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Pediatric Epilepsy Surgery

Clinical cases and posters presented at the 2nd International Epilepsy Colloquium, Lyon, France, May 2009

ABSTRACT

Nearly 15 years after the Bethel-Cleveland 1995 symposium, exclusively dedicated to *Pediatric Epilepsy Syndromes and their surgical treatment*, the development of new concepts and techniques has led to earlier and more frequent use of surgical therapy for children with epilepsy. The spectrum of surgical possibilities for early-onset epilepsy has widened and includes not only lesional focal epilepsies but also MRI-negative epilepsies and more diffuse types. The 2nd International Epilepsy Colloquium, organized by the Institute for children and adolescents with epilepsy IDEE (Lyon, France), in collaboration with the Marburg (Germany) and University of Cleveland (USA) epilepsy centers, covered appropriate diagnosis and medical treatment of epilepsy syndromes, early identification of surgical candidates and appropriate pre-surgical evaluation approaches as means of curing epilepsy whenever possible, and issues related to the preservation of cognitive development and socio-professional integration. Abstracts of clinical cases submitted for discussion and posters presented are published in *Epileptic Disorders*.

Case discussion forum

1. A Case of fronto-parietal epilepsy with surgical failure

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Boy, born April 1990, the second child of uneventful pregnancy and delivery; birth weight 3 700 g; early psychomotor development normal. At 2.5 years of age episodes during which he turned his head and eyes to the left for 5-6 seconds several times a day. Seizures became frequent. EEG showed spike-wave complexes over the temporo-central right region. Brain CT scan (1992) was normal. Nearly all AEDs and a ketogenic diet trial failed. Later on attacks became more prolonged: he stared towards the left side, smiled and felt down on his right side. Brain MRI was reported as showing right mesial temporal sclerosis (1999). He attended regular school with good results but he was obliged to stop because of a high seizure frequency. Following pre-surgical evaluation (2006; Bielefeld, Germany) a right frontal resection was performed. Histology did not show focal cortical dysplasia. Following surgery he was on oxcarbazepine and valproate but seizures persisted. Postoperatively, during bad periods, he had focal and tonic seizures with abduction of the arms and fall, up to 30 a day. Generic lamotrigine was introduced and he was much better after 3 months. MR of the brain (december 2007) showed changes related to the surgical procedure. Currently he presents with 3 secondarily generalized seizures a day. He moves his eyes from the left to the right, then upwards, raises his arms and falls backwards, and then jerks of extremities ensue, about 1 min. His neurologic status shows alterations of fine motor functions; he is left-handed. He receives: oxcarbazepine 2 100 mg/d, sodium valproate 1 500 mg/d and 3 lamotrigine 200 mg/d for a weight of 58 kg.

Questions:
1. Is he a candidate for reoperation?
2. Should he be treated with medication only?

2. Drug-resistant epilepsy in a child with temporal lesion extended to the parieto-occipital junction: indications to grid implantation and definition of the area to be resected

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We present a girl aged 4 years who at 6 months began to present weekly episodes of motionless staring. At age 17 months daily seizures occurred: motionless staring, slow nodding with leg flexion, mouth deviation to the left. MRI: right temporal lesion. At 19 months, an anterior temporal neoplastic lesion was hypothesized, and a right anterior temporal lobectomy was planned. However, due to a subdural

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bleeding, lesionectomy was only partial. Histology: focal cortical dysplasia type IB. The child has been then seizure free for 8 months, after which seizures recurred: a) spasms upon awakening; b) staring with arrest, followed by rightward head version; c) during sleep: respiratory modification, and non patterned limb movements. The ictal discharge was bilateral with right predominance. Neuropsychology demonstrated normal cognitive development. MRI was unchanged since operation. In order to define the relationship between lesional and epileptogenic zone(s), grids were implanted: seizures originated from the lesional area with early parietal involvement. Temporal lobectomy extended to the parieto-occipital junction was performed. The patient has been seizure free since then (9 months follow-up). Histology: type Ib dysplasia and astrocytoma grade III, the latter at the border of previous surgery.

Questions "to be answered":
1. Would it have been possible to perform a "direct" right temporo-occipital resection without invasive recordings?
2. In order to obtain a better surgical outcome, would it be necessary to also remove the area of early seizure propagation?
3. Does the association of dysplasia and high-grade astrocytoma influence seizure outcome?

3. Surgical treatment for Continuous Spike and Wave during Slow Sleep (CSWSS) in a child with infrequent seizures and moderate learning disability?

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This 10-year-old boy had his first seizure at the age of 4 years, consisting of a tonic gaze deviation to the left, followed by secondary generalization. A similar seizure occurred 4 months later. Continuous spike-waves during slow sleep (CSWSS) were then detected on EEG. Valproate and ACTH treatment was started with some improvement. When first seen at our centre at the age of 8 he had experienced similar seizures twice a year and frequent periods of CSWSS in spite of 4 courses of ACTH and lamotrigine therapy. Between the ages of 4 and 8 his learning abilities slowed down. He had moderate learning disability, especially in verbal skills. He is right handed, but dichotic listening demonstrated right-sided hemispheric dominance for language. An interictal video-EEG recording at age 8 showed frequent left fronto-temporo-parietal spike and wave discharges during wakefulness, which became continuous and diffuse during slow sleep. Ictal EEG showed frequent episodes of activity arrest with desynchronization of basal trace. A MRI scan showed left frontal cortical displasia. An interictal FDG-PET scan showed hypermetabolism in the dysplastic cortex. He has been subsequently treated with levetiracetam, topiramate and zonisamide with no improvement of CSWSS. He has seizures once a year, and the episodes of activity arrest had not had occurred for the last 2 years. EEG continue to show CSWSS, which is now more localised to frontal areas. The question to answer is whether this child should undergo epilepsy surgery in view of the localised brain lesion, intractable CSWSS and cognitive impairment but infrequent seizures.

4. Epilepsy after total congenital infarction of the left middle cerebral artery and additional cortical lesions on the right hemisphere

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Two children, Laureen and Kevin, show therapy resistant symptomatic epilepsy, a right hemiparesis and right hemianopia. They both had a total congenital infarction of the left middle cerebral artery.

Laureen had an additional watershed lesion of the right hemisphere (middle and posterior cerebral arteries); this lesion is very close to the presumed Wernicke's area. There are no obvious verbal problems.

Kevin had an additional partial infarction of the right middle cerebral artery; this lesion is very close to the presumed Broca's area. There are very evident verbal problems.

Laureen has many seizures including falls; Kevin showed already several life threatening episodes of status epilepticus.

What to do? Is it allowed to "complete" hemispherotomy on the left side, although knowing that both children will not get seizure free? How can we explain language representation? Is there a different strength to reorganization on the "healthy" hemisphere for Broca's and Wernicke's area?

5. Spasm-like seizures in a case of tuberous sclerosis

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We report the case of a 6-years-old boy with intractable epilepsy caused by tuberous sclerosis with

multiple tubers (left frontal, temporal and parietal and right temporal regions) who developed spasm-like seizures, with sudden head flexion, at 9 months of age. At 20 months of age, he presented isolated partial seizures with right dystonic leg movement and smiling followed by spasms. Neurological examination was normal but hyperactivity and attention deficit disorder were observed. Interictal scalp EEG showed paroxysmal slow, sharp and spike-waves over the bilateral central and parietal regions with predominance over the left hemisphere, more diffused during sleep. Ictal EEG showed a low voltage followed by bilateral spike discharges over the central and parietal regions and the vertex more predominant over the left hemisphere. During spasms we observed a diffuse sharp wave followed by polyphasic waves. SEEG was performed: 14 electrodes explored the left frontal (7), parietal (4) temporal (1) lobes and cingulate gyrus (2). Interictal epileptiform discharges were recorded on contacts located in the primary motor and sensitive area, in the SMA and the orbitofrontal cortex. The ictal SEEG showed a fast-activity discharge in the electrodes exploring post-central area, where was also located a tuber lesion. Stimulations of the somato-sensitive area elicited typical seizures. Focal left parietal resection was performed and the patient remained seizure free without neurological deficit.

Questions:
1. How to define these spasms-like seizures and what are their relationships with symptomatic partial seizures?
2. Take-home message focal resection in functional cortex, including dysplastic lesion, can be performed without neurological deficit?

6. Refractory cryptogenic frontal epilepsy: how to proceed?

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A 16-year old boy suffers from long-lasting, probably symptomatic frontal epilepsy. The disease started at the age of 3 years, with initial rare nocturnal tonic seizures. Up to 4 years of age they were not recognized and not treated. Thereafter, the seizure frequency increased and some diurnal tonic seizures and rare generalized tonic-clonic seizures manifested as well. Multiple awake EEGs showed generalized spike-wave discharges with frontal maximum and frontal lobe epilepsy was considered. The first brain MRI at 9 years was interpreted as normal. Nearly all antiepileptic drugs had no effect, a minimal reduction was observed under carbamazepine and oxcarbazepine. While the condition markedly deteriorated in the age 15-16 years, some focal ictal signs were also reported further suggesting frontal lobe origin of seizures, yet of unclear lateralization. Last summer the clinical status further worsened up to an epileptic status of tonic seizures in sleep with neurological deficit. The first sleep video-EEG monitoring registered multiple tonic seizures of various intensity and some longer seizures suggesting supplementary motor area onset or involvement, but next brain MRI did not reveal a clear pathology. The case is presented as an epilepsy surgery candidate with video- EEG sequences in order to clarify the seizure onset zone, the possibility and need for further neurophysiological and neuroimaging work-up.

Posters

P01. Evaluation of the Near Infared Spectroscopy (NIRS) to model changes in hemodynamics in relation to epileptic spikes

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Introduction. The relationship between epileptic spikes and local hemodynamism was evaluated by combined EEGNIRS recordings in rats.

Methods. Two ECoG electrodes, 2 light sources and 2 detectors were inserted bilaterally in the scalp above the SI cortex region of anaesthetized rats (n = 9). The epileptic spikes were induced by local application of of bicuculline methiodide to the left SI cortex. Data analysis for ECoG and hemodynamical activities were performed separately. The time of the spike peak was used for averaging ECoG and NIRS Data. A grand average was performed between the different rats.

Results. It is shown that the hemodynamic changes precede the epileptic spike in all rats. The hemodynamic changes are characterized by a biphasic pattern. The ″initial dip″ in oxygenated (HbO) and total (HbT) haemoglobin occurred before the spike onset and was followed by post-spike increasing in the HbO/HbT.

Conclusion. In accordance with results on fMRI (Gotman *et al.*, 2006), we demonstrate, using NIRS, hemodynamic changes which precedes the onset of spike activity. This suggests that such hemodynamic modulation has other than highly synchronized pyramidal cells origin. Thus NIRS analysis appears to be off interest in the evaluation of discrete brain dysfunction notably in presurgical evaluation in epilepsy.

P02. Variability of extreme isopotential areas in sequences of EEG maps containing seizure activity episode

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Introduction. The aim of the study was to evaluate the seizure activity dynamic by the analysis of the variability of extreme isopotential areas in sequences of EEG maps containing seizure activity episode.

Material and methods. The material comprised of the sequences of amplitude maps of records with the seizure activity for 17 subjects divided into two groups: clinically healthy subjects with the seizure activity and patients with epilepsy. The areas of isopotential regions in sequences of maps have been calculated. Estimation of the alternating variability of areas referring to - 20 mV (lowest) and 20 mV (highest) potential regions has been performed by visual analysis of the ratio and distribution of the values of both regions areas.

Results. In the group of patients the shape of curves of the ratios of both areas during seizure activity seemed to be regular and the both regions areas distributions are characterized by the regular dispersion. In the group of healthy subjects the shape of curves of the ratios of extreme areas during seizure activity is more variable than for patients and areas distributions tend to cluster. Based on the analysis of the extreme areas distributions coefficient differentiating the character of areas distributions for both groups of subjects has been defined.

Conclusion. The differences between analyzed relationships of extreme isopotential areas in EEG maps of healthy subjects and epileptic patients have been found. This was manifested by larger alternating variability of considered areas in the patient group. Application of the defined coefficient for two groups of subjects indicates that there is a moderate consistency between the quantitative results and the clinical assessment in differentiating between considered groups.

