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Review Article

An update on pediatric surgical epilepsy: Part I

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ABSTRACT

Epilepsy affects many children worldwide, with drug-resistant epilepsy affecting 20–40% of all children with epilepsy. This carries a significant burden for patients and their families and is strongly correlated with poor cognitive outcomes, depression, anxiety, developmental delay, and impaired activities of daily living. For this reason, we sought to explore the role of pediatric epilepsy surgery and provide an overview of the factors contributing to epilepsy surgery planning and execution. We review the necessary preoperative evaluations, surgical indications, planning considerations, and surgical options to provide a clear pathway in the evaluation and planning of pediatric epilepsy surgery.

Keywords: Drug-resistant epilepsy, Epilepsy surgery, Pediatric

INTRODUCTION

Epilepsy is estimated to affect 10.5 million children worldwide.[12] In the early pediatric population, achieving seizure freedom is critical to prevent developmental arrest or regression.[1,13] Nevertheless, these patients often require multiple antiepileptic medications, leading to additive side effects, without adequate seizure control. About 20–40% of children have drug-resistant epilepsy (DRE), persistent seizures refractory to two antiepileptic medications,[3,8] presenting significant social, economic, health, and developmental implications.[14]

Surgical treatment of DRE has been shown to be safer and more efficacious compared to medical management.[11] It is aimed to remove or disconnect the epileptogenic zone (EZ), the minimal amount of cortex to produce seizure freedom,[17] from surrounding normal brain while minimizing morbidity. Conventionally, epilepsy surgery focused on resections or disconnections: lobectomy, hemispherectomy, cortical excision, and corpus callosotomy. Overtime, the armamentarium has grown to include newer, less invasive approaches including neuromodulation and ablative techniques.

With advances in technology, there are now multiple indications for the different types of surgery to address pediatric epilepsy. We describe, in Part I, practices and advances in diagnostic workup and surgical strategies.

SURGICAL INDICATIONS

Surgical indications have evolved overtime to encompass a wider variety of epilepsy types, applying epilepsy surgery to more patients. Table 1 describes the evolution of indications for surgical evaluation in recent years.

SURGICAL PLANNING

Presurgical evaluation identifies the EZ, correlating it with function. Stepwise evaluation should include a detailed clinical history, interictal scalp electroencephalography (EEG), long-term video EEG, high-resolution structural

Table 1: Timeline of indications for epilepsy surgery.

Historically	Drug-resistant focal epilepsy impacting quality of life Absence of progressive neurological disease Presence of localizable focal epileptogenic zone[11]
American Academy of Neurology, 2003	Disabling complex partial seizures With or without secondarily generalized seizures Failed appropriate trials of first-line antiepileptic drugs[9]
International League Against Epilepsy, 2006	Evidence of focality or a potentially resectable lesion Presence of cortical dysplasia, tuberous sclerosis complex, polymicrogyria, hypothalamic hamartoma, hemispheric syndromes, Sturge-Weber syndrome, Rasmussen syndrome, Landau-Kleffner syndrome, and other pathologies with evidence of cortical injury[5]

Table 2: Presurgical epilepsy evaluation.

