EPILEPSY

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COMPLEX FEBRILE SEIZURES: NEUROIMAGING AND ELECTROENCEPHALOGRAM UTILITY IN EMERGENCY

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INTRODUCTION: Febrile seizures (FS) are the most common type of childhood seizures, affecting 2% to 5% of children. . A third of them correspond to the complex type. The majority occurs in those older than one month of age. Although reliable data exist about Neuroimaging and Electroencephalogram utility in the initial management of this entity, it is still in discussion.

OBJECTIVE: to analyze neuroimaging and electroencephalography utility in initial management of CFS.

MATERIALS AND METHODS: Descriptive, cross-sectional study. We reviewed 486 medical charts, from 1 month to 6 years, admitted with FS diagnosis from 2008 to 2013. Complex febrile seizures (CFS), is one with focal onset, one that occurs more than once during a febrile illness or lasts more than 10 minutes. 87 children were included.

RESULTS: 18% of the children presented with CFS, the median age was 2 years. Focal features 13, 7%, prolonged duration 56, 3% and recurrent episodes within 24 hours 60, 9%. The 81, 6% as the second episode of FS. 70% had a neuroimaging and were normal. Electroencephalogram abnormality was found in 12 patients (66.6% focal paroxysmal activity and 33.3% with diffuse slow activity). Patients with these findings showed prolonged and / or recurrent crisis.

CONCLUSIONS: CFS mostly occurred in children under 2 years of age equally in both genders. They were prolonged and recurrent. Neuroimaging nor EEG findings did not modify initial management at the Emergency Department.

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MULTIMODAL INTRAOPERATIVE MONITORING IN SPINAL SURGERY: RETROSPECTIVE MULTICENTRIC ANALYSIS AND RECOMENDATIONS BASED IN 1890 CASES.

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Introduction: Multimodal intraoperative monitoring (MIOM) is a combination of neurophysiological techniques that simultaneously evaluate ascendent and descendent spinal tracts, as well as nerve roots, to preserve their integrity during various spinal surgeries. Although these techniques had been used worldwide for many years, there are few publications on their use and diagnostic power in Latin America.

Objectives: To evaluate the sensitivity and specificity of the different techniques of MIOM in preventing mielorradicular damage in spinal surgeries.

Material and methods: Retrospective analysis of MIOM from 1890 spinal surgeries performed between 2002 and 2012 in different centers, 432 was pediatric patients with a mean of 12 years. MIOM included somatosensory evoked potentials (SSEP), motor evoked potentials (MEP), free run electromyography (EMG) with root or pedicle screws stimulation, and eventually, epidural potentials. True and false positives and negatives were analyzed.

Results: The surgeries performed were mainly scoliosis 57.5%, followed by cervical 21.3%, lumbar 10.6%, thoracic 8.3% and others, including tumors or tethered cord 2.3%. MIOM sensitivity was 93% and specificity 97.5 %.

Conclusions: MIOM, adapted in each case to the type of surgery and its potential neurological damage, can be performed easily and reliably, with high diagnostic power, so its use in spinal surgeries is essential. Having an unified MIOM protocol for each procedure, along with a good communication between the neurophysiologist and the surgical team, can reduce the morbidity of these procedures to a minimum.

P87

EPILEPSY PHENOTYPE IN PATIENTS WITH DOUBLE CORTEX: AN ARGENTINE SERIES

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Introduction: Genetic migration cortical disorders represent an important cause of epilepsy and developmental delay. Subcortical band heterotopic (HSB) or double cortex, is a disorders characterized by band of grey matter within the white matter, between the cortex and ventricular space.

Purpose: To analyse the clinical and neuroradiological features of patients whit double cortex.

Materials and methods: A retrospective descriptive study was performed of 10 patients whit diagnosis of HSB from January 2000 until June 2013.

Results: The mean age was 22 years, the average age of onset, 3.5 years, 60% were women. 40% had focal seizures, 20% generalized seizures and 40% both. Most patients had symptomatic focal epilepsy related to localization 50 %, followed by 30 % unclassifiable and 20% Lennox Gastaut syndrome. All had refractory epilepsy and developmental delay. Eight patients with moderate developmental delay and 2 severe. In the electroencephalogram, 50 % had focal discharges, 40% both and 10% not present. The MRI showed diffuse involvement, 3 with anterior predominance and 2 with back predominance. The average number of drugs used was 6. Valproic acid was the most AED indicated (9p), followed by lamotrigine and carbamazepine (8p).

The semiology was related to the focal dominance of the double cortex. (p 0.0039)

Conclusion: We found an association between the seizure semiology and focal predominance double cortex. No significant difference was found in other clinical variables and neuroradiological subclassification.

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MORBIDITY IN EPILEPSY SURGERY USING GRIDS OR DEPTH ELECTRODES OR COMBINED IMPLANTATIONS

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Introduction: Epilepsy surgery is a treatment option for drug-resistant focal epilepsies. A percentage of patients need invasive neurophysiology (NFI) for adequate resection of the epileptogenic zone (EZ) and avoid undesirable neurological deficit (DN).

Objective: To analyze the complications of patients who needed NFI, grids (ECoG) or depth electrodes (SEEG) or both (ECoG + SEEG).

Materials and Methods: Retrospective observational study that included 47 patients, between 2005 and 2012.

We analyzed demographic data and complications (medical and DN). The DNs were classified into temporary (less than 6 months) and permanent. The permanent ND, was further associated to the implant or resection procedure (expected or unexpected)

Results: Nineteen patients were male, mean age of 21.1y/o (1-45) and 19/47 were pediatric. All were chronically implanted, 7 (15%) with SEEG, 17 (36%) with ECoG and 23 (49 %) with SEEG+ECoG.

Nineteen patients had transient ND: ECoG: 9(motor: 2; language:3; neglect:2; altered level of consciousness:2); SEEG: 2(motor:1; altered level of consciousness:1); ECoG+SEEG: 8(motor: 3, language: 1, neglect: 1, altered level of consciousness:3).

Ten patients (21%) had permanent ND: ECoG: 4, SEEG: 1, ECoG+SEEG: 5, 3/10 had not expected ND. Seventeen patients, suffered medical complications, 59% (10p.) with ECoG+SEEG: 35% (6p.) with ECoG and 6% (1p.) with SEEG. A patient with bilateral combined implant died.

Conclusions: We observed more frequent morbidity with ECoG+SEEG. The unexpected complications were observed with ECoG+SEEG and ECoG.

One patient who underwent ECoG+SEEG implant, died.

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SURGICAL TREATMENT OF STATUS EPILEPTICUS

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Introduction: Resective surgery appears to be a viable treatment option in a small minority of patients with convulsive or nonconvulsive refractory super status (RSE) due to focal pathologies.

Materials: Review of medical charts.

Results: Patient 1: A previously healthy 8-year-old, right-handed girl started with stereotyped focal seizures at 6 y. She was admitted with malignant status epilepticus. The electroencephalogram showed right temporal ictal activity. The magnetic resonance imaging (MRI) showed right fronto parietal moderated encephalic hemiatrophy and amygdala and hippocampus T2 hyperintensity. She showed no response to multiple AEDs and ketogenic diet. After 15 days, epilepsy surgery was performed with Stereo EEG to localize epileptogenic zone. We performed an anterior temporal lobectomy with resolution of status epilepticus, persisting with occasional seizures and no neurological deficit, 1.7 year after surgery.

Patient 2: 21 year old female, with focal epilepsy since she was born and right hemiparesis. She was admitted due to complex partial RSE 48h of evolution, treated with Phenobarbital, carbamazepine, phenytoin, clobazam. The Electroencephalogam showed bifrontal ictal activity. The MRI showed left fronto-parietal polymicrogiria. She did not respond to treatment with multiples AEDs. After 20 days we performed a left hemispherectomy. She is seizure free with right hemiparesis since surgery, 6 months ago.

Conclusion: In both cases the surgery resolved RSE, allowing the discontinuation of anaesthetic drugs in one patient. The surgery is a treatment option for RSE in children and adult with defined epileptogenic area.

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REFLEX EPILEPSY IN CHILDHOOD

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Reflex seizures are those, which are specifically stimulus-triggered. The prevalence among patients with epilepsy is 5%. Even when visual stimuli are known to be the most frequent precipitating factors, they can be evoked by any somatosensory stimuli.

Purpose: to report a series of pediatric patients with reflex seizures -excluding those precipitated by visual stimuli- in a pediatric patient series with reflex seizures.

Method: we reviewed the chart of 15 pediatric patients (age range between 3 months and 15 year old) that were evaluated in a pediatric neurology institute between the years 2000-2013 with video-polygraphic and EEG confirmation of reflex seizures. Triggers stimuli, clinical features, neuroimaging, seizures types, ictal and interictal electroencephalographic records and therapeutic responses were evaluated.

Results: The precipitating stimuli were: hearing 13/15, touching 7/15, proprioceptive stimuli 1/10, emotional: 1/10.

Abnormal findings in neuroimaging were found in 12 out of 15 of the patients. They presented focal tonic daily seizures with poor to no response to antiepileptic drug therapy.

Two cases with massive myoclonic jerks and no evidence of structural damage in neuroimaging, showed a favorable response to valproic acid.

Conclusion: Reflex epilepsy provoked by any stimuli other than visual is rare in childhood. The outcome of reflex epilepsy is different regarding the presence of structural brain lesion. In case of structural brain lesion the response to antiepileptic drugs is poor.

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EEG MONITORING IN CRITICALLY ILL CHILDREN: INDICATIONS, FINDINGS AND IMPACT ON CLINICAL MANAGEMENT.

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Introduction: Continuous EEG (cEEG) monitoring is being used with increasing frequency in critically ill patients, most often to detect non-convulsive seizures.

Purpose: To describe the indications, findings and impact on clinical management of continuous EEG monitoring.

Methods: 50 cEEG were performed in the pediatric and neonatal intensive care units were reviewed. Age, gender, diagnosis, EEG background, epileptiform activity, ictal findings, time of onset, changes in treatment and survival were collected.

Results: Thirty two patients aged 1 day to 16 years (median 3 months) were monitored with cEEG for 3-24hs (median 4hs) from 2011 to 2013. Thirteen patients were monitored more than once during the study period which resulted in a total of 51 recordings. The indications were: seizures in 35 patients and suspected non-convulsive status epilepticus in 15.

We observed non convulsive seizures in 6 (12%) cEEG, electrical non convulsive status in 6 (12%), electro-clinical seizures in 15 (30%) and 2 (4%) paroxysmal non-epileptic events. In 12 cEEG we did not register events. Seizures were identified at the onset of cEEG in 6 (12%) and during the first hour in 17 (34%). EEG monitoring led to increase in AEDs in 28 (56%) patients and decrease in 3 (6%).

Conclusions: In this study, we observed non convulsive seizures in almost a quarter of the cEEG, half being non convulsive status.

The cEEG led to changes in AEDs in more than half of the patients.

We emphasize the importance of cEEG in pediatric patients for detection of non convulsive seizures.

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EPILEPSIA PARTIALIS CONTINUA: PRESENTING SYMPTOM OF ALPERS' DISEASE

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Introduction: Epilepsia Partialis Continua (EPC) is rare during the first year of life. Alpers' disease is an inherited autosomal recessive disorder characterized by defective mitochondrial DNA synthesis which leads to a fatal brain and liver degeneration clinically manifested as neurodevelopmental regression, liver failure and epilepsy. It's caused by mutations in POLG1, being a progressive disease with fatal outcome.

Case description and methods:

A 7 month-old previously healthy girl, with no family history of consanguinity, who develops initially febrile, focal clonic status epilepticus (EPC), progressing to chronic encephalopathy and multiorgan failure leading to death.

Results: Initial MRI was normal, follow up images showed restricted diffusion in the thalami, cortex and periatrial white matter, with low ADC signal. EEG had asymmetric amplitude, and later diffuse disorganized background activity with right centro-temporal acute waves. CSF and oligoclonal bands were normal, with persistent elevated liver enzymes and blood lactate. Other neurometabolic diseases were ruled out, with clinical suspicion of Alpers' disease POLG1 mutation analysis was performed with a positive result.

Conclusions: Although Alpers' disease is a rare disorder, due to its devastating outcome and wider genetic implications this diagnosis is important to consider in cases of Epilepsia Partialis Continua at a young age with suggestive clinical, EEG and MRI findings.

Confirming the diagnosis of Alpers' Disease allows accurate genetic counselling and avoiding the use of potentially hepatotoxic drugs (such as valproic acid for seizure treatment).

P93

TREATMENT WITH MODIFIED ATKINS DIET TYPE IN NINE PATIENTS WITH DRUG-RESISTANT EPILEPSY

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Introduction: The modified Atkins diet (DAM) is a therapeutic alternative in the treatment of drug-resistant epilepsy. It consists of a diet with a contribution of 60% fat, 30% protein and 10% carbohydrate.

Objective: To describe response, tolerance and side effects of a series of 9 patients diagnosed with refractory epilepsy of different etiologies, who were treated with AMD at our hospital.

Results: In our group of 9 patients obtained similar results to those reported by other authors with good adherence, good tolerance and effectiveness. Of the two patients achieved a reduction by more than 90% the number of seizures, 4 between 50-90%, 2 less than 50% control, only one did not respond to the DAM. No patient had increased the number of seizures and was well tolerated in all cases.

P94

KETOGENIC DIET NATIONAL CONSENSUS - KETOGENIC DIET ARGENTINIAN GROUP

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Objectives: to Standarize classic and modified Atkins ketogenic diet (KD) use between several Argentinians health centers.

Material and Methods: the KD National consensus was created between March 2012 and August 2013 by the KD national group from the Argentinian Child Neurology Society. This Group consists of Child neurologists, a nutritionist physician, pediatrician and professional nutritionist of 12 Argentine provinces from 13 health centers from where KD was used as treatment in drug resistant epilepsy. All published information and experience from each center was reviewed.

Results: Criteria was established as followed:

1. Patient selection: indications and contraindications.
2. Neurological exam and nutritional state previous to treatment
3. Laboratory test and complemmentary studies.
4. Food intake data and dietary requirements calcultation.
5. Diet initiation methodology.
6. Family training.
7. Follow up.
8. Efficacy and side efects registration.
9. End of treatment.

Conclusions: considering that KD has reemerged as a treatment in drug resistant epilepsy in our country and through out the world, guidelines are required in order to increase its efficacy and to minimize its side effects. the constitution of an interdisciplinary national group and this consensus publication brings out the possibility to other centers to implement it.

P95

EFFECTIVENESS AND SAFETY OF KETOGENIC DIET IN A POPULATION OF DRUG RESISTANT EPILEPSY

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Introduction: The ketogenic diet has been widely and successfully used for the past 90 years as a therapeutic option in patients with drug resistant epilepsy. Its effectiveness, tolerability and side effects has been reported in a series of publications, nonetheless its outcome once suspended has been not.

Objective: to describe the effectiveness and safety of ketogenic diet and their outcome once their treatment was concluded.

Material and methods. 82 medical charts from the ketogenic diet program of our Institution were reviewed.

Results: 10 patients achieved 100% seizures control, 20 achieved 90%, 30 managed to control between 50-90 % of seizures, 15 patients less than 50% of seizures and 7 patients had no response.

Regarding the outcome, 22 patients remain in diet so far, 26 were suspended (22 due to lack of response and 4 withdrawals because of side effects), 4 patients died and 30 patients concluded the 2 years period of treatment. From these 30 patients 12 increased seizures frequency, in 3 ketogenic diet was reinitiated with similar seizure control afterwards.

Conclusions. In our experience we report similar results from those published in terms of tolerability and efficacy of ketogenic diet. A high relapse rate was reported in those in which this treatment was suspended after the initial 2 years period, achieving acceptable seizure control once restarted.

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EPILEPSY DUE TO PROTOCADHERIN 19 GENE MUTATION: REPORT OF THE FIRST ARGENTINEAN CASE

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Introduction: Mutation in the protocadherin 19 (PCDH19) gene is an increasingly recognized cause of epilepsy in females. This disorder is frequently associated with mental retardation and psychiatric features. In our country, the molecular diagnostic test is not available.

Objective: To describe a patient with y epilepsy due to PCDH19 mutation with molecular confirmation.

Materials and Methods: Review of medical reports.

Results: Female, 4 years of age with epilepsy secondary to mutation of the gene PCDH19. Perinatal History: First child of non-consanguineous parents. She was born in term with adequate weight for gestational age

She was admitted to the hospital at 9 months due to febrile status that evolved into non-convulsive status. Brain MRI was performed without pathological findings, negative blood and CSF cultures were obtained. Negative viral serology. Neurometabolic studies in blood, CSF and urine revealed nothing abnormal. High resolution karyotype was normal. Remained seizure-free for 3 months, then she started with episodes of complex partial seizures with secondary generalization associated with mostly with fever. Myoclonic seizures were never noted. In her evolution she developed ataxia and difficulties in language acquisition and socialization. No mutation was found in the ***SCN1A*** gene. A C.1425_1426delGT heterozygous deletion (p.ser476Cysfs * 46) of the PCDH19 gene was found, producing a premature stop codon. Actually she is seizure free with Valproate, Topiramate and Clobazam treatment. She has rehabilitation therapies and school integration.

Conclusions: The phenotype of suspicion involves women with clinical features similar to Dravet Syndrome but with better cognitive development and epilepsy outcome.

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THE SCN1A GENE, EARLY EPILEPSIES, AND GENOTYPE/ PHENOTYPE CORRELATION

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INTRODUCTION: Mutations of the SCN1A gene generate variables epileptic phenotypes and in 90% of the cases are related to: Generalized epilepsy with febrile seizures plus, Dravet syndrome and its clinical variations and Intractable epilepsy of childhood with generalized tonic-clonic seizures that simulates Dravet Syndrome OBJECTIVES and MATERIALS: Description of 3 patients with mutations of the gene SCN1A at different alleles, ages 5 to 21 years, who have the common history of multiple seizures types, in clusters, refractory to AEDs, some of them related to fever or other precipitants, associated with late onset of different degrees of Mental Retardation, normal Neuroimaging and variable EEG abnormalities. And 1 patient with PCDH 19 gene mutation with similar clinical presentation as Dravet Syndrome.

DISCUSSION: 1. Variability of the mutations of the SCN1A gene in relation to genotype / phenotype. 2. Retrospective diagnosis in adults. 3. Mild forms of Dravet S. 4. Genetic Epilepsies not SCN1A mutation related simulating Dravet S. 5. Importance of gene diagnosis and future treatment implications

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TRANSCRANIAL DIRECT CURRENT STIMULATION TDCS IN REFRACTORY FOCAL EPILEPSY: CASE REPORT

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Introduction: tDCS is a non invasive, inexpensive and safe tool for neuromodulation. tDCS has been used in treatment of psychiatric disorders, pain and epilepsy. We report on a trial of tDCS in a child with refractory focal epilepsy (RFE).

Case report: A 3 year child with RFE, intractable to multiple AEDs and immunomodulation, on a gluten free diet for celiac disease, had tDCS over two weeks.

V-EEG was indicative of right centroparietal focus of seizure onset. MRI showed no definite focal lesion. Focal seizures with and without secondary generalisation occurred innumerable times per day.

Methodology: Cathodal tDCS was delivered using NeuroConn DC stimulator MC on four sessions per week for two weeks. There were no adverse effects and the child tolerated it well. V-EEG was undertaken before, during and after tDCS. The number of spikes, frequency of unilateral and bilateral spikes, the average maximum and minimum spike amplitude and the proportion of the EEG with spikes was evaluated for a duration of 5 minutes before, during and after tDCS. Seizure frequency was monitored by history.

Results: tDCS resulted in a significant reduction in the spike burden on EEG. However there was no significant improvement in clinical seizures.

Conclusion: tDCS may play a therapeutic role in refractory childhood epilepsy. In our child tDCS reduced the amount of epileptiform activity on EEG, but did not have an impact on her clinical seizures. Further studies are required to investigate different protocols of stimulation and evaluate efficacy.

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LACOSMIDE AS ADJUNCTIVE THERAPY IN CHILDREN WITH REFRACTORY FOCAL EPILEPSY

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Background: Lacosamide (LCM) is a novel anti-epileptic drug (AED) that enhances the slow inactivation of voltage-gated sodium channels. Randomised placebo controlled trials in adults with LCM as an adjunctive therapy with partial seizures report improved 50% responder rates (>50% reduction in seizure frequency) compared to placebo (RR=1.68)1. However, there is limited data on its efficacy and tolerability in children with refractory focal epilepsy (RFE).

Objective: In a single center audit, we aimed to assess efficacy and tolerance of LCM as add-on therapy in childhood RFE.

Methods: Efficacy was evaluated at 12 weeks after initiating LCM. Adequate Response (AR) was defined as ≥ 50% reduction in seizure frequency and partial response (PR) as >25% but <50% improvement. Poor response was defined as LCM discontinuation either due to inadequate seizure control (<25% reduction) or increasing seizure frequency.

Results: 23 children with RFE (multiple AED trials -all, previous epilepsy surgery-3, VNS-1) were included. The median age at initiation of LCM was 13.5 years (IQR, 7.5-15). An adequate response was observed in 3/23(13%) and a partial response in 5/23(22%). 15/23 (65%) had a poor response. The median dose was 6.8mg/kg, (IQR, 5.5-8) LCM was well tolerated in the majority, with minor side effects: drowsiness (17%) and mood change (8%).

Conclusion: LCM was well tolerated in children with RFE but appeared to have limited efficacy, achieving an adequate or partial response in one-third.

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NEW ONSET SEIZURE CLINIC FOR CHILDREN: NEED AND IMPORTANCE

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Background: New onset seizure (NOS) clinics are common in adult neurology, they are rare in paediatric neurology. It is known that single and recurrent seizures in childhood may be associated with impaired HRQOL, and cognitive, motor and behavioural impairment.

Objective: We established a NOS Clinic for Children (NOSC) in 2011, to provide expert service and to contribute to better understanding of NOS in childhood. The long term objective being development of targeted management strategies that cost effectively improve quality of care for children with NOS.

Methods: Details of the first 200 seen in the NOSC were analysed. Their demographics noted, seizures categorized and classified, investigations (EEG, Imaging) evaluated and comorbidities recorded. In the next phase we plan to administer QOL questionnaires, Child Behaviour Check Lists, Sleep questionnaires etc.

Results: Of 200 children (106 boys), 107 were <7 years of age. Seizures occurred in the wake state in 124, on arousal in 10, when asleep in 27, in both wake and sleep in 12, state was not recorded in 27. 117/192 (61%) EEGs were abnormal. Neuroimaging was obtained when clinically appropriate. At ~3 months follow up 80 were on regular AED, 17 had rescue medication only prescribed, 37 were on a plan to start AED if seizures recurred. More results will be presented. Families liked the NOSC and feedback was positive.

Conclusion: NOSC is useful and important. Long-term follow-up will identify predictors of seizure recurrence, associations and complications and help formulate early interventional strategies to minimize impact on HRQOL.

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EPILEPSY RISK FACTORS IN CHILDREN

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INTRODUCTION: Epilepsy risk factors in children may be of interest in the field of childhood epileptology.

OBJECTIVE: The aim was to compare clinical, EEG and neuroimaging data of patients with epilepsy and first unprovoked seizure and reveal epilepsy risk factors.

MATERIAL AND METHODS: In a prospective study 315 patients with epilepsy and 42 with first unprovoked seizure, aged from 2 months to 18 years, were included. We analyzed characteristics of the first seizure, perinatal history, family history of seizures, neurological examination. The patients underwent EEG and neuroimaging [Computed Tomography (CT) and/or Magnetic Resonance Imaging (MRI)]. Children were followed for 3 years from the time of first seizure.

