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Miscellaneous Topics 4

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Triplet repeat primed pcr for japanese patients with myotonic dystrophy type 1

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Background: Many patients with myotonic dystrophy type 1 (DM1) develop muscular atrophy and weakness accompanied with myotonia in their adulthood. Some patients with DM1 have cataracts and heart problems without muscular weakness. CTG expansion in 3' untranslated region of DMPK gene causes DM1. The detection of this mutation has been done using Southern blotting or PCR-Southern blotting. But these methods need many procedures. Recently, the triplet repeat primed PCR (TP-PCR) was developed to detect some nucleotides repeat expansions.
Objective: We tried to apply this TP-PCR methods to detect the Japanese DM1 patients and aimed to evaluate the efficacy for screening DM1.
Patients and methods: We took peripheral blood from 10 Japanese patients in our hospital with informed consent. Three of them had cataracts and abnormal ECG findings without muscular weakness. We designed primers and an anchor-primer according to the previous reports about TP-PCR for SCA36 and C9ORF72. We examined the TP-PCR and fragment analysis each with fluorescent-labeled primers by autosequencer, and did PCR-Southern blotting analysis.
Results: The PCR-fragment analysis revealed mild expansion within 100 repeats in three patients without muscular weakness. Seven other patients had smear-like abnormal band in the PCR-Southern blotting. TP-PCR showed smear-like-peaks in all 10 patients including three mild ones.
Conclusion: This method could detect all abnormal expansions. But above 100 repeats, all patients showed almost the same pattern and could not show the difference of the size of repeats. The triplet repeat primed PCR is a convenient and useful method for screening DM1.

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Ataxia and progressive myoclonic epilepsy associated with reduced ceramide synthase 1 (CERS1)

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Background: Ceramides and their sphingolipid have a wide range of biological functions. Ceramide synthase 1 (CERS1) catalyzes the synthesis of C18 ceramide and is mainly expressed in the brain. Alterations of sphingolipid metabolism are involved in the pathogenesis of many neurodegenerative disorders.
Objective: Report a case of myoclonic epilepsy and ataxia associated with homozygous variant in the gene CERS1, described in only one family previously).
Patients and methods: male, 22 years old with history of appendicular ataxia since 01 years old, followed by slight myoclonic jerks that progressively worsened despite treatment instituted and delayed psychomotor development. He had a previous episode of generalized tonic-clinic seizures (CTCSs). The electroencephalography (EEG) showed bursts of generalized polyspikes and slow wave discharges and Magnetic Ressonance imaging of the brain showed signs of pontine atrophy, cerebellum and cerebellar peduncle medium and higher, associated with the pontine cross sign. Genomix exonme's analysis demonstrated a homozygous variant of CERS1.
Results: It was found a homozygous variant of CERS1 gene in a patient with epilepsy, ataxia and progressive myoclonic epilepsy, a rare condition currently described as myoclonic epilepsy type 8.
Conclusion: Our case demonstrates that reduced levels of CERS1 could be associated with progressive myoclonic epilepsy and must be considered as a differential diagnosis, supporting that impairment of ceramide biosynthesis underlies neurodegeneration in humans. Futher studies may help to define the phenotypes more appropriately.

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Miscellaneous Topics 4
A novel homozygous missens mutation in capn3 gene detected in Saudi Arabian patient with limb-girdle muscular dystrophy type 2A

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Background: Limb-girdle muscular dystrophy type 2A (LGMD2A) is an autosomal recessive disorder caused by mutations in the CAPN3 gene, which is located on chromosome 15. Calpainopathy seems to be relatively frequent in comparison with other LGMD types. It is characterized by selective atrophy and weakness of proximal limb and girdle muscles.
Case: We report a Saudi Arabian family with initial presentations of weakness in limb girdle distribution. At presentation, waddling gait,

0022-510X/$ – see front matter.

positive Gowers' sign, and marked muscle atrophy in pelvic and leg muscles were noted.
Material and method: Sequence analysis of all coding exons (1 to 24) and all intron/exon boundaries of the CAPN3 gene.
Results: c.1699G > A variant was detected. It is a novel variant not previously described in other patients.
Conclusion: In silico predictions indicate that it is probably disease-causing mutation. To our knowledge, we report for the first time the c.1699G > A variant in CAPN3 gene which can be considered as a strong genetics factor disease causing of LGMD2A.

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Miscellaneous Topics 4
Finding words in the neural code of the GPI: Complex properties of neuronal activity in Parkinson's disease

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Background: Based on the calculation of the temporal structure function of spike trains, it can be determined whether information is encoded by time patterns, or they must be ruled out. Furthermore, the length of the information carrying units in the code (words) can be measured, and the time scales at which a rate or a time code take place can be determined (Andres et al. 2014, 2015).
Objective: To analyze the presence of complex time patterns vs. rate coding in the globus pallidus interna (GPi) from parkinsonian patients.
Patients and methods: Patient and Institutional Review Board approval was obtained. Twenty-two GPi single cell recordings from 6 patients with Parkinson's disease (PD) were analyzed. The temporal structure function was calculated based on interspike intervals (ISIS) time series. The algorithm is available at www.neurostruct.org.
Results: In 73% of the recordings, complex time patterns were found at short time scales. In 64% of the cases a co-existing rate code was detected at higher time scales (figure: upper panel). In 27% of the cells, no complex time patterns were found at short time scales, precluding any coding scheme other than a time averaged measure (figure: lower panel).
Conclusion: The co-existence of complex time patterns with a rate coding region at different time scales in the GPi shows the presence of a multiplexed neural code.

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Miscellaneous Topics 4
Are white matters changes in dm1 brain related to anosognosia?

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Backgound: Myotonic Dystrophy is a multisystem disorder where there is a prominent involvement of brain.
Patients and methods: We investigated 23 patients by neuropsychological testing, structural psychological interview, brain MRI and subdivided region of interest in frontal, parietal, occipital and temporal insular. We used the ARWMC score to classify white matter lesion as follow: 0 normal, 1 focal lesion below 5 mm diameter, 2 confluent lesions, 3 diffuse involvement. Anosognosia was defined as an inability of patient to be aware of their medical condition. All patients were genetically proven and MIRS was done. One patient refused MRI for claustrophobia.
Results: Our DM1 patients in about 80% exhibited lesion of the brain MRI and flair hyperintensity in temporo-polar or frontal regions. In most patients bilateral temporo-polar MRI intensity were seen and were compared with their neuropsychological parameters and CTG repeats. In our study most the prominent brain lesion were white matter lesion hyperintensity in temporo-polar region. Since, most of ours patients (65%) had anosognosia, we correlated the observe white matter lesion with this clinical features that were suggestive of correlation.
Conclusion: Most DM patient have evident neuropsychological abnormalities and 2/3 of them alteration of white matter in temporo-polar lesion. We proposed that anosognosia is correlated in DM1 with this changes. Whether these changes represent congenital changes or develop with time, a follow up of this patients will be necessary to address this issue.

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Miscellaneous Topics 4
Combining video games and neurology: The potential of brain computer interfaces

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Objective: Evaluating the feasibility of the Epoc + Brain Computer Interface system when developing applications and the potential of making a video game with that technology.
Background: Brain-computer interfacing research has been motivated by the wish to provide severely paralyzed people with new communication and motor abilities, for example Amyotrophic Lateral Sclerosis and Locked-in Syndrome. Recently more affordable BCI systems have been introduced in the market intended to be used at home like Epoc + by Emotiv Inc.
Design methods: We used 15 healthy participants, aged between 23 and 52. For facial recognition participants were told to smile, frown, wink, raise the eyebrows and smirk while wearing the Epoc+. Then, we used Epoc + with three different control actions. For emotion detection we made subjects listen to music used to trigger specific emotions.
Results: Facial expression detection was satisfactory with Epoc + having a 67.7% success rate in detecting the right expression. The control signal worked only when the subject was using facial expressions (90% success rate); without them it was quite disappointing (14% success rate). The same was true for emotion detection: It produced good results only when the participant was experiencing feelings triggering facial expressions for example fear and anger.
Conclusions: Based on both our results and the literature we believe that Epoc + mainly employs potentials from facial muscle contraction. However, it is a promising technology for future research with possible uses not only with patients with ALS and ILS but also in applications such as video games for healthy subjects.

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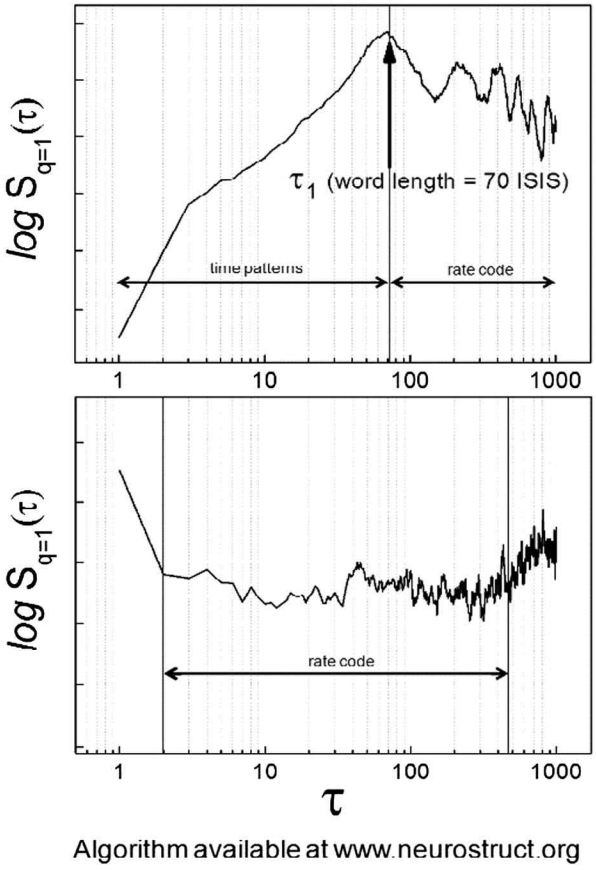

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Immune response and amino acids blood concentration during ultramarathon: A sportomics approach

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Background: Sportomics is non-hypothesis-driven holistic top-down research on individual metabolic changes during sports and exercise. Ultramarathon is a prolonged exercise and an important challenge for the Central Nervous System (CNS), making it an interesting protocol to study central and peripheral fatigue.
Objective: Here we evaluated the immune response and amino acids metabolism during an ultramarathon in an elite world-class male athlete.
Methods: The athlete ran for 8 hours (86 km). Venous blood samples were collected (fasting, before race, hourly, recovery - 60 min and 720 min) and ~75 cellular and biochemical parameters were measured. Major nitrogen compounds were corrected by creatinine concentration and evaluated using Pearson correlation.
Results: Leucocytes presence increase in blood was mostly due a neutrophil response to exercise (250%). We also observed a lymphocytes reduction. The exercise raise of both leukocytes and neutrophils were cooperative and fitted in sigmoid function with high correlation ($r = 0.97$). On the other hand their presence in blood declined during recovery following an exponential decay. We were not able to find an equation to fit the lymphocytes presence behavior. Ammonia increased more than 300%, almost due amino acid deamination (mainly Leu; Ile; Phe and Trp). A raise of both urea (200%) and urate (30%) was also measured.
Conclusion: The results may show an important metabolic adaptation during ultramarathon and demonstrate the kinetics of amino acids consumption as energetic molecules during exercise in a world-class athlete. The findings bring new information about the amino acids availability to the CNS during prolonged exercise.

