5; 20xPT: 94 ms for N=60 versus 93 ms for N=5). The sigmoid function fitted input-output curves well (i.e., $R^2=.67$ for N=60; $R^2=.65$ for N=5). Curve parameters did not significantly differ between the blocks.

Conclusions: Averaging less than 60 and as few as 5 traces adequately describes the relationship

between stimulus intensity and CSP duration. Future studies should determine the utility of this approach for studying CSP abnormalities in different patient populations.

Study supported by The Wilson Research Foundation, Jackson, MS

RELATIONSHIP BETWEEN PAIN AND THE STIMULUS DURATION IN NERVE CONDUCTION STUDIES

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Introduction: It is generally believed that an electrical stimulation with shorter duration is less painful for the subject in nerve conduction studies. However, whether this holds true for stimulations with the same physiological effects has not yet been verified.

Objectives: To examine the relationship between the level of pain and the duration of the stimulations that achieve maximal or supramaximal stimulation for the motor nerve.

Methods: The tibial nerve was stimulated at the ankle in 14 control subjects. Two settings were examined: 1) single supramaximal stimulation, and 2) 1 Hz train of five maximal stimulations. Stimulations were given that satisfied the above condition with two out of three durations of 0.05, 0.2 or 1.0 ms consecutively. The subject was asked which was more painful. Two scores were assigned to each duration, depending on the answer. A higher score was given for a more painful duration. Six such trials were repeated. The scores were totaled for each duration.

Results: The average scores for all the subjects were 4.4, 2.2 and 5.4 for 0.05, 0.2, and 1.0 ms durations, respectively in setting 1, and 4.0, 1.9 and 6.1, respectively in setting 2. The 0.2 ms duration was significantly less painful than the other two durations for both settings.

Conclusions: A shorter duration was not always found to be less painful. The 0.2 ms duration, which is widely employed in Japan, might be the appropriate duration. Furthermore, if a 0.05 ms duration is used, the intensity necessary for supra-maximal stimulation may easily exceed 100 mA for a pathological nerve with increased threshold.

DETERMINING WHICH TEST SHOULD BE USED LONG TERM IN SEVERE FORMS OF CARPAL TUNNEL SYNDROME

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Background: In long term hemodialysis patients, carpal tunnel syndrome (CTS) is the most common complication. In spite of the established diagnostic guidelines and many different methods of nerve conduction studies (NCSs), it is sometimes difficult to distinguish very advanced CTS (absent median motor and sensory responses) from uremic neuropathy. In the last few years, the 2LI-DML test has been accepted as a reliable method for the diagnosis of even the very advanced stages of CTS.

Objective: To evaluate the usefulness of NCSs based on the interlatency difference (2LI-DML) between the second lumbrical (2L) and second dorsal interosseous (2I) in the diagnosis of severe forms of CTS in long term hemodialysis patients with superimposed polyneuropathy.

Method: The 2LI-DML test was used in four cases of long term hemodialysis with severe damage to the median nerves (absent median motor and sensory responses) and with forearm arteriovenous fistulas and concomitant uremic polyneuropathy. The presence of forearm arteriovenous fistulas makes needle electrode test impossible to apply.

Results: The 2LI-DML test allowed for a confirmed clinical diagnosis of CTS in the patients studied. A mean difference of motor latency between the second lumbrical and the second interosseous muscles was significantly prolonged and amounted to 10.91 + 2.36 ms.

Conclusions: The 2LI-DML test is the only test that may be used to diagnose CTS in patients with severe damage to the median nerves and with forearm arterio venous fistulas and superimposed polyneuropathy. The test is efficient, simple, and well tolerated by such patients.

PEDICLE SCREW ELECTRICAL RESISTANCE HYDROXYAPATITE COATED VERSUS NON COATED

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Introduction: Stimulus evoked electromyography (EMG) testing has become a common tool for assisting in the confirmation of proper placement of pedicle screws during spine surgery. Hydroxyapatite (HA) coated screws have recently been introduced as a means of increasing "pullout" strength. It is the manufacturer's recommendation that HA coated screws not be stimulated due to inconsistent responses. There is no published data to confirm this recommendation.

Methods: Resistance measurements were obtained from a random sampling of 10 HA coated pedicle screws and ten noncoated screws. All screws were the same diameter (6.5 mm) and length (45 mm). Resistance measurements were taken from the hexagonal head slot through the shank of the screw to simulate surgical conditions, as well as at each thread. Surface resistivity measurements were also taken to determine voltage drop over its entire length.

Results: The non coated screws tested showed low resistive properties and proved to be an ideal conductor of electrical current. The resistive properties associated with the HA coated pedicle screws were found to be similar to those of commonly used insulators such as rubber and plastic, removing the effectiveness of evoked EMG testing.

Conclusions: This data suggests that the increased resistance value of the HA coated screw is large enough that any EMG response produced would be due to shunting of electric current from the non coated head of the screw into adjacent tissue and not through

the shank of the screw. These study results suggest that HA coated screws should not be stimulated as a means of assisting in pedicle screw placement.

TWO CASES OF POLIOMYELITIS WITH INTRAVENOUS IMMUNOGLOBULIN RESPONSIVE RADIAL NEUROPATHIES

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Introduction: Serological and pathological markers of inflammation are present in patients with a history of poliomyelitis, but neither intravenous immunoglobulin (IVIg) nor steroids has improved strength in placebo controlled trials of these patients. Two patients with a history of childhood poliomyelitis and late in life focal radial neuropathies responsive to IVIg are presented.

