

Pediatric Epilepsy Surgery

Posters and Video-Posters presented at the 5th International Epilepsy Colloquium, Lyon, France, May 2012

1. Epilepsy surgery in children with difficult-to-treat epilepsy in the southern region of Sweden: a long-term follow-up study

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Introduction. Resective epilepsy surgery in early childhood has become an important treatment option for selected infants and children with epilepsy. We describe experience and clinical outcomes in all children who underwent resective surgery for epilepsy in the southern region of Sweden. The study is a long-term follow-up of all children who underwent epilepsy surgery at the University Hospital in Lund from 1991 to 2007.

Methods. All children under 18 years of age who had resective surgery as treatment for epilepsy within the southern region of Sweden, as part of the Lund University Hospital epilepsy surgery program between the beginning of 1991 and 2007, were assessed for cognitive function, mental health, psychological, occupational and social function, QOL and expectations. Follow-up was between 5 and 21 years. Review of medical documents for demographic data and seizure-related characteristics such as aetiology, postoperative seizure frequency, pre- and postoperative cognitive function, long-term complications and repeat surgery rates were analysed by retrospective examination of the clinical records.

Results. Forty-seven children were included in our cohort with a median age at surgery of 8 years (range: 0.5-18.16 years). Surgical procedures comprised 15 lesionectomies, 16 lobar resections, 6 multilobar resections, 6 functional hemispherectomies, 2

callosotomies, 1 disconnection, and one subpial transection. Nineteen of 47 (40%) children underwent repeat surgery. Three of these were disconnections of hypothalamus hamartomas. Twenty-three of 47 (50%) children achieved seizure freedom. Six of 47 (13%) demonstrated more than 75% improvement in seizure frequency, 7/47 (15%) 50-75% improvement in seizure frequency and no children had an increase in seizure frequency. Postoperative complications included subsequent shunt procedure in 1/7 (14%) children undergoing hemispherectomy. One child had a preoperative haemorrhage without any sequelae and one an infraction with subsequent mild paresis. Two children died during follow-up. One died from SUDEP and one while having an epileptic seizure. **Conclusion.** Epilepsy surgery in children offers suitable candidates a good chance of significantly improved seizure outcome even after long-term follow-up when initial assessment is negative. At the individual patient level, computer-aided SPM analysis could serve as an additional observer and help to distinguish small or multiple hypometabolic areas. We advise to use more than one assessment method when performing a FDG-PET scan in these types of difficult patient cases since re-evaluation has the potential to increase eligibility for epilepsy surgery.

2. Clinical characteristics and outcome of surgery for temporal lobe epilepsy in children and adolescents

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Introduction. Although temporal lobe epilepsy (TLE) due to hippocampal sclerosis (HS) is the commonest cause of medically intractable epilepsy in adults and adolescents, in children TLE is responsible for less than 30% of the surgical resections.

Aim. To assess the clinical characteristics and postsurgical outcomes in children and adolescents with TLE.

Methods. Retrospective analysis of clinical characteristics, ictal VEEG, MRI, neuropsychology and pathology data was performed in 80 children and adolescents with refractory TLE who underwent surgery, and had at least one year postsurgical follow-up. Outcome was assessed according to Engel's outcome classification.

Results. There were 24 children (2-12 years) and 56 adolescents; 47 were males. The age at onset of epilepsy ranged from 4 months to 15 years and the duration of epilepsy was 1-15 (mean: 9.21) years. At the last follow-up visit, 18 (75%) children and 39 (69.6%) adolescents were seizure-free. Auras were more common in adolescents, while developmental delay, below normal IQ and secondary generalised and multiple seizures were more frequent in children ($p<0.05$). Interictal-EEG showed multifocal and generalised discharges significantly more frequent in children than adolescents ($p<0.001$). Hippocampal sclerosis was the commonest pathology, noted in 76.8% of adolescents and 62.5% children. Isolated focal cortical dysplasia was more frequent in children (21%) than adolescents (12.5%). Dysplasia was more common in children <10 years (38.4%). Acute postoperative seizures and non-lateralising ictal EEG findings were predictors of unfavourable outcome.

Conclusion. Children with TLE have distinct clinical EEG and pathological features when compared to adults. Favourable outcome can be obtained in children and adolescents after surgery for TLE.

3. Temporal and frontal lobe seizures in children who became seizure-free after surgery

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Aim. To compare semiology of seizures originating from the frontal lobe (FLS) and the temporal lobe (TLS) in children who became seizure-free after surgery.

Methods. We analysed 28 videotaped TLS from 11 patients and 25 videotaped FLS from 10 patients aged 3.5-16 years (mean: 8.1 years). For each patient, 2-3 seizures were assessed. Median follow-up was 18 months (range: 5-40 months). Lüders' semiological classification (1998) was used for seizure analysis.

Results. Simple motor seizures occurred exclusively in FLS (8 seizures from 4 patients) and frequently were observed in clusters (three patients). The mean seizure duration was longer for TLS than FLS ($p<0.05$), but did not differ significantly when complex motor seizures were compared. Version was always contralateral in

FLS (7 seizures of 3 patients) and had no lateralising value in TLS. Contralateral arm dystonic posture occurred earlier in the FLS than in TLS, whereas oral automatisms were observed later in FLS than in TLS. Association of contralateral arm dystonic posture with ipsilateral distal hand automatisms were seen only in the TLS (13 seizures of 5 patients), whereas for FLS, distal hand automatisms were usually bimanual (9 seizures of 3 patients) ($p<0.004$). Proximal automatisms were more frequent in the FLS (14 seizures of 5 patients) than in TLS (five seizures of 2 patients) ($p<0.001$) and hypomotor behaviour was observed more often in the TLS ($p<0.05$) group.

Conclusion. Semiological features of temporal and frontal lobe seizures differ in lateralising significance as well as in time of onset.

4. A 15-year follow-up of first unprovoked seizure: prospective study of 200 children

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Introduction. Epilepsy is defined as a tendency to have recurring unprovoked seizures and is associated with an extended spectrum of learning difficulties, behaviour and psychiatric problems. Prospective, population-based studies demonstrate the existence of these comorbidities prior to diagnosis of epilepsy and its persistence beyond the control of seizures. The purpose of this study was to investigate the natural history of children with first unprovoked seizures; characterising and comparing between those that develop epilepsy and those with a single seizure. We also wanted to identify significant prognostic factors for the development of epilepsy.

Methods. We prospectively studied 200 children under the age of 11 years who were admitted as a hospital emergency with a first unprovoked seizure. Neonatal seizures, myoclonus, absences, infantile spasms, and epileptic encephalopathy were excluded. Demographic variables, personal and family history, neurological examination, and electroencephalographic (EEG), psychiatric, cognitive, educational, and social profiles were analysed. We characterised the group that developed epilepsy (group 1), considering: time to relapse, remission rate, duration of epilepsy, neuroimaging, aetiology, epileptic syndrome, therapeutic regimen, and compared these with the single seizure group (group 2), 2 and 15 years after the first seizure. The data were statistically analysed using SPSS 20.

Results. Thirty percent of children who had a first unprovoked seizure developed epilepsy, the majority at school age. Forty-eight relapsed within the first three

months and 93% in the first year. Partial seizure type was a statistically significant variable for the development of epilepsy ($p=0.000$). Normal EEG after the first seizure was a predicting factor for early good prognosis ($p=0.000$). There was no statistically significant difference in cognition between the two groups. History of febrile seizures, problems in the neonatal period, family history of epilepsy, and neurological examination had no influence on recurrence rate. Benign childhood epilepsy with centrotemporal spikes was the main epileptic syndrome recognised in group 1 (30%). Currently, 77% of patients with epilepsy are seizure-free and are not taking therapy. Epilepsy remains active in 12 children (20%). Remission rate at 5 years was 59%. Fifteen years later, we observed a large number of psychiatric and academic comorbidities; the group with epilepsy had a 2.6-greater risk of comorbidities than the group without epilepsy ($p=0.011$). Group 1 had a 2.2-greater risk of requiring school support than the group without epilepsy ($p=0.036$). Antiepileptic or psychiatric drugs were used 4.7 times more in group 1 ($p=0.000$). The mortality rate in our study was 0.67% per year.

Conclusion. In the studied group, the long-term prognosis of epilepsy was favourable. Nonetheless, epilepsy remains active in 20% and is intractable in 11.5%. We found a large percentage of psychiatric and social comorbidities in a population with benign epilepsy and the persistence of these comorbidities well after the control/cure of epilepsy.

5. Unexpected marked improvement in epilepsy surgery candidates

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Introduction. Epilepsy surgery is performed based on the assumption that ongoing seizures will continue. Although this may be the case before surgery is undertaken, we set out to estimate how often this occurs during the period of epilepsy surgery evaluation.

Material and Methods. We retrospectively studied 173 children with medically refractory epilepsy referred for possible epilepsy surgery between January 1996 and December 2010 at the Dianalund Epilepsy Center. All patients were enrolled in the evaluation program based on one week of video telemetry, imaging studies, neuropsychological examination, and examination by a child psychiatrist. Because of waiting lists, this program usually lasted between 3 and 12 months, also providing the possibility for parents to consider epilepsy surgery or not. The same team (epileptolo-

gist/nurse) was in charge of the program during the observation period.

Results. Eight females and five males, with a mean age of 8.7 years (range: 4.3-14.6 years), withdrew from the epilepsy surgery programme during evaluation. All had marked improvement for at least one year (seizure-free or rare seizures) and none returned to the programme. Aetiology included: polymicrogyri (two patients), mesial temporal sclerosis (two patients), tuberous sclerosis (one patient), hypothyreosis of Hashimoto (one patient), infarct (one patient), cortical dysplasia (one patient), and five unknown with normal MRI. All children had tried between two and eight different relevant antiepileptic drugs when referred. Evaluation of possible reasons for seizure improvement revealed that improvement coincided with a febrile episode in four cases. In six cases, patients had a change in antiepileptic drug treatment and in five cases no reason was identified.

