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Late Breaking Poster Abstracts

1490
WFN15-1615
Late Breaking Posters 1
Polysomnographic findings in a sample of older adults with attention-deficit hyperactivity disorder

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Introduction: Previous studies in children and young to middle-aged adults with attention-deficit/hyperactivity disorder (ADHD) have shown impaired sleep quality with increased nocturnal motor activity. However, to our knowledge, there are no reports describing the characteristics of sleep and its disorders in older ADHD patients. The objective of this study was to describe polysomnographic findings in a sample of older adults with ADHD.

Materials and methods: Patients who were diagnosed as ADHD according to DSM IV criteria during 2014, and have one night polysomnographic recording , were included. We considered only those aged 50 and older, without psychiatric comorbidities or medication specific to ADHD.

Conventional polysomnographic parameters were calculated for each case. All subjects were assessed with questionnaires for subjective parameters to screen sleep disorders.

Results: We studied 21 patients, mean age was 61.57 years and 14 were females (66%). Periodic leg movements in sleep occurred in 7 (33%), REM sleep without atonia was found in 7(33%), and 7 patients had an apnea-hypopnea index greater than 5 (33%). The whole recordings showed a reduced sleep efficiency (73.7%), prolonged sleep onset latency (39.9 minutes) and REM sleep latency(132.7 minutes) , and a decreased percentage of stage 3 as well (3 %).

Conclusions: Our findings suggest that in older patients with ADHD, sleep disorders are a frequent and varied comorbidity that requires special attention. Future studies should determine the impact of these disorders on cognitive and behavioral performance of these patients.

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1492
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Late Breaking Posters 1
Cardiovascular and metabolic risk factors, for cognitive impairment in adults from Bogotá

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Introduction: The relationship between some metabolic and cardiovascular risk factors (CVRF) and cognitive impairment (CI), defined as mild cognitive impairment (MCI) and dementia are controversial.

Objective: Describe the cardiovascular and metabolic risk factors, that are associated with cognitive impairment in adults from Bogotá.

Material and methods: A cross-sectional study, where the state of cognitive functions (normal, MCI or dementia) was evaluated in two phases, in adults older than 50 years, autonomous, non-institutionalized, using neuropsychological tests and neuropsychiatric protocol. Its cardiovascular and metabolic risk by self-reported history and standardized anthropometric measurements were documented.

Results: Of 1045 seniors surveyed, 76%were women, 55% had hypertension (HT), 40% dyslipidemia, 37% were smokers, 36%were overweight, 28% presented hypothyroidism, 25% were obese, 17% drank alcohol and 16 % were diabetic. MCI was associated with incomplete high school education (OR: 1.74 95% CI. 1.23-2.45) and aged between 70 and 79 years (OR:1.93 95% CI. 1.47-2.53). Dementia was associated with lower education, incomplete primary (OR: 8.98 95% CI. 5.56-14.54), complete primary (OR: 6.23 95% CI.3.70-10.47), incomplete high school (OR: 2.50 95% CI.: 1.35-4.59), age over 80years (OR: 3.49 95% CI. 2.23-5.44); and hypertension (OR: 1.58 95% CI.1.12- 2.21).

Conclusion: Low education, older age and hypertension are in order, the most important factors for the development of dementia in our population risk. Adults between 70 and 79 with incomplete high school have higher risk of MCI.

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1493
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Late Breaking Posters 1
The effect of beta glucan of saccharomyces cerevisae in the parkinson's wistar strain rats (rattus norvegicus) model induced with rotenone

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Background: Parkinson's disease is a degenerative nerve disease due to damage in dopamine-producing neurons in the brain, particularly in the *substantia nigra pars compacta.*

Objective: Identify reduction of *alpha synuclein* expression on *substantia nigra* in Parkinson's rat model after given *Saccharomyces cerevisae.*

Patients and methods / Material and methods: This research applied true experimental design by in vivo with draft randomized post test only controlled group. The sample was divided into five groups, each of them consisted of 5 rats, namely, negative control group, positive control group, Treatment Group 1, 2 and 3 (Rotenone + *Saccharomyces cerevisae* 18 mg/kgBB), 36 mg/kgBW, 72 mg/kgBW which is given for 30 days). Variable measured is *alpha synuclein* expression.

Results: Significant difference in the number of expressions of *alpha synuclein* between positive control group and negative control group (p = 0000) and treatment group (p = 0.000; 0.000; 0.000); the addition of *Saccharomyces cerevisae* in Treatment 1, 2 and 3 (18mg/kgBW, 36mg/kgBW, 72mg/kgBW) caused a significant difference compared with positive control group (p = 0.000; 0.000; 0.000); the addition of *Saccharomyces cerevisae* in Treatment 1 (18 mg/kgBW) caused significant difference in the amount of *alpha synuclein* expression compared to the Treatment 2 (36mg/kgBW) (p = 0.981), but it showed a significant difference in Treatment 3 (72mg/kgBW) (p = 0000).

Conclusion: Addition of *Saccharomyces cerevisae* decreases *alpha synuclein* expression in the *substantia nigra* of Parkinson's rat models significantly with maximum results at the dose of 72 mg/kgBW.

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1495
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Late Breaking Posters 1
Primary supratentorial malignant melanoma: a case report with long survival

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Background: Melanomas are malignant neoplasms of melanocytes developing predominantly in the skin, but occasionally arising from eyes, mucous membranes, and the CNS. Primary CNS melanoma is a rare and aggressive malignant tumour.

Objectives: We report a rare case of a primary melanoma of CNS with no extracranial localization.

Material & methods: A 76 – year old man was admitted in January 2013 because of worsening symptoms of headache and vomiting. CT scan revealed an heterogeneous mass over the left temporo – occipital area, and MRI scan demonstrated an hyperintense on T1 images, and hypointense on T2 images lesion, with contrast enhancement. He underwent resection of the tumor, which composed of malignant epithelioid cells and was reported as malignant melanoma.

Results: A thorough dermatologic examination didn't show any abnormal pigmentation and fundoscopy revealed normal retinae. A PET – CT scan of the whole body was also normal. Based upon histological characteristics and the fact that there was no evidence of malignant melanoma elsewhere, the final diagnosis was primary melanoma of CNS. The patient subsequently received adjuvant radiotherapy, and more than two years postoperatively, is still alive with no evidence of disease in his follow – up.

Conclusion: One difficulty when discussing melanoma of the CNS is to decide which cases represent primary growths and which are metastases. Metastatic melanoma is more frequent and usually leads to an early fatal outcome. Our case was a primary solitary brain melanoma due to the negative metastatic work – up and a more than two – year disease – free state postoperatively.

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1496
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Late Breaking Posters 1
A systematic review and meta-analysis of clinical trials of bone marrow mononuclear cell therapy for patients with ischemic stroke

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Background: Bone marrow mononuclear cell therapy (BM-MNCs) has emerged as a potential therapy for the treatment of stroke.

Objective We performed a meta-analysis of available studies using BM-MNCs therapy in patients with ischemic stroke.

Methods: Literature was searched using MEDLINE, EMBASE, Trip Database and Cochrane library, clinicaltrial.gov to identify studies on BM-MNCs therapy in ischemic stroke till January 2015. Data was extracted independently by two reviewers. STATA version 13 was used for meta-analysis.

Selection criteria: We included non-randomized open label, single arm, and comparative study or randomized controlled trial if BM-MNCs were used to treat patients with stroke in any phase (acute, sub-acute or chronic).

Results: One randomized trial, one non-randomized comparative trial, and four single arm open label trials (total six studies) involving 122 subjects were included in the proportional meta-analysis. The pooled incidence for achieving the favourable clinical outcome (modified Rankin Scale score <2) was 29% (95% CI 0.19 to 0.43). Total 69 control subjects (taken from two comparative trials), those who did not receive stem cells had pooled incidence of favourable outcome (mRS <2) 10% (95% CI -0.09 to 0.29). After pooling the data of Barthel Index at six months ,reported in three comparative studies, no significant difference was found 0.12 (95% CI -0.19 to 0.42, P= 0.45). The pooled differences in the safety outcomes between MNCs group and controls were not significant.

Conclusion: Our meta-analysis suggests that BM-MNCs therapy is safe and feasible; however, its efficacy is debatable in the case of stroke patients.

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1497
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Late Breaking Posters 1
Narcolepsy type 1 clinical symptoms misdiagnosed for epileptic seizures

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Background: Clinical features of narcolepsy type 1 (narcolepsy with cataplexy) include unintentional sleep attacks and episodic muscle atonia triggered by emotion, which can be often accompanied by hypnagogic hallucinations, sleep paralysis and automatic behavior.

Objective: The aim of the work is to point out diagnostic difficulties between narcolepsy type 1 paroxysmal events and epileptic seizures, which can appear in clinical practice.

Patients and methods: Retrospective analysis of disease course in 31 adult individuals from the first symptoms until definitive diagnosis of narcolepsy type 1 according to ICSD-3 criteria.

Results: Narcolepsy symptoms were misdiagnosed and incorrectly treated as epileptic seizures in 7 patients (22,58%). Generalised cataplexy was misdiagnosed and treated as atonic epileptic seizures in 1 patient, localised cataplexy was managed as myoclonic epilepsy

in 3 patients. Automatic behavior and hypnagogic hallucinations were assessed as focal epileptic seizures. Incorrect assessment of narcoleptic symptoms as epileptic seizures delayed determination of adequate diagnosis by 3 years on average.

Conclusion: The most frequent mislead narcoleptic symptoms are catapletic seizures which might be considered as epileptic seizures. Targeted anamnesis for excessive daytime sleeepiness should be the basic question in differential diagnosis of paroxysmal events. Inadequate identification of narcolepsy symptoms as epileptic seizures leads to unnecessary diagnostic practice and wrong treatment, which delays statement of correct diagnosis even by several years.

An Institutional Review Board has waived the requirement for its formal approval of the study.

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1498
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Late Breaking Posters 1
Neurodevelopmental outcome of preterm infants born from mothers with and without chorioamnionitis

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Background: Neurodevelopmental outcomes in preterm infants have been widely studied. However, it has not been determined whether the presence of certain prenatal events related to premature birth, may affect the level of severity of the neurodevelopmental impairments in this population. According to some authors, preterm infants exposed to chorioamnionitis have increased risk to develop neurodevelopmental disorders due to pathophysiologic processes involved. However, behavioral studies about this topic have provided divergent results.

Objective: To determine whether neurodevelopmental outcome differs between preterm infants born to mothers with chorioamnionitis during pregnancy and preterm infants born to mothers without this pathology.

Material and methods*:* 16 preterm infants with clinical history of chorioamnionitis (CA group) and 16 preterm infants born to mothers without this pathology, but with other prenatal factors related to preterm delivery (NO-CA group) participated in this study. Both groups were matched for corrected age at the moment of neurodevelopmental evaluation and relation birth weight/gestational age (intrauterine growth). Neurodevelopmental outcome was assessed by the Bayley Scales of Infants and Toddler Development, Third edition (Bayley-III) that included five areas of neurodevelopment: cognitive, language, motor, socio-emotional and adaptive behavior.

Results*:* Both groups showed mild delay in cognitive, motor and language areas. Comparing both groups, the CA group presented lower scores in cognitive and fine motor skills than NO-CA group.

Conclusion: Although there were differences on neurodevelopmental outcomes between groups, these were not statistically significant. This suggests that chorioamnionitis, such as prenatal event related to prematurity, have a low impact on neurodevelopment of premature infants.

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1500
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Late Breaking Posters 1
Quetiapine for behavioral disorders management in Parkinson disease

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Patients with Parkinson's disease (PD) frequently present mental disorders such as hallucinations, delusions, confusion or agitation. Even after adjustment of antiparkinsonian therapy, it may be necessary to use neuroleptic drugs (better atypical than typical neuroleptics) to improve patient's mental symptoms without worsening his motor situation. Clozapine and quetiapine are the drugs that offer a surer profile for its use in PD patients.

Objective: The purpose of this article is presenting the obtained results after analyzing 36 PD patients who needed neuroleptic treatment because of behavioral disorders or psychiatric symptoms, and discussing underlying mechanisms of action.

Methods: It has been collected data of 36 patients diagnosed with idiopathic PD who need to be treated also with neuroleptic drugs. The data were obtained from medical histories and were focused on antiparkinsonian treatment received, the response to neuroleptic drug, its impact on motor function, and adverse events development.

Results: 80.6% of patients (n= 29) improved under quetiapine treatment, both isolated behavior disorders (3 patients) and 78.8% (n=26) of 33 patients who had developed hallucinations. The more frequent adverse event was somnolence, and none of them presented motor function worsening.

Conclusions: Data obtained after performing this work are concurrent to those obtained in other similar published investigations. Quetiapine shows an effective and sure profile to be used for the management of behavior disturbances or psychiatric symptoms that may be present in some PD patients, both due to dopaminergic treatment and duet o PD dementia development.

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1501
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Late Breaking Posters 1
Description of verbal and non-verbal ability in 280 patients with aphasia caused by stroke

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Background: Aphasia is a language disorder that, results from damage to the left brain hemisphere.The most common cause of aphasia is stroke. It is estimated that from 21 % to 38%of stroke patients are aphasic in the acute stage.

Objective: To describe verbal and nonverbal communication behavior in patients with aphasia caused by stroke.

Material and methods: Consecutive 280 aphasic patients recruited in a university center in Santiago, Chile and with clinical and imaging evidence of brain damage were included. All patients were evaluated with Porch Index of Communicative Ability and compared with 30 healthy controls.

Results: Aphasic participants had a lower performance in the Index Communicative Ability Porch than the control group. The level of communicative ability was assessed according to the type of aphasia. Those with global aphasia, non-fluent mixed aphasia, transcortical

sensory and transcortical motor aphasia demonstrated less efficient abilities than those patients with anomic, conduction or Broca aphasics. Demographic and cognitive variables, including age, education, severity, anosognosia and intellectual impairment, showed an effect on the communicative ability of patients.

Conclusion: Based on their level of communicative ability, aphasias can be coherently grouped into three categories, a moderate group, severe and very severe. The first group includes the anomic, conduction and Broca. The second corresponds to Wernicke, Transcortical Motor, Transcortical Sensory and non-fluent Mixed, whereas the third group corresponds to the global aphasia.

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1502
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Late Breaking Posters 1
Cognitive behavioral therapy for insomnia treatment: good results with trained nurses and a limited number of sessions

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Background: Chronic insomnia affects up to 20% of chilean adult population. Cognitive Behavioral Therapy (CBT-I) has been established as an effective first line treatment in these patients, especially in those who suffer primary insomnia.

Objective: To evaluate results of patients treated in a CBT-I program conducted by a trained nurse.

Patients and methods: All patients that entered the CBT-I program were included. Outcomes were: a) minimum number of sessions attended b) accomplishment of personal goals set at the beginning of therapy with subjective patient satisfaction.

Results: A total of 32 patients have been treated so far at the CBT-I program. Nine patients abandoned the program before the fifth session. Of those who completed at least five sessions (n=23), 82% (n=19) achieved excellent or good results, 9% (n=2) showed no significant improvement and 9% (n=2) are still under treatment. The mean number of sessions attended by patients that obtained good or excellent results were 6,7 (out of 8 programmed).

Conclusions: An insomnia CBT-I program conducted by a trained nurse is successful for the treatment of chronic insomnia in a significant proportion of patients. The program can be restructured to 6 or 7 sessions, with optional extra appointments if necessary. Both results open the possibility of a wider access to CBT-I, especially in a context of limited resources.

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1503
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Late Breaking Posters 1
Sleep quality evaluation in medical students

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There are certain factors that may influence quality of life and one of the most important is sleep.

Sleep deprivation has a significant impact in mood, alertness, cognitive functions, and motor activity.

The objective of this investigation is to estimate the sleep quality in medical students, the degree of chronic daytime sleepiness and if this is related or not with chronobiological factors.

43 out of 61 students answered the Epworth Sleepiness Scale questionnaire. 60.5% scored below 9 points, indicating a low degree of daytime sleepiness. 39.5% scored 10 or more points indicating a high degree of daytime sleepiness.

All students answered the Pittsburgh Sleep Quality Index questionnaire and 78.7% of them had bad quality sleep.