RESULTS: The risk factors for epilepsy were early seizure onset (before 6 month) (p<0, 0001), family history of seizures of first range relationship (p=0, 0466), neurological abnormality (including psychomotor retardation) (p=0, 0024), atrophic change defined by neuroimaging ($\chi 2$=11, 04; p=0, 0009). Using general regression analyses we revealed that early seizure onset (p<0, 0001) and neurological abnormality (p<0, 05) were the only variables significantly associated with recurrence and were cumulative risk factors for epilepsy in children comparing with first unprovoked seizure group.

CONCLUSIONS: This study suggested that early seizure onset and neurological abnormality may have an important diagnostic value in the prognosis of epileptic seizure recurrence in children. These results might help in developing of diagnostic protocol in epilepsy in children.

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EARLY EPILEPTIC ENCEPHALOPATHY WITH SUPPRESSION-BURST REVEALING RFT1-CDG SYNDROME: REPORT OF TWO NEW CASES.

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Background: RTF1-congenital disorder of glycosylation (CDG) syndrome is a very rare subtype of CDG syndrome (6 cases reported so far) associating early onset epilepsy, severe developmental delay and deafness. We aimed here at better characterize the electro-clinical presentation in reporting 2 new cases.

Cases report: Girl, born at term with irritability and axial hypotonia. Seizures appeared at 3 months. Video-EEG showed a suppression-burst pattern during slow sleep, and tonic seizures were recorded when awake. Auditory evoked potentials were absent and sialotransferrine electrophoresis suggested CDG-Ix. Seizures resisted to valproate but were controlled by vigabatrin. Direct sequencing of the RFT1 gene showed a homozygote mutation (p.K152E; c.454A>G).

Boy, born at term with irritability and axial hypotonia. Seizures appeared on day 2 of life. Video-EEG showed a suppression-burst pattern in slow sleep with high amplitude generalized spikes and sharp slow waves and both tonic and myoclonic seizures. At 3 months, partial seizures followed by clusters of spasms were recorded. After failure of several drugs, vigabatrin add-on led to an impressive decrease of seizure frequency. Auditory evoked potentials were absent and sialotransferrine electrophoresis was normal. DNA analysis showed that patient was heterozygous compound for RFT1 gene (c.1325G>A (p.R442Q) and c.110G>T (p.R37L)).

Conclusions: RFT1-CDG syndrome should be considered in the differential diagnosis of early epileptic encephalopathy with suppression burst even in the presence of a normal sialotransferrine electrophoresis. Other clues for diagnosis are congenital severe hypotonia and deafness. Vigabatrin should be used to treat seizures in this disease.

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VAGUS NERVE STIMULATION IN CHILDREN WITH DRUG-RESISTANT EPILEPSY: TWO CASE REPORTS.

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Introduction: Vagus Nerve Stimulation (VNS) is an adjunctive therapy used in intractable epilepsy. This procedure appears to be effective at reducing the frequency of seizures in adults. However, evidence in children is limited.

Methods: We report two cases of children with drug resistant epilepsy in whom VNS was implanted.

Case Description: Case 1: a 9 year old boy with gelastic epilepsy secondary to hypothalamic hamartoma who underwent a partial resection five years before, with only small decrease in seizure frequency. Case 2: a 2 year old girl with Ohtahara syndrome who was not candidate for surgical resection. Both were developmentally disabled and had multiple daily seizures refractory to antiepileptic drugs. They initially received low-output VNS with gradual increase to high-output parameters.

Results: Seizure frequency decreased in more than 70% in both patients with VNS therapy after a preliminary follow-up of respectively 18 and 4 months. Despite being not seizure-free, Case 1 improved significantly his language skills and social behavior and went back to school. Case 2 improved her daytime alertness and motor development. Both had no surgical complication and any side effect was observed.

Conclusion: VNS was safe, well-tolerated, reduced significantly seizure frequency in both cases and also improved mental and motor development.

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EPILEPSY SURGERY IN CHILDREN AND ADOLESCENTS WITH BRAIN TUMOR

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Introduction: Approximately 20-30% of long-standing medically intractable epilepsies are caused by tumors of neuroepithelial tissue, especially neuronal–glial tumors. In such cases, resection of the lesion is the treatment of choice with favorable seizure outcome. The aim of this study was to analyze the seizure outcome of children and adolescents who underwent tumor resections.

Methods: Retrospective data on 42 children and adolescents with tumors of neuroepithelial tissue who underwent extensive preoperative diagnostic studies according to a standard epilepsy protocol between 1991 and 2012 were collected. Postoperatively, Engel classification was used for seizure outcomes.

Results: The surgeries were performed between 2 and 18 years (mean 11), 15 (67%) were male. The age at seizure onset range from 1 month to 15 years (mean 6.2 years). The mean duration of follow-up was 5.7 years (1 to 18). The dysembryoplastic neuroepithelial tumors (DNET) were the most frequent (19 - 45.2%), followed by ganglioglioma 10 cases (23.8%) and other tumors represented 30.9%. At last clinical evaluation, 35 patients (83.3%) were Engel I, 2 (4.7%) Engel II, 4 (9.5%) Engel III and 1 (2.3%) Engel IV.

Conclusion: Surgical treatment for seizure control in children and adolescents with low-grade brain tumors can provide excellent results. For attaining favorable seizure and oncological outcomes, children with tumors and epilepsy must be promptly and thoroughly evaluated for early surgery, under a surgical epilepsy program.

P105
HEMISPHERIC SURGERY FOR EPILEPSY IN CHILDREN: OUTCOME REGARDING SEIZURES AND LANGUAGE

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Hemispheric surgery is an established treatment for medically refractory epilepsy due to diffuse hemispheric disease, with 60-80 % of seizure control, besides cognitive, behavioral and psychosocial improvement. Lateralization of linguistic functions, usually in the left hemisphere, is defined by the age of six years. Risk of postoperative aphasia due to dominant hemispheric surgery contributes for not operating children older than this. The aim of this study is to evaluate the speech of children who underwent left hemispherothomy by the time language dominance is theoretically well established.

Retrospective data on 18 patients older than 5 years old who underwent left hemispherothomy between 1998 and 2012, with at least 6 months follow up, were collected. Postoperatively, Engel classification and neurological examination plus Vineland Adaptive Behavior Scale were used for seizure and communication outcomes, respectively.

Ages at surgery were between 5 and 18 years (mean 9.9). Concerning the etiology, 6 (33,3%) presented Rasmussen, 7 (38,9%) acquired causes , 5 (27,8%) malformation of cortical development at the histopathology.

After a follow-up period from 6 months to 8 years (mean 3.5), 14 patients were re-evaluated for seizures and 10 (71,4 %) were classified as Engel I or II , 4 (28,6%) as III or IV. Regarding communication, postoperatively 17 patients (94,4%) remained unchanged and one presented speech deterioration.

Left hemispherothomy performed in children older than 5 years is associated with good seizure outcome and may not be deleterious on speech function previously acquired.

P106
PREVALENCE OF FEBRILE SEIZURE – A POPULATION-BASED STUDY

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Background: Febrile seizure is a type of seizure wich is more common in childhood, affecting approximately 2-4% of children.

Objective: Determine the prevalence of febrile seizures in children aged 1 month to 5 years of age, and to identify the clinical and epidemiological profile of the selected sample.

Methods: Cross-sectional study, door-to-door, conducted in two phases, in Barra do Bugres town, Mato Grosso state, from August 2012 to August 2013. The town has 2,726 inhabitants aged between 0-4 years and 11 months. We use the definition of febrile convulsion established by the International League Against Epilepsy.

Results: In the first phase 103 children with suspected seizure event were taken to the clinical center, these 18 (17.5 %) had febrile seizures, 50 % were male. The age of the children remained between 1-5 years. The prevalence of febrile seizures in the sample was 6.6/1,000 inhabitants aged 0-5 years. The age of first seizure ranged from 1-60 months (average 19, 3 months). Regarding the type of seizures 88.8 % of patients had generalized seizures. In 33.3% of cases we found family history of febrile seizures in 1st degree relatives and history of epilepsy in 11.1% of cases.

Conclusions: The prevalence in Mato Grosso showed to be lower than that found in another study in southern Brazil (16/1000), which can be justified by a geographical diversity and methodological differences between the studies.

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EPILEPSY SURGERY IN THE FIRST 3 YEARS OF LIFE: CLINICAL OUTCOME AND RISK-BENEFITS

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Up to one-tenth of the lifetime risk of epilepsy is realized in the first 3 years of life. Early-onset epilepsies might be associated with severe lifelong disability. In 30% of children medical treatment is unsatisfactory; surgery may be considered as an option. The purpose of this study was to provide seizure outcome of infants and toddlers who underwent surgical treatment for epilepsy, considering risks of the procedures.

Retrospective data on 33 patients younger than age 3 years who underwent epilepsy surgery between 1996 and 2013, with at least one year follow up, were collected. Postoperatively, Engel classification was used for seizure short and long term outcomes.

Ages between 2 and 36 months (mean 19.3), 18 males (48.6%). Mean follow-up was 6.1 years (1 to 16.8). Etiologies: malformation of cortical development 16 (48.5%), acquired causes 7 (21.2%), progressive disease 10 (30.3%). Twenty patients (60.6%) underwent hemispherotomy, 4 (12.1%) lesionectomy, 5 (15.2%) lobectomy and 4 (12.1%) VNS. At last clinical evaluation, 16 patients (50%) were Engel I, 3 (9.4%) Engel II, 9 (28.1%) Engel III and 4 (12.5%) Engel IV. Postoperative complications: mild motor deficit 6 (18%), treatable infection 6 (18.1%), intracranial hypertension 2 (6.1%), bleeding 2 (6.1%), liquoric fistula 1 (3%). There was one surgical mortality (3%).

Epilepsy surgery in infants and toddlers is effective for seizure control and can be performed with acceptable morbidity and mortality. Surgical intervention should be considered even in infants if seizures are poorly controlled.

P108

PREDICTORS OF RECURRENCE OF A FIRST UNPROVOKED SEIZURE IN CHILDREN. A PROSPECTIVE STUDY.

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198 children with a first unprovoked seizure were followed up from 6 months to 10 years. There were 122 boys and 76 girls. Age varied from 5 months to 16 years (mean=8.7y). All patients but 9 did not receive AED. The neurological exam and imaging were normal. In 101 (50.7%) the seizure never recurred. Mean age was 9 years. In 57 the seizure was partial and in 44 generalized. The EEG (n=99) was abnormal in 20. Positive family history for epilepsy was present in 42. Seizure lasted less than 15 minutes in 79. Recurrence occurred in 97 cases (49.2%). The mean age was 8.5 years; the seizure was partial in 49 and generalized in 48. EEG (n= 96) was abnormal in 42 cases. Positive family history was found in 46. The seizure lasted less than 15 minutes in 77. In 36 (37%) the second seizure occurred in the first 3 months; in 19 (19%) between 3 and 6 months); in 20 (20%) between 6 and 12 months, in 9 (9.3%) in the second year, in 7 (7.2%) in the third year and only in 5 (5%) after 3 years. All patients but 11 had AED after the second seizure. Eighteen patients (18.5%) experienced a third seizure. Seven had 4 seizures, and 6 more than 5. As a conclusion, from all parameters studied only abnormality of EEG was a predictive factor for recurrences in children with a first unprovoked seizure.

P109

HOT EXECUTIVE FUNCTION IN CHILDREN WITH EPILEPSY

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Purpose: The aim of this study was to evaluate the hot executive function in children with epilepsy and compare their performance with healthy children taking into account clinical variables of epilepsy.

Method: We evaluated 51 children, 17 with Rolandic Epilepsy (RE), 9 with Temporal Lobe Epilepsy (TLE) and 25 healthy controls. All children were assessed with the Iowa Gabling Task (IGT), the most used instrument to evaluate the decision-making style, and the Wechsler Intelligence Scale for Children (WISC-III) to investigate the intellectual level (estimated IQ). The clinical variables of epilepsy were: age of seizure onset and remission, type of seizure (focal versus generalized), use of AED (none, monotherapy or polytherapy), and seizure lateralization (right, left, and bilateral).

Results: No significant differences were found between the three groups. However, when analysing the RE and TLE groups, we observed that RE group (μ=-1, 44) had inferior results than the TLE group (μ=5, 5). The same was true when comparing the intellectual level of the two groups: the higher the IQ, the better the decision-making style.

Conclusion: We conclude that although RE is known as a benign entity, difficulty in decision-making ability may be observed. More studies are necessary to corroborate our findings.

P110

INFANTILE SPASMS: A SERIOUS NEUROLOGICAL DISORDER FOLLOWING WHOLE-CELL PERTUSSIS VACCINE

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Introduction: The adverse events following the whole-cell pertussis vaccination (WCPV) among children, include, infantile spasms (IS), or West Syndrome. IS is an important epileptic encephalopathy of childhood, with a electroencephalographic (EEG) pattern (hypsarrhythmia) and psychomotor delay. The cessation of spasms and normalization of the EEG, implicates in better response and prognosis. It consists of corticosteroids (ACTH, prednisone) and antiepileptic drugs (AED) as vigabatrin (VGB), topiramate (TPM), valproic acid (VPA), and pyridoxine. High dosage of pyridoxine, following the Japanese and Finnish protocols for ACTH has been used. There is a trend in using ACTH in smaller dosage.

Case report: A child at 2 months, received the WCPV and developed after 20 days IS. She underwent pyridoxine, ACTH, clonazepan, valproic acid, nitrazepan, topiramate and clobazam. The recent MRI showed subtle slowing of myelination at the right frontal and temporal. She is now with a normal neurological and psychomotor development. A partial epilepsy is present now, controlled with lamotrigine, oxcarbazepine and Nitrazepam.

Discussion: We think that the WCPV represent a harmful instrument to develop the spasms. Several protocols have been attributed to IS. Beside the other AED the ACTH was used. We conclude in this case, probably broken out by the WCPV, the rational polytherapy, according to universal protocoles was worthy. Despite a partial epilepsy has developed, her neurological exam and development are normal. Findings with delays in myelination are not uncommon in patients with West Syndrome.

P111

POSTERIOR CORTEX EPILEPSY SURGERY: SHORT AND LONG-TERM OUTCOMES

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Posterior cortex epilepsy (PCE) encompasses a group of epilepsies originating from the occipital, parietal, and/or posterior temporal lobes. A significant number of these patients will eventually develop medically refractory epilepsy. However, in these cases, good surgical outcome often can be achieved. The aim of our study was to analyze the short and long term seizure outcomes of children who underwent PCE surgeries.

A retrospective analysis of 22 patients who underwent PCE surgeries between 1998 and 2008 was performed. All patients had at least 5 years follow up. Surgical outcome was evaluated by using the Engel`s classification.

The subjects were 7 female and 15 (68.2%) male. The surgeries were performed between the age of 2 and 18 years (mean 10.4). Mean age at seizure onset was 3 years and the duration of illness lasted from 1 to 18 years (mean 7.4). The etiology was gliosis in 12 (54.5%) patients and cortical/vascular malformation in 10 (45.5%). In the first postoperative follow-up after 1 year of surgery, 16 (72.7%) patients were classified as Engel I, 1 (4.5%) as Engel II, 3 (13.6%) as Engel III, and 2 (9.0%) as Engel IV. After 5 years of surgery, during the postoperative follow-up, 15 (68.2%) patients were classified as Engel I, 2 (9.0%) as Engel III and 5 (22.8%) as Engel IV.

Surgical treatment can be effective in medically refractory patients with PCE. Nevertheless, up to now a few number of patients undergo a presurgical evaluation for epilepsy surgery. This evaluation could be decisive in identifying as early as possible patients that could benefit from epilepsy surgery in order to provide seizure control as well as to avoid disability, lower academic performance, and significant economic and personal costs.

P112

EVALUATION OF THE QUALITY OF LIFE OF PARENTS AND CARERS OF CHILDREN AND ADOLESCENTS WITH REFRACTORY EPILEPSY IN COMPARISON WITH PARENTS AND CAREGIVERS OF CHILDREN WITH DOWN SYNDROME

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Introduction: Epilepsy is the most common pediatric neurological disease. Conceptualizing quality of life is difficult, since it is a subjective concept of well-being in the best "analyst" is the patient's own.

Methods: In the period from 01-11-2011 to 01-08-2012, participated in the study (n = 64) parents / caregivers whose children / adolescents with epilepsy and Down syndrome to consult the HCSA; subdivided into 2 groups of 32 participants. For a significance level of 5 %, a power of 80 %, a standard deviation of 9.6 % in the total scores of the World Health Organization Quality of Life and a difference of 7.5 % between groups, we obtained a total minimum of 27 patients.

Results: The mean scores for each domain were compared between groups using the Student's t test for independent samples and considered significant if $p < 0.05$. The study showed no statistically significant difference between the quality of life of both groups in studies, in both groups, the environment domain had scores significantly lower

than in other areas ($p \leq 0.001$) .We notice that the group of parents and caregivers is remarkable predominance of females compared to males.

Conclusion: The present study, although it has not examined the quality of life of patients, found that no statistically significant difference between their parents and caregivers in both groups.

P113

DOWN SYNDROME: CLINICAL AND EEG CORRELATES DURING DEVELOPMENT

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Background: Down syndrome is the most common genetic cause of mental retardation and it is estimated that 5-13 % of persons affected with Down syndrome have seizures. In one series of 350 children with Down syndrome, seizures occurred in almost 8 % and one-third was infantile spasm. Seizures evolution and EEG findings in Down syndrome haven't been well investigated and studied.

Objectives: (1) To review the experience of Hospital for Sick Children on Down syndrome patients with seizure disorder over the last 20 years. (2)To follow and study the seizures' evolution (clinically and EEG) and responding to pharmacological treatment.

Methods: (1) A retrospective analysis of HSC's data including clinical information and EEGs result collected from 1990 to 2012 for Down syndrome patients with seizure. **(2)**All patients with Down's syndrome whose have MRI, EEG or presenting to ED or neurology clinic with seizures.

Hypothesis: Down syndrome patients with hypsarrhythmia would present with worse cognitive outcomes

Result: Total number of Down Syndrome patients with seizures seen at HSC between 1990 until 2012 was 71. 34 out of the 71 DS patients had infantile spasm (47 %) and 37 had other seizures type (53 %). Out of the 34 DS with infantile spasms; 23 patients had Hypsarrhythmia and 11 had no Hypsarrhythmia. 7 patients out of the 23 patients had severe developmental delay.

Conclusion: Severe developmental outcomes tend to be more prevalent in Down syndrome patients with hypsarrhythmia than those Down syndrome patients without hypsarrhythmia.

P114

"EPILEPSY TRANSITION: CHALLENGES OF CARING FOR ADULTS WITH CHILDHOOD-ONSET SEIZURES"

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Objectives: There is a striking deficiency of research in Epilepsy Transition. The objective of this work is to describe patients from a pediatric tertiary center referred to our Epilepsy Transition Program and to compare them with patients referred from community to our Adult Epilepsy Clinic. Additionally, we identified the level of confidence among neurologists receiving these challenging patients.

Methods: A cohort of age-matched patients (18-25y) from our Adult Epilepsy Clinic was retrospectively studied. Patients from our Epilepsy Transition Program (Group 1) were compared with the group referred directly from community physicians (Group 2). As well, we used a survey to evaluate neurologists dealing with childhood-onset epilepsies.

Results: Group 1 comprised 170 patients while Group 2 had 132. Patients in Group 1 had earlier seizures onset, longer epilepsy duration ($p<0.001$), more patients with symptomatic etiologies, epileptic encephalopathy and cognitive delay ($p<0.001$). Group 1 patients required more referrals to other specialties ($p=0.001$). Treatment with polytherapy ($p=0.003$), epilepsy-surgery ($p<0.001$), ketogenic diet ($p<0.001$), and vagus nerve stimulator were more common in Group 1 ($p<0.001$). Additionally, our survey applied to adult (n=86) and pediatric (n=29) neurologists indicated that adult neurologists have lower levels of confidence to diagnose and treat severe forms of childhood-onset epilepsies ($p<0.001$), as well as epilepsy associated with cognitive delay ($p<0.001$).

Conclusions: These findings suggest that: patients from tertiary centers present more complex healthcare needs and require more resources than age-matched patients from the community; and adult neurologists may not feel prepared to diagnose and treat adult patients with some childhood-onset epilepsies.

P115

ABSENCE STATUS EPILEPTICUS IN CHILDREN AS THEIR FIRST PRESENTATION OF ABSENCE EPILEPSY: A REPORT OF 4 CASES.

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Introduction: Absence status epilepticus in patients with absence epilepsy is uncommon. Absence status as the first presentation of absence epilepsy in children is even less common and not well documented in the medical literature.

Method: This is a retrospective study by means of chart review in a cohort of patients who presented with absence status epilepticus at the Children's Hospital of Eastern Ontario, Canada, between January 2008 and November 2013 inclusive.

Results: We identified 8 patients with a diagnosis of absence epilepsy and a history of absence status epilepticus. Half of the cases presented with absence status as their first presentation. In these, the patients presented to the ED with a 4 to 36 hour history of ongoing confusion. Absence epilepsy was confirmed by EEGs. All 4 responded to classical first line anticonvulsants for absence seizures and had no more confusional states during follow up.

Conclusion: Non convulsive status, especially absence status, should be considered in the differential diagnosis in children presenting in the ED with acute prolonged or recurrent confusional state, even in the absence of a prior history of epileptic seizures.

P116

INFRA-SLOW EEG ACTIVITY AND SLEEP SPINDLE EXPRESSION - POTENTIAL WINDOW INTO THALAMIC FUNCTION IN INFANTILE SPASMS

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Introduction: Analysis of infra-slow EEG activity (ISA) may provide insight into the pathophysiology of infantile spasms (IS). ISA is thought to be generated in part by thalamo-cortical (TC) networks which also are recognized to underlie the expression of sleep spindles, both of which may be pathologically expressed in patients with IS. We investigated a potential correlation of ISA and sleep spindles in patients with IS.

Methods: In 13 cases of IS, EEG recordings were evaluated for spasms-associated ISA to determine whether ISA changes, were generalized (g), lateralized (l), mixed (m) or associated with no- ISA. Results were correlated with the occurrence and expression pattern of sleep spindles.

Results: A total of 101 spasms were analyzed and 88/101 (87.1%) were associated with ISA. G-ISA was observed in 4/13 (30.8%), l-ISA in 1/13 (7%), m-ISA in 6/13 (46.2%) and no-ISA in 2/13 (15.4%) patients. Sleep was captured in all patients and spindles were observed in (7/13) 53% of patients: g-ISA 2/4 (50%; 50% symmetric), m-ISA 3/6 (50%; all asymmetric), no-ISA 2/2 (100%; all symmetric). Sleep spindles were not observed in the patient with l-ISA. Notably 10/11 (91.0%) of patients with spasm-associated ISA had either asymmetric or no recorded sleep spindles.

Conclusion: Spasm-associated ISA is likely pathologic, indicative of probable TC-dysfunction given the frequent co-occurrence of abnormal spindle expression and the normal spindle activity recorded in the two patients with no-ISA. Further research is needed to determine how TC dysfunction may lead to IS which may have both therapeutic and prognostic value.

P117

ALTERNATING HEMIPLEGIA EXTENDS BEYOND CHILDHOOD TO ADULTHOOD

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Introduction: Originally described as 'Alternating Hemiplegia of Childhood' (AHC), ongoing observation in some children now extends AHC into young adulthood, renamed AHCA.

Methods: Observational longitudinal studies in a small cohort of AHC subjects include phenotype, genotype, EEG and sleep, drug therapy and long-term follow-up. Variability was noted in age of onset, phenotypic

expression, seizure activity, EEG changes especially in sleep, and early response to medications: gamma-hydroxy butyrate, topiramate, phenytoin, flunarizine and ASA.