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The effect of training and Gallic acid on tumor necrosis alpha factor and anxiety in Alzheimer's rats

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Introduction: The aim of this study is to investigate the effects of endurance training and Gallic acid on tumor necrosis alpha factor(TNF-α) and anxiety behavior in rats with Alzheimer's disease.
Method: For this purpose, 70 Sprague Dawley rats (70 males) were randomly divided into seven groups, including: (1) control, (2) sham, (3) endurance training (4) Gallic acid 50 (5), Gallic acid 100, (6) Training and Gallic acid 50 and (7) Training and Gallic acid 100. During eight weeks, rats of groups 3, 6 and 7 ran on treadmill's without incline at a speed of 15 to 20 meters per minute for 15 to 30 minutes per session and five times a week. In addition, every day groups 4 and 6 were injectected with 50 mg/kg of Gallic acid and groups 5, and7 were injected with 100 mg/kg of Gallic acid. For statistical analysis of data used two way ANOVA and tukey post hoc tests.
Results: The findings showed that levels of tumor necrosis alpha factor, anxiety in all test groups were significantly lower than sham Group ($p \leq 0.05$).
Conclusion: Based on this finding endurance training, Gallic acid and a combination of these decrease tumor necrosis alpha factor, Anxiety in rats with Alzheimer's disease.

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And the words are meaningless- apahasia in Florence + The Machine's "all this and heaven too"

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Background: Fictional literature and poetry can be provide us with remarkable representations of neurological signs and symptoms and even give us a hint as to possible diagnosis. Similarly, this can be applied to the lyrics of indie/pop songs.
Objective: To analyze the lyrics of Florence + The Machine's song "ALL THIS AND HEAVEN TOO" and how these could be a poetical expression of aphasia.
Material and methods: The authors reviewed the lyrics of the song and how they can represent an aphasic patient throughout the onset of symptoms and recovery/rehabilitation.

Results: In Florence + The Machine's "ALL THIS AND HEAVEN TOO", the narrator reports a loss of communication skills (mostly oral) that can be interpreted as aphasia. Careful analysis and interpretation of lyrics provides us with what can be interpreted as aphasia, throughout the song, how it impacts on the subject/patient and conflicting emotions as functional adaptation to the deficit takes place.
Conclusions: Not only traditional fictional texts, but also lyrics for indie/pop songs can translate abnormal neurological symptoms and signs, namely aphasia.

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Migraine, subarachnoid haemorrhage, thunderclap headache and vertigo – Differential diagnosis in Florence + The Machine's "DRUMMING SONG"

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Background: Fictional literature and poetry can provide us with remarkable representations of neurological signs and symptoms and even give us a hint as to possible diagnosis. Similarly, this can be applied to the lyrics of indie/pop songs.
Objective: To analyze the lyrics of Florence + The Machine's song "DRUMMING SONG" and how these could be interpreted as migraine, subarachnoid haemorrhage, thunderclap headache or even bouts of vertigo.
Material and methods: The authors reviewed the lyrics of the song and how they can represent symptoms suggestive of different neurological conditions, namely migraine, subarachnoid haemorrhage, thunderclap headache or vertigo, discussing the possible differential diagnosis.
Results: In Florence + The Machine's "DRUMMING SONG", the narrator/patient reports what could be a throbbing, disabling headache, with loss of balance. Careful analysis and interpretation of lyrics provides us with diverse symptomatic descriptions that is used to formulate four possible differential diagnosis: migraine, subarachnoid haemorrhage, thunderclap headache or bouts of vertigo.
Conclusions: Not only traditional fictional texts, but also lyrics for indie/pop songs can translate abnormal neurological symptoms and signs. In this case, we discuss the differential diagnosis among four different conditions: migraine, subarachnoid haemorrhage, thunderclap headache and vertigo.

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Miscellaneous Topics 4
Tourette's syndrome in famous musicians

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Background: Tourette's syndrome (TS) is defined as a disorder characterized by multiple motor tics and at least one vocal tic that have lasted for at least one year. TS is a relatively complex neurobehavioral disorder, where the patients could have coexistent attention deficit hyperactivity disorder, obsessive–compulsive disorder or other behavioral comorbidities. Perhaps the cases of TS are that of the some musicians presented.
Objective: To discuss the possibility and the real diagnosis of TS in important musicians.
Material and methods: We selected the biography of many famous musicians with Movement Disorders to met features that could indicate the probability of the TS diagnosis.
Results: The tics and psychiatry disorders in genius Wolfgang Amadeus Mozart (1756–1791) and the rock star Kurt Cobain (1967–1994) may point to TS disorder. Some contemporary musicians have their clinical condition confirmed as TS, but after many years suffering with the disease.
Conclusion: The hypothetical diagnoses of TS in the cases of Mozart and Cobain have supporters with the possibility of tics and psychiatric comorbidities. There was a delay of TS diagnosis in the contemporary musicians. This situation and the controversies about the clinical case of Mozart show how difficult is the confirmatory diagnosis of this complex disease.

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Transcranial sonography of the substantia nigra and its correlation with DAT-SPECT in the diagnosis of Parkinson's disease

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Introductions: Transcranial sonography (TCS) of the substantia nigra is a new and promising method to diagnose Parkinson's disease (PD) but its effectiveness is controversial.
Methods: All 55 PD patients involved in the study underwent single photon emission computed tomography (SPECT) imaging using the labeled dopamine transporter radiotracer ^{99m}Tc-TRODAT-1 to assess nigrostriatal dopaminergic function. The echogenicity of the substantia nigra was measured by TCS in all patients who received DAT-SPECT scanning. Finally, statistical analysis was carried out to determine the diagnostic accuracy of TCS as well as its correlation with ^{99m}Tc-TRODAT-1 SPECT, its positive predictive value (PPV), and negative predictive value (NPV).
Results: Contralateral striatal ^{99m}Tc-TRODAT-1 uptake was significantly reduced compared to ipsilateral striatal uptake, and had a negative correlation with UPDRS-III($r = -0.334$, $p = 0.013$), disease duration ($r = -0.393$, $p = 0.003$) and H-Y stage ($r = -0.330$, $p = 0.014$). After TCS measurement, the contralateral SN echogenic area was similar to the ipsilateral SN echogenic area (27.77 ± 13.19 vs $25.98 \pm 11.94 mm^2$, $p = 0.402$, $n = 24$). No correlation was identified between TCS and UPDRS-III ($r = 0.383$, $p = 0.065$), disease duration ($r = 0.371$, $p = 0.075$) or H-Y stage ($r = 0.259$, $p = 0.222$). The sensitivity and specificity of SN TCS for the diagnosis of PD were calculated as 64.70% and 60% according to DAT-SPECT, respectively, while the positive predictive value and negative predictive value was calculated as 91.67% and 20%, respectively.
Conclusions: Compared to DAT-SPECT, TCS is a non-radioactive and convenient procedure to perform. In our investigation, TCS had no correlation with DAT-SPECT. However, the high positive predictive value of TCS highlights its possible utility as a routine diagnostic test.

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Miscellaneous Topics 4
Below-the-hairline EEG allows for the rapid evaluation of seizures in the neuro-ICU

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Background: Seizure is a commonly considered etiology of acute transient neurologic syndromes; however, obtaining an EEG has traditionally required trained technologists. The below-the-hairline (BTH) EEG can be placed by clinicians with minimal training.
Methods: We describe two cases highlighting effective use of BTH-EEG for the rapid evaluation of seizure in the neuro-ICU at Brigham and Women's Hospital where the use of the tool changed clinical management.
Results: A 55-year-old man with heart disease, HCV, and a complex psychiatric history presented with episodes of unresponsiveness and generalized limb shaking followed by confusion. Recent deterioration of his social support system prompted referral with a provisional diagnosis of non-epileptic seizures. Overnight BTW-EEG during one of his characteristic spells demonstrated bilateral seizure activity (Fig. 1). He was rapidly treated with anti-epileptic drugs and seizure activity promptly terminated. A 53-year-old woman with metastatic breast cancer was emergently transferred to the neuro-ICU for episodes of unresponsiveness with leftward gaze deviation concerning for seizures. Hairline EEG demonstrated only generalized cortical slowing during characteristic episodes. The diagnosis of seizure was promptly abandoned and a lumbar puncture revealed intracranial pressure >55 cm H2O, consistent with plateau waves due to leptomeningeal carcinomatosis.
Conclusions: The BTH-EEG is a fast and accessible tool that can be used effectively by clinicians without specialized technical training to evaluate for acute seizures.

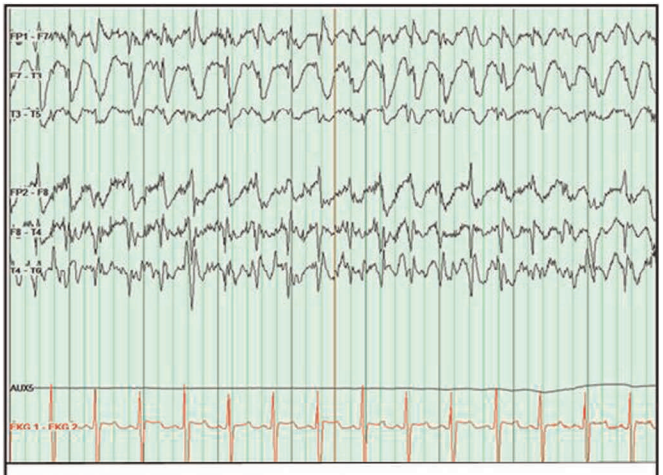

Fig. 1. BTH-EEG showing a generalized seizure in fronto-temporal regions.

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Establish a specialist in neurology program in Brunei Darussalam with the help of telemedicine

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Introduction: Neurology will be one of the major columns of medicine. With a worldwide lack of neurological knowledge there is an obvious high need for specialist in Neurology training as well as a need of neurological centres. Since 7/2010 a transcontinental cooperation between the Brunei Neuroscience Stroke and Rehabilitation Centre(BNSRC),Brunei Darussalam (BD) and the Department of Neurology, Krankenhaus Nordwest(KHNW), Frankfurt, Germany has been successfully implemented.BNSRC is the 1st neurological facility in BD, therefore local doctors has to be trained in the field of neurology to ensure that they can handle the patients as well as to get a curriculum of neurology in order to become specialist in neurology. This program is established and conducted by Universiti Brunei Darussalam,the University of Heidelberg,Germany,Johns Hopkins,USA,KHNW and BNSRC.
Methods: The total duration shall normally be at least 60 months full-time.We established a Teleteaching program including on site teaching. It consists of different modules;electrophysiology training, EEG education, clinical examination including scales for different diseases and the grand round.The grand round is basically a teaching ward round performed by Head of Department and consultant neurologists to discuss selected patients. The local doctor report the history, demonstrate physical symptoms, diagnostic work up and therapeutic strategies are discussed as well as all medical findings.
Results: 2 local doctors are enrolled, overall 7 doctors have taken part and past all assessments.
Conclusion: Neurological diseases are a major column and the prevention of stroke and other neurological diseases are more important than ever. As there is a need of specialists in Neurology worldwide, this specialist in neurology program can set a milestone in teaching of neurologic skills with the help of telemedicine to overcome distances.

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Miscellaneous Topics 4
Hereditary spastic paraplegia with thin corpus callosum (HSP-TCC): 3 new families from Saudi Arabia with 2 novel SPG-11 mutations

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Background: HSP-TCC is autosomal recessive with progressive spastic paraparesis, cognitive impairment, possible neuropathy and urinary bladder involvement caused by SPG11 (spatacsin) mutations.
Objective: To report 3 Saudi families with SPG11 HSP -TCC.
Patients/methods: Family 1. A 34-year-old male born to unrelated parents reported progressive gait abnormalities, sphincteric and cognitive impairment. His 31-year-old sister was affected. Family 2. A 26-year-old man born to consanguineous parents experienced childhood-onset cognitive impairment and gait difficulty. A brother

was affected. Family 3. An 18-year-old man, with 3 affected siblings, born to consanguineous parents developed cognitive impairment and progress weakness. All had clinical findings suggestive of HSP and underwent MRI, EMG and SPG-11 testing.
Results: Brain MRIs showed hypoplastic CC with deep white matter hyperintensities. In Family 1, EMG demonstrated severe bilateral median nerve entrapments and abdominal CT revealed polycystic kidneys. SPG11 testing (Athena Diagnostics) respectively revealed: a novel autosomal recessive, 1-base pair deletion of C, at codon 883(c.2694) resulting in homozygous frame shift mutation; an autosomal recessive homozygous 2-base pair deletion of AG at codon 2278 (c.6832_6833) resulting in pathogenic frame shift; and an autosomal recessive homozygous nonsense mutation c.6349 G > T resulting in p.Glu2117X predicted pathogenic. IRB approval was obtained to report these cases.
Conclusion: Three new Saudi families with SPG11 HSP-TCC are reported. One, with a novel c.2649 mutation, also had cystic kidneys and median nerve entrapments, possibly expanding the phenotype. Another had a novel nonsense mutation at c.6349. SPG11 HSP TCC is likely more prevalent in Saudi Arabia due to high consanguinity rates.