The morningness–eveningness questionnaire was completed by 49 of the 61 students. 2 indicated an extreme degree of eveningness, 15 had a moderate eveningness, 27 had an intermediate degree, and 5 had a moderate degree of morningness.

It can be concluded that 39,53% of the surveyed students have excessive daytime sleepiness and 78,68% have bad sleep quality.

Medical students whom are extremely evening persons have a bad sleep quality and excessive daytime sleepiness, probably due to long hours of study at night and packed schedules of class time starting very early in the morning. This should be taken into account when planning classes.

Is important to pay attention at the high prevalence of sleep disorders in this population, because of the negative impact on life quality, cognitive performance, and other associated health disorders.

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1504
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Late Breaking Posters 1
Ocular tilt reaction due to central nervous system (cns) paracoccidioidomycosis: case report

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Background: Ocular tilt reaction (OTR) is a rare syndrome caused by an unilateral interruption of the otolithic afferent pathway. OTR consists of head tilt, skew deviation, and ocular torsion combined with tilt of perceived verticality. Paracoccidioidomycosis (PCM) is a systemic mycosis that affects the CNS in 9-25% of cases.

Objectives: To report a case of ocular tilt reaction caused by *Paracoccioides spp*.

Patients and methods: We describe a case of a 50-year-old male rural worker from Paraguay with smoking and drinking habits. The patient's complaints were: headache, dizziness, vertical diplopia and gait instability. Physical examination revealed right dysmetria, head tilt to right, skew deviation and an ulcerated lesion on the palate. I have obtained patient and Institutional Review Board approval, as necessary.

Results: CNS Magnetic resonance(MRI): Nodular lesion on left cerebral peduncle hypointense in T2 and T1- weighted images, hyperintense in diffusion-weighted images, surrounding edema and annular enhancing after gadolinium injection. Cerebrospinal fluid: normal, with negative microbiological tests and cultures. Ophthalmology examination revealed torsion of both eyes to right. Biopsy of palate lesion and serology test positive for *Paracoccidioides spp*. The patient received antimycotic treatment to which he responded positively, besides, an improvement in the lesions was also shown on MRI.

Conclusion: It is important to consider PCM as a differential diagnosis in patients from endemic areas that show lesions in the CNS, given that is a treatable and reversible condition. To the best of our knowledge, there are no reported cases of OTR due to systemic mycosis in the literature.

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1505
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Late Breaking Posters 1
Deep convolutional neural networks for automatic identification of epileptic seizures in infrared and depth images

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Background: Epileptic seizures are often accompanied by strong patient motion that follows specific patterns. This pattern can be captured by a video-based system that will raise an alarm upon identification of an epileptic seizure, which is valuable for both inpatient as well as home monitoring scenarios.

Objective: The aim of this work is to determine a model of epileptic seizures in video data, which allows to automatically identify epileptic seizures in real time.

Patients and methods: In an epilepsy monitoring unit, 52 seizures were recorded with a combined infrared and depth imaging sensor at 15fps. We have obtained patient approval, as necessary. A deep convolutional neural network architecture (CNN) was trained on frames from the ictal as well as from the interictal phase. One CNN was used to process infrared frames and a second CNN to process depth frames. The output of both networks was combined to achieve a final decision for either ictal or interictal phase.

Results: Cross-validation revealed high sensitivity (87%) and specificity (81%) for general convulsive seizures. Taking into account also tonic and automotor seizures, the average sensitivity was 80% at a 59% specificity, while competing methods such as the average motion frequency or image gradient histograms scored worse.

Conclusions: Instead of a predefined model that expects only a certain type of motion, our model learns from raw input data and appears to capture more salient information from seizure recordings. Furthermore, including not only infrared but also depth data improved the ratio of correctly identified epileptic seizures.

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1506
WFN15-1717
Late Breaking Posters 1
Prednisolone and azathioprine are effective in dppx-autoantibody-positive autoimmune encephalitis

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Background: Antibodies to dipeptidyl-peptidase-like protein-6 (DPPX-Ab) have recently been described in patients with encephalitis. So far, only a few patients with DPPX-Ab have been reported and little is known about treatment responses.

Case report: A 40-year old woman presented with an unsteady gait, ataxia of the arms, a whole-body tremor, memory deficits, a profound startle response and panic attacks lasting for 1.5 years. She experienced a severe weight loss of around 80 kg (previous weight:160 kg) and had night sweats and diarrhea preceding the neurologic symptoms. Standard CSF was normal, with no evidence of intrathecal IgG synthesis. S100 protein was elevated in CSF. EEG and brain MRI were normal initially and 6 months later. No tumor was identified by repeated whole-body CT scans and FDG-PET. A highly positive IgG serum titer of autoantibodies against DPPX (1/10.000) was noted. Indirect immunofluorescence demonstrated typical patterns on DPPX-transfected cells and neuronal tissue sections. No further autoantibodies against 34 other neurological target antigens were identified.

The patient received IV methylprednisolone, followed by 80 mg prednisolone orally. All neurologic symptoms and cognition substantially improved over 6 weeks and completely resolved within 3 months. Prednisolone was slowly tapered over 6 months to 5 mg daily. Azathioprine was initiated at 50 mg/day and gradually increased to 300 mg/day. Within 6 months, the DPPX-IgG titer markedly declined. The patient remained clinically stable.

Discussion: Most patients with anti-DPPX-Ab positive encephalitis initially benefit from glucocorticosteroids but relapse during dose reduction and require prolonged immunosuppression. Our patient clearly and persistently improved after treatment with prednisolone together with azathioprine. We suggest that in anti-DPPX-Ab-positive encephalitis patients initially receive glucocorticosteroids alone, and if they respond, moderate immunosuppression by azathioprine be added during reduction of the glucocorticosteroid.

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1507
WFN15-1723
Late Breaking Posters 1
Unusual case of gliomatosis cerebri in a male patient with anaplastic astrocytoma

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Background: Gliomatosis Cerebri is a diffuse neoplasm of glial cells origin usually astrocytic infiltrating at least two cerebral lobes. The present symptoms of gliomatosis cerebri in literature are nonspecific.

Objective: A case-report of gliomatosis cerebri.

Methods and results: A 55-year-old man presenting insidious altered behavior, apathy and progressive memory loss. No motor or sensory deficit were noted. Headache complaint is present, non-specific. Personal and family medical history is unremarkable. Blood cell count, liver functions, renal function, toxicologic evaluation, thyroid hormones, vitamins and virus evaluation were normal. MRI revealed hyperintensities in T2 and FLAIR comprising right corpus callosum, thalamus, right basal frontal lobe, temporal lobe and diencephalon and white matter, without mass effect and with no Gadolinium enhancement (panel A). CSF analysis, autoantibodies studies, infectious diseases and systemic neoplastic evaluation were unremarkable. Empirically, while specific tests were being conducted, patient received antiviral therapy and systemic corticosteroids

with no results. A complementary Spectroscopy revealed high ratios of Choline/creatine and Choline, with peaks of 2.5 Cho/NAA (figure 1). This finding guided a stereotaxic cerebral biopsies in which was found a diffuse anaplastic astrocytoma grade 3 pathology.

Conclusions: Gliomatosis Cerebri should be included on the differential diagnosis when multiple brain abnormalities were found. Peaks of incident occurs between 40 and 50 years, slightly higher in males. The prevalence is underestimated. It is no recognizable clinical pattern. Gliomatosis invades predominantly white matter. slight contrast enhancement might be present.

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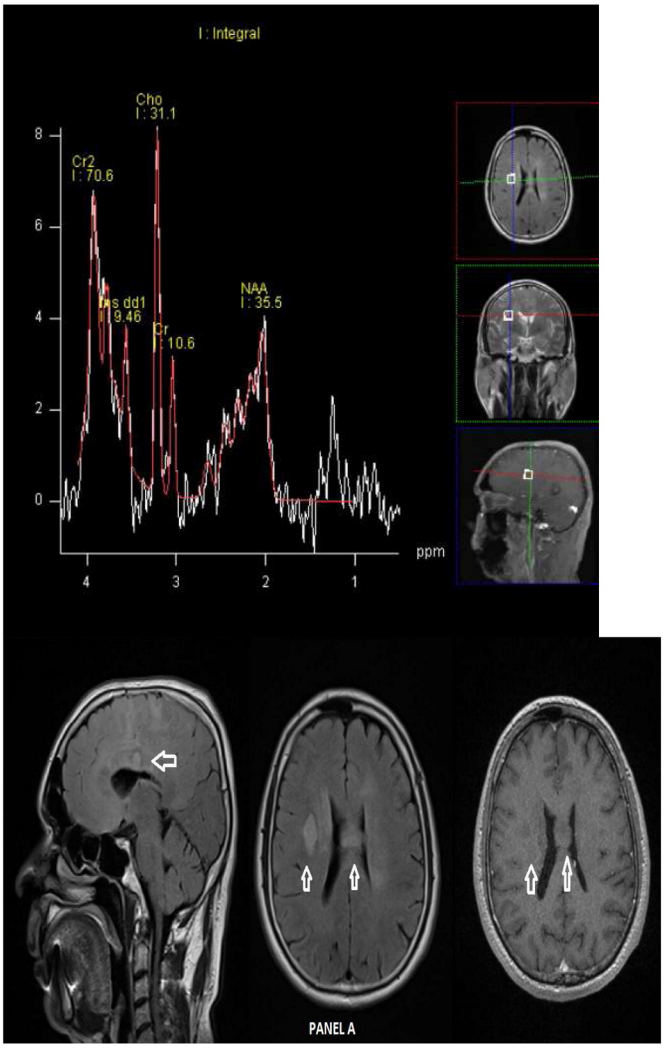

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1508
WFN15-1725

Late Breaking Posters 1
Epilepsy in patients with metabolic diseases, clinical features, study and treatment

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Introduction: Seizures are a common symptom in a great number of metabolic diseases. They occur as an occasional event secondary to metabolic decompensation or as a known epileptic condition. Epilepsy in these cases can be classified depending on physiopathology, time of onset and type of crisis.

Objective: To determine the main metabolic disorders associated with epileptic seizures, time of onset, form of presentation and response to treatment.

Materials and method: Retrospective study of 130 patients with metabolic diseases on our service. Review of clinical history.

Results: 23 metabolic diseases were evaluated, with a total of 130 patients, of which 68 (52%) presented epileptic seizures. Crisis presented between the first hours of life and 21 years of age (mean 60 months). Focal seizures predominated in 74%, followed by generalized seizures in 26%. Epilepsy was found in 100% of patients with: non ketotic hyperglycemia, gangliosidosis, lipofuscinosis, peroxisomal disorders of neonatal onset, sulfite oxidase deficiency, MELAS, MERRF and GLUT 1 deficiency. The most used antiepileptic drugs were phenobarbital, followed by carbamazepine and valproic acid. 22% of patients evolved with pharmacological refractoriness.

Conclusion: In our series, more than half of patients (52%) presented epileptic seizures. The main metabolic diseases associated to seizures were the ones described. Age of onset was more frequently during the first five years of life. Focal epileptic seizures with secondary generalization predominated and 22% of refractoriness was observed. The identification of epilepsy secondary to metabolic disorders is of great importance to establish an early and appropriate treatment.

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1509
WFN15-1727
Late Breaking Posters 1
Cerebral cavernous malformations in children: clinical features and outcome

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Introduction: Childhood cavernomas incidence is increasing. Its natural history, surgical treatment and clinical outcome are not well understood in children.

Objective: The aim of this study is to describe the clinical profile of patients with diagnostic of cavernomas in the Pediatric Neurology Department of our Hospital, between 1998-2010.

Methods: N=19 patients. Variables as age, clinical presentation, localization, treatment and evolution were described.

Results: 14 (73,7%) male, mean age of symptoms debut was 7,6 years (11 month to 14 years old), 31,5% of the patients were younger than 3 years and 26,3% older than ten years.

Clinical manifestations: seizures(63,2%), focal neurologic signs (31,6%),headache (31,6%). The onset of symptoms were related in 29,4% with radiological signs of bleeding. 84.2% were solitary lesions located at supratentorial level (73,7%), brainstem (10,5%) and spinal cord (10,5%). Of the supratentorial lesions 64,3% were left sided and were more frequent in temporal and frontal lobe (both 37,5%). CT sensibility was 53,4% and MR 88,2% . Surgical resection was practiced in 7 cases and no surgical management in 12 cases with favourable results in 4 and 9 cases respectively. Epilepsy was the most frequent complication (21,5%).

Conclusion: In our group cavernomas were more frequent in men and mostly presented as solitary supratentorial lesions. Epilepsy is the most frequent long term complication, similar to those published in other series.

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1510
WFN15-1729
Late Breaking Posters 1
Paroxysmal dyskinesias in pediatric patients

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Introduction: Paroxysmal Dyskinesias are involuntary intermittent movements, manifested with dystonia, chorea, athetosis, or a combination of them. They are secondary to ion channels defects.

Objectives: The aim of this study is to define the most frequent forms of presentation in our population and the management response.

Materials and methods: Retrospective study of medical records of 15 patients with paroxysmal dyskinesias treated at our centre between 1991-2010.

Results: 15 patients, 6 women and 9 men were evaluated, all of them presented normal development and no other pathologies. 4 had family history of abnormal movements (3 were brothers, who also had febrile seizures history). The average age of onset was 3.9 years. Dystonia: 6 patients generalized 3 patients hemidystonia (6 nonkinesigenic and 3 kinesigenic). Choreoathetosis: 5 generalized and 1 hemichorea, 4 of them were nonkinesigenic and 2 kinesigenic. All patienes had normal imaging, electrophysiological and metabolic studies. 7 patients received medical treatment with carbamazepine and 4 with acetazolamide, all of them with good clinical response.

Conclusion: In our study we found a male predominance in all dyskinesia patients. Dystonia was more common than choreoathetosis. Kinesigenic dyskinesia was predominant. There was a good response to symptomatic treatment. Three patients corresponded to a familial form of paroxysmal choreoathetosis with febrile seizures. We found no secondary causes.

Discussion: Although dyskinesias are often sporadic or familial and presented frequently spontaneous remission, it is important to look for secondary causes and discard epilepsy as the main differential diagnosis.

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1511
WFN15-1731
Late Breaking Posters 1
Neurotransmitter disease, clinical characteristics and outcome in a serie of chilean patients

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Introduction: The neurological manifestations of congenital metabolic diseases of aminergic neurotransmitters (NT) are diverse. The autosomal dominant Dopa-responsive dystonia (DRD), with deficiency of GTP cyclohydrolase1 (GTPCH1), is the most common type, with a satisfactory response to treatment. We describe clinical features, response to treatment and outcome of patients diagnosed with inborn errors of aminergic neurotransmitters in our center.

Methods: A retrospective descriptive study and a prospective follow-up of 17 patients. Review of clinical records.

Results: 17 patients. 16/17 exhibit DRD. 12/16 women. On 9/16 the average was 5 years age at onset and 9.5 years at diagnosis. In all patients the initial symptom was gait disturbance with diurnal fluctuation, lower limb (8/9) and upper limb (8/9) dystonia, trunk dystonia (3/9), tremor (3/9). Adult relatives (7/16) begin symptoms between 20 and 40 years: focal dystonia, parkinsonism. The mode of inheritance was autosomal dominant. The phenylalanine test was guiding. Diagnosis is confirmed with measurement of CSF levels of NT: low concentrations of neopterins, biopterins and 5HIAA HVA suggest deficiency of GTPCH1. Positive genetic study in 2 families. The response to levodopa treatment was satisfactory. One patient (1/17) shows a deficit of L-Dopa decarboxylase with severe global psychomotor retardation, fever, hypotonia, epilepsy, dystonia, and fatal outcome.

Conclusions: In our series predominates DRD, with clinical features and response to treatment classically described. Early diagnosis allows prompt treatment with improvement of symptoms and favorable course.

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1512
WFN15-1732
Late Breaking Posters 1
Clinical phenotype, genetic and imaging features in vanishing white matter disease/cach

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Introduction: VWM is an autosomal recessive hereditary leukodystrophy, linked to mutations in genes encoding the eukaryotic initiation factor 2B (eIF2B). Neurological features and clinical phenotypes are variable. The most common variant has a childhood onset and characteristic abnormalities on cranial MRI.

