DISCONNECTIVE procedures have been a common treatment for intractable epilepsy. For the last 2 decades disconnective procedures have evolved from anatomical hemispherectomy, to functional hemispherectomy, to hemispherotomy, which is the latest development in disconnective surgeries and represents a less invasive microsurgical procedure to disconnect the cortex of the affected hemisphere.

The move toward hemispherotomy started with Dandy’s technical description of anatomical hemispherectomy to treat brain tumors. Ten years later, McKenzie applied the technique to manage intractable epilepsy. However, anatomical hemispherectomy was not popularized until a case series published by Krynow demonstrated good results with seizure control and cognitive development. Subsequently, anatomical hemispherectomy became a popular procedure in the treatment of unilateral hemispheric epilepsy-related syndromes (such as neonatal infarcts, hemimegalencephaly, Rasmussen encephalitis, and Sturge-Weber syndrome). However, early and delayed complications from this procedure were increasingly reported, including excessive blood loss, metabolic imbalances, coagulopathy, superficial cerebral hemosiderosis, and development of hydrocephalus. Variations of the anatomical hemispherectomy technique were described to minimize the incidence of such complications, but hemidecortication and subdural space reduction techniques ultimately did not result in significant reduction of associated complications.

In 1974, Rasmussen introduced the functional hemispherectomy, which is a surgical method that involves removing less cerebral tissue and disconnecting the remaining tissue. Functional hemispherectomy is meant to achieve the same physiological goal of anatomical hemispherectomy in terms of seizure control. The overall goals of any functional hemispherectomy to achieve a complete disconnection are resection of the medial temporal structures, disruption of the internal capsule and corona radiata, intraventricular corpus callosotomy, and disruption of the frontal horizontal fibers. Based on a recent review, disconnective procedures have been categorized as Rasmussen, vertical, and lateral approaches. The lateral approach, or periinsular hemispherotomy, was derived from modifications on the functional hemispherectomy and involves removal of the temporal lobe mesial structures, exposure of the atrium via the circular sulcus, internal capsule transection under the central sulcus, intraventricular callosotomy, and frontobasal disconnection. The purpose of this article is to describe and illustrate in detail the anatomy and operative technique for periinsular hemispherotomy, as well as to discuss the nuances and issues involved with this procedure.

KEY WORDS • epilepsy • hemispherectomy • periinsular hemispherotomy
The Periinsular Hemispherotomy: Technical Description

This periinsular hemispherotomy is performed while the patient is under general anesthesia and receiving endotracheal intubation. With the patient supine, all pressure points are well-padded, and a small roll is placed under the ipsilateral shoulder. The head is turned 90° to the opposite site and is held using Mayfield 3-point pin fixation. Neuronavigation is registered and can be helpful in delineating the skin incision and craniotomy. We favor a reverse “question-mark” shaped incision centered over the sylvian fissure (Fig. 1). The surgical exposure should span the sylvian fissure, allowing access to the anterior and posterior temporal lobes, the lateral aspect of the frontal lobe, and the parietal lobe. The incision extends inferiorly to the zygoma, allowing adequate exposure of the temporal lobe. The superior limb of the skin incision should be extended at least slightly above the superior temporal line. The craniotomy should extend superior and posteriorly enough to provide adequate access to the anterior horn, body, and atrium of the lateral ventricle (Fig. 1). The dura mater is then opened in a C-shape and reflected anteriorly. The disconnective procedure is carried out in several steps: 1) an amygdalohippocampectomy is performed with or without an anterior temporal lobectomy (Figs. 2 and 3); 2) a transcircular sulcus exposure of the ventricular atrium is made (Fig. 4); 3) a transection of the internal capsule around the basal ganglia and thalamus under the central sulcus is completed (Fig. 5); and the 4) intraventricular callosotomy (Fig. 6); 5) frontotemporal disconnection (Fig. 7); and 6) insular aspiration are achieved.

