

# Hemispherotomy and other disconnective techniques

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The surgical treatment of intractable epilepsy has evolved as new technical innovations have been made. Hemispherotomy techniques have been developed to replace hemispherectomy in order to reduce the complication rates while maintaining good seizure control.

Disconnective procedures are based on the interruption of the epileptic network rather than the removal of the epileptogenic zone. They can be applied to hemispheric pathologies, leading to hemispherotomy, but they can also be applied to posterior quadrant epilepsies, or hypothalamic hamartomas.

In this paper, the authors review the literature, present an overview of the historical background, and discuss the different techniques along with their outcomes and complications. (DOI: 10.3171/FOC/2008/25/9/E14)

**KEY WORDS** • disconnective technique • epilepsy • hemispherotomy • hypothalamic hamartoma • posterior disconnection

**H**ORROR VACUI (“nature abhors a vacuum”) wrote Aristotle in 350 BC. Complications from a dead space will arise after epilepsy surgery when a large part of the brain is removed. Since the first hemispherectomy for epilepsy was done in 1938, surgical techniques have evolved to try to minimize the extent of resection while maintaining the same seizure outcome. In the past, patients died of hydrocephalus and hemosiderosis following anatomical hemispherectomies. Those late complications discouraged surgeons from performing this surgery in patients with refractory epilepsy. New surgical techniques, as well as a better understanding of the complications leading to appropriate treatment, have brought hemispheric epilepsy back to surgeons’ hands.

Disconnective procedures are based on the concept that interrupting the epileptic discharge-spreading pathway, by isolating the primary epileptogenic zone, would have the same effect as removing the focus. This is true for hemispherotomy as well as for multilobar disconnection, hypothalamic hamartoma disconnection, or corpus callosotomy.

A continual evolution of the disconnective techniques has helped improve outcome in epileptic lesions that are not well delimited from the surrounding structures, such as hypothalamic hamartomas or multiple unilateral foci that are responsible for intractable epilepsies. We will describe here some of the disconnective techniques, focusing on their different surgical techniques, with their outcomes and complications. Hemispherotomy, posterior disconnection, and hypothalamic hamartoma disconnection will be

reviewed. Corpus callosotomy is described in another article in this issue.

## Hemispherotomy

Anatomic hemispherectomy was first performed for the treatment of tumors by Dandy in the late 1920s.<sup>15</sup> With these operations, he proved that half the brain could be removed while leaving the patient alive. The first patient with epilepsy who underwent such a procedure was a pediatric patient; the surgery was performed by McKenzie in Toronto in 1938. The patient was seizure free after the procedure, which led more surgeons to try that approach. Subsequently, other cases were reported. The first larger series was reported by Krynauw<sup>37</sup> and consisted mainly of cases involving pediatric patients with infantile hemiplegia. Around the same time, White et al.<sup>66,67</sup> explored operative techniques for hemispherectomy on monkeys.<sup>65,66</sup> Two main techniques of hemispherectomy have been described: the removal of the hemisphere “en bloc” as performed by Rasmussen<sup>29,48</sup> and the removal in fragments as performed by Dandy and Penfield.<sup>15,37,43</sup> The operation seemed to cure most patients of their seizures, and was performed until the mid-1960s, when long-term complications were reported by Oppenheimer and Griffith.<sup>41</sup> They described progressive worsening of the neurological status of the patients a few years after surgery (an average of 8 years after surgery), leading to death in up to 30–40% of the patients. The autopsy revealed iron deposits on the brain surface, with a membrane lying over the hemispherectomy cavity and the ventricular wall; this condition was later called “superficial

Abbreviation used in this paper: CSF = cerebrospinal fluid.

cerebral hemosiderosis.” Their report was soon to be followed by more, describing the same late complications, and the procedure was abandoned.

Neurosurgeons have tried to understand the etiology of those complications and have developed technical variations on the surgeries. Alternative surgical strategies were developed by Adams,<sup>1</sup> who suggested decreasing the large CSF cavity that was created by the removal of the hemisphere by plugging the foramen of Monro with muscle, then suturing the dura mater to the falx and tentorium to create a large extradural space rather than a CSF-filled space. Peacock and colleagues<sup>42</sup> suggested leaving a shunt or a drain in the cavity, in order to evacuate the blood and debris from the surgery, as they were believed to cause the siderosis. Anatomical hemispherectomy is still performed at a few centers, in some selected cases.<sup>23,65</sup> After clipping and dividing of the major arteries, a corpus callosotomy followed by frontobasal disconnection and following the ventricle from the trigone to the hippocampus anteriorly, the hemisphere can be removed.<sup>27</sup> Hemidecortication was described as an alternative by Ignelzi and Bucy;<sup>33</sup> in this procedure, only the gray matter was removed, sparing the white matter and avoiding the opening of the ventricle. Again, the hemidecortication approach is still used in some centers, with subtle technical variants.<sup>9,68</sup> While some perform a standard temporal lobectomy followed by decortication of the rest of the hemisphere, others only remove gray matter over the entire hemisphere. In general, the basal ganglia and thalamus are left intact.

In parallel, functional hemispherectomy was introduced in the 1970s by Rasmussen, who modified the technique of the anatomical hemispherectomy,<sup>47,48</sup> to remove part of the central and temporal regions while disconnecting the rest. He realized that when some brain was left behind, the complications of the surgery seemed to be less. The aim of the surgery was to keep a comparable outcome with respect to seizures but to reduce the number of complications by removing less brain.

