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ADVANCES IN PEDIATRICS

Brain Surgery for Medically Intractable Epilepsy

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Keywords

• Epilepsy surgery • Outcomes • Medically intractable epilepsy • Surgical selection • Resection

Key points

- Children suffering from drug-resistant epilepsy must be quickly referred to a comprehensive pediatric epilepsy center for a thorough presurgical evaluation.
- Early referral allows for early treatment, which reduces the risk of secondary epileptic focus and increases the possibility of greater functional neuroplasticity following resective surgery.
- Hemispheric, lobar, or lesional epilepsies are often amenable to complete resection, which increases chances of seizure freedom after surgery.
- Bihemispheric and generalized epilepsies can benefit from palliative surgeries or from neuromodulation.
- Neuromodulation techniques infrequently allow for seizure freedom but can significantly reduce seizure burden and increase quality of life.

INTRODUCTION

In North America, an estimated 100/100,000 children are diagnosed each year with epilepsy [1], which is defined as recurrent seizures, and affects between 0.5% and 1% of the population. Of those patients, 50% will be controlled by

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a first antiepileptic drug (AED) and 15% will be controlled by a second AED [2]. After 2 AED, the chance of seizure control drops significatively, meaning that 15% to 30% of children [3] will be pharmaco-resistant [4] and suffer from intractable epilepsy.

Patients suffering from intractable epilepsy are more at risk of injury [5], low self-esteem [6], and even death [7] (sudden unexpected death in epilepsy or SUDEP). Since the first randomized clinical trial (RCT) [8] in adults demonstrating that temporal lobectomy could allow better seizure control than AED in certain cases, tremendous progress has been made to better define patient and surgical procedures that could help achieve seizure freedom. In a recent RCT by Dwivedi and colleagues [9], surgical management of various epilepsy disorders in children led to 77% seizure freedom at 12 months compared with 7% with medical treatment.

The most important steps are patient selection and procedure definition. This is achieved through the presurgical workup, which will classify, roughly, the patient between focal epilepsy (temporal lobe epilepsy [TLE] or extratemporal lobe epilepsy [ETLE]) versus generalized epilepsy as well as lesional versus nonlesional epilepsy. Once the epilepsy type has been well described, surgical procedures can be tailored to the patient and the epileptic region.

The authors divided the article as follows:

- Presurgical workup
  - Noninvasive investigations
  - Invasive investigations
  - Timing of surgery
- TLE
- ETLE
- Multifocal and generalized epilepsy
  - Palliative procedures
  - Neuromodulation

PRESURGICAL WORKUP

Noninvasive investigations

Once epilepsy has been diagnosed and failure of 2 AED has been noted, the diagnosis of *intractable epilepsy* is given [4]. At that point, patients should be referred to a comprehensive epilepsy center. The first step will be to confirm drug resistance. Indeed, the 2 AED given to the patient might have been inadequate for the epilepsy type [10].

Once drug resistance is confirmed, a complete workup is undergone to best define the epileptic zone based on anatomicoelectroclinical findings.

- Since the development of *electroencephalography* (EEG) in 1950, seizure diagnosis has always relied on it. This test can detect interictal or ictal epileptiform activity but can be negative in up to 50% [11] of cases because of its short duration. With improvement of technology and data-storage capacities,

video-EEG was developed, which allows recordings of EEG activities and clinical manifestations simultaneously over several days. Those recordings allow better electroclinical correlation and determination of the epilepsy type based on semiology and electric origin.

- As opposed to the video-EEG, which has good temporal accuracy but poor spatial accuracy, the field of intractable epilepsy has also significantly benefited from the progress in *MRI*. This diagnosis tool is the gold standard for epilepsy assessment that facilitates identification of an epileptogenic lesion, such as focal cortical dysplasia (FCD), glioneuronal tumors, or mesiotemporal sclerosis. The evolution to the 3-T magnet has increased spatial resolution and increased lesional diagnosis to 65% [12]. With the emergence of the 7-T magnet, one can expect even better spatial definition and increased diagnostic yield with as much as 43% more lesional cases identified [13].