Amygdalohippocampectomy With or Without Anterior Temporal Lobectomy

The initial step of a periinsular functional hemispherotomy depends on whether lateral temporal tissue is needed for pathological analysis. If tissue is required, the temporal portion of the procedure can be simplified by performing an anterior temporal lobectomy (Fig. 2). The initial incision is made along the superior temporal gyrus. The incision is inclined laterally, at a depth of 1 or 2 cm. The corticectomy is carried posteriorly to a distance of 4–4.5 cm from the temporal tip. The corticectomy is deepened into the white matter of the gyrus, angling slightly posteriorly until the temporal horn of the lateral ventricle is entered. The depth of the temporal horn from the cortical surface should be determined from the MRI scan and neuronavigation; it usually lies 3–3.5 cm from the surface. Once the ventricle is entered, a small cottonoid is placed in it. The neocortex is further released by extending the initial corticectomy along the superior temporal gyrus to the temporal tip and completing the orthogonal corticectomy to the floor of the middle cranial fossa. The lateral neocortex is then removed by dissection parallel to the cortical surface at the depth of the white matter just above the ventricle.

After the lateral neocortical resection is complete, the anterior tip of the temporal horn of the lateral ventricle is exposed. An incision is made from the tip of the ventricle to the temporal tip, and orthogonally to the lateral inferior pial edge. Each incision is extended to pia, and the pia over the dura is incised, resecting the parahippocampal cortex to obtain a specimen for pathological analysis. A subpial plane is then developed from the lateral edge medially until the carotid bifurcation is visualized thru the pia and the entorhinal sulcus is reached, confirmed either by inspection or by frameless stereotaxy. This specimen is taken as uncus. Caution should be used to maintain an intact pial membrane as a protective layer to avoid injury to the posterior cerebral artery and the third and the fourth cranial nerves lying deep to the subpial dissection. The amygdala is removed by resecting the tissue laterally and inferiorly to a plane drawn from the M1 segment of the middle cerebral artery visualized thru the pia and the choroidal sulcus (choroidal point). The hippocampus is resected by incising the ependyma over the blueish choroidal point (velum interpositum) between the hippocampus and the choroid plexus and extended posteriorly inferiorly to the hippocampal fissure. A second incision is made inferior to the hippocampus down to the pial membrane and extended to the hippocampal fissure. The hippocampus is then elevated laterally, exposing the skeletonized hippocampal fissure and allowing it to be cauterized and cut away from the choroidal fissure and hippocampus, delivered as an intact specimen.

If a cortical specimen is not required, the periinsular hemispherotomy is begun by an extensive dissection of the sylvian fissure, exposing the circular sulcus (Fig. 3). The cortex of the inferior circular sulcus is resected to the white matter of the temporal stem, and this incision is deepened until entrance into the temporal horn of the

Fig. 1. Artist’s illustration showing the skin incision (dashed line) and craniotomy (shaded area) for periinsular hemisphereotomy. The reverse question-mark incision extends superiorly from the anterior portion of the superior temporal line and inferiorly to the zygoma. The surgical exposure should include the lateral aspect of the frontal and parietal lobes, the sylvian fissure, and the majority of the temporal lobe. Adequate access is necessary to visualize the entire lateral ventricle, including the anterior horn, body, and atrium.
Transcircular Sulcus Exposure of the Ventricular Atrium

After removing the mesial temporal lobe structures, the atrium is identified. The dissection continues with the removal of the operculum of the supramarginal gyrus overlying the atrium of the lateral ventricle to expose the atrium. Branches of the middle cerebral artery that supply the parietooccipital area and large veins should be preserved to minimize cerebral edema or infarcts. Partial resection of the supramarginal gyrus may assist in exposure of the atrium (Fig. 4A–D). After the hippocampal structures have been resected, the free edge of the tentorium is followed posteriorly until its junction with the falx (Fig. 4E and F). As the tentorium ascends, several structures are identified: the tail of the hippocampus, the crus fornix, the calcar avis and bulb of the callosum (medial wall of the atrium), precuneus, and the cuneus. These structures should be disconnected behind the choroid plexus of the atrium. The medial wall of the atrium is disconnected by following the free edge of the tentorium up to the roof of the atrium, which is formed by the splenium of the corpus callosum. This disconnection can be confirmed using neuronavigation.

Internal Capsule Transection Under the Central Sulcus

The pia-arachnoid and cortex of the frontoparietal operculum is incised 1.5–2 cm above the sylvian fissure. The goal is to enter into the lateral ventricle perpendicularly, ideally above the caudate nucleus (Fig. 5A). At this point neuronavigation is useful to confirm the appropriate entrance into the ventricle (Fig. 5B). Conversely, the frontal and parietal opercula can be resected to lessen the need for retraction. Using a subpial dissection technique, the cortex and white matter are resected until the circular sulcus surrounding the insular cortex is exposed (Fig. 5C). Once the ventricle is entered, large cottonoids are placed in the ventricle to avoid blood entrance into the ventricle. This suprasylvian window is extended from the anterior-most aspect of the superior circular sulcus to its most posterior aspect at the level of the ventricle trigone. The cortisectomy should complete a C-shaped disconnection by joining with the prior incision of the posterior circular sulcus (Fig. 5D–F).
Intraventricular Callosotomy