Results: In three cases, one had onset in infancy now aged 8 years, two had childhood onset, are now young adults, still with acute hemiplegic episodes. Two continued to have partial-onset seizures requiring AED therapy with reasonable control. All had comorbidities: learning disorder, speech or memory deficits, and two have spastic hemiplegia. EEG studies had variable slow and focal spikes that changed with cerebral development, not specific to AHC. Two had the known gene mutations of ATP1A3. To date the prognosis is guarded for long-term outcome and quality of life measures are needed.

Conclusion: Although the genotype is known, phenotypic variability, EEG evolution and response to therapy showed variable evolution of AHC into adulthood. A name change is needed 'Alternating Hemiplegia of Childhood and Adulthood' (AHCA). The transition to adult neurological care should be optimised.

P118

CLINICAL SPECTRUM AND HANDLING IN 13 CHILDREN WITH ELECTRIC SLEEP STATUS

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

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

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

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

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

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ELECTROENCEPHALOGRAPHIC CHARACTERIZATION OF PATIENTS WITH PERVASIVE DISORDER DEVELOPMENT

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Introduction: The coexistence of epilepsy in patients with autism spectrum disorder (ASD) is a 30% and an electroencephalographic alteration reaches up to 90% according to series described. The implication that these alterations occur in language development, intellectual coefficient and the final prognosis is a matter of discussion.

Objective: To describe the electroencephalographic characteristics of patients with ASD monitored in the child neurology service.

Materials and Methods: A cohort study. Descriptive analysis of 59 patients admitted record to polyclinic pervasive developmental disorder, divided into 3 groups (Classic autistic disorder, Pervasive nonspecific and Asperger syndrome).

Results: n = 59 (boys n=52). Mean age 8.3 years (range 3-15). Clinical epilepsy in 3 patients. 41% abnormal EEG. Classic autistic patients 9/21 had abnormal EEG (generalized n=2, focal n=3, slow n=2, pharmacological n=2), 2/9 have epilepsy. In the pervasive nonspecific group, 13/31 had abnormal EEG (generalized n=1, focal n=9, slow n=2, pharmacological n=1), 7 anticonvulsant treatment them all with focal EEG. In all treated patients showed abnormal development of language. In Asperger syndrome group 5/7 with normal EEG, 2/7 with generalized abnormal EEG without current treatment.

Conclusion: Our series is similar to that described in the literature, with 41% of EEG abnormality. The decision of use antiepileptic drugs was based on the location of the electric bulb (frontal and/ or temporal) and its implication from theory in language development. However we cannot conclude if their response was only by drug treatment or as part of their evolution.

P120

WEST SYNDROME IN CHILD WITH DOWN SYNDROME: DESCRIPTION OF A NUMBER OF CASES.

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

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

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

Results: Out of 7 patients (4/7 women) the average first occurrence of spams is at 9 months of age. Delay diagnosis: average age 6.2 months. 7/7 present retardation PMD, 2/7 regression PMD. 7/7 Flexor spam. 6/7 received ACTH and valproic 1/7 valproic only. Post cure ACTH: 4/6 complete termination of spams with normal EEG (2/4). 2/6 partial remission with persistent spams hypsarrhythmia (1/2) and severely abnormal EEG non hypsarrhythmic (1/2). Outcome: Patients with good initial response to treatment, clinical and/or electrical (4/7) showed no recurrence or persistence of crisis at 6 months follow up.

Conclusions: Nearly 40% were unfavorable, not concordant with the literature.

P121

SEIZURES NOT RELATED TO HYPOGLYCEMIA IN TYPE 1 DIABETIC PATIENTS (DM1)

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

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

Method: Retrospective descriptive study, analysis of clinical records.

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

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

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

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EPILEPSY ASSOCIATED TO INBORN ERRORS OF METABOLISM, STUDY AND EVOLUTION OF 130 PATIENTS

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Introduction: Seizures are a common symptom in a number of metabolic diseases. Occasional occur as secondary metabolic decompensation events or as a set epileptic condition . Epilepsy in these disorders can be classified depending on the pathophysiology, age of onset and type of crisis.

Objectives: To determine the major metabolic diseases that are associated with seizures, age of onset, clinical presentation and response to treatment.

Materials and Methods: A retrospective descriptive study of 130 patients with metabolic diseases in our department. Review of medical history.

Results: 23 metabolic diseases were evaluated, with a total of 130 patients, of whom 68 (52 %) had seizures. Seizures occurred between the first hours of life and 10 years of age (mean 60 months). Focal seizures predominated by 74 % , followed by 26 % generalized. Epilepsy was presented in 100% of patients wiht Hyperglycemia nonketotic, gangliosidosis, lipofuscinosis , peroxisomal neonatal-onset deficiency of sulfite oxidase, MELAS, MERRF, Deficit GLUT 1. Most commonly used drugs were phenobarbital, followed by carbamazepine, valproic acid and thereafter. Evolved with pharmacologic refractoriness 22% of patients.

Conclusion: In our series the main metabolic diseases associated with seizures are described and more than half (52 %) were associated with seizures. Predominant seizures were focal with secondary generalization. They were refractory in 22% .

Comment: The identification of epilepsy secondary to metabolic disease is of great importance due to the possibility of early treatment and timely manner.

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DRAVET SYNDROME: DESCRIPTION OF 4 PATIENTS WITH GENETIC CONFIRMATION

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

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

MATERIALS AND METHODS: Descriptive retrospective analysis of medical histories, EEG and genetic studies with prospective follow-up.

RESULTS: 4 patients, 4/4 normal development before seizures, ¼ epilepsy in family. First seizure occurred between 4-6 months, ¾ were febrile, ¾ lasted over 15 minutes. 4 patients had all kinds of seizures except for tonic, being partial secondary generalized, myoclonic and complex-partial seizures the most frequents. 4 patients have refractory epilepsy with frequent status. 2/4 worsen seizures with Lamotrigine. All evolve with a moderate-severe developmental delay, ¾ with ataxia, 2/4 with pyramidalism, ¾ with autistic behavior. All had normal EEGs during first year of life, evolving with focal-multifocal activity and disorganized background activity. Photosensitivity (+) ¼. Normal brain MRI ¾, ¼ with diffuse cortical atrophy. All cases had punctual heterozygous mutations in exons 4, 17 and introns 5, 22 of SCN1A gene.

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

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

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FEBRILE SEIZURE RECURRENCE REDUCED BY INTERMITTENT ORAL LEVETIRACETAM

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Objective: Febrile seizure (FS) is the most common form of childhood seizure disorder, it is perhaps one of the most frequent causes of admittance to pediatric emergency wards worldwide. We aimed to identify a new, safe, and effective therapy for preventing FS recurrence.

Methods: A total of 115 children with a history of two or more episodes of FS were randomly assigned to the levetiracetam (LEV) and control (LEV: control ratio 2:1) groups. At the onset of febrile, the LEV group was orally administered with a dose of 15 mg/kg/d to 30 mg/kg/d twice daily for 1 week and then dosage was gradually reduced until totally removed in the 2nd week. The primary efficacy variable was seizure frequency associated with febrile events and FS recurrence rate during 48-week follow-up, the second outcome was the cost-effectiveness of two groups.

Results: The intention-to-treat analysis showed that 11 children experienced 148 febrile episodes and 15 FS recurrences in the LEV group and 19 children experienced 64 febrile episodes and 32 FS recurrences in the control group. There was significant difference between two groups in FS recurrence rate and FS recurrence/fever episode. The cost of LEV for prevention of FS recurrence is much lower than that of control group. During the 48-week follow up period, one patient in the LEV group exhibited severe drowsiness, no other side effects were observed in the same patient and in other children.

Conclusion: Intermittent oral LEV can effectively prevent FS recurrence, and reduce waste of medical resources.

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APPLICATION OF HYDROGEN PROTON MAGNETIC RESONANCE SPECTRUM IN INFANT SPASM

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Objective: Studying the brain biochemical metabolism of infant spasm through hydrogen proton magnetic resonance spectrum (^{1}H-MRS).

Method: 40 infant spasm in hospital and 10 age-matched normal infants were performed brain MRI plain scan and 1 H-MRS check by using 1.5 T superconducting MRI scanner. Choosing the same interesting region: Bilateral thalamus, basal ganglia region, temporal lobe, getting spectral graph of brain tissue and relative concentration and ratio of N-acetyl aspartic acid (NAA) choline (Cho) creatine (Cr).

Result: The ratio of NAA/Cho+Cr, NAA/Cho, NAA/Cr in Bilateral thalamus, basal ganglia region, temporal lobe of infant spasm group were lower than normal group, $P<0.01$, the difference were statistically significant. Compared bilateral symmetric areas of interest, the ratio of NAA/Cho+Cr, NAA/Cho, NAA/Cr had no significant difference in the two groups, $P>0.05$, the difference were not statistically significant.

Conclusion: ^{1}H-MRS can discover brain function change of Infant Spasm non-invasively; NAA in brain tissue of infant spasm were lower than normal group.

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ARRAY-CGH DETECTION OF CHROMOSOMAL ABNORMALITIESIN CRYPTOGENIC PATIENTS WITH INFANTILE SPASMS

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Infantile spasms (IS) is the most common form of catastrophic childhood epilepsies. Above 300 factors are associated with West syndrome, according to the etiology, this syndrome is now classified into two groups, symptomatic and cryptogenic. The aim of this study is to investigate underlying factors in cryptogenic patients with IS. Among 170 IS patients, etiology of 55 cases remain unknown. Array-comparative genomic hybridization (CGH) analysis (G-scanning) was performed on these 55 patients. The array CGH chip data were analyzed using the chromofluor image analysis system (Array Analysis; Macrogen, South Korea). The slides contained 1440 human BAC clones including specific loci of more than 40 chromosomal disorders and 356 cell growth related

genes from BAC libraries at a resolution average of 2.3 Mb for the entire genome. Whole-genome array CGH analysis revealed duplications of 2p22p24 and 4q31q32 in two patients, the positive rate is 3.64%. To our knowledge, it's the first report of these chromosomal abnormalities in IS. Previous study showed that duplication of the 2p23-2pter region is associated with neural tube development, while multiple doses of band 4q32.1 are relatively well-tolerated. The role of duplications of 2p22p24 and 4q31q32 in pathogenesis of IS deserves further study.

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THE CHANGE OF WNT3A AND WNT5A MRNA IN THE PROCESS OF EPILEPTOGENESIS IN THE KAINATE-INDUCED EPILEPSY MODEL

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To reveale the time course of Wnt3a and Wnt5a mRNA during epileptogenesis in the Kainate-induced epilepsy model and the effect of valproate acid on the genes expression. Sprague-Dawley rats were induced by kainic acid (KA) at postnatal day 30, Real-time PCR was applied to detect the expression of Wnt3a and Wnt5a mRNA in the hippocampus at different stages of epileptogensis. Meanwhile we investigated whether VPA could affect the expression of Wnt3a and Wnt5a mRNA during the epileptogensis. The results show that at the acute stage after kindling, Wnt5a mRNA of KA group significantly decreased compared with that of NS group ($P<0.05$). At the stable stage of chronic epilepsy, the expression of Wnt5a mRNA increased significantly, compared with those of the former two stages ($P<0.05$). At the chronic epilepsy stage, Wnt5a mRNA had a dramatic increase compared with that at the acute stage and the initial stage of SRS, the difference has statistical significance (both $P <0.05$). At different stages of epileptogenesis, Wnt5a mRNA has no significant differences between KA group and KA+VPA group. The difference of Wnt3a mRNA expression is not significant neither among the time courses nor the different groups. The conclusion is that in kainate-induced epilepsy model, Wnt5a may be involed in the epileptogenesis by the regulation of neurogenesis in hippocapus. The antiepileptic mechanism of valproate acid is not related to Wnt3a and Wnt5a in the KA model. Wnt3a has no effect on the neurogenesis and it has no relation with the epileptogensis of kainate-induced model.

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RISK FACTORS FOR PERINATAL ARTERIAL ISCHAEMIC STROKE: A CASE-CONTROL STUDY

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Introduction: Perinatal arterial ischaemic stroke (PAIS) is an important cause of morbidity and mortality. The pathophysiology and associated risk factors are no completely understood.

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

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

Results: We analyzed 40 patients (66, 7% male, 33, 3% female). The mean gestational age was 39 weeks. 21(52,5%) of cases were confirmed as perinatal arterial ischaemic stroke(PAIS) and 19(47,5%) presumed perinatal arterial ischaemic stroke(PPAIS), the mean age of diagnosis were 6 days and 2 years-3month, respectively. The most frequent clinical presentation were seizures (86%) for PAIS and focal neurologic signs (95%) for PPAIS. Strokes preferentially involved the MCA territory (88%), 95% unilateral, 65% in the left hemisphere. All patients presented some neurologic deficit in the following clinical controls, the most common were hemiparesis and epilepsy. Significant risk factors in the multivariate analysis ($p<0.05$) were nulliparity (OR 11.74; CI 3.28-42.02), emergency caesarean section (OR 13.79; CI 3.51-54.13) and Apgar score (5 min) ≤7 (OR 13.75; CI 1.03-364.03).

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

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CLINICAL DIAGNOSIS, TREATMENT, AND ALDH7A1 MUTATIONS IN PYRIDOXINE-DEPENDENT EPILEPSY IN THREE CHINESE INFANTS

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Objective: The present study analyzed the clinical features, treatment process and examined the **ALDH7A1** mutations in three Chinese children with Pyridoxine-dependent epilepsy (PDE).

Methods: Three patients whose clinical diagnosis and treatment process were carefully analyzed and were suspected PDE. **ALDH7A1**mutations by direct sequencing were screened.

Results: The seizures of the three patients were all resistant to multiple anticonvulsants. **ALDH7A1** gene mutation analysis revealed two heterozygote mutations in each case: c.410G>A (p.G137E) and IVS11+1G>A in case 1, heter c.952 G>C (p.A318P) and heter c.965 C>T (p.A322V) in case 2, and heter c.902A>T (p.N301I) and IVS11+1G>A in case 3. Each mutation was transmitted from one of the patients. Except heter c.902A>T (p.N301I), the other four mutations have not been reported previously.

Conclusion: This is the first time to report cases of Chinese patients diagnosed with pyridoxine-dependent epilepsy by molecular genetic analysis. Definitely diagnosis of PDE for the three patients helped to guide the correct treatment.

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CAN GENES PREDICT RESPONSE TO HORMONAL TREATMENT IN INFANTILE SPASMS

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Infantile spasms (IS) is an age-specific epileptic syndrome. ACTH has been the standard treatment for IS and a large and impressive body of experience has documented the efficacy of ACTH for IS. However, the mechanism of ACTH therapy is still unclear. Using case-control study we found that there was a statistically significant difference in the prenatal stress level among mothers of infants in the three groups. Prenatal stress exposure in children can trigger infantile spasms on the basis of leading causes of symptomatic epilepsy, such as brain dysplasia and inherited metabolic disease. The two may act in combination to cause the occurrence of infantile spasms. (2) It has been shown that the expression of ACTH binding sites on human adrenocortical cells is upregulated after exposure of the cells to ACTH. ACTH not only increases the transcriptional rate of MC2R message but also prolongs the MC2R mRNA half-life According to our results, the TCCT haplotype has not only protective effect on IS, but also has high responses to ACTH treatment. These results based on our in-vitro system may advance understanding of mechanisms of ACTH in IS treatment

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QUALITY OF LIFE IN CHILDREN WITH WEST SYNDROME IN SANTIAGO DE CUBA PAEDIATRIC HOSPITAL

Gelsy Naranjo Pelayo . Hospital Comandante Manuel Fajardo

The evaluation of the quality of life has been an important instrument to determine how effective treatments are in epileptic children, specifically in those who suffer from West Syndrome, one of the worst kind of epileptic syndromes. That is the main reason of this research paper.

The objective was to apply in Santiago de Cuba Paediatric Hospital a scale named CAVE to determine the quality of life of children admitted with this pathology, showing hopeless results. A retrospective and descriptive study was made and as a consequence, a multidisciplinary team was created to analyze and modify those results. Clinical files were used, hospital general files, the data base of every patient admitted there from 2005 until last year and the information at the web site of children with this syndrome. Forty-three patients were included in a program that combined the participation of Neurologist, Neurophysiologist, Neuropsychologists, Therapist and Pediatric evaluating children starting and after three months, time in which they had all kind of exercises to increase the plasticity of their brain and their capacity to include themselves to the society and their families environment. There was created a final program made of combined exercises and evaluation of the brain function by the Neurophysiologist to apply after the hopeless results showed at the beginning to modify the quality of life of every child admitted even with other kind of Epilepsy. The results of

this research paper were 97 percent successfully, a reference to every epileptic child admitted in our country.

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SERUM VITAMIN D AND SOME BONE MARKERS LEVELS IN in Epileptic Egyptian Children on Antiepileptic Drugs

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Objectives: The study was designed to estimate serum vitamin D level and some biochemical markers of bone turnover in a group of Egyptian children with epilepsy on antiepileptic drugs**.**

Patients and Methods: This case control cross sectional study was conducted on thirty children with epilepsy (19 males, and 11 females) on anticonvulsant therapy. They were divided into two subgroups according to their mode of therapy; 15 patients under polytherapy in group I, and 15 patients under monotherapy in group II. Twenty apparently healthy children of matched age and sex were recruited as control group

Results: Epileptic patients on polytherapy had highly significant low serum 25- hydroxy vitamin D level compared to those on monotherapy ($p<0.001$) with highly significant differences between patients versus controls ($p<0.001$). Differences between the cut off categories were highly statistically significant for patients versus controls ($p<0.001$), and among the polytherapy versus monotherapy subgroups ($p<0.001$). The mean serum calcium levels were found to be statistically significantly lower in epileptic patients compared to control ($p<0.01$) with no significant difference between the patients subgroups. The mean serum levels of alkaline phosphatase were statistically significant higher in the patients group compared to controls ($p<0.001$), with no statistically significant difference between patients subgroups.

Conclusion: Our results revealed high risk of vitamin D deficiency in epileptic children on antiepileptic drugs especially those under long term polytherapy. Routine monitoring of serum 25- hydroxy vitamin D is recommended with vitamin D supplementation on an individual basis.

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CORRELATION BETWEEN SOCIAL COMPETENCE AND NEUROCOGNITIVE PERFORMANCE IN CHILDREN WITH EPILEPSY.

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Introduction. Epilepsy may affect child's social competence and cognition. Important part of social competence is Theory of Mind – ability to understand others' mental states. The purpose of the study is to explore factors related with social competence and epilepsy affecting neurocognitive development and social competence.

Methods 35 children with epilepsy (M=10.46 yrs; SD=1.85): 25 with partial and 10 with generalized epilepsy, and 30 controls (M=10.26 yrs.; SD=1.88) participated. 17/35 children had newly diagnosed epilepsy and were tested before the beginning of antiepileptic treatment. Theory of Mind Tasks and Social Skills Rating Scale were used as social competence measures. Neurocognitive development was assessed using NEPSY battery.

Results: We revealed that children with epilepsy have better understanding of false beliefs than intentional lying ($p<.01$) and sarcasm ($p<.05$). Compared to controls, they had more behavioural problems ($p<.01$) and performed significantly worse in attention, executive, verbal and fine motor skills ($p<.05$). Children with better understanding of false beliefs also had better executive ($r=.6$; $p<.01$), verbal ($r=.45$-$.49$; $p<.01$) and visuospatial skills ($r=.34$-$.48$; $p<.01$). Children with generalized epilepsy had lower scores on sarcasm ($p<.05$) and memory ($p<.05$) compared to children with partial epilepsy. Age of epilepsy onset correlated positively with performance in Theory of Mind Task ($r=.42$; $p<.01$). Children who got AED had significantly lower fine motor and memory skills ($p<.05$).

Conclusion: Children with epilepsy have lower social competence and neurocognitive skills than healthy children. Furthermore, social competence is correlated with attention and executive abilities. Type of epilepsy and age of onset also affects social competence.

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THE USE OF CHROMOSOMAL MICROARRAY IN COMMON CLINICAL PRACTICE OF NEWLY DIAGNOSED EPILEPSY OF CHILDREN IN ESTONIA.

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Background: Epilepsy is one of the most common neurological disorders in humans with prevalence of 1% and lifetime incidence of 3%. Several chromosomal abnormalities have been described that may cause epilepsy and intractable epilepsy syndromes but the role of many copy number variants (CNV) is unknown.

Microarray-based genomic copy-number analysis (chromosomal microarray - CMA) gives a chance to detect very small chromosomal imbalances associated with different diseases. Our aim was to find out the frequency of using CMA in newly diagnosed epilepsy in common clinical practice in Southern Estonia.

Methods: the study group included all newly diagnosed children with epilepsy from 2009-2010 in Southern Estonia. All the preformed CMA-s as common clinical practice and other investigations were recorded in addition to general characteristics of the patients.

Results: From 122 children with newly diagnosed epilepsy CMA was performed in 15 (12.3%). From these 15 children 11 were boys (73.3%). 20% (3) of CMA had some changes, and 2 (66.6%) had mutations that can be related to epilepsy. One patient had 8p23 syndrome (intellectual disability, dysmorphic features and epilepsy), the other had microdeletion on 22q13.2 that consists the MCHR1 gene.

Intractable epilepsy or epileptic encephalopathy developed in 17 (13.9%) of 122 newly diagnosed epilepsy patients. As a first line diagnostic investigation CMA was performed in 8 (53,3%) of 17 children.

Conclusion: CMA is a useful diagnostic tool in detection of CNV's in epilepsy and it should be part of common clinical practice.

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DOES VIGABATRIN TREATMENT FOR INFANTILE SPASMS CAUSE VISUAL FIELD DEFECTS ? AN INTERNATIONAL MULTICENTER STUDY

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The aim of this study was to examine infantile spasms patients at school age for visual fields to see whether VGB treatment in infancy had caused defects.

This study included 35 children: ten children with tuberous sclerosis (TS), three with other symptomatic and 22 with cryptogenic aetiology for their spasms. Visual fields were examined by the Goldmann kinetic, by the static Humphrey or Octopus perimetry. Visual fields were re-evaluated by the ophthalmologist.

Typical VGB-attributed visual field defects were found altogether in 11/32 (34%) patients. Three of 35 patients were excluded because of VFD of other origin (2) or only with ERG study (1). The defects were mild in 5 and severe in 6 cases. One child out of 12 children (8%) who used VGB for less than one year (Group 1) had mild VFD. Three of the 9 patients (33%) using VGB for up to 22-24 months (Group 2), and 7/11 patients

(63%) using VGB for more than 2 years (Group 3) had VFDs. Defects were mild (1) and severe (2) in Group 2, and mild (3) and severe (4) in Group 3. The mean cumulative doses were 155 g (Group 1), 808 g (Group 2) and 2547 g (Group 3), respectively. The patients with TS had more VFDs (6/10 patients).

VFDs were found to occur at the same frequency as that reported in adults. The plasticity of an infant retina seems not to protect from damage. The risk/benefit ratio should always be carefully considered when using VGB.

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USING SELF-REGULATION CONCEPTS TO PREDICT SLEEP PROBLEMS IN ADOLESCENTS WITH EPILEPSY.

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Introduction: Multiple epilepsy and sleep related variables have been explored in order to elucidate the complex relationship between the two. We aimed to explore the extent to which epilepsy severity, gender, and self-regulation concepts (illness perceptions, autonomous treatment regulation, perceived autonomy support by parents) predict sleep problems in adolescents with epilepsy.