P03. Changes in filter settings during invasive video-EEG-monitoring help to define the seizure onset zone

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Objective. To provide evidence, that changes in filter settings can help to define the seizure onset zone during invasive video-EEG-monitoring.

Methods and results. Invasive long term video-EEG-monitoring was performed in a patient after implantation of subdural grid electrodes in the left mesial and centro-lateral perirolandic region. The electrode placement was based on data from a previous non-invasive monitoring with scalp electrodes. The sampling rate was initially 1 000 Hz and filter settings initially were LFF 0,53Hz and HFF 70 Hz. When the EEG analysis with these filter settings did not reveal a seizure pattern during clinical events the filter settings were changed to LFF 53Hz and HFF 300Hz and the EEG during the episodes was re-analysed.

Using these modified filter settings, regional ictal paroxysmal fast (ripple-) activity was now clearly detected in all myoclonic and tonic seizures of the right arm during the clinical events (electrodes A2-A4, A10-A12, E5, E6).

Conclusion. Modification of filter settings accompanied by a sufficient high sampling rate is important to uncover high frequency oscillations during seizures and may thus allow to deliniate the seizure onset zone in epileptic patients.

P04. Does MEG reflect interictal activity recorded during chronic subdural electrocorticography in children?

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We wanted to know whether interictal MEG accurately predicts the irritative zone in children, as established by the gold standard, chronic subdural electrocorticography (ECoG).

Eight children (4-16 years, 4 girls) underwent MEG and subsequent ECoG (5 left-sided implantations). MEG spikes were identified by a consensus method, clustered, averaged and modelled using the MUSIC algorithm. From chronic ECoG, all interictal spikes were sampled and localized. MEG and ECoG results were coregistred to the patient's surface MRI rendering.

Seven of 8 children had an MEG result. Of 9 interictal spike types, 90% showed at least one ECoG counterpart. Of 54 ECoG spikes, 45% were in the area indicated by MEG. MEG was most sensitive (> 75%) in the interhemispheric, parietal and central areas, and least sensitive (< 40%) in the mesiotemporal and frontal area.

MEG is not a substitute for ECoG, but can reliably be used for planning of grid placement. There seem to be regional differences in sensitivity of MEG. In the frontal lobe, MEG seems less sensitive than has been reported in adults.

P05. MEG/MSI evaluation of previously failed epilepsy surgery

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Objective. To review our experience with the contribution of MEG (magnetoencephalography)/MSI (magnetic source imaging) to the surgical evaluation of children who had previously had epilepsy surgery at our center or elsewhere, but failed to gain seizure control.

Methods. A medical record audit was performed to identify children who had MEG/MSI after a previously failed epilepsy surgery. The contribution of MEG/MSI to the decision whether or not to operate again to treat their epilepsy was reviewed.

Results. There were eight children, mean age at first surgery 8 years (range 2-17 years). All had MRI and ictal EEG recordings prior to the first surgery. Five children had their first surgical resection based on subdural electrode recordings of seizures, three on non-invasive evaluations and corticography during surgery. Seven patients had regions identified by MEG for additional resection. Some of these regions were adjacent to previously resected areas, others were in a different lobe. Of these seven patients, one is still considering surgery, and six have had additional surgery based on MEG/MSI findings. Four had topectomies and are seizure free. One had a hemispherectomy, and is seizure free. One patient with tuberous sclerosis has a greater than 90% decrease in seizures, following the second surgery. The eighth child was multifocal on MEG and not a surgical candidate.

Conclusion. MEG/MSI may help localize epileptogenic regions and define additional resection in children who have previously failed epilepsy surgery. In this small number of cases, re-evaluation of epilepsy surgical failures with MEG/MSI appears beneficial.

P06. Use of magnetoencephalography in epilepsies with continuous spike-waves during sleep

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Objectives. The electroencephalogram (EEG) pattern of continuous spike-waves during sleep (CSWS) is characterised by a diffusion of the spike-wave discharges (SWD) during non-REM sleep related to a mechanism of secondary bilateral synchrony from a focal cortical onset. The purpose of this study is to investigate the usefulness of magnetoencephalography (MEG) for localizing the source of epileptiform activity in children with CSWS.

Patients and methods. Four children (2 girls, aged 4 to 8 years) were investigated by MEG (Elekta Neuromag Vectorview 306) at the acute phase of CSWS. Two patients had Landau-Kleffner syndrome (LKS), one had global mental retardation and one had frontal syndrome. Spontaneous magnetic activity (lying position) was continuously recorded for 15 to 30 minutes during non-REM sleep and then visually inspected for epileptic events. The four patients were sedated. Equivalent current dipoles (g/% > 80%) corresponding to SWD were fitted in the patients' spherical head model and coregistered on their MRI. The onset of the SWD was determined using multi-dipole fitting.

Results. In the two patients with LKS, MEG showed that the SWD start in the right superior temporal gyrus and rapidly spread to the contralateral temporal region. In the two other patients, MEG revealed the existence of independent epileptic foci on both hemispheres with complex patterns of propagation.

Conclusion. MEG is a useful tool to identify the source of epileptiform activity and study its propagation patterns in children with CSWS. This technique provides valuable information to better understand CSWS pathophysiology and guide potential surgical treatment.

P07. Cortical mechanisms underlying late onset epileptic spasms associated with temporal lobe lesions

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Purpose epileptic spasms (ES) are characterized by sudden flexion or extension of the axial and/or proximal limb musculature, symmetrical or asymmetrical, with a wide range of ictal semeiology. Most often ES are age-related, usually beginning in the first year of life. The association with delay, arrest, or regression of psychomotor development, and with so-called "hypsarrhythmia" on the EEG, defines the classic triad of West syndrome. Several cases of late onset ES without any hypsarythmia and real cognitive regression have been described. The existence of such epileptic feature as an epileptic syndrome per se is still debated. Is it a late onset variant of west syndrome, an overlooked epileptic encephalopathy intermediate between West syndrome and Lennox-Gastaut syndrome, or focal seizures, with a specific, age related ictal phenomenology?

Methods. Two patients with late onset ES (seizure onset at 18 months and 4 years) without hypsarythmia were investigated by intracerebral recordings (SEEG). Temporal lesions suggestive of cortical dysplasia were located in the left mesial and anterior temporal region. Electrodes investigated premotor and temporal lobe structures. SEEG signal analysis used a new way to quantify the epileptogenicity of explored brain structures (Bartolomei, *et al. Brain* 2008; 131: 1818-30)

Results. In the two cases, ES corresponded to the delayed involvement of internal premotor cortex occurring with short delay after the temporal lobe onset. Maximal values of epileptogenicity were found in the temporal lobe.

P08. Anatomo-functional organisation of the insular cortex in epileptic humans: study using intracerebral electrical stimulation

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Introduction. Few data exist from human electophysiological studies performed in vivo by direct electrical stimulation of the insular cortex. The determination of clinical signs related to the participation of the insula permits a more accurate analysis of the electroclinical data.

Material and methods. This study focused on more precisely determining the anatomo-functional organization of the insula by studying the effects of intracerebral electrical stimulations during stereo-electo-encephalo-graphy exploration using an oblique approach. Twenty-five epileptic patients in whom at least one electrode was used to explore the insula were selected. A presurgical 3D-T1-weighted MRI scan and stereotactic software was used to target the insular cortex. A postoperative contrast-enhanced 3D CT scan enabled the fusion of the resulting image with the preoperative 3D MRI in the same stereotactic referenced system. The cortical location and anatomical position in reference to gyri and sulci could then be determined.

Results. All results were discussed in terms of their anatomical and electrical significance. Our data demonstrate a much more elaborate organization of the insular cortex in terms of sulci and gyri compared to the earlier simple distinction between anterior and posterior insula. We have been able to identify individual areas where direct electrical stimulation evokes language disorder, painful phenomena or sensory-motor responses. For the latter, we have identified a somatotopy.

Conclusion. The large sampling allowed by this exploration using an oblique approach has enabled us to draw up the first anatomofunctional insular cortex organization in terms of sulcal and gyral anatomy.

P09. Down-regulation of tubulin at postsynaptic density in rat temporal epilepsy and effect of tubulin stability on epilepsy seizure

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Numerous studies have show that temporal epilepsy (TLE) is associated with abnormity in neural network excitatory transmission, which is involved in molecular and functional change of three important parts, presynapse, synaptic cleft and postsynapse. Postsynaptic density (PSD) as super-signaling molecule complex in excitatory postsynaptic membrane, receives and transduces synaptic information. To date, the majority of studies examining synaptic protein profiles have focused on identifying the synaptic proteome. Only a handful of studies have examined the change of the PSD protein during TLE occurrance. We applied a quantitative proteomics analysis technique with MALDI-TOF-MS to quantitate relative changes in protein levels of the PSD in response to rat temporal epilepsy with chronic spontaneous seizure (CSS). We identified a total of twenty-six proteins of significant difference in the PSD, including cytoskeletal proteins α-tubulin and β-tubulin. Immunoblotting and immuno-electron microscopy studies showed that significant decrease of the two proteins in the PSD in CSS group compared to NSS group and controls, supporting the data from ITAT analysis. We further examed effect of disturbing tubulin stability on occurrence of TLE, through cochicine and paclitaxel treatment with intrahippocampal infusion. It was found a decrease of rate of spontaneous seizure and an increase of tubulin-labeled particles at PSD by immno-electron microscopy in. Taken together, these results suggest that, by changing the structure and composition of PSD to modulate synaptic plasticity, microtubule proteins α/β-tubulin at PSD are assotiated with the occurrence of TLE. And paclitaxel administration, polymerizing tubulin, may relieve the seizure.

P10. Vagus nerve stimulation in children with refractory epilepsy

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The vagus nerve stimulation (VNS) therapy is a neurostimulation technique used in treatment of patients

with refractory epilepsy. The studies in children are few. In the present paper we describe the results achieved until now in a group of children treated in our service with VNS for refractory epilepsy in Colombia (Medellin). This is uncontrolled, openlabel retrospective study. For the purpose of this study we included 13 children (9M/4F), between the 3 and 19 years with an age average of 10,2 years at time of implantation, and a mean post-implantation follow-up of 12,6 months (range 6-22). Diagnostic epileptic was refractory focal epilepsy in 7 children – of them 1 with sclerosis tuberosa –, Lennox-Gastaut syndrome in 5 children and mioclonic astatic epilepsy in 1 boy. The mean number of AEDs before implantation was 3 (range 1-4). Mean number of AEDs at maximum follow-up remained unchanged. The overall reduction in mean monthly seizure frequency was 75,5%. Mean seizure frequency before implantation was 272 seizures/month (range 5-600), and after implantation at maximum follow-up was 66,4 seizures/month (range 0-300). Responder rate was 62,6% (range -100% 100%). About 60% of patients had a seizure frequency decrease between 75% and 100%. Seizure freedom was obtained in 15.3% (2 children), while 23% had a seizure frequency decrease between 25-50%. One patient increase the seizure frequency and to other remained equal.

Conclusion. Our experience with VNS in children suggests that VNS is an efficacious adjunctive antiepileptic treatment for patients with refractory epilepsy.

P11. Ketogenic diet: alternative treatment option in patients with cortical malformations not eligible to surgery

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Objective. To evaluate the efficacy and safety of the ketogenic diet (KD) as add-on treatment in patients with neuronal migration disorders not eligible to surgery.

Methods. A retrospective analysis of all 64 patients with refractory epilepsy undergoing KD treatment from 1994 to 2009 identified 12 patients showing neuronal migration disorders not eligible to surgery. The classic KD was added to the baseline antiepileptic drugs and the efficacy was rated according to seizure type and frequency. During treatment, seizure frequency, side effects, urine and blood ketone levels and other parameters were systematically evaluated.