In the early 1990s, a new era in functional hemispherectomy was initiated with the introduction of the hemispherotomy,<sup>20</sup> and 2 different approaches were described almost simultaneously. A vertical approach was described by Delalande and colleagues,<sup>20</sup> while a lateral approach was described by Villemure and coauthors.<sup>59,62,63</sup> In hemispherotomy, the aim is to remove as little brain as possible by only disconnecting the hemisphere without creating a cavity. Since the first description of those techniques, a number of modifications have been made to reduce the number of complications. The latest variation of Delalande and Villemure’s techniques, as well as some other current techniques, will be described hereinafter.

### Indications and Selection Criteria

The success of hemispherotomy, like that of epilepsy surgery in general, largely depends on the selection of the patient. Different factors have to be taken into account, such as the type and localization of seizures, the intractability of the patient’s epilepsy, the etiology of the seizures, and the radiological and neurological findings.

Patients with unilateral epilepsy that is poorly controlled with medication are candidates for this type of surgery. The

cause of the epilepsy might vary and it can be either congenital or acquired. The pathology has to be unilateral and widespread, as in the following conditions: congenital hemiplegia from a prenatal vascular insult, Sturge–Weber syndrome, hemimegalocephaly or diffuse hemispheric cortical dysplasia, Rasmussen encephalitis, hemiconvulsion-hemiplegia-epilepsy syndrome, or a sequela of trauma or infection. However, even though patients with those different conditions can all be considered for hemispherotomy, their outcomes are not the same; this will be discussed later.

By disconnecting the motor cortex along with the rest of the hemisphere, hemispherotomy will create a contralateral motor deficit. In patients with hemiparesis and hemianopsia, this drawback of surgery is reduced, since no new deficit will arise, and these patients are offered surgery more readily. In patient with good neurological status but severe and intractable seizures, surgery will offer a relatively good seizure outcome but will impair neurological function. These cases have to be discussed thoroughly with the patient and his or her family to weigh the benefits and possible neurological outcomes. In progressive conditions, such as in Rasmussen encephalitis for example, the natural history will be a progressive worsening of the neurological function. An early surgery will acutely worsen the neurological status, but might preserve some cognitive functions; while waiting until hemiplegia occurs will reduce the risk of creating new deficits through surgery, but to delay the surgery might have a negative effect on cognitive outcome.<sup>34,51</sup> Timing of surgery becomes an important issue. Before a child undergoes any surgery of this type, it is important for the parents, and the child if he or she is old enough, to understand the expected residual handicap due to the hemiparesis.

Frequent seizures have a deleterious effect on the maturation of the brain and will affect the development of young children. Later in life, seizures will disrupt the ability of the child to learn, and a psychosocial decline might be seen. Uncontrolled epilepsy will also result in social exclusion of the child, with repeated absences from school, in addition to the secondary effects of medication, such as sleepiness and attentional deficits. There also is some evidence that brain plasticity might be greater at an earlier age, and therefore, early surgery might facilitate the other hemisphere’s involvement in different tasks.<sup>31</sup> Early surgery is also advocated by Vining et al.,<sup>65</sup> who suggests that constant firing in one hemisphere might interfere with the function of the other, which would explain the improvement in overall functioning of the child after a hemispherectomy/hemispherotomy.

### Preoperative Investigation

All patients with refractory epilepsy who are considered for a hemispherotomy should have a thorough clinical, neurophysiological (video-electroencephalographic), and radiological (MR imaging) workup.

The electroencephalographic abnormalities are generally multifocal and widespread throughout the diseased hemisphere. Since the patient will be functioning only with his or her remaining hemisphere, it has to be healthy; bilateral seizures are a contraindication for surgery. However, in rare cases, when a patient has severely debilitating seizures that always originate from the same hemisphere but electrical

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abnormalities on the contralateral side, the patient might be considered for an hemispherotomy. However, the presence of bilateral independent spikes remains an unfavorable prognostic factor.<sup>8,57</sup>

Magnetic resonance imaging should confirm the integrity of the “normal” hemisphere. Bilateral hyperintensities in the deep gray nuclei should raise a red flag, and mitochondrial or metabolic disorders are cause for concern. The imaging will confirm the diagnosis, as well as help the surgeon in visualizing the 3D anatomy of the brain, which is essential for the surgery. The procedure will vary slightly depending on ventricle size, brain atrophy or hypertrophy, and the configuration of the corpus callosum.

Neuropsychological evaluation helps to assess the presurgical status of brain function, and to determine the laterality of language in older children and adults. Presurgical assessments should test all of the domains of cognitive development if possible (motor skills, language development, academic skills, and behavior). Regression of the developmental quotient or of learning abilities is not unusual in patients with severe epilepsy. Very severe cognitive deficits might reflect bilateral involvement and thus indicate a poor prognosis with regard to seizure outcome.<sup>60</sup>

Depending on the patient’s need and the center’s facilities, additional testing such as neuropsychological assessment, the WADA test, SPECT, magnetoencephalography, and functional MR imaging can be performed. All data should be discussed in a multidisciplinary manner (neurology, neurosurgery, neurophysiology, and neuropsychology).

### Management Decisions

After the preoperative investigations, a recommendation can be made as to whether hemispherotomy should be undertaken.