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Miscellaneous Topics 4
X-linked-adrenoleukodistrophy with a new mutation in the ABCD1 gene

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Objective: ALD is a rare, X-linked peroxisomal disorder of beta-loxidation that results in accumulation of VLCFA in every tissue. It is caused by mutation in the ABCD1 gene that encodes an ABC transporter. A patient affected by X-ALD with a new mutation in ABCD1 gene is described.
Material and methods: 13-years-old male, without any personal or familiar history of neurological disease, with a progressive course consisting on vision impairment, dysarthria and right hemiparesis. The patient also has hyperpigmented skin. The laboratory test showed increased ACTH levels and a rise in plasma cortisol concentration following ACTH stimulation. EMG and visual evoked responses (VER) were normal. The cranial MRI showed severe bilateral occipitoparietal demyelination in cerebral white matter. The plasma concentration of VLCFA showed an increased ratio of C24:0/C22:0, despite a normal VLCFA concentration.
Results: The molecular genetic testing of the ABCD1 gene locus showed an unpublished hemizygous mutation c.430_432delGCT (p.Ala144del). His asymptomatic mother carried the same mutation.
Conclusion: We report a new mutation in the ABCD1 gene in a patient with X-ALD. This mutation means an amino acid deletion of an alanine which affects protein structure and may adversely affect its function.

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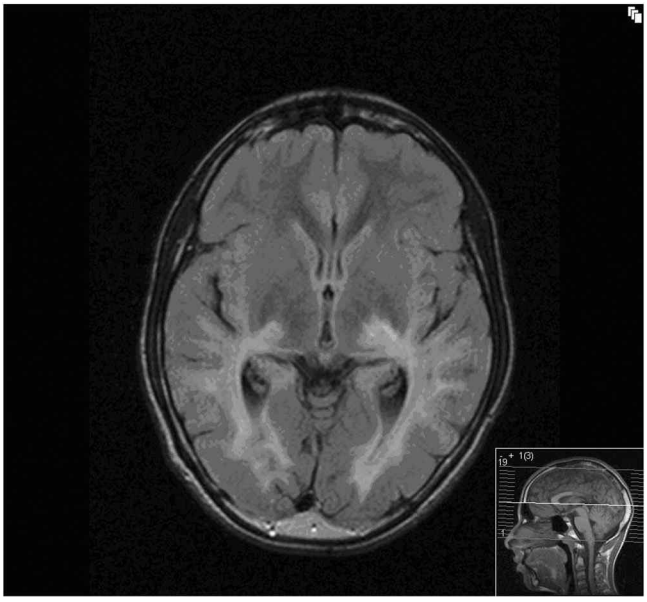

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WFN15-1166
Miscellaneous Topics 4
The electroencephalogram in the evaluation of school failure children - A comparative study

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Introduction: The real impact of electroencephalographic abnormalities in Learning Disorders (LD) has not yet been established, although it is usually required in these conditions. The aim is to describe the electroencephalogram (EEG) records findings in school failure group (SF) and not school failure (NSF) and, then, compare data.
Methods: A total of 71 students of SF group and 70 of NSF group, from a public school in Federal District in Brazil, were evaluated. Fifty-nine of SF group and 48 of NSF group was tested with EEG, which was interpreted by a blind neurophysiologist in this study. The association between the variables (normal and epileptiform) was tested with $\chi 2$ test and Fisher's exact test, case–control analysis. The level of statistical significance was 5% ($p < 0.05$).
Results: The mean age of SF group was 9.1 years and NSF group was 10.5 years. There was no epilepsy historic. In group SF, 10 (17%) were abnormal, 5 (50%) with epileptiform activity (EA) (all focal: 1 frontal, 1 occipital and 3 centro-occipital region). In group NSF, 4 (8%) were abnormal, of which 2 (50%) with generalized epileptiform activity (EA), 2 with focal EA and FIRDA. There was no significant association between the variables: EA and groups, even considering the whole sample ($\chi^2 = 0{,}104$, gl = 1, p (Fisher) = 0,999 [OR 1,25; IC95% 0,32 – 4,85]), or considering just abnormal EEGs.
Discussion: There were no differences between the groups, demonstrating the absence of benefits in routine EEG request as propaedeutic strategies in LD.

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772
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Miscellaneous Topics 4
Neurophysiologic correlates of depression and anxiety

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Background: Depression often combined with anxiety is the common comorbidity of neurologic disorders. However their diagnosis is based on clinical judgment and self-referential psychological questionnaires without any objective neurophysiologic parameters. In many cases they are not obvious and sometimes denied by the patients.
Objective: To determine neurophysiologic correlates of depression and anxiety.
Patients and methods: One hundred twenty patients with depression (including 75 in combination with anxiety) and 20 age and gender matched healthy volunteers were included in the study. Neurophysiologic examination included 19-channel EEG (with the analysis of relative power for different frequency rates), ECG (low to high frequency assessment, LF/HF), visual-motor reaction (standard deviation of reaction time) and speech signal parameters (temporal and non-linear). Mathematical model based on discriminant analysis was created.
Results: Neurophysiologic parameters were significantly different for patients compared to healthy volunteers (Fig. a, b, c).
Conclusion: Neurophysiologic parameters routinely collected in neurological practice can be of additional value in diagnostics and differential diagnostics of depression. Further studies are needed in this field.

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773
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Miscellaneous Topics 4
Different decremental pattern in Lambert Eaton myasthenic syndrome from myasthenia gravis. A case report

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Background: Lambert Eaton myasthenic syndrome is a rare autoimmune presynaptic disorder of neuromuscular transmission. The clinical picture include proximal muscle weakness, areflexia and autonomic features. The electrodiagnosis is made with 1) a low compound muscle action potential (CMAP) amplitud at rest, 2) a decrement response at low rate repetitive nerve stimulation and 3) an incremental response at high rate repetitive nerve stimulation. High rate repetitive nerve stimulation easily discriminates between Lambert Eaton and myasthenia gravis. However at low rates, both disease show a decremental of the CMAP size within the initial 4 to 5 responses but with a different pattern and decremental ratio.
Objective and results: Describe the ratio between the 1st and the 4th decrement and de 1st and the 9th decrement (¨late/early%¨) and compared with myasthenia gravis. 41 years old men with proximal weakness and sexual dysfunction. Without ocular impairment or neoplasic disease. CT Scan was normal. FDG PET was performed. The median nerve resting CMAP amplitude was 3.7 mV, with an increment of 10.3 mV post effort. The low rate decrement was more than 10% (33%) and the late/early% ratio was 118%. The high rate increment was more than 100% (635%). The late/early% ratio in a myasthenic gravis patient was 69%.
Conclusion: The progressive decremental pattern at low frequency testing can discriminate this myasthenic syndromes and it is a helpful test when the high rate stimulation cannot be performed.

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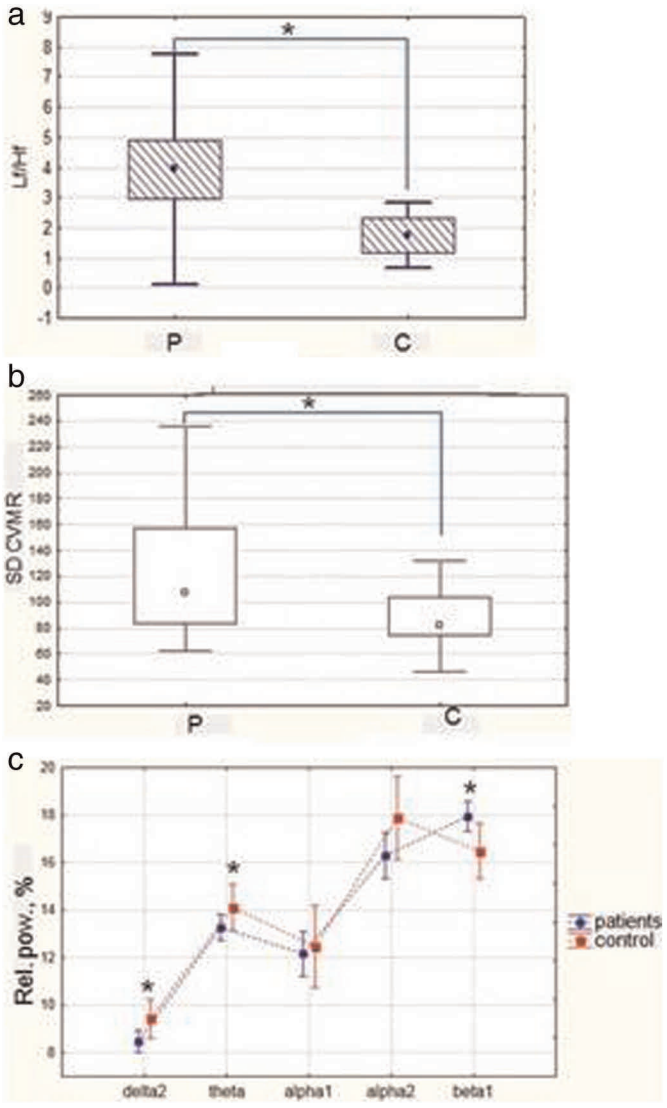

Figure. a. ECG (LF/HF). P – patients, C – controls. * - $p < 0.05$. b. Visual-motor reaction (standard deviation of reaction time). c. EEG (relative power for different frequency ranges).

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WFN15-0260
Miscellaneous Topics 4
Assessment of facial synkinesis with applied blink reflex test

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Background: Blink reflex could be a useful tool to differentiate facial synkinesis as one of complications of facial neuropathy, from volitional associated movements.
Method: We had performed applied blink reflex test for 43 patients with subjective or objective facial weakness. Protocol is as the following: routine nerve conduction velocity study of facial nerve and blink reflex, then stimulation of supraorbital nerve and recording of response from ipsilateral mentalis (Mn) muscles. Theoretical background is blink reflex is consisted of afferently maxillary branch of trigeminal nerve and efferently zygomatic branch of facial nerve, so that is intact, mandibular branch of facial nerve innervating Mn muscle does not contribute to blink reflex.
Results: R1 & R2 responses of blink reflex from ipsilesional Mn muscle are detected in 9 patients (20%) of the 43 patients. 6 (67%) of 9 patients show normal blink reflex response from OOc muscle but

positive blink reflex response from ipsilateral Mn muscle, and paradoxically only 3 (33%) both positive blink reflex responses from OOc and Mn muscles. R1 and R2 latencies from OOc muscle are able to be obtained from 8 of 9 patients except one unobtainable potential case, are faster than ones from Mn muscle by 0.72 for R1 and 0.94 for R2.

Conclusion: We concluded that a positive blink reflex from mentalis muscle is almost always suggestive of chronic facial neuropathy even in clinical silence of facial synkinesis, or an aberrant reinnervation after peripheral facial neuropathy, and does not electrophysiologically correlate with the severity of facial palsy.