Methods: Retrospective and prospective study: clinical, neuroimaging and genetic analysis. Patients were divided in groups according to age at onset: Group 1 (<2 years), group 2 (age 2-5) and group 3 (age >5), and according to disability scores (1=gait disturbance to 5=deceased).

Results: 10 patients (7male/3female), 3 siblings. 9/10 had prior normal development. Average age at onset was 7 (1-13years), with gait disturbance (6/10), development regression (3/10) and tremor (1/10). History of cranial concussion (4/10) and febrile infection (1/10). Group distribution: 1 (n=2), 2 (n=1), 3 (n=7). Disability score distribution: 1 (n=6), 2 (n=1), 4 (n=2), 5 (n=1). Phenotypes: late childhood/early childhood onset (7/3). Symptoms: spasticity (7/10), cerebellar ataxia (7/10). Clinical monitoring: 3-12 years, rapidly progressive (n=6), progressive (n=2), fulminant (1) evolution. Brain MRI: diffuse, symmetrical abnormal white matter signal (10/10), most with cystic degeneration. Genetic study (9/10): IF2B5-R113H mutation (homozygous/heterozygous=2/6), IF2B4-R373C homozygous (1).

Conclusion/Discussion: VWM is one of the most prevalent inherited childhood leucoencephalopathies, therefore it must be considered in the differential diagnosis. In this series the classical form was the most frequent, with characteristic white matter abnormalities in MRI, and the most with IF2B5- R113H mutation.

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1513
WFN15-1735
Late Breaking Posters 1
Effect of eslicarbazepine acetate on cognition in children with epilepsy

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Eslicarbazepine acetate (ESL) does not affect cognition in adults, but its effect in children is uncertain. This was a phase-II, randomized, double-blind, placebo-controlled study aimed to evaluate ESL effect on cognition in children with partial-onset-seizures (POS).

Methods: Patients (6–16 years) with POS (≥2 in the month before enrolment), receiving one-two antiepileptic-drugs (except oxcarbazepine), were randomized to ESL or placebo (2:1). ESL was titrated from 10 to 20 mg/kg/day over 4-weeks. If no intolerable AEs occurred, patients were up-titrated to 30mg/kg/day (maximum 1200 mg/day) maintained for 8-weeks. Down-titration was allowed only once. Primary endpoint was the change from baseline in composite Power of Attention (PA) measure. PA was defined as the sum of reaction time measures from attentional tasks (simple and choice reaction-times, digit vigilance-speed). A non-inferiority (NI) test was used with 121 milliseconds as limit. Efficacy was assessed as relative reduction in standardized-(/4week)-seizure-frequency (SF) and proportion-of-responders (≥50%SF-reduction) from baseline. Safety/tolerability included incidence of treatment-emergent AEs (TEAEs).

Results: 75 (90%) ESL and 37 (93%) placebo patients completed the trial. There were essentially no PA differences between treatments ($p>0.5$; Figure 1). However, non-inferiority failed to reject the null hypothesis as 95%CI lower-bound was below the NI margin. Mean SF relative change was -9% for placebo and -31% for ESL ($p<0.001$, Figure 2). Responders were 25% for placebo and 51% for ESL ($p=0.009$, Figure 2). Overall incidence of TEAEs was similar (45% for ESL and 48% for placebo). Most frequently reported TEAEs were headache, somnolence and vomiting.

Conclusions: In children with POS, adjunctive ESL treatment does not appear to have negative consequences for attention, information-processing and working-memory. ESL was efficacious, well tolerated and no new or unexpected safety findings emerged.

Figure 1. Effect on the magnitude of response [relative change (%) in SF] by increasing sample size (from ITT to mITT set)

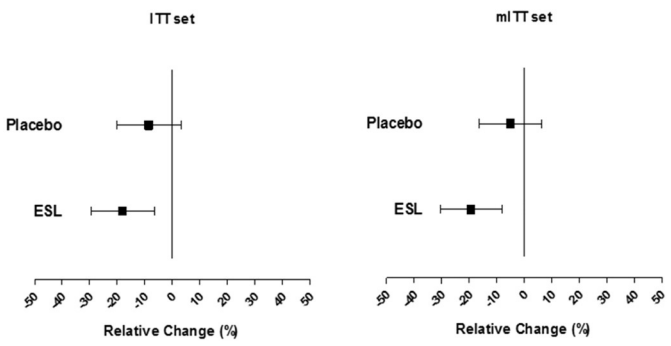
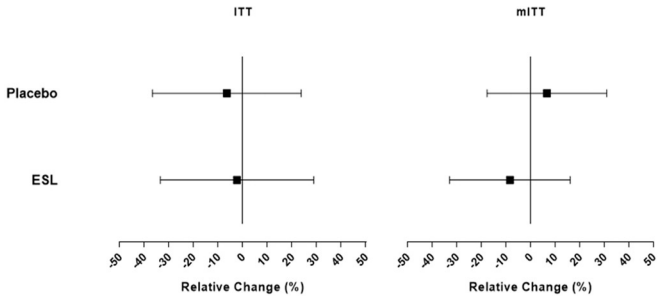


Figure 2. Relative (%) change from baseline in SF for the 2-6 years group (ITT and mITT sets)



doi:10.1016/j.jns.2015.09.073

1514
WFN15-1736
Late Breaking Posters 1
A placebo-controlled trial of eslicarbazepine acetate add-on therapy for partial seizures in children

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Eslicarbazepine acetate (ESL) is approved as adjunctive therapy for partial-onset-seizures (POS) in adults. Given its efficacy, good tolerability and convenience of once-daily dosing, ESL could also be beneficial for children. This was a phase-III, randomized, double-blind, placebo-controlled study aimed to demonstrate ESL safety and efficacy as add-on therapy in children with POS.

Methods: Children (2–18years) with POS (≥4 in the month before enrolment), receiving 1-2 antiepileptic-drugs (except oxcarbazepine), were randomized (1:1) to ESL or placebo. ESL started at 10mg/kg/day and uptitrated to 20mg/kg/day (maximum 1200mg/day). If tolerability and response were considered acceptable, 20mg/kg/day was maintained 12-weeks. If only tolerability was acceptable, patients uptitrated to 30mg/kg/day (maximum 1200mg/day) and maintained 12-weeks. Downtitration was allowed only once. Primary efficacy endpoints were relative reduction in standardized-(/4week)-seizure-frequency (SF) and proportion-of-responders (≥50% SF reduction) from baseline. Safety/tolerability included the incidence of treatment-emergent AEs (TEAEs).

Results: 87%-ESL and 89%-placebo patients completed the trial. Oral-suspension was recalled affecting few stratum-I (2-6years) subjects who were included in modified-ITT (mITT). An unbalanced seizure-frequency at baseline was observed (mean±SD: 62±186 and 37±72 for placebo and ESL, respectively). SF relative change was -18.1% for ESL and -8.6% for placebo (95%CI:-6.7,25.8; $p=0.2490$, ITT). For mITT, difference was larger (14.3%, 95%CI:-1.7,30.3, $p=0.0792$) (Figure 1). Responders were similar between groups. Stratum-I was affected more in variability (higher) and magnitude (less) of response (Figure 2). Incidence of TEAEs was 84% for ESL and 73% for placebo. Most frequently reported TEAEs were headache and somnolence.

Conclusions: In this study in children with POS, ESL was well tolerated but not superior to placebo. A high placebo response, unbalanced seizure-frequency at baseline and a potential suboptimal eslicarbazepine exposure for 2-6years group should be considered when analysing data.

Figure 1. Effect on the magnitude of response [relative change (%) in SF] by increasing sample size (from ITT to mITT set)

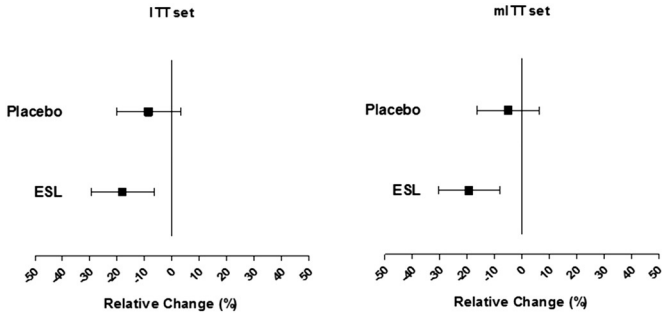
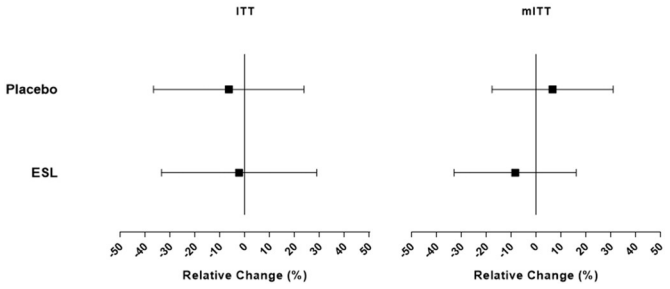


Figure 2. Relative (%) change from baseline in SF for the 2-6 years group (ITT and mITT sets)



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1515
WFN15-1746
Late Breaking Posters 1
West syndrome in patients with down syndrome. Clinical description and management

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Introduction: The prevalence of seizures among people with Down Syndrome (DS) is greater than in the general population (1-13%). A third of these cases present as West Syndrome(WS) epileptic encephalopathy, characterized by spams, regression/ retardation in the psychomotor development (PMD) and hypsarrhythmia. Typically these cases have a satisfactory outcome.

Objective: To analyze clinical characteristics, EEGs, outcome of patients with DS associated with WS.

Method: A retrospective descriptive study of 7 patients with DS associated with WS between January 2002 and May 2011 in our department.

Results: Out of 7 patients (4/7 women) the average first occurrence of spams is at 9 months of age. Delay diagnosis: average age 6.2 months. 7/7 present retardation PMD, 2/7 regression PMD. 7/7 Flexor spam. 6/7 received ACTH and valproic 1/7 valproic only. Post cure ACTH: 4/6 complete termination of spams with normal EEG (2/4). 2/6 partial remission with persistent spams hypsarrhythmia (1/2) and severely abnormal EEG non hypsarrhythmic (1/2). Outcome: Patients with good initial response to treatment, clinical and/or electrical (4/7) showed no recurrence or persistence of crisis at 6 months follow up. Conclusion: Nearly 40% were unfavorable, not concordant with the literature. I have obtained patient approval, as necessary.

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1516
WFN15-1748
Late Breaking Posters 1
Dravet syndrome patients with genetically confirmed diagnosis

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Introduction: Dravet Syndrome (DS) is an epileptic encephalopathy. It begins with febrile seizures, and then adds different types of seizures which are refractory to treatment. In more than 70% of cases a mutation in the voltage dependent sodium channel (SC-N1A) is identified.

Objective: Describe the clinical and electrophysiological characteristics of 4 patients with DS genetically confirmed.

Materials and methods: Descriptive retrospective analysis of medical histories, EEG and genetic studies with prospective follow-up.

Results: 4 patients, 4/4 normal development before seizures, 1/4 epilepsy in family. First seizure between 4-6 months, 3/4 febrile, 3/4 over 15 minutes. 4 patients had all kinds of seizures except for tonic, being partial secondary generalized, myoclonic and complex-partial seizures the most frequent. 4 patients have refractory epilepsy with frequent status. 2/4 worsen seizures with Lamotrigine. All evolve with a moderate-severe developmental delay, 3/4 with ataxia, 2/4 with pyramidalism, 3/4 with autistic behavior. All had normal EEGs during first year of life, evolving with focal-multifocal activity and disorganized background activity. Photosensitivity (+) 1/4. Normal brain MRI 3/4, 1/4 with diffuse cortical atrophy. All cases had punctual heterozygous mutations in exons 4,7 and introns 5,22 of SCN1A gene.

Conclusion: The 4 patients had the punctual mutations of SCN1A gene, they all presented the classic presentation of DS.

Discussion: One of the first national reports of the experience on molecular diagnosis of SD.

I have obtained patient approval, as necessary.

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1517
WFN15-1749
Late Breaking Posters 1
Risk factors associated with perinatal arterial ischaemic stroke: a case-control study

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Introduction: Perinatal arterial ischaemic stroke (PAIS) is an important cause of morbidity and mortality. The pathophysiology and associated risk factors are not completely understood.

Objective: Describe the clinical presentations of PAIS and to evaluate associated risk factors.

Materials y methods: Case-control study. We evaluated clinical data of patients with PAIS controlled in our center between 1993 and 2012. Each infant with PAIS was matched to three healthy controls. Risk factors were studied using univariate and multivariate conditional logistic regression analysis.

Results: We analyzed 40 patients (66,7% male, 33,3% female). The mean gestational age was 39 weeks. 21(52,5%) of cases were confirmed as perinatal arterial ischaemic stroke (PAIS) and 19 (47,5%) presumed perinatal arterial ischaemic stroke (PPAIS), the mean age of diagnosis were 6 days and 2 years-3month, respectively. The most frequent clinical presentation were seizures(86%) for PAIS and focal neurologic signs(95%) for PPAIS. Strokes preferentially involved the MCA

territory (88%), 95% unilateral, 65% in the left hemisphere. All patients presented some neurologic deficit in the following clinical controls, the most common were hemiparesis and epilepsy. Significant risk factors in the multivariate analysis ($p<0.05$) were nulliparity (OR 11.74; CI 3.28-42.02), emergency caesarean section (OR 13.79; CI 3.51-54.13) and Apgar score (5 min) ≤7 (OR 13.75; CI 1.03-364.03).

Conclusions: The principal clinical profile were seizures in PAIS and focal neurological signs in PPAIS. The prognosis was in general poor, all patients presented neurologic alterations. The risk factors nulliparity, emergency caesarean section, and Apgar score (5 min) ≤7 were found to be important risk factors in PAIS.

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1518
WFN15-1750
Late Breaking Posters 1
Small vessel disease: mirror symptoms, two different spectrum of the disease

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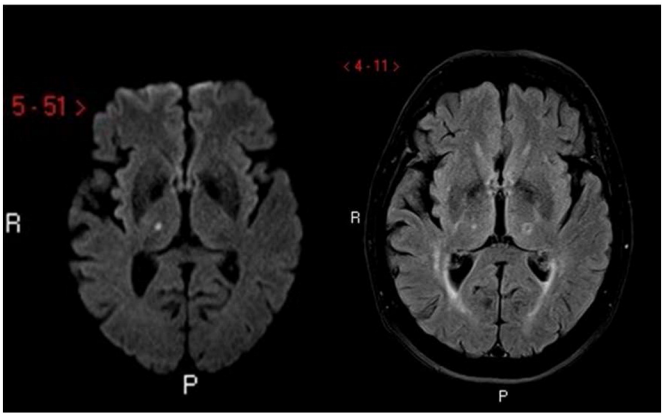
Background: Cerebral small vessel disease describes a range of neuroimaging, pathological, and associated clinical features. Clinical features range from none to insidious global neurological dysfunction and dementia. The main imaging features visible on MRI include acute lacunar infarcts or hemorrhages, white matter hyperintensities, visible perivascular spaces, micro bleeds and brain atrophy.

Case Report: A 64 years man with prior history of hypertension, abdominal aortic aneurysm, dyslipidemia, and thrombosis of a right popliteal aneurysm. In his first admission he came complaining of paresthesias in the third, fourth and fifth fingers of the left hand and in the left side of the perioral region, on physical examination he had diminished tactile sensation in the same regions. A week later, he returned with similar symptoms but in the right side, like “mirror symptoms”: paresthesias in the third, fourth and fifth fingers of the right hand on physical examination, as in the first admission, he had hypoesthesia in the related regions without any more signs.

On MRI, imaging suggestive of an ischemia in right thalamus, (event found in the first admission) and an acute hemorrhage in left thalamus.

Conclusion: Similar symptoms with different neuroimaging findings and the same disease: Cerebral small vessel disease and a challenge choosing preventive and treatment options.



doi:10.1016/j.jns.2015.09.078

1519
WFN15-1751
Late Breaking Posters 1
Clinical profile, risk factors and severity in patients with pediatric arterial ischemic stroke

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Introduction: Stroke is a cause of childhood disability. The aim of this study is to describe the clinical profile and risk factors in a series of patients.

Methods: Children aged between 29 days -18 years were enrolled (1989-2012). Clinical characteristics and risk factors were described by age group. PedNIHSS severity score was estimated and correlated with the presence of risk factors, by ordinal and linear regression models.