Conclusion. We found that 7.5% of children enrolled in an evaluation program for epilepsy surgery unexpectedly became seizure-free or had only rare seizures for more than one year, and never returned to the program.

6. Do children with severe impairments benefit from epilepsy surgery? A two-year follow-up after surgery

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The aim of this study was to assess seizure outcome two years after epilepsy surgery in a consecutive series of paediatric patients, with special focus on children with learning disabilities and other neuroimpairments, in addition to epilepsy.

Methods. Outcome two years after surgery was assessed in 110 of 125 children operated upon for drug-resistant epilepsy in Gothenburg in 1987-2006. The children were included consecutively and the surgical procedures included resective and disconnective surgery. In the 110 children followed, there were 33 temporal resections, 23 frontal resections, 12 parietal or occipital resections, 12 multilobe resections and 12 hemispherectomies. Callosotomy was performed in 14 children, hamartoma disconnection in two and multiple subpial transections in two children.

Results. More than half (59/110; 54%) of the children had learning disabilities (IQ<70), 43% motor impairments and 30% a neuropsychiatric diagnosis. Fifty-six per cent of those with an IQ<70 became seizure-free or had a >75% reduction in seizure frequency. Of those children undergoing resective surgery, 66% became seizure-free. The corresponding figure for those with more than 100 seizures per month was 48% (15/31), and 22% (7/31) had a 50-75% reduction in seizure frequency.

Conclusion. Learning disability, motor impairment and psychiatric morbidity should not be contraindications for paediatric epilepsy surgery. More than half of the children with learning disabilities had a worthwhile seizure outcome, with even better results after resective surgery. Children with drug-resistant epilepsy and additional severe neurological impairments should have the benefit of referral to a tertiary centre for evaluation for epilepsy surgery.

7. Temporo-parieto-occipital and parieto-occipital disconnection in patients with intractable epilepsy

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Aim. Temporo-parieto-occipital and parieto-occipital disconnection is described mainly in patients with diffuse posterior quadrant lesions (parietal, occipital and/or temporal lobes) and concordant electroencephalography (EEG) findings. The authors report on 16 intractable epilepsy cases with diffuse posterior quadrant lesions being treated with surgical disconnection, including 1 with tuberous sclerosis, 2 with pachygyria, and 13 with lesions caused by trauma or hypoxic ischaemic brain damage.

Methods. The authors conducted a retrospective review of clinical, neuropsychological, EEG, imaging, and histopathological data of 16 patients with intractable epilepsy who underwent temporo-parieto-occipital and parieto-occipital disconnection at their epilepsy centre between April 2004 and February 2011. Of the 16 cases, 12 were males and 4 were females. The age of seizure onset was from 0.1 to 27 years (average: 6.6) and disease duration of 0.1 to 18 years (average: 7.5). The surgery was performed between the age of 3 and 37 years (average: 14.1). Six patients underwent temporo-parieto-occipital disconnection, 5 patients parieto-occipital disconnection and 1 patient parieto-occipital disconnection and temporal lobectomy.

Results. After a mean follow-up of 2.8 years (range: 1-7.4 years), 13 patients (81%) were seizure-free, 1 patient had Engel Class II seizure outcome and one patient had Engel class III outcome. In the group of patients with posterior quadrant surgical disconnection, 1 patient developed hydrocephalus postoperatively and was

successfully treated by ventriculoperitoneal shunt. One patient exhibited severe brain swelling and a second resective operation was performed. One patient with postoperative acute intracranial haemorrhage underwent repeat surgery immediately and recovered well. None of the patients developed new motor deficits postoperatively and no death was reported in the study.

Conclusion. Temporo-parieto-occipital and parieto-occipital disconnection is a safe and effective surgical procedure for epilepsy patients with epileptic zone localisation to the posterior quadrant on one side. The results of surgical disconnection for posterior quadrant epilepsy have yielded excellent seizure outcomes in 81% of the patients in the series with no mortality or major morbidity.

8. Surgical treatment of patients with Rasmussen's encephalitis

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Objective. To describe the clinical, electrophysiological, neuroradiological, and histological findings of patients with Rasmussen's encephalitis (RE) and to evaluate the outcome of their surgical treatment.

Methods. This report was based on a consecutive series of 34 patients affected by RE studied and operated on at the Department of Functional Neurosurgery, Sanbo Brain Hospital from April 2004 to December 2010. The age at surgery was from 2.8 years to 17 years. Thirty-four RE patients were confirmed by presurgical evaluation including semiology, magnetic resonance imaging (MRI), interictal/ictal scalp video-electroencephalography (VEEG), intracranial recording, and biopsy. Presurgical evaluation modalities and surgical treatment were analysed retrospectively.

Results. Nineteen functional hemispherectomy (FH), nine anatomical hemispherectomy (AH), and six hemispherotomy procedures were undertaken. According to Engel's criteria, 28 patients (83%) achieved an Engel Class I status, 3 patients (9%) had an Engel Class II outcome, 2 patients (6%) had a significant decrease in seizure frequency (Engel Class III), and 1 patient (3%) had no change in seizure frequency (Engel Class IV). One patient showed contralateral seizure after AH and was diagnosed with bilateral RE. All of the patients except one case of bilateral RE had an increase in cognitive abilities, behaviour, and quality of life after the surgery. After the surgery, most of the patients could walk independently, but the fine movement of the hands were lost. The main early complications were fever (35%) after hemispheric surgery. Postoperative

hydrocephalus was observed one and a half years after AH in one patient. No death was reported in the series. **Conclusion.** Hemispherectomy and hemispherotomy were both confirmed as beneficial procedures in controlling seizures and improving quality of life in cases with RE.

9. Epilepsy surgery in infants and young children with tuberous sclerosis: 5 cases

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Introduction. Tuberous sclerosis complex (TSC) is often associated with early devastating and medically refractory epilepsy. Delineating the epileptogenic zone can be difficult in young patients with multiple tubers and multiple interictal EEG foci. We report five cases of early surgery in young children with TSC aged between 2 months and 4 years.

Patients and methods. Seventy-five children were followed before and after epilepsy surgery, between 1995 and 2010. Their charts were collected retrospectively (clinical and cognitive evolution, EEG, radiology, and anatomo-pathology). Among them, 5 young children with TSC were operated on in Strasbourg University Hospital, between 2004 and 2010.

Results. Presurgical evaluation included: repeated long-duration video EEG, MRI, CT, PET, psychological evaluation, and SEEG in 2/5 cases. The resection of a large cortical dysplasia was performed in two infants (2 months and 4 months of age) treated for neonatal focal drug-resistant epilepsy. A cortectomy with tuber removal was performed in three children treated for drug-resistant focal epilepsy following West syndrome (aged between 3 years and 4 years). Seizures were controlled in 3/5 cases (Engel Ia), reduced in 1/5 case (Engel II), and remained in 1/5 case (Engel III). In all the cases, a behavioural and cognitive progression was observed, with a better quality of life for the families.

Conclusion. The existence of multiple lesions in TSC must not exclude the surgical approach when the epilepsy is focal, very active, and medically resistant with severe cognitive impairment. Epileptogenic zone delineation may necessitate invasive EEG with depth electrodes, which is possible in young children. Early curative epilepsy surgery is technically feasible

in infants and young children, however, the question remains as to whether it correlates with a better behavioural and cognitive outcome, and whether it prevents secondary epileptogenesis in TSC?

10. Resective surgery for refractory epilepsy of childhood associated with glioneuronal tumours

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Introduction. Gangliogliomas (GGs) and dysembryoplastic neuroepithelial tumours (DNETs) constitute highly epileptogenic low-grade tumours of glioneuronal origin. Resective surgery is reported to effectively control tumour-related refractory epilepsy in children and prevent functional sequelae.

Methods. We retrospectively analysed the electroclinical, neuroimaging and neuropsychological data collected during presurgical evaluation and long-term follow-up of 30 children who underwent resective surgery for glioneuronal tumour-related refractory epilepsy at the Epilepsy Center, University Hospital Freiburg, in the years 2000-2010. Mean age at seizure onset was 8.2 years (range: 3 months to 15.5 years) and mean age at surgery was 10.6 years (range: 2.5 to 17.3 years). Four multilobectomies and 26 intralobar resections were performed, including 14 temporal and 12 extratemporal resections. Three children had their first operation in an external institution and underwent repeat surgery in the Epilepsy Center Freiburg due to seizure recurrence, and another 3 children underwent repeat surgery in follow-up due to tumour regrowth. In 7 cases, ECoG was performed intraoperatively, while 4 children underwent long-term intracranial EEG recordings due to lesion proximity to eloquent cortex. Aetiology comprised 13 DNETs and 17 GGs, with 5 children presenting focal cortical dysplasia and 3 hippocampal sclerosis.

Results. At 6 months after resective surgery, 27/30 (90%) children were seizure-free (Engel class I), with this ratio decreasing to 26/30 (87%) at the end of the second year and 23/30 (77%) at follow-up of 1-11 years, mean: 6.3 years. Recurring seizures appeared as late as

3 years after surgery, not directly coinciding with the discontinuation of antiepileptic drugs. Residual lesional tissue on MRI and interictal epileptic discharges in postsurgical scalp EEG correlated with poor outcomes. Developmental outcomes correlated with the duration of epilepsy prior to surgery.