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775
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Miscellaneous Topics 4
E-stroke: On-line stroke training for health teams

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Background: Continuing training of health teams is recommended to improve quality of care and stroke patient outcomes.

Objectives: To design and implement an online stroke training program for health care professionals working in the public health-care system and for Ñandu Project (Stroke Surveillance Project for the Ñuble province in Southern Chile) participants.The program aims to facilitate organizational improvements through modular, peer-reviewed, evidence-based training in the acute care of stroke patients in Chile.

Methods: The program includes the following stroke modules: urgent care, evidence-based acute care, and long-term care and rehabilitation of patients. Contents include national and international clinical practice guidelines recommendations. Each module contains interactive teaching material, academic forums with expert instructors (neurologists, nurses, epidemiologists), training evaluations, and access to complementary material.

Results: We designed and implemented the urgent care module, which included stroke symptoms identification, stroke code activation, and urgent care. Participants were physicians (43%), nurses (44%), and other health professionals (13%) from 12 regions of Chile. A total of 1,115 health professionals are registered in future courses. The retention rate was 93% and 74% passed the course. In all, 88% reported that the course helped them update their stroke care knowledge and skills, while 92% believed it would be useful in their clinical practice.

Conclusions: The preliminary results of this specifically-designed training program have been positive in terms of updating knowledge and modifying clinical practice.

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776
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Miscellaneous Topics 4
Early amygdala detection of intentional harmful actions

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Background: The amygdala has been proposed as a critical hub for the processing of intentional harms. A direct early involvement of amygdala has not been proved.

Objective: Determine the early role of the amygdala in detecting intentionality in harmful actions.

Patients: Three patients with intractable epilepsy took part of the study.

Methods: Local field potentials to stimuli were obtained with depth electrodes located in the amygdala and in other frontal, temporal and parietal regions while patients performed an adaptation of the intention inference task. Data analysis performed included: time-frequency charts, logistic regressions of trial by trial, a comparison between amygdala and the rest of the regions, seed analysis using weighted symbolic mutual information (wSMI) and brief time span functional connectivity.

Results: An early boost in the amygdala during the first 200 ms after stimuli at 1–40 Hz and along 0–1500 ms at 60–150 Hz (BB), discriminated intention and content as revealed by power activity. Trial-by-trial analysis's of the amygdala responses at BB predicted categorization. Compared to other brain regions, the amygdala was the only site that systematically discriminated and predicted the inference about intentionality of harmful actions. Both amygdala seed analysis using wSMI and frontotemporal connectivity at early stages showed enhanced network activity for intentional harmful actions.

Conclusion: This study provides unique temporal data on human amygdala regarding intentionality detection and its influence in subject's categorization, as well as accompanying early corticolimbic tuning during intentional harmful actions' observation (FONDECYT Regular 1130920 and 1140114, Foncyt-PICT 2012–0412, 2012–1309).

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777
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Miscellaneous Topics 4
Evoked potentials in a case of progressive multifocal leukoencephalopathy

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Background: Progressive multifocal leukoencephalopathy (PML) is a rare but fetal papovavirus infection of the central nervous system which primarily affects immunocompromised patients. Evoked potential (EP) findings associated with PML has rarely been reported. We report a case of human immunodeficiency virus (HIV)-associated PML with EP findings.

Case: A 32-year-old man was admitted to hospital with a 1-month history of anorexia, weight loss, and sluggish movement. Neurologic examination showed confused mentality, decreased fluency, mild dysarthria, impaired tandem gait, and mild rigidity and tremor. Brain MRI showed hyperintensity in the bilateral periventricular and middle cerebellar peduncle without gadolinium enhancement. Blood tests showed positive anti-HIV antibody. The plasma PCR for John Cunningham **virus** DNA was positive but negative in the CSF. Brainstem auditory and visual EPs were normal. Median nerve somatosensory EPs showed a prolonged central conduction time (right; 8.3 ms, left; 7.2 ms, nomal≦7.0 ms) and unusually broadened cortical responses following bilateral side stimulation. Tibial nerve somatosensory EPs showed normal lumbar potentials with unobtainable cortical responses. These findings suggested bilateral central conduction defects of the somatosensory pathways between the cervical cord and the brain. He was

treated with highly active antiretroviral therapy (HAAT) with levodopa/benserazide. Three months after admission, he was discharged with marked neurological improvement.

Conclusion: We report the favorable outcome of a patient with parkinsonism in an AIDS-related PML patient treated with a combination of HAAT and antiparkinson medication. We suggest that somatosensory EPs are would be helpful in the diagnosis and localization of the PML.

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778
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Miscellaneous Topics 4
Complete afferent trigeminal sensory neuropathy due to pontine infarction

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Background and objective: Isolated cranial nerve palsies are often attributed to lesions of the respective nerves. Small lesions of the brainstem can affect the trigeminal nerve only. A few cases of isolated trigeminal sensory change have been reported previously, but trigeminal sensory neuropathy has been rarely reported as a manifestation of pontine infarction.

Patient and methods: A 77-year old female patient who had a history of hypertension and hyperlipidemia admitted with sudden Left perioral numbness. Two days prior to admission, she complained of numbness on the left perioral area. On neurological examination, pain and light touch sensations were decreased in the left V2 and V3 area.

Results: Brain magnetic resonance imaging showed hyperintensity on left middle cerebellar peduncle in DWI & T2WI. No blink reflex potentials were recorded on the ipsilateral R1, R2 and contralateral R2 on stimulation of the left supraorbital nerve, while normal R1, R2 and contralateral R2 responses on the right side. Left Jaw jerk reflex were absent on masseter muscle.

Conclusion: These clinical and electrophysiologic findings of abnormal blink reflex, left V2 and V3 area sensory change and an absence of left jaw jerk responses supports that hypothesis that the lesion may involve the whole V1, 2, 3 trigeminal sensory afferent limbs including Ia fibers of motor part before entering the each nuclei. A few cases of isolated trigeminal sensory change have been reported previously, we report an interesting case of pontine infarction presenting complete trigeminal afferent defects with electrophysiologic data.

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779
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Miscellaneous Topics 4
The segmental conduction study on sural nerve would reveal progression of diabetic neuropathy

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Objective: The pathophysiology of diabetic metabolic neuropathy has a length dependent pattern. But in pathology some proximal axons showed demyelination lesion and distal part showed axonal degeneration. So we devised multi-segmental conduction study(NCS) to find progression of predominant pathophysiologic pattern (demyelinating or axonal injury) to select proper therapeutic measures.

Patients and methods / material and methods: We recruited 15 diabetic patients (30 sural nerves, male = 9, 52.6 ± 11.5 yrs) with some sensory symptoms and 12 normal controls (24 sural nerves, male = 10, 46.4 ± 13.3 yrs). Standard NCS on motor, sensory nerves in upper (median, ulnar nerves) and lower extremity (peroneal, sural, and tibial nerves) were done. Diabetics were classified into two groups (with or without polyneuropathy). Orthodromic sural segmental NCS was done; below lateral malleolus 7 cm, above malleolus 7 cm, and next 7 cm to calf (total 21 cm).

Results:

1. There is no difference between diabetic patients and control in age ($p > 0.05$). But, older subjects were more susceptible for diabetic neuropathy, but relevance was insignificant ($p > 0.05$).
2. There is significant difference of amplitudes in all three segments (esp, segment 2; 16.9 ± 8.4 μV, 12.9 ± 11.2, 4.5 ± 3.3, $r = -0.533$, $p < 0.05$).
3. Conduction velocity of diabetic neuropathy patients are affected in first segment (32.6 ± 3.8 m/s, 39.2 ± 3.5, 27.5 ± 9.3, $r = -0.299$, $p < 0.05$). In second, third segment, between three groups, results were not significant.

Conclusion:

1. In this NCS reconfirmed that diabetic neuropathy shows length dependent pattern neuropathy known as before.
2. If we use segmental NCS in sural nerve, we would obtain more relevant information for understanding pathophysiology of diabetic neuropathy.

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780
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Miscellaneous Topics 4
Characteristic development of GABAergic transmission in the mouse spinal cord

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In the mammalian central nervous system, gamma-amino butyric acid (GABA) is a predominant inhibitory neurotransmitter, whereas it acts as an excitatory transmitter in the immature CNS, and may be involved in morphogenesis. We have investigated the ontogeny of the GABAergic transmission by immunohistochemistry for glutamic acid decarboxylase (GAD), GABA transporters (GATs), vesicular GABA transporter (VGAT), and potassium chloride cotransporter2 (KCC2) in the mouse cervical spinal cord. In this session, we present the developmental changes in GABAergic transmission. (1) Before synapse formation, GABA may be extrasynaptically released by non-exocytotic system, and be transported into the processes of radial glia or immature astrocytes by GAT-3. (2) In the ventral horn, GABAergic neurons appear on embryonic day 12 (E12), synapses are formed after E13, and increased in number after E15. Synaptically released GABA was removed by only GAT-3 on the processes of astrocytes. (3) In the dorsal horn, GABAergic neurons are localized after E13, synapses are formed after E17, and increased in number after birth. Synaptically released GABA is removed by GAT-1 into presynapse and GAT-3 into processes of astrocytes. (4) GABA may act as an excitatory transmitter for several days before GABAergic synapse were formed in the embryonic spinal cord. (5) During

development, GABAergic synapses are decreased in the ventral horn, whereas glycinergic synapses are increased.

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781
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Miscellaneous Topics 4
Usefulness of median nerve residual latency to evaluate diabetic neuropathy

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Background and objective: Residual latency (RL) is the difference between the expected and measured terminal latencies. Terminal latency index (TLI) is the ratio between the expected and measured latencies. RL and TLI are indicators of the state of a progressive distal slowing of the nerve conduction produced at the distal short segment. Median mononeuropathy is frequently seen as only abnormal finding in diabetic patients and its relationship to polyneuropathy is not clear. Polyneuropathy interfere quality of life, and early detection needed. This study was performed to determine the usefulness of median nerve RL to assess distal slowing associated with median mononeuropathy in diabetic patients.
Material and methods: We included 12 patients with median mononeuropathy with diabetes. Control values were obtained from 21 healthy subjects. RL and TLI were used to compare the distal segment with the proximal segment conduction velocity.
Results: There was no significant difference for RL except median nerve between diabetic patients with median mononeuropathy and normal controls.
Conclusion: The ulnar, peroneal, and tibial nerve RL values were not different in diabetic patients with median mononeuropathy compared to normal controls. Distal slowing as described by RL has limited value for electrodiagnostic parameter for subclinical polyneuropathy.

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782
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Miscellaneous Topics 4
Transcranial direct current stimulation (tDCS) for treating chronic pain – Preliminary results of open-label, self-administered, at-home treatment

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Background: Accumulating evidence suggests that tDCS might effectively treat neurological conditions including chronic pain. As typical for non-invasive neurostimulation modalities, the effects are temporary, and stimulation must be repeated to maintain benefit. Some tDCS equipment is portable and use low current, thus long-term home use by patients is feasible but not yet studied.
Objective: To assess the feasibility, safety and efficacy of at-home tDCS treatment of chronic pain.
Patients and methods: Patients provided informed consent for 20-minute treatment sessions of 1.5-mA anodal tDCS directed to the motor cortex contralateral to patients' painful areas. Two days of in-hospital training preceded 12 days of unsupervised at-home use. Pain ratings were collected before, once-weekly during, and 3 weeks after the end of the treatment.
Results: The 11 patients studied so far (mean age 60.6 ± 16.3 y; 7 female) include 5 with postherpetic neuralgia, 3 with trigeminal neuropathy, 2 with fibromyalgia, and one with intramedullary cavernoma. At the end of two weeks of treatment 27% of patients (3/11) had ≥20% pain reduction. Mean pain reduction was 10.3% in the entire group; 35% among responders. Pain scores reverted to baseline within three weeks after stopping tDCS, and there were no serious adverse events.
Conclusion: These preliminary results suggest that at-home tDCS might safely treat neuropathic pain for some patients. Further controlled studies are warranted.