Case Reports: A 69-year-old man presented with 6 months of progressive painless wrist drop. Electromyography (EMG) demonstrated motor conduction block in the radial nerve below the spiral groove (normal radial sensory response). The second patient, a 54-year-old woman, presented with 2 years of progressive finger and then wrist drop. EMG demonstrated a posterior interosseous mononeuropathy. Both patients' radial neuropathies were found in the limb most severely affected by their childhood poliomyelitis. Serologic and spinal fluid investigations were unremarkable in both cases. Both patients responded to IVIg (2 months and 6 months of treatment, respectively) and are asymptomatic at 7 years follow up. The diagnosis of an immune mediated mononeuropathy akin to multifocal motor neuropathy is postulated, and has not been previously described in patients with a history of poliomyelitis.

Conclusion: These two cases suggest that 1) patients with late progression of poliomyelitis need electrophysiologic evaluation; 2) a subset of patients with late progression of weakness in poliomyelitis may respond to immunomodulation; and 3) revisitation of the relationship between poliomyelitis in childhood and autoimmune neuropathy late in life may be merited.

Study supported by Geisinger Medical Center

CHARACTERIZATION OF F WAVES BY RECURRENCE QUANTIFICATION ANALYSIS

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Introduction: Following the belief that most physiological variables are non linear, F-waves are inherently variable and arise from non linear and non stationary processes. Recurrence quantification analysis (RQA) is a technique designed to analyze non linear events. It is based on the likelihood that a dynamic system will return to a particular state. Recurrence plots can take a single physiologic variable and plot it in multiple dimensions to identify time correlations.

Objectives: To determine if RQA, coupled with principal component analysis (PCA), would be useful in differentiating patterns of F wave abnormalities.

Methods: Tibial F-waves were recorded from the abductor hallucis muscle using a standard protocol following 20 supramaximal stimuli and subjected to RQA. Such analyses were performed on seven control subjects and 45 patients seen in the Clinical Neurophysiology Laboratories at the Hines Veterans Affairs Hospital. The patients had established tibial nerve dysfunction due to polyneuropathies or radiculopathies as determined by history, physical examination, and electrodiagnostic information.

Results: RQA allows for description of recurrence in eight different variables, each of which speaks to a different quality of the recurrence pattern. Recurrence variables from all patients were assembled into a large matrix and normalized before being subjected to PCA. By plotting the first principal component as a function of the second, the data could be partitioned into separate clusters

Conclusions: The results underscore the potential of coupled RQA and PCA and provide a meaningful classification of F-wave abnormalities and peripheral neuropathic dysfunction.

ALTERATION IN MUSCLE ELECTRICAL ANISOTROPY AFTER SCIATIC NERVE CRUSH IN THE RAT

J. Li, L.L. Wang, S.B. Rutkove (Boston, MA)

Introduction: Anisotropy is a property that comes from applied high frequency electrical current that flows preferentially along, rather than across muscle fibers. Alterations in tissue structure after nerve or muscle injury may impact this characteristic and thus serve as a novel means of assessing neuromuscular disease. Limited human data suggests that such changes occur. Whether these changes can be modeled effectively in the rat is unknown.

Objective: To assess the effect of controlled denervation injury via sciatic crush to surface measured anisotropy in the rat.

Methods: Twelve young male rats underwent unilateral sciatic crush. The 2 kHz to 1 MHz electrical impedance of the gastrocnemius soleus complex was measured using a standard apparatus. The anisotropy was assessed by using small stainless steel electrodes placed at 0, 45, and 90 degrees relative to the major muscle fiber direction.

Results: Anisotropy, as measured across the three angles, increased shortly after sciatic crush. For example, the anisotropy difference (90 to 0 degree values) for phase at 100 kHz increased from a baseline value of 1.88 to 5.40 degrees at 1 week post injury. These alterations gradually resolved over a 2 month period of time, nearly returning to baseline with a value of 1.94 degrees.

Conclusions: Measurement of electrical anisotropy holds the promise of offering a novel, non invasive means for assessing neurogenic change in muscle.

Study supported by NIH/NINDS

VALIDATION OF ALTERATION ELECTROPHYSIOLOGIC SCALE IN PATIENTS WITH LUMBOSACRAL RADICULOPATHY

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Introduction: Electrophysiologic study is frequently requested in patients with suspicion of lumbosacral radiculopathy. One limitation of the evaluation is the absence of normatization for determination of the degree of abnormality.

Objective: Design and validate a scale to categorize the severity of qualitative and electrophysiological findings in patients with lumbosacral radiculopathy.

Methods: The scale was applied to ninety-one qualifying patients remitted for an electrophysiology study. These patients were evaluated clinically; with magnetic resonance imaging of lumbosacral spine. Quality of life scales related to health SF-36 and the questionnaire of the activities of Roland Morris were also given. The final scale was the product of the academic discussion in the research group and group external evaluators who work in the area of electro diagnostics.

Results: The authors discovered a significant correlation between the scale and prevalence of pain in extremity (p: 0,000), the level of abnormality in the neurological clinical examination (absent Achilles reflex p:0,011, weakness in muscles dorsiflexores or plantiflexores of neck of foot p:0,045 and 0,033, respectively), as with the scores in the domain of corporal pain of the SF-36 (p:0,048) and the degree of imaging abnormality (p:0,019).

Conclusions: The authors believe that the proposed scale can be applied to assess the severity of electrophysiological abnormalities in a patient with suspected lumbosacral radiculopathy by obtaining consistent and reliable results.

Study supported by National University of Columbia

NEUROPHYSIOLOGIC STUDY IN ADULTS WITH CARNITINE DEFICIENCY

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Introduction: Carnitine is an essential substance that transfers long chain fatty acids across the mitochondrial membrane for subsequent beta oxidation. The authors recently reported that carnitine deficiency (CD) in adults may cause peripheral and central nervous system (PNS and CNS) dysfunction. However, the neurophysiologic features in CD related PNS and CNS dysfunction are unknown.

Objectives: To evaluate the neurophysiologic features in adults with CD.