Conclusion. With completeness of resection predisposing favourable outcomes regarding oncological morbidity and seizure alleviation, the latency of surgery appears crucial in treating epilepsy and preserving developmental trajectories, thus making a clear case for early operative intervention.

11. Epilepsy risk in different types of supratentorial brain tumours

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Background. Symptomatic epilepsy is common in patients with brain tumours. Epileptogenesis depends on several factors including tumour histology, location, changes in the peritumoural brain parenchyma, and genetic predisposition. The aim of this study was to investigate the epileptogenic potential of supratentorial brain tumours of different histological types in children.

Methods. Among the paediatric patients investigated and treated in the Neurosurgical Department and Psycho-Neurological Department No.2 of the Russian Children Clinical Hospital and in the Department of Neurology, Neurosurgery and Medical Genetics of the Russian State Medical University (Moscow, Russia) during the period 2006-2011 years, 60 children with supratentorial brain tumours were revealed.

Results. The group of patients ($n=60$) consisted of 28 girls and 23 boys (age range from 5 months to 17 years). Forty-one patients (68.3%) had symptomatic epilepsy. Among the patients were revealed: 8 cases of fibrillary astrocytoma (all with epilepsy), 6 cases of pilocystic astrocytoma (three with epilepsy), 5 cases of pleomorphic xanthoastrocytoma (four with epilepsy), 1 case of desmoplastic infantile astrocytoma ([DIA] without epilepsy), 2 cases of protoplasmic astrocytoma (1 with epilepsy), 5 cases of ganglioastrocytoma (all with epilepsy), 3 cases of mixed glioma (one with epilepsy), 2 cases of mixed ganglioglioma (one with epilepsy), 1 case of anaplastic ganglioglioma (without epilepsy), 1 case of desmoplastic ganglioglioma (with epilepsy), 2 cases of glioblastoma multiforme (both with epilepsy), 1 case of oligodendroglioma (without epilepsy), 6 cases of anaplastic ependymoma (three with epilepsy), 3 cases of choroid plexus papilloma (without epilepsy), 1 case of atypical choroid

plexus papilloma (with epilepsy), 1 case of choroid plexus carcinoma (without epilepsy), 5 cases of dysembryoplastic neuroepithelial tumour ([DNET] all with epilepsy), 1 case of primitive neuroepithelial tumour ([PNET] with epilepsy), 1 case of atypical meningioma (with epilepsy), 3 cases of thalamic hamartoma (all with epilepsy), and 2 cases of meningioangiomatosis (both with epilepsy).

Conclusion. Tumour types consistently associated with epileptogenesis were: fibrillary astrocytoma, ganglioastrocytoma, DNET, PNET, hamartoma, meningioangiomatosis, desmoplastic ganglioglioma, glioblastoma multiforme, and atypical meningioma. Pleomorphic xanthoastrocytoma was epileptogenic in 80% of cases. Pilocystic astrocytoma, protoplasmic astrocytoma, mixed ganglioglioma, and anaplastic ependymoma were associated with a 50% risk of epilepsy. Tumour types such as DIA, mixed glioma, anaplastic ganglioglioma, oligodendroglioma, choroid plexus papilloma, and carcinoma demonstrated low epileptogenic risk.

12. Epilepsy unexpectedly revealing ganglioneuroma during follow-up for neuroblastoma, ten years later

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Neuroblastoma and ganglioneuroma are paediatric neuroblastic tumours arising in the sympathetic nervous system from neural crest cells. Outcome depends on the stage of disease and age at diagnosis.

We report a case of epilepsy that occurred ten years after chemotherapy for neuroblastoma treatment. The patient presented at birth with a heterogenous and bulky cervico-thoracic tumour without genetic 1p deletion or *N-MYC* amplification. The treatment choice was multimodal therapy, VP16/carboplatine and endoxan/oncovin with surgery; both allowed acquisition of developmental milestones and successful schooling despite palsies of the left leg and right arm. The occurrence of partial seizures, which increased in frequency, were localised to the temporo-occipital area and were resistant to antiepileptic drugs, prompted us to perform MRI. The latter revealed a new well-delimited cortico-subcortical tumour in the left temporal area. Fortunately, complete tumour excision was achieved by neurosurgery, with a decrease in seizure frequency. Both genetic markers and catecholamine assays were negative. An anatomopathological study of part of the tumour concluded a dysembryoplastic neuroepithelial tumour. The present follow-up consists of clinical examination and EEG every three months.

This case highlights once again the extreme variability in outcome of paediatric ganglioneuroma which are benign tumours of the sympathetic nervous system. We insist on the need to keep these children under a high level of clinical surveillance, comparable to that observed during their oncology care in the long-term.

13. Bottom-of-sulcus dysplasia in children with refractory epilepsy

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Objectives. To review children with bottom-of-sulcus dysplasia (BSD) and analyse electroclinical and imaging features, as well as outcome after epilepsy surgery. **Patients and Methods.** Twenty patients (2-19 years of age) with BSD were selected among all children with focal cortical dysplasia (FCD) admitted for presurgical evaluation (2008-2011), which included 3T-MRI and FDG-PET/MRI co-registration. Surgery was performed in 10 cases and after invasive evaluation (depth and subdural electrodes) in 8 cases.

Results. BSD represented 33% of our FCD cases. Most children had early-onset focal epilepsy (mean age: 3.9 years), and previous "negative" MRI. All patients had daily seizures (80% sleep predominance) and 35% focal epileptic status. Clinical deterioration occurred in 5 children. Scalp video-EEG showed highly informative localising data. 3T-MRI and FDG-PET/MRI showed mild gyral abnormalities mainly at the bottom of a sulcus, involving frontal, peri-rolandic or peri-sylvian regions, congruent with electroclinical data. Distinctive imaging findings included: 1) abnormal depth and/or thickness plus hypometabolism of the cortical ribbon, 2) mild juxtacortical FLAIR hyperintensity, 3) subtle blurring of grey-white matter junction, and 4) thin transmantle tail. Invasive EEG evaluation showed the characteristic epileptiform patterns of type II FCD in the deepest contacts by exploring the abnormal gyri. After gyrectomy/tailored resection, all patients were Engel class I. Pathology revealed ILAE type II FCD (50% IIB).

Conclusion. Children with BSD and severe epilepsy show highly informative focal electroclinical findings. Previously overlooked lesions can be revealed after high signal-to-noise 3T-MRI and FDG-PET/MRI co-registration. Excellent seizure outcome can be achieved after early epilepsy surgery, which, in addition, prevents neurological and cognitive deterioration.

14. Successful corpus callosotomy in a child with Dravet syndrome and SCN1A abnormality

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Introduction. Epilepsy surgery is rarely considered for children with underlying diffuse genetic/metabolic aetiologies. These children have dysfunction that will persist and seizure freedom following resection may be an unrealistic expectation. However, palliative procedures such as VNS or corpus callosotomy may be beneficial.

Methods. This is a case report of beneficial surgical treatment with corpus callosotomy in a 12-year-old girl with Dravet syndrome and SCN1A abnormality. Surgeries were performed prior to knowledge of the underlying aetiology of the seizure and epilepsy syndrome. Records were reviewed to document treatments and results.

Results. Seizure onset was at 3 months of age in August 2000. The initial seizure was a unilateral tonic-clonic seizure, lasting more than one hour, followed by postictal paralysis. EEG demonstrated right hemisphere slowing and right independent frontal, temporal, central, and parietal spikes. Treatment with carbamazepine (CBZ) was started. Within three weeks, there were numerous brief seizures, staring and loss of tone, and rare myoclonic seizures associated with diffuse spike-wave complexes. The CBZ was discontinued and treatment with phenobarbital initiated. She was seizure-free for two months. When seizures recurred, she was treated with topiramate (TPM) and had no seizures from November 2000 to March 2001. Seizures recurred; these were unilateral, either right or left, and occasionally generalised. Seizures increased in frequency and severity and staring with atonic head nods began. Primidone, phenytoin, lamotrigine, sodium valproate, felbamate, clonazepam and the ketogenic diet were tried. Seizures worsened; many generalised and were occasionally followed by postictal hemiparesis. The VNS was implanted and multiple settings were tried with no benefit for two years. Additional medications were tried. The VNS was turned off due to intolerance, dysphagia, and no benefit. By March 2004, at the age of 3 years and 10 months, she had been treated with 18 different antiepileptic medications, in combinations and monotherapies, the ketogenic diet, and the VNS in multiple duty cycles. She had staring with atonic head nods too numerous

to count (over 100 per day) and unilateral secondary generalised tonic-clonic seizures, 3-8 per week. There were 2-3 episodes of convulsive status epilepticus per month that did not respond to rectal diazepam, and required emergency room treatment, often intubation and hospitalisation. She had an anterior 2/3 corpus callosotomy with marked improvement. Within three months, seizures started to increase and by late August 2004 episodes of secondary generalised status epilepticus returned. In September 2004, the corpus callosotomy was completed. Since September 2004, there have been 4-8 unilateral seizures per month, a greater than 80% decrease, and one or two episodes of status per year that required hospitalisation, a greater than 80% decrease. The staring with atonic seizures also decreased. Parents state she improved cognitively, but she remains moderately impaired. In April 2010, the *SCN1A* abnormality in exon 26, gly→val change at C-5174, was found.

Conclusion. The effect of corpus callosotomy on seizures may be present even when the abnormality is diffuse such as for a channelopathy. Individuals with diffuse abnormalities may still respond to palliative surgical procedures.