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783
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Miscellaneous Topics 4
The VEMP score and walking in clinically isolated syndrome (CIS)

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Background: Balance, very important for walking, requires integration of 3 sensory systems, visual, somatosensory, and vestibular. Vestibular evoked myogenic potentials (VEMP), which directly investigate function of the vestibular system, are good measure of brainstem involvement of multiple sclerosis (MS). Also, worsening on Timed 25-foot Walk (T25FW) has a clinical impact on disability in multiple sclerosis (MS).
Objective: The aim of this study was to investigate the correlation of the VEMP score with the T25FW in patients with CIS.
Methods: We included 53 patients with CIS (41 females, mean age of 32,8 years, 16 with optic neuritis (ON), 17 with brainstem CIS (BC), 12 with transverse myelitis (TM), 3 cerebral and 3 multifocal). Cervical VEMP (cVEMP) and ocular VEMP (oVEMP) were analyzed in the form of the VEMP score. T25FW was performed two times consecutively.
Results: In the whole group, oVEMP score correlated with the T25FW ($r_s = 0.277$, p0,049), and there was a trend for VEMP score ($r_s = 0.263$, p0,062). When we performed subanalysis only on BC type of CIS, both VEMP score and oVEMP score correlated with the T25FW ($r_s = 0.711$, p0,002 and $r_s = 0.562$, p0,024, respectively). There was no correlation for any other type of CIS individually. More importantly there was no difference in the VEMP score between patients with BC type of CIS and all other patients ($p = 0.656$).
Conclusions: These results indicate that the VEMP score correlates well with walking speed in patients with CIS and further validates the VEMP score as a important measure of brainstem involvement in MS.

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784
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Miscellaneous Topics 4
Histopathological assessment of nerves in leprosy

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Background: Leprosy is a chronic granulomatous infection of the skin and peripheral nerves caused by Mycobacterium leprae. The

clinical presentation ranges from tuberculoid through borderline forms to lepromatous leprosy. Pure neuritic leprosy accounts for 5-10% of leprosy and it presents with asymmetrical nerve involvement and often no demonstration of lepra bacilli on slit skin smear.
Objective: To study the histopathological details of nerve biopsies from patients with leprosy.
Materials and methods: Patients presenting with mononeuritis multiplex with/ without skin lesions with suspected leprosy underwent nerve biopsy and slit skin smear examination for acid fast bacilli.
Results: Thirty one patients with mononeuritis multiplex were diagnosed clinically as leprosy. Skin smear examination was positive in 4 patients. Eighteen patients had isolated nerve involvement while 13 patients had associated hypoesthetic skin lesions as well. The histopathology of leprosy ranged from a spectrum of well-defined epitheloid granulomas and absence of lepra bacilli in tuberculoid type to plenty of bacilli in the endoneurium and foam cells in lepromatous leprosy. Leprosy was classified as tuberculoid in 12, lepromatous in 5 and borderline in 14 patients. Wade fite staining for lepra bacilli was positive in 14 patients. The diagnosis of leprosy could be confirmed in all patients and in addition the disease could be classified into various subtypes.
Conclusions: Nerve biopsy in leprosy offers not only a useful tool in the diagnosis of leprosy but also helps in classifying it.

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785
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Miscellaneous Topics 4
IFAP syndrome with corpus callosum atrophy (New Case Report)

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IFAP syndrome is an extremly rare congenital disorder with x linked transmission.

It is characterized by triad of Icthyosis Follicularis, Atrichia of the scalp, and Photophobia.
Objective: To report a new case of IFAP Syndrome with corpus callosum atrophy.
Patient and methods: An 11 year old male child issue of cansanguineous marriage with similar case in the family. He presented clinically severe psychmoteur retardation,with an inability to speak, generalized seizures, alopecia and photophobia MRI Brain showed Corpus Callosum atrophy. Diagnosis of the IFAP syndrome is based on the clinical features and the presence of mutation in the MBTPS2 gene.

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786
WFN15-1242
Miscellaneous Topics 4
Evaluation of medical students knowledge about brain death throughout meeting of academic leagues, organised by medical school in 2013, 2014

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Background: The knowledge concerning brain death (BD) criteria is not widely spread among medical doctors and students, as it is not a subject well covered in graduation. The lack of information about the ethical and legal aspects of this diagnosis generates incoherent attitudes and makes it difficult for potential donors to reach the patients that need the organs.
Objective: Evaluate the knowledge of medical students in Goiás on the BD protocol (Resolution No. 1480/97 of Federal Council of Medicine - CFM).
Materials and methods: A cross-sectional study with 136 medical students participating of a health promotion event for community, known as Meeting of Academic Leagues, was conducted by Academic League of Transplants, from Medical School of Federal University of Goiás in 2013 and 2014. A questionnaire with 20 questions was applied to evaluate knowledge of BD and organ donations. The participation was voluntary and anonymous.
Results: 39.7% never attended classes on BD. 48.53% considered going, as a criterion for early BD assessment, the Glasgow Coma Scale = 3. About 86% were aware of the requirement of neurologist / neurosurgeon participation at clinical examination. To 69.85%, the complementary exams are essential and 91.18% answered that it is mandatory to BD notification. The BD has value of clinical death for 75.74% and 86.03% trust their diagnosis.
Conclusion: This study confirmed the lack of information on BD among medical students, evidencing the necessity of inclusion of BD in academic curriculum, aiming to enable the future physicians to conduct such diagnosis.

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787
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Miscellaneous Topics 4
Inter- and intraindividual differences of vulnarability of recurrent laryngeal nerves under tensile stress in a porcine model

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Background & aims: Our aim was to analyse the impact of prolonged intraoperative tensile stress on the recurrent laryngeal nerve (RLN) in an animal model using continuous neuromonitoring (c-IONM) with the Saxophone Electrode®.
Methods: Constant tensile stress was applied to left and right recurrent laryngeal nerves in pigs (n = 14). In group I (n = 18 RLN) tensile stress of 1.2 N was applied until the signal amplitude was reduced by 85%. In group II (n = 10 RLN) 0.34 N were maintained for 10 minutes.
Results: In group I we found a great variation of the duration to attain 85% change of signal (COS) inter- and intra-individually, median 22 min (range 2–116 min). Left and right side do not differ significantly (3-68 min respectively 2–116 min $p = 0.18$). However in each individual animal there appear to be a vulnerable (2–14 min), and a less vulnerable nerve (30-116 min). These differences become highly significant at 85% COS ($p < 0.001$), where the vulnerability is 1.5 to 51 times higher on one side (mean 4.9 times).

In group II tensile stress of 0.34 N ± 0.07 for 10 minutes lead to decrease of signal by 27-96% (median 38%) in the more vulnerable nerve and 0-27% (median 7%) on the contralateral side. The more vulnerable side showed a signal decrease that was median 31% (26-68%) higher than the less vulnerable side ($p = 0.008$).
Conclusion: In our study each individual animal appears to have one RLN that was 4.9 times more vulnerable than the contralateral side.

The differences were highly significant at 85% COS ($p < 0.001$). The more vulnerable nerve does not seem to have a side preference.

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788
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Miscellaneous Topics 4
Does peritoneal dialysis prevent uremic neuropathy? Study of twenty patients

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Background: Neuropathy is a common complication of end-stage kidney disease (ESKD). Its prevalence ranges between 60% and 100%. The improvement of uremic neuropathy with peritoneal dialysis (PD) is controversial.
Methods: This trans sectional study was undertaken on 20 patients with ESKD treated by PD in the dialysis center. None of the patients had a history of exposure to toxins or other illnesses known to cause neuropathy such as diabetes mellitus, amyloidosis. All patients were reviewed with recording of history, neurological examination, biological assessment and nerve conduction studies (NCS).
Results: The mean age was 48.4 yrs with sex-ratio (M/F) 2.3. Continuous ambulatory PD was more frequent used than automated PD (57.9%). The mean dialysis duration was 26 months. Chronic interstitial nephropathy was dominant (30%). Neurological symptoms were found in 45% : distal paresthesias (5 cases) and signs of autonomic dysfunction(orthostatic hypotension, diarrhea, sweating) (4 cases). The examination noted distal hypoesthesia in 2 patients. No motor deficit was observed. Seven patients had axonal polyneuropathy, it was sensory in 6 cases (lower limbs: 4 cases, 4 limbs : 2 cases) and mixed sensory motor in one case. One patient had carpal tunnel syndrome. NCS were normal in 60%. Among the 5 patients with distal paresthesias only 2 had PN.
Conclusion: PD seems to be advantageous comparatively to hemodialysis, it reduces the risk and the severity of uremic neuropathy however, the prevalence remains high.

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789
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Miscellaneous Topics 4
Nerve Lesions In Knee Dislocations

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Nerve injury represents one of the most serious complications of knee dislocation. The incidence is higher than generally expected. Meta-analysis of the research so far reveals that among the patients with knee dislocations and lesions of menisci and ligaments of the knee, up to 25% suffer nerve injuries. The highest nerve & vascular injury prevalence was observed in knee injuries that involved the ACL, PCL, and medial collateral ligament (32%) and posterior dislocation (25%), respectively. Detailed clinical examination of motor strength, sensory function and trophic changes of correspondent muscles are crucial for right diagnosis. Standard electrodiagnostic (EDX) workout should follow, including electromyographic (EMG) testing of m. tibialis ant., m. extensor digitorum brevis, m. gastrocnemius and m. flexor hallucis, motor nerve conduction velocities (MCV) of common peroneal and tibial nerve, sensory nerve conduction velocities (SCV) of superficial peroneal and sural nerve, along with F-waves and H-reflex. The most common among nerve injuries is the lesion of common peroneal nerve (CPL) (25% of dislocations). The lesions may be complete or partial. Operative treatments include neurolysis, transposition, neurorrhaphy, nerve transfer and tendon transfer. Non-operative treatments include physical therapy and pain management. Earlier treatment yields better success rates, especially considering neurolysis and grafting procedures.

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790
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Miscellaneous Topics 4
A case of late onset Fragile X Syndrome

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Fragile X syndrome (FXS) is the most common inheritable cause of intellectual disability and genetic cause of autism spectrum disorders. It is a X-linked dominant disorder which is associated with expansion of CGG repeats affecting the Fragile-X mental retardation 1 gene (FMR1). Symptoms of FXS range from learning disabilities to more severe cognitive disabilities primarily in childhood. We report a 80 years old male who was a product of normal pregnancy and delivery and achieved all the developmental milestones. The patient was asymptomatic until 15 years ago when he first noticed difficulty with balance. His symptoms gradually progressed to a point where he was wheelchair bound by the age of 70 years when he was first seen at 75 years. His neurological examination revealed normal mental status and intact cranial nerves. Motor examination was normal but sensory testing showed absent vibration and proprioception in bilateral lower extremities. The reflexes were absent in ankles bilaterally with upgoing toes. He demonstrated slow shuffling gait with wide stance and frequent near-falls without support. Routine blood tests and CSF studies were negative for any infectious or inflammatory causes of peripheral neuropathy. Electrodiagnostic studies revealed sensory predominant axonal sensorimotor peripheral polyneuropathy. MRI brain revealed microvascular ischemic changes with generalized volume loss. This cervical and lumbar MRI showed diffuse degenerative changes. Genetic testing for heritable causes of ataxia revealed a premutation in fragile X gene (84 CGG repeats), confirming the diagnosis of fragile X-associated tremor/ataxia syndrome. We describe a rare case of late onset FXS.

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791
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Miscellaneous Topics 4
The effects of chronic administration of lithium chloride on anxiety and histopathological changes in the amygdala and hippocampus after trimethyltin intoxication

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Introduction: Trimethyltin (TMT) is an organotin neurotoxicant which causes hippocampal selective degeneration. TMT intoxication is the cause of psychological disorders in human and rodents.