Methods: N=100 adolescents (**Mage**=13. 9**, SD**=2.21, 59% boys). The Brief Illness Questionnaire, the Treatment Self- Regulation Questionnaire, the Perceptions of Parents Scales and the Athens Insomnia Scale were administered; multiple hierarchical regression analysis (5 models) was conducted.

Results: Most patients (91%) were well controlled on anticonvulsants; 6 % were pharmacoresistant; 3% had infrequent seizures. In model 1, gender significantly predicted sleep problems (R2=0.039, adjusted R2=0.029, F=3.955, df=1, P<0.05). Adding epilepsy severity to the regression (model 2: R2=0.041, adjusted R2=0.021, F=2.066, df=2, **n.s.**), the illness perceptions (model 3: R2=0.238, adjusted R2=0.189, F=4.844, df=6, **P**<0.001), the autonomous treatment regulation (model 4: R2=0.239, adjusted R2=0.181, F=4.121, df=7,**P**<0.001), and patients' autonomy support (model 5: R2=0.240, adjusted R2=0.173, F=3.583, df=8, **P**<0.001), a significant increase in explained variance occurred.

Conclusions: At a univariate level being a girl, having less severe epilepsy, expecting more consequences from epilepsy, believing that the disease will last longer, experiencing the symptoms intensely, being more concerned about epilepsy, regulating less autonomously the treatment and perceiving more autonomy support by parents were significantly associated with more sleep problems. However, at a multivariate level when the influence of consequences was taken into account all other relations vanished.

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USING A SELF-REGULATION FRAME OF REFERENCE FOR THE PREDICTION OF QUALITY OF LIFE IN ADOLESCENTS WITH EPILEPSY

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The link between self-regulation concepts, epilepsy and Quality of Life (QoL) remains inadequately investigated. We aimed to explore the extent to which epilepsy severity, gender, and self-regulation concepts (illness perceptions, autonomous treatment regulation, perceived autonomy support by parents) predict QoL in adolescents with epilepsy.

Methods: Interviews using the KIDSCREEN-27 Questionnaire, the Brief Illness Perceptions Questionnaire, the Perceptions of Parents Scales and the Treatment Self-Regulation Questionnaire were conducted in 100 patients ($\mathbf{M_{age}}$=13,9, **SD**=2.21, 41% girls). A multiple hierarchical regression analysis was performed.

Results: Most patients (91%) were well controlled on antiepileptics; 3% had infrequent seizures; 6 % were pharmacoresistant. In model 1, gender predicted significantly QoL (R^2=0.078, adjusted R^2=0.069, F=8.32, df=1, **P**<0.010). Adding epilepsy severity (model 2: R^2=0.117, adjusted R^2=0.099, F=6.42, df=2,**P**<0.010), illness perceptions (model 3: R^2 =0.339, adjusted R^2=0.281, F=5.83, df=8,**P**<0.001), autonomous treatment regulation (model 4: R^2=0.378, adjusted R^2=0.316, F=6.08, df=9, **P**<0.001), and patients' autonomy support (model 5: R^2 =0.423, adjusted R^2=0.358, F=6.52, df=10, **P**<0.001), a significant increase in explained variance occurred.

Conclusions: At a univariate level male gender, having less severe epilepsy, expecting less consequences from epilepsy, believing that the epilepsy will not last long, freedom, from symptom, being less concerned about epilepsy, the feeling of having more personal control over the disease and less treatment control, regulating less autonomously the treatment and perceiving more autonomy support by parents were significantly associated with better QoL. However, at a multivariate level it appeared that less concern, less autonomous treatment regulation and more autonomy support result in better QoL.

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CLINICAL PROFILE OF CHILDREN WITH LATE ONSET SPASMS - A REPORT OF 24 CASES

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Introduction: West syndrome is a common age-related epileptic encephalopathy with onset usually in the first year of life. This study aimed to evaluate the clinico-etiological profile and short term seizure outcomes in children with epileptic spasms with onset after 12 months of age.

Methods: The medical records of children diagnosed with West syndrome with onset of spasms after 12 months of age at a tertiary care, referral hospital in North India from January 2011 to June 2013, were retrospectively reviewed. The clinic-etiological profile and treatment response were studied.

Results: During the study period, 253 cases were diagnosed with West syndrome. 24 (9.5%) children had onset of spasms after 12 months of age. The mean age at presentation and onset of spasms were 20.4 months (SD-5.4) and 15.7 months (SD-4.8), respectively. 91.7% were males. Perinatal asphyxia was the most common etiology (16; 66.7%) followed by cryptogenic (3; 12.5%), tubercular meningitis (2; 8.3%), neonatal hypoglycemia sequalae (1; 4.2%), tuberous sclerosis (1; 4.2%) and bacterial meningitis (1; 4.2%). EEG showed hysparrhythmia (11; 45.8%) or modified hypsarrhythmia (13; 54.2%). The median duration of follow-up was 6 months (6-10). The response to oral prednisolone was: 100% (11; 45.8%), >50-99% (3; 12.5%) and < 50% (7; 29.17). At last follow up, 11 (45.8%) children were spasm-free, 7 (29.2%) had persisting spasms and 6 (26.1%) had other seizure types (2 evolved to Lennox-Gastaut syndrome).

Conclusions: Epileptic spasms may appear after 12 month of age. Perinatal asphyxia remains the most common etiology. The response to oral steroids appears to be good.

P138

ATTENTION DEFICIT HYPERACTIVITY DISORDER IN CHILDREN WITH EPILEPSY

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Introduction: Attention Deficit Hyperactivity Disorder (ADHD) is a common co morbidity experienced by children with epilepsy (CWE), and has a negative impact on their social functioning, quality of life, and represents a significant risk factor for academic underachievement.

Purpose: To assess the prevalence of ADHD and its characteristics and risk factors, in children with epilepsy.

Methods: Consecutive cases of active epilepsy, aged 6-12 years, were assessed for ADHD using Diagnostic and Statistical Manual of Mental Disorders-Fourth edition-Text Revision criteria. Exclusion criteria comprised an Intelligence quotient of 70 or less, associated other chronic disease. Children with co-morbid ADHD were compared with those who did not have it.

Results: Among 74 children (39 males and 25 females) with epilepsy, 17(22.9%) were diagnosed with ADHD. Of these 17 patients, 10 (58.8%) had ADHD predominantly inattentive type, 6 (35.3%) had ADHD

combined and 1(5.9%) had ADHD predominantly hyperactive-impulsive type. On univariate analysis male gender, epileptiform discharges on EEG and borderline intellectual functioning were associated with ADHD (**p**<0.05). On multivariate logistic regression only borderline intellectual functioning independently predicted ADHD (odds ratio 13.0; 95% CI 3.7 – 87.6; **p**< 0.001).

Conclusion: The demonstrated high frequency of ADHD in CWE suggests that pediatrician and pediatric neurologists should be sensitive to identification of ADHD in CWE.

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LONG TERM OUTCOME OF CHILDREN WITH WEST SYNDROME: A RETROSPECTIVE CASE RECORD ANALYSIS

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Introduction: West syndrome is an infantile epileptic encephalopathy with heterogeneous etiology and variable course. This study describes the clinical, therapy and outcome related characteristics of children with West syndrome in a tertiary care hospital in north India.

Materials and Methods: In this retrospective analysis, 310 children with West syndrome enrolled from January 2009 to March 2013 were reviewed.

Results: The median age at onset of spasms was 5 months and age at presentation was 18 months. The predominant spasm semiology was flexor [78.4%], perinatal cerebral insult [55.5%] being the commonest etiology. 91.6% (284/310) children received ACTH (140) or oral steroid (144) as first line therapy. Difference in proportion of patients showing complete resolution of spasm between ACTH (40%, 56/140) and oral steroids group (46.5%, 67/144) was not significant (p=0.27). Nineteen patients received ketogenic diet with 15 (79%) showing greater than 50 % seizure reduction. In 46.5% patients spasms resolved, 9% evolved to Lennox-Gastaut syndrome, 18.1% developed other seizure types and 26.5% had persistent spasms at last follow up (median follow up of 10 months). The median age at initiation of treatment of choice was significantly less (p<0.001) in patients who showed complete cessation of spasm (15 months) compared to those with partial or no response (20 months).

Conclusions: ACTH is the first line treatment in west syndrome, however oral steroids may be considered in resource-limited settings. Dietary therapy should be considered in pharmacoresistant cases. Early identification, appropriate management and good supportive care are critical for optimal neuro-developmental outcome.

P140

EFFICACY AND SAFETY OF ORAL TRICLOFOS AS SEDATIVE FOR CHILDREN UNDERGOING SLEEP EEG – AN OBSERVATIONAL STUDY

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Introduction: Triclofos is the most commonly used oral sedative in children. This study aimed to evaluate the efficacy and safety of oral Triclofos as sedative for children undergoing sleep EEG.

Methods: This observational study was carried out between 1st October 2013 and 30thNovember 2013 at a tertiary care centre in North India. Consecutive children aged 6 months to 5 years referred for sleep EEG evaluation who required triclofos for sedation were enrolled. After informed consent, clinical details were noted as per a pre-designed Proforma. Triclofos was administered orally at 50 mg/kg followed by EEG recording. Details of sedation, EEG recording and adverse effects were then noted.

Results: Sixty-one children were enrolled during the study period. The median age was 24 months (15-36) with 69% males. The mean duration of sleep in preceding 24-hours was 9.5 (2.5). The median sleep-onset latency after triclofos administration was 27.5 minutes (15-45). The median duration of sleep was 90 (35-120). 14 (23%) required second dose of triclofos. The EEG recording of at least 30 minutes was successful in 56 (91.8%) children. Two children (3.3%) had mild adverse effects within 24 hours (1-vomiting, 1-dizziness).

Conclusions: Triclofos is an effective and safe oral sedative for children undergoing EEG.

P141

SERUM TRACE ELEMENT LEVELS IN CHILDREN RECEIVING ANTIEPILEPTIC DRUG THERAPY: A CROSS-SECTIONAL STUDY

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Introduction: Few studies have evaluated the effects of antiepileptic drug (AED) therapy on trace element status in children and their results have been conflicting. The newer AEDs are considered to have a more acceptable safety profile, but this confidence is somewhat guarded in the absence of long-term data. This cross-sectional study was conducted to analyze serum trace elements levels in epileptic children treated with conventional and newer AEDs and compare them with healthy controls.

Methods: The study included 92 epileptic children and 28 healthy controls. The participant distribution was as follows, Group I: Phenytoin (PHT) (n=35), Group II: Valproate (VPA) (n=30), Group III: Valproate plus Levetiracetam (VPA+LEV) (n=27), Group IV: Healthy controls (n=28). Serum levels of seven trace elements i.e. zinc, copper, magnesium, manganese, iron, selenium and strontium were determined using inductively coupled plasma-atomic emission spectrometry (ICP-AES).

Results: Phenytoin monotherapy was associated with increased copper (1568.8µg/L vs. 1053.6 µg/L, p=0.009) and strontium (37.0µg/L vs. 30.7µg/L, p<0.001) concentrations & decreased manganese levels (1.5µg/L vs. 1.9µg/L, p=0.04). Valproate monotherapy treated children had decreased serum zinc (1010.5µg/L vs. 1242.9µg/L, p=0.01) and selenium levels (67.0µg/L vs. 84.7µg/L, p=0.02) as compared to healthy controls. However, in VPA+LEV group no significant differences were observed in trace-element profile as compared to healthy children.

Conclusions: A significant difference in trace element levels in VPA and PHT treated children suggests a possible association between AED therapy and trace element alterations. However, levetiracetam in combination with valproate was not associated with these alterations. These findings further support its favorable adverse effect profile as compared to conventional AEDs.

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HYPERHOMOCYSTEINEMIA: ESSENTIAL EVIL IN CHILDREN ON ANTI EPILEPTICS?

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Introduction: Elevated plasma Homocysteine concentration is associated with increased risk for vaso-occlusive disease like cerebrovascular stroke, coronary artery disease, and also the risk of resistance to anti-epileptics and refractory epilepsy. Hyperhomocysteinemia has been frequently associated with the administration of Anti epileptic drugs (AED). This study aims at evaluating the effect of anti-epileptic therapy on serum Homocysteine levels in children.

Methods: 53 children (Males – 32, Females – 21) with Seizures in age group of 6 months–14 years, were recruited from the Pediatric Outpatient and Inpatient department of a tertiary Hospital were included in the study. Serum Homocysteine (Hcy)levels of Children already on AEDs for >6months (Group A) were compared with Children before Initiation of Anti epileptic drugs (Group B).These children were followedup after 6 months of Anti epileptic therapy and Serum Homocysteine was compared (Group C).

Results: Average Hcy levels in subjects who had already received >6 months of antiepileptic drug therapy were 12.58±2.68 µmol/l, compared to 8.83±2.82µmol/l, at recruitment (p=0.001). Significant increased levels were also observed in children followed up after 6 months of AED –10.27±3.06 (µmol/l) compared to 8.63±2.90 (µmol/l) at initiation of AED. 9 children who received >1 AED had significantly higher levels – 14.15±2.56 (µmol/l) compared to children on monotherapy– 10.22±3.06 (µmol/l). Carbamazepine therapy for 6 months caused significant increase in Hcy 10.78±2.82 (µmol/l) compared to baseline of 9.30±2.70 (µmol/l) (p=0.016).

Conclusions: AEDs in children, particularly in those receiving multidrug or long duration treatment, cause hyperhomocysteinemia. This holds immense significance in malnourished populations.

P143

THE EFFECT OF DEPRESSION ON COGNITIVE PERFORMANCE IN CHILDREN WITH REFRACTORY MESIAL TEMPORAL SCLEROSIS (MTS)

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Aim: The aim of the present study focuses the difference between high and low depression and cognitive functions of Mesial Temporal Sclerosis (MTS) children, and to study the relationship between depression and cognitive performance.

Method: Thirty-six MTS children were included in the study, and the subjects were set up by a median split of the scores obtained on the Centre for Epidemiological Studies – Depression Scale for Children (CES-DC) high depression (N = 15) and low depression (N = 21). Standardized neuropsychological tests were employed in the domains of motor speed, mental speed, attention, working memory, executive functions and learning and memory.

Results: Group Analysis: Mean, SD, ANOVA and correlation analyses were used to interpret the data. **Individual analysis:** Scores were compared with age, education and gender specific norms.

High and low depression groups significantly differ on visual learning & memory [(CFT-copy, (**P**=0.019), CFT-IR, (**P**=0.01), CFT-DR- **(P**=0.008)]; and verbal learning & memory [(AVLT-Total (**P**=0.008), AVLT-IR (**P**=0.01), AVLT-DR (**P**=0.012); LM-IR (**P**=0.027)}. There is a significant relationship between high depression and the neuropsychological variables on CFT-DR (P=0.05), LM–IR (**P**=0.19), LM-DR (**P**=0.016) and DSST (**P**=0.024).

Conclusion: The MTS children who performed low in cognitive functions reported higher level of depression. MTS children's cognitive impairments are seen more in attention, processing speed, learning and memory functions. MTS children are not affected in the areas of focused & divided attention, motor speed, mental speed, comprehension and executive functions. It was concluded that high depression is associated with Bi-temporal involvement.

P144

COMPARATIVE STUDY OF FEBRILE SEIZURES VS GENERALISED EPILEPSY WITH FEBRILE SEIZURE PLUS SYNDROME

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Introduction: Generalized epilepsy with febrile seizure plus (GEFS+) described as autosomal dominant inherited epilepsy disorder. Currently no literature available from India.

Aims& objectives: To study and compare seizure profile and need for anti-epileptic drugs (AED) in febrile seizures versus GEFS group at tertiary care pediatric center.

Study design and setting: Prospective observational study, Non interventional study.

Results: Total 176 children enrolled in study, 75 were under GEFS group and 101 under Febrile Seizuregroup. Mean age of onset at 1st seizure was 17.9 months (± 12.45) in GEFS group and 19.87 months (+ 11.77) in febrile seizure group. Male: female ratio 1.58:1 in GEFS group and 1.72:1 in febrile seizure group. 45% had first febrile convulsion below 12 months v/s 21% in febrile seizure group. Longer duration of seizure >5minutes seen in GEFS group than febrile seizures (32% vs 17%). Recurrence in 52% of GEFS Vs 16% febrile seizure group. Recurrence of febrile and afebrile seizure 48% and 24% in GEFS, 15% and 1% in Febrile seizure. AED required in 33% of GEFS group v/s 1% of febrile seizure. AED (1 yr follow up) continued in 24% of GEFS v/s 1% Febrile seizure group. Neuroimaging and EEG were poor predictors of recurrence. Speech and behavioral issues noticed in 4% of GEFS, none in Febrile seizure group.

Conclusion: Children with GEFS plus had higher incidence of febrile & afebrile seizures recurrences, higher need of AED 33% v/s 1% in febrile seizure group. Speech and behavioral issues noted in GEFS group.

P145

CLINICO-ETIOLOGICAL PROFILE OF INFANTS WITH FIRST SEIZURE: AN OBSERVATIONAL STUDY FROM A DEVELOPING COUNTRY.

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Introduction: The risk of seizures is the highest in infancy but there is not much data from this region on infants with first seizure.

Methods: We studied 75 (61.3% males) consecutive infants (28 days-1 year) presenting with their first seizure to the pediatric emergency. Seizures were classified as per ILAE Classification, 1981. Seizure semiology was determined based on eye-witness account (77.3%), or direct observation. Routine biochemical studies, inter-ictal EEG, and developmental assessment were done in all infants. Neuroimaging was done selectively.

Results: Mean age was 5.8±3.4 month and 42.7% had seizures as their only complaint; fever was the most common co-morbidity. 57 (76%) patients presented with a first seizure. 93.3% infants had short-lasting (<15 min), and 72% generalized seizures. Biochemical studies were abnormal in 27 (36%), with hypocalcemia in 26. 12 CT scans and 10 MRI studies were done in 20 patients; in unprovoked seizures, only 31% of these provided any diagnostic information. Majority of the infants had provoked seizures (68%), 1/3[rd] of which were due to hypocalcemia. 29.3% had neuroinfections (pyomeningitis, 21.3%). Eight (10.7%) infants had febrile seizures and 5 had Benign infantile convulsions. Thirteen (17.3%) infants had developmental delay, with majority having moderate delay. Nine (12%) infants died during the duration of the study, 2 during the course of a seizure.

Conclusion: Metabolic derangements and neuro-infections were the commonest etiology. Existing management guidelines for infants with an initial seizure need to be modified for our region.

P146

KETOGENIC DIET CAN BE IMPROVED WITH A HIGH POLYUNSATURATED FATTY ACID CONTENT

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Introduction: The ketogenic diet (KD) is used to treat persons with uncontrolled epilepsy. One proposed mechanism of KD is the elevation of polyunsaturated fats (PUFA) resulting in increased resistance to seizures in ketotic brain tissue. The PUFA-enriched KD may achieve a greater level of ketosis than a traditional KD. We hypothesized that a PUFA enriched KD would help attain a greater degree of seizure control as compared to mixed fats KD (MFKD).

Method: This was a prospective open, non-blinded study of 27 patients of age 11 months to 16 yrs (mean= 7yrs) who had inadequate seizure control on MFKD. They were changed to high PUFA KD. MFKD consists of 32.1% PUFA while PUFAKD consists of 73% PUFA.

Results: A total of 10 (out of 27) patients had more than 90% reduction (of these 3 had 100% reduction) with a responder rate of 66% (> 50% reduction). No serious side-effects were seen.

Conclusion: PUFA KD could be tried in those who fail classical MFKD. Out of the four trials to date in one 5 out of 21 had a greater than 50% reduction in seizures while in three there was no change. A randomized control trial of PUFAKD against MFKD would help to decide whether PUFAKD should be used routinely in preference to MFKD.

Other studies: gave lesser or no improvement. This is probably because the period was too short or the amount of PUFA was inadequate.

P147

SIGNIFICANT MALE PREVALENCE IN CHILDREN WITH WEST SYNDROME IN INDIA

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Introduction: West syndrome is a peculiar age dependent epilepsy syndrome, characterized by constellation of clustered spasms and hypsarrhythmia pattern on EEG. There is data from Indian studies which suggest male preponderance in children with West syndrome. The objective of the present study was to investigate this skewed gender ratio.

Methods: We investigated this association via systematic review of published studies from India. We used a systematic review strategy as there is no national population based registry for West syndrome. We searched in PubMed, EMBASE, Scopus and Web of Science database for publications in English language from January 2001 to November 2013 using key terms: West Syndrome OR Infantile Spasms and India. Data on gender predominance and demographics were extracted from all the sources and incorporated in our study.

Results: We identified four eligible published studies. A total of 391 children with West syndrome in these studies were assessed for gender ratio. Male preponderance was observed in 72.9% of study population. There is also significant interregional variation in gender ratio.

Conclusion: Our study clearly demonstrated significant high male prevalence in children with West syndrome in India. There is a clear need of prospective multicenter study to confirm this observation and assess its implications.

P148

LONG-TERM OUTCOME OF MEDICALLY-TREATED DRUG-RESISTANT EPILEPSY IN CHILDREN

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Introduction: About 20% of patients develop drug –resistant epilepsy (DRE). There are only few studies on outcome of medically-treated DRE in children.

Methods: This observational study was carried out in a hospital-based child neurology clinic in a large Indian city. Patients included were 1) Onset age< 13 yrs 2) Mean frequency of 1 seizure / month for at least 2 years despite therapy with =>2 AEDs. 3) Minimum 5 years from epilepsy onset. Chart reviews and caretaker interviews were used. Progressive disorders, idiopathic epilepsies and surgical patients were excluded.The Kaplan Meier survival analysis was used to construct a life table to calculate the rate of remission per year, using <1 seizure per year as a definition for remission. Poor control was defined as <75% improvement while good control as > 75% control on day of data entry. The Pearson Chi Square test was used to test the association between various characteristics and poor control.

Results: Of 62 patients, follow-up information was sufficient in 55. Follow up was between 6-33 years (mean 16 yrs). Of the 55 patients 36(65%) had >75% improvement (22 patients were seizure free) and 19(35%) had <=75% improvement. The percentage of patients who remit per year was 2.84%. No characteristic showed a clear association but abnormal imaging & use of =>5 AEDs showed a tendency for association with <75% improvement.

Conclusions: The long-term outcome in medically-treated DRE in children is not as pessimistic as would seem. Our results however show a lower remission rate as compared to earlier publications.

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SAFETY, FEASIBILITY AND EFFECTIVENESS OF ORAL ZONISAMIDE MONOTHERAPY IN COMPARISON WITH INTRAMUSCULAR ADRENOCORTICOTROPIC HORMONE IN INFANTS WITH WEST SYNDROME

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Introduction: There is emerging evidence to suggest effectiveness of oral zonisamide therapy in children with West syndrome. The objective of the present study was to elucidate the safety, feasibility and effectiveness of oral zonisamide therapy in infants with West syndrome in comparison with intramuscular ACTH therapy.

Methods: Study design: Pragmatic, Randomized-controlled, open-label, non-inferiority, pilot study, concealed allocation, parallel-group assignment. Randomization was done via computer generated random table and allocation was concealed. Newly diagnosed 30 cases of West syndrome (age 6-12 months) attending the outpatient department were studied. Suspected or proven cases of tuberous sclerosis and neurometabolic disorders were excluded. The efficacy outcomes were cessation of spasms (as reported by parents) and resolution of hypsarrhythmia in EEG by day 14 after initiation of treatment and safety as incidence of side effects. The trial was approved with institute ethics committee and registered in clinical trial registry.

Results: Preliminary study results revealed oral zonisamide therapy was well tolerated. By day 14 of therapy, 4/15 children had cessation of spasms in zonisamide group, but 6/15 children had cessation of spasms in ACTH group. However, by day 21 of therapy, 8/15 children had cessation of spasms in zonisamide group, but 10/15 children had cessation of spasms in ACTH group.