Once the internal capsule transection has been completed, the medial disconnection can be initiated (Fig. 6). Medial landmarks include the septum pellucidum and the lateral ventricle. The slight angle formed by the junction of the corpus callosum and the septum pellucidum is identified and can be used to mark the site of the callosotomy (Fig. 6A and B). Typically the intraventricular corpus callosotomy is performed 3 mm off midline. By initiating the callosotomy with a small (2–3 mm) vertical incision in the superomedial aspect of the ventricular roof, the pericallosal arteries can be delineated. Once the pericallosal artery is identified (Fig. 6E), the ependyma of the ventricular roof is incised and the callosotomy is extended 1 cm at a time to follow the course of the artery. Preoperative T2-weighted coronal MRI is very useful for studying the relationship between the ipsilateral and contralateral pericallosal arteries and the corpus callosum, allowing the surgeon to predict arterial course changes and ensure that only the ipsilateral pericallosal artery is followed. The pericallosal artery is followed posteriorly to connect to the previously made incision in the splenium and anteriorly through the body, genu, and rostrum of the corpus callosum (Fig. 6).

Frontobasal Disconnection

After the intraventricular callosotomy is completed and after the rostrum of the corpus callosum is disconnected (which corresponds to the floor of the anterior horn), the most anterior portion of the interhemispheric fissure is identified. This can be accomplished by identifying the anterior cerebral artery or the anterior communicating artery. The disconnection continues anteriorly to the head of the caudate nucleus and inferiorly through the basal part of the frontal lobe. The edge of the sphenoid wing is identified and used as a landmark to indicate the

Fig. 3. Transsylvian amygdalohippocampectomy. A: Axial T1-weighted MRI of the brain with contrast administration in a 3-year-old child with Sturge-Weber syndrome and intractable epilepsy. B–G: Intraoperative images of the patient with Sturge-Weber syndrome (notice the hyperemic appearance of the cortex) undergoing transsylvian amygdalohippocampectomy. B: The lateral aspect of the frontal and parietal lobes and the temporal lobes have been exposed. The superficial portion of the sylvian fissure has been dissected. C and D: Dissecting through the white matter of the temporal stem into the temporal horn of the lateral ventricle. E and F: The temporal horn has been opened and the hippocampus (h) is seen. G: The head and body of the hippocampus have been resected. H and I: Intraoperative images from a different patient before (H) and after (I) the hippocampal resection.
Functional hemispherotomy

The initial part of the disconnection starts with the mesial temporal structures. There are 4 potential epileptogenic structures with efferent connections: 1) the anterior temporal cortex and paralimbic gyrus via the anterior commissure; 2) the hippocampus via the fimbria-fornix; 3) the amygdala complex via the stria terminalis and projections to the basal ganglia, thalamus, hypothalamus, and brainstem; and 4) the insular cortex via projections to the basal frontal lobe, basal ganglia, thalamus, hypothalamus, and brainstem. Due to the complex continuity of the amygdala and basal ganglia, its resection is mandatory and should be more aggressive. After the neocortical resection of the anterior temporal lobe, the only efferent fibers from the hippocampus are via the fimbria-fornix, and these are sectioned at the level of the atrium. The importance of resection of the insula is debatable and its removal was not found to result in higher seizure control, although resection in the setting of Rasmussen enceph-
alitis may be worth the extra operating time and blood loss.\textsuperscript{24} By opening the rest of the temporal horn posteriorly to the atrium, the sublentiform and retrolentiform components of the internal capsule are sectioned. The incision along the circular sulcus toward the lateral ventricle disconnects descending and ascending fibers of the internal capsule at the level of the corona radiata. The frontoparietal commissural connections are disrupted with a parasagittal intraventricular callosotomy that extends anteriorly to the rostrum of the corpus callosum. At this point the most rostral part of the anterior commissure is still intact. The frontal lobe still has some connections: 1) bifrontal caudal orbitofrontal cortices via the anterior commissure (anterior bundle); 2) projections of the frontobasal cortex via anterior sublenticular fibers; 3) cortical connections between orbitofrontal and insular cortices; 4) uncinate fasciculus; and 5) arcuate fibers (parietooccipital fibers). The frontobasal disconnection disrupts the connections described above.