15. Epilepsy associated with a *SCN1A* mutation and focal cortical dysplasia: therapeutic and prognostic considerations

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Brain magnetic resonance imaging (MRI) in patients with severe myoclonic epilepsy in infancy (SMEI) or Dravet syndrome has shown abnormalities in a small minority of patients. When present, brain abnormalities are mainly non-specific and more frequently observed in patients with SMEI or Dravet syndrome, not associated with mutations in the sodium channel gene *SCN1A*. Only a few cases of focal cortical dysplasia have been described in patients with epilepsy and *SCN1A* mutations.

We report the case of a girl aged 20 months, without a family history of epilepsy or febrile convulsions and without a previous personal history of disease. Epilepsy was diagnosed following prolonged hemiconic febrile convulsions with an onset at age 3 months. A *de novo* truncating mutation in *SCN1A* was found and MRI revealed focal cortical dysplasia over the right temporal region. Physical examination and EEG were normal. Up to now, she has not developed any other type of seizure, other neurological signs or retarded psychomotor development. Despite being on valproate postoperatively since age 5 months, she has

continued to present with seizures provoked by fever, and rectal diazepam is being administered in these cases.

We discuss some therapeutic and prognostic considerations: what drugs should be used or avoided in this case? Should we proceed to a presurgical evaluation despite the detection of a *SCN1A* mutation and a clinical profile that in the long term could ultimately be compatible with SMEI?

16. Epileptic encephalopathy in children and epilepsy surgery: a challenge based on an equivocal terminology

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Introduction. The concept of epileptic encephalopathy – “a condition in which the epileptiform abnormalities themselves are believed to contribute to the progressive disturbance in cerebral function” – was defined by the ILAE as a key term in 2001 (Engel JJ). It was formally recognised in the 2006 Report ILAE Core Group and was better clarified by Berg *et al.* (2010) within the Special Report of the ILAE Commission on Classification and Terminology. Nowadays, “epileptic encephalopathy embodies the notion that the epileptic activity itself (and/or recurring seizures) may contribute to severe cognitive and behavioural impairments above and beyond what might be expected from the underlying pathology alone....., and that these can worsen over time”. From a practical point of view, we know that the suppression of epileptic activity (and recurring seizures) may improve cognition and behaviour. Considering terminology, the first problem is that some syndromes are often referred to as epileptic encephalopathies. The best known and most common of these are: West syndrome, Lennox-Gastaut syndrome, Dravet syndrome, Landau-Kleffner-CSWS syndrome, Doose syndrome, EME, EIEE and epilepsy of infancy with migrating focal seizures (MFSI). In almost all of these conditions, the evolution is related to the underlying causes and patients with these disorders cannot be considered for epilepsy surgery. However, the encephalopathic effects of both seizures and epileptiform abnormalities may occur in association with any form of epilepsy, leading to an epileptic encephalopathic condition.

Methods. We evaluated the electroclinical and cognitive features of 6 children (age at onset/age at surgery in months: Patient 1: 1/29; Patient 2: 1/77; Patient 3: 28/160; Patient 4: 12/28; Patient 5: 9/135; Patient 6: 6/37) affected by focal epilepsy without any of the

above-mentioned syndromes but with an “encephalopathic evolution” of the disease. The anatomo-electroclinical characteristics fulfilled the criteria for different kinds and localisations of surgical treatments which were performed. After surgery we re-evaluated the electroclinical and cognitive features.

Results. In all 6 patients, epilepsy surgery improved the unfavourable evolution with regression of the encephalopathic condition. In two (Patients 1 and 4), seizures recurred. Patient 1 was operated on a second time at 77 months and has been cured since then (57 months), Patient 4 presented with rare clusters of seizures, but had a normal neurocognitive condition.

Conclusion. “*Epileptic encephalopathy*” is a misleading definition, both in lexical and in practical management terms. On one hand, the above-mentioned “*epileptic encephalopathies*” do exist as well defined epileptic syndromes. On the other hand, it must be kept in mind that focal symptomatic epilepsy, regardless of aetiology, may evolve into an encephalopathic drug-resistant condition, characterised by a developmental delay worse than expected for the epilepsy “*per se*”. This should be assessed early in order to propose a surgical treatment which may dramatically improve the prognosis with a regression of the encephalopathic condition. In this view, we propose that the term “*epileptic encephalopathy*” must be reserved for the classic syndromes, and “*functional epileptic encephalopathy*” could refer to the encephalopathic conditions linked to those focal epilepsies with a peculiar unfavourable evolution, for which a surgical evaluation and treatment must be considered early.

17. Hemispherotomy in patients with epileptic encephalopathy with continuous spike-waves during sleep and prenatal post-haemorrhagic hydrocephalus: a pre-and postsurgical neuro-functional study

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Aim. To study pre- and postsurgical neuro-cognitive functions and epileptic outcome in a child with a refractory epileptic encephalopathy with continuous spike-waves during sleep (CSWS), secondary to post-haemorrhagic three-ventricular dilatation and glyotic thalamic lesion, successfully treated by hemispherotomy.

Case report. We present a 6-year-old child with a family history negative for seizures or neurological disease. At 34 gestation weeks (GWs), cerebral ventriculomegaly was detected. Delivery occurred at 37.5 GWs, without perinatal distress. Neonatal MR showed bilateral ventriculomegaly (right>left), an extensive brain periventricular parenchymal loss involving right parietal and frontal lobes and including homolateral basal ganglia and thalamus. During the first months of life, neurological examination revealed asymmetric spontaneous motility with best performances on the right side. After 6 months of life, a motor developmental delay with a reduction of motor abilities of the left limb was observed. A definite left hemiparesis was diagnosed at 18 months. Sequential developmental assessment with the Griffiths Scales performed at 15, 19, and 33 months showed values in the normal range for all the subscales except that of locomotor. The first febrile focal seizure with secondary generalisation occurred at 25 months. From the age of 3 years and 6 months, the child presented with rare left motor seizures and daily atypical absences, while the EEG study showed the onset and a progressive increase of paroxysmal activity leading to a CSWS pattern, involving 85% of slow sleep. Despite antiepileptic drugs, as monotherapy and in different combinations, seizures and CSWS persisted, associated with a progressive deterioration of cognitive functions. At 6 years, neurological examination showed mild left hypertonia and hyper-reflexia with asymmetric strength (right>left), more evident on the upper limb, left hand mirror movements, which did not prevent the functional use of both hands, left hypohaesthesia, and homonymous hemianopia. Cognitive evaluation showed mild mental retardation (IQ: 60) and behavioural disorders with dramatic effects on school activities and family life. A presurgical sensory-motor functional study was performed specifically in order to ascertain the side of functional motor control and evaluate the potential damage of hemispherectomy. A non-invasive neuro-functional study consisted of motor evoked potentials and motor area mapping, and passive functional MRI in order to evaluate sensory-motor functional connectivity, as well as cortical-spinal tractography. The results of the study suggested an ipsilateral motor control of the left arm. At the age of 6 years, right peri-insular hemispherotomy was performed without surgical complications. After surgery, a full control of seizures and an immediate disappearance of CSWS occurred, without worsening of motor function. Neuropsychological evaluation showed also a progressive improvement of cognitive functions (attention, memory, and visuo-perceptive functions).

Conclusion. In our case, the indication of hemispherotomy to treat the epileptic disorder was not straightforward due to the presence of bilateral lesion,

prevalent on the right side, with good function of the affected left hand. The results of an extensive non-invasive sensory-motor functional study, suggesting bilateral control of motor function by the healthy hemisphere, have helped us to choose the surgical option, the effectiveness and safety of which appear to be confirmed by postsurgical follow-up.

18. Long-term outcome of hemimegalencephaly after hemispherotomy in infancy

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Introduction. Hemimegalencephaly (HME) is a rare brain developmental anomaly, characterised by unilateral enlargement of the cerebral hemisphere. Epileptic seizures occur in up to 90% of the patients and seizures often begin within the first days of life. Hemispherotomy is an effective procedure for the control of epileptic seizures. The analysis of the literature shows that duration of epilepsy prior to surgery is an important factor in determining adaptive outcome. The aim of our study was to analyse the long-term outcome in a series of infants with HME treated with hemispherotomy.

Methods. We retrospectively studied 12 infants with medically refractory epilepsy with HME who underwent hemispherotomy under 1 year of age at the National Center of Neurology and Psychiatry from December 2001 to February 2009 and who had a minimum follow-up of three years. The criteria for hemispherotomy were: intractable seizures lateralised or localised within the hemimegalencephalic hemisphere and the absence of seizure origin and structural abnormality in the contralateral hemisphere. Vertical parasagittal hemispherotomy was performed for all patients by the same neurosurgeon (TO). Post-operative seizure outcome was assessed using Engel's classification from the medical charts and telephone interviews with the parents.

Results. Seven males and 5 females with HME underwent hemispherotomy under 1 year of age. Seizure began at a mean age of 6 days (range: 1 to 140 days) and 11 patients had seizure onset at less than 1 month of age. Early infantile epileptic encephalopathy with suppression burst was observed in 11 patients. Five patients had neurocutaneous syndrome (hypomelanosis of Ito in 3, linear sabcutaneous gland birthmark syndrome in 1, and lipomatosis in 1). The mean age at operation was 4 months (range: 2 to 9 months). At a mean follow-up of 6.6 years (range: 3.0 to 10.1 years), there was no mortality. Complete

seizure control occurred in 8 (67%), and of these, all antiepileptic drugs (AEDs) were withdrawn post-operatively in two and significantly reduced in two others. In four of 12 patients, seizures recurred and persisted after hemispherotomy. In recurrent cases, video-EEG monitoring showed that seizures arose from the contralateral hemisphere. Patients with seizure-free outcome showed improvement in post-operative development, whereas all 4 patients with residual seizures showed no developmental progress and were bed-ridden.

