Modified periinsular hemispherotomy: operative anatomy and technical nuances

Technical note

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Surgical options for pediatric patients with marked dysfunction of a single epileptogenic hemisphere have evolved over time. Complications resulting from highly resective operations such as anatomical hemispherectomy, including superficial siderosis and secondary hydrocephalus, have led to the development of less resective and more disconnective functional hemispherectomy. Functional hemispherectomy has recently given rise to hemispherotomy, the least resective operation primarily aimed at disconnecting the abnormal hemisphere. Hemispherotomy is effective in decreasing seizure frequency and most likely decreases the risk of postoperative complications when compared with its predecessors. Hemispherotomy is a technically challenging operation that requires a thorough understanding of 3D cerebral anatomy to ensure adequate hemispheric disconnection without placing important structures at risk. The details of germane operative anatomy are not currently available because of the difficulty in exposing this operative anatomy adequately in cadavers to prepare detailed instructive illustrations. Using 3D graphic models, the authors have prepared 2D overlay illustrations to discuss the relevant operative nuances for a modified form of this procedure. Through hemispherotomy, experienced surgeons can effectively treat patients with unilateral epileptogenic hemisphere dysfunction while limiting potential complications.

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Key Words • hemispherotomy • intractable epilepsy • epilepsy surgery • disconnection

In 2006, the Commission on the International League Against Epilepsy recommended that patients who undergo two unsuccessful trials of antiepileptic medication, defined as drug-resistant epilepsy, be referred for surgical evaluation.2 Further indications for surgical consideration in patients with drug-resistant epilepsy include a defined epileptogenic focus and a low likelihood of new neurological deficit postoperatively.21 In adults, the most common type of seizures is medial temporal lobe epilepsy caused by mesial temporal lobe sclerosis.21 The most common epileptogenic origins in pediatric surgical candidates are low-grade tumors and malformations of cortical development. Severe unihemispheric dysfunction is a common finding in pediatric patients with medically refractory epilepsy who are selected for surgical treatment through hemispherotomy. Cortical dysplasia, hemimegalencephaly, polymicrogyria, posttraumatic epilepsy, Rasmussen encephalitis, perinatal stroke, and Sturge-Weber syndrome are some of the most common indications for hemispherotomy.10,11 The goal of surgery in these patients is to interrupt the spreading pathways of the epileptic discharge to isolate the epileptogenic zone.

In this article, we describe the technical nuances of hemispherotomy, a surgical procedure with the goal of disconnecting and isolating the affected hemisphere. Hemispherotomy is a technically challenging operation that requires a thorough understanding of 3D cerebral anatomy to ensure a safe and thorough hemispheric disconnection. The senior author (A.C.G.) describes the technical nuances of a modified form of this procedure. Through hemispherotomy, experienced surgeons can effectively treat patients with unilateral epileptogenic hemisphere dysfunction while limiting potential complications.

Methods

The simplified 3D brain and skull model was purchased from 3DScience.com and relevant cerebrovascular

Abbreviations used in this paper: ACA = anterior cerebral artery; EEG = electroencephalography; MCA = middle cerebral artery.
anatomy was remodeled to reflect accurate anatomy using 3D Studio Max (Autodesk, Inc.). The models were then exported as .obj files and imported into Cinema 4D. Specific anatomy was isolated and the materials were made semitransparent for a greater spatial understanding of the surrounding structures. The isolated 3D models were exported as Collada version 1.4 (.dae) files. Concurrently, a rough pencil sketch of the procedure had been created from surgical videos and scanned into the computer. Using Adobe Photoshop, we imported the isolated 3D models on top of the sketches and altered the scale and rotation to match the deeper operative anatomy through the more superficially visualized structures. This technique facilitated accurate illustration of the ventricles, A\textsubscript{1} branches, falx cerebri, corpus callosum, tentorium, and clinoïd in 3D space in relationship to the exposed operative anatomy.

**Historical Perspectives and Evolution of Techniques**

Hemispherectomy has evolved from hemispherectomy. Walter Dandy first performed the latter operation in 1928 for treatment of malignant gliomas.\textsuperscript{4,11} Hemispherectomy for treatment of epilepsy was first reported in 1938. The first case series, reporting excellent seizure control in 12 patients, was published in 1950.\textsuperscript{9,11,12} Superficial siderosis first case series, reporting excellent seizure control in 12 patients, was published in 1950. Superficial siderosis was later recognized as a potential long-term complication of this procedure. As many as 33\% of patients developed this condition at a median of 8 years after the procedure.\textsuperscript{11,14,18} Superficial siderosis occurs as a consequence of chronic granular ependymitis associated with multiple bleeding areas on the membrane that replaces the resected hemisphere in continuity with the ventricular system, leading to neurological decline, hydrocephalus, and in many cases, death.\textsuperscript{15}

