Hemispheric disconnection is an effective surgical option for carefully selected patients with disabling, medically refractory epilepsy. Ideal candidates are typically those with an etiology attributable to diffuse abnormalities of one hemisphere. This can include perinatal strokes, developmental or migration disorders, syndromes such as Sturge-Weber, or progressive processes such as Rasmussen’s encephalitis. Seizure freedom rates can be as high as 90% in the long term, depending on the pathology treated and the technique of hemisphere disconnection.

A progression of disconnection techniques has been described over the past century, each addressing efforts to minimize exposure and complications while still accessing target deep structures. Anatomical hemispherectomy involved full exposure and removal of an entire hemisphere but yielded an unacceptable complication profile including superficial cerebral hemosiderosis and hydrocephalus. Functional hemispherectomy reduced these complications by leaving some of the brain tissue in situ; the recognition that disconnection but not resection could produce results similar to those obtained with anatomical hemispherectomy provided the basis for Villemure’s periinsular hemisphereotomy and Delande’s vertical hemispherectomy. Many modifications have subsequently been described, all with the goal of minimizing both excision and exposure while maximizing disconnection. Regardless of the technique used, full hemispherotomy entails disconnection of the corpus callosum, frontobasal white matter, insular white matter,
and medial temporal lobe structures, including the hippocampus and fornix (Fig. 1). These approaches require a potentially large craniotomy for an adequate operative corridor and good visualization of the anatomy. This extensive surgical exposure can lead to increased surgical morbidity, including hypothermia, coagulopathy and blood loss necessitating transfusion, pain and swelling at the surgical site, complicated wound infections/revisions, and prolonged patient recovery.\(^5\)

In order to minimize the surgical exposure and decrease perioperative morbidity, several groups have reported the development of endoscope-assisted disconnection procedures ranging from corpus callosotomy to complete hemispherectomy (Fig. 2).\(^2,\text{ }4,\text{ }5,\text{ }19,\text{ }27\) The evolution of surgical instruments and techniques has made cranial surgery possible through smaller operative corridors. We aimed to investigate the anatomical feasibility of an endoscope-assisted surgical technique for hemispherotomy at our institution by beginning with a cadaveric study. We then transitioned our technique into clinical practice, and here we present two patients in whom hemispherotomy with complete disconnection was performed via our minimally invasive endoscope-assisted approach.

**Methods**

**Cadaver Study**

Three silicone-injected cadaveric heads were utilized in this study, which was conducted in our neurosurgical anatomy laboratory. Standard surgical instruments, microsurgical instruments, and endoscopic instruments were available for use in the anatomy lab. A Karl Storz (Karl Storz GmbH) 0° straight endoscope was selected for use, with high-definition images from the endoscope camera viewed on a screen. Video recordings and still photographs were captured.

We predefined the steps of the operation with disconnections including corpus callosotomy, frontobasal disconnection, insular cut (and temporal horn unroofing), and hippocampal (fornix) disconnection (Table 1). Thus, the major anatomical points of interest included the corpus callosum, insula, temporal horn, hippocampus, and territorial edge; the frontobasal disconnection would require visualization of the ipsilateral anterior cerebral arteries, middle cerebral arteries, and internal carotid artery. Surgical planning was based on anatomical landmarks and previously published measurements.\(^1,\text{ }5,\text{ }27,\text{ }28\) Disconnections were verified using the endoscope through a temporal burr hole.

**Clinical Application**

After successful hemispherotomy disconnection in three cadaveric heads in the neuroanatomy laboratory, the technique was applied in two pediatric patients with intractable epilepsy who were candidates for open hemispherectomy. Selected patients had had perinatal strokes leading to cystic spaces in the pathological hemisphere, thereby allowing a larger working corridor. The surgical technique is described, as are patient histories, images, and outcomes.

Cadaver lab study did not require IRB approval at our institution. Moreover, neither the use of endoscopic tools nor the anatomical goals of the hemispherotomy surgery were new or experimental. The way we approached, planned, and executed the procedure was technically novel to us. As surgical approach and technique often evolve over time, our endeavor was considered a technical shift and refinement rather than the discovery of a new treatment. Because we were not using a new tool, applying a new technology, or performing an entirely new procedure based on novel pathophysiological underpinnings, IRB approval was not required. Retrospective chart review of the clinical cases for this study did receive IRB approval.

