Pediatric Epilepsy Surgery (II): Surgical Strategies and Related Issues

Pediatrik Epilepsi Cerrahisi (II): Cerrahi Yaklaşım Stratejileri ve İlgili Hususlar

OĞUZ ÇATALTEPE

Institute for Neurology and Neurosurgery, Department of Neurosurgery
Beth Israel University Hospital, New York, NY, U.S.A.

INTRODUCTION

Children with intractable epilepsy constitute a special subgroup among epilepsy patients because of distinct challenges characteristic for this age. These age-specific characteristics, pre-surgical assessment techniques and selection criteria of surgical candidates were discussed in the first part of this review. Here, we will review special aspects of surgical approaches in this patient group. There are some critical questions which should be addressed by medical/surgical team and the patient's family before proceed for a surgical intervention: what is the chance for seizure control after surgery? what are the risks of surgery? what are the risks of not having surgery? what is the natural course of the underlying disease? what are the other medical management options? what are the surgical options? and finaly, what is the goal of the surgery? Sometimes, there are no straightforward answers to some of these questions. However, these questions should be discussed with the patient and family using all available data.

We strongly believe that pre-surgical and surgical approaches to children with intractable epilepsy should be tailored based on individual cases. Even among children, indications and surgical techniques are often different in infants/young children, preadolescent and adolescent groups. Therefore, this review is not a search for a general guideline for surgical treatment of epilepsy in children, but an effort to document and to discuss currently available approaches and techniques.

STRATEGICAL ASPECTS OF SURGICAL APPROACHES

One of the main challenges in epilepsy surgery arises from overlapping eloquent cortex and epileptogenic zone (7,8,14,40). The goal in surgery is to obtain optimal resection of epileptogenic tissue while preserving eloquent cortex. Although, there is no perfect solution for this problem, epilepsy surgeons frequently need to address this issue. There are two main options: removing epileptogenic zone along with a significant amount of eloquent cortical tissue or preserving whole eloquent cortex while removing much less epileptogenic tissue. The price of the first approach is causing new neurological deficits while having satisfactory seizure control, and the price of second approach is causing no new neurological deficit but no satisfactory seizure control neither. This theraupatic dilemma is not rare

especially in pediatric epilepsy surgery and challenge is to find optimal balance.

First step in surgical planning is to define sharper boundaries of epileptogenic zone and eloquent cortex. This is only possible by obtaining extensive data using all available cortical mapping tools. All data should be assessed and discussed throughly by an experienced pediatric epilepsy surgery team. The goal is to create a sharper cortical map of functionally important areas as well as safe areas for resection and to obtain a sharper map of epileptogenic zone (34). Epilepsy surgery team must also try to determine potential risks of surgical resection, to predict long-term recovery chance of possible functional loss and potential contribution of brain plasticity to recovery. In addition, other surgical techniques such as disconnection techniques and multiple subpial transection procedure should be considered based on available data. After reaching an agreement about surgical plan, pediatric epilepsy surgery team should discuss available data and surgical plan, including all potential risks and benefits of surgical approach, with the family and the patient, if he/she is mature enough. All other management options from most radical resection strategies to non-surgical approach should also discussed with them, and decision for a surgical intervention should be made together. The patient's family should be informed about available data regarding mortality and morbidity risks of planned surgical procedure. Although, perioperative mortality is not a high risk (1.2%-2) in this patient group, it still is most serious issue to be discussed (38,66). Pediatric epilepsy cases, especially infants, are surgically more vulnerable patients because of small blood volume, risk of hypothermia, anesthesiology complications and relatively more frequent indications for extensive surgical interventions such as multilobar resection and hemispherectomy (8,19,20,37).

Surgical approaches in epilepsy can be reviewed roughly in two groups: resective surgeries and other techniques, including corpus callosotomy, multiple subpial transection and vagal nerve stimulation. Here, we will briefly review each surgical technique and related complications.

Cortical Resection:

Temporal Lobe Resection: As I emphasized above, temporal lobe epilepsy has been encountered less frequently in children than adults and especially mesial temporal sclerosis is very uncommon in early childhood. Even temporal lobe lesions of children are less frequently localized to mesial temporal region comparing to neocortical involvement (10,18, 23,26,32,63). The plan for temporal lobe resection is determined individually for each patient based on EEG and neuroimaging results. Surgical approach might be tailored or standard anterior temporal lobectomy with/without amygdalohypocampectomy; selective amygdalohippocampectomy; pure lesionectomy or lesionectomy plus removal of epileptogenic area. The most frequent complication of temporal lobectomy is homonymous superior quadrantanopia. This complication is predictable to some extent if the length of the resection from the temporal pole is taken into account. Speech difficulties and memory dysfunction may also occur after temporal lobectomy on dominant side. Contralateral hemiparesis or hemiplegia is a rare complication and most likely to arise from a choroidal artery related vascular damage (15,28,42). Available data is variable about exact incidences of these complications in childhood. There is very rare mortality in temporal epilepsy surgery series. Polkey reported hemiparesis in 4.2% of patients with significant recovery (54). Exact percentage of visual field defects is not certain because of difficulty to judge in children, however superior quadrant defects were reported in 20-40% of the patients (18,26,35,45,55,57,63). It has been noted that school performances of children typically improve after temporal lobe resection (18).