Methods: Subjects with primary carnitine deficiency seen in the neuromuscular clinic from October of 2005 to January of 2010 were included. Subjects with secondary carnitine deficiency were excluded. Standard nerve conduction study and electromyography were performed on at least one arm and one leg. Motor neuron number estimation (MUNE) was performed using a statistical method. Conventional somatosensory, visual, and brainstem auditory evoked potentials (SSEP, VEP, and BAEP) were performed. VEP was recorded using both a video display with pattern reversal and goggles fitted with a mosaic of light emitting diodes.

Results: Six subjects with an age range of 38 to 53 years old (40.0 ± 5.5 , mean \pm SD), were studied. Four were male, the other two were female. One subject with cervical spondylosis/myelopathy was subsequently excluded. Neurophysiologic studies showed mild mononeuropathy (two of five); initially diffusely active denervation (two of five) and superimposed mild myopathy (two of five) both subsequently improved after carnitine supplementation. Delayed SSEP with median nerve stimulation (two of five), bifid or prolonged VEP (three of five), and normal BAEP (five of five) were seen. MUNE was normal (two of two). One subject had neurophysiological autonomic dysfunction.

Conclusion: This preliminary study demonstrates neurophysiologic evidence underlying the pathophysiology for adults with CD caused PNS and CNS dysfunction. A large scale study is warranted to validate the findings.

CLINICAL AND NEUROPHYSIOLOGIC STUDY ON IDIOPATHIC TRIGEMINAL NEURALGIA

J. Luo, M. He, F. Bumanlag, D. Laske, C. Miyamoto (Philadelphia, PA)

Introduction: Trigeminal neuralgia (TN) is characterized by sudden, severe, brief, stabbing, recurrent episodes of pain in the distribution of one or more branches of the trigeminal nerve. The responses to medical and surgical treatments in TN patients are diverse and range from effective to refractory. It is unknown whether a relationship exists between the selection of a therapy and the trigeminal excitability.

Objectives: To study whether trigeminal hyperexcitability plays a role in idiopathic TN.

Methods: The authors retrospectively reviewed the charts and neurophysiology databank to identify subjects with TN seen in a neurology clinic from April 1 of 2004 to January 31, of 2010. Subjects who fulfilled the American Academy of Neurology diagnostic criteria for TN were collected. Subjects with either neurologic deficits on examination or an identifiable structural lesion on neuroimaging studies were excluded. Neurophysiologic studies included bilateral nerve conduction on facial nerves, blink reflexes by separately stimulating unilateral ophthalmic nerve and recording on bilateral orbicularis oculi, and electromyography on the muscles of frontalis and masseters supplied by the facial and trigeminal nerve, respectively.

Results: Forty-five subjects (age: 55 ± 16 , range: 30 to 83 years; Female/male: 34/11=3.1/1) were included. Involvement of the V2 and V3 branches of trigeminal nerve was predominant (V1: 13.9%, V2: 19.4%, V3: 19.4, V1+V2: 5.6%, V2+V3: 30.6%,

V1+V2+V3: 11.1%). Initial treatments included carbamazepine (17/12/6, subjects: effective/ineffective/unknown); oxcarbazepine (1/2/1); gabapentin (0/2/0); baclofen (1/1/0); Lyrica(1/0/0/); Motrin (1/0/0); and amitriptyline (1/0/0). Three patients received surgical intervention. Six patients underwent neurophysiologic studies, which showed hyperexcitability in two subjects.

Conclusions: The results suggest that hyperexcitability may play a role in the underlying pathophysiology for some TN patients. Further prospective studies to confirm these preliminary findings are underway.

PERIPHERAL AUTONOMIC NERVE AND CENTRAL SPINOTHALAMIC DYSFUNCTION IN HEREDITY SPASTIC PARAPLEGIA 7

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Introduction: Hereditary spastic paraplegia 7 (HSP7), associated with SPG7 mutation encoding the mitochondrial protein paraplegin, is characterized by progressive lower limb weakness and spasticity with absent or variable sensory deficits. In one published clinical series, 0/22 affected individuals had loss of pin prick sensation, while 3/22 experienced some loss of vibration sense.

Case Report: A 49-year-old man presented with isolated stiffness of the lower limbs that caused difficulty running. Examination showed normal mental status, cranial nerves, and muscle strength. Other findings included distal hypoesthesia to pinprick, lower limb hyperreflexia, spasticity, and extensor plantar responses.

Genetic testing detected findings of hereditary spastic paraplegia 7, SPG7 variant 2 with gly349-ser at nucleoside position 1045.

Neurophysiological testing demonstrated normal nerve conduction studies, electromyography, somatosensory evoked potential, visual evoked response, and brainstem auditory evoked response

findings. Quantitative sensory testing showed elevated cooling detection thresholds and normal vibration detection thresholds. Autonomic reflex screen revealed mild patchy sudomotor and adrenergic dysfunction. Contact heat evoked potential stimulation showed low amplitude or absent responses with prolonged scalp latencies. Skin biopsy for epidermal nerve fiber density was normal in the calf and thigh.

Conclusion: In addition to corticospinal tract findings, the pattern of small fiber sensory deficits, physiological abnormalities, and normal epidermal nerve fiber densities suggest that this variant of HSP7 can include peripheral autonomic neuropathy and central spinothalamic pathway involvement.

CLINICAL AND NEUROPHYSIOLOGICAL FINDINGS OF BRACHIAL PLEXOPATHIES AT A LEVEL 1 TRAUMA CENTER

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Introduction: The brachial plexus is formed by the C5 to T1 nerve roots and supplies the upper limbs. Trauma is the most common cause of brachial plexopathy, although non traumatic causes may include idiopathic, infectious, cancer related, or iatrogenic processes.