Although these tests are sufficient for a subset of patients, others require more advanced technologies to help identify and delineate the epileptogenic focus.

- *PET-computed tomography* and *ictal*–single-photon emission computed tomography rely on abnormal brain metabolism secondary to the seizure focus during the interictal or ictal phase, respectively. This is usually used to confirm the affected lobe when MRI and EEG are incongruent, or when the lesion is subtle.
- *Magnetoencephalography (MEG)* detects magnetic dipole parallel to the brain surface. Its advantage compared with the EEG is the absence of artifacts generated by the skull and the scalp, but the depth resolution is poor. Unfortunately, very few centers have access to MEG. However, this technology is a good add on when EEG cannot well define the boundaries of epileptogenic cortex.
- *Functional MRI* and *tractography* are advanced MRI sequences that allow visualization of eloquent brain regions as well as main white matter tracts. Those sequences are useful when epileptic zones are near or in eloquent cortex.
- A more thorough *neuropsychological testing*, especially in TLE, might also prove useful to better localize cognitive dysfunction, such as memory and language deficits.
- *Biological and genetic testing* might help in refining the diagnosis and establishing a prognosis for the patient as well as a treatment strategy. These tests are improving each year, and more than 265 genes [14] have already been linked to various epilepsy syndromes.

Once the workup has been completed, a *multidisciplinary meeting* is organized involving epileptologists, neurosurgeons, neuroradiologists, neuropsychologists, and nursing staff. The purpose of this meeting is to answer 2 questions: (1) Can the epileptogenic zone be identified? and if so, (2) Is it safe to remove that region of the brain? In other words, does the deficit associated with removing that region of the brain result in an acceptable deficit to the patient? The best treatment plan is then defined in regard to all that information. Table 1 lists frequent diagnoses [15] and epilepsy type related to it.

Table 1
Common drug resistant epilepsy causes

Common causes	Epilepsy type
Focal cortical dysplasia	Lesional/lobar/multilobar
Hippocampal sclerosis	
Tumor: Ganglioglioma, DNET, glioma, hypothalamic hamartoma	
Tuberous sclerosis complex (hamartoma)	
Polymicrogyria	
Hemimegalencephaly	Hemispheric epilepsy
Hemispheric cortical dysplasia	
Sturge-Weber syndrome	
Rasmussen encephalitis	
Stroke	
Encephalopathic epilepsies: Lennox-Gastaut syndrome, West syndrome	Generalized epilepsy

DNET, Dysembryoplastic Neuroepithelial Tumor

If the noninvasive evaluation is inconclusive or not sufficiently congruent, a phase 2 investigation through invasive EEG evaluation can be performed to better define the seizure-onset zone and identify surrounding eloquent brain regions.

Invasive investigation

In some instances, the noninvasive assessment is insufficient to determine with precision the extent of the epileptogenic zone. This may often be the case in FCD or nonlesional epilepsy whereby MRI is usually not adequate to visualize the boundaries of the epileptogenic region or when the presumed epileptogenic region is adjacent or involving eloquent cortex. In these instances, representing up to 40% of surgical candidates, *invasive EEG* may help in understanding the epileptic network [16].

Invasive recordings can be performed in the following different ways:

- *Subdural grids* has long been the most frequent technique for invasive evaluation in North America. In this procedure, the EEG grids are placed on the surface of the brain through a craniotomy and a large dural opening. This allows the surgeon to place a large number of electrodes directly on the cortex under direct vision. When this procedure is performed, a potential resection procedure can be planned a few days after the recordings, making this a two-stage procedure. In those instances, subdural EEG grids can also be used during the surgery to guide resection.
- *Depth electrodes* have been the preferred approach over the past several decades in Europe, especially under the French and Italian school of epilepsy. The advantage compared with subdural grids is the possibility of depth analysis, accessing both sides of the brain and sampling over a wider area of the brain. In addition, these electrodes are placed in a percutaneous fashion obviating a craniotomy [17]. The main disadvantage is the inability to perform brain mapping, which allows identification of eloquent brain

regions. The appearance of *surgical robots* has facilitated its widespread use through North America more recently. Contrary to subdural grids, the depth electrodes can be removed through a very short operation, and the definitive surgical procedure is often delayed for about 6 weeks to minimize the risk of infection.