Extratemporal Resection: Extratemporal seizures are much common in infants and younger children and they may cause devastating, very frequent and intractable seizures. Therefore, extratemporal resections constitute most common epilepsy surgery procedure among infants and very young children. Extratemporal epilepsy characterized with earlier onset and high frequency daily seizures. Developmental abnormalities, especially cortical dysplasia, are very common pathologic findings in pediatric epilepsy patients. This also presents a significant diagnostic and surgical challenge because of understated neuroimaging findings of these cases. Locating a seizure focus in an extratemporal region is one of the most challenging tasks for a clinical neurophysiologist. In addition, large cortical surface area, very widespread and functional neural pathways permitting very fast spread of discharges within and outside of a lobe complicates the challenge more. Therefore, extratemporal epilepsy

cases more frequently need invasive EEG monitoring, functional neuroimaging studies and extensive work-up for better definition of borders of epileptogenic zone as well as eloquent cortex. The complications of extratemporal epilepsy surgery are mostly related to functional characteristics of resection area. There are a few pediatric epilepsy surgery series regarding the complications of extratemporal epilepsy surgery cases (1,2,24-26,37,46,57). Unfortunately, these series mostly report combine complication rates in both extratemporal and temporal cases. The complication rates are between 12 and 50% in these series and vary based on resection site. We reviewed our pediatric extratemporal surgery cases (n:51) which were operated at the Cleveland Clinic between 1991 and 1996 (8). The neurological complications in this series include increased or new motor deficit in 8 patients, sensory deficit in 3 patients and visual field cuts in 6 patients. In general, the type of extratemporal complications closely related to resected anatomical araes such as paracentral cortical area, supplementary sensorimotor area (SSMA), remaining frontal lobe, parietal lobe and occipital lobe resections. Resection of hand sensory-motor cortex cause severe, permanent fine motor deficit and abnormal position sense, whereas resection of motor leg area leads to flaccid leg paralysis with a reasonable chance of incomplete recovery. Resection of non-dominant primary face area does not cause any significant deficit, because of the bilateral innervations of facial muscles. However, the feasibility of extensive resection of dominant face motor cortex is controversial. Some authors reported postoperative dysarthria in these cases, especially if adjacent opercular area was removed (27,40,44,48). Resection of supplementary sensorimotor area leads to transient contralateral neglect and/or weakness, mutism in various severity without cognitive impairment, diminished spontaneous movements and apraxia. Post-operative findings resolve within days or weeks with no gross deficit at long-term follow-up. The intensity of deficits are much less in children after the surgical intervention in SSMA (8,14,27,33,40-42,44,52,56). Resection of Broca's area is only feasible using slow, staged removal of premotor opercular frontal cortex and with frequent intra-operative language testing under local anesthesia (4). Transient motor dysphasia may be seen if resection is extensive (8). Non-dominant parietal lobe may be removed extensively without any clinically significant sensorimotor deficit except a small risk (0.5%) of hemiparesis (44,48). However, Olivier recommends that parietal resections should not exceed intraparietal sulcus as inferior limit of the resection (53). Angular gyrus (Wernicke's area) should be respected to avoid postoperative aphasia and alexia (30,44,48,50,56). If resections are carried into the underlying white matter of parietal operculum, contralateral lower quadrantic or hemianopic visual field defect can be seen. Occipital resections may produce contralateral homonymous hemianopsia and language disorders in the dominant hemisphere (8).

Hemispherectomy:

Hemispherectomy is the procedure of choice for patients with Rasmussen's encephalitis with progressive hemiparesis, Sturge-Weber syndrome, hemimegalencephaly, hemispheric atrophy with infantil spasms and infantile hemiplegia syndromes (16,22,29,30,49,50,58,61,62). These patients generally have frequent, severely debilitating, medically intractable seizures arising from a structurally abnormal hemisphere and originating from multiple epileptogenic foci which are too many and/or too extensive for focal/lobar excision. Therefore these patients are not candidates for a more restricted surgical resection. The typical surgical candidate is a patient who already has hemiparesis with absent fine finger and foot tapping movements, but with preserved shoulder, elbow, wrist, and some finger flexion, and with ability to walk. In addition other hemisphere should be structurally and functionally normal. In good selected patients, hemispherectomy is very gratifying procedure by yielding complete or near complete seizure control (up to 85%) with no new neurological deficits. It should be noted that although seizures are decreased significantly, developmental gains are relatively poorer in hemimegalencephaly cases. This is most likely because of dysfunctional non-megalencephalic hemisphere. Hemispherectomy modalities are anatomical or functional hemispherectomy and hemidecortication. Original anatomical hemispherectomy included en bloc removal of the entire hemisphere down to basal ganglia. However, serious early and late onset complications were reported in the 70s, such as superficial hemosiderosis caused by numerous acute/chronic hemorrhages from the fragile capilleries within a subdural membrane, obstructed hydrocephalus, bleeding into the hemispherectomy cavity, or brain stem shift with progressive neurological deterioration. Several previous studies with long term follow-ups (up to 20 years) documented the incidence of these complications as 17-35% with high mortality rates. However, much better complication rates were