Results. Patients have been treated for 3-24 months (mean 10.6 months). 9/12 patients resulted responders with a reduction in seizure frequency between 50% and 90%. 2 patients resulted seizure free after 3 months of treatment. A relationship between diet efficacy and seizure type was noted: 71% of patients with spasms were responders with a complete disappearance of them in 42% and a reduction in frequency between 50% and 75% in the other. Partial seizures with secondary generalization disappeared completely in 2/2 patients. No significant relationship between diet efficacy and age at diet onset or sex. Adverse effects occurred in 5 patients (41%), but were generally mild and transient. Only 1 patient developed a metabolic failure with severe low level of potassium after 6 months requiring immediate diet withdrawal.

Conclusion. This retrospective study illustrates the efficacy and safety of KD in complex migration disorders and suggest its role as an alternative treatment option in those patients with refractory epilepsy not eligible to surgery.

P12. Effectiveness of corpus callosotomy for intractable epilepsy on development in young children

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Objective. To evaluate the efficacy of corpus callosotomy for intractable epilepsy on development, we investigated correlations between developmental quotient (DQ) before surgery and developmental velocity (DV) after surgery for young children with medically intractable epilepsies who were not candidates for epileptic focus resection.

Methods. Subjects comprised 63 children under 10 years old. The average age of surgery was 34.2 months (range 5-106). The kinder infant development scale (KIDS) was used to assess development before surgery and 1, 6, 12 and 24 months after the surgery. DQ and DV (difference of estimated developmental month between before and after surgery, divided by each follow-up point) were calculated for each patient. Epilepsy classifications were as follows: West syndrome, n = 33; Lennox-Gastaut syndrome, n = 2; symptomatic generalized epilepsy, n = 24; frontal lobe epilepsy, n = 2; and others, n = 2. Surgery involved total corpus callosotomy (n = 58) and anterior callosotomy (n = 5).

Results. Mean DQ before surgery was 21.6 (range 0-89). 30 of 63 children (47.6%) had preexisting development delay before seizure onset. DQ was gradually increasing for 33 children without preexisting delay (before surgery; 29, 1 month after; 30, 6 months; 31, 12 months; 32, 24 months; 35) and DV

also was stable (0.30, 0.31, 0.23, 0.32, respectively). In contrast, DQ declined in 30 with preexisting delay (13, 13, 11, 9, 8) and DV was lower (0.14, 0.03, 0.07, 0.04), respectively.

Conclusion. Corpus callosotomy yielded favorable outcomes in terms of development, particularly for young children without preexisting development delay. Corpus callosotomy should be recommended as early as possible when development delay is apparent.

P13. Seizure outcome after corpus callosotomy in children with learning disabilities and refractory drop attacks

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Background. Drop attacks (DA) are commonly associated with significant developmental delay and refractory to AEDs. Corpus callosotomy might ameliorate DA frequency and other generalized seizures. Methods: Post-surgical seizure outcome in epileptic children (< 18 years) with moderate to severe learning disabilities and refractory DA from 2 tertiary centres is evaluated after one stage total resection of corpus callosum with average follow up of 4 years (2 months-10 years).

Results. Seven consecutive patients (5 males) are included. Age at time of surgery ranges: 7-14 years (mean 10.66 ± 2.4); 2 with cryptogenic Lennox-Gastaut Syndrome, 2 postmeningitic bilateral infarctions, 2 refractory myoclonic epilepsies and 1 symptomatic to focal lissencephaly. All had injurious DA as their predominant refractory seizures (6-15 AED failed), were diagnosed in preschool age (< 4 years) without history of infantile spasms. Atypical Absence (AA) and GTCS were common in 6, myoclonic and focal seizures in 3, one case had nocturnal apnoeic seizures. MRI was normal in the 4 non-symptomatic children. DA disappeared with no change in other motor seizure frequencies in 4 cases. Nocturnal apnoea stopped as well. All seizures but AA disappeared in both postmeningitic children, with further improvement in their communication & motor power (follow-up: 7 months, 2 1/2 years). Mild initial improvement with secondary relapse was noticed in a single case. Surgery was complicated by: transient hemiparesis, angio-oedema of tongue (both totally resolved) and hyperactivity; each in single child.

Conclusion. Corpus callosotomy is a safe, probably effective surgical procedure in refractory drop attacks, yet long term neuropsychological outcome needs to be verified.

P14. Multiple subpial transections in medically refractory epilepsy: a single center experience in 63 consecutive patients

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Purpose. To present our results using multiple subpial transections (MST) for the treatment of medically refractory epilepsy (MRE) with epileptic foci in eloquent areas.

Methods. Between January 2003 and January 2009, 63 patients with MRE and epileptic foci in eloquent areas were treated by MST in rays, either isolated (MSTo group, n = 12, F/M = 4/8, mean age 21 y) or associated with resection or disconnection of other cortical areas (MST+ group, n = 51, F/M = 18/33, mean age 20 y). Our first 36 patients have a follow-up of at least two years, nine in the MSTo group and 27 in the MST+ one. Four outcome grades based on a modified Engel classification were used: seizure-free (100% seizure reduction; Grade I), important seizure reduction (75%-99%; Grade II), moderate reduction (50%-74%; Grade III) and at last no significant reduction (< 50%; Grade IV). Results. Regarding the potential MST related complications, we report 15 (24%) occurrences in the all series of which only one (1.6%) is permanent (right limb paresis, M4). Considering the seizures postoperative evolution at two years, in the MSTo group (n = 9), two (22%) patients are in grade I and six (66%) in grade II or III, while in the MST+ group (n = 27), nine (33%) patients are in grade I and 13 (48%) in grade II or III. In all patients, we observed at least seizure reduction associated with behavioral or cognitive improvement. Conclusion. MST in rays in MRE can be associated with significant seizure reduction (83.3%) and a low permanent morbidity rate (1.6%).

P15. Surgical efficacy and safety in subhemispheric multilobar pediatric epilepsy

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Objective. To demonstrate the safety and efficacy of surgical management in multilobar pediatric epilepsy with subhemispheric involvement. The prognosis for seizure freedom after epilepsy surgery is thought to be poorer in the setting of several epileptogenic zones involving multiple lobes, where hemispherectomy is contraindicated. We describe 13 patients whose age, language, and/or sensorimotor capabilities precluded hemispherectomy, who underwent multilobar

resections and disconnections in the setting of medically intractable seizures.

Methods. Ten children and three adults, ages 18 months to 46 years, with medically intractable epilepsy since childhood, were followed at the Comprehensive Pediatric Epilepsy Center at Beth Israel Medical Center in New York. Pre and post-operative evaluations included: EEG recordings with video monitoring, MRI, PET scans, and fMRI. All patients were treated with staged operations spaced by one week using invasive monitoring and functional mapping with grid and strip electrodes. Pre-operative MRI findings included perinatal infarction, post-traumatic encephalomalacia, cortical dysplasia, mesial temporal sclerosis, and a treated arachnoid cyst. All patients experienced disabling partial complex seizures and impediments to language, motor, academic, and social development.

Results. Seven children underwent a combination of functional and anatomic posterior quadrantectomy. In two of these patients, a partial frontal resection accompanied the quadrantectomy. One child with a dominant hemisphere perinatal parieto-occipital stroke underwent a temporal and occipital resection. The remaining five patients had combined temporal and frontal resections. Language and sensorimotor functions were preserved in all cases. All patients have had Engel I outcomes (12 Engel IA, 1 Engel 1C) with follow-up ranging from 7 months to 3 years.

Conclusion. Children and adults with multilobar epilepsy due to multiple pathogenetic mechanisms, who are not candidates for hemispherectomy, can be safely and effectively treated with epilepsy surgery. Staged operations with invasive mapping are necessary to comprehensively delineate the epileptogenic zones and distinguish them from functional areas.

P16. Epilesia partialis continua (EPC) in children

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Objectives. To define the electro-clinical spectrum of EPC in children in relation to MRI findings and etiology, and to evaluate epilepsy surgery in this context.

Methods. We reviewed 20 children with EPC studied at the pediatric video-EEG monitoring unit. Epilepsy features, neurological/neuropsychological and neuroimaging data were analyzed according to etiology.

Results. Five etiological categories were distinguished: 1) Rasmussen syndrome (RS, 5 cases), 2) mitochondrial diseases (MD, 6 cases), 3) malformations of cortical development (MCD, 5 cases), 4) destructive vascular lesion (1 case), 5) oncological disorders (3 cases). All patients showed focal motor/sensitive-motor seizures, and pyramidal cortical deficit. MRI lesions involved frontal/parietal cortex in 18 cases, white matter in 16, basal ganglia in 7. Patients with RS and MD showed rhythmic EPC with inconstant EEG correlation involving different muscular territories, progressive neurological deterioration, EEG background supression/slowing, multifocal epileptiform activity, and polymorphic seizures. These features were unilateral in RS and bilateral in MD. In patients with MCD, EPC manifested itself as an irregular muscular twitching associated with continuous regional/hemispheric epileptiform activity, aggravation of rolandic/opercular seizures, and functional motor derangement. Epilepsy surgery was performed in 6 children (3 RS, 3 MCD) with good results in 5 (seizure freedom or more than 90% seizure reduction).

Conclusion. Mitochondrial disorders, Rasmussen syndrome, and malformations of cortical development are the most significant etiologies of EPC in children. Video-EEG monitoring discloses distinctive features in each condition. Epilepsy surgery is an optimal treatment for selected children with EPC.

P17. Full-term newborn with TSC1 mutation, hemimegalencephaly and left hemispherectomy at 1 week of life

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This is a 1 year old female with of Tuberous Sclerosis (TSC1 mutation), who presented at birth with intractable rightsided seizure activity and left hemispheric hemimegalencephaly on MRI. Her EEG revealed nearly continuous electrographic seizures arising from the left frontal, central and parieto-occipital regions independently, often with clinical correlate that consisted of right arm and face clonic activity. At 1 week of life, she underwent the first of three operative procedures over 12 days for functional left hemispherectomy. By 3 weeks of life she was feeding by bottle, moving all extremities and had no further seizure activity. At two months of age, she began to have daily episodes bilateral flexor spasms that resembled infantile spasms, accompanied by generalized bursts on the EEG. These events resolved completely with Vigabatrin. At 3 months her partial right-sided motor seizures returned, involving frequent head and eye deviation to the right and right arm clonic activity. On EEG she had frequent electrographic and clinical seizures arising from the left frontal region. She returned to the

operating room for complete anatomical left frontal lobectomy at the age of 3 months. She has been seizure-free since that time. Currently, she is 12 months old and smiles frequently and reactively, sits without support, makes babbling sounds and has a resolving mild right hemiparesis.

Discussion/take-home message. Hemispherectomy can be performed safely in infants with hemimegalencephaly, and can result in drastically improved developmental outcome by preventing prolonged intractable seizure activity in the early months of life.

P18. Hemispherotomy for hemimegalencephaly with early infantile epileptic encephalopathy: surgical result of 9 cases

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Hemimegalencephaly (HME) is a rare brain malformation characterized by unilateral hypertrophy of cerebral hemisphere, sever drug-resistant seizures, and EEG abnormalities such as suppression burst. Although hemispherectomy/hemispherotomy is the most effective treatment to control seizures, surgical therapy of HME in infants is still burdened by a high complication rate and mortality risk. We have experienced hemispherotomy for 9 infants of HMC with early infantile epileptic encephalopathy with the post-operative follow-up from 1 to 5 years. In all the cases, seizures started in neonatal period succeeded by daily frequent spasms, suppression burst EEG abnormality and developmental delay. Surgery was performed within 3 to 7 months after birth. Hemispherotomy was performed vertically through a cortical window of 4 cm x 2 cm placed at the fronto-parietal cortex and whole pathological cortex was disconnected from the thalamus, basal ganglia, brain stem and the contra-lateral hemisphere. Complete seizure freedom was obtained in 6 cases, whereas monthly seizures remained in 1 case and daily seizures in 2 cases. No significant complication was experienced except one case with post-operative progressive hydrocephalus requiring VP-shunt. Developmental quotient (DQ) at the time of follow-up was better in seizure free group; *i.e.*, DQ 10 to 63 (av. 34.5) comparing to residual seizure group; *i.e.,* DQ 5 to 29 (av. 13.3).

Conclusion. Favorable surgical outcome can be obtained for HMC in infants associated with EIEE by using vertical hemispherotomy.