Conclusions: Zonisamide appears safe and feasible in infants with West syndrome but probably slower and weaker in efficacy.

P150

A STUDY ON UNIQUE ASSOCIATION OF POLYMICROGYRIA, SLEEP-RELATED ELECTRICAL STATUS-EPILEPTICUS AND INTRACTABLE DROP-ATTACKS

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Introduction: An association of polymicrogyria, drop attacks and electrical status-epilepticus during sleep is rare. The objective of the present study was to investigate this association in tertiary center setting and elucidate its profile.

Methods: Retrospective record review of 5 years from Pediatric neurodevelopment unit and epilepsy clinic was done. Clinical details, neuroimaging, serial electroencephalogram recordings and outcome were recorded for children with this entity.

Results: Two children fulfilled the diagnosis. Both boys had global developmental delay, hemiplegic cerebral palsy and focal seizures. Age of onset of focal epilepsy was 1.5 year and 2.5 year. Neuroimaging showed right sided polymicrogyria in both the cases. The onset of drop-attacks was at the age of 4 and 3.5 year respectively. Corresponding electroencephalogram showed electrical status-epilepticus during sleep. Both had associated cognitive decline and behavioral abnormalities with the onset of electrical status-epilepticus during sleep. The condition remained refractory to multiple anti-epileptic drugs and immune-modulatory therapy.

Conclusions: The association of electrical status-epilepticus during sleep and polymicrogyria is rare in childhood and refractory to anti-epileptics.

P151

USE OF THE MODIFIED ATKINS DIET IN LENNOX GASTAUT SYNDROME

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Purpose: Lennox-Gastaut syndrome (LGS) is a catastrophic epilepsy syndrome affecting children. There is scanty data regarding the efficacy and tolerability of the modified Atkins diet in children with LGS.

Materials and Methods: This study was a retrospective review of all children with LGS treated with the modified Atkins diet at our center from May 2009 and March 2011. The diet was initiated in those children who persisted to have daily seizures despite the use of at least 3 appropriate anti-epileptic drugs.

Results: Twenty five children with LGS were started on the modified Atkins diet, restricting carbohydrate intake to ten grams/day. After 3 months, two patients were seizure free, and 10 children had > 50% reduction in seizure frequency. At 6 months, 14 patients were still on the diet. Out of these, 3 were seizure free, and 10 had > 50% reduction in seizure frequency. At one year, nine children were on diet and all had > 50% reduction in seizure frequency. The side effects of the diet were mild and manageable.

Conclusions: The modified Atkins diet was found to be effective and well tolerated in children with LGS.

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FEASIBILITY OF A PARENTING TRAINING PROGRAMME IN PEDIATRIC EPILEPSY

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Introduction Pediatric epilepsy poses significant challenge to the normal development of affected children. Ranging from managing the seizures to providing as near normal development experience to the child as possible, parents are faced with different tasks in caregiving. The present study aims to assess parenting practices and feasibility of parenting training programme in pediatric epilepsy**.**

Methods A pretest post test experimental design is used .Parents of 50 children with Epilepsy attending outpatient department of a tertiary referral center for Neurology are studied. All the children had the diagnosis of Seizure disorder, both male and female, in the age range of one to eighteen years. Father, mother or both parents are included in the programme. All the parents received individualized intervention targeting specific areas of psychosocial needs identified, parenting

stress and parenting practices based on psycho educational model of family intervention.

Results Preliminary analysis shows that there is reduction in parenting stress reported by the parents, and healthier parenting practices. Dimensions specifically showing significant changes are over involvement from parents restricting children's regular activities, drug compliance, normalization of family process and child's development experience. Further analysis is being done in order to consolidate the above findings and will be reported.

Conclusion Psychoeducational Parenting training programme is effective in reducing parenting stress, promoting healthier parenting practices and thus contributing towards better disease management and Quality of life of children with Epilepsy

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SCN1A MUTATIONS IN INDIAN PATIENTS WITH SCN1A-RELATED EPILEPTIC DISORDERS

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INTRODUCTION: Dravet syndrome (DS) and variants are epileptic disorders of infancy and childhood associated with mutations in the SCN1A gene. There is no literature on such mutations in Indian patients with DS and related syndromes.

METHODS: Patients were recruited from a child neurology hospital-based clinic in a large city hospital in India both from a database and prospectively. Patients clinically suspected to have DS and related conditions using standard clinical criteria were offered genetic testing. Approval from the institutional review board was obtained and informed consent taken. Direct sequencing using an automatic sequencer screened mutations in the SCN1A gene. Multiplex ligation-dependant probe amplification (MLPA) was performed on those with no mutations or only missense mutations. 100 healthy controls were also analysed.

RESULTS: A total of 86 patients were included. 13 DS, 27 severe myoclonic epilepsy of infancy borderline (SMEB), 12 GEFS+, 9 febrile seizures (FS) and 25 other intractable cryptogenic epilepsies. 41 patients with mutations were detected including 4 nonsense, 8 frame-shift, 4 splice-site and 18 missense (7 were thought to be benign polymorphisms). 7 patients could not be analysed as information was incomplete. 14 mutations were novel. Mutations were noted in 5/12 (42%) GEFS+, 16 /27 (59%) of SMEB and 13/13 (100%) of DS patients. There was no correlation found between different phenotypes and mutation types.

CONCLUSION: SCN1A mutations were frequently seen in Indian DS / SMEB patients and several of these were novel. Not unexpectedly no genotype phenotype correlation was found as has been reported earlier.

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PREVALENCE OF HAEMOSTATIC ABNORMALITIES IN EPILEPTIC CHILDREN ON VALPROATE MONOTHERAPY: A CROSS SECTIONAL STUDY

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Background: Valproic acid (VPA) is a commonly used drug in children in the management of childhood epilepsies. This cross sectional study was conducted to evaluate the prevalence of various haemostatic abnormalities in children on valproate therapy, as there are no previous studies from India.

Methods: Sixty four consecutive children, aged 3-15 years were enrolled after ethics approval between April 2012 and December 2012. Blood sample was collected after obtaining a detailed history and clinical examination for the analysis of haemostatic parameters, liver function test and serum valproate trough level. Platelet count, prothrombin time (PT), activated partial thromboplastin time (APTT), thrombin time (TT), clot solubility test (CST), d Dimer, PF3 release with ADP at 0 and 20 minutes were performed. In children with prolonged PF3 release at 20 minutes, platelet aggregation test (PAT) was performed.

Results: Prevalence of thrombocytopenia, platelet dysfunction and elevated d dimer in children on VPA monotherapy were 20.6%, 69.3% and 85.7% respectively. There was a significant negative correlation between VPA level and platelet count. Five children with prolonged PF3 release at 20 minutes were observed to have an unclassified platelet function disorder on PAT. PT was prolonged in only one patient; APTT, TT and CST were normal in all the children.

Conclusion: Qualitative and quantitative platelet dysfunction were prevalent in children on VPA monotherapy but none were symptomatic. It is essential to screen the children on VPA therapy for haemostatic abnormalities during stress, major trauma, infections, surgery, bleeding symptoms and high VPA level.

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THE EFFECTIVENESS OF INTRAMUSCULAR MIDAZOLAM COMPARED TO RECTAL DIAZEPAM FOR ANTICONVULSANT IN CHILDREN

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Background: Seizures are one of the commonest neurological problem in Children. Diazepam is one of the most frequently used benzodiazepines both intravenously as well as rectally. Midazolam has 3-4 times better potency than diazepam and can be given IV, rectal or intramuscular. The effectiveness and easier route of these drugs administration are important choice to make faster seizure cessation.

Objective: To determine the effectiveness of intramuscular midazolam compared to rectal diazepam for terminating seizures in children.

Methods: The subjects were children with age from 1 month to 18 years who presenting with acute seizures. Patients were randomised into 2 groups with either received intramuscular midazolam or rectal diazepam for terminating seizures. Time interval from drug administration to cessation of seizures was compared. Log rank analysis was used for statistical analysis. Side effect of both drugs were evaluated.

Results: There were 66 patients, 33 in each groups enrolled the study. The median time interval for seizures cessation with intramuscular midazolam was 45 seconds, otherwise in rectal diazepam group was 180 seconds. There was statistically significant difference time interval between two groups ($p<0.01$). Intramuscular midazolam worked faster in patients with febrile convulsions, epilepsy and encephalitis. Five patients had time cessation of seizure more than 5 minutes. None of the both groups had any side effects which was statistically significant.

Conclusion: Intramuscular midazolam is effective for terminating seizures in children. It can be used as an alternative treatment for acute seizures in patients with intravenous or rectal route difficulties.

P156

VIGABATRIN EFFICACY AND SAFETY IN IRANIAN EPILEPTIC CHILDREN

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Background: Vigabatrin is an antiepileptic drug that its visual field defects are usually asymptomatic and only detectable by perimetry. Electroretinogram response amplitude to full-field 30-Hz flicker shine has been offered to be more specific in predicting visual field defects. This study was designed to determine the efficacy of Vigabatrin in Iranian children with intractable epilepsy and its visual adverse effects.

Method: Children with epilepsy taking vigabatrin referred to Children's Medical Center and Mofid Children Hospital between November 2012 and October 2013 were enrolled in the study. Seizure periodicity and severity were evaluated within 3–4 weeks after Vigabatrin initiation. Full-field electroretinography was conducted for all patients every 3 months.

Result: This study included 67 patients. The average age of patients was 3.1+2.6 years. The age of seizure onset was 1 day to 5 years, and the mean age at seizure onset was 8.57 + 12.8 months. Daily seizures were reduced by ≤56% and 54.5% after three and six months of Vigabatrin therapy. The responder rate, which means a reduction of >50% in seizure severity and duration, was 67.2% and 56.7% at three and six months of Vigabatrin therapy, respectively. Electroretinographic surveys showed normal range parameters despite 3 months of vigabatrin treatment, and just 3 (4.47%) children had been visually impaired at the end of 6-month treatment.

Conclusion: In conclusion, this study confirms the short-term efficacy and safety of Vigabatrin in children with refractory epilepsies. We suggest that vigabatrin is secure for short-term Pediatric antiepileptic treatment, with few cases of visual impairments and that are often reversible.

P157

ATTENTION IMPAIRMENTS IN BENIGN CHILDHOOD EPILEPSY WITH CENTROTEMPORAL SPIKES AND CHILDHOOD ABSENCE EPILEPSY

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Introduction: Benign childhood epilepsy with centrotemporal spikes (BCECTS) and childhood absence epilepsy (CAE), are well-known and clearly defined childhood epileptic syndromes. Both conditions have a number of features in common, including an early age of onset, a good prognosis, and a genetic component. Attention problems are commonly reported in children with BCECTS and CAE, and can interfere with academic performance and daily functioning. The aim of this study is to compare attentional functions in BCECTS and CAE using a computerized test battery.

Methods: 21 children with CAE (10 boys, 11 girls), 21 with BCECTS (12 boys, 9 girls) and 24 controls (12 boys, 12 girls) with comparable IQ (>80) matched for age and sex were enrolled. All participants were tested with a computerized test battery based on a multicomponent model of attention, which included tasks measuring selective attention, impulsivity, focused attention, divided attention, alertness, and vigilance.

Results: Results showed that children with CAE and BCECTS have attentional impairments in several measures of selectivity of attention (divided attention, focused attention and selective attention) associated with impulsivity as compared with healthy controls. Posthoc analysis revealed no difference between CAE and BCECTS patients.

Conclusion: Children with CAE and BCECTS showed impairments in divided, focused and selective attention compared with control subjects. Furthermore, patients with CAE and BCECTS did not show significant difference between them. Our results support recent evidence that BCECTS and CAE share some common pathophysiological mechanisms.

P158

EARLY EEG MONITORING FOLLOWING PRENATAL DIAGNOSIS PREDICTS EPILEPTOGENESIS IN TUBEROUS SCLEROSIS

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Objectives. To describe the role and the usefulness of a close EEG monitoring in children with tuberous sclerosis complex (TSC), following a prenatal diagnosis.

Materials and Methods. We present 3 patients with negative familial history, in which fetal ultrasound revealed multiple cardiac rhabdomyomas. We therefore performed a fetal MRI, which was able to detect the presence of subependymal nodules or tubers, allowing a definite prenatal diagnosis of TSC. Soon after birth, all these children began a close EEG monitoring, with awake/sleep EEG performed every 3-4 weeks. All the families have been informed of the high risk of epilepsy of TSC infants in the first year of life, and have been educated to recognize focal subtle seizures and epileptic spasms.

Results. EEG monitoring allowed us to identify focal epileptiform abnormalities early and to observe their evolution towards multifocal abnormalities. In the child who developed epileptic spasms we observed a further evolution toward secondary bilateral synchronization and a tendency toward a pre-hypsarrhythmic pattern the day before the appearance of spasms. The other two children didn't show seizures at 6 and 7 months of age.

Conclusions. Our data suggests that epileptogenesis is a slow process, taking several weeks or months from the first EEG epileptiform abnormalities to the first seizure. The close EEG monitoring allows us to detect subtle changes in the EEG pattern which might indicate an evolution towards epilepsy. The immediate detection and treatment of seizures is crucial to minimize the possible long-term neurodevelopmental sequelae of early life seizures.

P159

RUFINAMIDE AS ADJUNCTIVE DRUG IN REFRACTORY EPILEPSY DUE TO NEURONAL MIGRATION DISORDERS

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Objective: To evaluate the efficacy and tolerability of add-on Rufinamide in children with refractory epilepsy symptomatic of neuronal migration disorders.

Materials and Methods: We recruited 69 patients in a prospective, open-label, add-on treatment study from 8 Italian centers for pediatric and adolescent epilepsy care according to the following criteria: age 3 or above; focal or generalized seizures refractory to at least three previous antiepileptic drugs (AEDs), alone or in combination, secondary to neuronal migration disorders; two or more seizures per month in the last 6 months; use of another AED, but no more than three, at baseline. Informed consent from parents and/or caregivers was obtained at the time of enrolment.

Results: We enrolled 69 patients with a mean age of 15 years (range 3-43). Forty-three patients (62%) had a 50-99% seizure reduction, and two (3%) became seizure-free. Seizure frequency was unchanged in 18 (26%) and worsened in 6 (8.7%). Twenty-nine patients (42%) reported adverse side effects, whilst taking rufinamide. Irritability was the most common side effect (11 patients), followed by decreased appetite (10), mood shift (6), vomiting (5), drowsiness (4), and decreased attention (2). Blood levels of concomitant anticonvulsive drugs were transiently abnormal in 5 patients.

Conclusion: In our population of severely refractory epilepsy due to neuronal migration disorders, Rufinamide appeared to be effective and generally well tolerated.

P160

EARLY PREDICTORS OF STATUS EPILEPTICUS-ASSOCIATED MORTALITY AND MORBIDITY IN JAPANESE CHILDREN

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Introduction: Reliable early predictors of status epilepticus (SE)-associated mortality and morbidity are not well known in children.

Methods: We conducted a prospective multicenter study of clinical findings and laboratory data acquired at SE onset, and assessed outcomes at the last follow-up examination. In-hospital death during the acute period and neurological sequelae were classified as poor outcomes.

Results: Of the 201 children who experienced their first SE episode, 16 exhibited poor outcomes. Univariate analysis revealed that the following were associated with poor outcomes: young age (≤24 months); need for mechanical ventilation; seizure duration >90 min; seizure intractability; biphasic seizures; Glasgow Coma Scale (GCS) score<13 at 12 h after SE onset; abnormal blood glucose levels; serum aspartate aminotransferase (AST) ≥56 U/L; and C-reactive protein (CRP) levels>2.00 mg/dL. On the other hand, multivariate analysis revealed that GCS score <13 at 12 h after SE onset and age ≤24 months were statistically significant. To examine the earliest predictors of poor outcome when SE was first treated, we reanalyzed the parameters after exclusion of the GCS score. Age ≤24 months, AST levels ≥56 U/L, and CRP levels >2.00 mg/dL were determined to be statistically significant.

Conclusions: Prolonged unconsciousness and young age were highly predictive of poor outcomes in pediatric patients with SE. Moreover, elevated AST and CRP levels were predictors that might be closely associated with the etiology of SE in Japanese children.

P161

REFLEX PERIODIC SPASMS ARE A SEIZURE TYPE CHARACTERISTIC OF 5P- SYNDROME

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5p-syndrome is a genetic disease resulting from a deletion of the short arm of chromosome 5. The most characteristic clinical feature is high-pitched cry. In 5p- syndrome, epilepsy is a quite rare complication, and there have been few case reports with epilepsy. We describe 2 cases of 5p-syndrome, who suffer from a series of epileptic spasms. In case 1, 27-year-old male, seizures were provoked by eating. In case 2,

23-year-old female, seizures were provoked not only by eating but also by micturition. In both patients, ictal EEGs showed a periodic pattern characterized by diffuse high voltage slow waves. The symptoms and EEG findings of them were consistent with periodic spasms (PS) reported by Gobbi et al in 1987. Among few reports of 5p- syndrome with epilepsy, three cases including ours have reflex periodic spasms. We suggest that reflex periodic spasms are specific seizure pattern to 5p- syndrome. Conclusion Periodic spasms are specific seizure pattern to 5p- syndrome.

P163

THE ATTENTION DEFICIT HYPERACTIVITY DISORDER IN CHILDREN WITH EPILEPSY

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Introduction: Attention Deficit Hyperactivity Disorder (ADHD) is known to be more common in children with epilepsy than in the general population. Thirty one to forty percents of ADHD are accompanied with epilepsy. Few studies regarding this matter have been reported in Korea. This study was aimed to evaluate the comorbidity of ADHD in children with epilepsy.

Methods: This is a two big center based, retrospective and controlled study at Cheongju in Korea. Thirty four ADHD children with epilepsy and 38 ADHD children without epilepsy from Chungbuk National University hospital and Cheonju St. Mary's hospital were recruited from January 2005 to June 2010.

Results: In ADHD children with epilepsy, twelve (35.2%) had partial seizures, 11 (32.2%) did generalized seizures and 11 (32.2%) were unclassified. EEG abnormalities were found in the frontal lobe (15 cases), in the central lobe (7 cases), in the temporal lobe (6 cases), and in the occipital lobe (3 cases). In ADHD children with epilepsy, the combined type was majority (76.4%) and otherwise in ADHD children without epilepsy, the inattentive type was main (50.5%) (P=0.004). Learning disability was more common in ADHD with epilepsy than in ADHD without epilepsy (P=0.01).

Conclusion: This study showed that ADHD children with epilepsy are more likely to have combined type (76.4%) and learning disability as compared with ADHD without epilepsy.

P164

A CASE OF VAN DER KNAAP DISEASE PRESENTING SEIZURE AS THE FIRST SYMPTOM

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Introduction: Van der Knaap disease is diagnosed by clinical features and MRI findings. We report a case of Van der Knaap disease in infant.

Case description: A 5 month-old female infant presented with seizures of simple partial motor type. Personal and family history was negative. Investigations for Inborn error of metabolism done at birth were negative. Head circumference was 41.5 cm which was about 60 percentile for age. The psychomotor development was not delayed. The neurological examination was unremarkable except mild left upper and lower limb weakness with the power of grade 3, which improved soon.

Results: Blood tests including CBC, liver function test, electrolytes, lactate and pyruvate were normal. EEG showed a high amplitude delta slow waves of right parietotemporooccipital areas. Brain MRI revealed the oval cystic lesions in the right parieto-occipital white matter and thalamus.

Conclusion: This is the case of Van der Knaap disease presenting seizure as an initial manifestation and needs the follow up in terms of the development of neurological deficits and psychomotor delay.

P165

A COMPARISON STUDY ON SIMPLE AND COMPLEX FEBRILE SEIZURES IN KOREA

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Introduction: Febrile seizures can be classified as simple and complex type, the latter being characterized by increased risk of recurrence of febrile seizures itself and progression to epilepsy. The present study was conducted to compare the clinical characteristics of simple and complex febrile seizures in Korea.

Methods: Between January 2009 to December 2012, 545 children were diagnosed as febrile seizures. Their medical records were retrospectively reviewed for comparison between simple and complex febrile seizures.

Results: The ratio of male and female was 1.74:1. The age was distributed from 3 months to 8 years. Simple febrile seizures were 431 cases, four-fold larger than complex febrile seizures (114 cases). The causes of their febrile illness were upper airway infection (56%), pneumonia (4.2%), otitis media (4.2%), and acute gastroenteritis (3.9%). Family history of epilepsy was more frequent ($p<0.05$) in children with complex febrile seizures (3/114, 2.6%) compared to simple types (7/431, 1.6%) although no significant differences were found between the two types of febrile seizures in most clinical parameters such gender, age, and the cause of febrile illness.

Conclusion: Although febrile seizures is known as benign, which does not leave any sequelae in most cases, some view it as a basic type of epilepsy caused by fever. An understanding of the natural history and prognosis will enable the physician to reassure the families and provide appropriate counselling and management while avoiding unnecessary diagnostic and therapeutic intervention.

P165

A RETROSPECTIVE ANALYSIS OF PATIENTS WITH FEBRILE CONVULSION(FC) FOLLOWED BY UNPROVOKED SEIZURE ON JEJU ISLAND, SOUTH KOREA

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Introduction: We performed this study to confirm the known risk factors and to identify possible new risk factors for subsequent unprovoked seizure after febrile seizure (FS) on Jeju Island, South Korea.

Methods: A population-based retrospective study of 204 children with FS, whose first FS developed between March 2003 and August 2011, and who were seen in the Pediatric Department at the Jeju National University Hospital.

Results: Two hundred four children (136 boys and 68 girls) were enrolled in this study. The average age at the first FS was 19.2 months. The average total number of FSs was 4.3. A family history of FS or epilepsy was found in 29.4% and 7.8% of patients, respectively. Abnormal findings of EEG were observed in 35.8%. Complex features in the first FS were noted in 28.9%. Subsequent unprovoked seizures occurred in 23.0%. Univariate analysis showed that low parental educational level was one of several variables that were significantly related to unprovoked seizure. Multivariate analysis identified the following factors as significant predictors of unprovoked seizure: late onset of FS at age >3 years, complex features in the first FS, family history of epilepsy, abnormal findings on EEG, and FS developed at a body temperature of <39 °C.

Conclusions: We confirmed the known risk factors for subsequent unprovoked seizure and found that low parental educational status may be a new univariate prognostic indicator. However, further investigation using larger populations and a prospective design is needed to confirm that this is a valid prognostic factor for FS.

P167

CHARACTERISTICS OF HYPOXIC ISCHEMIC ENCEPHALOPATHY WITH SEIZURE IN CHILDREN

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Aim: This study was aimed to evaluate the clinical features of hypoxic ischemic encephalopathy (HIE) in children with and without seizures.

Methods: Fifty five children who had been diagnosed as HIE at our hospital from June 1999 to December 2011 were enrolled in this study. Subjects were divided into two groups according to the presence/absence of seizures and their medical records were retrospectively analyzed.

Results: Among the 55 cases, 34 patients (61.8%) had seizures, while 17 patients (32.2%) did not. Male to female ratio is was 1:1 for the 'seizure' group, the 'no seizure' group was a 2.5:1. The onset age was 9.7 months for the 'seizure' group and 10 months for the 'no seizure' group. The most common risk factor was birth asphyxia (17.7%) for the 'seizure' group, and prematurity (23.8%) for the 'no seizure' group. The most common symptom other than seizure was respiratory arrest for both groups. On radiologic imaging studies of the brain, main lesions were most commonly observed in the cerebral cortex in both groups. The neurologic deficits or death were detected in 67.7% of the 'seizure'

group, and 76.3% of the 'no seizure' group. There were no statistically significant differences in risk factors between the two groups.