Results: From 63 patients (average age 3,52 years) 67% were male. The most common symptoms were motor deficit (78%) and seizures (47%). The main risk factors were arteriopathy (60%) and infection (40%). Prevalence of systemic conditions was higher in children <3 years ($p < 0,05$). An average PedNIHSS score of 7.4 (0-17) was obtained. There is no significant relationship between the number of risk factors and PedNIHSS score. In a linear regression model, the presence of acute disorders of the head and neck was significantly correlated with PedNIHSS score ($p < 0,05$).

Conclusions: We confirmed male predominance and motor deficit at debut as previously described. We found at least one risk factor in all patients with complete information. We confirmed arteriopathy as the main risk factor, but we did not find any case of focal transient arteriopathy, may be because of lack of follow up neuroimaging. Searching for all risk factors is important, because the number of them is not predictable by severity score, but is important for recurrence risk. Acute disorders of the head and neck are predictors of higher PedNIHSS score.

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1520
WFN15-1752
Late Breaking Posters 1
Amyloid spells: a diagnostic challenge

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Background: Cerebral amyloid angiopathy is characterized by amyloid beta peptide deposits within small to medium-sized blood vessels of the brain and leptomeninges. Although it is usually asymptomatic, it is an important cause of primary lobar intracerebral hemorrhage and it may present with transient neurological symptoms, an inflammatory leukoencephalopathy, as a contributor to cognitive impairment, or with incidental microhemorrhages or hemosiderosis on MRI.

Case report: A 79 years old man, with history of hypertension, ischemic heart disease, an abdominal aneurysm treated with endovascular stent and former smoker. He was admitted complaining of paresthesia in the left lower and upper limbs lasting 5 to 10 minutes. After that episode, he presented another three similar episodes with the same duration. Neurological examination was normal. Investigated like a transient ischemic attack, the MRI found superficial siderosis in the parietal and frontal lobes and a subarachnoid hemorrhage in the medial region of the right central sulcus, suggesting a cerebral amyloid angiopathy.

Conclusion: As cortical superficial siderosis has been reported to present with transient focal neurological episodes it is necessary to differentiate it from acute transient ischemia, a misdiagnosis may induce the use of anti-platelets or anticoagulants and finally to intracranial hemorrhage.

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1521
WFN15-1754
Late Breaking Posters 1
Seizures not related to hypoglycemia in type 1 diabetic patients (Dm1)

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Introduction: In recent years there has been much attention to elucidating existence of an association between DM1 and epilepsies in children. Previous reports identify higher prevalence in DM1, others similar to the genera lpopulation, postulating an association between elevated levels of anti- glutamic acid decarboxylase (anti-GAD) antibodies in patients with DM1and epilepsy.

Objective: clinical characterization patients with DM1 and seizures not associated with hypoglycemia, and search partnership with anti GAD antibodies. Material

Method: Retrospective descriptive study, analysis of clinica lrecords.

Results: 8 patients, 7 women- 1 men (fig.1), mean age start DM1 6.1 years, and presentation first seizure 7.6 years (table 1). 7cases the diagnosis of DM1 precedes onset of seizures, 4 with partial seizures, 4 with generalized seizures, 5 have altered EEG, 5 treatment with valproic acid, 7 have normal neuroimaging (Table 2). The GAD antibody levels measured in 7 patients were positive in 6 (Table 3).

Discussion: identify seizures in patients with DM1 is complicated by the possible association with hypoglycemia. We agree with previous reports, which in most cases DM1 precedes seizure. We found no difference in number of patients with focal and generalized seizures. Most have positivity for anti GAD antibodies, which would support the existence of suspected autoimmune basis of the disorder.

Conclusions: In this series shows a relationship between patients with DM1 and no hypoglycemic seizures, may be involved an autoimmune component, what would be the common factor in both disorders.

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1522
WFN15-1756
Late Breaking Posters 1
Clinical and genetic manifestations of chilean patients with DNA mitochondrial disease

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Introduction: Mitochondrial diseases are a group of maternally inherited disorders, clinically heterogeneous produced by mitochondrial DNA mutations. Clinical features related to a specific mutation are usually variable and multisystemic.

Objective: To evaluate clinical manifestations, genogram, testing and evolution of patients diagnosed in our centre with mitochondrial diseases and their phenotypic characteristics in relation to the genotype with different point mutations of mitochondrial DNA(A3243G, G11778A, A8344G).

Methods: Retrospective descriptive and monitoring of all patients with mitochondrial DNA mutations confirmed. Review of clinical records.

Results: 45 patients were studied,9 present with A3243G mutation,33 G11778A mutation and 3 A8344G mutation. In patients with A3243G mutation, average age of onset symptoms was nine years: headache (5/9), stunting (9/9), sensorineural deafness (8/9), cardiac disorders (2/9). They present stroke-like episodes(9/9) between 6 to 21 years, generalized tonic-clonic seizures (9/9). Study:elevated lactic acid plasma-CSF relation (9/9), ragged-red fibers (RRF)(7/9), CT/MRI: basal ganglia calcification(8/9), areas of infarction (strokelike) temporoccipital(9/9). Evolution:progressive, 3 died. Their relatives were affected by deafness, diabetes, heart disease. 33 patients with G11778A mutation, 12 symptomatic. Presentation mean age 16 years, visual impairment (9/12), optic atrophy (7/12), impaired gait (4/12), dystonia (3/12). CT/MRI: putamen necrosis (4/12). Evolution: stable (5/12), slowly progressive (7/12). 3 brothers with A8344G mutation, average age presentation 10.6 years: all with myoclonic epilepsy, neuropathy, ataxia and FRR(+). Evolution: progressive.

Conclusions: In our series, A3243G mutation was related to mitochondrial encephalomyopathy with lactic acidosis and stroke-like episodes (MELAS) phenotype, G11778A mutation with Leber's optic neuropathy (LHON) phenotype and mutation A8344G with myoclonus epilepsy with ragged red fibers (MERRF) phenotype. Clinical manifestations, tests and maternally inherited form were the classically described for these phenotypes.

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1523
WFN15-1757
Late Breaking Posters 1
Pediatric arterial ischemic stroke: outcome and prognosis features

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Background and aims: arterial ischemic stroke is an uncommon but severe cause of pediatric disability. Our aim is to evaluate prognosis and evolution in patients diagnosed with arterial ischemic stroke between 1 month and 18 years in San Borja Arriarán Hospital in Santiago, Chile.

Methods: study design was descriptive-retrospective by search on clinical records from 1989 to 2012.

Results: were included a number of 64 patients with arterial ischemic stroke between 1 month and 14 years. 55/64 patients was a single episode and 9/64 had recurrence of the event. 3 patient no had sequels. Of the 61 patients with sequels, 65 % (40/61) evolved with hemiparesis, 4/61 with tetraparesis and 1/61 diparesis. 6 patients had extrapyramidal symptoms and 4 with cerebellum symptoms. Epilepsy was evident in 23/61 patients, while 17 patients presented cognitive impairment in different grades.

Conclusions: is evident the high degree of sequelae in our population, mostly in the motor área. The mortality was lower than international reports. Is necessary to find new methods to prevent or ameliorate the impairment of children affected with the disease.

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1524
WFN15-1758
Late Breaking Posters 1
Acute allergic reaction following injection of generic, government-provided botulinum toxin a (GGP-BTA) for hemifacial spasm

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Background: Allergic reactions after administration of Botulinum Toxin A are rare, unpredictable complications and may vary from rash to anaphylaxis. These have been reported with the use of commercially available formulations, but not with generic formulations. These are probably due to allergic reactions to albumin, lactose or gelatin use in the formulation of BTA.
Objective: To report the case of a patient who developed facial angioedema one hour after receiving intramuscular Generic, Government-provide Botulinum Toxin A (GGP-BTA).
Patient and method: Case report.
Result: A patient with left hemifacial spasm, who had already been treated with commercially available BTA (Botox®) with no significant side-effects, was treated with intramuscular injections of GGP-BTA on the affected muscles of the left side and additional injections in the periocular region of the right side for aesthetic symmetry. About one hour following the procedure he developed a purplish-red oedema of the eyelids, as well as the left nasolabial fold (Figure). He was treated at first with IV Methylprednisolone, followed by oral 60 mg qd of Prednisone. As there was minor improvement with corticosteroids, following evaluation by the Dermatology team, he received intramuscular Adrenaline and was started on oral Diphenhydramine with resolution of the angioedema.
Conclusion: Although rare, allergic reactions to commercially available BTA may vary from rash to life-threatening anaphylaxis. We reported a case of facial angioedema following injection of GGP-BTA, with resolution after adequate systemic treatment with anti-allergic medications.

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1525
WFN15-1766
Late Breaking Posters 1
Phenotypic description of Rett syndrome patients with MECP2 gene mutation

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Introduction: there are several clinical phenotypes related to mutations in MECP2 gene, being Rett syndrome in females the most frequent in its classical and variant forms, and syndromes in males going from encephalopathy to mental disability.
Objective: to establish the clinical phenotype and evolution of patients with mutations in MECP2 gene and to describe its relation to the phenotype.
Materials and methods: descriptive and retrospective study of patients with mutations in MECP2 gene encountered in our Service between 2005-2009. Cases with positive mutations are studied from the protocol applied to girls with Rett syndrome phenotype.
Results and discussion: 11 girls with Rett syndrome phenotype, 9 classical forms and 2 variants. The 9 girls with the classical form had normal initial development, regression between 6 to 18 months, stereotypies emergence, 7 lost hand propositivity, 6 had epilepsy, 7 acquired microcephaly, 4 achieved gait and none had language. 6 patients had frequent mutations R255X, R168X y R306C with similar clinical severity. The 2 patients with variant forms had normal development until 3 years old, slow progression of language, normocephaly, epilepsy, late stereotypies, preserved gait, with R294X and TRUNC293 mutations.
Conclusion: phenotypes of MECP2 gene mutations manifest as usually is described in females with Rett syndrome. Most are classical forms, with the exceptional gait achievement as highlight. The most frequent mutation is R255X and the severity of the phenotype is not related with a mutation in particular.

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1526
WFN15-1768
Late Breaking Posters 1
Allocentric neglect strongly associated with egocentric neglect during intraoperative eletrophysiological stimulation in low grade glioma

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Abstract

The authors show a case of 34 year old female patient with a tumor - Astro grade II in supramarginal gyrus and and angular gyrus on the right side, who presented as initial clinical picture a partial motor and complex seizure, of difficult control. During the craniotomy the angular area was stimulated with 3 volts, with a monopolar tip, as well as the supramarginal gyrus, during intraoperative neuropsychological testing. The response was curious, being allocentric neglet strongly associated with egocentric neglect. There is an established tradition for considering these two phenomena as both behaviorally and anatomically dissociable. However, several studies and some theoretical work have been suggested that these rather reflect two aspects of a unitary underlying cause. An consistent and important work of Yue et al. (2012), suggests that allocentric behavioral deficits might appear only in conjunction with egocentric deficits as well as there would an large corresponding overlap for the anatomical regions associated with egocentric and with allocentric neglect. The authors discuss how different anatomical and behavioral findings can be explained in a unified physiologically plausible framework, whereby allocentric and egocentric effects interact.

Our findings suggest strong association between egocentric and allocentric neglect.

Figure 1- A- MRI showing the glioma in supramarginal gyrus , also in B-perfusion we can identify the cold area of low grade glioma

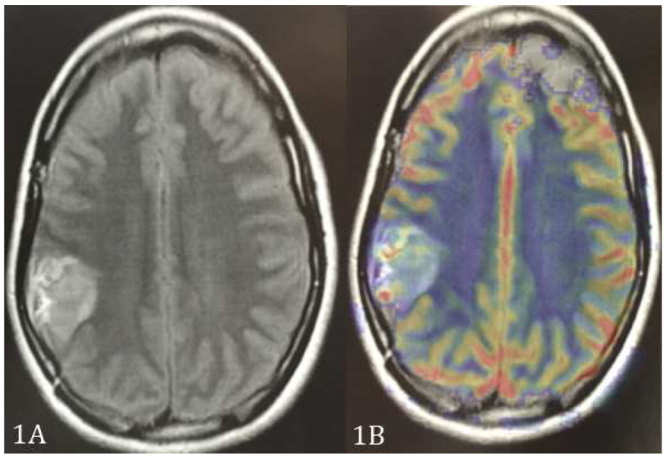
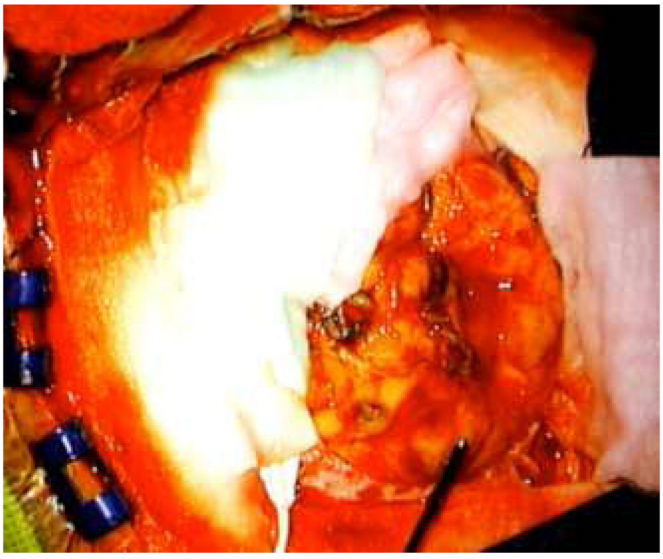


Figure 2- intraoperative cortical stimulation



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1527
WFN15-1769
Late Breaking Posters 1
Why a normal intraoperative motor evoked potential of temporal glioma surgery be paradoxal comparing to post operative?

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The neurophysiological motor monitoring for left temporal cortical low grade gliomas in awake surgery and non awake show disappointing results in 3 % of the cases with permanent and severe disabilities. A problem that brings to us an important question is when the intraoperative motor potential is showing no motor impairment, and just after the patient the patient became contralateral hemiplegic to operated side. It is a big frustration for all team and for the patient beyond any doubt. The physiopathology of this complications has to be taken in concern and discussed based on literature and own experience. Many hypothesis can be thought including late vascular obstruction, basal edema of inferior portion of central core, lesion of inferior frontal occipital fibers, and wrong interpretation of recorded waves. Details regarding resection of temporal lobe, anatomical landmarks of central core, and the main motor cognitive syndromes will be discussed facing an illustrative report case.

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1528
WFN15-1770
Late Breaking Posters 1
Electric sleep status: clinical and electroencephalographic description

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Introduction: Electrical status epilepticus of Sleep (ESES) is characterized by a continuous spike-wave activity during the slow sleep. This pattern is described associated with Landau Kleffner Syndrome (SLK), Epilepsy with Continuous Wave Sleep Spike Slow (EPOCSL) evolutions and atypical Rolandic epilepsies (ER-EA). Cognitive impairment, behavioral and / or motor is part of the ESES.

Objective: Describe the clinical spectrum, etiology and management of 13 children with ESES.

Methods: Review of clinical data and EEG / video-EEG of patients with ESES encountered at the HCSBA Neuropsychiatry Service, from 2005 to 2013. We considered ESES if the continued spike-wave activity was seen in at least 85% of the slow sleep.

Results: 13 patients were analyzed. The mean age of seizure onset was 5 years and 6 months, the mean age of ESES onset was 6 years and 6 month. Related epileptic syndromes were ER-EA (8), EPOCSL (3) and likely SLK (2). With the onset of ESES 7 patients presented change of seizures pattern and all showed cognitive impairment and / or behavioral. 5 patients responded to treatment. Showed efficacy: the addition of benzodiazepines (2), clobazam, levetiracetam association (1), valproic (1) and prednisone(1). The duration of the ESES was variable (1 month-7 years).

Conclusion: ESES can be seen in various epileptic syndromes. We observed a change in the seizures pattern and behavioral cognitive impairment when ESES started. We observed response to most of treatments, however the deterioration was kept in most of the cases. The persistence of the impairment is associated with duration of ESES.