**Results**

**Cadaver Study**

Mayfield pins and head holder were used for fixation and positioning of the cadaveric heads. The positioning was supine with slight flexion. Palpation and anatomical landmarks were used to locate the coronal suture, and a straight 5-cm incision was marked extending 3 cm ante-

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**FIG. 1.** Disconnections for functional hemispherotomy. A: Callosotomy and insular disconnection. B: Frontobasal and mesial temporal lobe disconnections. Green lines indicate lines of disconnection. Image created by Katherine Relyea, MS, CMI, and printed with permission from Baylor College of Medicine. Figure is available in color online only.

**FIG. 2.** A: Incision and mini craniotomy (solid red line) compared to the standard incision for open hemispherotomy (dashed red line). Green circle indicates burr hole. Ghosted light green burr hole shows a supplemental access point to the temporal horn through the middle temporal gyrus. B: Supine, fixed patient position for endoscope-assisted procedure. Image created by Katherine Relyea, MS, CMI, and printed with permission from Baylor College of Medicine. Figure is available in color online only.
rior and 2 cm posterior to the coronal suture (Fig. 3A). A standard question mark–shaped incision for a traditional frontoparietotemporal craniotomy was provisionally marked, with the inferior vertical limb anterior to the ipsilateral ear down to the level of the zygoma (Fig. 3C). This temporal incision would allow a small craniotomy for access to the temporal horn of the lateral ventricle if needed, as well as provide a corridor for visual confirmation of the frontobasal disconnection. A 4 × 2-cm keyhole craniotomy was made with a high-speed drill, with the medial border exposing the midline (Fig. 3E). The dura mater was opened, and the flap was retracted toward the sagittal sinus.

Corpus Callosotomy

We performed an interhemispheric approach to the corpus callosum with endoscopic visualization and dynamic retraction using a Penfield instrument (Fig. 4A). The pericallosal arteries were identified overlying the corpus callosum, and the midline surgical corridor was defined between the vessels. The corpus callosotomy was done with controllable suction, microsurgical instruments, and bipolar cautery. The lateral ventricle was entered, with visual identification of the ipsilateral intraventricular anatomy landmarks, including the septal vein, thalamostriate vein, choroid plexus, and foramen of Monro.

We completed the callosotomy with the anterior extent exposing the anterior cerebral arteries via subpial resection of the white matter of the genu (Fig. 4B) and the posterior extent visualizing the pineal cistern and internal cerebral veins via subpial resection of the white matter of the splenium (Fig. 4C). The anatomy was easily identifiable during this portion of the study, as in an open callosotomy or a transcalsal approach to ventricular pathology.

Disconnection of Insular White Matter

The view from inside the ventricle requires identification of intraventricular landmarks, including the foramen of Monro, caudate nucleus, choroid plexus, atrium, and temporal horn. The temporal horn was accessed from the atrium of the lateral ventricle (Fig. 4D). We followed the choroid plexus and ependymal walls and advanced a rectangular cottonoid patty from the atrium to the ipsilateral temporal horn.

We used bipolar cautery, microscissors, microinstruments, and controllable suction to unroof the temporal horn of the lateral ventricle and disconnect the cortical projection fibers, beginning along the ventricular margin of the caudate. This anatomically corresponds with disconnection of the internal capsule and proceeds from inside the ventricle outward. As the disconnection progressed, especially in specimens without significant ventriculomegaly (and thus a smaller working area), it was imperative to continually identify known landmarks (i.e., falx, choroid plexus, ependyma) to maintain orientation. The aforementioned cottonoid patty served as an additional point of reference in the temporal horn of the lateral ventricle. In the anatomy lab, neuronavigation was not available, though stereotactic navigation would be an additional intraoperative instrument.

Hippocampectomy

After visualizing the entire lateral ventricle, including the temporal horn, and completing the insular disconnection, we proceeded to the hippocampectomy. We used bipolar cautery, microscissors, microinstruments, and controllable suction to remove the hippocampus and fornix, following the anatomy and landmarks identified during the callosotomy.
tion, the hippocampus was identified. The white matter of the hippocampus was disconnected via subpial resection, visualizing the posterior cerebral artery and the third cranial nerve past the tentorial edge. The white matter disconnection proceeded posteriorly in the subpial plane to the tail of the hippocampus, continued to the posterior fornix, and extended until it met the area where the splenium had been resected.