P19. Modified vertical parasagittal hemispherotomy for intractable hemispheric epilepsy

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Objective. To present our methodological modification and experience on the Delalande's vertical parasagittal hemispherotomy.

Methods. Vertical hemispherotomy was performed for 4 patients with intractable hemispheric epilepsy, including 3 infants with hemimegalencephaly or extensive cortical dysplasia, and one adult with traumatic brain lesion. The step for the anterior disconnection is modified. Otherwise our technique basically follows the Delalande's original method. After paramedian frontal craniotomy, complete section of corpus callosum is placed first. Entering the lateral ventricle, small amount of the cingulate gyrus is aspirated, and the dorsal aspect of the thalamus and the stria terminalis are identified via interhemispheric space by retracting the frontal lobe laterally. The incision is made lateral to the thalamus to disconnect the descending projection fibers, which is advanced until the inferior choroidal point is identified by following the inferior horn. Then, the incision is completed by connecting the inferior choroidal point and the foramen of Monro. Finally, the fornix is cut at the level of trigone.

Results. No surgical complications were seen. Excellent seizure outcome was obtained in 3 patients. Complete disconnection was confirmed in post-operative MRI.

Conclusion. Compared with the original Delalande's method, our modification further reduces the amount of corticectomy and the length of brain incision to complete the hemispherotomy. Vertical hemispherotomy can be accomplished through a small cortical window with low risk for surgical complication. This is especially efficacious in infants with low body weight.

P20. Movement disorders in three children after hemispherectomy for refractory seizures

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Objectives. Hemispherectomy is treatment option in patients with intractable epilepsy and widespread hemispheric abnormalities or processes. The more common postoperative complications of hemispherectomy include hydrocephalus and infections. In a

previous study, dystonic or bilateral ballistic movements have been reported in one adult patient years after an anatomic hemispherectomy. We review the incidence of post operative movement disorders in children who underwent hemispherectomy in our Institution.

Methods. Medical records of seventeen children who underwent hemispherectomy for refractory seizures at the Children's National Medical Center from March 1996 to January 2009 were retrospectively reviewed from a surgical database.

Results. Three of seventeen patients who had a transient or persistent movement disorders following hemispherectomy. Patient 1 presented with refractory seizures at age 10 and Rasmussen's encephalitis and underwent left hemispherectomy at age 12. She developed left resting tremor and parkinsonian features after the surgery. Her symptoms have improved and the resting tremor has abated with medications. MRI showed an infarct in the left thalamus and an intial edema on the left basal ganglia; subsequent MRI showed a persistent signal abnormality in the left caudate. Patient 2 underwent left hemispherectomy for hemimegalencephaly at 5 months of age. He developed jerking movements of the right upper extremity on day 1 postoperatively with no electrographic correlate on video EEG. The movements resolved the next day. Patient 3 had an initial right functional hemispherectomy at age 27 months secondary to an extensive left cortical dysplasia, then an anatomic hemispherectomy 2 years later for continued seizures. She developed hand postural tremors that lasted for 1 month seven months after the second surgery which eventually resolved.

Conclusion. Transient and persistent movement disorders were seen in 3 children who underwent hemispherectomy. Movement disorders may be a little recognized complication of hemispherectomy and may be related to perioperative involvement of the basal ganglia.

P21. Seizure outcome and complications after vertical parasagittal hemispherotomy in children

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Objective. Hemispherectomy and hemispherotomy are recognized as effective treatments for severe epilepsy caused by large lateralized epileptogenic lesions, but there is only one previous report of vertical parasagittal hemispherotomy (Delalande, *et al.* 2007). We analyzed our experience in using this technique.

Methods. Twenty-four children underwent vertical parasagittal hemispherotomy in 1995 through 2008. The mean age at operation was 6.8 (range, 0.4 to 18) years. Preoperative work-up included video-EEG and MRI. Twenty-two children had daily seizures and two had fewer seizures but electrical status epilepticus in sleep. Etiology was vascular or other de-structive lesions in 14 children, brain dysgenesis in five, chronic encephalitis in three and Sturge-Weber in two.

Results. Four children (17%) continued to have seizures postoperatively; three of them had a reoperation to complete the disconnection. At two years of follow-up, 18 children (90%) were seizure free (including two reoperated). All four children with a shorter follow-up (two to 12 months) are seizure free. There was one serious permanent complication in a child who was operated at three years in a grossly malformed hemisphere while the other side appeared structurally normal. He had postoperative hypothalamic dysfunction with hypersomnia, hyperphagia and multiple hormonal deficiencies. Five children (21%) required shunt placement postoperatively. One child had transient diabetes insipidus.

Conclusion. Seizure outcome after vertical parasagittal hemispherotomy seems to be similar to other hemispherotomy techniques. Severe hypothalamic dysfunction was observed in one child in both the previous and the present patient series; this has not been reported after other disconnecting procedures.

P22. Predicting motor function of arm and hand after hemispherectomy

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Objective. Most children maintain their ability to walk after hemispherectomy. Arm and hand functions, however, often deteriorate. We investigated whether the type and timing of the epileptogenic lesion predicted changes in motor function of the arm after hemispherectomy.

Methods. Standardized assessments of muscle strength and tone before, 6 months, and 2 years after hemispherectomy in 35 children (≤ 16 yr). Etiologic classification was based on timing of epileptogenic lesions: "early" (< 3rd trimester; n = 14) *versus* "late" (> 3rd trimester; n = 21); or on type of pathology: "hemispheric malformations" (n = 13), "vascular" (n = 12), or "other" (n = 10). Hand function was dichotomized as "good" or "poor".

Results. In children with early lesions, preoperative hand function was good more often than in late lesions (93% *vs* 38%, p = 0.001), but at 6 months distal strength decreased more frequently (79% *vs* 38%, p = 0.036). In 85% of children with hemispheric malformations distal arm strength decreased postoperatively, compared to 17% of vascular patients. Early postoperative reduction of strength was predictive for a permanent decrease (p = 0.003). The eventual postoperative dichotomized hand function did not differ between etiological subgroups, and was "poor" in approximately 80% of all cases.

Discussion. Children with early developmental lesions have a better preoperative hand function, which is frequently lost after hemispherectomy, implying preoperative motor innervation from the epileptic hemisphere. Children with late lesions typically had poor hand function preoperatively, and did not deteriorate. Thus, they probably already depended on ipsilateral innervation before surgery. Eventual postoperative motor function did not differ between early and late lesions on a group level.

P23. The influence of contralateral EEG and MRI abnormalities on seizure outcome and cognition after hemispherectomy

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Objective. To study how structural and functional abnormalities in the contralateral hemisphere affect seizure and cognitive outcome after hemispherectomy.

Methods. Of the 40 children (age at surgery 0-16 years) who underwent a hemispherectomy between 1991 and 2007 with a minimum postoperative follow up of two years, all preoperative EEG's were reviewed retrospectively for the existence of (inter)ictal epileptic or other abnormalities in the contralateral hemisphere. Preoperative MRI's were evaluated for the existence of contralateral abnormalities. In 23 children, intelligence or developmental quotients (IQ/DQ's) were obtained pre- and 2 years postoperatively. Seizure freedom was defined as Engel 1A-C. We tested the correlation between contralateral EEG and MRI abnormalities and postoperative seizure freedom and cognitive outcome. Results: Postoperatively, 31 children achieved seizure freedom (78%). Contralateral epileptic EEG abnormalities correlated with the presence of contralateral MRI lesions, and with underlying etiology (hemispheric malformations: 47%, vascular lesions: 71%, other etiologies: 100%). EEG abnormalities in the contralateral hemisphere did not correlate with seizure outcome. Contralateral MRI abnormalities significantly predicted seizure recurrence (p = 0.034). Seizure outcome significantly related to insulectomy (p = 0.029), and to etiology (recurrence risk in malformations: 6%, vascular lesions: 24%, other etiologies: 67%, p = 0.014). The existence of contralateral EEG and MRI abnormalities was not associated with pre- and postoperative IQ/DQ's, nor with the difference between pre- and postoperative IQ/DQ's.

Conclusion. Preoperative contralateral MRI abnormalities increase the risk of seizure recurrence after hemispherectomy. Contralateral EEG abnormalities do not predict seizure outcome. MRI or EEG abnormalities in the "unaffected" hemisphere do not correlate with cognitive recovery.

P24. Epilepsy surgery in children with epileptic encephalopathies

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Objectives. To review the electro-clinical presentation, neuroimaging findings and outcome in children who underwent resective surgery for refractory epilepsy and had an epileptic encephalopathy documented during preoperative video-EEG.

Methods. From our resective epilepsy surgery series, we identified 25 children with prominent interictal epileptiform discharges and/or frequent subintrant seizures during preoperative video-EEG.

Results. 18 patients (72%) had malformations of cortical development (MCD): 8 hemispheric/subhemispheric MCD, 4 focal MCD, 4 tuberous sclerosis and 2 neuroglial tumours; 6 children (24%) had destructive hemispheric lesions and one had a hypothalamic hamartoma (HH). Epilepsy onset was during neonatal period in 11 children (6 hemispheric/subhemispheric MCD), 8 of them with focal motor seizures; between 3-12 months in 7 patients, 6 of them with infantile spasms; and after the first year of life in 7 cases, all of them with epileptic spasms during evolution. Most patients (88%) had a dominant EEG focus, concordant with lesion location; EEG discharges had no predominance in 3 children (one hemispheric MCD, one hemispheric destructive lesion and one HH). We recorded seizures with focal motor features and/or focal EEG onset in all but 3 patients, who had other evidence of focality. At 29 months' mean postoperative follow-up, 68% were either seizure free (11 patients) or had a reduction of seizure frequency > 90%. Two out of 5 patients with no significant improvement, were seizure-free for 2.5 years before seizures relapsed.

Conclusion. Children with epileptic encephalopathies and focal lesions on neuroimaging can be good surgical candidates, despite abundant epileptiform abnormalities on EEG or non-localizing seizures.

P25. Anatomic or idiopathic epilepsy?

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Partial epilepsies in children are often idiopathic, particularly those involving the centro-temporal regions. Strict criteria are necessary for diagnosis: clinical, electrophysiological, and developmental.

We report the case of a six year-old boy whose seizures were considered as suggesting a benign Rolandic or occipital epilepsy. Clinical examination, developmental assessment and EEG were compatible with benign epilepsy. Family history was negative. He had no difficulties at school; his neuropsychological evaluation (WISC) was satisfactory. Aggravation was reported, characterized by an increasing frequency of nocturnal seizures. Transitory right arm and speech deficiency were observed on awakening. Interictal EEG pattern evoked idiopathic focal epilepsy without ESES. At the age of 8 years, his state slowly worsened: disability poor elocution and sleep-quality; he became more and more troubled with dribbling. EEG suggested an area of epileptogenic cortex, spikes and spike-waves arising from the left temporal, frontal and rolandic regions. In spite of several AED trials, focal epilepsy remained drug resistant, and led to a new neuroimaging investigation (MRI) that showed a bilateral schizencephaly of type I (fused lips): two CSF intensity clefts, nearby the calcarine fissures, extending from the lateral ventricles to the temporal cortex and lined by gray matter. This dysplastic area constitutes an epileptogenic region that can explain intractable epilepsy. Diagnostic issues related to idiopathic *versus* symptomatic epilepsy are discussed.

P26. Hippocampal sclerosis in idiopathic epilepsy and its implications for epilepsy surgery

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Objective. To provide evidence that hippocampal sclerosis can occur as result of recurrent seizures in patients with idiopathic epilepsy and to discuss different treatment options on grounds of electrophysiological data.

Case 1. A 20 year old patient with history of febrile status at 6 months of age presented for presurgical evaluation with a refractory epilepsy with epigastric auras, automotor and generalized tonic-clonic seizures. Long term video-EEG-monitoring revealed left temporal sharp waves as well as left temporal EEG seizure onset. On discontinuation of anticonvulsive medication frequent generalized spike-wave-complexes were recorded. On MRI a left sided hippocampal sclerosis was present. A diagnosis of left temporal lobe epilepsy and idiopathic epilepsy was established.