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1529
WFN15-1774
Late Breaking Posters 1
Eslicarbazepine acetate as add-on therapy for partial seizures in children: an integrated evaluation

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Eslicarbazepine acetate (ESL) is approved as adjunctive therapy for partial-onset-seizures (POS) in adults. Given its efficacy, good tolerability and the convenience of once-daily dosing, ESL could be beneficial for children.

Methods: A meta-analysis was conducted for absolute and relative change in standardized-(/4weeks)-seizure-frequency (SF) from double-blind, placebo-controlled studies (harmonized population >6years). A POPPK model was developed (2-18years) as one-compartment model parameterized for absorption, renal-clearance and volume-of-distribution. Influence of age was considered as a structural-covariate. Finally, an integrated evaluation was piloted from available data.

Results: In meta-analysis, absolute or SF relative-reduction was higher for ESL. Confidence-intervals (95%CI) were >0 indicating that ESL reduces significantly SF in children >6years (95%CI: -7.21,-0.25,p=0.036; -32.98,-1.21,p=0.035; absolute and relative, respectively). From POPPK, plasma-exposure for 7-11 and 12-18years was similar but 2-6years presented relevant lower values (Figure_1A). Based on simulated-PK, a minimum dose of 27.5mg/kg/day is necessary for 2-6years to achieve the 20mg/kg/day (>6years) exposure (Figure_1B). By integrating children and adults data, when SF relative-reduction is plotted against ESL-dose or exposure, no dose- and exposure-dependency is observed due to children magnitude of response (Figure_2A). Considering that ~1/3 subjects aged 2-6years received ESL below 27.5mg/kg/day (ITT) and the high variability due to small sample-size, when adjustments are made, SF relative-reduction improves (Figure_2B) and a dose- and exposure-dependency is observed (Figure_2C).

Conclusion: In children with POS aged >2years, adjunctive ESL is considered effective. Despite absence of a significant difference to placebo in the pivotal study, by increasing the sample-size and dose-optimization, similar relationship between exposure and SF is comparable between children and adults. Therefore, >6years the expected therapeutic doses are 20 and 30mg/kg/day, up to 1200mg/day. Between 2-6years, the expected therapeutic doses are 27.5 and 40mg/kg/day, up to 1200mg/day.

Figure 1. Mean (+), median, and range of eslicarbazepine PK parameters using ESL reference dose of 20 mg/kg/day (**A**) and, median and range of dose finding PK scenarios by fixing the ESL reference dose of 20 mg/kg/day for above 6 years (**B**).

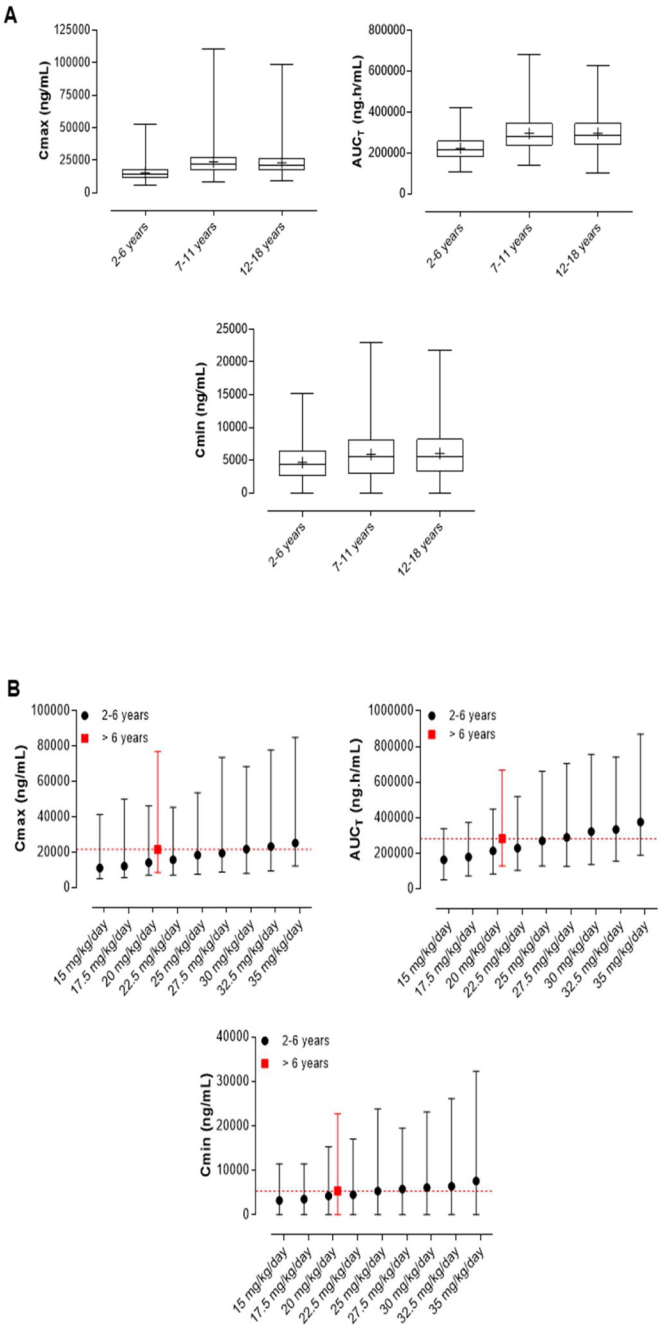
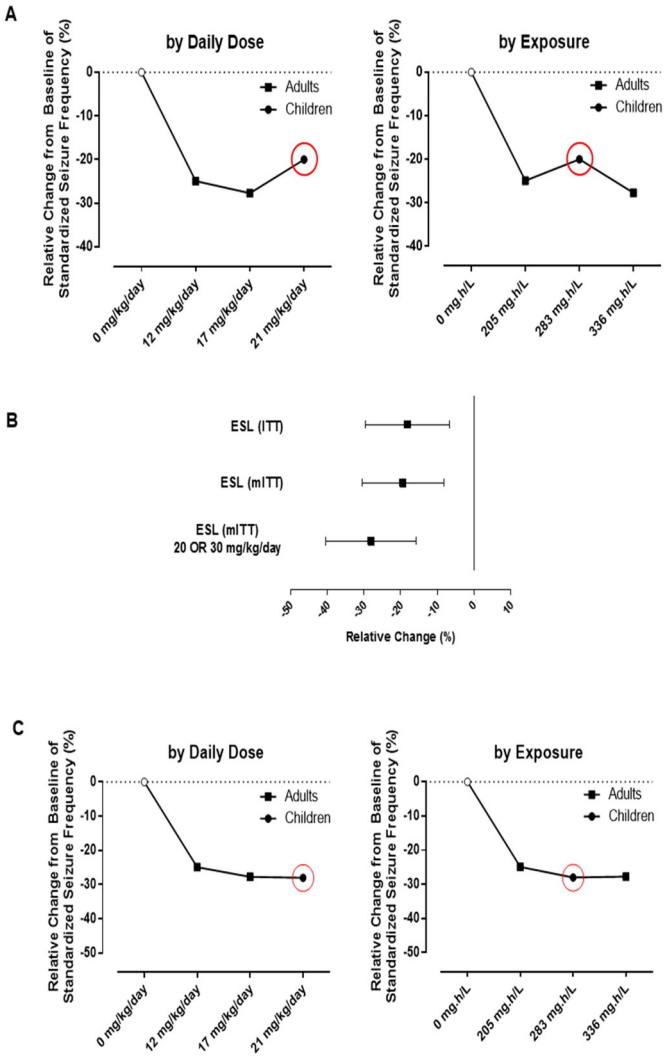


Figure 2. Exposure-effect relationship in adults and children aged 2 to 18 years (**A**, ITT set; **C**, mITT set who received 20 or 30 mg/kg/day) and, effect on ESL efficacy by increasing sample size (from ITT to mITT set) and optimising dose (only 20 or 30 mg/kg/day considered) (**B**).



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1530
WFN15-1628
Late Breaking Posters 2
Ace gene polymorphism and of MTHFR parameters effects on migraine

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ABSTRACT:

Background: Recently; there have been some proofs between angiotensin converting enzyme (ACE), methylenetetrahydrofolatereductase (MTHFR) gene mutation and migraine. The aim of this study; to compare between ACE, MTHFR with migraine control groups and to reveal its connection.

Material and methods: This study was managed between September 2012 and January 2013 in the department of neurology at Dicle University Medical Hospital. This study consisted of 219 people those 119 (mean age 39,4-/+ 10,8) were migraine patient and the rest 100 (mean age 35,5-/+11,6) were controls group. The polymerase chain reaction (PCR) was used for the evaluation of ACE genotype, but the real time PCR technique was used.

Results: While in the patient group DD/ID/II genotype frequency is %33.0, %61.5,%5.2, control group's is %53.6, %43.6, %2.6. It has been statistically found out that the ID genotype in the patient group was highly meaningful compared with control groups. No risk difference has been witnessed both with aura and without aurain migraine group compared to ACE gene DD/ID/II polymorphixm and MTHFR mutations.

Conclusion: In this study the ID genotype in the migraine (with aura and without aura) has been frequent. No correlation has been detected between both aura and without aura compared to ACE gene DD/ID/II polymorfizsm and MTHFR mutations. It hasn' t caused a meaningful increase in the D allele in ACE gene and T mutation in MTHFR in our study population.

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1532
WFN15-1640
Late Breaking Posters 2
Intrathecal baclofen therapy in a patient with hallervorden - Spatz disease

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Background: Hallervorden - Spatz disease, is a rare neurological disorder due to abnormal iron metabolism in the brain and iron deposition in the globus pallidus and the pars reticulata of the substantia nigra. It is characterized by dystonia, rigidity, intellectual impairment and optic atrophy. The onset is during the first two decades of life and the course of the disease usually proceeds over 10 years.

Objectives: We report a rare case of a patient with Hallervorden – Spatz disease treated with intrathecal baclofen (ITB) pump implantation.

Material & methods: A 26 – year – old woman whose illness spanned seven years was presented with a significant hypertonia of all four limbs, more apparent in lower extremities. Dystonia, speech disturbance, vision problems and mild cognitive disorders were also present. A previous effort for rehabilitation combined with botulinum toxin injections failed. The patient was referred for consideration of ITB therapy and after a successful trial test was found to be an appropriate candidate.

Results: A programmable ITB infusion pump was implanted, and in next days, spasticity and dystonia were significantly reduced. Titration dose of baclofen was programmed at 120 μg/day. ITB therapy resulted in a progressive improvement in patient's activity and quality of life. After two weeks she could be sent for further rehabilitation.

Conclusion: ITB pump is an effective and safe treatment option for intractable spasticity in Hallervorden – Spatz syndrome. Rehabilitation is meaningful even in the most affected cases. Short life expectancy and low intelligent quotient should not be a contraindication for rehabilitation.

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1533
WFN15-1641
Late Breaking Posters 2
Spasticity – related pain in amyotrophic lateral sclerosis treated with intrathecal baclofen therapy

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Background: Baclofen, an agonist of $GABA_B$ receptors binds to a number of spinal and cerebral sites and depresses the excitability of motor neurons. Intrathecal baclofen (ITB) infusion is a widely well accepted therapy for the treatment of severe spinal and cerebral origin spasticity. ITB therapy may also provide adequate pain relief in patients with pain associated with spasms and spasticity not controlled with medication and physical therapy.

Objectives: We report our experience of a small group of amyotrophic lateral sclerosis (ALS) patients with intractable spasticity and pain.

Material & methods: From 2008 to 2011, three patients (mean age 41.8 years; 2 men, 1 woman) with ALS underwent an ITB pump implantation under local anesthesia for pain associated with intractable spasticity at our department. Patients' selection criteria were severe and intractable spasticity associated with ALS, patients who exhibited slower disease progression and longer expected duration of life, and positive response to a previous ITB test (50 μcg baclofen applied intrathecally).

Results: All patients reported reduction of preoperative pain score. Spasticity was also significantly reduced. There were no major postoperative complications. All patients survived for more than 12 months following implantation.

Conclusion: ITB pump is an effective and safe treatment option for intractable spasticity and pain. It might be worthwhile, in selected ALS patients suffering from severe spasticity and pain, to consider early ITB administration.

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1534
WFN15-1645
Late Breaking Posters 2
Endovascular treatment of spinal dural fistula

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Background: Spinal dural arteriovenous fistula is the most common cause of spinal vascular disease, constituting on average 70 to 80 % of all spinal vascular malformations. Diagnosis is made of late, with a form of relatively nonspecific presentation, for which a high level of suspicion is required.

Objective: Presenting the spinal dural arteriovenous fistula is a treatable cause with the endovascular approach in our center.

Patients and methods: For a man of 17 years, with 1 year episodes of acute urinary retention and gait disturbance occurs. Evaluated by the urology area where urodynamic studies were performed and results consistent neurogenic bladder, medicated with Tamsulosin no improvement. Referred to the neurology area where proximal and distal paraparesis, hyperreflexia, plantar cutaneous reflex present with urinary sphincter and rectal disorder, without sensory level is found. The MRI showed signal voids causing thoracolumbar myelopathy and spinal angiography showed a malformation type perimedullary arteriovenous fistula supplied by posterior spinal artery that originated with the Adamkiewicz artery from the left segmental artery T11.

Results: The fistula was successfully eradicated by transarterial embolization with NBCA cyanoacrylate. The patient experienced progressive improvement, is without motor disorders, hyperreflexia persists, continue with urinary disorder, i with rehabilitation.

Conclusion: Spinal dural arteriovenous fistula should be considered in the differential diagnosis of progressive myelopathy in young patients where the endovascular approach is the treatment of choice for spinal AVMs in centers with experience.

Keywords: Fistulas spinal dural arteriovenous malformation, endovascular therapy, myelopathy.

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1535
WFN15-1650
Late Breaking Posters 2
Restless legs syndrome and spastic paraplegia in a Brazilian family with SPG4 mutation

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Background: Restless Legs Syndrome (RLS) is characterized by unpleasant sensation in legs and urge to move limbs during rest. Primary forms are usually familiar and secondary forms can be temporarily restricted or associated with other conditions. Hereditary spastic paraparesis (SPG) are a neurodegenerative disorders group characterized by progressive spasticity and legs weakness. SPG type 4 (SPG4) is the most common subtype.

Objective: Report a family case with mutation in SPG4 presented with RLS and SPG.

Patients and methods: Reviewed data from patients and families followed at our Hospital.

Results: 66 year-old male admitted at age 57 with weakness complaints in legs 15 years ago. Onset was insidious, progressive, reported paresthesia and discomfort in legs in late afternoon. Medical examination: spastic paraparesis. Pedigree: Figure1 and Table1. Laboratory tests, brain/spinal cord MRI and ENMG: normal. DNA test: heterozygous mutation missense c.1442T>C in SPG4.

Conclusion: There is no clear etiological link between RLS and SPG4. Increased prevalence of RLS symptoms in patients with SPG was described. Only one Italian family has this mutation and this does not present RLS. In our patients, the symptoms of RLS preceded onset of paraparesis; this may be casual or suggest that RLS is part of a clinical phenotype spectrum of SPG4. It is the first description of a family with mutation in SPG4 with RLS.

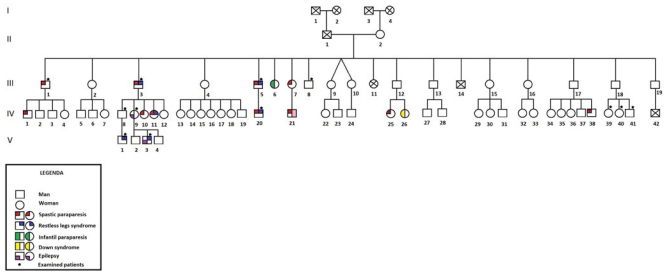

Table1

Patient	Sex	Age	Parentage	Symptoms	Ashworth	IRLSS	Ferritin (28-397)	Medications	Comorbidities
III5	M	66	Index case	SPG/ RLS	4	29	326	-	Hypertension/ smoking
III1	M	71	Brother	SPG	-	-	398	-	-
III3	M	69	Brother	SPG/ RLS	4	37	461	Pramipexol	-
III8	M	62	Brother	Leg´s pain	-	-	44	-	-
IV20	M	27	Son	SPG/ RLS	1	21	72,6	-	-
V1	M	1,5	Nephew	RLS?	-	-	-	-	-
V3	M	19	Nephew	Restless hands	-	21	52,2	Fenitoin	Epilepsy

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1536
WFN15-1651
Late Breaking Posters 2
Use of antiepileptic drugs combined with antiretroviral drugs in patients with aids in an infectious diseases center in Brazil

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Background: AIDS and epilepsy are prevalent problems in developing countries. Antiepileptic drugs (AEDs) are used in the treatment of epilepsy and other neurological conditions. The interaction between antiretroviral drugs (ARVs) and AEDs can have a negative impact on HIV control and there are few studies in developing countries.