Conclusion: Although the characteristics between patients with and without seizures associated with HIE revealed no significant differences, HIE mainly results in death or permanent disability in childhood. Therefore, permanent brain damage may be minimized by early suspicion and treatment in these patients.

P168

CLINICAL AND GENETIC CHARACTERISTICS OF FEBRILE INFECTION-RELATED EPILEPSY SYNDROME

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Background: Febrile Infection-Related Epilepsy Syndrome (FIRES) is a catastrophic condition characterized by a refractory status epilepticus following a febrile illness in normally developing children. Many attempts to find the related genes have been tried without success. This study was aimed to evaluate its clinical and genetic characteristics.

Method: Fifteen patients with FIRES were involved in the study. Their clinical features, lab findings, treatment and outcome were retrospectively reviewed. We also performed whole exome sequencing on 5 out of the subjects. Among hundreds of epilepsy candidate genes, rare (<0.5% in 1000 genomes population), nonsynonymous, loss-of-function and splice-site variants were prioritized.

Results: They initially presented with either partial seizures or partial seizures with secondary generalization. Half of the patients had pleocytosis in CSF. All patients showed diffuse slowing or epileptic discharges in various regions in EEG and normal on initial brain MRI. The mainstay therapy consists of phenytoin, midazolam drip and high dose phenobarbital during the acute phase. Patients who underwent whole exome sequencing revealed shared mutations in eleven genes, which are functionally important in various epilepsy syndromes. No significant variants were identified in **SCN1A, PCDH19, POLG,** or**ABCB1**.

Conclusions: Genetic analysis can be a valuable method for patients with febrile status epilepticus to predict its courses. Further knowledge about the pathogenesis and identification of the molecular/regulatory pathways in FIRES will enable the optimal target to the therapeutic potential of genetic-based drugs.

P169

RELATION TO CORTICAL BLOOD FLOW AND ELECTROGRAPHIC ACTIVITY IN CHILDHOOD-ONSET SEIZURES: CORRELATION BETWEEN MRI-SWI AND EEG

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Aim: To evaluate the relationship between cortical perfusion or venous flow and electrographic activity in the children with seizure using susceptibility weighted imaging (SWI) and electroencephalogrphy (EEG).

Methods: Children presenting with seizures who underwent MRI-SWI and EEG within 24 hours of seizure onset were retrospectively reviewed. The localized area of increased cortical venous flow (SWI+) was assessed using SWI while the abnormal activities such as slowing or epileptiform discharges (EEG+) were investigated on EEG recordings. We defined three groups of patients according to the correlation between MRI-SWI and EEG: (A) no increased venous flow and no abnormal discharges, (B) discordant finding between the SWI+ and EEG+ area, (C) concordant finding between the SWI+ and EEG+ area.

Results: We identified 297 children (194 in group-A, 76 in group-B, and 27 in group-C). The mean age among the three groups was similar (group-A: 3.8±4.6, group-B: 5.0±4.5, group-C: 4.6±4.8 years). The greatest difference among these groups was in seizure frequency and underlying disease. Multiple seizures were revealed more frequently in group-C (12/27, 44.4%) than in group-A (47/194, 24.2%, p=0.026) or group-B (18/76, 23.7%, p=0.041). The incidence of newly-diagnosed epilepsy was significantly higher in group-C (14/27, 51.9%) than in group-A (59/194, 30.4%, p=0.026) or group-B (22/76, 28.9%, p=0.032). By contrast, there were no significant differences in the previous seizure history, seizure types or duration among the three groups.

Conclusion: Seizures with concordant findings between increased venous flow on MRI-SWI and abnormal electrographic activities are more likely to more frequent or real epileptic seizures.

P170

EEG STUDY IN MYANMAR CHILDREN WITH COMPLEX FEBRILE SEIZURES

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INTRODUCTION: Since complex febrile seizure (CFS) is associated with increased risk of future epilepsy, EEGs are frequently ordered in clinical practice. This study aims to examine the effect of various clinical variables on the EEG abnormalities.

METHOD: The study was conducted in Yangon Children Hospital, Myanmar. History, examination, review of medical records and EEG request forms were done, and then standard EEG was obtained for all study children. The individual clinical variables associated with EEG abnormalities were analyzed using multivariate analyses.

RESULTS: Out of 44 children, 21 (47.7%) children were infancy, 18(40.9%) between 1 to 3 years and above 3 years were 5 (11.4%). Only 6 children (13.6%) had EEG abnormalities (95% CI: 0.04-0.24). EEG abnormalities included focal epileptiform discharges (2 children), generalized epileptiform discharges (1 child) and others (3 children). Generalized epileptiform discharges included slow spikes waves in 3/ second in one case and that child with absence seizure came to hospital only because of CFS. There were significant associations between abnormal EEG and two clinical variables -age older than 3 years and focal seizure (p =0.005 and 0.014 respectively). Other clinical variables like sex, seizure duration, postictal deficit, seizure frequency, interval between seizure and onset of fever, previous history and family history of febrile seizure and interval between EEG recording and seizure were not statistically significant.

CONCLUSION: EEG abnormality was more common in children over 3 years and focal seizures. Finding of EEG abnormality may result in closer clinical follow up to monitor for recurrent afebrile seizures.

P171

EPILEPSY: BELIEFS ON CAUSES AND TREATMENT MODALITIES AMONGST CAREGIVERS IN SOUTHERN NIGERIA.

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Introduction: Epilepsy is a chronic brain disorder characterized by recurrent seizure with highest incidence in the first decade of life. Parental beliefs and treatment given, significantly impact adjustment and quality of life of the child. In Nigeria, beliefs on the causes of epilepsy among caregivers, is currently unscientific. Treatment modalities are mainly unorthodox with stigmatization being a major problem. It is more difficult to overcome problems related to the social stigma of epilepsy than the disease condition. Lack of information and inappropriate beliefs contribute to stigma, discrimination, poor self image and access to healthcare.

A**im:** To elicit beliefs on causes that caregivers of children with epilepsy have about the disease and treatment modalities given.

Materials: This is a prospective study from 1st October 2012 to 31st September 2013 in the Neurology clinic of UPTH. 211 caregivers of children with epilepsy were recruited. They were allowed to express themselves on beliefs on causes and treatment options.

Results: All participants had some knowledge prior clinic visits. Common etiological responses were: witchcraft 137 (64.93%), evil spirit 111(52.61%), inheritance 98(46.45%), demons 87 (41.23%), head injury 77(36.49%), insanity 61 (28.91%) and unknown 44(20.85%). Treatment modalities given included: instilling onion juice 203 (96.21%), crude oil into the eyes 198 (93.84%), cow urine to drink 88(41.71%), application of hot spices on the body of a convulsing child 137(64.9%), orthodox medication 48(22.75%) and unknown 8(3.79%).

Conclusions: To reduce morbidity and mortality of children with epilepsy in Nigeria requires educating caregivers with updated evidence- based information.

P172

RISK FACTORS FOR INTRACTABLE EPILEPSY IN CHILDREN

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Background: Intractable epilepsy is defined as lack of seizure control on more than 2 first line antiepileptic drugs with an average of >1 seizure per month for 18 months and no more than 3 consecutive months seizure free during the interval.

Objective: The purpose of the study was to determine the risk factors for intractable epilepsy in children

Method: A retrospective study was conducted with all the patients diagnosed as epilepsy. Files were reviewed from January 2013-July 2013. Univariate analysis was done to assess the risk factors of intractable epilepsy. Results are reported as the odds ratios (OR) with respective 95% CI.

Results: There were a total of 119 patients. Males were 58%. Age ranged from 1 month to 16 years (mean 5.6 years). 40 % had normal and 59 % had abnormal EEG. Of children with abnormal EEG's, 31 were receiving a single drug and 35 were receiving more than 3 drugs. Developmental delay had an OR of 3.759 (95% CI 1.69-8.34). Onset of seizures in children before 2 years had an OR of 4.64 (95% CI 1.639-13.17). Polymorphic seizures had an OR of 2.78(CI 95%2.187-3.549) and generalized seizures an OR of 1.941 (CI 95% 0.912-4.13). Abnormal EEG increased the risk of intractable epilepsy (OR 3.99 CI 95% 1.68-9.44). MRI abnormality did not increase the risk of intractable epilepsy

Conclusion: Early onset of seizures, developmental delay, seizure type and abnormal EEG increases the risk of intractable epilepsy in children

P173

CLINICAL AND EPIDEMIOLOGICAL CHARACTERISTICS OF CHILDREN WITH REFRACTORY EPILEPSY TREATED AT A TERTIARY HOSPITAL IN LIMA, PERU.

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Objective: To describe the clinical and epidemiological characteristics of pediatric patients diagnosed with refractory epilepsy treated at the Hospital Nacional Cayetano Heredia in Lima, Peru.

Methods: This is a prospective and observational study. Consecutively attending patients were enrolled according to the following criteria: (1) aged under 14; (2) established diagnosis of epilepsy; (3) pharmacoresistance to two or more appropriate AEDs. This study was initiated on October 1, 2013 and will end on March 31, 2014.

Results: At the time of this preliminary report, a total of 27 patients were enrolled representing 25 % of children treated for epilepsy. Mean age was 8.2 years. 37% were female. The majority of patient (70.4%) had symptomatic or cryptogenic epilepsies: Brain malformations were the most common etiology (18.5%), followed by congenital infections (7.4%) and stroke (7.4%).

The most common type of epilepsy among 27 children was non-syndromic generalized epilepsy (30.8%), followed by focal epilepsy (26.9%), Lennox-Gaastaut Syndrome (19.2%), and West Syndrome (7.7%).

At enrollment, 25.9% of patients were on AED monotherapy, 51.2% of children received two AEDs and 25.9% received three AEDs. Valproic acid (66.7%), carbamazepine (37%), lamotrigine (33.3%), and phenobarbital (22.2%) were the most commonly prescribed AEDs.

Learning disorders (88.9%), mental retardation (85.2%), and cerebral palsy (40.7%) were the most frequent comorbidities.16% of patients had at least one hospitalization for status epilepticus in the last year.

Conclusions: We describe the clinical and epidemiological characteristics of the first reported series of children with refractory epilepsy in Lima, Peru.

P174

QUALITY OF LIFE OF FILIPINO CHILDREN WITH EPILEPSY AGED 5-12 YEARS OLD USING PEDSQL TM 4.0

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Background: Epilepsy is a chronic neurological condition with numerous social and psychological consequences. In children, epilepsy can cause low self esteem compared to children with other chronic conditions such as asthma or diabetes. The use of quality of life measures in epilepsy is relatively recent compared with that of other chronic conditions particularly within the pediatric population

Objectives: This study aims to assess the health related quality of life among children with epilepsy aged 5-12 years at Philippine Children's Medical Center.

Methods: This is a cross sectional study. Health related quality of life was measured using PedsQL generic score questionnaire.

Statistical Analysis: Mean scores per domain and overall mean scores were obtained and compared. T- test, Chi square and logistic regression was used for the analysis of data and association and determination of predictors for poor quality of life.

Results: A total of 117 children with epilepsy aged 5-12 years old and their caregiver were evaluated. Total overall mean PedsQL was 70.44 while for parents was 67.66. Physical functioning has the highest score while school functioning has the lowest mean score. The means scores are comparable with that of patients with chronic Illness.

Conclusion: Almost 50 % of children with epilepsy had poor quality of life. There was good correlation between child self report and parent proxy report. Predictors to a poor quality of life were male sex, on Phenobarbital and uncontrolled seizures.

P175

SIMULTANEOUS ACTH AND ANTIVIRAL THERAPY IN WEST SYNDROME

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Objectives: Use of adrenocorticotropin hormone (ACTH) is the treatment of choice in West syndrome. However, in infantile spasms caused by human cytomegalovirus (HCMV) or herpesvirus type -1 (HSV1) ACTH therapy could be dangerous. The results of simultaneous treatment with ganciclovir (GCV) and acyclovir (ACV) in West syndrome are presented.

Material and Methods: 26 infants (12 male, y 14 female) were simultaneously treated with intramuscular steroid and intravenous antiviral regimen. Thereafter therapy with oral antiviral drugs was continued. Cerebrospinal fluid examinations were performed for DNA HCMV and DNA HSV 1.

Results: The most early onset of the combined therapy was 4 months. 6 infants have confirmed neuroinfection (5 with CMV and 1 HSV1). 15 were treated with antiviral drugs before simultaneous regimen. In 6 infants combined ACTH and GCV therapy started without previous antiviral treatment. The mean duration of the ACTH and GCV, ACV intravenous regimen was 4 weeks. All infants received 2 or 3 antiepileptic drugs. In 10 patients (one after HSV 1 encephalitis) cessation of seizures was achieved. In 16 infants reduction of spasms frequency was higher than 50%. There was no significant differences with other West syndrome cases treated with ACTH (p>0, 05). Only one patient required ketogenic diet and treatment with vagus nerve stimulation. Despite the active HCMV, HSV1 infection none of the patients had side effects of the ACTH and antiviral regimen.

Conclusion: Simultaneous ACTH and antiviral therapy can be used even in early drug-resistant epilepsy caused by cytomegalovirus and herpersvirus type 1 neuroinfection.

P176

LACK OF EFFICACY OF MONOTHERAPY IN EPILEPSY OF THE 1ST YEAR OF LIFE- POSSIBLE CORRELATION WITH ETIOLOGY?

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Objective: During the 1st year of life epilepsy is most frequent in childhood. The epileptic seizures in very young infants may be severe and drug resistant. The aim of the study is to estimate epilepsy treatment results in epilepsy started during the 1st year of life.

Material and methods: 134 infants (60 female, 84 male, 4 premature) treated in 2010-2013. All infants have video-electroencephalography, magnetic resonance imaging, metabolic examinations and some of them also genetic tests.

Results: Structural-metabolic cases are established in 39(29.1%) infants with tuberous sclerosis complex, congenital cytomegalovirus, toxoplasmosis, hypoxic-ischemic encephalopathy, stroke, brain

malformations and tumor, isobutyric acidosis. Chromosomopties were stated in 4(2.98%) and Dravet syndrome in 2(1.49%) patients. The diagnosis of the West syndrome was established in 12(8.9 %) patients. Only in 2 infants benign myoclonic epilepsy in infancy was diagnosed. Antiepileptic drug (AED) monotherapy (most frequently with valproic acid) was effective only in 18 (13.4%) patients. 3 infants were seizure-free on vigabatrin and 2 on levetiracetam monotherapy. In 83 (61.9%) patients treatment with 2 antiepileptic drug was effective, but 25(18.6%) required adrenocorticotropin regimen. Only in 3 infants 3 AED were used. The antiepileptic drug monotherapy was significantly less frequent ($p<0.05$).

Conclusions: 1. Antiepileptic monotherapy is very rare effective in epilepsy started during the first year of life. 2. In early onset epileptic seizures etiological factor is frequently unknown.

P177

EPISTOP – INTERNATIONAL, LONG-TERM, PROSPECTIVE STUDY EVALUATING CLINICAL AND MOLECULAR BIOMARKERS OF EPILEPTOGENESIS IN A GENETIC MODEL OF EPILEPSY – TUBEROUS SCLEROSIS COMPLEX

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Epileptogenesis is barely studied in humans, as patients usually present AFTER the seizure onset. EPISTOP is the first prospective study of epileptogenesis in humans, beginning BEFORE seizures and continuing through age 2+ years, permitting detailed analysis of the onset, drug-resistance, and comorbidities of epilepsy in a homogenous group of patients with prenatal or early infantile diagnosis of Tuberous Sclerosis Complex (TSC). Project is funded by European Union within 7th Frame Programme.

AIM: The aim of EPISTOP is to examine the biomarkers of epilepsy and to identify possible new therapeutic targets to block or otherwise modify epileptogenesis in humans.

METHODS: Biomarker analysis will be performed by a multidisciplinary, comprehensive approach which consists of: 1/ prospective study of epilepsy development in infants with TSC, including analysis of clinical, neuroimaging, and molecular, blood-derived biomarkers at predefined time points; 2/ prospective study of blood-based biomarkers in infants with TSC treated with antiepileptic drugs prior to seizure onset in comparison to children treated only after clinical seizures appearance; 3/ analysis of biomarkers of epileptogenesis and drug-resistant epilepsy in brain specimens obtained from TSC patients who have had epilepsy surgery and TSC autopsy cases.

RESULTS: EPISTOP should permit to confirm or discover role of genes and proteins involved in the epileptogenesis. The mechanisms of drug-resistancy should be also elucidated.

CONCLUSION: EPISTOP is the first project aimed to assess the epileptogenesis in humans.

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PREDICTORS OF POST-STROKE SEIZURES IN POLISH PAEDIATRIC PATIENTS

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Background: Ischaemic stroke in children, a rare disease occurring in about 3 per 100 000 children per year, is related to various neurological complications, most commonly: motor and speech impairment and intellectual regression. Additionally, the incidence of post-stroke seizures and epilepsy in paediatric patients is quite high. Previous data demonstrated that the incidence of seizures during 24 hours after ischemic stroke may be almost 18 times higher in children than in adults. The aim of the present study was to analyse predictors of post-stroke seizures in Polish paediatric patients.

Methods: The study group comprised 78 paediatric patients suffering from ischemic stroke and recruited in the Department of Neuropediatrics in Katowice (Poland). Among them, three subgroups were separated: patients with early seizures (n=13; mean age: 7.6±5.3), patients with late remote seizures (n=7; mean age: 2.7±2.3), and seizure-free patients (n=58; mean age: 8.9±5.4). Data were analysed using Statistica 9.0.

Results: The total anterior circulation infarct (TACI) subtype was the most common in patients with late seizures compared to seizure-free cases (71% vs 26%, p=0.014, OR=7.17). We also found higher prevalence of focal cerebral arteriopathy (FCA) among patients with late seizures than in patients without seizures (100% vs 51%, p=0.015). Patients with late remote seizures were younger than patients with no seizures. Analysing data with multivariate Cox analysis we found that age at time of stroke, FCA and number of infarct foci are predictors of post-stroke seizures (p=0.013,p=0.014,p<0.001,respectively).

Conclusions: Younger age, FCA as well as number of infarct foci are strong predictors of seizures occurring after childhood ischemic stroke in Polish patients.

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EPILEPSY IN TUBEROUS SCLEROSIS COMPLEX.

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Introduction: Tuberous Sclerosis Complex (TSC) is a major genetic cause of epilepsy. The aim of this work was to characterize epilepsy in a large cohort of TSC patients.

Material and methods: The clinical data of 428 patients with definite TSC were analysed. Eighty patients were followed prospectively from early infancy, before the onset of epilepsy.

Results: Selected results are presented. Epilepsy was present in 366 (85%) patients. Mean follow-up in the prospective group was 4.5 year (range: 3 months - 13 years) and in this group 73% of patients had epilepsy. Drug resistant seizures were observed in 61% in the whole cohort of epileptic patients and in 42% of epileptic patients in the prospective group. Infantile spasms were present in 38% of patients in whole cohort and in 17% of patients in the prospective group. In 47 patients, EEG data before the onset of epilepsy were available. In 223 patients mutational analysis was performed and revealed TSC1 mutation 22%, TSC2 mutation in 69%, and no mutation in 8.5% of patients. Epilepsy was more severe and earlier in patients with TSC2 mutation.

Vigabatrin was effective in 53.3% of cases. In 12 patients with drug resistant seizures everolimus treatment for SEGA was introduced and in 6 (50%) resulted in significant improvement of epilepsy control.

Conclusions: Early diagnosis and treatment improves outcome of epilepsy in TSC. Vigabatrin was effective as a first line drug for focal seizures and infantile spasms in TSC. mTOR inhibitor was effective in some patients with drug-resistant epilepsy.

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EPILEPTIC ENCEPHALOPATHY WITH STXBP1 MUTATIONS - EXPANDING THE ELETROCLINICAL PHENOTYPE

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Introduction: STXBP1 mutations are associated with early infancy epileptic encephalopathy and a characteristic phenotype has been described. We present two new patients with STXBP1 epileptic encephalopathy with different clinical phenotypes.

Case Description:

Patient 1

A 12 years-old male presented at day 3 of life with focal seizures and spasms, evolving to a West syndrome treated with vigabatrin. After a period of 6 years without seizures he started to have refractory epilepsy. The patient evolved with severe intellectual disability and motor delay. Currently he has an ataxic gait, no speech and he presents several stereotypies: hands and head stereotypies, bruxism and hyperventilation alternating with apnoeic spells. He had a **de novo** STXBP1 mutation.

Patient 2

A 7 years-old female began focal seizures with automatisms on the first days of life, without burst suppression. At 1 month she presented epileptic spasms with interictal EEG showing anterolateral paroxysmal activity. She stayed seizures-free during 5 years on monotherapy. The patient acquired independent gait at 3 years-old and presently she combines two words. The microarray showed a deletion on long arm of chromosome 9 involving STXBP1 gene.

Discussion: The first patient illustrates the broad phenotypic spectrum of STXBP1 mutations and the need to consider this diagnosis in patients with a Rett like phenotype. The second patient has a milder phenotype than those previously described. She didn't have Ohtahara or West syndromes and evolved to an easily controlled epilepsy and moderate intellectual disability. These case reports introduce diversity on STXBP1 mutations epileptic encephalopathy phenotype.

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INVESTIGATION OF CALCIUM CONTENT AMONG CHILDREN WITH EPILEPSY

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104 children with epilepsy (56 boys and 48 girls) and 28 children of control group (14 boys and 14 girls) were included in the study. Age 5 month-17 years. Statistical analysis of calcium content in boys and girls of control group didn't reveal any difference. Boys with epilepsy were divided into 2 groups: before 10 years old (1 group) and 10-17 years old (2 group), in which calcium content in hairs significantly (P=0,90) differ. Confidence interval (P=0,90) of calcium content in 1 group was (609.9, 1219.8)ug/g and in group 2 was (507.3, 763.8)ug/g, of girls with epilepsy (643.15, 833.15)ug/g, of control group: boys (673.55, 1226.45)ug/g, girls (752.4, 1054.5)ug/g. Thus, the main value of calcium content in children with epilepsy was higher, then in control group. Calcium content in hairs of boys with epilepsy of group 1 was significantly higher, then in group 2, confidence interval of differences of mean values was (28.5, 530.1) ug/g. The study of significance of differences in calcium content among boys with epilepsy and control group revealed that in group 2 with epilepsy calcium content was significantly (P=0,95) low, then in control group and confidence interval was (-58.9, -570)ug/g. Significant difference (P=0,90) in calcium content was revealed among girls with epilepsy and control group - (-6.65, -323.95)ug/g. Significant difference in calcium content among boys and girls with epilepsy not found. **Conclusions:** Calcium content in hairs of boys upper age and girls with epilepsy significantly lower (P=0,95) in comparison to control group. This study underlines the role of epilepsy in calcium metabolism and need of antiepileptic therapy choice and correction based on identified dates.

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PARENT'S KNOWLEDGE AND ATTITUDES TOWARD CHILDREN WITH EPILEPSY

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Background: Parent's attitudes toward children with epilepsy are influenced by the degree of their knowledge. Misinformation and misconceptions should be identified and corrected for optimal care and management. Our objectives were to study the parent's knowledge and attitudes and identify contributing factors to negative attitudes.

Methods: Consecutive parents were included prospectively from 15 February until 15 August 2012 through the paediatric neurology out-patient clinic of King Abdul aziz University hospital, Jeddah, Kingdom of Saudi Arabia. A structured 40-item questionnaire was designed to examine their demographics, knowledge and attitudes toward children with epilepsy.