reported at more recent series (16,22,29,30,50,56,58-62). Functional hemispherectomy was developed by Rasmussen to overcome these long-term complications. Functional hemispherectomy is an anatomically limited, but functionally complete hemispherectomy which aims complete functional disconnection of damaged hemisphere from remaining brain. Functional hemispherectomy includes removal of pre- and post-central cortical regions, transection of all frontal, parieto-occipital white matter and corpus callosum, and disconnection or resection of the temporal lobe including amydala and hippocampus (62,49,59,60). Villemure et.al. reported 54 functional hemispherectomy cases with early hydrocephalus in 7.4% of the patients and mortality rate of 3.7 percent (60). Hemidecortication consists of removing whole cortical tissue of the hemisphere while preserving the white matter and not entering ventricle. Several series reported hydrocephalus in 20-33% of the cases and severe per-operative haemodynamic disturbances in these cases (16,50,59,60).

Lesionectomy:

Some patients have distinct lesions which cause epilepsy such as tumor and vascular malformation. In these cases, surgical decision must be based on not only concern of seizure control, but also potential behavior and natural history of the lesion. It should be confirmed that epileptogenic discharges originate from the lesion, and gold standart for this assessment is long-term non-invasive or invasive EEG-Video monitoring depend on the characteristics of the case. Main controversy is extent of the resection in these cases: removing just lesion without resecting surrounding brain tissue or extending resection area beyond the boundaries of the lesion. In general, accessibility of the lesion and its relation with eloquent cortex determine the extent of the resection. Available data shows better outcome in extratemporal cases with strict lesionectomy. However strict lesion resection is less effective in temporal lobe lesions. If there is any tumor involvement, active epileptogenic activity in mesial structures or hippocampal atropy, best strategy is resecting mesial temporal structures along the lesion. However, mesial structures may be preserved in a patient with pure lateral neocortical lesions. If tumor borders were not well defined, or epileptogenic area is much larger than tumor, temporal lobectomy is recommended. Borders of resection site may be tailored based on electrophysiological, neuroimaging studies or histopathologic criteria. Extended lesionectomy, which is resection of lesion with surrounding epileptogenic area, if functionally feasible, is our preference as in most epilepsy surgery centers (7,8,9,21,40,41).

Corpus Callosotomy:

Corpus callosotomy is not a definitive surgical intervention. The procedure does not aim to eliminate seizure focus. The purpose of surgery is to alter the propagation pathways of epileptic discharges and to prevent the spread of discharges to the opposite hemisphere. The surgical candidates typically have symptomatic generalized epilepsy arising from multiple areas bilaterally and present with multiple seizure types (atypical absence, myoclonic, tonic, atonic, tonic-clonic). The procedure is usually reserved for patients who are not candidates for resective surgery but severly disable with very frequent seizures, particularly"drop" attacks. There is no firm criteria that can be used to predict good outcome after corpus callosotomy. However, it has been believed that normal intelligence, focal EEG abnormalities, focal radiological abnormalities, the presence of generalized tonic-clonic, tonic and atonic seizures, and hemiparesis are good predictors for better outcome. Pre-surgical MRI should be assessed to see the presence, length and thickness of corpus callosum. The procedure can be performed with different techniques such as sectioning entire corpus callosum with or without sectioning anterior commisure and fornices, two stages complete corpus callostomy with/without cutting the anterior commissure, and one stage anterior one-half or two thirds corpus callosotomy. Although surgical approach should be decided individually, partial callosotomy, if it is effective in that group of patients, frequently preferred to one stage sectioning of whole corpus callosum. Several complications were described after corpus callosotomy. The patients with acute disconnection syndrome exhibit decreased spontaneity of speech which may range from simple initiating problems to complete mutism, variable degrees of paresis, forced grasping on the non-dominant side, incontinence and antagonism in basic manual tasks because of persistent antagonism between two hemispheres. Failure in recent memory function has also been noted in minority of callosotomy patients. Split-brain syndrome is related to the extent of callosotomy. Sectioning of splenium leads to deficits of tactile and visual transfer. Speech deficit usually improves, however permanent language disturbances may also occur (3,6,11,42,67) Cendes et.al. reported persistent postoperative

dysarthria, diminished speech output, and gait dyspraxia in 13 out of 34 callosotomy patients (11).

Multiple Subpial Transection:

MST is a surgical alternative for management of seizures arising from epileptogenic zones overlapping the eloquent cortex. The idea behind this surgical technique is to prevent lateral spread of the epileptogenic discharges over the cortex in an eloquent area while the columnar circuitry and central projections of the cortex, consequently normal physiological function of the brain tissue, are preserved. Although, this technique is not as efficient as resective surgery on seizure control, it has been used successfully in well selected and carefully mapped cases. In general, candidates for this procedure are the patients with Landau-Kleffner syndrome, epilepsia partialis continua originating from eloquent cortex, focal sensory-motor seizures secondary focal cortical dysplasia on eloquent cortex. Appropriate cortical areas for MST are pre-and post-central gyrus, Broca's and Wernicke's areas. The particular difficulty to choose appropriate surgical technique (cortical resection vs MST) in children arises from excellent synaptic plasticity and functional recovery potential of children frequently seen after resective surgery. The complications of this procedure are related to hemorrhage and direct injury of neuronal structures. The incidence of permanent morbidity, including hemiparesis, aphasia and visual field defects, was reported about 7% in addition to transient deficits in 7% of the children (36,42).