Objective: To describe the use of AEDs combined with ARVs for epilepsy and other neurological conditions in patients with AIDS.

Patients and methods: Descriptive analysis of 356 patients from 2010 to 2014, from IPEC/Fiocruz/ Brazil.

Results: Of the 356 patients analyzed there were 165 patients using AEDs for epilepsy prophylaxis, followed by neuropathic pain (59), peripheral neuropathy (72), headache (35) and psychiatric disorder (48). The analysis of anticonvulsants use in the general group (592 samples) / epilepsy group (165 patients) revealed: valproic acid (45%/73%), followed by gabapentin (29%/5%), phenytoin (16%/11%) carbamazepine (5%/5%) and phenobarbital (5%/6%). Most epilepsy cases were due to opportunistic infections. The most prescribed ARVs were lamivudine, efavirenz, tenofovir and others. Valproic acid was the most widely used oral drug and it was last used by patients who switched drugs.

Conclusion: The study was realized in a infectious diseases center, what explains the high number of patients taking valproic acid. Most modern and safe drugs are not available in other Brazilian centers or other developing countries. More detailed studies on interactions between ARVs and AEDs need to be made in developing countries.

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1537
WFN15-1653
Late Breaking Posters 2
Reference parameters of facial nerve stimulation using nerve conduction study among sudanese population

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Background: Facial Nerve Stimulation Study is regarded as a valuable tool for assessing facial nerve function.

Objectives: The study was meant to set reference physiological parameters of facial nerve stimulation [latency, amplitude, total duration and unit area of the compound muscle action potential (CMAP)] in Sudanese population. The study also aimed at looking into the correlation between the parameters of both the right and left facial nerves and assessing the best parameter for comparison.

Methods: This Prospective cross sectional departmental based study was conducted at the Department of Physiology, Faculty of Medicine, University of Khartoum. Sixty normal healthy individuals(35 females and 25 males)were included in the study, with mean age (34.1± 15.98,range *17-65*years). Bilateral stimulation of the facial nerve anterior to the ear lobe and recording CMAP from the nasalis muscles were done using Digital Medelec Synergy EMG (Electromyography) machine. Ethical clearance was obtained from the Ethical Committee at the Faculty of Medicine, University of Khartoum. Subjects' informed consent was obtained.

Results: Analysis of data yielded mean values(±SD) and *ranges* for latency, amplitude, unit duration and unit area as follows:[2.87± 0.56ms(*2.31-3.43*),2.74±1.12mv(*1.63-3.87*),11.45±14.67ms(*3.22-27.12*) and 12.48±16.65mv/ms(*4.17-29.13*) respectively]. The mean distance between the stimulating and recording electrodes was found to be 8.21±0.65cm(7.56–8.86). The highest significant correlation between the parameters was found to be for the amplitude (P=0.000).

Conclusion: The parameters of facial nerve stimulation in Sudanese population were comparable to those of other populations. It was suggested that the amplitude is the best parameter for comparison between the right and left facial nerves in cases of facial nerve dysfunction.

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1539
WFN15-1665
Late Breaking Posters 2
Selection of a neuropsychological battery for vascular cognitive impairment-no dementia in China

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Background: Our goal was to find an optimal combination of neuropsychological tests for detecting VCI-ND in a Chinese population.

Methods: Between April 2012 and March 2013, we compared performance on the NINDS-CSN-VCIHS test battery between fifty-six people with VCI-ND and fifty control individuals. We used the receiver operating characteristic (ROC) curve to compare tests normally found in the NINDS-CSN-VCIHS with additional neuropsychological tests commonly used in China with respect to their ability to detect individuals with VCI-ND. The tests encompassed cognitive screening (MMSE), memory (AVLT), executive function/attention (STT, SCWT, RME, DOT-A, SDMT), language (ANT, BNT, FFT, ST, VFT), and visuospatial function (CFT).

Results: Individuals with VCI-ND performed significantly worse than those in the control group in terms of MMSE scores, memory, executive function/attention, and language ($P < 0.05$). The tests with the largest predictive power with respect to VCI-ND (area under the curve) were the AVLT-LR memory test (0.72), the STT-B test of executive function (0.87), the SDMT test of attention (0.90), the BNT language test (0.75), and the CFT-copy test of visuospatial function (0.56).

Conclusions: We identified several neuropsychological tests that are suitable for assessment of VCI-ND in a Chinese population. We found that individuals with VCI-ND patients had poor memory, executive function/attention, and language ability compared with people in the control group. We believe that a test battery including the AVLT-LR, STT-B, SDMT, and BNT would be optimal for detection of VCI-ND in a Chinese population.

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1540
WFN15-1673
Late Breaking Posters 2
New mutation in the CPOX gene associated with hereditary coproporphyria

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Background: Hereditary coproporphyria is an acute hepatic porphyria characterized by the occurrence of neurovisceral manifestations. This rare metabolic disorder is due to deficient activity of the mitochondrial enzyme coproporphyrinogen oxidase (CPOX) and it's usually inherited in an autosomal dominant pattern.

Objective: We report a case of hereditary coproporphyria caused by an undiscovered mutation in the CPOX gene.

Patients and methods: A 38-year-old woman with a medical history of anxiety-depressive disorder and bariatric surgery in 2003. She has no known family history. She consulted the Emergency Department for intense abdominal pain. Exploratory laparotomy was performed without pathological findings. Physical examination revealed an ascending pattern of progressive paralysis which prevented her from walking and even standing, as well as urinary and fecal incontinence. There were no signs of skin lesions. We performed full blood and CSF analysis (including plasma porphyrins and serologies), a brain and spinal cord MRI scan and an echocardiogram, which revealed no abnormalities. EMG showed a sensorimotor axonal polyneuropathy. Urinary ALA, PBG and coproporphyrin III were elevated, as well as fecal coproporphyrin III. The motor neuropathy progressed to a respiratory muscle weakness, so the patient was transferred to the ICU. The patient's condition improved after treatment with intravenous hemin was started.

Results: Genetic testing revealed a heterozygous mutation results in a valine to methionine substitution at codon 205 in the CPOX gene (c.613G>A (p-Val205Met)).

Conclusion: This never-before-reported mutation leads to a change in the amino acid sequence of the protein, therefore alters the protein's features and compromises its function.

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1541
WFN15-1674
Late Breaking Posters 2
Application of progressive aphasia severity scale (pass) in patients with primary progressive aphasia

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Background: Primary progressive aphasia (PPA) is due to a neurodegenerative process affecting the language areas of the dominant cerebral hemisphere, which is manifested by an insidious and progressive deterioration of language skills and eventually leads to dementia. The PPA is classified in four different variants: agrammatic (Gv), semantic (Sv), logopenic (Lv) and lexical (Lexv).

Objective: To describe the verbal performance of the patients with PPA on the Progressive Aphasia Severity Scale (PASS).

Materials and methods: The PASS was applied to 58 patients diagnosed with PPA based on current diagnostic criteria and after a thorough neurological and clinical examination. Each patient underwent a comprehensive language evaluation and non-verbal and neurocognitive testing. All participants conducted brain imaging scanning.

Results: We observed that SvPPA cases were impaired in word retrieval, single word comprehension, and their writing and reading demonstrated some level of deterioration suggesting semantics involvement. In the LvAPP, the most affected linguistic function was word retrieval, associated with impaired repetition. This group also showed some level of deterioration in single word comprehension, reading and writing. GvPPA demonstrated articulatory errors, impaired syntax and grammar in spontaneous speech, and evidence of impairment in word retrieval, repetition, writing, all of which impacted the functional communication. LexvPPA was the group that showed mainly word retrieval deficits.

Conclusion: PASS is a clinical instrument to assess the severity of the different domains of speech and language. It can be used to determine the differential diagnosis and monitoring the change overtime of the different variants.

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1542
WFN15-1676
Late Breaking Posters 2
A case of progressive apraxia of speech and progressive agraphia apraxic

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Background: Progressive Apraxia of Speech (PAOS) is an insidious condition characterized by a gradual deterioration of speech caused by neurodegenerative disease focused on premotor cortex, Broca and Insula. The most common pathologies associated are corticobasal degeneration (CBD) and progressive supranuclear palsy (PSP).

Progressive Agraphia Apraxic (PAA) is a disorder is less described entity, which is manifested by a continued deterioration of the production of graphemes.

Objective: To describe a case presenting with both syndromes.

Patients and methods: A right-handed 46 year old woman with, higher education. She started with apraxic disabilities characterized by writing difficulties, followed by alterations in speech characterized by difficulty in initiating, distortion,articulation breaks, syllabic segmentation, slowly and foreign accent syndrome; without compromise of language, consistent with PAOS. Neurologic examination showed unilateral parkinsonism in the right hand. Trans cranial sonography showed echogenicity of the substantia nigra suggesting an atypical parkinsonian syndrome. MRI showed diffuse cortical atrophy, affecting especially the left hemisphere perisylvian areas.

Conclusion: PAOS and PAA are disorders that can be presented isolated but also as part of other cognitive syndromes. It has been suggested that PAOS represents a different entity from APP. However, our case suggests that both entities can co-exist and eventually evolve to the agrammatic variant of primary progressive aphasia. The patient is currently being treated based on compensatory strategies.

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1543
WFN15-1677
Late Breaking Posters 2
Clinical characterisation of primary progressive aphasia cases using western aphasia battery (WAB-R)

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Background: Primary progressive aphasia (PPA), a neurodegenerative process affecting the language areas of the dominant cerebral hemisphere, is manifested by an insidious and progressive deterioration of language skills, which eventually leads to dementia. It is classified in four different variants: agrammatic (vG), semantic (vS), logopenic (vL) and lexical (vLex). Western Aphasia Battery – Revised (WAB-R) is widely used battery, mainly used for aphasia caused by stroke and limited information is available on its application on other settings.

Objective: To describe PPA variants based on deficits detected with WAB-R.

Materials and methods: The WAB-R was applied to 58 patients with a diagnosis of PPA after a complete neurological examination and brain imaging studies.

Results: The SvPPA showeds impaired comprehension and severe anomia, but preserved repetition. LvPPA presented with language partially fluent, severe impairment in repetition, and moderate deficits in comprehension and naming. GvPPA exhibited non-fluent language output, associated with impaired repetition and naming. LexvPPA showed a fluent language and mildly impaired comprehension and repetition. The naming was the most affected function.

Conclusion: The WAB-R is a suitable tool to determine the clinical characteristics of the different variants of PPA. It allows establishing the characteristic profiles of each variant, mirroring the classic classification of aphasias. Accordingly, SvPPA is concordant with transcortical sensory aphasia, LvPPA with conduction aphasia,the GvPPA with Broca's aphasia and LexvPPA with anomic aphasia.

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1544
WFN15-1686
Late Breaking Posters 2
Rhombencephalitis related to listeria infection in a myasthenic patient

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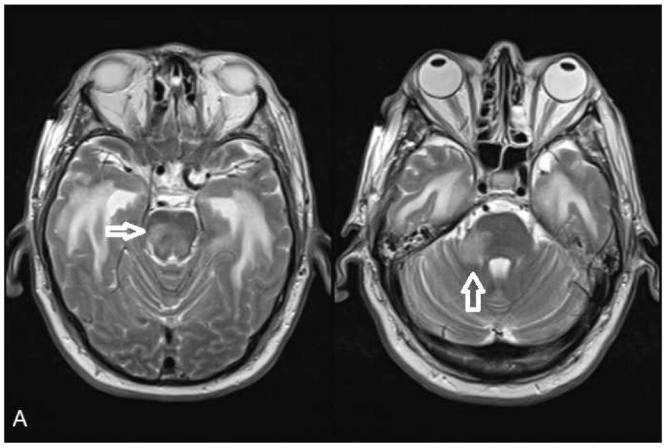
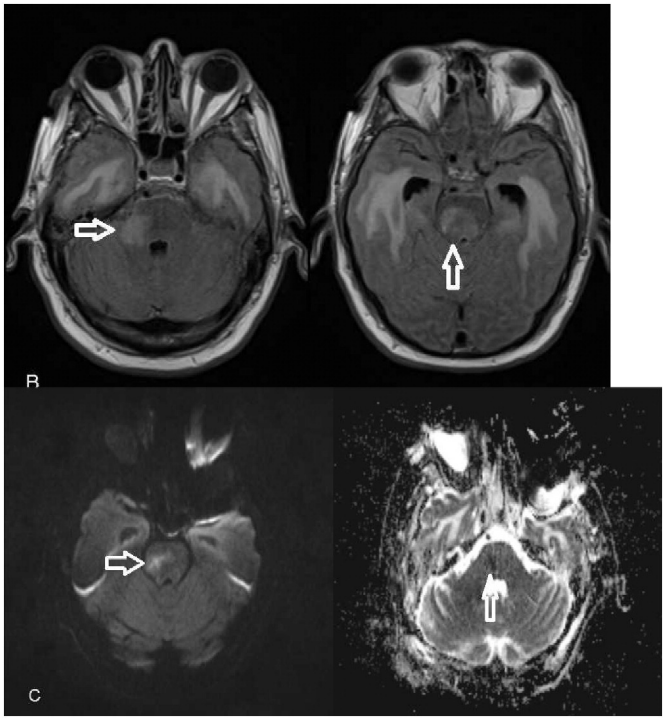
Background: *Listeria monocytogenes* infection in CNS most frequently occurs in immunocompromised and older adults. The most common manifestation in CNS is meningoencephalitis and progress to brain abscess and rhombencephalitis are less common.

Objective: A case-report of listeria infection rhombencephalitis.

Methods and results: A 75-year-old man, with *myasthenia gravis*, chronically using Azathioprine and Corticosteroid, hospitalized for myasthenic outbreak secondary to limb erysipelas, progressing with unfavorable clinical course requiring tracheal intubation, even after infusion of human immunoglobulin. During the extubation, the neurological exam showed brainstem signs. Head CT was performed and showed communicating dilation of the ventricular system. Submitted to external ventricular drainage, which intraoperative CSF showed 05 cells lymphocytic pattern, protein in 96mg/dl, glucose 64mg/dl. Subsequent brain MRI showed hyperintense lesion on T2 and FLAIR with tenuous restriction of diffusion of water molecules in cerebellar peduncle and right lateral portion of the pons (panels A,B,C). CSF analisys with positive PCR for Listeria guide the treatment.





Conclusions: Initiated treatment with Ampicillin in recommended dose, nonetheless with poor clinical response. It evolved with adjacent infectious and clinical complications, occurring irreversible brain injury, and it is currently in a persistent vegetative state.

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1545
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Late Breaking Posters 2
N200 non target responses in cognitive decline

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In p300 event related potentials (ERP) the target stimulus and non target stimulus generate two responses with similar configuration but dissimilar latencies in N2 and P3 waves.

It is known that In cognitive decline the N2 latency of target response is longer than in normal subjects.

It is also known that the N2 latency is shorter in target response than in non target response.

It is our objective to estimate: 1) If the N2 latency of non target response is also affected in cognitive decline and 2) If the difference of N2 latency between target and not target is modified by the cognitive decline; for this purpose we studied 81 subjects with cognitive decline and 81 subjects with normal cognition matched by age and instruction level.

Results: N2 latency (target) 233,11 ms – 284,25 ms (p=0,001), N2 latency (non target) 279,88 ms – 280,87 ms (p=0,8775) N2 latency difference 46,77 ms - -3,69 ms (p=0,001)

Conclusion: in this population the cognitive decline only affects the N2 latency in the answer target, taking place therefore a modification of the difference between the latencies of N2 target versus N2 not target, which tends to be annulled.

It could be assumed that the facilitatory mechanisms that allow a quicker processing when the stimulus agrees with the subject expectation are lost in cognitive deterioration.