Results: A total of 117 parents were interviewed, 57% were mothers. The ages of their epileptic child ranged from 1-16 years (median 6.6), mostly (65%) boys. Although most parents (70%) felt informed about epilepsy and recognized various treatment modalities, many believed that epilepsy is a mental disorder (48%),correlates with evil (44%), and affects the child's intelligence (38%). Up to53% admitted that they treat their epileptic child differently and avoid upsetting or punishing him/her. This behaviour was less likely if they achieved college or university education (p=0.01). Some parents (29%) admitted to using non-medical treatments, usually traditional herbs and religious practices. Those parents were more likely to believe that epilepsy is a mental disease (p=0.002) or correlates with evil (p=0.015).

Conclusions: The level of knowledge and understanding about epilepsy among parents of epileptic children needs improvement. Many parents have significant misconceptions, negative attitudes, and poor parenting practices. Increased awareness and educational programs are needed to help improve the quality of life of these patients and their families.

P183

PUBLIC AWARENESS AND ATTITUDES TOWARDS EPILEPSY

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Background: There is a considerable misconception about epilepsy in our community and it is commonly linked to evils possession. The study was conducted to obtain the perception about epilepsy in the capital city, Riyadh.

Methods: Using a questionnaire survey in Arabic (native language) that contains 10 items regarding epilepsy, we interviewed, face-to-face, a sample of 749 persons, divert in (age, sex and educational backgrounds), selected randomly in public places.

Results: Most respondents (77.4%) mentioned that they had a prior knowledge about epilepsy. Although 52% believed that epilepsy is an organic disease, 15% of individuals linked devils to epilepsy. This correlated with their educational level as those with high education were more likely to link epilepsy organic causes (p= 0.008). Many respondents (37%) preferred spiritual and religious rituals as a treatment either solely (20%) or concomitant with medications (17%). Although most respondents (61%) would accept an epileptic patient for a job, 71% mentioned that they would refuse marrying someone with epilepsy.

Conclusions: Although the majority of the public know about epilepsy, many still link it with evil spirit possession. Religious healers were chosen either alone or with medical treatment as a preferred method for treatment by considerable number of them. More education and epilepsy campaigns are needed in our community.

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PRIMARY SCHOOL TEACHER'S KNOWLEDGE AND ATTITUDES TOWARD CHILDREN WITH EPILEPSY

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Background: Primary school teacher's knowledge and attitudes toward epilepsy can have significant impact on the performance and psycho-social development of the epileptic child. Our objectives were to study teacher's knowledge and attitudesand identify areas in which further teacher training and education arerequired.

Methods:A stratified random sample surveyinvolving a group of primary school teachers in Jeddah, Saudi Arabia included private/publicschools designated for male and female students. A structured 37-itemquestionnaire was used to examine their demographics, knowledge, attitudes, andexperience with epilepsy.

Results: Six hundred and twenty primary school teachers working in public (58%)or private (42%) schools were included with ages ranging between 21-59 years(mean 36). Most teachers (79%) were of Saudi Arabian nationality and 66% had acollege or university degree. Their years of experience ranged from 1 to 35(mean 13.5). Only 17% of the teachers felt very well informed about epilepsy. Teacherswith higher education were more likely to have goody knowledge (p=0.009). Teachers of Saudi nationality were also more likely to report good knowledge,independent of their educational level (p=0.013). Overall, teachers with goodknowledge were less likely to have negative attitudes

including minding to havean epileptic child in their class (p=0.028) or thinking that they should beplaced in a special classroom (p=0.029).

Conclusions: Primaryschool teacher's knowledge needs improvements.Their attitudes correlated highly with their knowledge. Educational campaigns about epilepsy are needed to develop a wellinformed and tolerant community.

P185

THE ROLE OF MELATONIN IN THE EFFECTIVE ATTAINMENT OF ELECTROENCEPHALOGRAMS IN CHILDREN IN A SUB-SAHARAN AFRICAN SETTING

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Introduction: The paucity of access to electroencephalograms (EEGs) in sub-Saharan Africa results in a high patient load attending the few centres with neurophysiology units. Sleep state for EEGs performed on children improves yield and reduces artefact. Melatonin induces "natural sleep" without the risk of airway compromise. This study aims to evaluate the effectiveness of oral melatonin in attainment of useful electroencephalogram in South African children.

Method: Consecutive children booked for routine EEG that were either uncooperative or referred for sleep EEG received oral melatonin (3mg < 15kg; 6mg > 15kg) (September-December 2013).

Results: 58 children were recruited, 30 (51.7%) male, median age 4.3 years (range 0.4 – 12.11 years). Fifty-four (93.1%) children successfully slept for the study, whilst three (5.2%) were drowsy but cooperative. EEG could not be performed in one child who remained awake and uncooperative. The quality of the EEG (background activity and detection of abnormal findings) did not differ when compared to children in who slept spontaneously. Fifteen (26%) of the 57 successful EEG studies detected abnormalities. There were no adverse events, and no child needed their study deferred due to intercurrent illnesses. Children showed no signs of post sedation irritability or persistent drowsiness. They were awoken and were immediately able to go home.

Conclusion: Melatonin is effective and safe in inducing sleep for EEG recording in our setting.

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DOOSE SYNDROME: REVIEW AND EVOLUTION-RELATED FACTORS IN 20 CASES

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INTRODUCTION: Known as Doose Syndrome (DS), myoclonic-astatic epilepsy is a childhood-onset encephalopathic epilepsy characterized by several types of seizures (mostly generalized, myoclonic and atonic) and producing cognitive retardation and ataxia in a previously normal child. Our aim is to describe several clinical aspects in children with DS and look for factors related to a worse evolution.

MATERIAL AND METHODS: We reviewed several clinical aspects of 20 children diagnosed of DS in our Department and studied their relationship with final seizure control and the motor and cognitive development achieved.

RESULTS: We found a statistically significant relationship between final cognitive development (CD) and types of seizure at onset ($p<0.045$), epileptic discharges in EEG ($p<0.059$) and types of seizures developed ($p<0.063$). The same was associated with motor development ($p<0.049$, $p<0.007$ and $p<0.015$, respectively). Epilepsy outcome was related to: types of seizures developed ($p<0.041$), epileptic discharges in EEG ($p<0.002$) and MRI ($p<0.026$).A normal CD (45% of patients), absence of ataxia (60%) and seizure-free status (65%) were related to the absence of frontal discharges in EEG studies and not presenting absences nor focal frontal seizures as the disease develops. In addition, lack of myoclonic seizures at onset was associated with better cognitive and motor development, and a normal MRI (without atrophy) was related to a better seizure control.

CONCLUSIONS: Some DS patients have a bad evolution (mental retardation, ataxia, refractory epilepsy). We found in our patients some factors related to this that could help us identifying them early and improving their management..

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ATOMOXETINE TREATMENT IN PATIENTS WITH ATTENTION DEFICIT HYPERACTIVITY DISORDER AND EPILEPSY

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Introduction: The attention deficit hyperactivity disorder (ADHD) is a chronic neurobiological disorder. There is strong evidence of a higher prevalence of ADHD in children suffering from epilepsy. There are very few studies of pharmacological treatments in this patients. One of the current pharmacological options for this disorder is atomoxetine (ATX).

Objective: To describe the response, epilepsy behavior and the electroencephalogram (EEG) after treatment with ATX, also its tolerability.

Patients and methods: A sample of 22 patients under 18 years old, diagnosed of ADHD and epilepsy, and treated with ATX was reviewed. In all cases clinical information was collected before and after treatment with ATX. Data reviewed came from journal of seizures, parents, teachers and psychologists reports, DSM-IV questionnaire, Conners test, EDAH test and CBCL questionnaire.

Results: Clinical improvement was observed in 59% of patients, 4 worsened (patients previously diagnosed of secondary focal epilepsy and epilepsy with continuous peak-wave during sleep), and in 5 cases there was no change. Regarding the number of seizures and EEGs evolutionary control: no change in 20 patients and in only 2 worse seizures (unchanged in the EEG). Main side effects were: drowsiness, anorexia and headache. In 5 patients treatment was withdrawn (4 cases due to irritability and 1 supraventricular tachycardia case).

Conclusions: ATX is an efficient and safe treatment in the clinical control caused by ADHD showing no impact in the epilepsy behavior or EEG

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STATUS EPILEPTICUS IN INFANTS AND CHILDREN

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Introduction: Status epilepticus is a medical emergency associated with substantial morbidity and mortality. The outcome is directly related to the underlying etiology. The aim of this study is to analyze the etiology of status epilepticus in infants and children.

Methods: From July 2000 to Oct 2013, 311 patients exhibiting with status epilepticus were admitted to the tertiary hospital. The definition of status epilepticus is seizure activity lasting longer than 30 minutes or unconsciousness between two seizure episodes.

Results: Variable etiologies could be identified. 42 patients (13.5%) exhibited infectious CNS diseases. 32 patients (10.3%)exhibited prolonged febrile seizures. 27 patients (8.7%)were fever-induced refractory epileptic encephalopathy. 89 patients (28.6%) had cryptogenic epilepsy exhibited status epilepticus, with poor drug compliance, aggravation by infectious disease, or disease process. 51 patients (16.4%) were categorized metabolic diseases related status epilepticus, including mitochondrial diseases, lysosomal storage diseases, lipid storage disease, or Reye syndrome. 41 patients (13.2%) had underlying CNS diseases presenting with status epilepticus. 16 patients (5.1%) had acute brain insults-related status epilepticus, including intracranial hemorrhage, cerebrovascular infarction, or hypoxic-ischemic encephalopathy. Seven patients had developmental defect, three patients had chromosomal disorders, and three had drug-related status epilepticus.

Conclusions: Multifactorial etiologies of status epilepticus in childhood are noted. Individual treatment strategies for status epilepticus must be initiated after initial general principle of medical treatment.

P193

DISPERSION DURATIONS OF P-WAVE AND QT INTERVAL IN CHILDREN TREATED WITH KETOGENIC DIET

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Background: Limited data are available for the effects of ketogenic diet on dispersion duration of P-wave and QT interval measures in

children. We aimed to search for the changes in these measures with serial electrocardiograms in patients treated with ketogenic diet.

Methods: Twenty-five drug resistant epileptic patients treated with ketogenic diet were enrolled in this study. Electrocardiography was performed in all patients before the beginning and at the sixth month of ketogenic diet. Heart rate, maximum and minimum P-wave duration, P-wave dispersion, maximum and minimum corrected QT interval and QT dispersion were manually measured from the 12-lead surface electrocardiogram.

Results: Minimum and maximum corrected QT and QT dispersion measurements showed non-significant increase at month 6 when compared to baseline values. Other previously mentioned electrocardiogram parameters showed no significant changes either.

Conclusions: A 6-month duration ketogenic diet has no significant effect on electrocardiogram parameters in children. Further studies with larger samples and longer duration of follow-up are needed to clarify the effects of ketogenic diet on P-wave dispersion, corrected QT and QT dispersion.

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MEDITERRANEAN STYLE KETOGENIC DIET: EFFICACY AND SAFETY

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We report the efficacy of the Mediterranean Style Ketogenic diet (KD) in refractory epilepsies focusing on outcomes with regard to epilepsy syndromes and etiology in children with refractory epilepsy. 76 consecutive children were retrospectively enrolled from 2012 to 2013

Method: The Mediterranean Style Ketogenic diet was initiated on an outpatient basis with dietary ratios ranging from 2:1 to 4:1 fat to carbohydrate and protein. The hospital records of the patients given KD due to resistant epilepsy in the pediatric neurology department of our hospital between June 2012 and March 2013 were examined retrospectively. All data recorded as age, sex, diagnosis, seizure type, epilepsy syndrome, associated Autism, physical activity, number of antiepileptic drugs(AED), previous ACTH treatment, period of KD therapy, KD rate applied, Ig G, Ig A, Ig M ,Selenium,Carnitine, fT3,fT4,TSH, Total cholesterol, Total cholesterol levels at least 1 week before KD and 1, 3,6,9,12 months after the therapy started. Moreover, in order to determine the side effects of the treatment. We follow up the efficacy of the diet with blood ketone levels.

Results: At the first month of KD %38.4 of patients seizure free and %38.4 0f patients have %50-99 seizure reduction. Blood ketone level at which patients became seizure-free was found mean 4.45±0.70. We found immobility ($p<0.001$), previous ACTH treatment ($p<0.001$), constipation ($p<0.001$) have negative effect on ketogenic diet. Side effects (constipation%12.5, Hyperlipidemia %35, renal stones %0) were found lower than literature.

Conclusion: Mediterranean Style KD could be a new challenge more than classical KD

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THREE-YEAR FOLLOW-UP ON THE INTAVENOUS IMMUNOGLOBULIN THERAPY IN LANDAU-KLEFFNER SYNDROME

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Landau-Kleffner Syndrome (LKS), is an epileptic disorder characterized by acquired aphasia and auditory agnosia, focal/multifocal spikes and spikes-waves over the temporal region. In 70-80% of the cases epileptic seizures may occur. There are no controlled clinical trials that have investigated treatment in LKS. We describe three-year follow-up of four cases of LKS in 4 to 7 years old children, who improved after a 400 mg/kg /day dose of intavenous immunoglobulin (IVIG) for five consecutive days, after various antiepileptic therapy had failed. In conclusion, IVIG therapy should be considered in cases of LKS who did not respond to traditional antiepileptic treatment.

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THE EFFECT OF KETOGENIC DIET ON THYROID FUNCTION IN CHILDREN WITH REFRACTORY EPILEPSY

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Background: The ketogenic diet remains a valuable therapeutic option for patients with intractable epilepsy. However no data are available for the influence of ketogenic diet on thyroid function in children.

Purpose: The aim of this study was to investigate the effects of ketogenic diet on thyroid functions.

Methods: Between 2011 and 2013, a total of 31 children who received a ketogenic diet for a 6-month-period were enrolled in the study. Serum free thyroxine (fT4) and thyroid-stimulating hormone (TSH) levels were measured before and at first, third and sixth months of therapy. The data were compared to those of a matched group of 45 children with newly onset epilepsy from our previous study.

Results: While average serum fT4 concentrations at baseline were significantly lower in patients receiving ketogenic diet compared to controls ($p = 0.012$), TSH concentrations were similar between the 2 groups. Serum fT4 levels increased significantly at month 3 and month 6 when compared to month 1 measurements ($p = 0.022$ and $p = 0.016$, respectively). Serum TSH levels nonsignificantly increased in patients at the first month of ketogenic diet as compared to baseline levels ($p = 0.754$). A significantly reduced serum fT4 concentrations and increased TSH concentrations were found in the controls at month-6 ($p = 0.026$, $p = <0.001$, respectively).

Conclusion: The ketogenic diet has no deleterious effect on thyroid function in children with refractory epilepsy.

P197

NODDING SYNDROME PATIENTS IMPROVE WITH SYMPTOMATIC TREATMENT

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Objectives: Nodding syndrome (NS) is a poorly understood neurologic disorder in Eastern Africa. In Uganda, a symptomatic treatment intervention of sodium valproate for seizures, management of behaviour and emotional difficulties, nutritional and physical rehabilitation was introduced in 2012. We examined the outcomes of this intervention after a minimum 12 months and compared these outcomes to patients with other convulsive epilepsies (OCE).

Methods: The study was conducted from July–September 2013. Clinicians completed case record forms from a review of records, inquiry from carers, and on physical exam. The primary outcome measure was the proportion achieving seizure freedom (≥1 month without seizures). Secondary outcome measures included reduction in seizure burden, resolution of behaviour and emotional difficulties, performance of activities of daily living and return to school.

Results: We assessed 484 patients with NS and 476 with OCE. The intervention resulted in marked improvements in symptoms and function; 121/484(24.7%) of patients with NS became seizure free and there was >70% reduction in seizures; behaviour and emotional difficulties resolved in 194/327(59.3%); 193/484(39.9%) were attending school; 80% had achieved independence in self care and 75% could assist in home care activities. These improvements were however less than that in patients with OCE in who 243/476(51.1%) were seizure free and there was 86% reduction in seizures.

Conclusions: Ugandan patients with NS have improved with symptomatic treatment suggesting that NS may not necessarily be a progressive neurodegenerative disorder. Epileptic seizures may be a major contributor to the cognitive decline in NS.

P198

VISUALLY INDUCED EPILEPSIES

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Background/objectives: The aim of the study was to define the spectrum of the epileptic syndromes and epilepsies that can be associated with fixation-off sensitivity (FOS) and delineate the electro-clinical types of FOS.

Design and methods: Clinical and video EEG data of all our patients with FOS over the last 6 years were reviewed using FOS technique described by Panayiotopoulos.

Results: The results show that from January 2005 to December 2012, 14 of about 1,990 patients had had one or more video-EEGs with FOS (0.70%). From the 14 patients with full clinical and EEG data available, 8 had various epilepsies that included: symptomatic or probably symptomatic focal (8), cryptogenic generalized (4), and two had no seizures. Four patients (28%) were photosensitive of which a twin with Jeavons syndrome and one was scotosensitive. FOS EEG abnormalities were occipital in 6 patients, and generalized in two.

Two showed atypical forms. One boy of normal intelligence showed abnormal behavior associated with disorientation and confusion and postictal amnesia. His video/EEG evaluation unexpectedly documented the presence of FOS. Another patient was diagnosed with atypical benign partial epilepsy, and his repeated video/EEG recordings showed FOS. His sister was diagnosed with epileptic encephalopathy with continuous spike and wave complexes. Four patients were diagnosed as childhood absence epilepsy.

Conclusions: Irrespective of classification, routine video-EEG monitoring for documenting FOS suggested by Panayiotopoulos should be offered to selected patients with epilepsy. Unusual and rare cases within the spectrum of benign childhood seizure susceptibility syndrome can explain the atypical cases.

P199

THE SPECTRUM OF BENIGN NEONATAL AND INFANTILE SEIZURES

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Background/objectives: To highlight the characteristic features of benign neonatal and infantile seizures and demonstrate the benign nature of the syndromes. These syndromes include many new entities. The aim of this review is to describe these entities, discuss their nosological aspects and demonstrate the characteristic electroclinical features.

Design and methods: A case series of 310 patients with epileptic seizures who fulfilled the criteria for benign neonatal and infantile seizures were followed over a period of 4 years and 7 months which included patients with benign neonatal seizures, benign infantile seizures and benign infantile seizures with mild gastroenteritis between the period from February 2008 to August 2013. Serial video-EEG, MRI/CT brain were done in addition to appropriate blood/urine tests when indicated.

Results: In 2009, Saadeldin et. al. documented for the first time the presence of benign infantile seizures in Saudi Arabian and Arab populations. In 2010, Saadeldin analyzed the electroclinical features of patients with benign infantile seizures with mild gastroenteritis and demonstrated the benign nature of this entity. Recently, in 2013, Saadeldin et. al published for the first time in United Arab Emirates and Gulf region a novel KCNQ2 mutation in a large Emirati family with benign familial neonatal seizures

Conclusions: Increasing the awareness of clinicians regarding the existence of these syndromes and their benign nature in children will limit unnecessary investigations.

P200

SEIZURE AND DEVELOPMENTAL OUTCOME OF INFANTILE (EPILEPTIC) SPASMS STARTING AS FE IN EARLY INFANCY

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Introduction: This study sought to compare the clinical course, seizure and developmental outcome of infantile spasms(IS) that start off as focal epilepsy(FE) with those who present with IS from the outset.

Method: Retrospective observational study.

Results: 26 (37.1%) out of 70 patients with IS presented with FE before evolving into IS and 6 (8.5%) with multifocal epilepsy. Majority from FE group had concordant electroencephalographic (EEG) changes. 20 (77%) out of 26 patients in FE group had an abnormal MRI compared to 22 out of 38 (58%) in IS group. Median age of diagnosis of IS was 24 weeks in FE group compared to 20 weeks of IS group. In 38 patients with IS as initial presentation, 37 showed hypsarrythmia and 1 showed modified hypsarrythmia. Patients with structural-metabolic abnormalities were more common in FE group than IS group. Infants with FE were more resistant to treatment for IS and had worse developmental outcome at the last follow-up.

Conclusion: Our study looks at potential predictors in infants who initially present with FE before evolving into IS. This, in turn, could influence their early treatment to prevent epileptic encephalopathy. We have identified that patients with structural-metabolic aetiology presenting with FE in infancy are at an increased risk of developing IS, are more resistant to usual treatments and have worse developmental outcomes. This begs the question of treating this group of patients earlier with steroids before the emergence of IS to avoid the bad outcome(s) associated with epileptic encephalopathy.

P201

CHRONIC PANCREATITIS IN CHILD TREATED FOR REFRACTORY EPILEPSY WITH ZONISAMIDE

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Introduction: Zonisamide is a broad spectrum anti-epilepsy drug (AED), licensed for treatment of adults and children over the age of 6 years with focal epilepsy. We report the occurrence of pancreatitis in child treated with Zonisamide and believe it may have a causal relation.

Case: We present a 5-year boy suffering from refractory generalised chronic epilepsy, characterised by generalised tonic, tonic clonic seizures and absence seizures compatible with symptomatic Lennox Gastaut syndrome. He had acquired microcephaly, severe 4-limb spastic cerebral palsy, cerebral visual impairment and severe cognitive impairment. He was fed by gastrostomy. He had suffered severe perinatal hypoxic ischaemic brain injury at term gestation. Brain MRI after age 2 years showed extensive multicystic encephalomalacia.

He had been treated with multiple antiepilepsy drugs over the years. He was commenced on Zonisamide. Co-medications included Phenytoin, Phenobarbitone, Clobazam. In the months after commencing Zonisamide he was frequently troubled by severe irritability and excessive crying and his parents felt he was suffering from severe abdominal pains. 8 months after commencing Zonisamide, a CT abdomen showed him to have a pseudo pancreatic cyst. His serum Amylase levels were normal.

Discussion: Pancreatitis has been listed as a rare but serious adverse effect of Zonisamide therapy. This case report illustrates this potential risk.

P202

HEAD-TO-HEAD COMPARISON OF KETOGENIC DIET AND VAGUS NERVE STIMULATION IN PAEDIATRIC POPULATION WITH PHARAMACORESISTANTEPILEPSIES

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Objective: To compare efficacy, response in specific epilepsy syndromes, sustenance of response, effect on quality of life (QOL) and adverse effects between vagus nerve stimulation (VNS) and Ketogenic diet (KD).

Method: Retrospective review of case notes and direct patient interaction to discuss QOL.

Results: Pre- and post intervention data were available on 50 and 30 children for KD and VNS respectively. Follow up data, with reducing numbers, were available for upto 5 years for VNS and 2 years for KD. With VNS, 53% (16) patients reported over 50% reduction in seizures at 1y, 48% (14) at 2y, and 55% (10) at 5y. With KD, 40% (20) reported over 50% reduction at 1y and 38% (19) at 2y. KD was very effective in all patients with myoclonic astatic epilepsy (MAE) whereas VNS seemed more efficacious in unclassified multifocal epilepsies. Discontinuance rate was 16% for VNS due to infection and inefficacy and 46% for KD due to inefficacy and intolerance.

Conclusion: In our cohort, VNS was more effective than KD as a long term treatment for pharmacoresistant epilepsies. However, KD was extremely effective in MAE. Family feedback suggested an improvement in QOL with VNS even without significant seizure reduction, whereas with KD this effect was achieved only through reduction of seizure burden. Our results potentially could help in patient selection as both modalities of treatments pose their own risks and affect QOL of child and family.

P203

KETOGENIC DIET EFFICACY IN THE TREATMENT OF INTRACTABLE EPILEPTIC SPASMS

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Objective: To determine the efficacy of the ketogenic diet (KD) in controlling Epileptic spasms (ES) after failing traditional therapy.