Timing of surgery

Persistence of seizures despite medical treatment is sometimes seen as trivial by the families and even caretakers, especially in the case of focal seizures or well-known stereotypical seizures. This might lead to a delay in referral to surgical epilepsy centers. This delay added to the time-consuming workup can lead to a significant delay before appropriate treatment can be initiated.

The importance of early surgical intervention is well documented. Children operated before 12 months of epilepsy history seem to have a better psychomotor prognosis [18]. With the absence of seizure control, a secondary epileptogenesis can develop leading to a propagation of seizure onset zones [19]. With time, new independent epileptic foci will appear and transform a curable epilepsy into an incurable epilepsy [20]. Furthermore, younger patients will benefit from neuroplasticity [21], and surgery around eloquent zones can be more aggressive with limited long-term impact (see later discussion).

Even though a definite timing for treatment has not yet been defined, a widespread consensus for treatment is "as soon as possible."

TEMPORAL LOBE EPILEPSY

TLE accounts for 15% to 20% of epilepsies in children [22]. Its prognosis compared with extratemporal epilepsy is usually better. As in all epilepsies, the presence of a lesion in relation to the epilepsy or abnormal MRI as well as the absence of generalized seizures usually indicates better prognosis of seizure freedom [23].

In contrast with adults whereby TLE is the most frequent cause of epilepsy, pediatric TLE is less frequent and usually involves more complex networks [24]. Indeed, adult TLE is frequently related to mesiotemporal sclerosis [25], whereas TLE in children is usually caused by FCD [26], glioneuronal tumors [27] or both, which involves more lateral cortex and sometimes other lobes.

A full workup is done to confirm the electroclinicoanatomic origin of the seizures. Involvement of surrounding regions, the frontal operculum and the insula, in particular, needs to be excluded. Neuropsychological workups are especially useful to rule out memory impairment before surgery and determine if memory function in the contralateral hippocampus is sufficient. Functional MRI is also useful to determine the dominant hemisphere and exclude language areas surrounding the epileptogenic foci.

- The typical surgery is the *anterior temporal lobectomy,* which consists of a resection of the anterior temporal lobe as well as the mesiotemporal structures comprising the amygdala, the hippocampus, and the parahippocampal gyrus, which have all been shown to be involved in mesial TLE. In the dominant

hemisphere, the superior temporal gyrus is usually spared to avoid language impairment.
- The resection can be tailored with the use of *electrocorticography* (ECoG), which consists of intraoperative recording of interictal activities with subdural grids. The strength of this approach is to better determine, intraoperatively, which regions should be resected and which regions should be spared. The weakness is the lack of ictal recordings as well as the limited duration of the recordings, which might lead to false negatives. The literature is still inconclusive on the added benefit of ECoG.
- Apart from the traditional tailored anterior temporal lobectomy, a newer procedure using *laser interstitial thermal therapy (LITT)* has emerged [28]. The main indication in temporal epilepsy is to target the mesiotemporal structures while minimizing collateral damage to the nearby optic radiations and language networks, which course in the lateral part of the temporal lobe. Selective approaches in pediatric neurosurgery are exceptional because the epileptic network usually involves the lateral cortex but might still be indicated in older children with networks closer to those of the adult population.
- In some cases, the neuropsychological assessment shows that most of the memory function is on the side of the temporal epilepsy. In those cases, hippocampus resection cannot be performed, and the multidisciplinary teams will need to resort to *neuromodulation techniques*, which is discussed later in the article.

Overall, prognosis after lobectomy is good in children, with 76% achieving seizure freedom [23], with a trend toward better results in cases of lesional epilepsy where total resection is possible.