In making the decision, the results of the tests relate to one of the following areas: 1) the presence of a structural lesion that affects one entire hemisphere or at least 3 lobes; 2) the presence of a seizure focus or foci that arise unilaterally; and 3) the presence of a neurological deficit (hemiparesis or hemianopsia).

With those results, the multidisciplinary team can discuss the necessity and timing of such an operation.

### Surgical Anatomy

The common goal of all of the hemispherotomy techniques, as well as the earlier hemispherectomy and functional hemispherectomy interventions, is the interruption of the corpus callosum, the internal capsule and corona radiata, and the mesial temporal structures as well as the frontal horizontal fibers.

A short review of the association and commissural fibers will help in understanding the 3D anatomy and the disconnection procedures.

The major long association bundles are the superior and inferior longitudinal fasciculi, the superior and inferior occipitofrontal fasciculi, the uncinate fasciculus, and the cingulum. The arcuate fasciculus connects the frontal lobe and the parietotemporooccipital region. It arches around the posterior end of the sylvian fissure and joins the superior longitudinal fasciculus.

The uncinate fasciculus connects the inferior temporal

lobe to the orbital surface of the frontal lobe. The cingulum connects the cingulate gyrus, the parahippocampal gyrus, and the septal area; it is part of the limbic system.

Commissural fibers connect the 2 hemispheres. The main commissures are the corpus callosum, the anterior commissure, and the hippocampal commissure. The corpus callosum is composed of the anterior genu (connecting the frontal lobes), the rostrum, the body (connecting the posterior portions of the frontal lobes and the parietal lobes), and the splenium (temporal and occipital connections). The anterior commissure connects the 2 temporal lobes, but it also contains decussating fibers from the olfactory tracts, and is part of the neospinothalamic tract. The hippocampal commissure joins the 2 crura of the fornix.

All connections between the diseased hemisphere and the healthy one have to be interrupted in order to achieve a good outcome in hemispherotomy.

### Surgical Techniques

The vertical parasagittal hemispherotomy (described by Delalande et al.<sup>18</sup>) and the latest version of the periinsular hemispherotomy (described by Villemure and Daniel<sup>60</sup>) are illustrated in the right and left hemispheres, respectively, in the 3 illustrations in Fig. 1.

*Vertical Parasagittal Hemispherotomy.* The patient is placed supine, in a state of general anesthesia, with the head in a neutral, slightly flexed, position. A linear transverse incision is performed, allowing for a small parasagittal frontoparietal craniotomy (3 × 5 cm, 1–2 cm from midline, 1/3 anterior and 2/3 posterior to the coronal suture).

A limited cortical resection (3 × 2 cm) of the frontal cortex is performed to reach the ependyma of the lateral ventricle. Upon entering the lateral ventricle, the surgeon identifies the foramen of Monro and the posterior aspect of the thalamus. The corpus callosum is found by following the roof of the lateral ventricle mesially.

The body and splenium are resected to the roof of the third ventricle and the arachnoid cisterns are exposed. The midline is identified as the falx cerebri and is close to the upper part of the corpus callosum, and anteriorly, the pericallosal arteries are seen. Posterior disconnection of the hippocampus is achieved by cutting the posterior column of the fornix at the level of the ventricular trigone. The vertical incision is performed lateral to the thalamus, guided by the choroid plexus of the temporal horn, then following the temporal horn from the trigone to most anterior part of ventricle, keeping the incision in the white matter.

The callosotomy is then completed by resecting the genu and the rostrum of the corpus callosum to the anterior commissure. The next step is the resection of the posterior part of the gyrus rectus, which will allow the visualization of the anterior cerebral artery and optic nerve and provide enough space for the last disconnection step—a straight incision anterolaterally through the caudate nucleus from the rectus gyrus to the anterior temporal horn. The disconnection of the diseased hemisphere is then complete (Fig. 2).

*Periinsular Hemispherotomy.* The patient is placed supine, in a state of general anesthesia, with a cushion under the ipsilateral shoulder and the head turned almost horizontally. A “barn-door” incision is made, centered on the insula, with a bone window from the coronal suture, to