Frontobasal Disconnection
The frontobasal disconnection and insula cuts were made anterior and lateral to the caudate. Subpial resection was carried along the ipsilateral anterior cerebral artery to the bifurcation of the internal carotid artery. Anteriorly, this was connected to the previously unroofed temporal horn.

As the procedure progressed, the disconnected hemispheric cortex required some dynamic retraction by the surgical assistant to allow passage of the working surgical instruments. We frequently referred to known normal anatomical structures such as the major arteries, intraventricular ependymal surface, falx, tentorial edge, and choroid plexus to ensure the maintenance of surgical orientation. At this point, a complete hemispherotomy had been per-
formed, with endoscopic visualization of all disconnections.

Temporal Horn Access

As a possible alternative for access to the temporal horn and hippocampus in the event that full hippocampectomy and frontobasal disconnection could not be completed, we tested a middle temporal gyrus approach. The vertical limb of the traditional question mark–shaped skin incision was opened, and a 2 × 2–cm burr hole superior to the zygoma was created with a high-speed drill. The dura was opened in a cruciate fashion. A corticectomy was performed in the middle temporal gyrus, parallel to the middle fossa floor, until the temporal horn was entered. The opening into the ventricle was extended until it was possible to inspect the hippocampal resection bed and to visualize full resection from the head to the tail. The tentorial edge, posterior cerebral artery, and third cranial nerve were visualized through the arachnoid from this approach via endoscopic visualization. Posteriorly, we visualized the choroid plexus in the atrium of the ipsilateral lateral ventricle. Access and visualization anteriorly to the internal cerebral artery was also ergonomic and feasible through this approach should it be needed for disconnection.

Thus, a complete hemispherotomy disconnection was achieved. The workflow and progression of disconnections are shown in Fig. 5. We recognized that a solo surgeon was not able to easily perform the disconnections with instruments through the endoscopic working channel, nor to have safe control of blood vessels for hemostasis. Thus, we developed the endoscope-assisted method, a three-hand technique in which the surgical assistant holds the endoscope, leaving the surgeon free to operate with both hands. With the endoscope-assisted method, all steps of the surgery could be achieved in the cadaveric study. We confirmed the anatomy and disconnections with endoscopic visualization. Finally, we finished by performing a standard craniotomy on the cadaver models to directly verify anatomy and disconnections.

Clinical Application

The endoscope-assisted technique was translated into
clinical practice in two patients who had been referred for hemispherotomy surgery. We recognized the steep learning curve when converting a large open craniotomy surgery to a minimal access surgery, and this factored into patient selection. Given favorable anatomy with a generous ventricular size and relative paucity of cortical tissue, our patients were appropriate candidates for application of the endoscope-assisted hemispherotomy procedure.

In practice, a multidisciplinary epilepsy conference is convened weekly for surgical epilepsy candidates to review their history, physical examination, imaging, epilepsy workup, and treatment options. Three pediatric epilepsy neurosurgeons attend this conference. A spectrum of surgical options for any proposed procedure is reviewed, taking into account known evidence, specific anatomy, and case details, as well as surgeon and family preferences. The possibility of a smaller cranial opening for hemispherotomy was introduced first in detail to the team and then incorporated into the discussion.

Rationale for surgery and surgical options were reviewed by the epileptologist along with the family. The same information together with additional surgical details and the expected perioperative timeline was discussed at the neurosurgery appointment. The overall goal of surgery was to safely achieve the disconnective cuts for hemispherotomy. The option of a smaller opening with the use of an endoscope was discussed with the family. We were open and direct about the relative novelty of the application of this surgical technique involving a smaller craniotomy opening and use of the endoscope. We gave the parents the information, with the traditional standard frontotemporoparietal-temporal craniotomy represented as the standard surgery. The rationale for the endoscope-assisted hemispherotomy, pros and cons, and the contingency plans for converting to open craniotomy were reviewed. The families knew that their cases were the first and second to be undergoing this specific technique modification at our institution and they shared the decision-making by opting into this modified surgical technique.