Advantages of Periinsular Hemispherotomy

The original anatomical hemispherectomy has undergone several modifications to avoid late complications. One of the main complications that typically led to death was superficial cerebral hemosiderosis. This complication appeared to be related to size of the postsurgical cavity, where multiple hemorrhages and subdural membranes tend to occur.\textsuperscript{14,24} All the most recent functional hemispherotomy techniques have the reduction of this cavity in common. Periinsular hemispherotomy leaves a large amount of viable nonfunctional hemisphere. Anatomical hemispherectomy and hemidecortication typically require a large skin and bone flap, usually reaching the midline. This type of craniotomy is related to blood loss and its complications (hypovolemia and coagulopathies), especially given the blood volume in young children.\textsuperscript{2} The small craniotomy required for periinsular hemispherectomy decreases blood loss and operative
time. Another early or late complication of anatomical hemispherectomy is hydrocephalus, occurring in as many as 50% of the patients, which is related to the elimination of subarachnoid space over the operated convexity.7,12, 23 Periinsular hemispherotomy spares a substantial amount of subarachnoid space, significantly decreasing the incidence of hydrocephalus.

Disadvantages of Periinsular Hemispherotomy

Disadvantages related to the periinsular hemispherectomy come from difficult anatomical orientation, postoperative brain swelling, and transventricular tissue manipulation. Incomplete disconnection is a significant risk; it has been observed to be a risk with all types of functional hemispherectomies.16,19 Early postoperative MRI is useful for demonstrating adequate disconnection (Fig. 8); it shows a layer of blood reaching the mesial or basal arachnoid membrane as a sign of complete disconnection.22 Early hydrocephalus is noted in 5%–64% in all types of functional hemispherectomy23.22 Death related to a functional hemispherectomy is very rare; a death related to the procedure was reported,25 but in that case other factors might have contributed, such as aspiration due to mental obtundation secondary to brain swelling. In patients with hemimegalencephaly, we advocate a certain degree of brain resection to allow space for postoperative swelling, temporal lobectomy, or resection of the insular or frontal operculum.

Other Types of Functional Hemispherectomy

Other types of functional hemispherectomy or hemispherotomy use partial resection and hemisphere disconnection and use callosotomy and disconnection of the frontal and occipital lobes. The Rasmussen type involved a resection of the temporal lobe and central part of the frontal and parietal lobes, approximately the length of the corpus callosum, and a subpial disconnection of the frontal and occipital lobe. The vertical approach described by Delalande et al.5 begins with a parasagittal parietal corticectomy down to the lateral ventricle. The corpus callosum is then identified, the roof of the lateral ventricle followed, and its fibers disconnected. Next, the temporal horn is reached through the corona radiata, and the medial temporal structures and the frontal and occipital lobes are disconnected. Recently, Schramm and colleagues21 modified the original technique and used the transsylvian keyhole functional hemispherectomy. Shimizu and Maehara22 also modified the original technique with a modified periinsular hemispherotomy. They proposed a superior window between the inferior frontal gyrus and
the superior part of the insula, and through this space, the medial cerebral arteries are coagulated, the mesial temporal structures are resected, and the rest of the hemisphere disconnected. Recently, Bahuleyan et al., in a cadaver study, demonstrated the feasibility of a minimally invasive endoscopic transventricular hemispherotomy using only 2 bur holes. However, this technique has not been applied to humans.

Conclusions

Periinsular hemispherotomy is an effective procedure that allows complete disconnection and isolation of the diseased hemisphere with a minimal amount of brain resection. Complications from an anatomical hemispherectomy, such as superficial cerebral siderosis and hydrocephalus, are less frequent.

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: Rangel-Castilla, Hwang. Acquisition of data: Rangel-Castilla, Hwang, Al-Shamy, Curry. Analysis and interpretation of data: all authors. Drafting the article: all authors. Critically revising the article: all authors. Reviewed submitted version of manuscript: Jea, Curry. Study supervision: Curry.

Acknowledgments

The authors would like to recognize the assistance and effort provided by Lily Chun in the preparation of the manuscript and acknowledge the work of Kathy Relyea in producing the illustrations.

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Functional hemispherotomy


Manuscript submitted November 16, 2011.
Accepted January 10, 2012.
Please include this information when citing this paper: DOI: 10.3171/2012.1.FOCUS11331.
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