Objective: To examine the differences in the extent and distribution of clinical and neurophysiologic involvement caused by traumatic and non traumatic injuries to the brachial plexus.

Methods: A retrospective review of electromyography (EMG) charts of the past 5 years from an electrodiagnostic (EDX) laboratory of a large urban county hospital was conducted. Nerve conduction studies and EMG data were reviewed. All cases meeting diagnostic criteria for brachial plexus involvement were identified. Clinical data regarding presentation and course was obtained from the medical record.

Results: A total of 678 EDX files from 2004 to 2009 were reviewed; 47 cases of brachial plexopa-

thy were identified. A vast majority were traumatic, followed by idiopathic plexopathies. Mechanisms of traumatic injury included motor vehicle accidents, gunshot wounds, and falls. Sixteen cases involved the upper trunk, 13 involved the lower trunk, and 9 involved all trunks. Most EDX studies were performed within the first 3 months of symptom onset. Serial studies were done in a few select cases. The main referral sources were surgical subspecialties.

Conclusions: Most brachial plexopathies at a level 1 trauma center were traumatic in etiology, and the majority had significant axon loss injury with variable outcomes. Further longitudinal studies are needed to better determine clinical and electrophysiologic factors that predict long-term outcomes.

CEVICAL RADICULOPATHIES DIAGNOSIS WITH DERMATOMAL SENSORY EVOKED POTENTIALS

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Introduction: Electrodiagnostic (EDX) tests are performed across the world to detect cervical radiculopathies in patients with symptoms related to the cervical spine.

Objectives: To demonstrate the diagnostic yield of dermatomal evoked potentials (DEP), and electromyography (EMG) in patients with cervical radiculopathies.

Methods: The study was a self controlled trial performed on 301 patients who were referred to a specific neurodiagnostic laboratory for the evaluation of possible cervical radiculopathies. Each patient had DEP with surface electrical stimulation over anatomically defined dermatomes. The electrical stimulation would only stimulate the specific dermatome. Recordings were made from Erb's point, cervical spine and cortical electrodes (P1) with measurement of their latencies. C4-C8 dermatomes stimulated separately and bilaterally with standard averaging with each stimulation. EMG

was performed on 167 patients in this group using a disposable monopolar needle. During EMG testing, cervical paraspinal muscles and C4-C8 innervated muscles in the upper limbs were evaluated.

Results: The DEP was abnormal on 203(301) patients 67.4%. The EMG was abnormal on 84 (167) 49.7 %. The t test had a P value of <0.0001 and ANOVA single factor analysis showed P < 0.0001.

Conclusion: EMG examination has been routinely used for evaluating radiculopathies, yet changes must occur in motor fibers for this test to show the abnormalities. Patients with sensory fiber related symptoms may have a negative result. This study indicates that DEP can be useful and may be superior to EMG for detecting radiculopathies. A combined use of EMG and DEP may enhance diagnostic abilities in patients with cervical radiculopathies.

CHARACTERISTICS OF MYOTONIC DISCHARGES IN NEUROGENIC DISORDERS

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Introduction: Myotonic discharges (MDs) are muscle fiber action potentials that wax and wane at rates of 20 to 80 HZ. Although they are frequent in myotonic dystrophies, they also occur in other myopathies and neurogenic disorders. Neurogenic MDs are often thought to be of short duration and primarily waning, although this has not been systematically studied.

Objective: To measure the characteristics of MDs that occur in neurogenic disorders.

Methods: MDs were recorded as part of a larger, institutional review board approved, consented study of MD characteristics during standard electromyographic (EMG) testing for neuromuscular disease. MD recorded in patients with well defined neurogenic disorders were stored electronically and studied offline. The authors measured the number of spontaneous and evoked individual MDs per muscle. The following were measured for each discharge: a) total duration, b) maximum and mini-

mum rate of firing, c) maximum amplitude, d and e) number of times the rate and amplitude change, and f) direction of each change. MD characteristics were compared with clinical findings.

Results: Thirty-seven MDs in 24 muscles from 12 patients (radiculopathy, brachial plexopathy, and polyneuropathy of 6 to 480 months) were studied. The average duration of neurogenic MD was 9.3 seconds (0.6 to 51); 29 of 37 MDs were waxing in frequency (exponential in all); 29 of 37 showed change in amplitude (eight increased); maximum and minimum rates were 20 to 125 and 4 to 83.

Conclusions: Neurogenic MDs may be long or short in duration, with many lasting over 10 s. Waxing and waning discharges, similar to those in myopathies were seen. No unique features were identified. None were recorded in patients with less than 6 months of disease.

ULNAR NERVE COACTIVATION THROUGH MEDIAN NERVE STIMULATION AT THE WRIST: A STUDY IN 62 HANDS OF 31 HEALTHY VOLUNTEERS

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Objective: To determine the threshold level of ulnar nerve coactivation by median nerve stimulation in healthy subjects and to correlate coactivation with wrist circumference, body weight, and body mass index (BMI).

Methods: Median nerve stimulation at the wrist was done in 62 hands of 31 healthy volunteers. Compound muscle action potentials (CMAP) were recorded from abductor pollicis brevis and abductor digiti quinti (ADQ) simultaneously. Coactivation was present if a reproducible CMAP could be recorded from the ADQ by median nerve stimulation. A t-test was performed to compare the group with coactivation to the group without coactivation in regards to wrist circumference, body weight, and BMI.

Results: In 35 hands, coactivation of the ulnar nerve was elicited by stimulation of the median nerve. The average stimulation intensity was 45 mA (standard deviation (SD) 13,1, range 20 to 70 mA). Stimulation intensity was an average of 27,4 mA (SD 13,7, range 2 to 60 mA) higher than the supramaximal stimulation intensity for the median nerve. There was a significant difference for BMI ($p < 0,001$), body weight ($p < 0,001$), and wrist size ($p < 0,003$) between the group with coactivation and the group without coactivation. BMI and wrist circumference were significantly lower in the coactivation group.