Discussion of each case was then held at the pediatric neurosurgery indications conference with the faculty, fellow, resident, and perioperative group to confirm surgical candidacy and evaluate the proposed surgical approach. Perioperative care was coordinated during the high-risk case conference with anesthesia staff and operating room nursing staff.

Translation to the Operating Room

After general anesthesia was induced, patients were positioned supine with the head fixed in a Mayfield head holder, with pin sites selected to allow for the paramedian incision over the coronal suture, with the option for conversion to an open question mark–shaped incision for frontotemporoparietal craniotomy. BrainLab (BrainLab AG) neuronavigation registration was undertaken. We verified that external landmarks corroborated with the BrainLab registration. We marked the midline and coronal suture for external reference. Using the BrainLab in-line trajectory view, we verified that the intended craniotomy opening could reach the genu, splenium, and ipsilateral temporal horn.

A paramedian 5-cm incision was marked 2 cm off the midline. We modified the incision to a paramedian location, as this facilitated the craniotomy. This line was typically over the coronal suture with approximately 3 cm anterior and 2 cm posterior to the coronal suture (Fig. 3B). A full question mark–shaped standard craniotomy for hemispherotomy incision was also marked, with the vertical line of the question mark falling within 1 cm of the ear, ending anterior to the tragus at the zygoma (Fig. 3D). This traditional open-incision marking would be used if the surgery needed to be extended to an open craniotomy. The vertical limb of the question mark shape drawn above the zygoma would serve as the area of access should the second burr hole for accessing the temporal horn become necessary. This entire standard question mark–shaped incision was prepped and draped into the sterile field.

The procedure was performed as previously described, without adverse events. After the scalp was opened and cranial sutures on the skull surface were visible, a 4-cm-long by 2-cm-wide craniotomy was undertaken, with burr holes made on the coronal suture and sagittal suture (Fig. 3F) to expose the dura overlying the sagittal sinus and carefully free up the dura underneath the bone. Neuronavigation was used, with the surgical team being cognizant that distortion after access into the ventricle could lead to a decrease in accuracy. Intraoperative ultrasound was available in the event that additional verification of anatomy was required beyond direct endoscopic visualization and could be used to identify the ventricular anatomy, tentorial edge, and skull base. Though the equipment was on and ready, the use of intraoperative ultrasound was not required in our two clinical cases. Moreover, the temporal burr hole access was not needed; thus, the objectives of surgery were achieved entirely from the single paramedian exposure.

The cortical brain parenchyma in vivo was quite mobile after performing the callosotomy and entering the ventricle because of the large ventricular anatomy and thin cortical ribbons. Thus, the cortex could be gently retracted away from the operative field with wet cotton balls placed between the pathological hemisphere and the falx. The view through the endoscope was much more likely to become blocked by tissue in vivo than in the cadaveric anatomy lab. Therefore, we added controllable irrigation (ClearVision, Karl Storz GmbH), which allowed on-demand clearing of the endoscopic view. In order to maximize the limited working space, an angled endoscope camera (P3, Karl Storz GmbH) was used. This ergonomic addition kept the assistant’s hands away from the working field and corridors of the operative surgeon while still allowing manipulation and dynamic positioning of the endoscope.

After the procedure, a ventriculostomy catheter was left in place in the ipsilateral lateral ventricle via the interhemispheric exposure, exiting from an existing burr hole and tunneled away from the incision. Blood transfusions were not required. Both patients were extubated and observed in the pediatric intensive care unit setting overnight and transferred to a surgical unit on postoperative day 1 to begin mobilization and therapy evaluations. Both recovered well; they were discharged to home within 1 week without the need for CSF shunting and without demon-
strating a need for inpatient rehabilitation. Neither patient experienced postoperative fever. Postoperative MRI with diffusion tensor imaging showed complete disconnection of the fiber tracts (Fig. 6). At the 6-month follow-up, both patients had remained seizure free since surgery.

