in Gardner–Medwin–Walton and Vignos scales. A significant correlation ($p < 0.05$) was detected between the mean MFF of all muscles and Gardner–Meldwin–Walton ($r = 0.84$) and Vignos ($r = 0.60$) scale. This correlation was also found separately in the muscles of thigh (anterior $r = 0.83$, posterior $r = 0.75$) and calves (anterior $r = 0.65$, posterior $r = 0.82$).
**Conclusion:** Muscle fat content measured with 3-point Dixon MRI provides a high correlation with clinical severity of ambulant LGMD2A patients and could be a useful biomarker.

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**Abstract - WCN 2013**
**No: 1233**
**Topic: 7 - Neuromuscular disorders**
**Time from onset to treatment and prognosis in patients with CIDP: A 3-year follow-up of 29 cases**

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**Background:** Time from onset to treatment may be important for CIDP prognosis, although this variable has not been systematically assessed.
**Objective:** To investigate the association between time from onset to treatment and prognosis in CIDP patients.
**Patients and methods:** We retrospectively enrolled 29 consecutive CIDP patients [15 males, median age, 58 years (range, 15–82 years)] who were followed-up for 3 years after initial treatment. Twenty-seven patients fulfilled the EFNS/PNS definite criteria. Two patients categorized to probable CIDP were diagnosed using sural nerve biopsy. The primary outcome was good prognosis, which was defined as stable disease activity ≥2 years and Hughes grade of 0–1 at 3 years. Clinical characteristics, time from onset to treatment, and Hughes grade at initial treatment were evaluated as predictors of good prognosis.
**Results:** Patients were first treated with immunoglobulin infusion (57.1%), corticosteroids (32.2%), and plasmapheresis/immunoabsorption (10.7%). Twenty-five patients (86.2%) responded to the treatments. Patients showed monophasic (17.2%), relapsing (51.7%), and chronic (31.1%) clinical courses. Twelve patients (41.4%) showed good prognosis. Six of 8 (75.0%) patients treated <8 weeks and 6 of 14 (42.9%) patients treated from 8 weeks to 1 year after onset showed good prognosis, whereas none of 7 patients treated ≥1 year after onset demonstrated good prognosis (Fisher's exact test, $P = 0.01$). Hughes grade at initial treatment was not significantly associated with good prognosis (Fisher's exact test, $P = 0.67$).
**Conclusion:** Time from onset to treatment was suggested to be important for CIDP prognosis, regardless of the disease severity at initial treatment.

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**Abstract - WCN 2013**
**No: 1379**
**Topic: 7 - Neuromuscular disorders**
**Rare case of leprosy, where initial presentation is carpal tunnel syndrome**

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33-year-old woman from village area of country, had history of right arm pain with tingling like sensation, especially aggravated in night time. Clinically suspected carpal tunnel syndrome confirmed with nerve conduction study. Apart patient did not have any other symptoms related to skin or other nerve involvement. 9 months post decompression surgery patient had developed hypo pigmented rash on forearm and leg, as well as trophic ulcer in hands. With no apparent exposure to leprosy, clinical and histological evidence of tuberculoid leprosy was found. A particular characteristic of this case was leprous neuritis involving the median nerve which was diagnosed clinically as carpal tunnel syndrome, and scheduled for surgical treatment. Under treatment with dapsone and rifampicin, however, the condition cleared up completely.

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**Abstract - WCN 2013**
**No: 1370**
**Topic: 7 - Neuromuscular disorders**
**Cholinergic neuromuscular hypersensitivity in musk antibody positive myasthenia gravis**

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**Background:** The patients with MuSK antibody (MuSK-Ab) positive myasthenia gravis (MG) show distinct responses to acetylcholinesterase inhibitor (AChEI), such as less effective therapeutic response to AChEI and more frequent nicotinic side effects and negative result of diagnostic AChEI test. Some MuSK-Ab positive MG patients experience overt worsening after AChEI treatment. In addition, the compound muscle action potential with extradischarges (CMAP-EDs), elecrophysiologic feature of cholinergic neuromuscular hyperactivity, may develop in the MuSK-Ab positive MG patients with usual therapeutic dose of AChEI.
**Objective:** We investigated the clinical and electrophysiologic features of cholinergic neuromuscular hypersensitivity in MuSK-Ab positive MG patients.
**Patients and methods:** We retrospectively reviewed the medical records and electrodiagnostic findings of seventeen MG patients (MuSK-Ab positive: 10, MuSK-Ab negative: 7) who underwent electrodiagnostic test before and after neostigmine test.
**Results:** The MuSK-Ab positive patients had higher frequency of intolerance to oral pyridostigmine bromide (50 vs 0%, $p = 0.044$) and lower maximal dose of oral pyridostigmine (90 vs 480 mg/day, $p = 0.023$) than the MuSK-Ab negative. The frequency of positive results of neostigmine test was significantly lower in the MuSK-Ab positive patient the in the MuSK-Ab negative (100 vs 40%, $p = 0.035$). The nicotinic side effects of neostigmine were more frequent in the MuSK-Ab positive patients (80 vs 14.3%, $p = 0.015$). The CMAP-EDs were more frequently developed after neostigmine injection in the MuSK-Ab positive patients than the MuSK-Ab negative (90 vs 14.3%, $p = 0.004$).
**Conclusion:** Cholinergic neuromuscular hypersensitivity is a distinct characteristic of MuSK-Ab positive MG.

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**Abstract - WCN 2013**
**No: 1324**
**Topic: 7 - Neuromuscular disorders**
**Effect of Alpha-Lipoic Acid on the postural stability of patients with diabetic peripheral neuropathy**

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**Background:** Diabetic peripheral neuropathy (DPN) is a common complication of diabetes mellitus, that affects the postural stability.