Conclusion: Coactivation of ulnar nerve could be elicited in young healthy subjects with stimulation intensities used to examine injured nerves. This source of error must be considered when stimulating the median nerve at the wrist, especially in slender and smaller people.

THE INVESTIGATION OF SYMPATHETIC SKIN RESPONSE IN PATIENTS WITH DIABETIC NEUROPATHY

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Introduction: Diabetic polyneuropathy is one of the most commonly encountered polyneuropathies in the world. Sympathetic skin response (SSR) test is a technique used for assessment of the sympathetic functions and measuring the sudomotor activity of the skin.

Objective: The aim of this study is to investigate sympathetic skin response and to establish its diagnostic value for autonomic dysfunction in patients with diabetic neuropathy.

Methods: In 78 diabetic patients (33 females and 45 males) and in 30 normal control subjects (20 females and 10 males), SSR were recorded. Nerve conduction studies and SSR test were performed in all patients: motor nerve conduction velocity (MNC) of median, ulnar, peroneal and tibial nerves and sensory nerve conduction velocity (SNC) of median,

ulnar and digital plantar medial nerves. The SSR test was performed by electrostimulation of median and tibial nerves and recording with surface electrodes from the palm and the sole. The latency and the amplitude of the SSR were measured.

Results: The SSR was present in 100% of the control subjects with: mean latency 1.36 ± 0.14 sec from the hands and 2.21 ± 0.19 sec from the feet and mean amplitude 1.51 ± 0.58 mV from the hands and 1.21 ± 0.53 mV from the feet. The SSR was absent in 35 patients from the 78 investigated.

Conclusions: The results of this study suggest that SSR is noninvasive and simple test for evaluation and early determination of autonomic dysfunction in patients with diabetic polyneuropathy. It can be used as a routine method in electrophysiological examination.

MOTOR RESPONSES ELICITED BY DIRECT MECHANICAL STIMULATION OF PERIPHERAL NERVES IN AMYOTROPHIC LATERAL SCLEROSIS PATIENTS

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Introduction: Previous studies have demonstrated that amyotrophic lateral sclerosis (ALS) patients have widespread disturbances in axonal excitability. The evaluation of motor responses elicited by compression or mechanical stimulation of peripheral nerves (MREC) could be used to quantify motor axon excitability. There are no previous studies of MREC in ALS.

Objectives: To describe the electrophysiological features of MREC in ALS patients.

Methods: Twenty-one ALS patients and 17 healthy controls were studied. The MREC were elicited using two types of stimulation to the peroneal nerve at the fibular head (1) mechanical compression: briskly sliding the index and middle fingers over the nerve trunk, transversally, like pulling a guitar string and (2) tapping the nerve with a reflex ham-

mer. Motor potentials were recorded from the extensor digitorum brevis muscle. Peak to peak amplitude and the percentage of occurrence of MREC, over 10 trials, were measured.

Results: The percentage of occurrence of MREC was significantly higher in ALS patients than in controls: Mechanical stimulation ($75.5 \pm 21.3\%$ vs. $25.5 \pm 12.2\%$, $p=0.009$); percussion: ($35.5 \pm 11.3\%$ vs. $12.5 \pm 5.2\%$, $p=0.04$). ALS patients showed significantly increased MREC amplitudes than controls: Mechanical compression: (1200 ± 250 uv. vs. 900 ± 236 uv., $p=0.001$); percussion: (650 ± 145 uv. vs. 450 ± 175 uv., $p=0.001$).

Conclusion: These findings demonstrate that ALS patients have increased peripheral motor axon excitability, which can be identified with MREC, particularly with mechanical stimulation. The clinical evaluation of MREC, just by compression or percussion of the nerve and observing the elicited muscle twitch, could be used as a simple, replicable and widely available surrogate tool to test axonal excitability in ALS patients.

Study supported by Cuban Ministry of Public Health

INCIDENCE OF MYASTHENIA GRAVIS BASED ON SCREENING DATA OF MGTX STUDY

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The MGTX study, now on its 5th year, has been re-funded by NINDS for another 5 years. It is a single-blinded, multicenter, two-arm trial that aims to determine whether in non-thymomatous MG extended transsternal thymectomy (ETTX) plus prednisone compared to prednisone alone result in a greater improvement in myasthenic weakness. We aim to recruit 150 MG eligible patients based on

our inclusion and exclusion criteria. The primary outcome measure will be the total prednisone exposure of the two treatment groups over the three years conditional on the overall clinical state.

Sixty six centers from around the world are participating in this study. We have screened over 6000 patients. We describe the incidence of MG over time based on our screening data. In particular, we look at the gender and age distributions. We find that before 1980s, females diagnosed with MG have higher proportions than males. However, this difference in proportion starts to decrease over time. Comparing the age when first MG symptoms were observed, the number of patients over 50 showing first symptoms has increased over time while the proportion of patients in the age group of 30 or younger showing first symptoms has decreased over time. Furthermore, females tend to show first MG symptoms at an early age (younger than 30) while males tend to be diagnosed at a later age (older than 50).

HOW MYASTHENIA GRAVIS ALTERS THE SAFETY FACTOR FOR NEUROMUSCULAR TRANSMISSION

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Introduction: Endplate Na⁺ channels increase the efficiency of neuromuscular transmission by reducing the threshold depolarization for triggering an action potential (EAP). In MG, AChRs and Na⁺ channels are lost from the neuromuscular junction. We previously demonstrated in moderate to severe MG (MGFA IIIa) that Na⁺ channels as well as AChRs were lost from the endplate membrane. In this study, we examined how the loss of Na⁺ channels and AChRs impacts the safety factor for neuromuscular transmission (SF) in 2 patients with MGFA IIa grade MG.