Vagal Stimulation:

Vagal nerve stimulation is relatively a new technique. It has been used with a reasonable degree of seizure relief in patients who are not good candidates for resective surgery. Although it is not certain, the technique probably works by globally reducing epileptogenicity in brain and locus seruleus plays a crucial role in this. Most common complications are persistent cough (15%), voice alteration (55%), dyspnea (13%), pain, paresthesia and headache. There were 2 deaths among 195 patients (17).

SURGICAL OUTCOME

Available data regarding overall surgical outcome of children with epilepsy shows comparable seizure control with adults. It appears that young age does not adversely effect good surgical outcome. One of the biggest series in children came from Montreal Neurological Institute and showed favorable outcome in 56/118 patients with a median follow up duration of 15 years (23). It showed that temporal lobe cases had better seizure free outcome after temporal lobe resection than extratemporal cases as in adults. Several recent studies provided much detailed overall outcome data which shows much better results in children (12,20,38,65,66). These recent series documented that 60 to 65% of infants had seizure free outcome while another 13 to 20% of patients had rare seizures. These rates are 59 to 67% and 11 to 19% respectively among preadolescent children and 69% and 20% among adolescents. These rates are variable secondary to type of epileptic syndrome and type of surgical procedure. Fish et. al. reported 73 pediatric temporal surgery patients from MNI with seizure free outcome in 34 patients (23). Another study from Oslo, Norway reported significant reduction (more than 95%) of seizure frequency in 60% of the children with temporal lobe epilepsy (26). Both studies also agree that lesional cases had better outcome than non-lesional cases in this group. Tuxhorn et.al. published another temporal epilepsy surgery series from Bethel, Germany (55). This series includes 36 patients with the mean age of 8. Seventy five percent of the patients had seizure free outcome and 11% had significant seizure reduction (>90%).

We reviewed extratemporal epilepsy surgery and hemispherectomy cases distributed from the age of 3 months to 18 years (mean 11.3 year old) (n: 66) in Cleveland Clinic (8). Fifteen of the patients underwent functional hemispherectomy and 51 patient underwent various extratemporal resection operations. In extratemporal resection group, 69.1% of the patients had Engel Class I outcome, 3.8% Class II, 11.5% Class III and 15.3% Class IV outcome. In hemispherectomy group, 75% of the patients were seizure free after the operation (Class IA), 12.5% were Class IIB and 12.5 % Class IIIA. Villemure et.al. reported similar results with 75% seizure free outcome rate in 54 hemispherectomy patients (60).

Blume reviewed several corpus callosotomy series (total 513 patients) and documented that outcome varies based on seizure types (6). Favorable surgical outcome was reported in 70% of children with drop attacks whereas this rate droped to 63% in tonic-clonic seizures and to 54% in absence and complex partial seizures. Morrell reported surgical outcome results in 99 patients who underwent

multiple multiple subpial transection (36). Fifty two percent of the patients were seizure free, 11% of the had rare seizures and 21% of the patients had seizured reduction more than 90% after MST. One of the largest long-term prospective studies regarding vagal nerve stimulation (n:195 cases) showed more than 50% seizure reduction in 35% of the patients and more than 75% seizure reduction in 20% of the patients at 12 months (17).

There is a limited data regarding epilepsy surgery in infants, however the results of this data shows that epilepsy surgery in infants may be gratifying in a group of well selected patients. The Cleveland Clinic reported 12 cases of infants (2.5-24 months of age) with focal cortical dysplasia, Sturge-Weber, hemimegalencephaly, and neoplastic lesions (64). They documented that six patients were seizure free (Engel Class I), three had rare seizures (Engel Class II) and two patients had worthwhile improvement (Engel Class III). No patient had a new neurological deficit after surgery. Several patients had marked "catch-up" developmental progress. In another series from UCLA, 15 patients were reported. Sixty five percent of the patients were seizure free, 13% had seizure reduction more than 90% in seizure frequency (Engel Class II (64,66). Chugani et.al also reported 23 patients with intractable infantile spasms (13). These patients underwent cortical resection (n:15) and hemispherectomy (n:8) and 15 patients became seizure free, 3 has 90% seizure control and one had 75% seizure control. Four patient did not have any benefit. Another important data from these studies show lower seizure free outcome in children with cortical dysplasia (52%) comparing to children with low grade tumors (82%) (65).

SURGICAL TECHNIQUE AND CONSIDERATIONS

Extensive pre-surgical assessment, right and well planned surgical approach, and appropriate surgical technique constitute keystones of good outcome in epilepsy surgery. Appropriate epilepsy surgery techniques for adults are also relevant for children. In addition, pediatric neurosurgery techniques and principles should also be followed strictly during epilepsy surgery procedures in children. Here, we also would like to emphasize some important technical points in pediatric epilepsy surgery cases.