There are researches about the effect of Alpha-Lipoic Acid (ALA) on the reduction of pain and improvement of neuropathic deficits.
**Objective:** This study evaluated the effect of treatment with ALA on the postural stability in patients with type 2 DPN.
**Patients and methods:** Sixty patients and 20 healthy age-matched subjects took part in this investigation. All patients had a good glycemic control. The two schemes of treatment were applied: the first — with 600 mg ALA (5 day infusion and 60 day oral dose), and the second — ALA, benfotiamin, pyridoxine and cyancobalamine together. The postural stability was evaluated using static posturography under two visual conditions (eyes open and eyes closed) on stable and soft surfaces. The investigations were made on the first, 5th and 60th day after the drug therapy.
**Results:** Before the treatment with ALA the all posturographic parameters, for both patient groups were significantly higher than in healthy subjects. After treatment the sway velocity and sway path decreased. The most pronounced decrease was observed in sway path during stance on foam support with closed eyes. The changes of the posturographic results after the combined therapy were better than with ALA only.
**Conclusion:** Treatment with combined therapy (ALA, benfotiamin, pyridoxine and cyancobalamine together) showed stabilizing effect on the quiet upright stance, that leads to improvement of the quality of life of patients with type 2 DPN.

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**Abstract - WCN 2013**
**No: 1355**
**Topic: 7 - Neuromuscular disorders**
**Classification of sporadic lower motor neuron disease in Jeju Island**

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**Objectives:** The sporadic forms of lower motor neuron disease (LMND) constitute a heterogeneous group of diseases with largely unknown pathogenesis. Clinical, genetic, electrophysiological, and immunological findings can help to distinguish patients with LMND who never develop amyotrophic lateral sclerosis (ALS) from patients with typical ALS.
**Methods:** We studied the clinical and electrophysiological features of 23 patients with sporadic LMND in a cross-sectional study. Disease duration was more than 4 years to exclude the majority of patients with ALS. Based on the pattern of weakness, we classified patients into three groups: with generalized weakness (group 1); with non-generalized asymmetrical weakness of the arms (group 2); with non-generalized asymmetrical weakness of the legs (group 3).
**Results:** We identified two patients in group 1, eighteen patients in group 2, and three patients in group 3. Distinctive features of group 1 were an older age at onset, more severe weakness and muscle atrophy and more widespread abnormalities on needle EMG. In groups 2 and 3, age at onset was younger than patients in group 1. However, needle EMG findings also suggested a more widespread disease process. Retrospectively, the prognosis of sporadic LMND seems to be relatively good after a median disease duration of 8 years.
**Conclusion:** The clinical phenotypes of the different subgroups described in this study may help to differentiate the several LMND forms from each other. However, prospective studies are needed to investigate whether specific clinical or pathogenic variables may help to identify patients with a more benign form of LMND.

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**Abstract - WCN 2013**
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**Topic: 7 - Neuromuscular disorders**
**A rare case of an African–American male with AOA2**

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**Background:** Ataxia with oculomotor apraxia type 2 (AOA2) is an autosomal recessive cerebellar ataxia caused by mutation of the senataxin gene (SETX) on chromosome 9q34 and characterized by cerebellar atrophy, axonal sensorimotor neuropathy, oculomotor apraxia, and elevated serum alpha-fetoprotein (AFP).
**Objective:** We describe the case of a 32 year African–American male with classic clinical features of AOA2 with positive SETX gene mutation.
**Methods:** At birth the patient was delivered without complications and had normal development. At 7–8 years of age he developed difficulties with coordination and became wheelchair bound by age of 12. Since then he has been suffering from progressive cognitive impairment, polyneuropathy and hypercholesterolemia. However, family history is unclear. Neurological examination is significant for moderate cognitive impairment, dysarthria, dysconjugate gaze, jerk nystagmus and alternating strabismus. Motor exam revealed normal tone, reduced bulk in all the extremities and reduced strengths in bilateral lower extremities. Dysmetria on limb movements and marked ataxia were noted. Reflexes were absent in all the extremities.
**Results:** NCS revealed progressive sensorimotor axonal polyneuropathy and EMG showed evidence of acute & chronic denervation. MRI revealed marked cerebellar atrophy and degenerative changes in the cervical spine. Genetic testing was performed for AOA2 and Fragile X syndrome was positive for the SETX gene, confirming the diagnosis of AOA2.
**Conclusion:** AOA2 is a rare condition frequently seen in Europe, North Africa and West Indies. Most of the cases encountered in the US are of French descent. Our case is unusual in that the patient was of African–American origin.

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**Abstract - WCN 2013**
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**Topic: 7 - Neuromuscular disorders**
**Nerve ultrasound score in distinguishing chronic inflammatory demyelinating polyneuropathy from Guillain–Barré syndrome**

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**Background:** GBS is defined as an acute-monophasic polyradiculoneuropathy, while CIDP may also present (16% of the cases) with a subacute-monophasic course. Their differential diagnosis is of great importance, as their therapies differ significantly.
**Objective:** The aim of this study was the development and evaluation of a new ultrasound score to distinguish these two nosologies.
**Materials/methods:**

Phase-1 (development):
20 GBS (mean-age 58.3y, SD ± 13.8; 10 women), 20 CIDP-patients (mean-age 56.7y, SD ± 13.5; 6 women), 75 healthy-subjects (mean-age 53.5y, SD ± 14.8; 30 women) underwent nerve-ultrasound, a mean