Conclusion. Hemispherotomy in infants with intractable epilepsy related to HME is associated with an excellent chance of seizure control, reduction or cessation of AEDs, and consequently, accelerated neurodevelopment.

19. Cortical involvement during epileptic spasms: an intracerebral EEG study

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Introduction. The pathophysiological mechanisms of epileptic spasms are still currently poorly understood. The role of the subcortical structures has been put forward mainly on the basis of non-localised EEG features and experimental data. The description of asymmetric spasms associated with lateralised EEG patterns have challenged this view and raised the issue of a focal cortical origin. Several studies based on ictal SPECT have provided arguments for a cortical origin in some patients. This study was aimed at analysing the cortical pattern recorded during intracerebral EEG recordings of children with spasms.

Material and Methods. Eight children for whom a presurgical evaluation was performed for drug-resistant epilepsy, in which epileptic spasms was the main ictal feature, were studied. In all these patients, several features (EEG, Clinical, and MRI) suggested a possible focal origin. All had a complete non-invasive Phase I evaluation followed by an intracerebral electrode investigation (iEEG). Cortical involvement was visually analysed and studied using the "Epileptogenicity Index" (EI) method. In this way, we quantified the capacity of each explored cortical region to generate high frequency activity (beta-gamma) relative to the delay of involvement of each region.

Results. The mean age of the patients was 8 years and 9 months at the time of iEEG exploration. Seven of 8 had a cortical lesion identified by MRI. All but one showed an involvement of cortical areas during the

ictal activity (spasms). In 7/8, the EI showed maximal values either in the temporal ($n=2$), the parietal ($n=1$) or the frontal cortex ($n=4$). During the spasms, the iEEG generally showed an involvement of premotor cortex ($n=7$). In 1 patient, no cortical involvement was found.

Conclusion. In the majority of these patients, a cortical focal origin of epileptic spasms could be observed. Thus a scenario in which the epileptic discharge is primarily driven by the cortex was the most plausible. In 1 patient, with no clear focal origin, the role of subcortical regions could be prominent.

20. Acute intralesional recording in hypothalamic hamartomas: description of four cases

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Introduction. Hypothalamic hamartomas (HH) are well-recognised congenital malformations of the brain and represent a cause of medically intractable focal epilepsy. Interictal and ictal scalp EEG is usually poorly informative in most cases. **Aim.** To report the results of acute intralesional recording of the interictal activity arising inside the HH in four cases and to compare this with concurrent scalp EEG.

Methods. We have reviewed the medical records of 17 children affected by drug-resistant focal epilepsy associated with HH, referred between January 1990 and December 2011 to the Neurology Division of the Bambino Gesù Children's Hospital in Rome and "Carlo Besta" Neurological Institute in Milan. All clinical notes were reviewed in order to evaluate the age at onset, the seizure type at onset and during the follow-up, treatment, and epilepsy outcome. HHs were defined according to the classification proposed by Delalande *et al.* (Delalande *et al.*, 2003). In 4 of 17 patients studied, an intraoperative endoscopic intralesional recording was performed. In 3 cases, intraoperative stereo-EEG was carried out using foramen ovale electrodes (Dixi Medical ACS-798S, 5 contacts) and DBS leads in 1 case (Medtronic 3389, 4 contacts). Scalp-EEG was performed

in all patients (referential montage, lacking frontal and central leads for surgical reasons).

Results. In 10 of 17 cases, a surgical approach was performed: 4 of these underwent a one-stage stereo-endoscopic disconnection, three cases a double-stage stereo-endoscopic disconnection, and 1 case required a multistage stereo-endoscopic disconnection. A bilateral deep brain stimulation (DBS) protocol was performed for 1 patient, 1 a surgical resection, 1 radiotherapy, and 1 radiosurgery. Following the Delalande classification, HH were mostly of type 2 (8 of 17), followed by type 3 (6 of 17) and lastly type 4 (3 of 17). No cases were classified as type 1. All patients (4 cases) presented gelastic seizures, associated with focal and secondary generalisation. Seizure frequency ranged from multiple per day to multiple per week. Seizure duration was between a few seconds to 60 seconds. Following Delalande classification, 2 patients had type 2 HH and another 2 had type 4 HH. Interictal scalp EEG showed epileptiform abnormalities in all patients; in 2 cases these were evident on the temporal region, in 1 over the bilateral parieto-occipital region, and in 1 over the central and parietal regions. In all patients, intraoperative scalp EEG showed synchronous interictal epileptic discharges which were recorded from the same side of the lesion. From the depth electrode, high-amplitude fast activity in sequences of variable duration was recorded. In all cases, the predominant side of the EEG abnormalities was ipsilateral to the activity recorded within the hamartoma.

Discussion. We detected the electrical activity of the lesion and compared it with the scalp EEG activity recorded at the same time. Our data confirm the epileptogenicity of HH. Clinical and neurophysiological findings in all described cases suggest that the cortical activation is secondary to the epileptogenic activity of HH. Intraoperative electrographic recording may be considered as part of a strategy of transendoscopic surgery for HHs.

21. Long-term efficacy and hospitalisation rate in children with refractory focal epilepsy treated by vagus nerve stimulation

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Aim. Evaluation of long-term efficacy in children with refractory focal epilepsy treated by vagus nerve stimulation.

Methods. We retrospectively analysed 15 children with intractable focal epilepsy treated by vagus nerve stimulation. The inclusion criteria were: age younger than 18 years at the time of VNS implantation, confirmed electroclinical diagnosis of refractory focal cryptogenic or symptomatic epilepsy without the possibility of resective surgery, and at least 12 months follow-up after VNS implantation. We analysed the effectiveness at 1, 2 and 5 years of follow-up. We counted the average number of urgent hospitalisations per year for each patient before and after the VNS implantation. The study population consisted of 15 children; 6 males and 9 females. The median age at time of implantation was 14.6 ± 2.5 years and median duration of epilepsy before implantation was 11.0 ± 3.4 years. The preoperative investigation identified symptomatic aetiology in 10 patients (66.7%) and cryptogenic aetiology in the remaining 5 patients (33.3%). We were also able to determine the probable localisation of the seizure onset zone during the preoperative investigation. Eight patients had extratemporal epilepsy (53.3%), 1 patient temporal lobe epilepsy (6.7%) and in the remaining 6 patients (40%) the multifocal ictal onsets were revealed. Of the children with extratemporal epilepsies, 7 had frontal lobe epilepsy (of whom four were without any clear lateralisation, 2 with onset on the left and 1 on the right side) and 1 patient had seizure onset in the operculo-insular region on the right side. Nine of 15 patients (60%) had partial seizures (PS) only, 5 patients (33.3%) had a combination of PS and generalised tonic-clonic seizures (GTCS) and 1 patient had GTCS only.

Results. The mean seizure reduction was 42.5% at 1, 54.9% at 2, and 58.3% at 5 years. The number of responders was 7 (46.7%) at 1 and 9 (60%) at both 2 and 5 years, respectively. The average monthly frequency of all epileptic seizures preoperatively was 71.9 ± 52.7 . We observed the gradual decrease of seizure count postoperatively (43.7 ± 46.1 at 1, 35.9 ± 38.4 at 2, and 29.0 ± 36.4 at 5 years, postoperatively) ($p < 0.001$, in comparison to the preoperative period). The mean number of urgent hospitalisations per one patient was 1.0 ± 0.6 per year, preoperatively and 0.3 ± 0.5 per year, postoperatively ($p < 0.0001$). A serious adverse event was observed in one patient (late-onset asystole).

Conclusion. Vagus nerve stimulation is an effective and relatively safe method for treating children with refractory focal epilepsy.

22. Application of a carbon dioxide laser in corpus callosotomy

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Introduction and Methods. Here we describe the application of a flexible carbon dioxide (CO₂) laser for corpus callosotomy in three children with epilepsy. These cases illustrate the use of this device in completing a corpus callosotomy safely and efficaciously. **Results.** In all cases there was clean lesioning of the tract with preservation of the ependymal plane. The low profile laser fibre tip is well suited for working in the depths of the interhemispheric fissure with decreased brain retraction. The flexible CO₂ laser allows a more precise and controlled callosal lesioning due to its dual cutting and hemostatic properties.

Conclusion. We believe this flexible, low profile CO₂ laser is an excellent additional tool for the neurosurgeon performing corpus callosotomies.

23. A systematic comparison of covert and overt verb generation during presurgical fMRI scanning in paediatric epilepsy patients

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Introduction. We aimed to assess the feasibility of imaging overt (articulated) speech for presurgical mapping of expressive language with functional magnetic resonance imaging (fMRI) in paediatric epilepsy patients.

Methods. Thirty-eight medically intractable symptomatic paediatric epilepsy patients (6-17 years) performed a covert (non-articulated) and overt verb generation (VG) task during continuous fMRI scanning. Measures of data quality (task performance, in-scanner movement and quality ratings) were compared between covert and overt speech conditions. The neural substrates engaged during covert and overt VG were also compared, as were laterality indices in regions of interest in frontal and temporal cortex and fMRI signal strength.

Results. In-scanner movement increased during overt VG, but did not reduce data quality; good quality data were found in 50% of patients during covert speech and in 68% during the overt speech condition. Patients performed more poorly inside the scanner, especially younger patients and those with larger brain lesions. Verbal IQ and practice performance related to better fMRI data quality and there was more movement in younger patients. Similar brain regions were engaged in both conditions, with overt speech also involving the dominant premotor cortex. fMRI laterality indices were in agreement across conditions, despite a trend for reduced values during overt speech. Categorical judgments of language dominance were also equivalent between conditions.

Conclusion. Imaging overt speech with fMRI is feasible in paediatric epilepsy patients and valid in the presurgical setting; providing measures of language laterality which are in agreement with the current non-invasive gold standard. Importantly, overt paradigms provide the advantage of monitoring performance in the scanner, which appears crucial.