Case 2. A 10 year old patient presented for presurgical evaluation with a refractory epilepsy with visual auras, ictal vomiting, automotor and generalized tonic-clonic seizures. Long term video-EEG-monitoring revealed bi-occipital sharp waves and generalized spike-wave-complexes. The EEG seizure onset was either right or left occipital. On MRI a right sided hippocampal sclerosis and no occipital pathology was detected. A diagnosis of idiopathic focal occipilal lobe epilepsy was established.

Conclusion. Idiopathic epilepsy can lead to secondary hippocampal sclerosis which may be of relevance for further management. In case 1 clearly left temporal seizures were the culprit for refractory epilepsy and a selective amygdalahippocampectomy was considered. In case 2, the hippocampal pathology was not associated with the epileptogenioc zone and surgical treatment would be not beneficial.

P27. Bitemporal depth electrode investigation in children and adolescents with intractable temporal lobe epilepsy

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Objectives. Limited data exist on the usefulness of bitemporal depth electrodes for the presurgical evaluation of children and adolescents with temporal lobe epilepsy.

Methods. A retrospective study on patients under 19 years of age who went through an invasive video-EEG recording using bitemporal depth electrodes in CCF from 1997 to 2007 is presented. For the invasive EEG recording the patients had six depth electrodes implanted stereotactic bitemporally targeting the amygdala, the anterior and the posterior hippocampus. For the present study, clinical and imaging data were collected from the medical records and the EEG data was re-evaluated. The two-year postoperative outcome is presented according to Engel *et al.* (1993).

Results. Ten patients, 11 to 18 years of age, went through a bitemporal depth electrode recording. The main indications for the invasive studies were bitemporal MRI findings in four, normal or subtle findings in

MRI in four and bilateral video EEG and semiological patterns in two patients. FDG-PET was either localizing or lateralizing in nine patients. One patient had an uncomplicated bleeding during implantation of depth electrodes. Bitemporal interictal spikes were seen in all patients. Seizure onset in 3 patients was unilateral mesial temporal, bitemporal independent in 6 and unilateral neocortical temporal in one. Seven patients underwent temporal lobe resection. Seizure outcome 2 years postoperatively was Engel Ia in 6 patients and Engel 1c in one patient.

Conclusion. Bitemporal depth electrode recordings provided clinically useful data in children and adolescents with temporal lobe epilepsy. Seizure outcome of patients who then underwent temporal lobectomy was excellent.

P28. Ictal and interictal EEG patterns in children with temporal lobe epilepsy - correlation with surgical outcome

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Objective. To identify ictal and interictal EEG patterns in children with temporal lobe epilepsy (TLE) and to correlate with surgical outcome.

Methods. One hundred and fifty five ictal video EEG recordings and 24 hour awake and sleep interictal EEG of 28 children with TLE who underwent surgery and had at least 1 year post- surgery follow-up were reviewed. Ictal EEG patterns were classified according to (1) frequency (theta, delta, alpha, others) (2) spatial distribution (focal, regional, hemispherical, others) (3) duration of persistence of rhythm (< 5 sec and > 5sec) and (4) morphology (Ebersole and Pacia, 1996). The outcome was assessed according to Engel's outcome classification (ClassI- favourable outcome and class 2-4 unfavourable outcome). Interictal EEG was classified as preponderantly unilateral (> 75% discharges unilateral) and those with bilateral interictal epileptiform discharges.

Results. The age range was 6-16 (mean 13.3) years; Engels class-I outcome was noted in 17 (61%), class-II in 3 (11%) and class-III in 8 (28%). Histopathology revealed hippocampal sclerosis (HS) in 16, dual pathology-2, tumoral lesions-4, gliosis-2 and neuronal loss in 4. Ictal EEG showed typical rhythmic theta pattern at onset in 18 (64%), focal/regional distribution in 19 (68%) patients. Ebersole's type IA pattern was seen in 12 (43%), IB in 7 (25%), IC in 6 (21) and was uncertain in 2 patients; 21(75%) had persistence of rhythm for > 5 sec. The predictors of good surgical outcome, OR (95%CI) were focal/regional distribution 2.88 (1.021-13.985), Ebersole's type 1 pattern 1.889 (1.05-33.891), and persistence of rhythm for > 5 seconds 19.2 (1.844-19.937) on ictal EEG and unilateral spikes on interictal EEG 21 (2.913-151.408). Predominant posterior temporal spikes are a predictor of poor outcome ($p < 0.05$). A unilateral interictal discharge is a predictor of ipsilateral seizure origin in 90% patients ($p \leq 0.05$).

Conclusion. Focal/regional distribution, Ebersole's type1 pattern and persistent rhythm for > 5 sec on ictal EEG and unilateral spikes on interictal EEG are predictors of favourable outcome. Predominant posterior temporal spikes are a predictor of poor outcome.

P29. Resection of intraventricular subependymal giant cell astrocytomas in tuberous sclerosis complex

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Introduction. Subependymal giant cell astrocytomas (SEGAs) occur in about 10% of tuberous sclerosis complex (TSC) patients. Associated morbidity occurs mostly due to obstructive hydrocephalus. Diagnostic criteria to differentiate SEGAs from subependymal nodules and indications for resection remain controversial. We present our experience treating patients with intraventricular SEGA.

Methods. We retrospectively reviewed of all patients with TSC that underwent resection of a SEGA. Indications for surgery, size of lesion(s) and ventricles, surgical technique, pathological findings and clinical/radiologic follow up were reviewed.

Results. Eleven patients underwent 13 surgeries. Mean age at surgery was 11 years old. Indications for surgery were tumor growth (10), intratumoral hemorrhage (1), and mild ventricular enlargement associated with tumoral growth (2). Two patients had been previously shunted due to hydrocephalus, two had mild ventriculomegally, and seven had small ventricles. Ten patients were asymptomatic at the time of surgery; one had acute symptoms of increased intracranial pressure and two had mild symptoms. The surgical approach was transcallosal (12) and transcortical (1), and included total gross resection (TGR) (9), near TGR (3), and one subtotal resection. Postoperative morbidity included transient somnolence in one patient. Three patients required new shunts, and one required a septum pellucidotomy. At last follow-up (mean 23 months) all patients were neurologically stable and had no radiological evidence of recurrence.

Conclusion. Resection of SEGA is associated with low morbidity and no mortality, and should be considered

if there is radiological evidence of tumor growth or associated hydrocephalus. However, even after resection, hydrocephalus may develop.

P30. Triple pathology in epilepsy: two cases with coexistence of cavernous malformation, cortical dysplasia, and other lesions

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Cavernous malformations, cortical dysplasias, arteriovenous malformations including venous angiomas, and hippocampal sclerosis are frequent histopathological substrates in patients with medically intractable epilepsy. While cortical dysplasias may not be uncommon in conjunction with hippocampal sclerosis, coexistence of cortical dysplasia and cavernous malformations has rarely been reported. We reviewed our surgical specimens from patients who underwent surgery for drug-resistant epilepsy between 2003 and 2008, and identified two cases with both cavernous malformations and cortical dysplasia. In addition, each of these patients had a third form of a potentially epileptogenic lesion: hippocampal sclerosis in one patient and a venous angioma in the other. The finding of multiple malformative pathological abnormalities in this setting of medically intractable epilepsy suggests as possible common underlying developmental cause or process contributing to this coincidence. Analyses of further clinical experience and investigation of pathological mechanisms underlying impaired neocortical morphogenesis is necessary to elucidate the significance of the cases reported herein.

P31. Giant subcortical heterotopia involving the temporo-parieto-occipital region: a challenging cause of drug-resistant epilepsy

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Gray matter heterotopia is a cortical malformation which may cause intractable epilepsy and cognitive delay. Giant subcortical heterotopias have only occasionally been described. Surgical removal has been proposed for the management of seizures. Good results are reported after complete excision, but one concern comes from a possible functional role of the heterotopic tissue. Three patients affected by giant temporo-parieto-occipital subcortical heterotopia underwent surgical treatment. Two subjects got rid of seizures after complete excision. The third patient underwent a subtotal excision, in order to space the motor cortex possibly involved in the malformation, achieving only a reduction of seizures. An improvement of development and behaviour without neurological worsening was observed in all three patients. Patients affected by giant subcortical heterotopias seem to present a favourable prognosis after the surgical removal, even in cases of long-lasting epilepsy, in comparison with other extensive cortical malformations for which adjunctive disconnective procedures are often advocated.

P32. Correlation of non-Taylor type focal cortical dysplasia (FCD) MRI criteria with neuropathology: preliminary observations

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Background and purpose. Epilepsy associated classical focal cortical dysplasia (FCD), first described by Taylor (1), is a distinct structural abnormality with cortical laminar disorganisation interspersed with dysplastic neurons and associated with so-called "balloon" cells. Subsequently, heterogenous lesions were added generating a spectrum of Taylor and non-Taylor type FCDs. Several classification schemes have been proposed for FCDs integrating electrophysiology, MRI, and neuropathology (2, 3). Non-Taylor type FCDs were charcterized by core atrophy and/or focal brain atrophy with moderate white matter signal intensity alterations (3). We present preliminary neuropathological findings in a consecutive series of pediatric epilepsy surgery specimens showing MRI characteristics of non-Taylor type FCD according to (3).

Patients and methods. Twenty children with drug-resistant partial epilepsy, who underwent surgical resection of the temporal lobe, were reviewed. MR examinations were performed on a 1,5 Tesla system with sequences as follows: axial FLAIR sequence of the entire brain, (5 mm), paracoronal (perpendicular to the course of the hippocampus) turbo SE T2w sequence (2 mm), Inversion-recovery and FLAIR sequence (3 mm). MRI was reviewed with respect to: presence of hippocampal atrophy/sclerosis, small temporal lobe, blurring of the gray-white matter junction, signal changes of the subcortical white matter.

Results. In 8/20 cases atrophy and/or signal changes of the hippocampus were associated with blurring of the gray/white matter interface, and/or small ipsilateral temporal lobe and were thus classified as non-Taylor type FCD. In 5/8 (62,5%) cases histologic diagnosis was

hippocampal sclerosis with neuronal loss and reactive gliosis. 2/8 (25%) showed unspecific reactive changes with gliosis and inflammatory infiltrates, and 1/8 (12,5%) displayed no pathologic changes at all. In all cases, the white matter showed sparsely neurons in the subcortical white matter. The number of white matter neurons did not exceed that of normal controls. None of the cases displayed laminar disorganisation of the cortex.

Conclusion. Based on our preliminary experience, prospective use of Colombo *et al*'s MRI criteria for non-Taylor type FCD seems not strictly associated with the neuropathological finding of cortical disorganisation.

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P33. Surgical treatment of complex partial status epilepticus caused by cortical dysplasia in a 3-year-old girl

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Objective. Urgent resective surgery appears to be an option in treatment of patients with convulsive or nonconvulsive refractory status epilepticus due to focal brain pathologies although it is not often used in clinical practice. We present a case report of urgent surgical treatment of complex partial status epilepticus caused by cortical dysplasia in a 3-year-old girl.

Methods. Video EEG monitoring, MR imaging, psychological examination, surgical treatment, and postsurgical histological evaluation were performed.

Results. We present a case report of a girl who first seizures occurred at the age of 1 year and 9 months. She was treated with AEDs without any effect, by contrast there was a slow progressive worsening in daily number of seizures. The frequency of seizures increased and the complex partial status epilepticus started at the age of 3 years without any effect of standard treatment with AEDs and thiopental coma. The MRI scans showed extensive focal cortical dysplasia of right frontal lobe. The urgent surgical intervention (the lesionectomy) was performed at the age of 3 years and 2 months and the histological examination approved the cortical dysplazia Ib (Palmini 2001). Since the surgery was performed, seizures have occurred only occasionally with frequency of 2 partial simplex seizures per year and the neuropsychological examination has confirmed the continuation of the psychomotor development.

Conclusion. Urgent epileptosurgery of lesional complex partial status epilepticus is an option of treatment and we think that it is necessary to take it into consideration in patients suffering from urgent epileptic conditions.