EXTRATEMPORAL LOBE EPILEPSY

Most lobar epilepsies are extratemporal in children. They have been traditionally separated from temporal epilepsy because lobectomies are often partial owing to the presence of eloquent cortex (central region and visual cortex), which leads to less robust seizure control [29]. ETLE is further divided depending on the affected lobe, including the frontal, parietal, and occipital lobe in decreasing order of frequency.

The workup is especially important in those epilepsies to analyze the involvement of the eloquent cortex, which mostly limits resections. Functional MRI and tractography may be of special interest for this aspect. Second-phase investigation with intracranial recordings is often performed to completely rule out involvement in other regions, which has a direct impact on prognosis, as well as strictly defining the location and involvement of eloquent cortex. Intraoperative ECoG is often a useful adjunct to help tailor resections based on interictal EEG recordings.

Lesionectomy

More frequently than in temporal epilepsy, the resection might be tailored to a lesion when present and when the epilepsy assessment confirms that the seizure onset zone is limited to that lesion. Surgical technique is comparable to a

tumorectomy and involves diverse techniques of microdissection, which is beyond the scope of this article. Prognosis for those lesional epilepsies is significantly better than nonlesional epilepsies.

- Specific lesions to consider are *FCDs*. FCDs have been classified into 3 groups [30]: type 1 is related to architectonic disorder only; type 2 includes architectonic disorder as well as dysmorphic neurons (balloon cells); and type 3 is the association of an FCD with another lesion (tumor or scar tissue). The diagnosis of FCD has significantly improved with the increase of spatial definition on MRI [31]. Typically, FCD is seen on the MRI as blurring of the gray-white junction of the neocortex. Other signs such as a transmantle sign can be seen but are not always present. The imaging abnormality can be extremely subtle so advanced MRI sequences such as voxel-based morphometry can be promising [32]. The main difficulty remains the delineation of the boundaries of the FCD, which will influence the extent of resection. Type 2 FCDs are usually more focal and better delineated than type 1 FCDs, which makes seizure control more likely [33]. Again, ECoG during surgery might help in determining resection boundaries, but results in the literature are not always consistent.
- Another special condition is *hypothalamic hamartoma*. This deep-seated lesion can cause a specific type of seizure called gelastic seizures, which are characterized by involuntary laughter. Those seizures are often intractable but respond well after resection [34]. Unfortunately, direct resection can cause harm to the hypothalamus, which in turn can lead to complications, such as obesity or diabetes insipidus. Because this lesion is benign from an oncologic standpoint, the risk-benefit balance is not always in favor of resection. Nowadays, LITT is the preferred approach to these lesions, allowing for maximal ablation while reducing the risks of approach-related morbidities [35].

Altogether, surgery for lesional ETLE achieves seizure freedom in about 66% of cases versus only 34% for nonlesional ETLE [36].

Frontal lobectomy

In medically intractable frontal lobe epilepsy without an identifiable lesion, a frontal lobectomy can be considered. The main determinant is hemisphere dominance, which will indicate the presence of language function. The functional limits are the primary motor cortex posteriorly and the language cortex inferiorly if present. The trend nowadays is to tailor the resection instead of doing standard lobectomy. For this purpose, these cases are commonly done with ECoG or preresection invasive EEG evaluations. Care must be taken to avoid injuring major veins and arteries, as morbidity often comes from those lesions. Transient neurologic impairment is frequent after such surgeries but resolves in the following weeks.

Posterior quadrant resections

A posterior quadrant resection consists of parieto-occipital resection that can be extended to the temporal lobe if needed. Although the posterior quadrant resection is an uncommon procedure, most children undergoing a large

multilobal resection are less than the age of 2 years and have an identifiable lesion, commonly FCD, that spares the frontal lobe.

This resection spares the central area and, in the absence of a vascular lesion, should thus not lead to hemiparesis. On the contrary, a complete hemianopia is a rule but is often present before surgery owing to the underlying disease. In the well-selected patient, seizure freedom can reach 75% [37].