It is recommended to make a large craniotomy in resective surgeries to have better exposure and access to critical anatomical landmarks and functionally important areas such as post-central gyrus and parieto-temporal language areas. This will help to assess satisfactorily the borders of resection site and to apply electrical stimulation and mapping techniques if necessary (27,41,44,48,50,56).

Intraoperative ultrasonography and especially frameless stereotaxy became part of the standard armamentarium of pediatric epilepsy surgery cases. Frameless stereotaxy is especially important in corpus callosotomy cases for pre-operative localization of bridging veins and determining full extent of resection during surgery. Frameless stereotaxy is also very helpful for resective surgery in paracentral and central cortical areas. Finally, its place in hemispherectomy cases is particularly important if surgery has been performed through narrow exposures. Frameless stereotaxy especially critical in hemimegalencephaly cases which have very distorted anatomy and small ventricles.

Subpial resection technique is the main surgical technique to avoid the complications in the epilepsy surgery. This technique helps to recognize the anatomical location, extend and depth of the various sulci and gyri in the resection area at every stage of the surgery. It also provides excellent anatomical guidance to remove pre-determined cortical area without disturbing the vascular supply of adjacent cortex. The endopial resection technique provides excellent hemostasis while resecting the tissue and prevents development of meningo-cerebral scar tissue and distal ischemic insult. The surgeon selectively protect the pia by peeling it from cortex and/or aspirating cortical tissue without perforating or disturbing the pia and pial vessels. Thanks to endopial resection technique, all arteries, veins, sinuses and cranial nerves around the resection area are kept subpially throughout the surgery and left intact. The most helpful surgical tool in subpial dissection of the cortex is a low setting aspirator or ultrasonic dissector set at a very low parameter of suction and vibration (8,15).

Stripping the brain and pia-arachnoid from sinuses, especially during parietal and occipital resections, can cause postoperative bleeding and hydrocephalus secondary to decreased CSF absorbtion. We use strictly suppial resection technique in these cases and leave the pia-arachnoid membranes attached to sinuses. Protecting all vascular structures and vascular supply of surrounding cortex is another important surgical

principle. Especially, the watershed territory between the anterior and middle cerebral arteries should be protected from any surgical damage as much as possible and surgeon should not leave any devascularized tissue behind in this area to prevent postoperative swelling which may be catastrophic.

Another critical issue for a successful surgical resection is determining the borders of functional areas. Best tools for this purpose are using intra- or extra-operative cortical stimulation techniques, evoked potential responses, functional neuroimaging studies and stepwise resection of eloquent cortex in an awake patient by continuously testing sensory-motor functions during resection. Recognizing central artery when it exit from the sylvian fissure has central importance during surgery of this region. It can be recognized easily by its typical localization. The central sulcal artery exits from sylvian fissure and makes a loop over the central operculum before penetrating deep into the lower part of the central sulcus. Utmost care must be taken to protect central artery and all other vascular structures in Rolandic, precentral and postcentral sulci during the surgery.

Resection of eloquent cortex is a challenging task. Beforehand knowledge of potential deficits is important. Therefore, meticulous mapping has critical importance for resection on and around motor cortex, especially mapping of tongue, thumb and lip areas. One of the biggest challenges during paracentral resection in children is identification of central region. Classical anatomical landmarks, although helpful, may not be always reliable. Frequently, tubers and dysplastic lesions distort paracentral area. In these cases, using SSEP/MEP techniques has critical importance to determine central sulcus by phase reversal signal and to evoke specific motor responses. Postoperative language dysfunction can be seen if adjacent pial banks, underlying white matter or ascending vascular supply are manipulated or if opercular areas are removed in this region. (24,50,56). The surgeon also should not extend the resection boundaries at motor cortex to the point of 2-3 mm below the lowest elicited thumb response, if the patient has no arm weakness (50). Stimulation studies and mapping of speech areas may be challenging. At times, producing speech interference with stimulation in Broca area may be very difficult. Olivier (40) as well as Rasmussen (48) emphasized that speech interference with stimulation in the Broca area is often negative with currents well above sensory and motor thresholds and a negative response does not necessarily indicate absence of function. Therefore, the posterior 2.5cm of inferior frontal gyrus or at least three opercular gyri in front of the lower end of the precentral gyrus must be preserved to avoid speech disturbances (40,48). In addition to resection of this area, venous occlusion of ascending frontal veins draining into the superior longitudinal sinus in the dominant hemisphere may cause transient motor dysphasia as well. Children have significant advantage for recovery after resection of eloquent cortex because of neural plasticity in the developing brain. The same mechanism is also important in tumoral cases. If the tumoral mass is in opercular region in small children, it can be expected having an already moved speech area. This should be kept in mind during mapping studies.

Even if cortical tissue is preserved, manipulation or removal of underlying white matter may produce significant deficits. Therefore, utmost care not to compromise functionally critical axonal tracts should be taken during the resection of cortical tissue.