P34. Focal cortical dysplasia type 2B may produce motor responses by non invasive mapping: a pediatric case study

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Objective. We report surgical outcome in a nine-year-old patient with refractory epilepsy, who had a focal cortical dysplasia (FCD) in the left frontal lobe anterior to the motor gyrus. Hand motor responses were obtained by noninvasive methods from the FCD.

Methods. Preoperative work-up included motor representation mapping with navigated transcranial magnetic stimulation (nTMS); localizations of sensorimotor areas and epileptic activity with magnetoencephalography (MEG); hand representation by functional magnetic resonance imaging (fMRI); pyramidal motor tracts depicted in probabilistic tractography; and localizations of sensorimotor cortex and epileptic activity with chronic subdural grid recording and electrical cortical stimulation (ECS). nTMS, MEG, and fMRI were repeated 6 months postoperatively.

Results. Preoperatively, noninvasive methods localized motor and sensory representations more anteriorly than expected, and overlying the FCD visible on MRI. ECS localized the same functions more posteriorly to the region which by anatomical landmarks was estimated to be the motor gyrus. Tractography showed one bundle of motor tracts connected to the precentral gyrus, and another to the FCD. Resection of the FCD was carried out without any postoperative motor deficit. Pathology showed FCD2B. Postoperative nTMS and fMRI revealed a large scattered motor representation, with no clear boundaries between hand muscles, posterior to the sites activated preoperatively, and MEG showed posteriorly shifted sensory responses. These representations were shifted towards the areas depicted preoperatively by ECS. The patient is seizure-free.

Conclusion. FCD2B may contain non-essential but functional motor tracts, which may be overrepresented in noninvasive mapping methods. Hyper-excitability of the epileptogenic cortex in FCD may inhibit primary motor areas.

P35. Symptomatic focal epilepsy due to focal cortical dysplasia type IIB: a case report

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Objective. We present the case of 8 years old boy with normal history of pregnancy, birth, postpartal period and psychomotor development until the age of 3 years. At the age of 20 months he experienced first convulsiones (febrile) but 4 months later a febrile seizure occur. Antiepileptic drug was introduced and child was seizure free for next 6 months when seizures relapsed. In the following years the frequency of seizures was changing from cluster with up to 20 seizures per night to seizure free periods lasting weeks or even month. At the age of 5 years he had up to 30 seizures per day. Almost all antiepileptic drugs were given in mono- and polytherapy but with no success. Poor control of the epilepsy was accompanied by retarded speech development, hyperactivity and attention deficit (ADHD).

Methods. Semiology of the seizures occuring mainly during first night sleep sometimes during day describes most often eye deviation to the right side, version to the right, clonic movements of the right corner of the mouth lasting 20 to 30 seconds with a loss of urine. EEG recorded focal discharges over both hemispheres becoming more intensive on the right side. Worsening of the disease manifested with further deterioration in the EEG. High amplitude slow activity appeared in the right frontal region with bilateral focal discharges. Several brain MR scans with spectroscopy showed suspicious prolipherative tumorous lesion in the right frontal lobe. Regarding the suspected tumor patient had two stereotactic biopsies whose histopathology finding revealed cortical dysplasia type IIB. Soon after a partial resection of the lesion took place and the diagnosis was confirmed.

Results. The boy had immediately after surgery only few abortive seizures in the first sleep and was seizure free ten months after. Unfortunately seizures started again with high frequency in spite of antiepileptic therapy. Postoperative MR scan diagnosed the residual parts of cortical dysplasia in the right frontal lobe.

Conclusion. With the diagnosis of focal cortical dysplasia type IIB the chance to become seizures free with medication alone is very small and a second resection is recommended. Before performing another surgery seizure onset zone should be identified through prolonged Video and EEG monitoring. High resolution brain MR scan and ictal and interictal SPECT will be done as well as methionine and FDG-PET. The diagnostic results will be requested and discussed with neurosurgeon, pediatric neurologist and radiologist hoping that well tailored second surgery might free the child from epilepsy.

P36. Epilepsy surgery in tuberous sclerosis complex

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Objectives. Which patients are good surgical candidates and how to select operative approach for TSC patient are still perplexity. We want to discuss the two questions by reviewing our experience.

Methods. We retrospectively analyzed clinical datum of 19 cases with TSC and secondary intractable epilepsy who were underwent surgical therapy in our department from September 2001 to May 2007. Preoperative assessment include physical examination, initiative sign of seizure, video-EEG monitor, PET/SPECT, MRI and CT, IQ test. There were 8 patients with focal epilepsy and 11 patients with bilateral or multi-zone epilepsy, and contex EEG were recoded in 10 patients with multi-zone epilepsy, and 6 cases can find focal epileptogenic zone. IQ of 9 patients were low than 70, and 3 patients' IQ were norm, 7 cases had light deficiency of intelligence. Surgical procedures included 1 corpus callosotomie, 3 lesionectomies, 5 lobar restections and 5 lesionectomy and lobar resections, 2 corpus callosotomies and lesionectomies, 1 corpus callosotomies and lesionectomy and lobar resections.

Results. Of the total sample 11 patients had Engle Class I outcome, 5 had class II outcome, 3 class III or IV. For the 4 patients with uncertain of epileptogenic zone, nobody reached seizure free, and there were significance of difference in surgical outcome between cases with localizable epileptogenic zone and unlocalizable ones. The sex, age at seizure onset and IQ were not associated with outcome.

Conclusion. There were excellent surgical outcome for TSC with focal epilepsy, which patients are reasonable candidates for epileptic surgery. Validity of surgery for TSC with multiple epiletogenic zone were less than patients with focal epilepsy, but surgery for those patients is availability when introcranial electrode were used and epileptogenic zone were localized. Lesionectomy and lobar resection are essential procedures for TSC.

P37. Identification of the epileptogenic zone in patients with tuberous sclerosis: concordance of interictal and ictal epileptiform activity

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Purpose. The interictal EEG often shows multifocal interictal epileptiform activity in patients with tuberous sclerosis complex (TSC) and its value in identifying

the epileptogenic zone has been considered of little value in this patient group. Recent studies have demonstrated the presence of one dominant and consistent interictal epileptogenic focus in a selection of patients. We hypothesize that dominant interictal epileptiform foci are concordant with the ictal onset zone in TSC patients.

Methods. Consistency and dominance of focal interictal epileptiform activity was assessed in 19 TSC patients. The ictal onset zone was identified. Concordance between interictal en ictal findings was analyzed. If concordance was found epilepsy surgery was proposed.

Results. We found a dominant, consistently present interictal focus/lobe in 74% of the patients. Concordance between the dominant interictal foci and the ictal onset zone was found in 79% of these patients. Six patients underwent surgery. Three patients achieved seizure freedom (50%), and one a reduction > 90% after a follow-up of 2.8-7 years.

Discussion. This study illustrates that a dominant as well as a consistent interictal focus is concordant with the ictal onset zone in the majority of patients. Therefore, in patients in whom the ictal onset zone can not be assessed with certainty, the identified dominant and consistent interictal focus may indicate the localization of the epileptogenic zone and epilepsy surgery may offer seizure freedom or seizure reduction in these patients.

P38. Epilepsy surgery for children with tuberous sclerosis complex and multifocal EEG findings

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Rationale. Tuberous sclerosis complex (TSC) is associated with medically refractory epilepsy and developmental delay in children. Some patients may have a single focus and may benefit from cortical resection, however, many children with TSC develop uncontrolled seizures in association with multiple epileptogenic tubers with poorly localizable/lateralizable VEEG findings.To identify patients who might benefit from surgery, and to maximize outcome, some subjects undergo bilateral implantation of subdural and/or depth electrodes. This study examines the utility of bilateral subdural electrode studies in medically refractory TSC.

Methods. A retrospective analysis of all 52 patients undergoing epilepsy surgery from 2000 to 2008 for TSC identified 20 patients with bilateral strip implantation. The preoperative surgical workup, intracranial EEG findings, surgical intervention and outcome was performed to identify the features which predict successful lateralization. Surgical outcomes for patients at least six months following resection were classified utilizing the Engel system.

Results. 14/20 patients undergoing bilateral strip surveys went on to have a focal resection. One is pending further resection. In 7/14 patients, multiple resections were undertaken. 5 patients had multifocal, non-resectable or nonlateralizable onsets and no further surgery was performed. Of the patients undergoing a resection, 7/14 have Engel Class I surgical outcomes with a median follow-up of 25 months (6 months-4 years).

Conclusion. This retrospective study illustrates the role of invasive monitoring in medically refractory TSC for patients with poorly lateralized interictal and ictal VEEG data. Despite non-lateralizable/non-localizable pre-operative evaluations, 74% of the patients studied invasively had focal (or multifocal) resectable regions and, of those, 50% are now seizure-free.

P39. Epilepsy surgery in tuberous sclerosis complex: successful circumscribed resection within the primary motor area without deficit

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A 2-year-old girl with clinical diagnosis of tuberous sclerosis complex (cortical tubers, subependymal nodules, facial angiofibromas; no mutation detected in TSC-1 or TSC-2) presented with atonic seizures followed by tonic elevation and cloni of her right arm. She was left-handed and active use of her right arm and leg was clearly reduced with additional postictal loss of function. Seizure frequency was up to 12/day. Cognitive development was in a normal range. Video-EEG-monitoring gave the following interictal results: Intermittent slowing left central, left parietal and right parietal region. Sharp waves and polyspikes left posterior central, and central with maximum left hemisphere, as well as rare spikes right frontocentral. Ictal EEG showed initial fast activity in the left central region. MRI showed bilateral cortical tubers with a large confluent hamartomatous area in the left lateral occipito-parietal region and a tuber in the left central region. Intraoperative assessment revealed a large area of cortical malformation including the anatomic pre- and postcentral region. Consistent with electroclinical preoperative assessment electrocorticography showed

circumscribed activity within this area, localized in the presumed precentral region. Guided by electrophysiological data and tactile evaluation the most solid part of this tuber within the hamartomatous area was resected. Postoperative examination showed transient loss of function in the right hand and arm. Six months after surgery the patient is seizure-free and shows functional improvement of her right hand and arm. Cognitive development is again in the normal range (with subtle hints to even better function than preoperatively). In conclusion, epilepsy surgery in patients with TSC can be successful even without complete resection of a hamartomatous area. Tubers within eloquent regions may be resected without loss of function.

P40. Cognitive outcome after temporal lobe surgery in childhood: a long-term follow-up study

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Purpose. Temporal lobectomy (TL) is a successful surgical treatment for patients with intractable temporal lobe epilepsy. However, little is known about long-term cognitive outcome after TL in children.

Methods. 41 TL patients were tested (24 left- and 17 right-sided surgery) of whom 16 had developmental tumors and 25 had hippocampal sclerosis). Mean age at surgery was 13.8 years. In addition we assessed a group of 11 patients with lesional focal epilepsy who underwent the same presurgical evaluation but who did not undergo surgery (non-TL group). Minimum follow-up time was 5 years in both groups. General cognitive function was measured using standardised tests of intelligence. Additionally, structural MRI data were acquired and used to extract total brain white and grey matter (GM) volumes.

Results. 86% of TL and 36% of non-TL patients were seizure free at follow-up. Surgical and non-surgical groups were well matched for initial full scale IQ (FSIQ). Most surgery patients showed an increase in FSIQ post-surgically, with the most marked increase in those with below average preoperative intellect. No change in FSIQ was found in the non-surgery group. An improvement of > 15 IQ points was seen in 10 TL patients and none of the non-TL group. Both left and right TL cases improved in non-verbal intelligence, while the left TL group also showed an improvement in verbal IQ. Change in FSIQ in TL patients was positively correlated with change in total brain grey matter volume, regardless of size of the resection.

Conclusion. Temporal lobectomy in childhood results in good seizure control and appears to facilitate intellectual and brain development.

P41. Neurological and psychological outcomes of children with cryptogenic localization related epilepsy

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Objectives. About one third of the children epilepsy population is diagnosed with cryptogenic localization related epilepsy (CLRE). Few studies were interested in the short and long term evolution of these patients and no conclusive study concerning the impact of CLRE on neuropsychological functioning is yet published. In this study we determined neurological and neuropsychological outcome of 6 children with CLRE who had been referred to our department of paediatric neurology and we tried to explore which epilepsy factors (seizure type, seizure frequency, age at onset, duration of epilepsy, and drug load) were influencing the cognitive profile of CLRE.