Insulectomy

The insula is a deep-seated and quite thin lobe, underneath the Sylvian fissure. Its access is difficult and puts the main vascular structures of the brain at risk. Nevertheless, the insula is at a junction between the frontal, temporal, and parietal lobe through their opercular regions. It is commonly involved in limbic system epilepsy.

Its general involvement is probably underrecognized, as its investigation is difficult. Surface EEG can rarely distinguish insular activity from the surrounding opercular region, and invasive EEG implantation is thus frequently needed [38]. Subdural grids are difficult to place owing to the narrow space, even in the case of a large Sylvian split. Depth electrodes are the preferred technique, but targeting, owing to the vessels, might be complicated [39]. All the technical difficulties make the insula an understudied and less understood lobe compared with the others.

Failures in TLE are thought to be related to more extended seizure onset zones commonly referred to as temporal lobe plus epilepsy. The involvement of the insula in this entity is thought to play an important role [40].

Technically demanding, the insulectomy can be limited to the anterior or posterior lobe. A large Sylvian fissure split is required to expose the lobe, and resection is performed between the Sylvian vessels, or part of an operculum (usually the frontal operculum) is resected. Nevertheless, favorable outcome is possible with up to 69% achieving seizure freedom [41]. In order to avoid the vascular risks, some groups have resorted to the use of radiofrequency ablation or LITT for a more minimally invasive approach [42], but definite proof of superiority is still lacking.

Multiple subpial transection

If the seizure focus is located in an eloquent brain region, resection may not be an option, especially if it will lead to an unacceptable neurologic deficit, such as hemiparesis or aphasia.

In these situations, multiple subpial transection (MST) has been described as a technique to isolate the epileptogenic cortex from surrounding cortex without compromising neurologic function [43]. Some variations to the technique have been described, but the concept remains the same: interrupt the horizontal connections in the cortex to limit propagation and recruitment of gray matter in seizure generation and spread. This technique maintains the longitudinal connections from the cortex, which allows conserving the functional activity of the pathologic cortex.

MSTs can be performed alone if the seizure-onset zone is strictly limited to the eloquent cortex, or in addition to a resective procedure such as a lesionectomy or a lobectomy if the seizure-onset zone extends to the eloquent cortex. Good outcomes (>95% reduction of seizure) are reported in up to 87% of patients if MST is performed in conjunction with resection and in up to 71% of patients if performed alone [44]. Some studies report 50% of seizure freedom at 5 years [45] after MST only in eloquent cortex.

Neuroplasticity

Even if resection of eloquent cortex is usually avoided, there are special nuances for infants, toddlers, and younger children with respect to neuroplasticity. Contrary to previous theories, studies have shown that language lateralization only starts around 4 years old and is usually finished by age 7 years with different timing depending on the region of the brain involved [46]. Hand dominance usually occurs by the age of 2. In children less than 2 years of age, resection can generally be considered regardless of the brain region involved, as almost all functions lost can be completely reacquired.

Furthermore, recovery from a craniotomy is generally quicker and more complete than in adults [47]. This ability to recover is most evident in patients undergoing hemispherectomy, which will be discussed later. Even though a complete hemisphere is disconnected, patients usually maintain walking capacity, gross motor arm function, and the ability to communicate.

It is important to consider that the brain of patients with drug-resistant epilepsy might be wired differently. This is especially the case in "catastrophic" or syndromic epilepsies with diffuse brain involvement whereby neuroplasticity might also be impaired [21].

MULTILOBAR AND GENERALIZED EPILEPSY

Palliative surgeries

When the epileptogenic zone involves homotopic regions in both hemispheres, or when the epilepsy is generalized from its onset, curative resections cannot be achieved. Nevertheless, targeting the most impairing seizure types can be useful. These surgeries are called "palliative" in that they are meant to lower the seizure burden but not necessarily provide seizure freedom.