24. Low-frequency electrical stimulation of white matter tracts in intractable mesial temporal lobe epilepsy

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Objective. To explore the tolerability and efficacy of low-frequency stimulation (LFS) of white matter tracts (WMT) in patients with intractable mesial temporal lobe epilepsy (MTLE).

Background. Deep brain stimulation has had limited success in epilepsy. However, WMTs have not been stimulated, and low frequencies have not been adequately explored for treatment of epilepsy.

Design/Methods. Seven adult patients with MTLE underwent implantation of depth electrodes in WMTs: the corpus of the fornix ($n=2$) and lateral to the splenium of the corpus callosum where fibres of the dorsal hippocampal commissure travel with the crus of the fornix ($n=5$). All patients had hippocampal electrodes. 1-Hz stimulation of the WMT in all subjects elicited hippocampal evoked potentials (EPs), confirming connectivity. The EP duration was approximately 200 msec, suggesting that 5-Hz stimulation would elicit continuous interference of hippocampal activity. LFS-WMT occurred in 4-hour sessions (1-3 sessions/patient at 5 Hz; current: 8 mA; pulse width: 0.2 msec) in the video-EEG unit.

Results. There were no complications and hourly mini-mental status examination scores showed a mean increase of 0.917 during LFS-WMT ($p<0.001$; Generalised Estimating Equations [GEE]-Identity Link Function [ILF]). LFS-WMT resulted in a significant reduction of hippocampal spikes ($p=0.001$; GEE-ILF), which persisted for the subsequent four hours ($p=0.05$). Seizure odds were reduced by 87% in 1-2 days following each 4-hour LFS-WMT session ($p=0.001$; GEE-Logit Link Function), without medication changes.

Conclusion. Our data show that LFS-WMT activates the hippocampus and other areas of Papez circuit, and reduces epileptiform discharges and seizures, without affecting memory. Stimulation of WMTs appears to be attractive because it interferes with spontaneous electrical activity of a relatively extensive cortical area and requires less current injection, implying more safety and a longer battery life. Our data illustrate the need for an exploratory trial to confirm safety and tolerability of LFS-WMT using different parameters and a chronic implantable pulse generator.

25. How to improve the yield of FDG-PET in the preoperative work-up for epilepsy surgery

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Introduction. Fluorodeoxyglucose PET (FDG-PET) is a non-invasive, interictal method to localise epileptogenic foci, marked by decreased FDG uptake. In temporal lobe epilepsy (TLE), FDG-PET is a sensitive marker of the epileptogenic focus, even when the MRI is negative. Particularly for focal extratemporal lobe epilepsy (extra-TLE), computer-aided analysis could show a higher yield over visual interpretation. In this study, we retrospectively compared three FDG-PET assessments: 1) initial clinical visual assessment (PET-clin), 2) blinded visual re-assessment (PET-re), and 3) computer-aided Statistical Parametric Mapping (SPM) analysis (PETspm). We investigated their clinical value by comparing results with the gold standard of ictal electrocorticography (ECoG).

Methods. Included in this study were 44 refractory focal epilepsy patients (mean age: 25 years; 19 female; 75% extra-TLE) who underwent chronic ECoG monitoring in the UMC Utrecht, The Netherlands. All patients underwent a static FDG-PET scan (ECAT EXACT HR+) during preoperative work-up, as did 38 healthy controls in the Hospices Civils de Lyon. All FDG-PET scans were evaluated for decreased FDG uptake. First,

clinical visual reports were collected from the clinical database. Second, a blinded visual re-assessment was performed by the same two experts (RD and EC). Third, the scans were analysed using a SPM(8) voxel-based analysis. Preprocessing involved co-registration of the FDG-PET scan to a 3D T1 MRI, smoothing, and normalisation. Subsequently, each patient PET scan was compared, at voxel level, to the healthy control dataset using a two-sample t test. After applying a liberal (1. $p_{none} < 0.001$) and strict (2. $p_{FWE} < 0.05$) threshold on whole brain level, resulting significant clusters were re-transformed and overlaid on the individual MRI. Chronic ECoG was performed with subdural electrode grids and strips covering the presumed focus. The ictal onset zone in the ECoG was determined. Finally, all results were classified into ten predefined brain anatomical regions. On the group level, inter-observer agreement and sensitivity, specificity and accuracy of the three strategies, compared to the ictal ECoG, were calculated. The added value of the visual re-assessment and SPM analysis was also evaluated at the level of individual patients.

Results. The number of scans without abnormalities for PETclin, PETre and PETspm1&2 were 5, 1 and 0 and 10, respectively, without overlap between PETclin and PETre. The highest inter-observer agreement was found between PETclin and PETre. Preliminary results indicate that PETre increases the overall sensitivity (>55%) compared to PETclin, maintaining equal specificity (>90%). PETspm resulted in multiple hypometabolic clusters in most patients, resulting in an overall higher false positive rate compared to the ictal ECoG. However, on the individual patient level, PETspm may reveal new or additional information regarding the epileptogenic focus when added to the other clinical information.

Conclusion. Blinded visual re-assessment should be considered

26. Definition of a stereotactic 3D model of the human insula as a neurosurgical approach (epilepsy and stereotaxic surgery)

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Objectives. To design a method for 3D reconstruction of the insula, including the gyri and sulci, in AC-PC reference, to be used individually for imaging or for epilepsy and stereotaxic surgery.

Materials and Methods. Morphometric study of MRI of normal insular regions in 56 subjects; 26 males and 30 females, 28 left and 28 right hemispheres. Stage 1: reconstruction in AC-PC reference of the insula

from 3D-T1-MRI slices 1 mm thick. Stage 2: digitalisation and superposition of data in 3D using PhotoStudio software (Photo Editing Software) system with PC as the centre of coordinates. Stage 3: MATLAB software (Mathworks Inc.) was used to transform, in colour, values of each pixel to obtain a colour scale corresponding to the probability of *insula sulci* localisation between 0 and 100%.

Results. Demonstration of highly significant correlations between the coordinates of the main insular structures (angles, sulci) and the length of AC-PC. This close correlation allows us to describe a method for 3D reconstruction of the insula on MRI slices that requires only the positions of AC and PC, and subsequently the inter-commissural (AC-PC) length. This procedure defines an area containing insula with 100% probability.

Conclusion. 3D reconstruction of insula will be potentially useful: 1) to improve localisation of cortical areas, allowing the differentiation between insular cortex and opercular cortex during stereoelectroencephalographic exploration of patients with epilepsy (SEEG) or in morphological and functional imaging; 2) as a microsurgical approach for the insula using neuronavigation techniques; and 3) to identify the insula during stereotaxic surgery (SEEG, biopsy).

27. Intellectual development before and cognitive outcome after frontal lobe epilepsy surgery in children

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Introduction. Cognitive outcome after temporal lobe resection (TLR) is relatively well known, but few data are available on the cognitive effects of epilepsy surgery in the frontal lobe, especially in children. We compared cognitive outcome after epilepsy surgery in children with frontal lobe epilepsy (FLE) and temporal lobe epilepsy (TLE). In addition, we investigated the effect of intractable FLE on intellectual development by tracing the developmental trajectory preceding the surgical intervention.

Methods. Nineteen 6 to 18-year-old children undergoing frontal lobe resection ([FLR], 5 left-sided and 12 right-sided) were compared with children undergoing TLR matched for side of resection and age at surgery (average: 12.5 years). A comprehensive neuropsychological assessment containing 12 cognitive domains was administered preoperatively and repeated two years postoperatively. Of the FLE children, 58% were seizure-free two years postoperatively. Fourteen FLR children had had their intellectual quotients (IQ)

assessed before the preoperative assessment. These longitudinal data were used to retrospectively trace the intellectual development of those children before surgical treatment.

Results. Preoperative cognitive performance was below average in FLR children and significantly lower than in TLR children in most cognitive domains. Analyses of variance revealed two significant interaction effects; verbal short-term and working memory declined significantly after FLR compared to improvement after TLR, and left-sided FLR was associated with decline in expressive language compared to improvement in other groups. Both FLR and TLR children improved in full scale IQ (FSIQ) and performance IQ (PIQ). In the subset of FLR children with available longitudinal data, change in IQ was negatively correlated with follow-up time.

Conclusion. Intractable FLE may affect intellectual development adversely over time in children. School-aged FLR children function on a lower general cognitive level than age-matched TLR children before surgery. FLR is associated with more cognitive risks than TLR, including decline in verbal short-term and working memory and, after left-sided FLR, decline in expressive language. On the other hand, both FLR and TLR are associated with improvement in FSIQ and PIQ.

28. Magnetic source imaging in paediatric epilepsy: clinical characteristics and postsurgical language outcome in patients with atypical language dominance

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Introduction. Magnetoencephalography/magnetic source imaging (MEG/MSI) is integral to presurgical evaluations for paediatric epilepsy patients, as it yields data regarding localisation of seizure onset (MEG) and lateralisation of cerebral language dominance (MSI). Delineating areas of eloquent cerebral cortex facilitates prediction of postsurgical language outcome. The current study presents MSI language lateralisation data for the largest series of paediatric epilepsy patients reported thus far. Preliminary data analysis reveals recognisable patterns of relevant clinical features among patients with atypical language dominance and stable or improved language functioning for the majority of patients assessed.