As we emphasized before, children with epilepsy have some specific patologies such as Sturge-Weber, hemimegalencephaly, infantile hemiplegic syndromes. These patologies have some characteristics which should be aware of to avoid trouble during surgery. The cortex of a Sturge-Weber patient generally has abundant tortuous capilleries and small veins. These vascular structures may be very thick at times, and may constitute several layers deep in subarachnoid space and may penetrate into cortex and subcortical white matter with accompaniment of calcifications. In addition, some large, tortuous veins frequently in unusual locations can be seen in these patients. Non-filling anterior sagittal sinus, thrombosed branches of middle cerebral artery or simply an area of avascularity is also not uncommon in these cases. The venous drainage may radiate toward the supependymal venous network, especially toward the superolateral ventricular edges. From surgical point of view, this extensively abnormal vascular anatomy can cause serious problems during surgery (4,30,47). Therefore, it is an absolute necessity to do adequate preoperative vascular evaluation and to use appropriate resection techniques in these cases.

Another peculiar and rare developmental pathology is hemimegalencephaly characterized by congenital hypertrophy with thickened, abnormal cortex of one hemisphere (53,54).

Hemimegalencephalic brains had poor nutrition, abnormal vascular supply and AVM-like vessels which can cause heavy bleeding problems and cardiac failure during surgery. Unusual anatomy and absence of anatomical landmarks might also cause serious orientation problems during these operations. Adequate pre-operative vascular work-up and detailed structural neuroimaging studies should be obtained and a meticulous surgical planning should be done based on this data. Frameless stereotaxy technique provide great help during surgery.

CONCLUSION

Epilepsy surgery in childhood is a safe and effective therapy with well selected cases and adequate surgical technique by an experienced surgical team. The outcome in seizure control is similar to that obtained in the adults. It may be a definitive treatment not just for controlling seizures but also for preventing their long-term harmful effects. Focal resective surgery is the procedure of choice if there is a focal zone of epileptogenic onset and/or zone of cortical abnormality concordant with seizure semiology and if epileptogenic zone is amenable for resection. Hemispherectomy is the procedure of choice if there is multifocal or generalized seizure onset limited only one hemisphere with associated significant contralateral hemiparesis. If resective surgeries are not appropriate, then corpus callosotomy, multiple subpial transection and vagal nerve stimulation should be considered. Satisfactory outcome can only be obtained using a team approach by an experienced pediatric epilepsy surgery group. Extensive and throughly assessed pre-surgical evaluation data, well planned surgical approach and strict adherence to appropriate epilepsy surgery and pediatric neurosurgery technics and principles are critical determinants of success in pediatric epilepsy surgery.

REFERENCES

1. Adelson PD, Black PM, Madsen JR et.al.: Use of subdural grids and strip electrodes to identify a seizure focus in children. Pediatr. Neurosurg 22;174-180, 1995
2. Beckung E, Uvebrant P, Hedstrom A, Rydenhag B: The effects of epilepsy surgery on the sensorimotor function of children. Dev Med and Child Neurol 36;893-901, 1994
3. Benbadis SR, Wyllie E: Evaluation of intractable seizures in children. In Albright L, Pollack A, Adelson D (eds), Principles and Practice of Pediatric Neurosurgery, New York: Thieme Med Pub, 1999;1095-1109
4. Bentson JR, Wilson GH, Newton TH: Cerebral venous drainage pattern of theSturge-Weber Syndrome. Radiology 102;111-118, 1971
6. Blume WT: Corpus callosotomy: a critical review, in Tuxhorn I, Holthausen H, Boenigk H (eds.), Pediatric Epilepsy Syndromes and Their Surgical Treatment, London: John Libbey, 1997;815-829
7. Cascino GD, Sharbrough FW, Trenerry MR et.al.: Extratemporal cortical resections and lesionectomies for partial epilepsy: complications of surgical treatment. Epilepsia 35; 1085-1090, 19948
8. Cataltepe O, Comair Y: Complications of extratemporal epilepsy surgery in infants and children, in Tuxhorn I, Holthausen H, Boenigk H (eds.), Pediatric Epilepsy Syndromes and Their Surgical Treatment, London: John Libbey, 1997;709-725
9. Cataltepe O, Comair Y: Focal resection in the treatment of neuronal migrational disorders, in Kotagal P, Luders HO (eds), The Epilepsies: Etiologies and Prevention, San Diego: Academic Press, 1999;87-92
10. Cataltepe O, Comair Y: Lesional temporal lobe epilepsy series from Cleveland Clinic (Unpublished data)
11. Cendes F, Ragazzo PC, Da Costa V, Martins LF: Corpus callosotomy in treatment of medically resistant epilepsy. Epilepsia 34;910-917, 1993
12. Chugani HT, Shewman DA, Peacock WJ et.al.: Surgical treatment of intractable neonatal-onset seizures: the role of PET. Neurology 38;1178-1188, 1988
13. Chugani HT, Shewnon DA, Shields WD, Sankar R, Comair Y, Vinters HV, Peacock WJ: Surgery for intractable infantile spasms: neuroimaging perspectives. Epilepsia 34;764-771, 1993
14. Comair YG, Hong SC, Bleasel AF: Invasive investigation and surgery of the SMA. Adv In Neurol 70;369-378, 1996
15. Comair YG, Choi HY, Van Ness P: Neocortical resections, in Engel J, Pedley TA (eds), Epilepsy, volume 2, Philadelphia:Lippincott-Raven, 1997;1819-1828
16. Davies KG, Maxwell RE, French LA: Hemispherectomy for intractable seizures: Long term results in 17 patients followed for up to 38 years. J Neurosurg 78;733-740, 1993
17. DeGiorgio CM, Schachter SC, Handforth A, Salinsky M, Thompson J, Uthman B, Reed R et.al.: Prospective long-term study of vagus nerve stimulation for the treatment of refractory seizures. Epilepsia 41;1195-1200, 2000
18. Duchowny M, Levin B, Jayakar P, Resnick TJ, Alvarez LA, Morrison G, Dea P: Temporal lobectomy in early childhood. Epilepsia 33: 298-303, 1992
19. Duchowny M: Epilepsy surgery in children. Curr Op Neurolog 8;112-116, 1995
20. Duchowny M, Jayakar P, Resnick T, Harvey AS, Alvarez L, Dean P, Gilman J, Yaylali I, Morrison G, Prats A, Altman N, Birchansky S, Bruce J: Epilepsy surgery in the first three years of life. Epilepsia 39;737-743, 1998