Methods: We used standard electrophysiological techniques to compare membrane currents and potentials in intercostal nerve-muscle preparations from controls and 2 MG patients (MGFA IIb).
Results: Endplate AChR loss decreases the size of

the endplate potential (EPP) by 25% from 40.2 ± 1.3 mV to 30.1 ± 1.9 mV ($p < 0.001$). Endplate Na⁺ channel loss in MG increased EAP by 41% from 13.5 ± 2.4 mV to 19.0 ± 2.5 mV ($p < 0.05$). The SF for neuromuscular transmission was reduced from 2.98 to 1.58. If the EPP was normal, the SF for MG fibers would have been 2.12. If the EAP was normal, the SF for MG fibers would have been 2.23. Decreased EPP accounted for 47% of the reduction in SF and the increase in EAP produced 53% of the SF reduction.

Discussion: Endplate Na⁺ channel loss accounts for about half of the SF reduction in 2 patients with MGFA grade IIa MG.

Study supported by Department of Veterans Affairs Research Service

MG-ADL: IS IT STILL A RELEVANT OUTCOME MEASURE?

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Introduction: The myasthenia gravis Activities of Daily Living scale (MG-ADL) is a quick and simple eight-item measure that requires no special equipment or training. It has been used in a variety of MG clinical trials and in routine practice for over a decade. The aim of this analysis is to examine the correlation of MG-ADL with newer, validated MG-related outcome measures and clinical improvement.

Methods: In a multicenter study, MG patients were assessed with several MG outcome measures on two consecutive visits. We analyzed the MG-ADL, MG Composite (MGC), 15-item Quality-of-Life (MGQOL15) and physician global impression of change. Statistical tests included descriptive analysis, Pearson correlation and sensitivity/specificity.

Results: A total of 87 patients completed MG-ADL, MGC and MGQOL15 on the first visit and 76 returned for the second visit. The mean initial MG-ADL score was 4.89 (SD 3.54) and 3.59 (SD 3.3) at the second visit. At the first visit, there was a strong positive correlation between the MG-ADL and MGC ($r=0.85$, $P<0.0001$) and between the MG-ADL and MGQOL15 ($r=0.76$, $P<0.0001$). Correlation in the delta MG-ADL score and physician global impression of change between the two visits was also strong ($r=0.70$, $P<0.0001$). Sensitivity/specificity analysis revealed a 2-point improvement in MG-ADL best predicted clinical improvement.

Conclusion: The MG-ADL correlates strongly with newer MG measures and an improvement in score correlates with clinical improvement. The MG-ADL is useful both as a research tool and in routine clinical management.

RELATIONSHIP BETWEEN THE QUANTITATIVE MYASTHENIA GRAVIS SCALE AND CLINICAL, IMMUNOLOGICAL AND ELECTROPHYSIOLOGICAL MARKERS OF DISEASE STATUS IN MYASTHENIA GRAVIS

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Introduction: The Quantitative Myasthenia Gravis Scale (QMGS) is a standard research tool used to assess outcomes in myasthenia gravis (MG) research trials. The relationship between QMGS and other clinical, electrophysiological and immunological markers of disease status in MG has not been comprehensively analyzed.

Methods: Patients included in this cross-sectional study took part in one of two recent randomized controlled trials evaluating the effectiveness of intravenous immunoglobulin (IVIG) vs placebo or plasmapheresis in MG. Baseline QMGS was analyzed with respect to a) MGFA classification status b) baseline acetylcholine (AchRAb) and muscle specific tyrosine kinase (MuSK) receptor antibody status and quantitative levels c) compound muscle action potential amplitude (CMAP) decrement on repetitive nerve stimulation d) single fiber electromyography jitter and abnormal or blocked pairs e) presence of thymoma

Results: 134 patients with MG from two randomized studies were included in the analysis. QMGS showed an excellent correlation with the MGFA Classification ($r=$, $p<0.001$). QMGS was non-significantly highest in the AchRAb positive patients (14.2) compared to the MuSK positive (13.0) and seronegative (12.8) MG patients ($p=0.13$). No significant relationship was found between the QMGS and quantitative AchRAb levels. The QMGS was higher in patients with thymoma compared to those without thymoma (15.1 vs 13.1, $p=0.04$). There was a moderate correlation between QMGS and all electrophysiological parameters: decrement on repetitive nerve stimulation (at baseline and after exercise) and single fiber EMG (jitter, blocking, abnormal pairs) ($r>0.3$ and $p<0.05$ for all parameters).

Conclusion: The QMGS correlates well with other clinical and electrophysiological parameters and is significantly higher in patients with thymoma. QMGS does not appear to correlate well with Ach-RAb levels or antibody status.

DETECTION OF MUSCLE SPECIFIC TYROSINE KINASE ANTIBODIES IN MYASTHENIA GRAVIS: COMPARISON OF AN ELISA AND RADIOIMMUNOPRECIPITATION ASSAY

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Introduction: Antibodies to muscle specific tyrosine kinase (MuSK) are detected in a variable proportion of myasthenia gravis (MG) patients without acetylcholine receptor antibodies¹. The commercial availability of MuSK antibody (MuSK-Ab) testing has revealed MuSK-Ab positive MG as a distinct MG subtype and is important for the diagnosis and management of MG patients.

Methods: 174 MG serum samples from the UBC MG Clinic, which had been measured for MuSK-Abs by radioimmunoprecipitation assay (RIPA) at the Institute of Molecular Medicine, in Oxford, U.K. (Prof. A. Vincent), were assayed by ELISA at UBC Neuroimmunology lab. The ELISA and RIPA results were compared and the sensitivity and specificity of the ELISA was calculated.