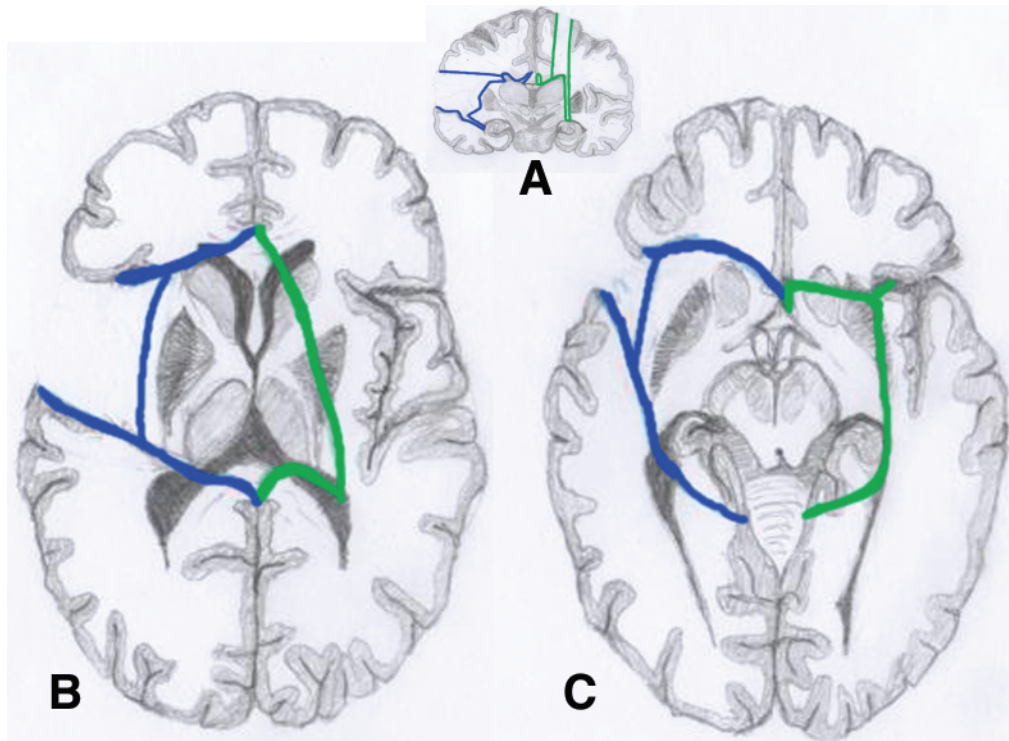


FIG. 1. Schematic drawings of the 2 hemispherotomy techniques. Each hemisphere shows a single technique with the left hemisphere (*blue*) representing the periinsular hemispherotomy and the right (*green*) representing the vertical parasagittal hemispherotomy. A: Coronal view. B and C: Axial views. (In order to have only 2 axial cuts, some structures are represented on the same schema even if they are anatomically at different levels).

3–4 cm posterior to the external auditory canal. The inferior part should be just above the middle fossa, and ideally should go high enough, to the mid-convexity, to provide access to the suprasylvian circular sulcus. Adequate exposure would provide access to the brain 2–2.5 cm below and above the sylvian fissure. The dura mater is reflected either caudally or rostrally. The resection and disconnection follows 3 major stages: the supraisular window, the infrainsular window, and the insular stage.

**The Supraisular Window.** The frontal and parietal opercular cortex are resected, allowing the surgeon to visualize the insula as well as the vessels of the sylvian fissure. The corona radiata is reached then transected on a perpendicular plane when reaching and opening the ventricle. The ventricle should be opened from the frontal horn to the trigone. Once the ventricle is opened, the corpus callosum can be identified. A full transventricular callosotomy is then carried out. The orientation and localization is confirmed with the falx and the pericallosal vessels.

At the level of the splenium, the extension of the medial incision anteriorly to reach the choroidal fissure will interrupt the fimbria-fornix and disconnect the hippocampus.

The last step of this stage consists of disconnecting the frontal lobe just anterior to the basal ganglia, going from the rostrum in the direction of the sphenoid wing, while staying in the frontal horn.

**The Infrainsular Window.** In this stage, T1 is removed from the uncus to the posterior insula. Again, the circular sulcus can be reached then transected when opening the temporal horn from its most anterior aspect to the trigone.

The uncus and amygdala are removed, as well as the anterior hippocampus to the choroidal fissure. In this way, the temporal lobe is disconnected.

**The Insular Stage.** The insula can be resected by subpial aspiration or undermined with an incision at the level of the claustrum/external capsule. The disconnection is completed (Fig. 3).

**General Considerations.** Irrespective of the technique, during the whole procedure, the resections are done subpially and the arteries and veins have to be preserved. Hemostasis is achieved. An external ventricular drain is not required in most cases, but it is usually left in place for 48 hours in patients with hemimegalencephaly. Subgaleal drains are sometimes also left in place and removed within 48 hours. Careful monitoring of the CSF outflow will prevent complications.<sup>17</sup>

**Variations.** Variations of these surgical techniques have been described, including the transcortical subinsular hemispherotomy,<sup>56,64</sup> the hemispheric deafferentation,<sup>53</sup> the transsylvian functional hemispherotomy or transsylvian keyhole functional hemispherectomy,<sup>7,54</sup> or the transopercular hemispherotomy. These variations follow the general principles of 1 of the 2 techniques described with small technical variations. For example, the transsylvian functional hemispherotomy consists of a transsylvian exposure and resection of the mesial temporal structures through the temporal horn. The lateral ventricle is then opened, allowing for disconnection of the frontobasal white matter and the transventricular callosotomy, as well as the occipi-

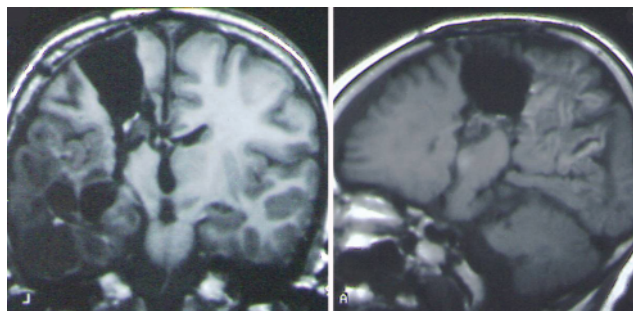


FIG. 2. Coronal (left) and sagittal (right) T1-weighted MR images of a vertical parasagittal hemispherotomy.

toparietal disconnection.