21. Fried I, Cascino GD: Lesionectomy, in Engel J, Pedley TA (eds), Epilepsy, volume 2, Philadelphia:Lippincott-Raven, 1997;1841-1850
22. Falconer MA, Wilson PJ: Complications related to delayed hemorrhage after hemispherectomy. J Neurosurg 30; 413-426, 1969
23. Fish DR, Smith SJ, Quesney LF et.al.: Surgical treatment of children with medically intractable frontal or temporal lobe epilepsy: results and highlights of 40 years experience. Epilepsia 34; 244-247, 1993
24. Goldring, S: Pediatric epilepsy surgery. Epilepsia 28 (Supp1);S82-S102, 1987
25. Green JR: Surgical treatment of epilepsy during childhood and adolescence: The Percival Bailey oration. Surg Neurol 8;71-80, 1977
26. Guldvog B, Loyning Y, Hauglie-Hanssen H et.al.: Surgical treatment for partial epilepsy among Norvegian children and adolescent. Epilepsia 35;554-565, 1994
27. Haglund MH, Ojemann GA: Extratemporal resective surgery for epilepsy. Neurosurg Clin of North Amer 4;283-292, 1993
28. Helgason CM, Bergen D, Bleck TP et. al.: Infarction after surgery for focal epilepsy: manipulation hemiplegia revisited. Epilepsia 28;340-345, 1987
29. Hendrick EB, Hoffman HI, Hudson AR.(1969): Hemispherectomy in children. Clin.Neurosurg. 16,315-327.
30. Hoffman HJ: Benefits of early surgery in Sturge-Weber syndrome, I Tuxhorn I, Holthausen H, Boenigk H (eds.), Pediatric Epilepsy Syndromes and Their Surgical Treatment, London: John Libbey, 1997;364-370
31. Holmes GL: Surgery for intractable seizures in infancy and early childhood. Neurology Suppl.(43)5;S28-S37, 1993
32. Holmes GL: Temporal lobe epilepsy in childhood, in Tuxhorn I, Holthausen H, Boenigk H (eds.), Pediatric Epilepsy Syndromes and Their Surgical Treatment, London: John Libbey, 1997;251-260
33. Laplane D, Talairach J, Meininger V et.al.: Clinical consequences of corticectomies involving the supplementary motor area in man. J Neurol Sci 34;301-314, 1977
34. Madsen JR, Adelson PD, Haglund MM: The future of pediatric epilepsy surgery. Neurosurg Clin North Amer 6;589-597, 1993
35. Meyer FB, Marsh WR, Laws ER,Sharbrough FW: Temporal lobectomy in children with epilepsy. J Neurosurg 64;371-376, 1986
36. Morrell F, Kanner AM, Whisler WW: Multiple subpial transection: application to pediatric epilepsy surgery, in Tuxhorn I, Holthausen H, Boenigk H (eds.), Pediatric Epilepsy Syndromes and Their Surgical Treatment, London: John Libbey, 1997;865-875
37. Morrison G, Duchowny M, Remnick T et.al.: Epilepsy surgery in childhood. Ped Neurosurg 18; 291-297, 1992
38. Munari C, Lo Russo G, Minotti L, Cardinale B, Tassi L, Kahane P, Francione S, Hoffman D,Benabid AL: Presurgical strategies and epilepsy surgery in children: comparison of literature and personal experience. Child's Nerv Syst 15;149-157, 1999
39. Ojemann GA: Individual variability in cortical localization of language. J Neurosurgery 50; 164, 1979
40. Olivier A: Extratemporal resections in the surgical treatment of epilepsy. In Spencer S, Spencer D (eds), Surgery for Epilepsy, Boston: Blackwell Scientific Pub, 1991;150-161
41. Olivier A, Awad I: Extratemporal resection. In Engel J Jr (ed), Surgical Treatment of the Epilepsies, New York: Raven Press, 1993; 489-500
42. Peacock WJ: Neurosurgical aspects of epilepsy in children, in Youmans JR, Neurological Surgery, fourth edition, Philadelphia:WB Saunders, 1996;3624-3642
43. Penfield W, Welch K: The supplementary motor area of the cerebral cortex: a clinical and experimental study. Arch Neurol Psychiatry 66;289-317, 1951
44. Pilcher WH, Rusyniak WG: Complications of epilepsy surgery. Neurosur Clin of N Amer 4;311-325, 1993
45. Polkey CE: The complications of temporal lobe surgery in children, in Tuxhorn I, Holthausen H, Boenigk H (eds.), Pediatric Epilepsy Syndromes and Their Surgical Treatment, London: John Libbey, 1997;345-347
46. Prats AR, Morrison G, Wolf AL: Focal cortical resections for the treatment of extratemporal epilepsy in children. Neurosurg Clin North Am 6;533-540, 1995
47. Probst FP: Vascular morphology and angiographic flow patterns in Sturge-Weber Angiomatosis:facts, thoughts and suggestions. Neuroradiology 20;73-78, 1980
48. Rasmussen T: Surgery for epilepsy arising in regions other than the frontal and temporal lobes. In Purpura DP, Perry KK, Walter RD, Advances in Neurology, New York:Raven Press, 207-226, 1975
49. Rasmussen T: Hemispherectomy for seizures revisited. Can J Neurol Sci 10;71-78, 1983
50. Rasmussen T: Extratemporal cortical excisions and hemispherectomy. In Engel J Jr (ed), Surgical Treatment of the Epilepsies, New York:Raven Press, 1987; 417-424
51. Rinthaka PJ, Chugani HT, Messa C, Phelps ME: Hemimegalencephaly: evaluation with PET. Peds Neurol 9;21-28, 1993
52. Rostomily RC, Berger MS, Ojemann GA: Postoperative deficits and functional recovery following removal of tumors involving the dominant hemisphere supplementary motor area. J Neurosurgery 75; 62-68, 1991
53. Salanova V, Andermann F, Rasmussen T et.al..: Parietal lobe epilepsy: clinical manifestations and outcome in 82 patients treated surgically between 1929 and 1988. Brain 118;607-627, 1995
54. Taha JM, Crone KR, Berger TS: The role of hemispherectomy in the treatment of holohemispheric hemimegaloencephaly. J Neurosurg 81;37-42, 1994