Objectives: To investigate the relationship among BDNF serum as well as hippocampal level, Amygdala and hippocampus histo-pathological changes and the occurrence of anxious and exploratory behavior in Sprague Dawley TMT-treated rats.
Method: First, TMT (8 mg/kg) was injected intra-peritoneally (i.p) to all rats. The test groups received 20, 40 and 80 mg/kg of lithium chloride (LiCl) respectively and the sham group received saline for 14 days. The elevated plus maze, dark–light box and open field tests were conducted in order to investigate the anxiety symptoms and exploratory behaviors. Then, the BDNF serum and hippocampal levels were measured using ELISA procedure. This was to study the correlation between the anxiety symptoms with the neurotrophin levels and histopathological changes and neuronal density in different hippocampus subdivisions, including CA1, CA2, CA3 and dentate gyrus (DG) and the amygdala in both hemispheres.
Results: Findings indicated an increase in anxiety behaviors and a decrease in exploratory ones. In addition, the BDNF hippocampal and serum levels were decreased in the TMT-treated rats. Also, an increase in necrotic areas and a decrease in neuronal density in different regions of the hippocampus and amygdala in both hemispheres were seen after Trimethyltin intoxication.
Conclusion: LiCl, having sufficient neuroprotective effects, could be used as a solution to manage the anxiety symptoms and exploratory behaviors comorbidity with hippocampal degeneration after TMT intoxication.

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792
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Miscellaneous Topics 4
Deficient neuroscience teaching in the basic science curriculum, is this the origin of the neurophobia?

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Background: The deficit in the teaching of neuroanatomy during medical school has been proposed as the main cause of neurophobia or the fear of neurosciences. This may compromise the care of patients with neurological conditions by physicians.
Objective: To determine the perception of the amount of teaching oriented to the nervous system provided during medical school in comparison to other systems in the basic science curriculum.
Material and methods: A cross sectional study was done in medical schools of four major cities in Ecuador (Guayaquil, Quito, Cuenca and Manta). A survey was sent to the students of these medical schools about the perception of the amount of teaching oriented to the nervous system in comparison to other systems in each of the following basic sciences: embryology, histology, physiology, pathology, pharmacology, pathophysiology and immunology. The students were asked to qualify the teaching as outstanding, acceptable, insufficient or non-existent.
Results: A total of 849 students completed the survey. Of these, 4.5% considered as outstanding the amount of teaching received oriented to the nervous system in the different basic sciences. 33.2% considered it acceptable, 40.2% insufficient and 22.1% as non-existent.
Conclusion: Most of the surveyed students consider teaching on the nervous system in comparison to other systems deficient. Almost two thirds considered the teaching they received either insufficient or non-existent. More research is needed to determine the contribution of deficient neuroscience teaching in the basic science curriculum to the origin of neurophobia.

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793
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Miscellaneous Topics 4
Tacs in theta range improves the hit rate and general accuracy in a spatial working memory task

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Background: There is a significant body of evidence that theta rhythm in the prefrontal cortex plays an important role in working memory processing. However, it is unclear whether direct magnification of theta oscillations in this region will impact accuracy of working memory performance.
Objective: To investigate the effect of the increased theta activity in the left DLPFC on visual-spatial working memory.
Material and methods: Sixteen healthy right-handed volunteers (9 females, age range: 20–26 years) participated in this randomized, placebo-controlled, crossover study. Each of them completed a 2-back match-to-sample visual-spatial memory test during transcranial alternating current stimulation (tACS; 10 min, 1 mA peak-to-baseline, 6 Hz) and sham stimulation. A Laplacian electrode-montage was used (5 electrodes, central position – AF3) to ensure focality of the stimulation. Resting state EEG was recorded before and after each session.
Results: We observed a significant increase of the hit rate in the working memory test during stimulation ($Hit_{control} = 70.8 \pm 3.5\%$, $Hit_{stimulation} = 74.3 \pm 3.5\%$; $p = 0.02$, $g = 0.25$ (paired T-test and Hedges' g statistics)) but no effect on the false alarm rate. General accuracy improved from $76.1 \pm 2.4\%$ to $78.0 \pm 2.5\%$ ($p = 0.04$, $g = 0.2$). Power analysis of the EEG revealed an increase in the low frequency range (6–12 Hz) after the stimulation in comparison to sham ($p = 0.045$, $g = 0.49$).
Conclusions: tACS at 6 Hz over the left DLPFC during the visual-spatial working memory test increases the theta power and improves memory accuracy.

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794
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Miscellaneous Topics 4
Intraepidermal nerve fibre density (ienfd) in the assessment of central nervous system disease

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Background: Reduction of intraepidermal nerve fibers density (IENFD) may be an early indicator of small diameter nerve fiber pathology. Skin biopsy to measure IENFD is a diagnostic method used internationally. There are few studies that have looked at IENFD in patients with central nervous system diseases with and without neuropathic pain.
Methods: 3 mm punch biopsies 10 cm above the lateral malleolus. 3 sections of 50um were analized using monoclonal antibody PGP 9.5 (UltraClone Ltd, UK). Nerve fibres crossing the dermoepidermal junction were counted and expressed as intraepidermal nerve fibre

density (IENFD) = Number of IENF/mm. The study recruited 22 healthy subjects (S) and 12 patients with central nervous system disease involving spinothalamic sensation (PC): 9 with neuropathic pain (PCD), 4 painless (PCSD). T tests were performed (Stata 12.0). The Bonferroni correction was used for multiple comparisons. The significance level was set at $p < 0.005$. Interobserver agreement was assessed with the Bland-Altman method.

Results: IENFD: (S): 6.89 +/− 2.14, (PC): 4.52 +/− 2.21, (PCD): 4.37 +/− 2.62 (CSDP) 4.84 +/− 1.36. Mean IENFD was significantly lower in PC group compared to controls ($p < 0.001$). There were no significant differences between patients with and without neuropathic pain ($p = 0.84$).

Conclusion: Patients with central nervous system diseases showed a decrease in IENFD compared to controls. A larger number of controls and patients is needed to compare values by gender and age and by patient subgroups.

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795
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Miscellaneous Topics 4
Multimodal neuroimaging analysis (tractography, volumetry and functional) in a series of tbi patients with neuropsychiatric sequelae

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Introduction: Mild TBI is under reported in epidemiological studies that neuropsychiatric sequelae are not as visible; however, the literature indicates severe neuropsychiatric disturbances in the medium and long term that significantly affect quality of life. The present study attempts to show that the multimodal analysis (clinical data and different types of neuroimaging) is useful to understand and explain the neuropsychiatric sequelae.

Methodology: An analytic observational case–control study was conducted. Psychiatric depth interviews were conducted; neuroimages were acquired of diffusivity (DTI), fMRI and structural. A multimodal neuroimaging analysis and fusion was performed. These data were subsequently associated with clinical and neuropsychology information.

Results: The multimodal analysis allowed creating an explanatory model of neuropsychiatric disorders in the sample. Tractography analysis identified that the frontal networks and the lower longitudinal fasciculus showed a decrease in the fraction of anisotropy. The volumetry and segmentation showed decrease in orbitofrontal regions and the temporal pole; fMRI analysis showed less activation in the superior temporal gyrus (posterior segment) in patients versus controls. These data variables were associated with behavioral change and empathy.

Discussion: The consequences of mild TBI affect a significant number of patients and decrease the quality of life. The use of multimodal techniques and clinical correlation are essential tools for the diagnosis and prognosis.

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796
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Miscellaneous Topics 4
The role of klotho in the regulation of adult hippocampal neurogenesis

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Introduction: Adult hippocampal neurogenesis (AHN) is a neuronal plasticity mechanism that depends on the proliferation and differentiation of neuronal progenitors cells. AHN decreases with age and is important for cognitive processes such as learning and memory. Klotho, an aging-related protein, is expressed in the adult hippocampus and in non-neuronal tissues has been shown to regulate progenitor cell proliferation.

Objective: To determine the effect of hippocampal Klotho expression on AHN in mice.

Methods: Klotho expression in mouse hippocampus was characterized by immunofluorescence. Lentiviral particles expressing short hairpin RNA (shRNA) to downregulate klotho expression were injected in the hippocampus of adult mice (Group 1 shRNA control; Group 2 shRNA-klotho-interferent-1; Group 3 shRNA-klotho-interferent-2; n = 4 mice/group:). AHN was quantified as the number of cells expressing markers Ki67 and doublecortin in the subgranular zone of the hippocampus. The percentage of BrdU-nestin cells in adult mouse neural progenitor cells in culture (AHNPC) stimulated with klotho recombinant protein was determined. Averages were compared using Mann–Whitney test.

Results: Klotho protein is expressed in the dentate gyrus of adult mice hippocampus. Inhibition of hippocampal Klotho expression decreased the number of Ki67 (shRNA control: 948 +/− 171, shRNA-interferent-1: 456 +/− 96, shRNA-interferent-2: 750 +/− 88 $p < 0.05$), and doublecortin (shRNA control 1821: +/− 372, shRNA-klotho-interferent-1: 69 +/− 11, shRNA-klotho-interferent-2 594: +/− 42 $p < 0.05$) positive cells. Recombinant Klotho protein stimulation increased the proliferation of AHNPC (13.4% +/− 4,2 control; 21.7% +/− 6.1 klotho stimulated, $p = 0{,}0079$).

Discussion: These results suggest that Klotho participates in the regulation of AHN in mice.

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797
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Miscellaneous Topics 4
Magnetic- stimulated motor evoked potentials in adult tethered cord syndrome pre-operative and post-operative evaluation

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Introduction: Tethered cord syndrome (TCS) is a progressive condition presenting with various neurological symptoms such as

bladder/bowel disturbance, motor involvement and pain along with cutaneous signs, orthopedic deformities.

The diagnosis of TCS is mainly clinical although radiological investigation, spinal somatosensory evoked potentials (SSEP). recordings and urodynamic studies are equally important for both diagnosing the condition also in decision making for the treatment.

Aim (objectives): In this study, aims to investigate the role of motor evoked potentials (MEP) in TCS which is to our knowledge never been investigated before .Twenty patient over 18- years old with definitive TCS diagnosis were included for the study.

Method: Maglite compact transcranial magnetic stimulation machine is used to in order to stimulating the spinal cord and motor cortex for MEP recording. Nerve Conduction Studies were performed in pre-operatively and post-operatively in order to exclude a motor neuropathy.

The motor conduction time to thenar and tibialis anterior muscles,the latencies and the amplitudes of the motor responses recorded and compared with the normal laboratory values.

Results and discussion: It has been found that all the preop-postop MEP values of the twenty patients were in normal limits. We conclude that magnetic stimulation test in TCS is of no use in evaluating the tethered spinal cord.

We propose that SSEP investigation is still of value in identifying and in follow up of TCS.

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798
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Miscellaneous Topics 4
Clinical-eletrographic discordance: A diagnostic and therapeutic challenge

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Background: It is established that electroencephalogram (EEG) is a useful tool to predict recurrence of seizures, and a EEG without abnormalities infers good prognosis. However, it is necessary caution regarding interpretation of interictal activity. The objective is illustrate three cases, with apparent seizures control, although evidence of electrographic worsening.

Results:

Case 1: LCM, male, 8 years, seizures started at five years old, described as recurrent vomiting, followed by loss of consciousness. Brain MRI was normal. EEG showed bilateral occipito-temporal epileptiform activity. Carbamazepine (CBZ) was introduced, reaching seizures free. EEG revealed subsequent increasing of epileptiform discharges incidence.

Case 2: DLS, male, 6 years, focal seizures, monomorphic, during seconds, started at three years old, characterized by behavioral stop and right oculocephalic deviation. Brain MRI was normal. EEG with frequent focal (temporal) and generalized epileptiform activity. Oxcarbazepine was introduced with resolution of seizures. EEG showed elecctrographic worsening.