**Illustrative Cases**

**Case 1**

An 11-year-old boy had a history of cerebral palsy and intractable epilepsy secondary to perinatal stroke since the age of 8 months. Despite medical optimization, he was having up to five seizures per day. Comprehensive multidisciplinary epilepsy workup concluded that he had seizures originating from the left frontoparietal and posterior temporoooccipital region propagating to the anterior temporal lobe. Magnetic resonance imaging showed structural abnormality of the left hemisphere with extensive cystic encephalomalacia consistent with the clinical history of infarction (Fig. 7A and B). He had atrophy of the left cerebral peduncle, and functional studies showed his language lateralized to the right. He was recommended for functional hemispherotomy; the large left-sided cyst was favorable to endoscopic intervention, as it allowed a larger working space without retraction of brain parenchyma.

**Case 2**

A 23-month-old boy had a history of perinatal stroke and medically refractory epilepsy and encephalopathy, leading to developmental delay and failure to thrive. Comprehensive evaluation by the epilepsy team included electroencephalography studies showing frequent high-amplitude multifocal epileptiform activity over the entire right hemisphere consistent with right hemispheric epilepsy. Magnetic resonance imaging showed cystic encephalomalacia of the right middle cerebral artery territory (Fig. 7C and D). He had atrophy of the right cerebral peduncle, and functional studies confirmed his language lateralized to the left. The large cyst made the patient suitable for minimally invasive endoscopic functional hemispherotomy.

**Discussion**

We demonstrated the anatomical feasibility of complete hemispheric disconnection via a minimally invasive endoscope-assisted technique in a cadaver study and subsequently translated this technique into clinical practice with encouraging preliminary results in carefully selected pediatric patients with intractable epilepsy.

Hemispherotomy has been a good surgical option in the treatment of medically refractory nonlesional epilepsy with seizures originating from one hemisphere. Classic techniques have required relatively extensive surgical exposure, which subjects children to the risks of bleeding, blood transfusion, fevers, postoperative aseptic meningitic symptoms, and prolonged recovery requiring extended hospitalization and inpatient rehabilitation stays. The advances of endoscopic and minimally invasive tools provide the opportunity for surgical techniques to also evolve in order to decrease perioperative morbidity and improve patient outcomes.

The anatomy of hemispherotomy has been well described. A thorough understanding of and comfort with this anatomy is essential, as the endoscopic approach uses trajectories and views structures from perspectives different from those previously described; thus, anatomical views may be unfamiliar and disorienting. However, we demonstrated that it is possible to clearly see and disconnect all critical structures using our cadaveric model. In that model, we also verified feasibility by developing back-up approaches to safely access required anatomical structures through additional burr holes and finally through standard craniotomy as necessary; these strategies were not needed in our two patients.

The callosotomy is the relatively straightforward component of the disconnection, as the working corridor is
straight down along the falx, as in a standard interhemi-
spheric approach to ventricular pathology.21 The anatomy
is easily identified at the first ventricular entry into the ip-
silateral ventricle, especially as most pediatric neurosur-
geons are familiar with intraventricular neuroendoscopy.
As the disconnection continues, maintaining orientation
and identifying anatomical landmarks becomes impera-
tive, as neuronavigation may lose accuracy with shifting of
the brain tissue. A long rectangular cottonoid in the tem-
poral horn serves as an indicator of this anatomical land-
mark, especially as the anatomy becomes distorted once
CSF has been drained and the thin cortex of the affected
hemisphere may be sagging from disconnection.

Our experience in the cadaver models allowed optimi-
zation of the technique and instrument manipulation
in a bloodless field. As subpial resection via microsurgi-
tical technique requires clear visualization of specific fields
with a rigid endoscope (genu of corpus callosum anteriorly,
splenium posteriorly, temporal horn of the lateral ventricle
lateral and deep), a dynamic working relationship between
surgeon and operative assistant is essential. For instance,
certain portions of the surgery require the endoscope an-
gled from the anterior margin of the craniotomy with the
surgeon working from the posterior portion of the opening.
Conversely, the frontobasal disconnection was most easily
performed with the endoscope entering from the posterior
edge of the craniotomy and aimed anteriorly, with the sur-
geon’s instruments working above the endoscope along the
anterior edge of the craniotomy. All portions of the opera-
tive field were easily reached with our 4-cm craniotomy.