## The Use of Neuronavigation

Since the normal anatomy can be distorted in some patients, and therefore, the landmarks of hemispherotomy more difficult to find, some centers use neuronavigation. The addition of neuronavigation to the surgical planning can also help in the reduction in size of the craniotomy. In patients with large CSF-filled cavities, such as in cases of perinatal ischemic insults, it must be kept in mind, however, that some shift might occur after the opening of the dura, reducing the precision of neuronavigation.

## Postoperative Management

Depending on the age of the child and the etiology of the seizures, postoperative management may vary. In general, the patient would spend the first night in the intensive care unit. In young children, a blood transfusion might be required. Low-grade fever can be seen as well as other symptoms of “aseptic meningitis” such as lethargy, decrease in appetite, and irritability following the procedure. This can be explained by the contamination of the CSF by blood during the procedure.<sup>18,61</sup> In patients who had only partial motor deficit preoperatively, there is a sudden postoperative worsening of symptoms, which will decrease with time; patience and physiotherapy will enable the child to regain his ambulation as well as some motor function in the upper extremity.

Anticonvulsant treatment should be pursued during the period of hospitalization and for at least 3 months afterward. If there are no seizures by then, the medication dose can be slowly tapered; however, at some centers the same regimen is maintained for at least 1 year after surgery.

## Surgical Outcome

There are different points of view in assessing the outcome of surgery. Seizure outcome is the primary concern, but the morbidity associated with the different procedures—new neurological deficits, requirement for CSF diversion procedures, blood loss, and quality of life after surgery—has to be taken into account.

Outcomes vary depending on the etiology of the seizures. In reports of large case series (> 10 cases) involving hemispherotomy, published since 1995,<sup>7,9,10,18,21,22,35,54,55,60,65</sup> between 43 and 90% of patients have been described as seizure-free (Engel Class I) after surgery (Table 1). It is

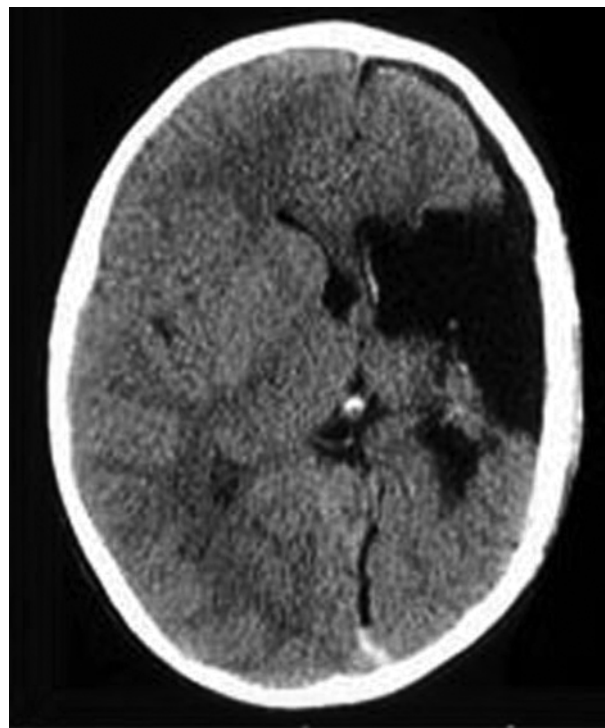


FIG. 3. Computed tomography scan of a periinsular hemispherotomy.

difficult to compare the different series, as the populations differ not only in terms of demographic variables (age, sex), but also with respect to seizure duration and, most importantly, with respect to the etiology of the seizures. In addition, while some authors use the Engel classification when assessing outcome, others do not and instead report patients being either seizure free or not.

In all of the case series in which outcomes were analyzed with respect to etiology,<sup>18,21,32,35,45,55,60</sup> there was agreement on the wide difference in outcomes between the different pathological conditions. In publications on hemispherotomy, the percentage of patients with Rasmussen syndrome, Sturge–Weber syndrome, and infantile hemiplegias who are reported to be seizure free postoperatively (73–93%) is higher than those with multilobar dysplasia or hemimegalencephaly who are seizure-free postoperatively (63–80%, Table 2).<sup>7,18,60</sup>

A multicenter study reported by Holthausen et al.<sup>32</sup> in 1997 included 333 patients who underwent hemispherectomy at 13 different centers. The authors reported a higher seizure-free percentage in the “hemispherotomy” group, with 85.7% of patients being seizure free compared to hemispherectomies. Again, Rasmussen and Sturge–Weber syndromes and vascular insults had a better prognosis (94.6% of patients became seizure free) than did multilobar cerebral dysplasia and other etiologies (68% seizure free).<sup>9,10,18,21,22,34,36,55,60,65</sup>

When comparing different techniques, the seizure outcome seems to be constant. Another case series comparing anatomical hemispherectomy, functional hemispherectomy, and hemispherotomy<sup>12</sup> showed no significant differences between the 3 groups, with 71% of patients overall

TABLE 1  
Comparison of surgical techniques and their outcomes\*

Authors & Year	Technique	No. of Patients	% Engel Class I	Morbidity/ Mortality Rates	Mean FU (yrs)
Binder & Schramm, 2006	transsylvian hemispherotomy	49	90	4% hydrocephalus; 2% mortality	4.3
Delalande et al., 2007	parasagittal vertical hemispherotomy	83	74	16% hydrocephalus; 4% mortality	4.4
Schramm et al., 2001	keyhole transsylvian hemispherotomy	20	88	5% mortality; 5% infection	3.8
Villemure & Daniel, 2006	periinsular hemispherotomy	43	90	2% hydrocephalus; 2% mortality; 5% hemorrhage	9
Cats et al., 2007	functional hemispherectomy	28	78	4% mortality; 7% hydrocephalus	3.3
Devlin et al., 2003	functional + anatomical hemispherectomy	33	52	9% hydrocephalus	3.4
Kestle et al., 2000	functional hemispherectomy	11†	73	0% hydrocephalus	1
Shimizu, 2005	functional hemispherectomy	44	66*	16% hydrocephalus	>1
Vining et al., 1997	hemispherectomy	58	54	7% mortality; 28% hydrocephalus	6.2

\* Patients with Engel Class I and II outcomes combined. FU = follow-up.