Results: Of the 14 MuSK-Ab positive MG samples by RIPA, 12 were determined MuSK-Ab positive by ELISA. Of the 160 MuSK-Ab negative MG samples by RIPA, 151 were MuSK-Ab negative by ELISA. All 37 healthy controls were MuSK-Ab negative by ELISA. ELISA sensitivity and specificity compared to the RIPA were 86% and 94%.

Discussion: The ELISA offers a simple and rapid screening test for measuring MuSK-Abs in human serum. The technique provides the advantage of increased safety by eliminating the use of radioactive materials. A strong agreement can be seen between the ELISA and RIPA MuSK-Ab results. The finding of “false positive” might in fact indicate greater

sensitivity than the RIPA. The finding of false negative indicates a need to improve the technique.

References:¹Hoch W, McConville J, Helms S, Newson-Davis J, Melms A, Vincent A. Auto-antibodies to the receptor tyrosine kinase MuSK in patients with myasthenia gravis without acetylcholine receptor antibodies. *Nat Med* 2001; 7: 365-368.

OUTCOMES IN A LARGE COHORT OF MUSK ANTIBODY POSITIVE PATIENTS

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Introduction: Long term outcomes and optimal immunosuppressive management of patients with MuSK-antibody positive MG (MuSK-MG) remain uncertain. This study examines outcomes and pharmacotherapy in a large cohort of well characterized patients.

Methods: We analyzed demographics, maximum MGFA class, immunosuppressive medications, side effects, and MGFA post interventional status (PIS) in 110 MuSK-MG patients seen in the MG Clinics at Duke University (40 patients) and the Catholic University, Rome (70 patients).

Results: 85% were female and average age at symptom onset was 36.6 years. 85% had maximum MGFA Class of III or greater, and 28% had at least one crisis. During acute exacerbations, 61% improved after intravenous immunoglobulin, while 93% improved after plasma exchange. Cholinesterase inhibitors were tried in 98%: 43% showed no significant improvement and 31% had adverse effects. Patients received the following medications at some point: corticosteroids - 101; azathioprine - 53; mycophenolate - 27; cyclosporine - 18; and rituximab - 6. At last follow-up 92% had a PIS of “Improved” or better (54% Minimal Manifestations or better). Immunosuppressive treatment was ultimately withdrawn in 12 patients (11%); immunosuppressive monotherapy was achieved in 32%: prednisone - 26; mycophenolate - 5, azathioprine - 3; cyclosporine - 1). All 6 patients who received ritux-

imab improved with reduced immunosuppressant dosages; 4/6 achieved Minimal Manifestations.

Discussion: MuSK-MG patients have more severe disease than non-MuSK MG but most improve significantly with aggressive immunosuppression. Combined immunosuppressive therapy is often required. Response to plasma exchange is better than to intravenous immunoglobulin. Rituximab is a promising therapy for refractory MMG patients.

THE MG-QOL15 FOR FOLLOWING THE HEALTH-RELATED QUALITY OF LIFE OF PATIENTS WITH MYASTHENIA GRAVIS

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Introduction: The MG-QOL15 is helpful in informing the clinician about the patient’s perception of the extent of and dissatisfaction with MG-related dysfunction. The aims of this study were to determine the usefulness of the MG-QOL15 for following individuals with MG and to guide clinical researchers planning to use the MG-QOL15.

Methods: We assessed sensitivity and specificity of the MG-QOL15 for detecting clinical change in a multi-center, MG Composite and MG-QOL15 scale validation study. We also evaluated test-retest reliability of the MG-QOL15 at one center.

Results: Sensitivities and specificities of various cut-points of change in MG-QOL15 scores are presented. The psychometric properties of the MG-QOL15 were influenced by disease severity at visit 1. For example, the sensitivity and specificity of a 5-point improvement in MG-QOL15 was 69.4% and 75.3% when all patients were included and 82.9% and 60.0% when only patients with initial MG-QOL15 ≥ 10 were included. Also presented are means and standard deviations of MG-QOL15 scores for all patients and for subgroups of patients. The test-retest reliability coefficient was 98.6%.

Discussion: The MG-QOL15 has acceptable longitudinal construct validity. We consider this instrument to be most useful for informing the clinician about the patient’s perception of MG-related dysfunction and tolerability with that dysfunction. We recommend that more objective measures, such as the MG Composite, be used to follow disease severity over time or in clinical trials.

This study was supported in part by the Myasthenia Gravis Foundation of America.

THE OCULOBULBAR FACIAL RESPIRATORY IS A USEFUL TOOL TO ASSESS BULBAR FUNCTION IN MYASTHENIA GRAVIS

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Introduction: The oculobulbar facial respiratory (OBFR) score is a tool that objectively assesses bulbar function in myasthenia gravis (MG) patients. The OBFR score consists of five clinical items including facial muscle assessment, palatal contractility, tongue muscle appearance, swallow time and forced vital capacity. We studied the correlation between the OBFR and other MG related scores including the MG composite (MGC), MG-QOL15 and MG-ADL scores.

Methods: We enrolled 42 MG patients in two centers (Glasgow and University of Virginia) and assessed them on two occasions, circa 6 months apart, using the MGC, MG-ADL, MG-QOL15 and OBFR scores. In one center, facial muscle assessment was carried out by two assessors in 13 patients to assess inter-rater variability.

Results: The OBFR score correlated with the MGC ($P < 0.0001$ for both assessments) and with the sum of bulbar items of the MGC ($P < 0.0001$ for both). The OBFR score correlated with the MG-ADL ($P < 0.0001$ and $P = 0.0013$ respectively) and MG-QOL-15 scores ($P = 0.0005$ and $P = 0.05$ respectively), and with the sum of the bulbar items for both scores ($P < 0.0001$ for all except MG-QOL15 on second assessment, where P was 0.02). The “modified” OBFR score, derived from the original OBFR score, assesses three instead of five facial muscles, omitting corrugator supercilii and buccinator, because of higher inter-rater variability for these two muscles. The modified OBFR score also correlated significantly with all three MG-related scores.