Corpus callosotomy

In some epileptic syndromes, such as Lennox-Gastaut syndrome, the most debilitating seizures are drop attacks in which the patient suddenly loses tone of the lower limbs, resulting in a fall. These seizures can cause serious injuries and can significantly decrease quality of life. In those cases, a *corpus callosotomy* can be performed to stop the epileptic activity from spreading from one hemisphere to the other. This can be very effective in eliminating drop attacks.

- This technique is a *partial callosotomy,* as it involves transection of the anterior two-thirds of the corpus callosum. By doing so, motor fibers, at the origin of the atonic seizures, are interrupted, but other fibers connecting both hemispheres

are maintained, decreasing the risks of disconnection syndromes [48]. The most serious concern with a disconnection syndrome is the loss of bihemispheric communication. If drop attacks or other serious seizures originating from the spread of the epileptic activity between hemisphere persist, the callosotomy can be completed in a second-look surgery.

- In more debilitated patients with severe mental delay and those that are nonverbal, a *complete corpus callosotomy* can be performed in a single step, as the chance of controlling seizures, usually more complex in those cases, outweighs the risk of disconnection syndromes, usually less clinically relevant in this population.
- In a systematic review [49], 88% of patients with a complete corpus callosotomy experienced worthwhile seizure reduction (Engel class III) versus 58% in anterior two-thirds partial corpus callosotomy, but with 18% of patients developing a disconnection syndrome in the case of complete corpus callosotomy versus 0% in case of partial corpus callosotomy.

Some teams [50] have suggested performing this procedure with an endoscope to reduce blood loss and incision length, but no clear superiority to open procedures exists. Some investigators have also performed a callosotomy with LITT [51] with, again, lack of clear advantage compared with the classic open technique.

Functional hemispherotomy

Some patients suffer from a diffuse, even hemispheric, but unilateral epilepsy. Usual causes are Rasmussen encephalitis, hemimegalencephaly, large perinatal hemispheric stroke, or Struge-Weber syndrome. In those cases, the affected brain is not fully functional, and a full hemisphere can be disconnected to eliminate clinical seizures with limited impact on neurologic function. Even though this procedure might seem aggressive, it allows development of the normal hemisphere, which usually leads to functional improvement. Nevertheless, this surgery is obviously reserved for select candidates. The authors' research group has recently developed a prognosis score [52] based on a multicenter experience, which demonstrated that older patients at the time of seizure onset (>3.5 years), those without generalized seizure semiology, those without previous resection or contralateral hypometabolism, and those with epilepsy originating from poststroke gliosis had the highest likelihood of seizure freedom.

Different approaches have been described and can be roughly divided into 2 groups: lateral perisylvian hemispherectomies (developed by Villemure) and vertical parasagittal hemispherectomies (developed by Delalande). Again, some investigators have developed endoscopic disconnection to reduce invasiveness, but this is still not used on a widespread basis. In a large multicenter review [53] performed on 672 patients, seizure freedom was reached in 88% of patients at 1 year but decreased to 62% at 10 years with more durable seizure freedom in those that underwent a vertical hemispherotomy.

Neuromodulation procedures

Apart from corpus callosotomies, neuromodulation procedures can still be considered for palliative reduction in seizures. To date, no epilepsy types nor epileptogenic syndromes are formal contraindication for those procedures, which makes them a useful tool for the epilepsy neurosurgeon.

Those techniques rely on the introduction of electric current in the neurologic system to modulate brain activities. Even though these technologies rarely bring complete seizure freedom, they usually improve epilepsy control, which in turn increases quality of life.

The main complexity of those procedures comes from the adjustment of settings (ie, the adaptation of the current) after the surgeries, which requires a trained specialist. Most of the time, this task is performed by the treating epileptologist.

Deep brain stimulation

Deep brain stimulation (DBS) has initially been developed for the treatment of movement disorders. The aim is to modulate brain circuiting by stimulation at particular nodes of the circuit. The type of stimulation used can drive or inhibit the circuit. Interest in treating other condition such as epilepsy has rapidly emerged.