To avoid this complication, different less-resective modifications of the operation were implemented, such as functional hemispherectomy.\textsuperscript{11,15,18} In this procedure, the frontal and occipital poles are disconnected, but not removed, with the goal of achieving similar seizure control to hemispherectomy while preventing the delayed complication of superficial siderosis.\textsuperscript{15} This operation has continued to be modified in an attempt to minimize cerebral resection and also decrease intraoperative blood loss, while maintaining its effectiveness, leading to hemispherotomy techniques (Table 1).\textsuperscript{2,11}

**Preoperative Preparation**

As previously stated, children suffering from refractory seizures who are using 2 appropriate trials of antiepileptic medications should be referred for surgical evaluation. Pediatric patients suffering from epilepsy may develop developmental regression or arrest.\textsuperscript{2} An early referral for surgical evaluation may avoid these adverse effects as a direct result of epileptic syndrome.\textsuperscript{2}

Magnetic resonance imaging remains the most sensitive neuroimaging test available for localization of epileptogenic foci and exclusion of any structural abnormality in the intact hemisphere.\textsuperscript{17} All patients should undergo video electroencephalography (EEG) recording in an attempt to confirm the epileptogenic hemisphere and exclude any potential seizure activity from the intact hemisphere.

**Surgical Technique for Hemispheric Deafferentation**

Multiple variations of hemispherotomy have been described, with the most common ones being perinsular hemispherotomy, modified perinsular hemispherotomy, and vertical parasagittal hemispherotomy.\textsuperscript{22} As described by Morino et al.,\textsuperscript{13} all hemispherotomy techniques have 4 commonalities: resection of medial temporal structures, interruption of the fibers forming the internal capsule and corona radiata, transventricular corpus callosootomy, and disruption of the frontal horizontal fibers. The surgical technique described below is a modification of the procedure first developed by Schramm and colleagues in an attempt to decrease operative time and blood loss, remove less brain tissue, perform a smaller craniotomy, and achieve similar seizure relief when compared with functional hemispherectomies. Wen et al.\textsuperscript{22} advocate using this technique over other hemispherotomy techniques because access to the lateral ventricle is gained through the temporal horn, a technique that is common and familiar to epilepsy surgeons.

**Operative Technique**

**Patient Positioning and Skull Clamp Placement**

The patient is positioned supine on the operating table. The single pediatric pin of the skull clamp is placed behind the ipsilateral ear above the superior nuchal line (Fig. 1A), while the double pin is placed on the contralateral superior temporal line (Fig. 1A inset). This configuration allows for generous exposure of the ipsilateral hemicranium. Skull clamps are generally used in children who are 2 years of age or older and the clamp allows neuronavigational systems to be fixed to the patient’s head.

**TABLE 1: Comparison of hemispherectomy and hemispherotomy techniques**

<table>
<thead>
<tr>
<th>Technique</th>
<th>Description</th>
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<tbody>
<tr>
<td>hemispherectomy</td>
<td>largest blood volume loss; superficial siderosis; hydrocephalus w/ highest rate of postoperative shunting\textsuperscript{2}</td>
</tr>
<tr>
<td>functional hemispherectomy</td>
<td>decreased average blood loss when compared to original hemispherectomy procedure\textsuperscript{2}; hydrocephalus w/ lowest rate of postoperative shunting; higher rate of recurrent seizures &amp; need for reoperation\textsuperscript{2}</td>
</tr>
<tr>
<td>hemispherotomy</td>
<td>hydrocephalus w/ moderate shunting rates\textsuperscript{2}; residual seizures (lower when performed by an experienced operator); postoperative hemorrhage distant to operative site; lowest blood loss; shortest intensive care unit stay; lower overall complication rate\textsuperscript{2}</td>
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We find neuronavigation very helpful, especially during intraventricular dissection, to guide the extent of the genu callosotomy and the medial extent of the frontal fiber tractotomy. A large gel pad is used under the ipsilateral shoulder to avoid significant torsion of the neck while allowing the head to turn approximately 60° from midline.

Incision and Craniotomy

The skin incision is a standard question mark–shaped incision centered on the periinsular region. The temporalis muscle is reflected forward in a single layer with the skin flap, and a craniotomy is performed to expose the periinsular region and temporal lobe. Following the craniotomy, the dura is open and reflected in the direction of the skin flap (Fig. 1B).