Discussion: This study confirms construct validity of the OBFR and modified OBFR scores. They are useful in the assessment of MG patients with predominant bulbar weakness.

RITUXIMAB IN REFRACTORY GENERALIZED MYASTHENIA GRAVIS

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Introduction: Myasthenia gravis (MG) is an immune-mediated disorder associated with autoanti-

bodies against neuromuscular junction molecules. Rituximab, a chimeric monoclonal antibody to the CD20 cell marker, is indicated to treat B-cell non-Hodgkin lymphoma and rheumatoid arthritis. Experience with rituximab in MG is limited.

Method: We performed a retrospective chart review of patients treated with rituximab for refractory generalized MG at the University of Kansas Medical Center and the University of Texas Health Science Center-San Antonio. We assessed safety based on patient-reported adverse effects. We abstracted efficacy using changes in the MG functional assessment (MGFA) grade and in concomitant medications.

Results: We identified 3 women and 2 men with a mean age of 55 years and mean disease duration of 12 years. Two patients had acetylcholine receptor antibodies, and another had MUSK antibodies. All 5 cases were refractory to prednisone and various adjuvant immunosuppressive agents. Three patients were on scheduled plasmapheresis and one on IVIG. At induction, we mostly administered 2 doses of rituximab 1 gm each, and in 3 cases a 3rd dose 6-8 months later. None of our patients experienced any serious adverse effects. We stopped plasmapheresis in 2 cases, and increased the interval between IVIG treatments in 1 case. There was an improvement in the MGFA grade in 4 cases and we were able to reduce the prednisone dose in the fifth case.

Discussion/Conclusion: Our preliminary data indicates that rituximab is safe in MG patients. A larger randomized controlled study is needed to confirm the safety and efficacy of rituximab in MG patients.

THE COST OF MISDIAGNOSIS IN MYASTHENIA GRAVIS

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Introduction: The diagnosis of myasthenia gravis (MG) in seronegative patients remains challenging. We are commonly referred patients diagnosed as unsubstantiated “seronegative MG” who have re-

ceived very aggressive immunotherapy, often with chronic intravenous immunoglobulin (IVIg) infusions. After thorough evaluation, an alternative diagnosis is frequently found. Here we present such a case, introduce IVIg overuse, and emphasize the importance of proper diagnosis.

Methods: A representative patient seen in the Duke MG clinic will be presented. Pharmacotherapy costs, including IVIg, were obtained from a cohort of 1,322 patients diagnosed with MG who were identified from the Accordant Health Services comprehensive nationwide medical and pharmacy claims database, which covers approximately 27 million lives.

Results: The mean age of the MG cohort was 59.8 and there was a female predominance. Average annual total/pharmacotherapy costs per patient based on claims were: \$11,656/\$2,076 (age 0-19), \$18,310/\$21,670 (20-39), \$13,498/\$11,044 (40-64), and \$13,281/\$6,620 (65+). The most frequently prescribed drugs used by the cohort were cholinesterase inhibitors (77%), corticosteroids (48%), nonsteroidal immunosuppressants (32%), and IVIg (11%). Total MG-related pharmacotherapy costs were \$10 million. IVIg accounted for 67.3% of MG specific pharmacotherapy costs (\$6.7 million), while nonsteroidal immunosuppressives and cholinesterase inhibitors accounted for 22.6% and 9.8%, respectively. Corticosteroids represented a mere 0.4%.

Discussion: Patients with symptoms suggestive of MG require a thoughtful, comprehensive evaluation. When the diagnosis is in doubt or patients fail to respond to aggressive therapy, referral to a neuromuscular specialty center should be considered. The misdiagnosis of patients with MG-like symptoms carries considerable healthcare and psychological costs.

INCREASING INCIDENCE OF ANTI-ACHR SEROPOSITIVE MYASTHENIA GRAVIS IN BRITISH COLUMBIA, CANADA

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Introduction: The primary mechanism for clinical symptoms of myasthenia gravis (MG) is an antibody-mediated attack against nicotinic acetylcholine receptors (AChR) at the neuromuscular junction. Anti-AChR antibodies are detected in 80-90% of generalized MG patient sera and 50-60% of patients with ocular MG.¹ Their high specificity makes anti-AChR seropositivity an excellent surrogate marker for MG. This study aimed to evaluate the epidemiology of anti-AChR antibody-seropositivity in British Columbia (BC), Canada.

Methods: The Neuro-Immunology Laboratory at the University of British Columbia is the sole laboratory in BC that receives sera for diagnostic purposes to measure anti-AChR antibodies. We use a radio-immunoprecipitation technique. A population-based study of anti-AChR seropositivity in BC for the period of January 1984 to December 2008 was carried out by retrospectively identifying all first-time cases of anti-AChR antibody-seropositivity.

Results: Between January 1984 and December 2008, 1243 individuals were identified to be anti-AChR seropositive (648 women, 587 men, 8 unknown). In women the age at the first positive serum sample had a bimodal distribution with peaks at 45-55 and 70-85 years, while in men there was a single peak at 70-80 years. The average annual incidence of new anti-AChR seropositivity for the period of 1984-2008 was 13.2/million/year. The incidence had dramatically increased in the elderly (≥ 65 years), from 21.4/million (1984-1988 average) to 52.9/million (2004-2008 average).

Discussion: Our results indicate an increased incidence of late-onset anti-AChR seropositive MG in BC which is not due to increasing longevity. This study is important for the allocation of resources and future health care planning.

Drachman DB. Myasthenia gravis. N Engl J Med 1994;330:1797-1810.

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