Case 3: MESP, female, 12 years, reporting febrile seizures that started at 8 months. At age 11, he used to have daily seizures, described as behavioral stop and oculo-automatism, during a few seconds. Brain MRI revealed right hippocampal atrophy. EEG with frequent right temporo-occipital epileptiform activity. CBZ was introduced, and seizures free was reached. EEG control revealed electrografic worsening.

Conclusion: These cases illustrate that not always is possible establish an association between clinical and electrographic worsening, which should generate questions about accuracy of parents information, and limitations of EEG. Thereby, Ictal Monitoring in therapy decision is mandatory.

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799
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Miscellaneous Topics 4
For effective utilization of dat-spect with 123i-ioflupane in clinical practice

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Background & objective: Effective utilization of DaT-SPECT with ^{123}I-ioflupane (DSI), approved in Japan in Jan 2014, has not been discussed on clinical grounds. Considering its expensiveness and time-consumingness, we think it important to select patients who most profit from it.

Patients and methods: Medical charts of patients who had taken DSI from March '14 to April '15 were retrospectively analyzed about reasons of its ordering, disease duration, its results, MIBG data if available, and latest clinical diagnosis. Evaluation of DSI was performed visually based on Food & Drug Administration prescribing information.

Results: Ninety-six (male 46, female 50) patients have taken DSI. Numbers of patients who showed decreased striatal uptake/total patients taken DSI in each diagnosis were as follows; 41/41 in Parkinson's disease (PD), 1/8 drug-induced parkinsonism (DIP), 7/7 Lewy body dementia, 6/6 progressive supranuclear palsy, 3/6 parkinsonism of undetermined etiology, 5/5 multiple system atrophy, 3/5 vascular parkinsonism, 3/3 corticobasal syndrome, 1/3 essential tremor (ET), 1/3 normal pressure hydrocephalus (NPH), 2/2 primary progressive freezing gait, 1/2 idiopathic REM sleep behavior disorder (iRBD), and 1/5 in other diseases. Of PD patients, 8/22 had disclosed preserved H/M ratio (≧1.7 in late phase) in MIBG study and 3 have been transformed from ET.

Conclusion: DSI is useful in confirming organic damage to nigrostriatal system under restriction of poor discrimination among various parkinsonian disorders. Our data suggest probable efficiency of DSI in differentiation of DIP from depression associating with PD and in evaluation of NPH sometimes mimicking parkinsonism or iRBD and ET both frequently progressing into synucleinopathy.

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800
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Miscellaneous Topics 4
Evaluation of microchimerisms in neurodegenerative disorders

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Neurodegenerative Disorders constitute a key public health problem that affects nations worldwide. Previous studies suggest the presence of microchimerisms in brain tissue as a protective factor against Alzheimer´s Disease (AD), due to the fact that these are fetal cells in immature stages of specialization and their function may be compared to that of stem cells. Nonetheless, microchimerims have

been related as a risk factor in numerous other diseases. This descriptive study, compares the frequency and distribution of microchimerisms in brain tissue with neuropathologic diagnostic of AD and Fronto Temporal Dementia (FTD), with healthy brains. Samples were taken from 15 brain regions related to neuropathology. 34 brains from patients diagnosed with AD and FTD, along with a control group were selected for histological analysis and recollection of clinical information in order to determine the state of the disease. Next, the experimental group was genotyped to detect microchimerisms through specific X and Y chromosome markers without concluding results. Nowadays, microchimerisms are being detected through the DYS 14 gene and autosomic markers DIP-STR. Further, an association between prevalence of microchimerisms and the state of the disease will be made through a chi-square. The expected results are a high number and distribution of these cells in healthy brains, with tendency to descend, as the characteristic neuropathological damage of neurodegenerative disorders increases.

An Institutional Review Board (IRB) and/or Animal Use Committee have waived the requirement for their formal approval of the study.

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801
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Miscellaneous Topics 4
Including analysis of acid alpha glucosidase in screening for muscle disease in Cuiabá, Brazil

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Introduction: Pompe disease is an autosomal recessive disorder that caused by enzymatic deficiency, resulting in accumulation of glycogen in lysosomes, and changing particularly, cardiac, respiratory and skeletal muscle tissues. Clinical manifestations may appear in all age ranges. Late form can be similar to other neuromuscular diseases, making it hard to diagnose.
Objective: Identify, among patients with progressive muscle weakness treated at HGU neurology outpatient service, those with Pompe disease.
Methodology: It is a descriptive analysis with observational clinical trial by means of a cross-sectional study of 150 patientsoutpatient clinics ofchild neurology, neurology and genetics followed in *Hospital GeralUniversitário*(HGU) *from Cuiabá* – Brazil 2010/2015. We conducted a screening program in all patients with suspected of neuromuscular disease. The DBS was used in all patients. Inclusion criteria were conditions of progressive muscular weakness, frequent fallings, hyper-CK-aemiaand that showed even the slightest decreased muscle strength at the physical examination.
Results: The sample consists of 150 patients with mean age of 15,4years (1 month/73 years old), 54,6% were male. The screening identified eleven patients with Pompe Disease (7,3%) and 6 patients (4%) who had borderline results. The DBS test was negative for the remainder of samples, these 27,7% were diagnosed with progressive muscular dystrophy, 1,3% with myasthenia gravis, 4,6% Spinocerbelar Ataxia and 10% with spinal muscular atrophy.
Conclusion: The late onset form of Pompe disease is similar to other neuromuscular diseases; the research of acid alpha glucosidase in this group must be reviewed, since studies show that once the enzyme replacement therapy is started, disease prognostic is changed.

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Miscellaneous Topics 4
Electroencephalogram profile in a brazilian private clinic – Analysis of 2437 examinations

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Background: The EEG is a non-invasive and widely used test in neurology, mainly performed to determine the pattern of brain activity, its symmetry and epileptiform abnormalities.
Objective: To describe the EEGs findings in a cohort of Brazilian patients, highlighting the percentage of epileptiform activity according to the indication of the test.
Methods: This is a retrospective study. We analyzed 2437 EEGs performed from 2013 to 2015 in a private neurological clinic in Brasília, Brazil.
Results: The mean age in the study was 19 years, 59% of the patients were masculine and 41% were feminine. From the 2437 EEGs, epileptiform activity was present in 203 EEGs (8.3%), mainly focal (71%) and in central regions (79%). Of the 546 EEGs performed as a part of a health examination for work or school entry, 1% showed epileptiform discharges (ED). Of the 509 patients with suspected attention/hyperactivity disorder (ADHD), 10% had abnormal EEGs and 6% showed ED. Of the 296 patients with headache, 4% had showed ED. Of the 52 patients investigating syncope, 3% showed ED. Of the 27 patients with autism spectrum disorder, 15% showed ED.
Conclusion: The results showed 84.2% of normal EEGs. The results of this study confirm the findings in the literature of the percentage of asymptomatic patients with epileptiform activity in the EEG (1%). Patients with autism spectrum disorder and ADHD had a lower percentage of epileptiform findings than the reported in the literature.

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803
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Miscellaneous Topics 4
Electrophysiological visual pathway assessment in correlation with brain mr and mr-spectroscopy in patients with hashimoto's disease

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Background: In Hashimoto's disease (HD) the disturbances of central and peripheral nervous system could be seen even in euthyroid status.
Aim: The aim of the study was the assessment of visual pathway in visual evoked potentials (VEPs) in euthyroid HD patients without clinical neurological deficit, and ophthalmological abnormalities.
Material and methods: 100 euthyroid patients with HD (92 women, 8 men, mean age 46.3 years old) and 50 healthy controls, sex and

age-matched were studied. Clinical neurological and ophthalmological examinations, VEPs, MRI, and MR-spectroscopy were performed.
Results: In about 25% we revealed abnormal VEPs. In the study group the mean P100 latency was significantly longer, and the mean amplitude P100/N145 was significantly higher than in controls. There were no correlations between VEP parameters and thyroid hormones, and anti-thyroid antibodies levels, but in most patients incorrect VEPs correlated with MR-spectroscopy abnormalities.
Conclusions: In euthyroid HD patients without clinical central nervous system impairment the visual pathway disturbances could be found.

Significantly higher amplitude of VEPs in HD patients may indicate the cortex hyperactivity.

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Miscellaneous Topics 4
Four days in Ghana: Teaching a neurology continuing medical education course to Ghanaian physician assistants

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Background: In high-income countries there are approximately 3 neurologists per 100,000 population whereas in lower income countries that proportion drops to 3 neurologists per 10,000,000. Thus, in much of the world primary care providers are left with the burden of treating neurological disease.
Objectives: To evaluate the effectiveness of a neurology continuing medical education course taught by University of Utah neurology faculty to Physician Assistants in Ghana.
Methods: Pre- and post- course surveys assessed self-perceived confidence in ten areas of clinical neurology including the neurological exam, stroke, epilepsy and central nervous system infections. A Likert Scale allowed participants to rate from 1 ("not confident") to 5 ("very confident") their comfort with topics before and after the course was completed. The Exact Wilcoxon signed rank test was used to analyze survey data.
Results: A total of seven ($n = 7$) Ghanaian physician assistants enrolled in a four-day neurology course. Total increased self-confidence in the ten topics surveyed pre- and post- course almost reached statistical significance ($p = 0.08$). The one area that did achieve statistical significance was increased self-confidence with neurological exam skills after course completion ($p = 0.02$).
Conclusions: The provision of education in the diagnosis and treatment of neurological conditions to primary care providers is one intervention that can address the limited supply of neurologists in lower income countries. This study suggests that a continuing medical education course can increase participant self-confidence in major topics in neurology and that the most valuable contribution of an educational intervention could be instruction in the neurological exam.

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805
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Miscellaneous Topics 4
Impact of hemodialysis length and duration on neurophysiological parameters

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Goal: To examine the effect of the hemodialysis duration of on neurophysiological parameters in patients.
Material and methods: The study was a prospective, observational and analytical, conducted at the Neurology Clinic, Clinical Center of Sarajevo University. The study included 38 patients treated with hemodialysis at a Clinic for Hemodialysis and all have placed an AV fistula. All respondents underwent EMG analysis of peripheral nerves in the limbs which included: estimation of parameters of motor median, ulnar, tibial and peroneus nerve, F wave and H reflex as well as the sensory neurographs-neurogram of the median, ulnar, peroneal and sural nerve.
Results: HD duration is correlated with distal motor latency of median nerve rho = 0.307; $p = 0.017$. Duration of HD is correlated with MBP median nerve rho = −0,275; $p = 0.007$. Values of MBP decreases with years spent on HD. Also HD duration is correlated with MBP of the ulnar nerve rho = −0.233; $p = 0.042$. MBP values of ulnar nerve are slowing down with the years spent on hemodialysis. HD duration is correlated with amplitude in tibial nerve rho = −0.352; $p = 0.030$. The values of the amplitude in tibial nerve decreases with years on HD. Length of HD is correlated with the H-reflex of the sural nerve rho = 0.485; $p = 0.012$. Values of H reflex latency extending with HD duration.
Conclusion: The duration of hemodialysis treatment has an impact on the neuro physiological. The reason may be the effect of beta 2 microglobulin in these patients and are consistent with other known studies.

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Miscellaneous Topics 4
Case report of a young adult whit vanishing white matter disease (VWM)

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VWM is the most prevalent inherent childhood leucoencephalopathy. It is a slow progressive disease whit a wide variation on phenotypic presentation. His MRI findings are characteristic. The basic genetic defects reside in one of the five subunits of eukaryotic translation initiation factor eIF2B. We present a 24 years old female patient, with secondary amenorrhea since she was 14 and some learning disability in high school years. She presented in the last six months fluctuating gait instability. Recently, working under high temperature conditions, she suffered severe bifrontal headache, associated with gait instability, mental confusion and speech problems. A brain MRI was performed showing whole brain atrophy and a diffuse leukoencephalopathy whit small cavitation next to the frontal horn of the right lateral ventricle. No gad enhancing lesion was detected. The rest of the laboratory tests (including CSF) were normal. Normal saline hydration and iv NSAIDs were started. 24 hours after, the patient was normal again. The genetic study is still pending.