† +5 w/ hemispherectomy.

being seizure free at 2 years after surgery. There was a slightly better outcome in the hemispherotomy group (83%) compared with the functional (73%) and anatomical (59%) hemispherectomy groups.

Hemiparesis is generally more important in the upper than in the lower extremities. In one case series in which the authors studied quality of life after hemispherotomy,<sup>18</sup> 84% of the children were able to walk either alone or with help, and all children who were able to walk before surgery retained the ability to walk, as shown in other series as well.<sup>39</sup> The spasticity was found to be greater in the Rasmussen and ischemic-vascular sequelae groups than in the other groups.<sup>18</sup> Of these, 14% had voluntary movement of their hands, 36% could use their hands to hold an object, and 50% had no finger movement. Children who underwent right hemispherotomy had an overall better communication outcome than did the children with a left hemispherotomy.<sup>18</sup> The duration of epilepsy before surgery seemed to be correlated with scores. The age at seizure onset was not significant except in association with motor skills, with a better outcome in those patients with later onset of epilepsy. Delalande and colleagues<sup>18</sup> demonstrated a correlation between the preoperative delay and the Vineland Adaptive Behavior score, encouraging earlier surgery.

An increasing number of publications show that hemispherotomy, or hemispheric surgery, can improve the post-

operative development of children by decreasing the number of seizures they experience.<sup>69,71</sup> There are numerous studies in which authors have assessed the cognitive abilities of patients after hemispheric surgery. While some show no change,<sup>4,45</sup> others show improvement in scores after hemispherectomy.<sup>5,38,39,58</sup> The presurgical developmental level seems to be important not only for the capacity of the brain to improve,<sup>2</sup> but also for seizure outcome.<sup>60</sup>

Again, patients with severe cortical dysplasia or hemimegalencephaly consistently show worse postsurgical outcomes than do those with other etiologies.<sup>3</sup> Brain plasticity is challenged with an operation such as hemispherotomy. There seems to be a higher improvement of verbal skills as compared with nonverbal skills, independent of the affected hemisphere,<sup>21,24,42</sup> and a late plasticity for language has been shown after left-sided hemispherotomy.<sup>31</sup> Early surgery has been suggested to have a positive effect on cognition.<sup>13,70</sup> In infants with epilepsy, even though we might think that plasticity is greater (and therefore functional recovery should also be greater), seizures might interfere with functional reorganization during a critical period, and therefore, if surgery is delayed, the outcome might not be as good as expected. Communication skills might be better in children who undergo surgery on the right side than those who undergo surgery on the left.<sup>18</sup>

The rate of shunt placement for hydrocephalus varies

TABLE 2  
Percentage of patients with Engel Class I outcomes in large case series, stratified by underlying pathological condition\*

Author & Year	Op	Follow-Up	Cortical Dysplasia	HME	Rasmussen	Vascular	Sturge-Weber
Carson et al., 1996	hemidecortication	mean 7.4 yrs	35†	35†	73	NA	50
Cats et al., 2007	functional hemispherectomy	mean 3.3 yrs	90	0	50	90	NA
Delalande et al., 2007	hemispherotomy	mean 4.4 yrs	63†	63†	80	73	80
Devlin et al., 2003	functional hemispherectomy, hemispherectomy	>3 yrs	27†	27†	40‡	100	40‡
Di Rocco et al., 2006	hemispherectomy	mean 9.9 yrs	NA	75	NA	NA	NA
Jonas et al., 2004	hemispherotomy, functional hemispherectomy, hemispherectomy	5 yrs	60	33	63	71	NA
Kossoff et al., 2003	hemispherectomy	range 3 mos–22 yrs	57	40	65	81	67
Shimizu, 2005	functional hemispherectomy	NA	31	80	NA	67	NA
Villemure & Daniel, 2006	hemispherotomy	mean 9 yrs	80†	80†	90	93	NA
Vining et al., 1997	hemispherectomy	mean 6.2 yrs	38†	38†	67	50	NA

\* HME = hemimegalencephaly; NA = data not available.

† The authors did not differentiate between cortical dysplasia and HME in their analyses in these series.

‡ Devlin et al. did not differentiate between Rasmussen syndrome and Sturge-Weber syndrome in their analysis.



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between 2 and 16%, depending on the case series. When analyzed separately, it seems that it is also highly variable depending on the etiology. For example, patients with hemimegencephaly or other multilobar cortical dysplasia will require more CSF diversion procedures than would the patients with ischemic-vascular sequelae. In Delalande et al.'s series,<sup>18</sup> with an overall shunt placement rate of 16%, only patients from those 2 groups required a shunt.