Patients and methods. For all patients we determined the clinical, therapeutic and evolution characteristics of seizures. All patients had at least one EEG and MRI. Seizure frequency was used as the clinical outcome variable and full scale IQ, psychomotor delay and educational delay were used as neuropsychological outcomes.

Results. Six patients (5 boys and 1 girl) with average age of 12 years (7 months-18 years). Psychomotor development and clinical examination were normal for all patients. The age of onset of epileptic seizures was ranged between 2 months and 8 years. All of the children had one type of seizures, frontal (3 cases) and temporal seizures (3 cases). All of them had a high seizure frequency (7 seizures per day in 2 cases, 3 seizures in 1 case and uncountable seizures per day for the others). MRI was normal for all patients. Two patients were seizures free with one AED and two needed association of AED with a low frequency of seizure. The mean full scale IQ was 61 (range 48-100). The average educational delay was approximately 1 school year related to memory disorders.

Conclusion. The knowledge about the prognosis of CLRE is poor. In general, CLRE has a non-predictable course; but have several type of seizures, a delay of diagnosis and treatment were predictive for poor outcome. The patients may benefit from early detection of CLRE and consequent epilepsy treatment including pedagogical and academic support.

P42. Neuropsychological outcome in preadolescent children after epilepsy surgery in the temporal lobe

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Rationale. Cognitive outcome after temporal lobe epilepsy surgery is relatively well known in adults but less data are available in preadolescent children. We studied postoperative cognitive outcome in 19 pediatric patients with temporal lobe epilepsy.

Methods. 19 children underwent surgery (ten left and nine right sided) in the temporal lobe at a mean age of ten years (range: 6.5-12.6): 13 standard lobectomy, three standard plus lesionectomy, one amygdalohippocampectomy, and two lesionectomy with preserved hippocampus. The mean age at seizure onset was 4.7 years (range: 0.3-9.8). The mean preoperative VIQ and PIQ were 83 (range: 41-113) and 82 (range: 36-130). 14 children (74%) were seizure free (Engel class I) at two years postoperatively. All children were tested preoperatively and at two years after surgery using WISC-III, NEPSY and VMI. Postoperative changes were analysed controlling for duration of epilepsy.

Results. Two significant statistical interactions were observed. VIQ scores improved significantly in seizure-free children compared to poor seizure outcome regardless of side of surgery. Verbal memory scores improved after right-sided surgery while memory decline was observed in the left sided group. There were no significant changes in PIQ, visual memory or linguistic, visuo-spatial, executive or sensorimotor functions.

Conclusion. Children who become seizure-free show postoperative improvement in verbal intelligence compared to those whose seizures persist. Similar to adults, left sided surgery is associated with decline and right sided with improvement in verbal memory scores.

P43. Long-term cognitive outcome of children with drug resistant temporal lobe epilepsy after surgical intervention

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Purpose. To study the long term neuropsychological performance of children and adolescents with drug-resistant temporal lobe epilepsy (TLE) after epilepsy surgery.

Methods. Intellectual functioning, concentration, flexibility in thinking, verbal and non-verbal memory were assessed three months after surgery, and then once a year. To determine group differences, data were analyzed separately for left TLE (LTLE) and right TLE (RTLE) patients. Individual changes in intellectual functioning and verbal memory, as well as predictors for postoperative outcome were also evaluated.

Results. 40 patients aged 6-18 years (21 RTLE, 19 LTLE) were examined. The long term data analysis ranged from 2 years to 10 years postoperatively. Long-term neuropsycholgical performance revealed improvement in intellectual functioning, concentration, and verbal and nonverbal memory in both TLE groups. Tendencies indicate a slight decrease for left TLEs in verbal memory performance shortly after epilepsy surgery, follow up study showed that they catch up and improve steadily. Analysis of individual changes revealed a significant improvement especially in verbal learning capacity and attention. Predictors for cognitive outcome were age at seizure onset and slightly duration of epilepsy. Long-term data showed seizure freeness in about 85% of our children.

Conclusion. Children and adolescents with drug-resistant TLE show very good long-term cognitive development after epilpsy surgery. Early intervention therefore seems to be advisable and supposes a better base for rehabilitation.

P44. Mapping language cortex in children with focal epilepsy using intracranial event-related potentials

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Electrical cortical stimulation (ECS) is the standard technique for identifying eloquent cortex during intracranial EEG recording but in children is often restricted by poor cooperation. We explored if event-related potentials (ERP) to language stimuli could be recorded reliably from subdural electrode grids in patients undergoing invasive extra-operative EEG monitoring for epilepsy surgery.

ERP stimulation included auditory presentation of common nouns (< 600 ms duration, presented every 3-5 s, blocks of 100 stimuli each). Children were asked to either repeat words aloud or generate verbs, in separate blocks. For posterior temporal grid locations stimulation included visual word presentation (reading) and naming of objects. During separate runs stimuli were presented without overt response (passive control condition). Additional recordings included AEP (pure tones and syllables) and SEP.

Reproducible ERPs were recorded in 18/22 children (10 left implants and 8 right, age range 3-18 years) despite the presence of interictal EEG changes.

Language ERP were distinctly different from short-latency potentials to tones and syllables and were recorded focally in the inferior and middle frontal and precentral gyri, as well as from superior temporal and inferior parietal cortex. Language ERP consisted of a sharp negative wave at 300ms followed by a slow positive-negative wave complex (latency of 400 and 1 000 ms, respectively), peak amplitudes ranged from 20-100 ms. ERP amplitudes were larger over the left hemisphere. Language fMRI and ECS and ERP co-localised in a small number of cases.

We conclude that intracranial language ERP can be recorded in children and may provide corroborating evidence about the location of eloquent cortex.

P45. Language lateralisation and reorganisation in early-onset focal epilepsy: evidence from fMRI

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Purpose. We investigated language lateralization using functional MRI (fMRI) in children suffering from left-sided, medically refractory, childhood-onset focal epilepsy and determined factors associated with atypical lateralization.

Methods. Twenty-four children suffering from drug-resistant, left-sided, early-onset epilepsy underwent fMRI-scanning using a covert verb-generation task. Twenty-nine healthy volunteers, matched for age and gender, served as controls. Images were analyzed using SPM5 and lateralization indices (LI) were calculated within regions of interest (Broca's area, temporal lobe, cerebellum) using the LI-toolbox (Wilke *et al. J Neurosci Methods* 2007; 163: 128-36). Typical language lateralization (left-sided) was defined as LI > 0.2 and atypical language (right-sided or bilateral) as LI < 0.2. Factors contributing to atypical language distribution were investigated including: age at seizure-onset, seizure frequency, handedness, lesion location, lesion size, size and asymmetry of the planum temporale.

Results. There was no statistically significant correlation between age at seizure-onset, seizure frequency, handedness, lesion location or lesion size and presence of an atypical language lateralization ($p > 0.05$). However, the correlation between PT-asymmetry and the fMRI language lateralization index was highly significant ($r = 0.823$, $p < 0.001$). A stepwise linear regression analysis including all variables revealed that asymmetry of the planum temporale was the only significant predictor for language lateralisation, accounting for 50% of the variance (Beta = 0.723, $p < 0.001$).

Conclusion. In the presence of left-sided pathology the asymmetry of the planum temporale appears to indicate the efficacy of language reorganisation to the right hemisphere.

P46. Quality of life outcome in children after temporal lobectomy-longterm follow-up

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88 patients underwent temporal lobectomy, with or without extratemporal procedures, between 1992 and 2008 at the Comprehensive Epilepsy Center at the University of Texas/Memorial Hermann Hospital. Here we present long-term follow-up of these patients, including quality of life measures as well as seizure outcome. Methods include telephone interviews with patient families, with a standardized questionnaire with questions regarding seizure control, social and cognitive functioning, independence, and emotional status.

Results. With a post-operative follow-up period of between 1 and 16 years, all patient families interviewed report an improvement in overall quality of life, with most significant and consistent improvements seen in social functioning, independence and energy/activity level.

Conclusion. Temporal lobectomy in children results in long-term improved quality of life in the majority of patients, as well as improved seizure control.

P47. Influence of IQ/DQ on outcome in epilepsy surgery of children

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Purpose. Operations of children with intellectual disabilities have been questioned by some because of suspicion of a poor outcome.

Material and methods. Retrospective study of all Danish children (n = 65) operated between 1996 and 2007. 25 children had an IQ/DQ above 70 (group A) and 40 had IQ/DQ of 70 or below (group B). Mean age at operation in group A were 13 years and in group B 9 years and 1 month. In group A the majority (17) were operated in Denmark, and in group B the majority (16) were operated abroad mainly Cleveland Clinic, USA. In group A, 4 had hemispherectomy or multiple lobe resection whereas in group B 19 children had this kind of operation. Results: Seizure free outcome (Engel 1 A or B) were found in 68.0% (group A) and 67.5%

(group B) at one year follow-up. In group A 27.3% had an increase and 18.2% a decrease of 10 IQ/DQ or more compared to 10.0% and 14.0% in group B one year after the operation. Behavioural changes (mood,agression) were registered by the parents at a visit 6-8 weeks after the operation. In group A, 45% of the parents reported an improved situation, compared to 34% in group B. Only 10% of the parents in group A reported an improved "attention and energy" compared to 47% in group B.

Conclusion. Seizures and developmental outcome after epilepsy surgery did not differ in a Danish cohorte comparing children with intellectual disabilities with normal children.

P48. A dichotic listening to assess modifications of hemispheric asymmetry in partial rolandic epilepsy

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We assessed the impact of unilateral foci in benign idiopathic partial epilepsy with rolandic discharges (BECT), on performance and hemispheric specialization in lateralized cognitive functions. Of first data suggest that the profiles of dysfunction depend on the lateralization of focus (Bedoin *et al.* 2006). Children with left-sided focus (BECTL), children with right-sided focus (BECT-R), and control children were tested in dichotic listening. We adapted this experiment to the children and we establish standards by age. Two different auditory stimuli are presented to the participant simultaneously, one to each ear, and the task is to call back the heard word. For right-handed adults, but also children in a weaker measure, words sent to the right ear are more often reminded (REA: Right Ear Advantage), what testifies of a left hemispherical dominance to treat the word. The originality of our test is to show that this REA is stronger for the treatment of the phonetic feature of place than the feature of voicing. Our hypothesis is that REA of patients is only reduced in case of BECT-L, and particularly for the treatment of the place. On the other hand, for the voicing, both hemispheres can participate effectively in the treatment, and the lateralization of focus should not infer particular effect. The first data consolidate this hypothesis and suit to previous data obtained with a test of reading in divided field. Subclinical epileptic discharges thus modify the functional hemispherical asymmetry, in particular for language, in a different way according to the lateralization of the focus.

P49. Two years outcomes of a multicenter cohort of non-surgical and post-surgical children with intractable epilepsy: Impact on family functioning and quality of life

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Objective. To date, no studies have assessed the impact of epilepsy surgery on the child development or social adaptation, family functioning or parental well-being. In a multicenter cohort of children with intractable epilepsy, candidates for resective surgery, we assessed these aspects according to the clinical outcomes.

Population and methods. Children with partial medically intractable epilepsy, potentially operable, were followed every 6 months over two years. The clinical outcome was the proportion of seizure-free patients (1a-1b Engel's classification). Socio-economic data and the impact of epilepsy on quality of life (QoL) of child and family (Impact of Childhood Illness, ICI Scale, Hoare, 1994) were also measured.