Hemispherotomy

The dissection begins with a standard temporal lobectomy (Fig. 1B). This step includes generous neocortical resection (6 cm from the temporal tip) and removal of the medial temporal structures (amygdala and hippocampus). This resection provides the surgeon with a conduit to access the atrium of the lateral ventricle (Fig. 2A). The corpus callosum is now in view and a callosotomy can be performed (Fig. 2C); the location of the corpus callosum, anterior cerebral artery (ACA) complex, and third ventricle have been overlaid in Fig. 2C at the depth of the surgical field to provide an appreciation of the complex operative anatomy and the working angles necessary for the callosotomy.

Neuronavigation may assist in this step to confirm the correct plane of disconnection. We have not used ultrasound guidance due to the narrow transventricular operative corridor. As the accompanying illustrations illustrate the operative view, the operator’s working depth in reality is often deep. The dissection is carried through the corpus callosum until the pericallosal segment of the ACA is identified within the callosal sulcus, lying just inferior to the cingulate gyrus (Fig. 3A). It is often helpful to create an incision in the coronal plane to identify the artery and

Fig. 1. A: The patient is positioned supine on the operating table. The single pediatric pin of the skull clamp is placed behind the ipsilateral ear above the superior nuchal line, while the double pin is placed on the contralateral superior temporal line (Inset). A large gel pad is used under the ipsilateral shoulder to avoid significant torsion of the neck while allowing the head to turn approximately 60° from midline. B: A scalp flap is turned and the dura is opened to expose the hemisphere of interest. The craniotomy must be large enough to expose the frontal, parietal, and temporal opercula, as well as the temporal pole and portions of the frontal lobe lying superior to the sphenoid wing (all outlined in dashed red line). The dissection begins with a standard anterior temporal lobectomy and amygdalohippocampectomy, exposing the temporal horn. The portion of the ventricular system that needs to be later unroofed is shown in light blue. C: Following anterior temporal lobectomy and medial temporal lobe resection, the supramarginal gyrus is excised. This provides access to the atrium of the lateral ventricle. Care should be taken to avoid damaging the bridging branches of the MCA, which are preserved to avoid ischemia to the distal convexity cortices. Copyright Aaron A. Cohen-Gadol. Published with permission.
then continue with an incision in the parasagittal plane to start the callosotomy.

Identification of the pericallosal segment of the ACA ensures that the dissection has not crossed the midline, placing the contents of the contralateral intact hemisphere at risk. Following identification of more proximal A2 branches, the genu callosotomy is extended anteriorly while following the A2 branches through their encasing arachnoid membranes. Due to significant thickness of the genu, this arterial landmark and neuronavigational guidance is important to facilitate surgical orientation. The genu callosotomy is stopped at a coronal plane approximately 5 mm short of the foramen of Monro to avoid injury to the diencephalic structures.

The callosotomy is then carried posteriorly through the body and splenium of the corpus callosum. Aside from the pericallosal arteries, we use the falx cerebri as an important landmark that can be pursued while performing the body and splenium callosotomy (Fig. 3B). This landmark is especially effective during transection of sple-
nium as small distal A\textsubscript{3} and A\textsubscript{4} branches are not easily identifiable within the interhemispheric fissure. Following completion of callosotomy, we turn our attention to the posterior extent of our hippocampectomy, where the horizontal edge of the tentorium is exposed and the tail of the hippocampus is transected subpially. White matter dissection through the calcar avis and medial wall of the atrium allows us to pursue subpially the horizontal edge of the tentorium leading to its ascending segment (Fig. 4A). The disconnection proceeds superiorly following the ascending aspect of the tentorium until the last portion of the splenium of corpus callosum, calcar avis, tail of the hippocampus, crus fornices, cuneus, and precuneus are disconnected during this step\textsuperscript{22} (Fig. 4). Pursuing the junction of the tentorium to the falx cerebri further assists the surgeon with confirming complete white matter disconnection along the splenium. The horizontal and ascending segments of the tentorium, as well as the falx cerebri, have been imported on Fig. 4B to allow for the operator’s orientation.

Care should be taken to stay subpial during this transection and preserve branches of the posterior cerebral artery crossing through the medial and inferior arachnoid membranes.\textsuperscript{22} The dissection within the atrium of the lateral ventricle must be performed posterior to the choroid plexus to ensure preservation of the thalamus. Once the posterior deafferentation is completed, the basal surface of the frontal lobe is disconnected. For this portion of the dissection, the lesser wing of the sphenoid bone can be used as a landmark to guide tractotomy of basal frontal fibers.\textsuperscript{22} We limit our dissection to the lateral part of the clinoid to avoid injury to the hypothalamus (Fig. 5A). Anterior cerebral arteries are skeletonized to the level of the skull base to assure adequate transection. At the completion of this step, the operator exposes the frontal horn of the lateral ventricle and the area of previous genu callosotomy through the frontal lobe. The bony clinoid has been illustrated at the depth of the field and a green arrow outlines the route of disconnection (Fig. 5A). Finally, the insular cortex is removed while MCA branches are protected (Fig. 5B).\textsuperscript{22} We have previously described the use of intraoperative scalp EEG to confirm hemispheric deafferentation.\textsuperscript{8} It is also important to confirm visualization of the pia on the medial side of all the disconnection routes to ensure complete hemispheric deafferentation. We do not routinely place a catheter within the ventricle for temporary postoperative drainage unless perfect hemostasis is not possible. The ventricles are copiously irrigated to remove debris.

