Parietal and Occipital Resections
- Parietal resections may be limited to the superior parietal lobule in the dominant hemisphere by sensory cortex and motor cortex.
- Occipital resection should not extend into the angular gyrus of the dominant hemisphere.
- The calcarine region (mesial occipital lobe) is usually responsible for seizure onset.
  - Visual field risk should be discussed with the patient.

◆ Postoperative

- Intensive care unit observation for 24 hours
- Head of bed at 30 degrees
- Venodyne
- Regular diet
- Postoperative MRI scan to confirm extent of resection; may be done in first 48 hours or 2-3 months postoperatively, after resolution of postoperative changes.
- Continue anticonvulsants in conjunction with patient's epilepsy neurologist
- Antibiotics continued for 24 hours
  - Dexamethasone tapered over approximately 1 week

Complications
- Infarct, venous/arterial thrombosis, inadequate seizure relief, unintentional neurologic deficit, wound infection, meningitis, intra- or extra-axial hematoma, cerebral abscess, cerebral edema

Outcomes
- Pediatric and adult patients with extratemporal epilepsy with a nonlocalizing MRI or with nonlesional epilepsy are less likely to be cured of their seizures compared with patients with a focal epileptogenic lesion.
- Seizure freedom after surgical resection is between 30% and 50%.
- Focal cortical dysplasia is the most common finding in surgical specimens.
- Tumor, vascular malformation, nontumor changes, and microdysgenesis account for the remainder of pathologies identified on histology.
- Focal EEG seizure onset in noneloquent cortex identified by invasive recordings may predict a better seizure-free outcome.
- Common reasons for failure include diffuse EEG onset (failure to localize seizure onset) and/or focal onset in eloquent cortex (failure to resect seizure onset).
- Chronic extraoperative mapping is mandatory to define seizure onset zone and functional cortex in majority of nonlesional epilepsy cases.
- Resective strategies are tailored to individual patients with specific neurologic risk depending on anatomic location of epilepsy (motor/sensory, vision, language).

62 Transsylvian Functional Hemispherectomy
Devin K. Binder and Johannes Schramm

Indications
- Unihemispheric syndromes associated with intractable epilepsy
- Common diagnoses:
  - Infantile hemiplegia (e.g., posthemorrhagic)
  - Perinatal cerebral infarct
  - Sturge-Weber syndrome
  - Rasmussen encephalitis
  - Hemispheric cortical dysplasia
  - Hemimegalencephaly
- Hemispherectomy is primarily a pediatric operation, as perinatal, congenital, and early developmental pathologies account for the majority of conditions leading to intractable unihemispheric epilepsy and transfer of hemispheric function works best under the age of 2 years

◆ Preoperative

Overview
- Hemispherectomy refers to a variety of operations that functionally isolate the cerebral cortex of one hemisphere from the rest of the nervous system.
- Modern functional hemispherectomy procedures aim at greater degree of disconnection and less resection than original anatomic hemispherectomies.

Operative Planning and Preoperative Work-up
- In addition to intractable epilepsy, these patients typically present with a contralateral spastic hemiparesis.
- Because no portion of the hemisphere is to be spared functionally, preoperative or functional localization has no specific role; however, confirmation of transfer of language and motor function into the nonaffected hemisphere will affect risks of surgery.
- Structural neuroimaging (especially magnetic resonance imaging) plays an integral role in preoperative evaluation and may reveal malformations, vascular or posttraumatic hemispheric injury, from atrophy to porencephalic cysts, or other disease-specific imaging findings.
- Electroencephalography should lateralize disease to the radiographically abnormal hemisphere (lateralization, not intrahemispheric localization, is important).
- Hemispherectomy is not employed if less extensive surgical therapy (callosotomy or focal or multilobar resection) may be similarly effective.
- Older children with dominant hemisphere disease and some preserved language function should undergo assessment of lateralization of language (e.g., with the intracarotid amobarbital [Wada] test—lack of transfer to healthy hemisphere constitutes a contraindication to surgery).
Timing of surgery determined by severity of epilepsy, the age of the patient, the natural history of the disease, and the adequacy of therapeutic trials of anticonvulsant medications.

Patient Preparation and Anesthetic Issues
- Antiepileptic drugs are not withdrawn and are given the day before surgery.
- In hemimegalencephaly cases or cases with a near normal brain volume, dexamethasone (4 mg, six times daily) is given starting 1 to 2 days before surgery and tapered within 4 to 6 days after surgery.
- Premedication with midazolam (0.5 mg/kg) is followed by induction with thiopental (5 to 7 mg/kg); anesthesia is maintained with remifentanil (0.2 to 0.3 mcg/kg/min) and isoflurane or sevoflurane.
- At least two intravenous lines are placed and an intra-arterial line is placed in the radial or femoral artery; a central venous line is not used routinely but is useful in small infants with expected larger blood loss (e.g., hemimegalencephaly).

Intraoperative (Fig. 62.1)

Overview
- Primary goal of hemispherectomy is to achieve seizure control via complete disconnection of the cortex of the epileptogenic abnormal hemisphere from the "good" hemisphere.
- Many variations of hemispherectomy have been described; this chapter describes the surgical technique for transsylvian functional hemispherectomy, which entails four steps:
  - Linear incision and small craniotomy
  - Transsylvian exposure and mesial temporal resection (uncoamygdalohippocampectomy)
  - Transventricular callosotomy
  - Frontobasal disconnection and transsylvian-transventricular occipitoparietal mesial disconnection

Positioning
- Patient either in a lateral decubitus position or supine with the shoulder elevated
- Head is placed in three-point Mayfield pin fixation and turned so that the frontotemporal region is parallel to the floor and the vertex is tilted down slightly.