Results. 87 children with a mean age of 9 years (range 0.1-16) were included from 9 tertiary centers (40 operated, 40 medically treated, 2 not eligible, 5 lost of follow-up). Median duration of epilepsy is 6 years (SE = 4.0). Epilepsy is symptomatic in 75% of cases. At one year, seizure-free patients are 78% (n = 30/40) in the operated children and 21% (n = 8/40) in medically treated ($p < 0.001$); at two years, they are respectively 78% (n = 25/37 whose 60% without treatment) and 32% (n = 9/33, 0% without treatment). In the medical group, there are no significant variations of ICI scores. In the surgical group, at 2 years, significant improvement are observed for Impact of epilepsy and its treatment ($p = 0.045$), Impact on the family ($p = 0.04$) and total score ($p = 0.06$) but not for impact on child's development and adjustment.

Conclusion. The good results of surgery epilepsy have an impact on specific QoL domains and family functioning.

P50. Pediatric epilepsy surgical series of primarily ECoG-guided resections

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We reviewed our experience with our first 100 patients with refractory epilepsy treated with epilepsy surgery.

For outcome data we restricted our analysis to those with more than one year of follow-up (n = 87). Few of our patients (n = 11) have had the use of chronic implanted electrode recordings (invasive). The majority (n = 76) underwent a single surgery guided by intraoperative electrocorticography (ECoG). Outcome at last follow-up was rated using the Engel scale. A favorable outcome was defined as Engel Class I or II. Of those who underwent invasive monitoring 55% had a favorable outcome, whereas 87% had a favorable outcome with ECoG-guided resections ($p < 0.02$). When we compared those with favorable outcome to those without in the ECoG group we found that shorter duration of epilepsy (3.4 *versus* 6.5 years) and higher full scale intelligence quotient (91.6 *versus* 74.8) were associated with a better outcome. Otherwise, outcome did not appear to differ strongly based upon location of surgery or pathology between the two groups. Patient selection is the most obvious explanation for the differences between the invasive and the ECoG-guided resection groups, but the favorable outcome for many patients in the ECoG group is encouraging and furthermore, it appears that earlier operation in children with higher IQs might be particularly predictive of a favorable outcome. These observation need to be substantiated in a prospective multi-institutional study, but they may have implications for the selection of optimal candidates for pediatric epilepsy surgery.

P51. Seizure outcome after hemispherectomies in swedish children operated 1990-2004

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Purpose. To describe the results from the two year follow-up after hemispherectomies in children with pharmacoresistant epilepsy, on a national level in Sweden.

Methods. Data from the Swedish National Epilepsy Surgery register 1990-2004 were analysed. The register includes data from all surgical procedures within the epilepsy surgery programme in Sweden and is prospective since 1995.

Results. During the period, 616 procedures were performed, and all had a two-year follow-up. Forty-two were hemispherectomies, 37 of which were performed in children < 19 years, 18 complete and 19 partial. Epilepsy onset was at 1.6 years of age (median 6 months), age at surgery was 5.7 years (median 4.9 years) and the duration of epilepsy was 4.1 years (median 3.4 years). Seizure freedom was achieved in 76% with complete hemispherectomy and in 21 % with partial hemispherectomy. Another 11% and 37%, respectively, had more than 75% reduction of seizure frequency.

Conclusion. In the Swedish series more than 75% of the children who underwent complete hemispherectomy became seizure-free. Subtotal hemispherectomy was a much less successful procedure and should be preceded by a careful risk/benefit analysis whether a complete hemispherectomy might not be considered a better alternative.

P52. Outcome of temporal lobectomy in childhood

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Introduction. Surgery is an effective treatment for medically resistant temporal lobe epilepsy. Few reports, however, have defined cognitive and seizure outcome in children. This study aimed to evaluate post operative seizure and cognitive outcome related to preoperative variables.

Methods. Data from children who underwent temporal lobectomy between 1992 and 2006 were reviewed: Age at onset of habitual seizures, duration of epilepsy, presence of secondary generalizations, status epilepticus, febrile seizures, EEG focus, preoperative seizure frequency, pathology, side of lesion, and preoperative IQs were recorded. Postoperative seizure outcome was documented at 1 and 3 years along with postoperative IQ scores.

Results. 109 children were included (61 left). Means (in months) for age at onset of habitual seizures and average duration of epilepsy prior to surgery were 46.24 (0.3 to 168), and 90.2 (8.2 to 204.6), respectively. Pathology revealed hippocampal sclerosis (46), tumors (41), tuberous sclerosis (3), cortical dysplasia (8), Rasmussen's encephalitis (1), Sturge-Weber syndrome (2), nonspecific gliosis (1) and no abnormality (2). A history of febrile seizures was present in 21 cases, secondary generalization in 38 (39.6%) and status epilepticus in 25 (26%). EEG was localizing in 85 (86.7%). 60/94 (63.8%) were seizure free at 12 m and 41/65 (63.1%) at 36 m. IQ scores were available in 80 preoperatively (mean FSIQ 79, VIQ 81, PIQ 85) and 74 postoperatively (mean FSIQ 77, VIQ 77, PIQ 82). Preoperative predictors of outcome were analyzed with multiple regressions.

Conclusion. Temporal lobe resection is a safe and effective procedure for children with epilepsy.

P53. Outcome of epilepsy surgery in children evaluated with non-invasive protocol

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Objective. To assess outcome of epilepsy surgery in children with medically refractory epilepsy evaluated with noninvasive protocol.

Methods. Retrospective analysis of ictal VEEG, MRI, SPECT, neuropsychology and pathology data was performed in 45 children (< 16 years) with refractory epilepsy who underwent surgery after presurgical evaluation with non-invasive protocol, and had at least one year post-surgery follow-up. WADA, depth/grid monitoring was not done. Outcome was assessed according to Engel's outcome classification.

Results. Mean follow-up was 28 (12-58) months; 26 (58%) were males; 23 (51%) were < 12 yrs. Mean age at surgery was 11.2(3-16) years. Intaoperative electocorticography was used in 6. Surgery was AH-ATL in 25 (55.6%), lesionectomy-8 (17.7%), functional hemispherectomy-6 (15.6%), resection of hypothalamic hamartoma (HH)-4 (8.8%) and VNS implantation in 2 patients. Pathology revealed hippocampal sclerosis (HS) in 16, tumour lesions-6, hamartoma-4, gliosis-6, cortical dysplasia-3 and dual pathology in 2 patients. None had major post-surgery complications and 4 had minor (visual field) deficits. The total cost was Rs. 60 000/-(1 200 US $). At follow-up 27 (60%) were in class-I, 5 (11%) class-II and 13 (29%) in class-III. Favourable outcome was noted in 73% with TLE-HS group, 83% with functional hemispherectomy, 80% with lesionectomies and 50% with HH. The predictors of favourable outcome (OR [95% CI]) were lesion on MRI 10 (1.056-94.67), typical ictal SPECT perfusion pattern 1.275 (1.851-1.912), a concordant ictal EEG 1.6 (1.285-8.984) and for unfavourable outcome were acute post-operative seizures 10(1.801-55.526) and secondarily generalized seizures 3.4 (1.971-11.905).

Conclusion. Favourable outcome after epilepsy surgery can be obtained for patients with TLE-HS and lesion related epilepsies in developing countries with limited resources, if careful presurgical evaluation is planned.

P54. Seizure outcome following resection of epileptogenic lesions in swedish children operated between 1990 and 2004

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Purpose. To describe seizure outcome following resection of epileptogenic lesions in children with pharmaco-resistant epilepsy on a national level in Sweden.

Methods. Data from the Swedish National Epilepsy Surgery register from 1990-2004 were analysed. The register includes data from all surgical procedures within the epilepsy surgery programs in Sweden and is prospective from 1995. Children were defined as less than 19 years old at surgery. The pathoanatomical diagnoses considered as lesions for this purpose were gangliogliomas, DNET, low-grade astrocytomas and cavernomas.

Results. During the period 1990-2004 616 procedures were performed in Sweden with a two-year follow-up. Of these 152 procedures concerned epileptogenic lesions as above and of these 52 were in children. Epilepsy onset was at 7,6 years of age (median 8 year), age at surgery was 12,9 years (median 13,6 years) and epilepsy duration was 5,3 years (median 3,5 years). There were 34 gangliogliomas or DNET, 17 low-grade astrocytomas and one cavernoma. 79 % of the patients became seizurefree (astrocytomas 65%, gangliogliomas and DNET 85%), including the patient with a cavernoma.

Conclusion. Resection of epileptogenic lesions in children with pharmaco resistant epilepsy results in a high proportion of seizure freedom, as expected with better results for gangliogliomas and DNET than for astrocytoma.

P55. Neurologic evolution in pediatric patients with simple craniosynostosis following surgery

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Objective. Evaluate the neurodevelopment in pediatric patients follow surgery for simple craniosynostosis in a mexican population.

Methods. Since year 2000 we evaluated 96 children operated on for simple craneosynostosis in the Hospital Infantil de Mexico. Six months following surgery all patients were submitted to clinical and neurodevelopment evaluation and EEG.

Results. 66 males and 30 females were evaluated. Age at surgery was 14 months. The most frequent form of craniosynostosis was scaphocephalia in 35% of the patients, followed by trigonocephaly and brachicephaly. Only 5 patients had an abnormal EEG characterized by inter-hemispheric asymmetry, all of them with an abnormal neurodevelopment. Almost all patients with surgery after 13 months showed some delay.

Conclusions. The early surgical correction in a simple craneosynostosis (before 12 months of age) is correlated with a better neurodevelopment evolution.

P56. Surgical management of patients with pediatric onset epilepsies from Slovenia

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Purpose. Two million population is too small for an epilepsy surgery program. Non-invasive evaluation is performed by our team to select candidates for pre-surgical evaluation and surgery abroad (12 surgery centers).

Methods. Retrospective analysis of referrals and outcomes (1985 to 2008).

Results. 43 patients were referred abroad: 14 were declined, 27 had surgery, 2 wait phase II. 15 patients wait for non-invasive evaluation. Surgeries: temporal (14/27), frontal (8/27), parietal lobes (1/27), 3 callosotomies, 1 hypothalamic hamartoma surgery. FCD was the most common, followed by lowgrade tumors, gliosis, postencephalitic changes, hippocampal sclerosis. Seizure outcome: 17 Engel I after 1y, 12 after f-up (min 2,5 y, mean 4,5 y). 6 patients switched from Engel I to Engel II. Worst Engel III-IV: 2 FCD without complete resection and hypothalamic hamartoma. Callosotomies: 3 benefits: Engel II, II-III, III-IV. Complications: 8 of 23, transient 7, persistent mild hemiparesis 1. Psychological outcome: no adverse influence on intelligence. Some cases: improved performance IQ: in younger with higher intelligence. Attention might improve after temporal, not after frontal surgery.

Conclusion. High proportion of declined patients reflects severe pediatric epilepsy despite pre-selection by our Center. Positive trends: total number, earlier referrals. Center selection depends on pathology, patients' age, logistics (coordination, planning professional work abroad, family support, accompaniment). Rehabilitation after surgery is needed for better quality of life and social integration.

P57. To be or not to be an orphan: views and proposals from a small country on managing epilepsy accross borders

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Background. Managing surgically amenable epilepsies in a small country - Slovenia (2 million inhb) due to small population without a quartary center - requires networking models to manage epilepsy accross borders, if not accepting to put patients in a position of a "double orphan" disorder: multiplying the effect of relative rarity with absolute small demography.

Methods. Retrospective analysis of managing pts with severe pediatric onset epilepsies referred to 12 foreign epilepsy centres.

Results. Evaluation of the practice over 25 years based on 43 patients' referral data confirms the need to re-assess the situation for efficient use of existing and novel services in international networking with the aim of: sharing roles in international collaboration, definition of standardized steps at national, regional, relay and transEU centres, agreed protocols, improved referrals, second opinion services, management of complications and legal issues related to crossborder care, discussion of additional safety risks related to mobility in patients with language and culture - sensitive comorbidities. Other suggested modalities with proven value: international expert visitors, real life clinical teaching seizure conferences, e-medicine, teleconferencing. Other issues to discuss: shared learning, research and development of epileptology at the referring site. Needed attention to administrative, financial agreements; linguistic aspects and practical patient-oriented guidelines. Activity to be recognized at the referring and receiving institutions.

Conclusion. The importance of the above issues require collaboration: a questionnaire shall be distributed, suggestions welcome to: <epilepsija@epilepsija.org>.