Limbic epilepsy, which is very common in adult epilepsy syndromes, commonly involves the circuit of Papez. The most commonly used target in this circuit seems to be the *anterior thalamic nucleus (ATN)*. Other targets have been tried, such as the centromedian nucleus of the thalamus, the mamillary body, or the hippocampus.

This surgery needs careful planning and targeting, which is made possible using a stereotactic head frame. The use of the frame is challenging in children under 4 years of age because of the skull thickness. Once the frame is positioned, 2 electrodes are inserted to the target and then connected to a generator in the pectoral region. This generator can later be interrogated, and current settings can be adjusted in order to optimize epilepsy control usually over the course of several months.

The group SANTE has performed [54] the largest RCT in adults to date and shows 56% seizure reduction at 2 years with 54% of patients having more than 50% of seizure reduction and 12.7% being seizure free. These outcomes improve at 5 years [55] with 69% mean seizure reduction and 68% of patients with more than 50% of seizure reduction, and further at 7 years [56] with 75% mean seizure reduction. The main side effect of ATN targeting is the occurrence of subjective depression and memory impairment in some cases, which might require therapy. However, these symptoms are not found following formal neuropsychological assessment [57]. Comparable data for children are still lacking, but a recent systematic review [58] found 12.5% seizure freedom and 85% seizure reduction in patients.

Even though this technology has been available for a few years, its use is still limited probably because of its falsely perceived greater aggressiveness

compared with vagus nerve stimulation (VNS) without major added value proven yet.

Vagus nerve stimulation

VNS consists of placing an electrode around the vagus nerve in the neck. The electrode is connected to a stimulator under the skin in the pectoral region. The exact mechanism is still unclear. When the stimulator activates, an antidromic discharge travels backwards along the nerve and is thought to release neurotransmitters inside the brainstem, mainly in the locus coeruleus. Those neurotransmitters in turn are thought to reduce cortical excitability that leads to epileptogenesis.

This intervention is quite simple and can be performed in same-day surgery, which has made it easily available even in patients with severe disability. The results are somewhat limited with 50% of patients achieving greater than 50% seizure reduction. Better outcomes can be achieved in those with later age of seizure onset and fewer AED tried before VNS, probably meaning less severe epilepsy [59]. Still, this technique is frequently considered given the ease of implantation and the severity of treated patients.

Future developments are related to seizure detection in the hopes of developing closed-loop systems. Presently, the system can detect tachycardia, often related to seizure onset. It can use this indirect biomarker of a seizure to send an additional stimulation to try to abort the forthcoming seizure. This seems to improve seizure control but is not yet a true closed-loop system.

Responsive neurostimulation

Responsive neural stimulation is a closed-loop system in which 1 or 2 electrodes, inserted close to or inside epileptogenic focus, are connected to a generator. These electrodes contain detecting contacts, and when an ictal pattern is detected, a stimulation is induced.

Indication is usually limited to 2 epileptogenic foci, which may limit patient selection, but results for this selected population are quite good.

Similar to DBS, most of the data are from the adult population, but the authors conducted a recent retrospective multicenter analysis [60] on its use in children and young adults with 35 patients, which showed that 60% of patients have at least greater than 50% of seizure improvement and 6% of patients achieve seizure freedom. Only 3 patients presented complications (2 infection and 1 hemorrhage), and 4 patients had concurrent resection.

SUMMARY

Intractable epilepsy is a frequent diagnosis in children. A comprehensive assessment is necessary, which is initiated after referral to a dedicated pediatric epilepsy surgery center. Multiple surgical procedures are available and need to be tailored to each patient to achieve the best possible outcome. Curative procedures usually consist of resection, whereas palliative procedures comprise neuromodulation and disconnection techniques.

CLINICS CARE POINTS

- There is an increasing number of children that can benefit from epilepsy surgery.
- A surgical resection or disconnection offers a chance to cure.
- There is a shift toward minimally invasive procedures and neuromodulation.
- Individualized treatments are required for each patient to achieve the best outcomes.

Disclosure

The authors have nothing to disclose.

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