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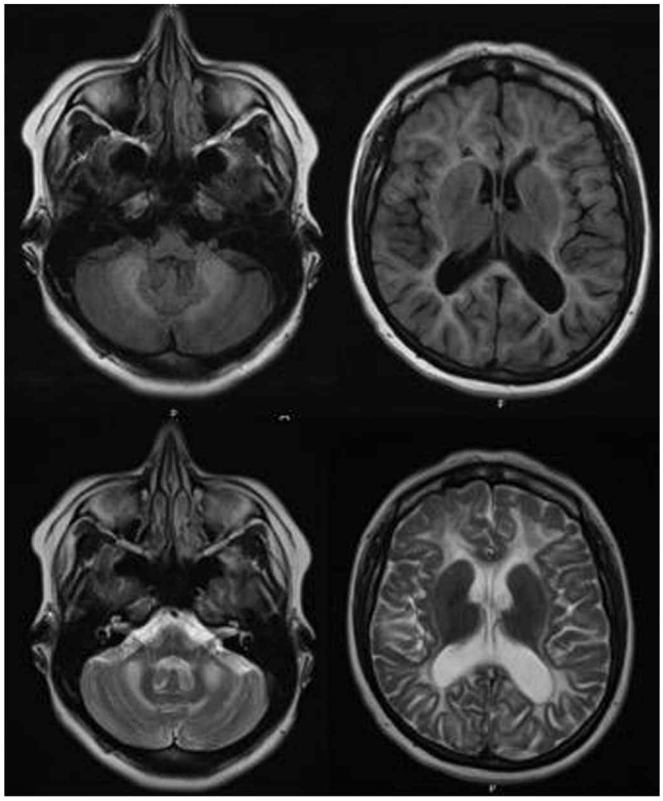

807
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Miscellaneous Topics 4
III Nerve palsy caused by intracraneal hypotension secondary to lumbar laminectomy. A case report

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Intracranial hypotension syndrome (IHS) appears with a decrease in pressure of the cerebrospinal fluid (CSF) below 65–60 mmH2O. IHS can be spontaneous (idiopathic) or symptomatic (post-lumbar puncture, post-surgical, post traumatic or in systemic disease). Orthostatic headache is the most common symptom. HIS is usually associated to VI nerve palsy and III nerve affection is exceptionally rare. Only a few case reports of intracranial hypotension associated with oculomotor nerve palsy are described.

We report a case of a 54 year old man with recent history of lumbar disc hernia surgery consults for one week history of progressive orthostatic headache and diplopia. Neurologic examination showed medial rectus palsy, ptosis and less reactive midriasis plus L5 left radiculopathy. Ocular inspection of lumbar region put over a tender, circumscribed inflammatory lesion in the surgical site.

Brain MRI showed a diffuse leptomeningeal thickening and right frontal cortical vein thrombosis. Lumbar Spine MRI evidenced a pseudomeningocele secondary to a lumbar fistula in the surgical site. Anticoagulation was started and rest was indicated. However, a month after the pseudomeningocele increased in volume, and surgical repair was made.

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Miscellaneous Topics 4
Morphological changes of the retina and visual functions in demyelinating diseases of the cns

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Background: New findings demonstrate an involvement of gray matter from the early onset of the multiple sclerosis (MS). In patients with optic neuritis (ON) reducing of the neural layer of the retina (retinal nerve fiber layer, RNFL) compared with healthy controls is presented. This is on the average about 20.38 um. RNFL thinning is registered even in patients with MS without ON (7.08 um).The reduction of the RNFL in association with a reduction in visual acuity is measured by Snellen chart.
Objectives: The aim of the work is postgraduate longitudinal monitoring of visual acuity, contrast sensitivity, VEP induced stimulations type of pattern reversal and motion onset RNFL in patients with demyelinating disease (with a focus on the initial stages) to establish the links between the monitored parameters and the clinical status of the patient and any prediction of its development.
Methods: File so far examined patients comprise a total of 18 patients, 6 males (age 33 ± 10 years) - 14 patients with a first clinical demyelinating attack (of which 3 ON) and 4 more episodes (2 ON).
Results: Preliminary statistical analysis of the relationship between morphological and functional findings demonstrated a statistically significant negative correlation between RNFL thickness and VEP latency (Spearman R from -0.65 to -0.57) and positive relationship with the amplitude (R from 0.48 to 0.62).
Conclusions: Negative correlation of reduction RNFL and P100 latency is consistent with literature .Correlation between reduction of amplitude P100 and RNFL was not confirmed.

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809
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Miscellaneous Topics 4
Unusual blink reflex: Absent r1 response on ipsilateral supraorbital nerve stimulation but present during contralateral stimulation

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Background: Early or R1 motor response from the orbicularis oculi (o. oculi) muscle is obtained only unilaterally from ipsilateral supraorbital nerve stimulation, while the late or R2 response is present bilaterally during ipsilateral and contralateral stimulation.
Case material: A 29-year-old woman developed horizontal diplopia, left facial numbness, and occasional left eyelid twitching. Otherwise, she was healthy. Examination showed left 6th cranial nerve palsy and decreased left facial sensation. There was no blepharospasm, myokymia, or facial weakness.
Electrophysiologic findings: Peripheral motor nerve conduction study of both facial nerves and needle EMG of left facial muscles showed no abnormality. Blink reflex study revealed no R1 response from the left o. oculi muscle on ipsilateral supraorbital nerve stimulation, but present during contralateral stimulation (latency = 11.4 ms). R2 responses from the left o. oculi muscle showed normal latencies during contralateral (31.2 ms) and ipsilateral stimulation (30.8 ms) stimulation. R1 latency from the right o. oculi muscle was 10.0 ms, and the R2 responses showed normal latencies during contralateral (31.0 ms) and ipsilateral (31.9 ms) stimulation.
Conclusions: Absent R1 response during ipsilateral stimulation suggests abnormalities in the main sensory nucleus of the pons.

Presence of an R1 response from the o. oculi muscle during contralateral supraorbital nerve stimulation is unusual. This may indicate cross excitation of the facial nucleus from contralateral trigeminal sensory nerve impulses that are not involved in the usual multi-synaptic pathways responsible for eliciting the R2 contralateral response. Perhaps this is a result of focal CNS demyelination.

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810
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Miscellaneous Topics 4
The effect of glutamate on the neuronal differentiation of neural stem cells

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Background: Our previous study showed that NSCs in peri-injured cortex of adult rats with TBI could differentiate into immature neurons, but they failed to grow up to mature neurons.
Objective: To investigate effect of glutamate (Glu) on neuronal differentiation of NSCs.
Methods: NSCs were cultured 14 days in four groups: control group, Glu group, Glu + MK-801(0 h) group and Glu + MK-801(24 h) group. Differentiated neurons were observed by DCX or MAP-2 immunofluorescence, cells apoptosis was detected by TUNEL kit. The levels of NR1 mRNA and protein of cells in control and Glu + MK-801(24 h) groups were detected by real-time PCR and Western Blot.
Results: After 1 day, the number of DCX^+ cells in the Glu and Glu + MK-801(24h) group was more than that of control and Glu + MK-801(0h) group. After 7 days many DCX^+ cells were still found in the Glu + MK-801(24h) group and more than that of other three groups. After 14 days, many $MAP\text{-}2^+$ cells were found in the Glu + MK-801(24h) group and more than that of other three groups. The percentage of $TUNEL^+$ cells in the Glu group peaked at 3rd day then decreased and was still more than that of the other three groups. The expression of NR1 mRNA or protein in the Glu + MK-801 group increased gradually and peaked at 7th day then kept in a sable state and was more than that of control group.
Conclusion: Glutamate can promote NSCs differentiation into immature neurons then cause cells apoptosis which maybe results from increasing expression of NMDA receptor in the neuronal development.

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Miscellaneous Topics 4
Association of drd3 gene polymorphisms with parkinson's disease (PD)

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Background: PD is one of the most serious and widespread neurodegenerative diseases characterized by chronic progressive course, dysfunction of the basal ganglia and severe disability of patients. Genetic susceptibility plays a significant role in the development of PD.
Objective: To investigate associations between DRD3 dopamine receptor gene tag single nucleotide polymorphisms(SNPs) and the risk of PD development.
Material and methods: In this study ten tag SNPs(rs963468, rs2134655, rs9817063, rs324035, rs11721264, rs1800828, rs3773678, rs167770, rs167771, rs7633291) within DRD3 have been genotyped in 143 PD patients and 96 healthy individuals in Russian population. The diagnosis of PD was made using the criteria established by UK Brain Bank. The severity of PD was estimated using Hoehn&Yahr Scale. PD patients and control group were homogeneous by age, race and ratio of men and women(mean age: 69.8 ± 8.6; mean PD duration: 6.9 ± 3.6; women:men = 4:3).
Results: Significant associations of 4 polymorphisms(rs11721264, rs3773678, rs167771, rs324035) with PD have been found (Table 1).
Conclusions: Our study confirms polymorphic features of dopamine receptor's genes and their involvement in the pathophysiology in PD.

Table 1
The genotype's distribution of polymorphic variants of DRD3 gene in PD patients and healthy individuals.

Polymorphisms	Genotype	PD patients(n,%)	Healthy control subjects(n,%)	χ2	P
rs11721264	AA	14(11,2%)	22(28,9%)	10,265	**0,006**
	AG	54(43,2%)	28(36,8%)		
	GG	57(45,6%)	26(34,2%)		
rs3773678	AA	4(2,8%)	0(0%)	17,846	**0,0001**
	AG	30(21%)	44(45,4%)		
	GG	109(76,2%)	53(54,6%)		
rs167771	AA	103(73,6%)	60(67,4%)	6,368	**0,041**
	AG	31(22,1%)	29(32,6%)		
	GG	6(4,3%)	0(0%)		
rs324035	AA	5(3,5%)	0(0%)	8,018	**0,018**
	AC	32(22,5%)	35(36,1%)		
	CC	105(73,9%)	62(63,9%)		

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Miscellaneous Topics 4
Runx1t1 regulates the neuronal differentiation of neural stem cells from the rat hippocampus

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Background: Our previous studies showed expression of Runx1t1 increased during hippocampal neurogenesis. It was hypothesised that Runx1t1 involved the neural development in hippocampus.
Purpose: To investigate the function of Runx1t1 in the process of hippocampal neurogenesis.
Methods: Expression and localization of Runx1t1 were detected by real-time PCR, Western blot and immunocytochemistry during differentiation of hippocampal neural stem cells(NSCs) in vitro. Runx1t1 in hippocampal NSCs was knocked down by small interfering RNA (siRNA), and Runx1t1 overexpression was induced by LV-Runx1t1 in vitro, to explore the effects of Runx1t1 on the neuronal differentiation of hippocampal NSCs.
Results: Expression of Runx1t1 mRNA and proteins in hippocampal NSCs decreased significantly after their differentiation, mainly located in the immature neurons and mature neurons and was not observed in astrocytes. Only 3.6% NSCs differentiated into MAP-2 positive neurons after Runx1t1 knocked down, less than that of the control groups. Moreover, the length and number of neuronal process were also significantly reduced. In contrast, up-regulating

Runx1t1 expression in the NSCs, 32% cells differentiated into MAP-2-positive neurons with more and longer processes. The results indicated that Runx1t1 involves the neural differentiation of hippocampal NSCs.

Conclusions: Runx1t1 can promote the neural differentiation of hippocampal NSCs.

Keywords: Runx1t1, Hippocampus, Neural stem cells, Neuron, Differentiation

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