In many series, hemimegencephaly patients have been reported to have the worst seizure and cognitive outcomes as well as the highest rates of complications.<sup>3,34,69</sup>

The nature and rate of complications varies depending on the technique and the case series. In comparison with anatomical hemispherectomy or hemidecortication, both the vertical parasagittal and the periinsular hemispherotomy have relatively small exposures, leading to decreased blood loss, shorter operations, and fewer complications.

If the results of hemispherotomy are compared with the results in some large series of functional or anatomical hemispherectomy (Table 1), there seem, in general, to be fewer complications associated with hemispherotomy. For example, in one series involving patients with hemimegencephaly, up to 53% of patients required a second surgery for a complication<sup>23</sup> or shunt placement. Reporting on their series of pediatric patients who underwent hemispherectomy at Johns Hopkins, Vining et al.<sup>65</sup> described 4 deaths related to surgery (7%) and the need for shunt placement in 16 patients (28%). Carson and colleagues<sup>9</sup> reported 52 cases of hemidecortication, with 96% of the patients being seizure free postoperatively, but there was a mortality rate of 6% related to surgery and death in another patient related to his epilepsy.

### Other Disconnective Techniques

#### *Posterior Disconnection*

Patients suffering from intractable epilepsy due to extensive posterior lesions that do not involve the whole hemisphere can also benefit from disconnective surgeries instead of large resections.

A review of surgical cases from Canada, Switzerland, and India was reported by Daniel and colleagues;<sup>16</sup> these cases involved a total of 13 patients who underwent surgery for a refractory temporoparietooccipital epilepsy.

The surgical technique consists of an extended anterior temporal lobectomy, with partial removal of the hippocampus. There are an additional 4 steps to disconnect the parietooccipital region. A posterior incision is made on T1, keeping the vein of Labbé intact, to reach the ventricle up to the trigone. The incision is then curved, crossing, but preserving, the sylvian vessels, and extending just posterior to the postcentral gyrus to the vertex. The incision is then deepened to reach the falx, transecting all the white matter from the corpus callosum to the sagittal sinus. The fornix is cut later, just anteroinferior to the splenium, interrupting the hippocampic connections.

A variant of the technique is also described; in this variant the surgeon disconnects the temporal lobe instead of performing a temporal lobectomy.

In the series presented, 85% of patients were seizure free (Engel Class I outcome) with a mean follow-up of 6.7

years. One patient had a transient worsening of neurological status, but no other complications were reported.

Four patients who underwent posterior functional hemispherectomy were described by D'Agostino et al.<sup>14</sup> The outcome was not as good in this group, with Engel Class I attained in only one case.

#### *Temporal Disconnection*

There is one series of temporal disconnection procedures that involved 47 patients.<sup>11</sup> The 2-year follow-up data showed 85% of the patients to be seizure free, which compares to outcome after temporal lobectomy. These results are promising but will have to be reproduced to determine whether this procedure is a valid alternative to temporal lobectomy.

#### *Hamartoma Disconnection*

Hypothalamus hamartomas are congenital lesions in the region of the third ventricle and tuber cinereum, their prevalence is 1 in 50,000–100,000. They consist of neuronal and glial cells, are highly epileptogenic, and are not always well controlled by medication. They are sometimes associated with cognitive impairment and endocrine disturbances. Different types of seizures can be seen in cases of hypothalamic hamartoma. The most frequent are gelastic or dacrystic seizures; however, some patients have simple or complex partial seizures, and some generalized tonic or tonic-clonic seizures<sup>28</sup> have been reported as well. While the origin of the gelastic/dacrystic seizures is clearly inside the hamartoma as revealed by invasive monitoring,<sup>6,30</sup> the origin of the other types is less clear.

Different surgical techniques and approaches to the hypothalamic hamartomas have been described, such as pterional, frontotemporal, subtemporal, or transcallosal interforniceal. Since these hamartomas are benign lesions, there is no need for resection and the lesions can be disconnected instead. Some nonsurgical techniques have also been published.<sup>46,49</sup>

Intraventricular hypothalamic hamartoma disconnection was first reported by Delalande and Fohlen<sup>19</sup> (Fig. 4). Since then, a number of case series have been reported in which either endoscopic disconnection or resection was performed.<sup>25,26,40,50,52</sup> The feasibility of the disconnection depends on the plane of insertion. The Delalande classification consists of 4 lesion types: Type 1 has a horizontal implantation plane and may be lateralized on one side; Type 2 has a vertical insertion plane and intraventricular location; Type 3 is a combination of Types 1 and 2; and Type 4 includes all giant hamartomas<sup>19</sup> (Fig. 5). Types 1 and 3 (the intraventricular lesions) can be disconnected endoscopically by an intraventricular approach.

Larger lesions might require more than one disconnection procedure, and some might require a multistep surgical approach, possibly associated with a pterional approach. In lesions that are not approachable by endoscopy, a microsurgical disconnection can be performed.