Methods: All Infants with epileptic spasms who were referred for KD treatment at our hospital between 2009 and 2012. All subjects had ES with evidence of hypsarrythmia or severe epileptic encephalopathy on electroencephalogram (EEG) despite appropriate medication treatments. The diet efficacy was assessed through clinic visits, phone communications and/or EEGs every 3 months. Quality of life improvement was charted based on the care giver's perspective.

Results: 20 infants (15 males) were included in the study. The mean age at seizure onset was 4.5 months. Age at KD initiation mean 1.20, SD 0.78. 15 patients had cryptogenic ES. 15 failed to respond to ACTH and/or vigabatrin before going on the KD. The remaining 5 patients tried at least 3 other antiepileptic medications before the diet was started. Three months after starting the diet >50% seizure reduction was achieved in 70%. These results were maintained at 6 and 12 months intervals. At 24 months follow up all patients had >50% seizure reduction. At least 90% seizure reduction was reported in 20% of patients at 3 months, 22% at 6 months and 35% at 12 months. Majority of patients (63%) achieved improvement of their spasms within one month after starting the diet. All care givers reported improvement of the quality of life at 3 months visit. This ratio was 94% at 6 months and 82% at 12 months.

Conclusion: KD is safe and potentially effective treatment for patients with ES; especially those who failed medication therapies.

P204

THE SIMULTANEOUS USE OF KETOGENIC DIET AND VAGUS NERVE STIMULATOR IN PATIENTS WITH PHARMACOLOGICALLY REFRACTORY EPILEPSY

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Rationale: About one third of patients with epilepsy have pharmacologically refractory epilepsy. This subset of patients pose a significant challenge to the epilepsy community, with negative impact on development, co-morbidities, and economic burden. Both the ketogenic diet (KD) and Vagus Nerve Stimulator (VNS) have been well studied and established effective treatments for patients with refractory epilepsy. This study assesses the efficacy of the combined treatment using VNS and KD.

Methods: Retrospective chart review assessing seizure control in patients who are treated with both VNS and KD. Particular attention was given to the seizure control achieved with both treatments to assess if combined therapy provided improved efficacy. Seizure outcome was defined as a 50% or more improvement in seizures from baseline.

Results: A total of 473 patients have VNS implanted and 150 patients are treated with KGD. 13 patients had both VNS and KD treatment. Of these patients, 10 (Group A) had KD started after VNS had been implanted for at least 18 months. And 3 patients (Group B) had VNS implanted after being treated with KGD for at least 6 months. Response from a seizure control perspective was assessed 6 months after the introduction of the second therapy. [No change; < 50% ; 50%-70%; >75%]

Group A : 2 (20%) 1(10%) ; 4(40%) ; 3 (30%)

Group B: 0, 1 (33%) , 2 (67%)

Conclusion: The simultaneous treatment with both VNS, and KD potentially provides synergistic anti-epileptic effect that is cumulative and seems sustainable over a 12 months period after the introduction of both therapies

P205

AUTOIMMUNE ENCEPHALITIS WITH ANTI-VGKC ANTIBODIES MIMICKING HSV ENCEPHALITIS IN A TEENAGE BOY

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Introduction: New onset refractory status epilepticus can have various reasons as etiology. Autoimmune encephalitis/encephalopathy is becoming more and more recognizable. This entity includes anti-neuronal surface protein antibodies, such as anti-VGKC, anti-NMDAr, anti-AMPAr; and intracellular antibodies, such as anti-GAD and onconeural antibodies.

Case Description: A 13-year-old boy presented with confusion accompanied by fever and focal seizures. MRI of brain showed abnormal signals from temporal lobes and CSF showed mild pleocytosis. HSV encephalitis was highly suspected. The patient was treated with acyclovir, but seizures were difficult to be controlled despite multiple anti-seizure medications. Eventually the patient was placed in a pentobarbital-induced coma, which necessitated a high dose to eliminate the seizures. PCRs for HSV twice were negative as well as other viral searching. Repeat MRI of brain did not show any progressive abnormalities and demonstrated stable signals from temporal lobes. Immune-modulating treatment was initiated 10 days after admission. Autoimmune work-up showed positive anti-VGKC antibody. The treatment included IVIG, high dose of steroids and plasmapharesis. Now, 7 weeks after admission, the patient's seizures are still not well-controlled.

Discussion: Anti-VGKC antibodies associated intractable seizures are rare. Encephalitis associated with VGKC antibodies presented with status epilepticus and focal epilepsy can be confused with herpes encephalitis. Early recognition of autoimmune epilepsy is critical. Since most of the cases are non-paraneoplastic, more systematic and aggressive immune-modulating treatment should be initiated as soon as possible in order to improve outcomes

P206

DESIGNING A CLINICAL STUDY OF CLOBAZAM FOR DRAVET SYNDROME USING PHARMACOKINETICS/ PHARMACODYNAMICS MODELING

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Introduction: We are initiating a study of the efficacy and safety of clobazam for pediatric patients ≥1 to ≤16 years of age with Dravet syndrome (DS). Lack of clobazam dosing information for patients <2 years makes this challenging. A published RCT of stiripentol with clobazam and valproate demonstrated the combination's efficacy in DS.1 However, as stiripentol substantially increases clobazam concentrations, clobazam's contribution to efficacy was unclear.1

Methods: We employed PK/PD modeling, using previous clobazam studies,[2-4] to determine the appropriate clobazam dosage for patients 6 months to 2 years of age. Results from the stiripentol with clobazam and valproate study in DS4 were used to design the study.

Results: Maximum clobazam dosage was determined to be 1.5 mg/kg/day for patients 6 months to 2 years of age, similar to dosing for older patients. Based on statistical analysis, we estimated 54 patients would be needed to demonstrate clobazam's efficacy (percentage decrease in seizure rate vs. placebo, 85% power) in a clinical study in DS. This estimate was with the assumption that 50% of stiripentol response in the previous study1 was due to clobazam.

Conclusion: An innovative pharmacometrics modeling approach eliminated the need for a Phase I trial to justify Phase III study dosage and will provide supportive evidence of efficacy for clobazam in DS. This approach may facilitate other drug studies in ultra-orphan indications.

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P207

QUANTIFICATION OF CONNECTIVITY STRENGTH BETWEEN EPILEPTOGENIC CORTEX AND REMOTE FDG PET ABNORMALITIES IN CHILDREN WITH ETL EPILEPSY

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Introduction: Our objective was to quantify the connectivity strength (based on probabilistic DTI) of fiber tracts that originate in the seizure onset region (based on intracranial ECoG) and that project to remote cortical areas characterized by F18-deoxyglucose (FDG) PET abnormalities that are electrophysiologically normal.

Methods: We developed a computational framework that allows quantitative assessment of the spatial relationship between multi-modality neuroimaging data (PET, DTI, ECoG). The framework is based on

the parcellation of the cortical surface in native space using landmark-constrained conformal mapping, which yields finite cortical elements (FCEs) that are homotopic across subjects. The FCEs were used as source/target regions for probabilistic fiber tracking (55 direction), allowing the calculation of a connectivity score (CS). We applied this approach to 7 young children (3–12 yrs) with non-lesional epilepsy and compared the obtained connectivity pattern against 12 age-matched normal children.

Results: CS reproducibility was assessed in the normal group for three major fiber tracts (arcuate, SLF and ILF), yielding a COV of 5.3 + 4.7%. Comparison of the CS between the normal group and the patient group showed either unchanged or decreased CS for major fiber tracts, with the decreases more pronounced on the ipsilateral side (22% vs. 14%). Connectivity strength analysis between the epileptogenic cortex and remote FDG PET abnormalities showed a similar pattern with highly significant decreases in both ipsi- (>50%) and contralateral (~20%) hemisphere.

Conclusion: Our findings indicate that fiber connectivity between the primary focus and remote FDG PET abnormalities is significantly decreased as compared to controls.

P208

CAN EPILEPSY ETIOLOGY PREDICT RESPONSE TO KETOGENIC DIET IN CHILDREN?

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Introduction: The study purpose is to determine the impact of etiology on ketogenic diet (KD) response in children with intractable epilepsy.

Methods: Children initiated on KD January 2009 through December 2012 were identified from the KD diet database at Mayo Clinic Rochester. Charts of those with known etiology (genetic, structural, or both) were reviewed to determine seizure frequency (a) prior to KD onset, (b) 3 months after KD onset and (c) at final follow-up on KD. Significant response was defined as >50% decrease in seizures.

Results: 49 children were identified (62% male). Etiologies included genetic (33%), structural (49%), both (18%). Median age of KD initiation was 30 months (range 3-194 months) and treatment duration was 21.5 months (range 3-120 months). Structural or genetic etiologies responded equally, 46% and 44% significant response, respectively. Only 22% of children with both structural and genetic etiology had significant response. However, 2 of 4 children with tuberous sclerosis complex (TSC) had a significant response, whereas none of those with structural and genetic etiology not due to TSC (n=5) had significant response at final follow-up. The non-TSC structural and genetic etiology patients were less likely to respond when compared to all others (p=0.05).

Conclusions: From etiology alone, it is difficult to predict which children will respond to KD. Therefore, it is recommended that most children with intractable epilepsy be given a trial of KD. However, those with both genetic and structural etiology for epilepsy, especially those without TSC, may be less likely to respond to KD.

P209

AN EASY TO USE DIAGNOSTIC TOOL FOR PEDIATRIC EPILEPSY IN LOW-RESOURCE REGIONS

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Introduction: Epilepsy affects 50 million people worldwide, and is disproportionately prevalent in regions where specialist care and diagnostic tests are scarce. To improve diagnostic accuracy, we created a simple questionnaire with the aim to discriminate between focal versus generalized epilepsy, and guide medication choices.

Methods: Through literature review, key semiology features were identified. These were then validated by retrospective chart review in a Western tertiary care setting to identify 15 key questions. The refined questionnaire was translated into Kiswahili and validated prospectively in Tanzania. The performance of the questionnaire was compared to an electro-clinical diagnostic standard, from comprehensive clinical evaluation by a visiting pediatric neurology provider and locally obtained interictal EEG, read remotely.

Results: Children aged 6 months to 18 years with suspected or active epilepsy were included. On retrospective validation, the overall predictive value of the questionnaire was 85%**.** During prospective validation, two questions were omitted as they translated poorly. The refined questionnaire had a positive predictive value of 78% for diagnosis of focal epilepsy, and 100% for generalized epilepsy.

Discussion: Our questionnaire provides a simple method to improve diagnostic accuracy, with the ultimate goal of guiding medication choices and improving treatment outcomes, with good predictive value in different health care settings. It may help bridge the diagnostic gap in pediatric epilepsy in resource-limited regions, and once validated may be easily implemented in various forms for use by any pediatric healthcare provider.

P210

HUMAN PAPILLOMAVIRUS INFECTION IS NOT CAUSALLY RELATED TO FCD IIB

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Introduction: Focal cortical dysplasia (FCD) is increasingly recognized as a cause of symptomatic epilepsy in children. Recently, it has been proposed that human papillomavirus (HPV) infection during fetal development plays a causal role in the pathogenesis of FCD type IIB, characterized histologically by the presence of dysmorphic neurons and balloon cells. Two groups have shown evidence of HPV-16 (by DNA PCR and immunohistochemistry) in a very high proportion of surgical resection specimens meeting criteria for FCD IIB, but in virtually no brain tissue specimens without FCD (1,2). We sought to examine the correlation between HPV-16 infection and FCD IIB in an independent surgical cohort.

Methods: We queried a database of surgical biopsy material for cases of FCD between 1995 and 2013. Twenty cases met criteria for FCD IIB on neuropathological review, and 6 samples were tested for HPV-16 by DNA PCR. Immunohistochemistry for HPV P16 and BCL2 was performed in a subset of samples.

Results: None of the samples tested (0/6) was positive for HPV by DNA PCR. Immunohistochemistry showed no P16 staining of balloon cells or other neuronal cells, using cervical transformation zone as a positive control. In one sample, P16 labelled rare small cells which appeared to be lymphocytes. BCL2 highlighted a reactive astrocytosis.

Conclusion: These results do not support the hypothesis that HPV-16 infection is causally related to the pathogenesis of FCD IIB.

P211

ASSESSMENT OF THE IMPLEMENTATION OF THE EPILEPSY QUALITY MEASURES IN THE COMMISSION FOR CHILDREN WITH SPECIAL HEALTH CARE NEEDS (CCSHCN)

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Rationale: The purpose of this study was to assess the application of the recently developed 8 epilepsy quality metrics in a primarily rural and Appalachian system of clinics conducted at the Kentucky (CCHCN).

Methods: Retrospective review of clinical charts from the CCSHCN between November 2013 to January 2013. Measures assessed were: seizure type and frequency, epilepsy etiology or syndrome, electroencephalogram (EEG), brain imaging, antiepileptic drug side effects, surgery referral, safety counseling, folic acid supplementation, and counseling of childbearing potential patients.

Results: We identified 81 children with epilepsy; aged 1 to 21 yo (mean age 9.6 years) 46 males and 35 females. The seizure type and frequency were addressed in 98.8%. Epilepsy etiology or syndrome in 96.3%. EEG reviewed or requested in 97.5%. Brain imaging was reviewed or requested in 79%. Antiepileptic drug side effects were addressed in 59.3%. Epilepsy surgery evaluation and referral was 3.7% of the 23 patients with drug resistant epilepsy. Documentation regarding safety counseling was 25% of patients. Of the 19 patients with childbearing potential counseling was documented in 40% and folic acid supplementation was addressed in 33%.

Conclusion: In this study, a sub-set of quality measures had a high rate of compliance. For other measures with lower rates of compliance it is unclear if the measures, as currently formulated, are appropriate for pediatric clinics. While charts routinely included information about child educational status, it is not included in the quality guidelines.

We suggest that pediatric clinics require a revised sub-set of quality guidelines for epilepsy.

P212

DOES ANTIEPILEPTIC DRUG (AED) FAILURE FOR LACK OF EFFICACY AFFECT LONG-TERM PROGNOSIS IN CHILDREN WITH GENERALIZED EPILEPSY?

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AIM: To assess impact of AED failure for lack of efficacy on long-term outcome in children with generalized epilepsy.

METHODS: Children with generalized epilepsy followed for >2 years after diagnosis were identified from a 30-year database of new-onset pediatric epilepsy in Olmsted County, MN. Clinical details abstracted from the medical record included seizure outcome, use of AEDs and number of AEDs failed for lack of efficacy at final follow-up. An AED was defined as failing for lack of efficacy if seizure control was not achieved despite maximum tolerated dose.

RESULTS: Ninety-six children were identified (51% male). Median age at seizure onset was 7.8 years (IQR 3.6, 13.2). Duration of follow-up was 10.0 years (IQR 4.9, 15.4). A defined electroclinical syndrome was identiifed in 67 (70%). At final follow-up, 65 (68%) were seizure-free for >1 year, 35 (54%) of whom were also off AEDs. Seizure freedom was seen in 44/59 (75%) who failed no AED, 10/20 (50%) who failed one AED and 11/17 (65%) who failed two or more AEDs for lack of efficacy (p=0.12). Predictors against seizure freedom included intellectual disability (p<0.001), lack of febrile seizures (p=0.001), abnormal neurological exam (p=0.03) and prior neonatal seizures (p=0.04).

CONCLUSIONS: While there was a trend for children who had never failed an AED for lack of efficacy to have a higher likelihood of seizure freedom, 57% who had failed medication still achieved this outcome. Prognosis was more concerning for those with intellectual disability, neurological exam abnormalities, prior neonatal seizures or lack of febrile seizures.

P213

STROKE IN CHILDHOOD LEAD EPILEPSY. IS IT TRUE?

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Introduction: Etiological factors causing childhood epilepsy have wide variety, including strokes. One of the features of stroke in childhood is manifest of vascular accident as seizures.

Objective: To analyze link between manifest of stroke with seizures and post-stroke epilepsy.

Methods: We examined 150 children from birth up to 14 years with stroke. Conducted MRI, EEG studies.

Results: Among managed patients 60% (90) has hemorrhagic stroke, 40% (60) ischemic. 53% (80) of children has perinatal stroke. In 60% (39) of hemorrhagic stroke, vascular accident began with seizures, epilepsy formed in all this patients. During recovering period in 21.5 % (14). Refractory epilepsy formed in 25.6 % (10), others corrected with medicines. Type of seizures: mainly partial 32.3% (20), with secondary generalization 21% (13) - dominantly tonic convulsions 21% (13), 16.1% (10) myoclonic.When ischemic stroke was analyzed, 34% of patients has convulsions (33) and they are mainly represented by partial seizures 44.1 % (15) and secondary generalized seizures with tonic component 32.5% (11). From the amount of ischemic stroke convulsions in the most acute period convulsions appeared in 45.5% (15), in the late period in 54.5% (18). Later 48.5% (16) were observed as post-stroke epilepsy, which controlled by medicines.

Conclusion: Hemorrhagic stroke characterized with earlier onset of seizures 46% (30), ischemic with late 54.5% (18). Post-stroke epilepsy occurred in 60% (39) with hemorrhagic stroke and in 48.5 % (16) with ischemic.The severity of neurological deficiency, seizure types, epileptic focus localization impact on the overall outcome of stroke.

P214

SOME CLINICAL FEATURES OF MESIAL TEMPORAL LOBE EPILEPSY IN CHILDREN

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Purpose: To analyze the main clinical and neuroimaging findings, postsurgical results in children with mesial temporal lobe epilepsy (MTLE) with and without presence of hippocampal sclerosis (HS).

Material and methods: We analyzed the clinical histories of 30 patients with MTLE and with minimum 12 month of postsurgical follow up.

Results: All patients were divided for 4 groups according to the age (from 7 to 15 y.o.) when MTLE diagnose were made. The mean age of patients with MTLE was 12, 5 y.o. Gender tendencies were higher in female. Only 2 patients presented family history of epilepsy (none of these relatives had MTLE). In 30% of patients we found the pathological background, in 2% - previous encephalitis as a possible trigger cause of MTLE and the other patients -presented mental retardation, as possible sequence of MTLE. In 32% of patients the history of febrile seizures (simple febrile seizures) had confirmed. Among these patients – 80% had positive biopsy result for HS and positive MRI findings compatible with HS. Clinically 62% of patients had aura and in 71% patients the seizure semiology was presented by ipsilateral automatisms with secondary generalization. In 14 patients MTLE findings were positive; in 33% of patients MRI was normal. In 33% of cases the biopsy of HS was confirmed. According the Engel scale 71 % (15) of patients were seizure free.

Conclusions: The history of previous febrile seizures confirmed in 32% of children with MTLE. The surgical treatment was successful in most patients with early presurgical evaluation.

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MUTATIONAL ANALYSIS OF TSC1 AND TSC2 IN KOREAN PATIENTS WITH TUBEROUS SCLEROSIS COMPLEX: GENOTYPE AND EPILEPSY

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Background: Tuberous sclerosis complex (TSC) is an autosomal dominant disorder involving multiple organs and tissues, which is caused by a mutation in either TSC1 or TSC2, tumor suppressor genes. TSC has wide spectrum of clinical manifestations, which might be associated with genetic heterogeneity and incomplete penetrance. It has been reported that various neurocognitive phenotypes are correlated with genotype of TSC. However, to date, there are only a few reports correlating the genotype with epilepsy.

Patients and Methods: We performed mutational analyses on 60 unrelated patients diagnosed as definite TSC. Genetic testing was performed using direct DNA sequencing and/or multiplex ligation-dependent probe amplification. Clinical data of the patients were obtained by retrospective chart review.

Results: We identified pathogenic mutations in 45 patients (75%). There were 10 TSC1 mutations and 35 TSC2 mutations. Six patients had unclassified variants. No mutation was identified in nine patients. TSC1 mutations included 7 frameshift (70%) and 3 nonsense mutations (30%). TSC2 mutations included 9 frameshift (25.7%), 9 nonsense (25.7%), 4 splicing mutations (11.4%), 4 large deletions (11.4%), 6 missense mutations (17.1%) and 3 in-frame deletions (8.6%). All hamartin interaction domain mutations were protein-truncating. Fifty-one patients had epilepsy (85%). Of patients with epilepsy, 14 of 45 developed refractory epilepsy. Epilepsy was significantly associated with cognitive impairments (p=0.03). Eighteen of 51 epileptic patients had a history of infantile spasms (35.3%). No significant difference was observed in the phenotype and outcome of epilepsy among patients with TSC1, TSC2 and no mutations. Unexpectedly epilepsy was frequently involved in the patients with no mutations, although most were focal epilepsy.

Conclusions: We could not find any significant differences among TSC mutation groups. However, the group with no mutations identified had epilepsy, notably focal with higher frequency than expected. Patients with TSC2 mutations are more likely to have infantile spasms than patients with TSC1 mutations

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THE KNOCK IN MOUSE MODEL OF ALTERNATING HEMIPLEGIA OF CHILDHOOD

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Background: Recent research at Duke with collaborators has shown that de novo mutations in ATP1A3 gene cause alternating hemiplegia of childhood (AHC; Heinzen et al, 2012). Missense mutation in ATPA13-D801N is the most frequently seen in AHC patients.

Goals: Create and characterize a mouse model carrying the most common mutation causing AHC in humans.

Methods: ES cell approach followed by homologous recombination, microinjection in blastocysts and breading of chimeric mice with wild type to generate the knock in mice.

Results: A small plasmid containing of our gene of interest, with the desired mutation, was removed from bacterial artificial chromosome and linearized construct introduced into ES cells (129SvEv) by electroporation resulting homologous recombination at the shared sequence in one allele, during which two crossover events replaced the wild type gene with the targeting construct. Using genomic southern blot the desired mutation in the ES cells DNA was confirmed. Heterozygous ES cells were expanded, microinjected into blastocysts harvested from C57BL/6 mice, and implanted into uterus of pseudopregnant females. Male chimeras were mated with C57BL/6 females. PCR genotyping confirmed heterozygote agouti mice which were further mated with Flp mice to remove antibiotic(neo) cassette. The resultant litters were subsequently mated to make a colony for behavioral and neurophysiological studies.

Conclusions: We have developed knock-in mouse model that has the most common mutation affecting humans with AHC.

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INFANTILE SPASMS AND FOCAL SEIZURES: A PREVIOUSLY UNREPORTED PRESENTATION OF A WDR45 MUTATION

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Introduction: WDR45 mutations have recently been reported to cause Neurodegeneration with Brain Iron Accumulation (NBIA). Such patients present with global developmental delay in early childhood followed byextrapyramidal symptoms in early adulthood. Seizures may occur in childhood, and absence, atonic, febrile, and myoclonic seizures. Presentations as seizures in infancy, or as infantile spasms, however, have not been reported.

Case Description: This patient first presented with a prolonged, focal-onset seizure at age 3 months. Phenobarbital was given then weaned off. The patient re-presented within fantile spasms at age 11 months. EEG showed high voltage, slow background with multifocal and generalized spikes and polyspikes. Zonisamide stopped the spasms, but they recurred a few months later together with tonic seizures. Neither clonazepam, nor levetiracetam, or rufinamide helped. Spasms continued and consisted of flexion of the head and extension ofthe extremities with 7-10 clusters per day mostly upon awakening. Atage 4 years she has a developmental age of 9 months, no extrapyramidal signs, ten spasms per day, weekly tonic seizures, and hypsarrhythmia in sleep. Extensive neurometabolic workup and two MRIs are normal with no "eye ofthe tiger" sign. Whole exome sequencing shows a heterozygous deleterious mutation c.400C>T (p.R13X) in WDR45 which is not present in her parents.

Conclusion: We report the first case of WDR45 mutation presenting with focal seizures and infantile spasms during the first year of life. Testing for this mutation should be considered inpatients with infantile spasms of unclear etiology.