Several modifications were made in the transition to
clinical application. Optimal positioning is essential for vi-
sualization of critical structures with the rigid endoscope.
The head should be sufficiently flexed to allow for the ap-
propriate angles required to visualize the anterior and pos-
terior disconnection, but with vigilant assessment of the
airway in conjunction with the anesthesia team to avoid
obstruction. It is essential to plan for and prepare the field
for a possible full craniotomy in the event that endoscopic
access is suboptimal and if the goals of the surgery cannot
be achieved safely. Neuronavigation is used, but surgeons
should always be cognizant that technology can be unreli-
able, especially as anatomy shifts during surgery. Intraop-
erative ultrasound can theoretically be a useful adjunct in
identifying known landmarks. However, no surgical navi-
gation technology can substitute for experienced, intimate
knowledge of the surgical anatomy. An angled camera on
the endoscope is useful for simplifying the limited work-
ing space for the traffic of instruments and hands in the
surgical corridor.

Previous studies have shown that patients in whom
hemispherectomy has failed are likely to have remaining
hemispheric connections, particularly anteriorly, and can
benefit from reoperation to complete the disconnection.21
This new technique may raise concerns that complete dis-
connection may be inadequately evaluated with the endo-
scopic view compared to the direct visualization provided
by classic open procedures; therefore, we have high stan-
dards of close inspection and verification of every discon-
nection step. We also carefully reviewed the intraoperative
endoscopic video of every surgery and verified disconnec-
tions radiographically with postoperative tractography in
our patients.

Blood transfusions were not required. Postoperatively,
hemispherotomy patients have external ventricular drain-
age to allow egress of any postoperative intraventricular
blood and protein in the CSF. In our two cases, the drains
were weaned and removed without incident, with rapidly
decreasing output and low intracranial pressures. Patients
recovered quickly, without the postoperative frontotempo-
ral flap soft-tissue swelling and pain that typically
accompany open craniotomy. The paramedian incisions
obviated temporalis muscle disruption. Both patients were
discharged home without new neurological deficits and
without the need for inpatient rehabilitation.

Recognizing limitations of the novel approach and
equipment, as well as strengths and limitations of the op-
erative team, is a crucial area of insight and understanding.
For instance, patients with Sturge-Weber syndrome and
hemimegalencephaly would not have been appropriate for
initial deployment of this endoscope-assisted technology
given our understanding that these cases are challenging
even in standard open craniotomy approaches. Our first
patients for this relatively novel technique had generously
sized ventricles and thin cortical mantles. This allowed for
a larger operative corridor and more working room than
would be possible in the aforementioned pathologies. A
learning curve is expected in the development of any new
technique, and we expect to apply our technique to in-
creasingly varied patients as workflow and surgical team
skill improve.

Proper patient selection with thorough workup by a
multidisciplinary epilepsy team is crucial in any epilepsy
surgery. Careful patient selection is also essential in tran-
slating innovation in surgical technique into clinical prac-
tice. Establishing contingency plans to ensure the achieve-
ment of surgical goals, as well as head off any possible
complications, is imperative. In surgery, the patients were
positioned and marked for extending to standard open cra-
niotomy, with defined intraoperative checkpoints for anat-
omy visualization and strict goals for complete discon-
nection. The surgical and anesthesia teams were prepared
for possible complications as the surgeons worked around
the venous sinuses and cranial vessels. Recognizing pos-
sible limitations and complications while applying rigor
and safety to the development of this endoscopic surgical
technique with subsequent translation into clinical care
has been paramount to this process.

Conclusions
Full hemispheric disconnection can be completed with
endoscopic assistance through a mini craniotomy, mini-
mizing operative exposure. Magnetic resonance imaging
with diffusion tensor imaging allows for verification of
fiber tract disconnection. Further studies and long-term
follow-up are needed to determine seizure freedom and
complication rates compared to those in standard open
craniotomy.

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Disclosures
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
Conception and design: Lam. Acquisition of data: Lam, Wagner, Vaz-Guimaraes. Analysis and interpretation of data: Lam, Wagner. Drafting the article: Lam, Wagner. Critical revision of the article: Lam, Wagner. Reviewed submitted version of manuscript: Lam, Wagner. Approved the final version of the manuscript on behalf of all authors: Lam. Administrative/technical/material support: Lam, Vaz-Guimaraes, Camstra. Study supervision: Lam.

Correspondence
Sandi Lam: Baylor College of Medicine, Texas Children's Hospital, Houston TX. sklam@texaschildrens.org.