Methods. Data from 27 paediatric epilepsy patients (aged 3-17 years) was collected *via* retrospective record review. All patients were being considered for epilepsy surgery, due to focal intractable seizure disorder. All patients produced valid language lateralisation data during the receptive language (CRM) task of the MSI protocol, which involved listening to a list of words

and identifying target words among foils during six trials of a recognition task. The magnetoencephalography (MEG) unit consisted of a 148-channel Magnes 2500 WH system and data were analysed using the single equivalent current dipole (ECD) model across each whole hemisphere. Mean age was 11 years (range: 3-17). Twelve patients were female. Seven patients were left-handed. Six patients had FSIQ<70. Sixteen patients had primarily left-hemisphere seizure focus and 11 patients had primarily right-hemisphere seizure focus. **Results.** MSI data demonstrated left language dominance for 11 patients (41%), right language dominance for 6 patients (22%), and bilateral language for 10 patients (37%). Of the 16 patients who showed atypical (non-left) language dominance, 13 (81%) had seizure onset <5 years of age, 12 (75%) had duration of epilepsy >4 years, 10 (63%) had primarily left hemisphere seizure focus, 4 (25%) were under age 8 years at the time of MSI, 6 (38%) were left-handed, and 2 (13%) had FSIQ <70. Ten of the 16 atypical language patients underwent focal resective epilepsy surgery (6 left, 4 right). Eight of these patients underwent postsurgical neuropsychological evaluation. Seven of these 8 patients demonstrated stable or improved performances on three separate tests of postsurgical language functioning. Three of these patients were classified as having right hemisphere language dominance per MSI and had surgery in the left (non-dominant) hemisphere. These patients would not have been expected to experience language decline. The remaining 5 patients who were classified as having bilateral language showed no language decline after focal resection in a hemisphere with language representation on MSI.

Conclusion. Preliminary data analysis implicates younger age at seizure onset, longer duration of epilepsy, and left hemisphere seizure focus as prominent characteristics for the majority of patients with atypical language dominance on MSI. Focal resection in a hemisphere with MSI-demonstrated language representation did not cause measurable decline in language in this sample.

29. Personality profiles of children before and after epilepsy surgery

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Introduction. A large corpus of research has established an increased prevalence of psychopathological and psychiatric disorders among children after epilepsy surgery. Up till now, personality profiles of children with (medically intractable) epilepsy have remained

underexposed. We charted the personality profiles of children who were candidates for epilepsy surgery using an instrument designed for the general childhood population, *i.e.* not intended to detect behaviour problems or pathology.

Methods. Twenty-four children (mean age: 10.0 years; 15 girls) were consecutively included in a country-wide study of neuropsychological functioning shortly before epilepsy surgery, and 6 children, 12 and 24 months after epilepsy surgery. For every patient, a healthy, age- and gender-matched control was assessed at similar intervals. In this study, data obtained from parents before, and six months after, surgery is presented. The questionnaire "Individual Differences between Children" (Dutch "Blikvanger") (Elphick *et al.*, 2002) mapped the personality profiles of children. This instrument was based on a lexicon formed by free parental child descriptions. The 128 items of the "Blikvanger 5-13" provide information of five domains and 16 subdomains: extraversion (subdomains: initiative in contact, positive emotionality and sociability), friendliness (subdomains: dominance, compliance, altruism and affection), carefulness (subdomains: carefulness and impulsiveness), emotional stability (subdomains: emotional stability, self confidence and manageability) and development (subdomains: curiosity, school performance, creativity and autonomy). The questionnaire has sufficient reliability and construct validity.

Results. In contrast to the main domains, differences between patients and controls existed in subdomains. Very low scores (deciles 1 and 2) indicating poor functioning were more common in patients than in controls for the subdomains: autonomy (patients 75%, controls 13%), school performance (patients 67%, controls 17%), sociability (patients 58%, controls 17%), impulsiveness (patients 54%, controls 17%), and altruism (patients 33%, controls 4%). Very high scores (deciles 9 and 10) in the dominance subdomain were more frequent in patients (33%, controls 8%). Six months after surgery, school performance, impulsiveness and altruism had not changed significantly, but fewer patients scored low on sociability (38%, with a slight increase to 25% for controls). Although the very high percentage of low scores for autonomy in patients decreased to 58, this was still very high and greater than that for controls (21%). Very low scores in the impulsiveness subdomain increased in patients who had not become seizure-free ($n=6$) and decreased in seizure-free patients ($n=18$).

Conclusion. For all domains and subdomains, a subgroup of children with medically intractable epilepsy were shown to have normal functioning. However, dominance was high, and autonomy, school performance, sociability, impulsiveness, and altruism were poor in children with medically intractable epilepsy

and remained so after surgery. Seizure freedom seemed to have a positive effect on impulsiveness. Few patients changed from normal to low deciles after surgery. Hence, epilepsy surgery had no additional negative effect on personality. Except for school performance, all marked subdomains concerned the interaction between the child and his/her environment. Whether the tendency for improvement in some subdomains continues will be reported after completion of the two-year follow-up. The contribution of the role of the child's environment to these improvements should be studied.

30. Visual habituation as an index of memory in very young children before and after surgery

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Introduction. Research in adults and older children has demonstrated that epilepsy surgery may affect memory and learning. However, almost nothing is known about the effects of epilepsy surgery on memory and learning in very young children, probably partly due to the difficulty of memory assessment in these children. Experimental research frequently employs visual habituation paradigms to probe memory functions in infants and toddlers. Visual habituation refers to the decrease in looking time (attention) when a stimulus is presented again. Habituation is thought to reflect encoding, *i.e.* the storing of an internal representation of the stimulus. We developed a method to measure visual habituation in order to investigate memory in infants and toddlers who undergo epilepsy surgery.

Methods. Twenty computer-manipulated face photographs were presented on a computer monitor for 20 seconds each, as ten pairs with 200 ms between the images of each pair. The child was seated in front of the monitor on a suitable chair or on the caregiver's lap and allowed to look or not look (freely) at the photographs. Using this method, we found significant habituation in a reference sample of 23 normally developing children (mean age: 21 months [2-43 months]). In the context of a larger longitudinal, controlled study, the method was administered to 12 epilepsy surgery patients with mental age ≤ 36 months (mean age: 25 months [5-48 months], 10 with hemispherectomy, 1 with temporal resection, 1 with central resection) and 11 normally developing children (mean age: 17 months [6-29 months]). Because there was no relation between habituation and gender, calendar age, developmental

quotient or mental age in the reference sample, we combined these additional controls with the reference sample for increased power in further analyses.

Results. Prior to epilepsy surgery, patients habituated significantly less than healthy controls. Patients did not improve after surgery. In fact neither before nor after surgery could significant habituation be demonstrated in patients at the group level. This appeared mainly to be due to lack of habituation in the most delayed patients, and in fact relative development (developmental age compared to calendar age) significantly predicted habituation in patients, with the highest habituation scores in those with the least delayed development.

Conclusion. Visual habituation is reduced in infants and toddlers with intractable epilepsy and does not improve after surgery. This is in line with research on general cognitive development after hemispherectomy. In patients with severely delayed mental development, little or no habituation was found both before and after surgery, whereas patients with less delay appeared to show at least some habituation, again independent of surgical status. Assuming that habituation is indeed an index of stimulus encoding, this suggests that deficient habituation plays a role in the emergence and perpetuation of delayed cognitive development in these children.

31. Cognitive improvement after epilepsy surgery in children under 3 years

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Introduction. Epilepsy surgery in children under 3 years is effective in reducing seizure frequency and is well tolerated. However, in most of the children, cognitive improvement is mild as their development is maintained on the same trajectory, although some children may have a dramatic improvement in their development during the years following surgery. In this report, we have tried to specify the main characteristics of these patients.

Methods. Seventy-five children were followed before and after epilepsy surgery (performed in several epilepsy centres), between 1995 and 2010. Their charts were collected retrospectively (clinical and cognitive evolution, EEG, radiology, and anatomic-pathology).

Among them, 13 children had resective surgery between 2 months and 2.6 years. In these children, developmental trajectory was different and the children could be divided into two groups in order to compare their characteristics. In group 1 (7/13), mental deficiency was observed and there was no significant change between preoperative and postoperative DQ. In group 2 (6/13), normal or subnormal cognitive development was observed with postoperative DQ highly superior to the preoperative DQ.

Results. In group 1 (7/13) there was little cognitive improvement; the epilepsy was focal and associated with an epileptic encephalopathy in four cases. The preoperative DQ ranged from 20 to 50. The aetiology was a malformation of cortical development (MCD) in all the cases: hemimegalencephaly, multilobar dysplasia, and tuberous sclerosis complex (TSC) with dysplasia. The surgery was performed from the age of 4 to 25 months with a delay varying from 2 to 19 months. In all the children, the cortical resection or functional hemispherotomy was incomplete and 5/7 children received surgery again, successfully. The Engel status was II or III after the first surgery, except for one case where the first surgery was successful, with a larger brain malformation. In group 2 (6/13), there was high cognitive improvement; the epilepsy was focal and associated with an epileptic encephalopathy in three cases. The presurgical DQ ranged from 20 to 50 (70 in one case). The aetiology was MCD in all the cases; focal dysplasia or DNET, TSC with dysplasia, and posterior megalencephaly. The surgery was performed from the age of 2 to 30 months, with a delay varying from 2 to 21 months. For all the children, it was successful with a complete focal resection in 5/6 cases and an early hemispherotomy in 1/6 (4 months). The Engel status was I after the surgery and the treatment was withdrawn in 5/6.

Conclusion. In this small population, we could observe a good cognitive prognosis in nearly half of the patients, a relatively high rate when compared to similar series of young patients. The good cognitive prognosis does not seem to correlate with the initial cognitive status, nor the severity of the epilepsy, as it may be observed for older children. It appears to be more closely linked to the aetiology (focal dysplasia or DNET), to the early successful resective surgery, and to the shorter duration of epilepsy.