Test	Purpose	Strengths/weaknesses
Interictal scalp EEG	Identification of interictal electrical abnormalities (e.g., spike and sharp wave or focal rhythmic slow-wave activity).	Inexpensive; sensitive to cortical currents in all orientations. Low spatial resolution; low diagnostic yield; attenuated by skull/scalp.
Long-term video EEG	Analysis of ictal semiology and correlation with ictal EEG.	Rules out nonepileptic seizures; allows better classification of seizure type and localization. Low spatial resolution; attenuated by skull/scalp.
High-resolution MRI	Detection of structural epileptogenic lesion.	Protocoled to detect hippocampal sclerosis and focal cortical dysplasia. Nonexpert reading fails to detect subtle lesions.
Neuropsychological/Neuropsychiatric assessment	Evaluation of cognitive capabilities and functional deficits.	May detect subtle deficits as well as cognitive reserve. Dependent on skill of administering clinician and cooperation of subject.
Magnetoencephalography	Identification of magnetic fields produced by interictal epileptic discharges.	Sensitive to smaller cortical sources than EEG; lack of attenuation by skull/scalp. More sensitive to superficial cortical activity; sensitive only to currents tangential to scalp surface.
Functional MRI	Identification of eloquent cortex by mapping language, motor, and memory tasks.	Acceptable concordance with Wada testing in language localization. For sensorimotor tasks, electrocortical stimulation more reliable; dependent on patient cooperation and education.
Interictal positron emission tomography	Detection of interictal glucose hypometabolism within epileptic foci.	Can detect MRI-negative focal epilepsies (e.g., cortical dysplasia, temporal lobe epilepsy). Distribution of hypometabolism is wider than the seizure focus; less reliable in extratemporal epilepsy.
Ictal single-photon emission computed tomography	Detection of region of increased cerebral blood flow induced by a seizure.	Well correlated with ictal focus. Yield depends on timing of tracer injection; not feasible if seizures infrequent.

EEG: Electroencephalography, MRI: Magnetic resonance imaging

Table 3: Open surgical options for drug-resistant epilepsy.

Surgery	Clinical application	Outcomes	Considerations
Resection of lesion	DRE originating from cortical-based lesions[21]	Over half of pediatric patients obtain seizure freedom following lesionectomy of cortical lesions.[4,7] Cure can occur if seizure originates from single lesion.[24]	Multifocal epilepsy may necessitate multiple EZ resections or disconnections.[24] Favorable seizure outcomes correlate with MRI confirmation of resection.[4] Cortical mapping may allow for maximal resection while minimizing neurological impairment. Intraoperative electrocorticography may guide extended lesionectomy of tissue adjacent to the primary pathology if necessary.[7]
Lobectomy	Temporal lobe epilepsy is the most common surgically amenable cause of DRE in children (e.g., 20% of cases).[20]	Anterior temporal lobectomy, with selective amygdalohippocampectomy, provide improved seizure outcomes in 60–80% of pediatric patients.[20,31]	Associated risks of homonymous superior quadrantanopsia, language deficits, and rarely contralateral hemiparesis. Early temporal lobectomy in children with DRE of temporal lobe origin may improve neurocognitive outcomes without impacting cognitive functions.[16]
Hemispherectomy	Disorders are characterized by severe unilateral hemispheric damage: porencephaly, Sturge-Weber syndrome, and Rasmussen encephalitis.[15]	Hemispherectomy provides excellent seizure freedom with anywhere from 54% to 90% of Engel 1 seizure freedom.[15,18]	Functional over anatomic hemispherectomy may avoid complications such as superficial hemosiderosis and hydrocephalus.[15] Various techniques and endoscopic approaches allow functional disconnection with minimal cortical removal.[29]
Corpus callosotomy	Palliative procedure for patients with generalized seizures or rapid secondary generalization, particularly for drop attacks (atonic, tonic, or myoclonic seizures).[26]	Improvement in seizure frequency has been noted in 65–85% of pediatric patients.[10]	Anterior callosotomy with preservation of the splenium may avoid disconnection syndrome with improved seizure control and lower relapse after complete callosotomy.[10,26] Endoscopic techniques have been recently introduced to more focally reduce seizure burden.[26]

DRE: Drug-resistant epilepsy

magnetic resonance imaging (MRI), and neuropsychological/neuropsychiatric assessment[22] and also included magnetoencephalography (MEG), functional MRI (fMRI), interictal positron emission tomography, and ictal single-photon emission computed tomography [Table 2]. Conventionally, the Wada test had been used for cortical stimulation mapping; this may be supplemented or supplanted with fMRI[19] as well as MEG or resting state fMRI.[6]