**Surgical Technique.** Endoscopic disconnection can be undertaken with any rigid endoscope. In the case series reported by Procaccini et al.,<sup>44</sup> the disconnection is described as being performed with the help of the NeuroMate Stereotactic Robot; such a device is not necessary as long as there is an arm, or other fixation device for the endo-

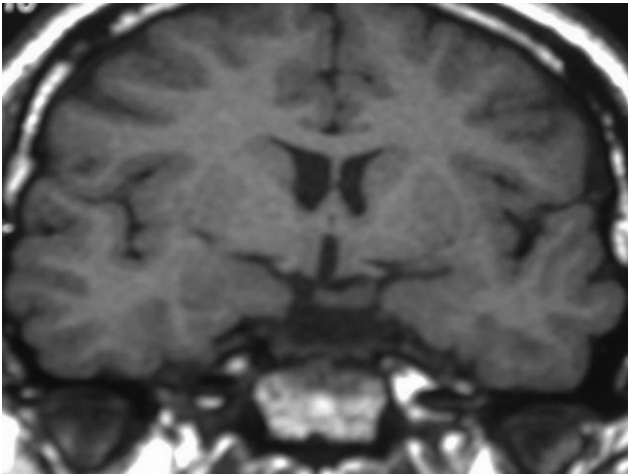


FIG. 4. Coronal T1-weighted MR image obtained after disconnection of a Type 2 hamartoma.

scope, available. Because the ventricles are generally small, neuronavigation is useful in order to find the ventricle and to have a perfect trajectory.

The patient is supine, the head slightly flexed, in a neutral position, either fixed in the robot or in a Mayfield head-fixation device. The entry point is determined by neuronavigation. A small linear incision is performed. A bur hole is drilled, and then the dura mater is opened. The trocar and the endoscope are inserted in the lateral ventricle, following the trajectory. The endoscope is then advanced, through the foramen of Monro, into the third ventricle, and the hamartoma is visualized. The disconnection's target is the base of implantation of the hamartoma. Again neuronavigation, or computer-assisted surgery, is useful, as the limit between the hamartoma and the hypothalamus is not always easy to see. The disconnection is achieved with several monopolar coagulations, under direct vision, up to a depth defined by the diameter of the hamartoma's base along the trajectory on the imaging. In the future, stereoelectroencephalography might help to better understand and find the plane of disconnection.

The case series reported by Procaccini et al.<sup>44</sup> included 33 patients who underwent endoscopic hypothalamic hamartoma disconnection. Of these, 49% were seizure-free and another 49% experienced significant improvement postoperatively. While 54.5% of patients had a single operation, the rest had multistage operations (39.4% had 2 surgeries and 6.1% had 3). There were 2 complications, both related to a combined pterional approach.<sup>44</sup>

Endoscopic resection has been described in a few published series. Ng et al.<sup>40</sup> reported on a series of 37 patients who underwent endoscopic resection. Of these, 49% were seizure free after a mean follow-up of 21 months. However, 3 patients (8.2%) had permanent short-term memory loss. Other series of cases involving resection of a hypothalamic hamartoma have shown comparable results: Rosenfeld et al.<sup>52</sup> described a series of 5 patients who underwent a transcallosal interforaminal approach with no major complications except some appetite increase; 60% experienced seizure-freedom. Feiz-Erfan et al.<sup>25</sup> reported on a series of 10 cases in which various approaches were used (6 transcallosal interforaminal, 1 endoscopic, and 3 frontotemporal).

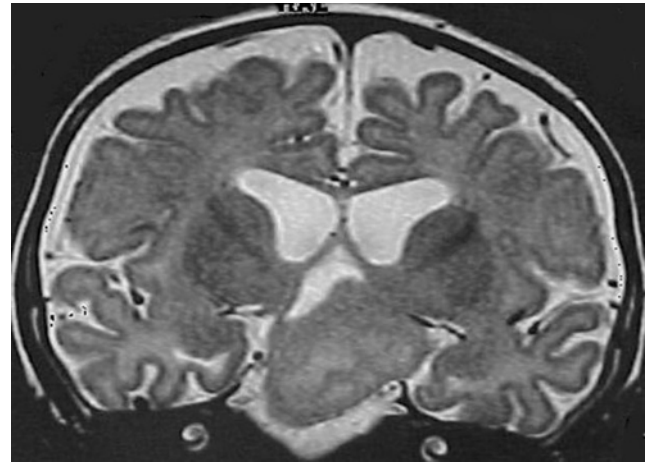


FIG. 5. Coronal T2-weighted MR image demonstrating a giant hypothalamic hamartoma (Type 4), which would require more than 1 disconnection procedure.

ral), with a mean follow-up of 16 months; 60% of the patients experienced seizure free. In this series, 2 patients had mild short-term memory deficit while 2 had moderate short-term memory deficit (20%).

Comparing the disconnection series with the resection series, there is no significant difference in seizure outcome; the percentage of permanent complications, however, seems to be higher in the resections.

The surgical approach depends on the anatomical features of the hamartoma. The endoscopic transventricular approach is a low-risk, well-tolerated procedure, which, when coupled with neuronavigation or computer-assisted frameless stereotaxy, becomes even safer.

## Conclusions

Disconnection procedures are the product of a modification of resection procedures and are based on the concept that interruption of the epileptic discharge pathway would have the same effect as removing the focus. Understanding the 3D anatomy of the brain and its interlobar/interhemispheric connections is essential for success in disconnection procedures. Resecting only a minimal amount of cortical structure is associated with a decrease in short- and long-term complications. The procedures are in general shorter, with a decreased amount of blood loss.

In the treatment of refractory epilepsy, disconnection procedures have a comparable success rate to resection, with respect to seizure outcome. They are, however, generally associated with lower morbidity.

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