55. Tuxhorn I, Pieper T, Holthausen H, Pannek H: Seizure outcome after temporal lobectomy in childhood, in Tuxhorn I, Holthausen H, Boenigk H (eds.), Pediatric Epilepsy Syndromes and Their Surgical Treatment, London: John Libbey, 1997;334-344
56. Van Buren JM.(1987): Complications of surgical procedures in the diagnosis and treatment of epilepsy. In Engel J Jr (ed), Surgical Treatment of the Epilepsies, New York: Raven Press, 1987; 465-475
57. Ventureyra ECG, Higgins MJ: Complications of epilepsy surgery in children and adolescents. Pediatr Neurosurg 19;40-56, 1993
58. Vigevano F, Bertini E et.al.: Hemimegalencephaly and intractable epilepsy: benefits of hemispherectomy. Epilepsia 30;833-843, 1989
59. Villemure JG, Adams CBT, Hoffman HJ et.al. (1993): Hemispherectomy. In: Engel J Jr, Surgical Treatment of the Epilepsies, New York: Raven Press, 1993;511-519
60. Villemure JG: Hemispherectomy techniques: a critical review, in Tuxhorn I, Holthausen H, Boenigk H (eds.), Pediatric Epilepsy Syndromes and Their Surgical Treatment, London: John Libbey, 1997;729-738
61. White HH: Cerebral hemispherectomy in the treatment of infantile hemiplegia. Confin Neurol 21;1-50, 1969
62. Wilson DH: Cerebral hemispherectomy for infantile hemiplegia: report of 50 cases. Brain 93;147-180, 1970
63. Wyllie E, Chee M, Granstrom M-L, DelGiudice E, Estes M, Comair Y, Pizzi M, Kotagal P, Bourgeois B, Luders H: Temporal lobe epilepsy in early childhood. Epilepsia 34;859-868, 1993 64-Wyllie E, Comair YG, Kotagal P et.al.: Epilepsy surgery in infants. Epilepsia 37;625-637, 1996
65. Wyllie E, Comair YG, Kotagal P, Bulacio J, Bingaman W, Ruggieri P: Seizure outcome after epilepsy surgery in children and adolescents. Ann Neurol 44;740-748, 1998
66. Wyllie E: Catastrophic epilepsy in infants and children: identification of surgical candidates. Epileptic Disorders 1:261-264, 1999
67. Zentner J: Surgical aspects of corpus callosum section, in Tuxhorn I, Holthausen H, Boenigk H (eds.), Pediatric Epilepsy Syndromes and Their Surgical Treatment, London: John Libbey, 1997;830-849