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1546
WFN15-1688
Late Breaking Posters 2
When celiac disease touches neurons: a report of four patients

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Background: Celiac disease (CD) is an auto-immune disorder characterized by gluten hypersensitivity and presence of anti-transglutaminase and anti-gliadin antibodies. Clinical manifestations, mainly digestive, are various but neurological involvement is rare.

Aim of the study: To describe clinical, para clinical and outcome parameters in patients presenting neurological manifestations of CD.

Patients and methods: A retrospective study, including patients followed for CD with neurological involvement at the neurology department in Sfax, Tunisia, since its creation in 1991.

Results: Three women and a man were included in this study. Mean age of neurological symptoms onset was 20 years (7 to 36 years). Neurological manifestations were inaugural in one case. One patient presented peripheral nerve damage and central nervous system (CNS) was found in 3 patients (cerebellar ataxia, seizures and cerebral venous thrombosis). All patients received a gluten-free diet with a favourable outcome.

Discussion / Conclusion: Neurological complications of CD are but various. They may be revealing a CD in adults. They affect either central or peripheral nervous system. Peripheral neuropathy and white matter abnormalities are the most common paraclinic findings. The use of brain imaging or electromyography, guided by clinical assessment, is highly recommended in patients with CD, as well as a strict follow up and observation of a gluten-free diet.

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1547
WFN15-1691
Late Breaking Posters 2
Examination of the influence factor in cognitive impairment in Parkinson's disease

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Background: In late years, cognitive function is important issue in Parkinson's diseases (PD). Therefore we focused on a detailed evaluating cognitive function in PD by Wechsler Adult Intelligence Scale, 3rd Edition (WAIS-III). We have special result in their issue.

Objective: We investigate that influence factors in cognitive impairment of PD according WAIS-III.

Patients and methods: We examined 162 cases (68.9±9.2 years old) of PD patients using WAIS-III.

Result: I performed a multiple regression analysis as a purpose variable at each sub test scores, as an explanation variable at disease severity (Yahr), age, onset age, and duration of illness. As a result, scores of "Similarities" "Digit Symbol-Coding " "Block Design" decreased as duration of illness became long. Score of "Comprehension" "Arithmetic" "Block Design" "Letter-number sequencing" declines as age is higher. Score of "Picture completion" "Block Design" declines as the Yahr is higher.The other, age of onset is no influence factor for each sub tests.

Conclusion: As a result of multiple regression analysis, it is clear that age, duration of illness, and Yahr are declined score of sub test in WAIS-III. Because score of "Block Design" is related with each factors; age, onset age, and Yahr, it is considered plural factors cause visual-perceptual disabilities in PD. Aging, prolongation of duration of disease, and aggravation of PD are factors to cause a decline of sociality in PD. Therefore it suggested the possibility that intervention to be maintained sociality to maintain cognitive function of PD was valuable.

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1549
WFN15-1695
Late Breaking Posters 2
Forecasting for the diagnosis of carpal tunnel syndrome

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Carpal tunnel syndrome (CTS) is a clinical syndrome of numbness, weakness, and pain in the fingers and wrist, associated with median nerve compression at the wrist. CTS is basically diagnosed on clinical examinations and electromyography (EMG). Patients often have Tinel's sign, Phalen's sign and positive provocative tests. The conventional standard EMG for CTS requires demonstration abnormal median nerve conduction tests across the wrist. Many patients still have negative EMG.

Purpose: The aim of this study is to assess whether artificial compression tests will improve the diagnostic rate of carpal tunnel syndrome (CTS).

Methods: We recruited 36 hands of 18 CTS patients, and 40 hands of 20 controls. We performed artificial compression test in the remaining symptomatic hands and the non-CTS hands.

Results: To forecast the development of carpal tunnel syndrome enough to use two indicators electromyography. Latency of motor fibers and a sensitive response amplitude during the test artificial compression provides 100% sensitivity and specificity.

Conclusion: For CTS patients with normal results from the standard methods, artificial compression test is a good additional comparative test to further improve diagnostic rate. Our study founded that one more simple test can contribute significantly to the diagnosed rate of CTS.

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1550
WFN15-1701
Late Breaking Posters 2
Stroke patterns in rural south India 3 years data from a rural stroke centre

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Objective: Our Aim was to study the incidence of stroke among general neurological patients in rural population, To identify the stroke subtypes, prevalent risk factors, the number of patients arriving within the time window and the number of patients eligible for thrombolysis.

Methods: In this retrospective study, data wereobtained from our hospital registry from 2011-2014.

Results: The total number of patients admitted in our hospital from 2011-2014 was 5715. Among them Stroke patients were 2577. Ischemic strokes were 1728, Male 1164, Female 564,Age range(2-91yrs),Risk factors were DM in 414,SHT in 324,Dyslipidemia in 327,RHD in 72, Tobaccoand alcohol users in 696, 225(13%)patients had come within the time window and 126 of them(7.5%) thrombolysed successfully.

ICH were 483, pure parenchymal ICH in 363, ICH with IVH in 87,Pure IVH in 33,Age range(14-80yrs), Risk factor were SHT in 300, DM in 75. CVST were 228, Risk factors were post partum in 36, Alcoholism in 93, Fever with dehydration in 60, OCP intake in 15 and CNS infection in 6 patients. SAH were 63, Age range(23-80yrs), aneurysmal bleed in39, SHT in 24. SDH were 75, Age range was (26yrs-80yrs), post traumatic in 69 and 6 patients were anticoagulant induced.

Conclusion: The predominant stroke was Ischemic (67%), Alcohol and tobacco were the major risk factors in the younger age group and low socio-economic status. 13% of the Ischemic stroke patient came within the time window and 7.5% thrombolysed successfully. CVST accounted for9%, alcohol was major risk factor. ICH accounted for 18% and SHT was major risk factor. SAH accounted for 3%, and SDH in 3%.

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1551
WFN15-1702
Late Breaking Posters 2
Recurrent myelitis with normal imagem as an unusual systemic lupus erythematous (SLE) -probable first manifestation

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Background: Medical literature describe Systemic Lupus Erythematous (SLE) and others autoimmune diseases as a cause of myelitis, and it should be considered over diagnostic process.

Objective: report a case of myelitis by probable SLE.

Methods and results: 39-year-old man, with unremarkable previous medical history, was admitted to hospital with a one month of insidious and progressive unsteadiness, gait impairment and vertigo. General medical evaluations was normal. Neurological examination revealed spastic paraparesis with clonus, bilateral Babinski, no sensory abnormality, brisk reflexes at inferior members. Pos spontaneous micturition volume was 300mL. Vestibular and cerebellar proves were normal. Superior members have no abnormalities. MRI scan revealed a slight leptomeningeal enhancement al cerebellum and no abnormalities at spinal cord and nerve roots. Cerebrospinal fluid analysis was no pleocitosys and high protein concentration (210mg/dL). Further evaluation reveled anti-nuclear antibody 1/640, anti-SM 259U/mL and Anti-SCL70 172U/mL. Considering the news diagnostic criteria for SLE 2012 this pacient has three positive criteria. A treatment with 1gm per day endovenous Methylprednisolone was performed with undoubtedly improvement. At the end no clonus and spasticity were noted. Recurrence occurred in the third week post treatment, and a new corticoids cycle was performed. The patient under follow-up taking immunosuppressives.

Conclusion: the formal SLE diagnose was not performed, however the autoimmune etiology was confirmed by the following-up and response to treatment. Autoimmune diseases should be considered on differential diagnosis of myelitis. We obtained patient and Institutional Review Board (IRB) approval.

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1552
WFN15-1703
Late Breaking Posters 2
Different clinical presentations of neurocysticercosis in developing countries

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Cysticercosis is a zoonotic disease caused by parasite, *Taenia solium*, producing cysts in different parts of the body, having predilection for the CNS resulting in neurocysticercosis. Human infection occurs when ingesting cyst in water or food contaminated with feces of animals carrying the parasite (fecal-oral transmission). In endemic regions such as Latin America, Africa and Asia there is a 2-4% incidence in general population.

Most frequent clinical manifestations are: epileptic seizures (50-65%), headaches (40%) and focal deficits.

There are active and silent ways of presentation, the first ones (50-80%) have two different types of cysts: extra parenchymal, racemosus in subarachnoid space or base tanks and intra parenchymal, cellulose where the scolex could be seen.

The objective of this investigation is to present 3 cases with different ways of presentation.

Case 1: male, 65 years, bolivian, admitted for sensory loss and slight right cruris-braquio hemiparesis. A multiseptate cystic supra and infratentorial image in the MRI with ventricular dilatation and cerebral aqueduct compression.

Case 2: male, 22 years, argentinian, debuts with a generalized tonic-clonic seizure.

Case 3: female, 47 years, paraguayan. Admitted for right cruris-braquio hemiparesis,headache, dizziness and dysarthria of one week evolution. MRI: lesions in mesencephalic aqueduct, white substance, right parietal left and mesencephalic cerebral peduncle where the scolex is seen.

Conclusion: Even though neurocysticercosis is a public health problem in developing countries, in Argentina is not so frequent. It is important to consider the extra parenchymal forms to have an early diagnose and treatment for setting a good prognosis.

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1553
WFN15-1713
Late Breaking Posters 2
Guillain-Barre syndrome and the clinical importance of a early diagnosis for a better prognosis

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Background: Guillain-Barré Syndrome (GBS) is an acute demyelinating polyradiculopathy autoimmune inflammatory, characterized by motor dysfunction, sensory and autonomic, probably secondary to immune process against myelin antigens. It usually occurs two to three weeks after viral infection or bacterial nonspecific. It is described as progressive muscle weakness, areflexia and increased protein in the liquor. It is characterized by ascending, progressive and usually symmetrical peripheral involvement, in which the motor manifestations predominate over sensory.

Objective: To report a case of GBS and its outcome. We had obtained patient approval.

Patients and methods: MMO, 25, female, resident of Wanderlândia-TO (Brazil); started in February/2015 with paresthesia and paresis of lower and upper limbs, difficulty walking, suddenly after intestinal infection early. It was referred to the Araguaína Regional Hospital where it evolved with sharp widespread pain, constipation, ascending

progressive motor loss, areflexia, dysphagia and dysphonia. She received noninvasive monitoring and oxygen therapy. It was requested electroneuromyography and began treatment with human immunoglobulin. There was return speech, painful abdominal distension, emesis and anuria, use of nasogastric tube relief with substantial elimination of gastric fluid. Evolved with improvement of movements, swallowing and breathing. Enema was performed to remove feces. Applied second dose of human immunoglobulin, followed by hospital discharge.

Results: We present the case of GBS where treatment with immunoglobulin was started late causing complications.

Conclusion: It is a self-limiting disease, but diagnosis and treatment are mandatory for the best restoration of the patient and reduction of lethality in more severe cases.

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1555
WFN15-1724
Late Breaking Posters 2
Neuroleptic malignant syndrome induced by quetiapine

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Background: Neuroleptic malignant syndrome (NMS) is a life threatening neurologic emergency associated with the use of neuroleptic agents, a class of medications that block dopamine transmission, and characterized by a distinctive clinical syndrome of mental status change, muscular rigidity, Hyperthermia and dysautonomia. The tetrad of NMS symptoms typically evolves over one to three days.

Objective: To describe the case of a 57 year-old patient who developed compatible symptoms with NMS after initiate Quetiapine use, a neuroleptic agent, used in the handling of psychiatric comorbidity.

Case report: A 57-year-old man with Depression and Panic disorder who has developed mental status change, visual and auditory hallucinations, muscular rigidity and autonomic instability, such as tachycardia, 3 days after starting Quetiapine use for psiquiatric disorder. While many consider fever to be an essential feature of the diagnosis, cases are reported where it is absent, as in this case.

Conclusion: There is debate in the literature about milder or atypical cases of NMS, those associated with lower potency agents or those diagnosed early on. From a practical clinical point of view, it seems reasonable to consider the diagnosis when any two of the tetrad of symptoms are present in the setting of an offending agent. However, there are related disorders that must be reminded as differential diagnosis, such as Malignant Hyperthermia, Malignant Catatonia and Serotonin Syndrome.

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1557
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Late Breaking Posters 2
Effects of functional and analytical strength training on upper limb motor and functional recovery after stroke: a randomized clinical trial

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Background: The benefits of strength and functional training in chronic stroke patients have been described separately, while the combination of both on muscle strength and/or motor fuction is incipient.

Objective: To determine the effects of functiona l(FS) and analytical (AS) strengthening in the motor function recovery and muscle strength in patients with chronic hemiparesis after stroke.

Methods: 28 subjects were recruited, stratified for maximum strength of shoulder flexors and randomly assigned to two intervention groups: FS or AS (n=14). FS group performed reaching-to-grasp movements against resistance and the AS group trained upper limb strengthening without functional goal. The protocols were performed 3 times a week along 5 weeks. Primary outcome measure was Upper Extremity Performance Test (TEMPA). Secondary outcome measures included shoulder and grip strength, motor recovery (Fugl Meyer Scale) and muscle tone (modified Ashworth scale).

Result: The FS group showed the largest increase in unilateral tasks analysis (43%; p=0.04) and in unilateral total score (46%; p=0.04) (table 1). Both groups exhibited a similar and significant improvement in hand grip and shoulder flexors strength and motor recovery in the paretic upper limb, without increase on spasticity (table 2).

Conclusion: Both protocols of strength muscle training presented positive results for of the upper limb rehabilitation in patients with chronic hemiparesis post stroke. Therefore, the FS could be a best choice for rehabilitation regarding the movement quality in the paretic side.

Tabel 1. Clinical Primary Measurements

TEMPA	Scores						Change Scores (%)		
	FS (n=13)			AS (n=14)			FS (n=13)	AS (n=14)	
	Pretest	Posttest	P-value[a]	Pretest	Posttest	P-value[a]	Post-pre	Post-pre	P-value[b]
Functional Graduate (FG)									
Unilateral Tasks	-2 (-11/0)	-2 (-11/0)	0.01	-4 (-12/0)	-4 (-12/0)	0.04	36	23	0.68
Bilateral Tasks	0 (-4/0)	0 (-2/0)	0.08	-0,5 (-8/0)	-1 (-8/0)	1	17	7	0.40
Task Analysis (TA)									
Unilateral Tasks	-7 (-43/-1)	-3 (-43/0)	0.002	-11(-48/0)	-3,5(-48/0)	0.06	43	13	**0.04**
Bilateral Tasks	-3 (-21/0)	0 (-22/0)	0.05	-6,5 (-30/0)	-4 (-30/0)	0.04	52	16	0.28
Unilateral Total score	-9 (-54/-2)	-5 (-54/0)	0.002	-15 (-60/0)	-13,5(-60/0)	0.04	46	13,5	**0.04**
Bilateral Total score	-3 (-25/0)	0 (-24/0)	0.03	-7 (-38/0)	-5 (-38/0)	0.04	54	17	0.26
Unilateral and bilateral tasks scores combined	-16(-79/-4)	-3 (-78/0)	0.001	-22 (-38/-4)	-13(-38/-4)	0.001	45	11	0.07

Values are median (min/max) or mean (±SD). Wilcoxon and Mann–Whitney U Tests were used for intra-group and inter-group comparisons, respectively. aWithin-group comparison. bBetween group comparison

Table 2. Clinical Secondary Measurements

	Scores						Change Scores (%)		
	FS (n=13)			AS (n=14)			FS (n=13)	AS (n=14)	
	Pretest	Posttest	P-value[a]	Pretest	Posttest	P-value[a]	Post-pre	Post-pre	P-value[b]
Strength measures in the paretic side									
Shoulder Flexors (kg)	3.6 (0.6/3)	6.7 (2.2/21)	0.001	2.3(0.5/3.4)	6 (2.5/28)	0.001	79	162	0.43
Hand Grip (pounds)	31 (±12)	38 (±14)	0.03	24 (±13)	30(±15)	0.002	17	32	0.32
Fugl-Meyer Scale Motor Function (66)	55 (27/63)	62 (38/66)	0.001	44 (13/60)	53(25/66)	0.001	18	16	0.83
Muscle tone (Modified Ashworth scale 0 - 4)	1 (0-2)	1 (0-2)	0.04	1+ (0/3)	1 (0/3)	0.03	33	24	0.9

Values are median (min/max) or mean (±SD). Paired samples tests and Wilcoxon test were used for intra-group comparisons. For inter-group comparisons, Mann–Whitney U Test was performed. [a]Within-group comparison. [b]Between group comparison

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1558
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Late Breaking Posters 2
Clinically mild encephalitis/encephalopathy with a reversible splenial lesion (MERS) associated with hupoglycemia. Case report and literature review

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Mild encephalitis/encephalopathy with a reversible splenial lesión (MERS), its a syndrome characterized by mild neurological manifestations and the evidence on magnetic resonance imaging (MRI), of a reversible lesion with restricted diffusion and low signal in ADC map, in the central portion of the splenium of the corpus callosum. MERS has been reported to be associated with several conditions including infectious disease and electrolyte disturbance. Nevertheless, the pathogenesis of this syndrome remains unclear. There are few reports about MERS associated with hypoglycemia. We report the case of an adult female patient with hypoglycemia who was diagnosed with MERS. This is the first case report reported in south América. We made a literature review about clinical implication and MRI, electroencephalographic and laboratory findings. We also discuss possible hypothesis of the pathogenesis of MERS.