32. Memory outcome after temporal lobe resection in childhood

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Objective. Temporal lobectomy (TL) is a successful surgical treatment for patients with intractable temporal

lobe epilepsy (TLE). So far, little is known about the long-term outcome of memory functions after TL in children.

Methods. Forty-two patients were followed after temporal lobe surgery (25 left [L-TL] and 17 right-sided [R-TL]) for developmental tumours ($n=16$) or hippocampal sclerosis ($n=26$). In addition, we assessed 11 patients with lesional TLE who underwent the same presurgical evaluation but did not undergo surgery (non-TL control group). Mean age at surgery was 13.8 years in the TL group, and average follow-up time was 9 years (minimum 5 years) in both groups. Memory function was assessed using the Wechsler Memory Scale Revised ([WMS], pre- and postoperative) and the Doors and People Test ([DPT], postoperative only), Semantic memory was measured using a composite of vocabulary, information and comprehension scales on the Wechsler Adult Intelligence Scale ([WAIS], postoperative only). Structural MRI data were used to determine the volumes of remaining temporal neocortex and hippocampus.

Results. Pre- to postoperative comparisons showed gains in WMS verbal memory (immediate and delayed) scores in the R-TL group, while L-TL did not differ from non-TL controls. Visual memory scores (immediate and delayed) improved in the L-TL, and remained unchanged in R-TL and non-TL controls. Gains were independent of changes in IQ reported previously (Skirrow *et al.*, 2011). Comparison of postoperative DPT scores showed an interaction ($p=0.030$) of side of surgery and task (verbal, visual). Post-hoc differences in verbal memory (covariate IQ) were found between L-TL and non-TL controls, while visual memory did not differ between groups. There were no group differences in DPT recall or recognition performance. Partial correlation analyses of postoperative memory scores and temporal lobe volumes showed robust associations (covariate IQ) between volume of the anterior/inferior temporal lobe and semantic memory in the L-TL group only ($R=0.68$, $p<0.001$), while the volume of the residual left hippocampus predicted delayed verbal recall at follow-up ($R=0.72$, $p<0.001$). No significant correlations were found for the R-TL group.

Conclusion. At the group level, temporal lobectomy in childhood resulted in improved memory functions which are presumed to be subserved by the unoperated temporal lobe. At the individual level, the extent of resection of the left anterior temporal lobe predicted semantic memory scores at follow-up. The volume of the residual left hippocampus predicted verbal episodic memory, similar to findings in adults (Baxendale *et al.*, 2000).

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33. Asymmetry of planum temporale predicts language lateralisation in children with left-sided focal epilepsy

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Introduction. The proportion of atypical language lateralisation, *i.e.* language being either bilateral or right-lateralised, is increased in patients with early-onset left-sided lesional epilepsy compared to healthy controls. We investigated potential clinical and neuro-anatomical predictors of functional reorganisation resulting in an atypical pattern of language representation.

Methods. Fifty-one children (mean age: 13.3 years; 53% female; 14 non-right-handed) with drug-resistant lesional focal epilepsy and 36 healthy controls matched for age and gender (5 non-right-handed) underwent fMRI using a covert verb generation task. Images were analysed using SPM8. The fMRI-laterality index within Broca's region was correlated with clinical predictors and neuroanatomical measures including the volume of temporal lobe structures and the length of the planum temporale. Furthermore, the influence of the location of a lesion was analysed using Voxel-based Lesion-Symptom Mapping (VLSM) analysis.

Results. Atypical language lateralisation was seen in 19 of the 51 patients (37%) and in 4 of the 36 controls (11%). Atypical language lateralisation in epilepsy patients was associated with several clinical and brain structural measures, including handedness, location of seizure onset, lesion volume and location (all $p<0.05$). Asymmetry of the planum temporale was the strongest predictor of atypical language laterality in patients ($R=0.70$, $p<0.0001$) accounting for 48% of the variance, while handedness accounted for a further 5% in a final regression model.

Conclusion. Asymmetry of the planum temporale is not related to language lateralisation in healthy children but may indicate the potential for inter-hemispheric language reorganisation in the presence of a left-sided epileptogenic lesion.

34. Cognitive development from two to ten years after paediatric epilepsy surgery

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Introduction. Pharmacoresistent epilepsy in children has been shown to affect cognitive development over time and early surgical intervention may have the potential to reduce the severity of cognitive impairment. Neuropsychological assessment is therefore an important part of the pre- and postoperative work-up in order to investigate the cognitive consequences of epilepsy surgery in children. Most follow-up studies focus on cognitive outcome one to two years after surgery, which is a short time when the neurodevelopmental aspect is taken into account. The aim of this study was prospectively to investigate the development of cognitive functions and the sustainability of seizure control between two and ten years after epilepsy surgery in children.

Methods. Twenty-one children and adolescents underwent epilepsy surgery in 1998-2000 at Sahlgrenska University Hospital in Gothenburg. Twenty had an extensive neuropsychological assessment before and two years after surgery. One had repeat surgery before the 10-year follow-up and 2 refused to participate. Seventeen individuals were prospectively included and followed for two and ten years after epilepsy surgery. The age at surgery was 4 to 20 years and the age after follow-up 14 to 30 years. Normal intelligence defined as full-scale IQ over 69 was found in 10, mild learning disability defined as full-scale IQ between 50 and 69 was found in 2, and severe learning disability defined as a full-scale IQ of <50 was found in 5 children. A comprehensive neuropsychological test battery of intelligence, learning and memory, working memory, attention and executive functions was part of the presurgical evaluation and was repeated two and ten years after surgery.

Results. IQ was stable between two and ten years. Learning capacity improved with a greater increase in verbal than figurative learning. Verbal memory improved in half of the subjects and declined in half, whereas figurative memory declined in most patients. Working memory improved, as did attention regarding sustained attention and impulse control. In contrast, reaction times were longer and the auditory attention span was shorter. Executive functions were not affected. Six subjects were seizure-free at ten years of follow-up and a seizure reduction of more than 75%

was achieved in 13. Seizure control improved in 5 and seizure recurred in 2 subjects between two and ten years of follow-up.

Conclusion. This study indicates that IQ remained stable between two and ten years after epilepsy surgery in children and that learning capacity may improve as well as working memory and sustained attention. Memory, especially figurative memory, frequently declined. Seizure control may improve further, but a recurrence of seizures may occur.

35. Preliminary results from a study of changes in attentional function in a selected group of children in the Danish epilepsy surgery program

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Aim. The aim of the study was to investigate possible changes in attentional function in children who undergo epileptic surgery.

Method. The sample consisted of 14 patients who had undergone epileptic surgery and who were evaluated pre- and postsurgically by a neuropsychologist. This was a retrospective study of Danish children in the age group 7-14 years ($n=14$) who underwent epileptic surgery in 2006-2009. Five children had an IQ above 80 (group A) and 9 children had an IQ of 79 or below (group B). The mean age of children in group A was 12.1 years and in group B the mean age was 10.5 years at the time of surgery. All children had undergone surgery in Denmark and had partial or single lobe resections. Measures and descriptions of attentional functions were registered presurgically and one and two years after surgery. The results were obtained by a parent interview and in connection with postsurgical neuropsychological testing.

Results. Seizure-free outcome (Engel 1A or 1B) was found in 80% of group A and 55% of group B at two years of follow-up. The parents of the children in group A reported attentional functions as being the same as before surgery and test results supported the parent assessment. Results point to no increase of attentional functions in the majority of group A. Test results in group B point to an increase of attentional functions in 55% ($n=5$) of the patients while 45% ($n=4$) of the patients appeared to have the same level of attentional functions before and after surgery. In 22% ($n=2$) of the cases, the parents reported better attentional functions while testing showed no increase.

Conclusion. Results indicate that children who have undergone epileptic surgery have some chance of improved attentional functions.

36. Accompanying patients with epilepsy to have surgery abroad

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Aim. To present a study of professional and lay accompaniment of patients with epilepsy and their parents referred abroad due to inexistent quaternary epilepsy care in Slovenia (2 million inhabitants), in collaboration with foreign epilepsy centres.

Methods. Historical overview and assessment of the project.

Results. Since the start of the project, there have been about 60 referrals abroad for epilepsy surgery for Slovenian patients with epilepsy, which started in childhood (most often children, adolescents, rarely also young adults), and their parents. Conceptualised and initiated by professionals (specialists in paediatric epileptology and neuropsychology) at the Centre for Epilepsy, Department of Child Neurology, Division of Paediatrics, UMC Ljubljana in the late 1980s as a financially unsupported activity, the project later expanded and diversified, including participation of lay voluntary accompanying persons, with a modest funding from

a non-governmental source (via the Slovenian Chapter of ILAE). Finally, a contribution from the NHI made it possible to provide professional accompaniment (by a junior neuropsychologist or medical doctor) for diagnostic procedures (especially during Wada testing and phase II recordings, stimulations), where a professional speaking the young patient's mother language is required to be present. Advantages and shortcomings of the project have been assessed with a very favourable general estimate and it has become possible to offer young patients access to excellent epilepsy surgical management, especially in cases where, due to social and cultural reasons, this could not have been possible. Collaborative activities have stimulated better quality of care in the tertiary unit. The experience was later also successfully developed in the same way by the adult tertiary epilepsy centre of the UMC Ljubljana, in collaboration with the national ILAE Chapter.

Conclusion. The project has proved to be an important element supporting smooth collaboration between Slovenian tertiary and foreign quaternary epilepsy centres, creating a good example of cross-border professional mobility, international collaboration, and better service to the patients. The experience, so far thus obtained, in blending professional and non-governmental inputs has proved successful and has been considered worthy to be continued.