Where the EZ cannot be characterized with noninvasive testing, or noninvasive testing yields contradictory results, Phase 2 assessment utilizing intracranial EEG monitoring may be pursued. Implantation of subdural grids and depth electrodes allows more accurate localization of the EZ than scalp EEG. Functional zones may be identified through cortical stimulation mapping. However, invasive electrocorticography may carry a complication rate of up to 20% (e.g., intracranial hematoma).[30]

For those who are not candidates or have failed surgery, vagus nerve stimulation may palliatively reduce seizures by 50–75%.[2,28] Targeted, responsive neuromodulation is also an option, discussed in Part 2.

SURGICAL OPTIONS

Several surgical options exist based on the seizure type, lesion type, size and location, and EZ characteristics. Lesionectomy is favored for singular cortical-based lesions and can be curative. Lobectomy is used for more focal lesions and proven superior in cases of temporal lobe epilepsy over medical management (Class I evidence).[31] Hemispherectomy, reserved for lesions affecting an entire cerebral hemisphere, has evolved to focus on tissue disconnection rather than resection. Corpus callosotomy palliatively prevents synchronization of epileptic activity between hemispheres and is reserved for those most affected by generalized DRE. The clinical application, outcomes, and considerations for each approach are detailed in Table 3.

SURGICAL CONSIDERATIONS IN PEDIATRICS

Pediatric epilepsy is more diverse in etiology and semiology with migrational disorders, congenital epileptic syndromes, and extratemporal epileptogenic foci more common in children. Therefore, cortical excisions and hemispherectomies are perhaps more common than temporal lobectomies in the pediatric population versus adults. In addition, DRE impacts neurodevelopment in children. Early surgical intervention limits the time on intolerable medications, minimizes cognitive delays and learning disabilities, and improves psychomotor development.[27] Children brains exhibit greater plasticity versus adults, increasing the potential for rehabilitation following even extensive resective or disconnective procedures.[27]

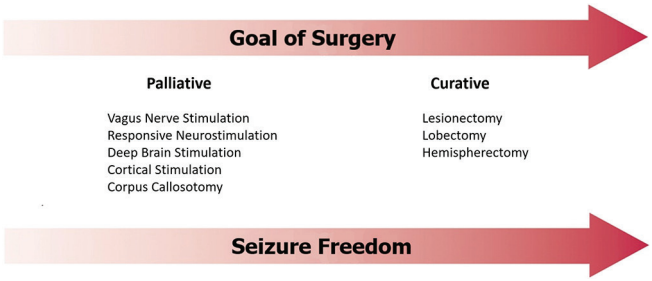

Figure 1: Flowchart demonstrating strategies in the surgical management of drug-resistant epilepsy. The goal of epilepsy surgery can range from curative to palliative, with various available techniques to achieve a range of seizure outcome.

IDENTIFICATION OF CANDIDATES FOR EPILEPSY SURGERY

It is critical to identify the best candidates for epilepsy surgery. The goals include cure or palliation and may warrant a variety of open versus stereotactic techniques [Figure 1]. With growing technology, there is enhanced ability tailor treatment to individual patients.

Despite the growing appreciation for the deleterious developmental and psychosocial effects of pediatric DRE, there are too few surgical referrals,[23] with <1% of patients with DRE referred to epilepsy centers. This may be explained by limited access, cost, and misconceptions regarding who may benefit from evaluation.[25] With continuing innovation in the field of pediatric epilepsy surgery, it is imperative that continued strides be made in patient recruitment and referral to enhance clinical outcomes.

CONCLUSION

Here, we reviewed, summarized, and synthesized important practices and advances in diagnostic workup and surgical strategies of epilepsy surgery. Future increased awareness of the role of epilepsy surgery in children with DRE is critical to increase the breadth of impact.

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Conflicts of interest

There are no conflicts of interest.

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