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1559
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Late Breaking Posters 2
Antiphospholipid syndrome and polymyositis: a rare multiple autoimmune syndrome

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Background: Antiphospholipid syndrome (APS) can be primary, secondary to several conditions or occur simultaneously with another autoimmune condition resulting in a multiple autoimmune syndrome. Its association with Polymyositis (PM) is extremely rare.

Case report: We report the case of 52 y.o man complaining of a rapidly progressive proximal muscle weakness. Laboratory analyses showed increased levels of muscle enzymes, electromyography and muscle biopsy findings led to retain inflammatory myopathy. Based on the hypothesis of PM, patient was put on 1mg/kg/day of prednisone with clinical improvement. One year later, he had lower limb deep venous thrombosis and acute mesenteric ischemia. Etiological investigation revealed increased rate of anticardiolipin IgM confirming the diagnosis of APS. He was put on prednisone and warfarin. The evolution was marked by an episode of arterial stroke 6 years later.

Discussion/Conclusion: Although APS may be associated with a wide range of autoimmune disorders, only few cases, describing the association between APS and PM, were reported. Pathological mechanisms of this association as well as its impact on PM prognosis are still unclear. Treatment should include oral anticoagulants and immunosuppressive agents in order to prevent further vascular event as well as major clinical relapse of any associated autoimmune disorder.

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1560
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Late Breaking Posters 2
Rapidly progressive dementia by neoplasia

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A rigorous routine should always be followed in the investigation of mnemonic deficits, with a careful history, supplemented by neuroimaging and laboratory tests. We bring a case that could exemplify this importance. Male patient, 78, pediatrician, still active in his office, evolving over the past six months with Dysnomia, episodic memory deficit and difficulties in their work routines, such as slowing to remember the name and dose of medicine, referring to two episodes of transient global amnesia short. Complains still anhedonia, anergy and asthenia, but without sadness complaint. History of systemic arterial hypertension under control. General physical and neurological examination without significant changes. Screening test (MOCATEST) normal for their schooling.

Neuropsychological assessment carried out about 15 days after the initial consultation already demonstrated significant deficits in auditory -verbal aspects of memory and anomie, and depressive symptoms.

In the investigation by neuroimaging (MRI) we find a left temporo-insular infiltrative lesion, without contrast enhancement, suggestive of low-grade glioma.

The patient was then referred for cancer treatment.

Although initially demonstrate very suggestive features of cognitive impairment by a depressive disorder, further investigation demonstrated a more serious cause for this commitment.

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1561
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Late Breaking Posters 2
Botox in a pediatric patient with severe intractable migraines with severe opioids and acetaminophen allergies

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12 year old girl that started with hedaches at age six. Diagnosed with migraines with aura and treated with amitriptyline and propranolol as preventive therapy. After age ten, migraines started to be more frequent and stronger. On multiple hospitalizations and trials for breakthrough pain medications she was found allergic to topiramate, acetaminophen, valproic acid, hydromorphone, tramadol, morphine, hydocodone, oxycodone and fentanyl. Nerve block done on supraorbital nerves, auriculotemporal nerves with frontalis and temporalis muscles with lidocaine, helped for breakthrough pain on her last hospitalization. She was schedule after the nerve block for Botox injections on frontalis and temporalis and she gets these every four months consecutively without needing any medication for breakthrough pain.

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1562
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Late Breaking Posters 2
Neurosyphilis: an old and new disease

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The incidence of Syphilis decreased in the late 90s, reaching its lowest levels in 2000. However, cases are increasing annually, suggesting a greater circulation of treponema.

Objective: Describe two cases with different presentations of neurosyphilis.

Case 1: Male, 41-yo, diagnosed with HIV. In April 2014, he presented right-side hemiparesis which he reported to be due to ischemic etiology. Patient didn´t continue attendance. On admission, in May 2015, he kept complete and disproportionate right-side hemiparesis. CT scan of June 2015 showed a left frontoparietal hypodensity lesion in the middle cerebral artery region of ischemic character, without expansive effect or enhancement by contrast. CT brain angiography

and echocardiography didn´t show alterations. Patient had negative treponemic tests in 2009. In 2012, VDRL 1/4 with negative TPHA, without treatment. In 2015, VDRL 1/128 and TPHA reactive. The cerebrospinal fluid showed VDRL 1/32. Patient started treatment with crystalline penicillin. Case 2: Male, 37-yo, without any known comorbidity, admitted in the emergency room complaining of acute blurred and doubled vision of sudden onset. Furthermore, he presented a two-month history of ulcerated genital lesions. On examination: bilateral optical papillitis with no other alterations. HIV non-reactive. Serum VDRL: 1/256,positive blood TPHA and VDRL: ½ in the cerebrospinal fluid, among other modifications. The patient also received crystalline penicillin.

Early diagnosis and treatment of neuro-syphilitic ischemic stroke or ocular manifestations of neurosyphilis are crucial to a favorable prognosis. Syphilis continues to present itself as a silent disease and therefore cases are still not diagnosed and treated properly.

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1564
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Late Breaking Posters 2
Neuropathic pain treatment and quality of life: results in leprosy patients

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Introduction: Leprosy is a chronic infectious disease caused by Mycobacterium leprae. In addition with functional impact and social stigma, it causes neuropathic pain.

Objective: Evaluate the impact of two treatment regimens in health-related quality of life in patients with neuropathic pain due to leprosy presenting active neuritis.

Methods: Randomized pilot study with 10 patients with leprosy neuropathy were included following the WHO criteria, among the age range of 18-75 years and with neuropathic pain for more than 1 month and less than 5 years. Peripheral nerve biopsy was done for histopathological evaluation of the etiology of pain. Patients with active neuritis were randomly divided into two groups. Group 1 (4 patients) received oral corticosteroid up to 1 mg/kg. Group 2 (6 patients) received oral corticosteroid therapy at 1 mg/kg and oral azathioprine at 1 to 3 mg/kg, both for six months. The SF-36 scale was applied at the beginning and end of the study.

Results: Statistical analysis of the results showed that Group 1 had an improvement in comparison to Group 2, proven from the overall mean of all SF36 criteria. Group 1 had an initial mean of 49.87 and final mean of 63.58 (27.49% improvement) while Group 2 had an initial of 55.5 and a final of 73 (31.53% improvement).

Conclusion: This study shows that the correct treatment in leprosy impacts in quality of life basically in the physical domains, more specifically in the relief of pain and highlights the importance of this underestimated symptom in the disease. Besides that, this pilot study identified a superiority of Group 2 compared to Group 1, preliminary results of a research line that is still in progress.

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1565
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Late Breaking Posters 2
99mTc-DPD cardiac scintigraphy in familial amyloidosis

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Introduction: Familial amyloidosis is a group of diseases characterized by tissue deposition of amyloid fibrils. There are three types of familiar amyloidosis: transthyretin (TTR), apolipoprotein A1 and gelsolin. Cardiac involvement is a leading cause of morbidity and mortality; a new mutation strongly related with isolated cardiac amyloidosis is the TTR Val122Ile. The discovery of tests that allow early diagnosis of cardiac involvement in amyloidosis, and inferred that the etiology of the disease is of utmost importance.

Objectives: In a cohort of patients with different types of familiar amyloidosis (TTR and gelsolin), we aimed to assess the role of 99mTc-3,3-diphosphono-1,2-propanodicarboxylic acid (99mTc-DPD) in detecting myocardial amyloid infiltration.

Methods: We enrolled six patients diagnosed with late familiar amyloidosis, which mutations were documented at deoxyribonucleic acid analysis: three patients with TTR Val30Met mutation, two patients with gelsolin mutation and one patient with TTR Val122Ile mutation. Five patients were asymptomatic for cardiac involvement and one patient (Val122Ile mutation) had diagnosis of heart failure. Myocardial uptake of 99mTc-DPD scintigraphy was assessed at five minutes and three hours.

Results: The uptake of 99mTc-DPD highly demonstrated amyloid in cardiac area in two out the three cases of TTR Val30Met and in TTR Val122Ile, and no uptake in gelsolin cases. TTR Val122Ile case presented the highest uptake with exclusive deposition of amyloid in cardiac area resulting in severe heart failure.

Conclusion: In hereditary TTR amyloidosis, including the mutations TTR Val30Met and Val122Ile, 99mTc-DPD cardiac scintigraphy can identify infiltration even in asymptomatic individuals, allowing early diagnosis of cardiac compromise. As expected, there wasn't uptake in patients with the gelsolin mutation. We can consider that this non-invasive test would be a tool for the demonstration of cardiac amyloid deposition.

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1566
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Late Breaking Posters 2
Polysomnographic study in patients with duchenne muscular dystrophy

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Introduction: Duchenne Muscular Dystrophy (DMD) is a progressive neuromuscular disease of X-linked recessive inheritance caused by a defect in the dystrophin gene. Sleep-disordered breathing is common in these children, being ten times more prevalent than general population. The objective of this study is to analyze the polysomnographic characteristics in children with DMD.

Methods: Cross-sectional study of 29 patients with DMD who underwent a polysomnographybetween 2005-2011.

Results: Total of 29 patients, mean age 9 years (2-18). 20/29(69%) with preserved ambulation, of these, 19 being treated with corticosteroids (18 prednisone, 1 deflazacort). 9/29(31%) had lost gait (mean age 13 years), of these, 7 receiving corticosteroids (6 prednisone, 1 deflazacort). 3/29 were users of non-invasive ventilation (NIV). 13/29(45 %) had reduced sleep onset latency, 14/29(48%) decreased sleep efficiency, 8/29(27%) with sleep fragmentation. 13/29(44%) with increased superficial sleep and 11/29(37%) decreased REM sleep. Respiratory disturbance index (RDI) was increased in 18/29(62%), 6/18 showed obstructive apneas and hypopneas, 11/18

central apneas and 2/18 a mixed pattern. Of the 9 patients in wheelchair, 3 had an increased RDI. 2/29 with PCO2>53 at the time of the exam. Snoring was present in 7/29(24%), of which two were associated with obstructive events. Mean baseline oxygen saturation was 97% and desaturation <90% present in 13/29(45%) patients.

Conclusions: The prevalence of sleep-disordered breathing in children with DMD is relevant. In this study the wheelchair-bound patients didn't experience more respiratory events than patients who maintained ambulation. The number of patients receiving NIV was low.

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1567
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Late Breaking Posters 2
Pompe disease: distal myopathy in a previously unreported heterozygous variant

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Introduction: To report a previously unreported heterozygous variant in exon 8 of the GAA gene (**c.1198G>A p.V400I**) associated to a **distal myopathy** presentation.

Background: Pompe disease (PD) is an autosomal recessive, rare disease, caused by a deficiency on lysosomal enzyme alpha-glucosidase (GAA). It causes progressive muscular weakness and may lead to respiratory impairment. Presentation may occur as soon as in newborns or later in children, adolescents or adults. There are genotypic and phenotypic variations on disease. We describe two cases (mother and daughter) with previously unreported mutations and clinical heterogeneity.

Methods: Patient 1 is a 48-year-old female presented with a 5-year disease that started with distal limb weakness, foot dorsiflexion impairment. She has recently started to present dyspnea. Electroneuromyography (EMG) showed normal neuroconduction and an unequivocal distal myopathy pattern. Patient 2, her daughter (23 y.o.) presented dyspnea and no evident weakness at neurological examination; a pericardic effusion was observed at ultrasound examination. Parents are consanguineous. EMG findings suggested a proximal myopathy. Both patients were tested genetically for PD (CENTOGENE®).

Results: Genotype for both patients are presented: Patient 1 - Tandem MS alpha-glucosidase (Pompe) - 1,7 μmol/l/h (reference: > 3 μmol/l/h). **Gene sequencing GAA - heterozygous variant (c.1198G>A p.V400I)**. Tandem MS sphingomyelinase (control) - 1,9 μmol/l/h (reference: ≥ 2 μmol/l/h). Patient 2 - Tandem MS alpha-glucosidase (Pompe) - 2,7 μmol/l/h (reference: > 3 μmol/l/h). Gene sequencing GAA - heterozygous variant (c.1198G>A p.V400I).

Conclusion: Our patients present the phenotypic heterogeneity of PD under the same genotypic variation in the same family, being ours the first distal myopathy case reported. We also detected a previously unreported heterozygous variant in exon 8 of the GAA gene (c.1198G>A p.V400I). PD is a rare, heterogeneous condition that may mimic many neuromuscular conditions.

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1568
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Late Breaking Posters 2
Inflammatory radiculomyelopathy associated with sapho syndrome

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Introduction: SAPHO Syndrome (Synovitis, Acne, Pustulosis, Hyperostosis and Osteitis) includes a variety of osteoarticular disorders associated with skin conditions. It is a rare and often unrecognized disease and its diagnosis is even more difficult when atypical sites are involved and there are no skin lesions. Spinal lesions in SAPHO syndrome are described in 50% of the cases but generally show a good prognosis and rarely cause neurological deterioration.

Objective: To report a case of radiculomyelopathy in a patient with SAPHO Syndrome.

Results: A 73-year-old man referred difficulty to walk 18 months ago, becoming paraplegic in the very first month. At the time, he was admitted in the ICU. He improved over a year but didn`t recover the ability to walk. Neurologic examination showed spastic paraparesis with bilateral Babinski sign, sensory level on D6, which was probably associated with SAPHO (despite the lack of skin lesions, the diagnosis was made with histopathological study of the D6 vertebral body, bone MRI and scintigraphy findings) spinal injury. Laboratory analysis showed an inflammatory reaction: high levels of C-reactive protein and erythrocyte sedimentation rate, even though the leucocyte count was normal. Tests for rheumatoid factor and HLA-B27 antigen were negative, also supporting the diagnosis. There was no evidence of malignancy. He presented intense pain with neuropathic characteristics in the D6 site that persisted and spread to lower limbs, being treated with corticosteroids followed by Azathioprine. The treatment resulted in pain control and improvement of the walk disability.

Conclusions: We would like to call neurologists' attention to this association, since there are only a few reports of SAPHO syndrome with neurological deficit, showing possible misdiagnosis owing to the rarity and limited knowledge about this disease.

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1570
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Late Breaking Posters 2
Frequency of epilepsy in patients with neurocysticercosis, in the service neurology and neurosurgery mother-child hospital

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Abstract: Cysticercosis of the central nervous system is the most important of human neurological diseases of parasitic origin. It generates considerable morbidity, is known to be one of the major causes of epilepsy, with serious social, physical and psychological consequences.

Objective: To assess the frequency of epilepsy in patients with neurocysticercosis, treated at the Mother and Child Hospital.

Type of study: Study descriptive. Study Area: Neurology and Neurosurgery Department of the Mother and Child Hospital the city of La Paz – Bolivia.

Universe: work was done with the total of the universe, 352 patients diagnosed with epilepsy treated during the period from 2007 to 2010.

An instrument to collect: Workbook statistics and medical histories.

Results: Of the total 115 patients had such as base diagnosis neurocysticercosis in 43 cases (37 %) were asymptomatic patients and in 72 cases (63 %) patients had epilepsy.

Conclusion: Of 352 patients diagnosed with epilepsy in 72 cases (20 %) epilepsy was secondary to neurocysticercosis, followed by 31 cases (9 %) secondary epilepsy to AVC.

Keywords: Neurocysticercosis and epilepsy.

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