Complications

Residual Seizures and Potential Reasons for Suboptimal Disconnection

Residual seizures most often occur when deafferentation is incomplete. In a retrospective analysis performed by Shimizu,\textsuperscript{17} 3 (7%) of 44 patients who underwent hemispherotomy were found to have incomplete disconnection of the corpus callosum. The possibility of this complication may be minimized by exposure of the pericallosal arteries and falx cerebri as described above. The use of intraoperative EEG on the ipsilateral occipital lobe and the contralateral hemisphere may assist with confirmation of callosotomy.\textsuperscript{8}

Inadequate basal frontal lobe disconnection and insular resection have also been implicated in seizure recurrence.\textsuperscript{19} We have defined the extent of our basal resection along the clinoid to ensure complete disconnection. In addition, inadequate tractotomy along the splenium and medial wall of the atrium may lead to suboptimal seizure outcome. We have therefore modified the previous techniques to pursue the horizontal and ascending edges of the tentorium to ensure complete disconnection in the region. Partial genu callosotomy is another potential reason for inadequate hemispheric disconnection; neuronavigation,
along with $A_2$ branches, provide adequate surgical roadmaps.

Patients suffering from recurrent seizures should undergo MRI and diffusion tensor imaging evaluation to assess the completeness of hemispheric deafferentation. We do not routinely perform postoperative MRI. Due to working space limitations within the ventricles, patients with hemimegalencephaly are likely to be at a higher risk of inadequate disconnection.14

**Postoperative Hemorrhage**

Villemure and Daniel20 described 2 cases of postoperative intracranial hemorrhage at sites distant from the original operation (periinsular hemispherotomy). The authors hypothesized that these hemorrhages resulted from excessive drainage of CSF resulting in sudden changes of intracranial pressure dynamics. They suggest avoiding rapid CSF drainage through external ventricular drainage systems and keeping drainage systems set to slightly positive pressures to mitigate the risk of this complication.20

**Hydrocephalus**

Although hydrocephalus occurs less commonly after hemispherotomy than hemispherectomy, it continues to complicate approximately 2%–15% of operations.5,6 Hydrocephalus necessitating ventriculoperitoneal shunt placement is more likely to occur when the underlying pathology is hemimegalencephaly or another multilobar cortical dysplasia.5

**Outcomes**

A number of studies have confirmed the effectiveness of hemispherotomy in appropriately selected patient populations. In a retrospective study evaluating the outcomes of 49 pediatric patients who underwent either functional hemispherotomy or periinsular hemispherotomy, Limbrick et al.10 found that 38 patients (77.6%) were free of disabling seizures (Engel Class I outcome) at a mean follow-up of 28.6 months. Of the 11 patients who continued to have seizures postoperatively, all demonstrated at least worthwhile improvement from their preoperative status (Engel Class III outcome).10 In a similar study evaluating periinsular hemispherotomy in 43 pediatric patients, Villemure and Daniel20 found that 34 (90%) of 37 patients remained free of disabling seizures (Engel Class I outcome) after 9 years. The other 3 patients experienced only rare disabling seizures (Engel Class II outcome).20 In this study, patients suffering from infantile hemiplegia secondary to stroke (unilateral internal carotid artery or MCA) and Rasmussen’s encephalitis had the highest likelihood of attaining a favorable outcome (93% and 90%, respectively), while patients presenting with neuromigration disorders were least likely to attain a favorable outcome (80%).20 The authors attributed this difference to the purely unilateral nature of stroke and Rasmussen’s encephalitis.20

**Conclusions**

Hemispherotomy is an effective treatment for pediatric epilepsy resulting from marked dysfunction of a single cerebral hemisphere. Although technically more challenging than previously developed operations for unilateral hemispheric epilepsy, the procedure has the potential to provide adequate relief from seizures while limiting postoperative complications.
Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: Cohen-Gadol, Kovanda, Rey-Dios. Acquisition of data: Cohen-Gadol, Kovanda, Rey-Dios. Analysis and interpretation of data: all authors. Drafting the article: all authors. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Cohen-Gadol. Study supervision: Cohen-Gadol. Created the illustrations: Travnieck.

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