Skin Incision and Craniotomy
- Slightly curved incision (10 to 12 cm) is marked from just anterior to the tragus to the superior frontal area short of midline.
- Incision is opened, Raney clips placed, temporalis fascia/muscle split along incision and retracted
- Bone flap is planned with the following approximate borders:
  - Anterior: limen insulae
  - Superior: corpus callosum

Fig. 62.1 Schematic illustrating the operative exposure for a transsylvian functional hemispherectomy.

- Posterior: pulvinar
- Inferior: 0.5 cm below the level of the ascending M1 branch (i.e., overlying the temporal operculum slightly)
- Neuronavigation may be used for ideal placement of the bone flap

Dural Opening and Fissure Dissection
- Standard dural opening and sylvian fissure dissection
- Overall length of dural opening does not need to exceed the length of the corpus callosum, in experienced hands up to 2.5 cm shorter
Ascending M1 branch, the meninx insulae, and insular cortex with M2 and M3 branches are exposed, and the superior and inferior insular sulci (together termed the circular sulci) are identified.

Temporo-Mesial-Basal Resection

- Temporal horn is opened via 25 to 35 mm incision through the inferior circular sulci, revealing the hippocampus, choroid plexus, and choroidal fissure mesially.
- Uncal resection is then performed with following steps.
  - Uncus is resected with the ultrasonic aspirator leaving the arachnoid intact.
  - Mesial disconnection is performed from the choroidal point anteriorly through the tissue of the amygdaloid body and the entorhinal cortex to reach the empty uncual cavity.
  - Disconnection down to the base is then made lateral to the hippocampus using a dissector.
  - Fimbria is disconnected mesially, and the hippocampal vessels in the hippocampal fissure are identified.
  - Hippocampal vessels are coagulated and transected and the parahippocampal gyrus can then be subpially elevated.
  - Combined hippocampus/parahippocampal gyrus can be disconnected posteriorly with the ultrasonic aspirator, and the specimen can be removed en bloc.

Full Exposure of Lateral Ventricle

- Following the outline of the circular sulcus starting from the posterior margin of the opening of the temporal horn, ventricle opened from trigone to tip of frontal horn, leaving most MCA branches intact.
- Retraction of the operculum and/or basal ganglia/insular cortex block may be necessary; the entire lateral ventricle can be viewed through this U-shaped opening; finally, the insular cortex is removed again preserving MCA branches.

Mesial Disconnection

- Divided into three parts:
  - Frontotemporal white matter disconnection
  - Corpus callosotomy
  - Disconnection of mesial white matter of the occipital and parietal lobes
- Frontotemporal white matter disconnected by starting at the opened frontal horn and extending to the base of the frontal lobe just anterior to the M1 and deeper down anterior to the A1 toward the midline.
- Slit created between the tip of the frontal horn and the arachnoid at the base of the frontal lobe; M1 and A1 are visible through the basal arachnoid.
- Basal disconnection is carried medially to the interhemispheric fissure; one can open the arachnoid to make sure that both A2 segments are visualized.
- Transventricular callosotomy is then performed by continuing the disconnection around the genu of the corpus callosum following the pericallosal artery to the superior surface of the corpus callosum (resection of the frontoparietal operculum during this step is useful).
- Paramedian disconnection through the corpus callosum again follows the pericallosal artery, exposing but not violating the arachnoid of the cingulate gyrus.
- At the splenium of the corpus callosum, the inferior border of the falx and anterior rim of the falcotentorial junction become important landmarks that lead posterior to the splenium.

The disconnection is carried more posteriorly through the medial wall of the trigone.
- Calcar avis (ventricular impression of the calcarine fissure) is identified and the posterior cerebral artery crossing the tentorial rim slightly more inferiorly is identified and spared.
- Temporomesially, the final step of the disconnection is facilitated by following the choroidal fissure from the posterior margin of the hippocampal cavity dorsally toward the trigone.
- The temporal disconnection line is then extended posteriorly and superiorly through the collateral emience to reach the mesial disconnection line from the callosotomy; during this step, ependymal veins can be carefully coagulated and divided.

Closure

- All arachnoid surfaces are left intact, hemostasis in the mesial white matter disconnection is easily achieved with Surgicel, copious irrigation is used, and the wound is closed in standard fashion.

Postoperative

- Patient brought to the intensive care unit and extubated when fully awake
- Transfusion is rarely necessary for this procedure, mainly in babies or hemimegalencephaly
- Patients are monitored for seizures with attention to known preoperative semiology.

Complications

- Potential operative and postoperative complications include:
  - Hemorrhage
  - Infection
  - Hydrocephalus (late hydrocephalus rate in original anatomic hemispherectomies was ~50% but is much lower in modern functional hemispherectomy series)
  - Cerebral edema
- In a recent multicenter series, mortality was 1.5%.
- Important perioperative advantages of newer techniques include reduction in operative time and blood loss.

Management Pearls

- The transylvanian functional hemispherectomy technique described in this chapter is ideal for atrophic hemispheres with enlarged ventricles and is especially suitable for cases of perinatal infarction and cystic encephalomalacia. However, it is not ideal for hemimegalencephaly, in
Anatomic Hemispherectomy

Jorge Gonzalez-Martinez

Indications

- Ipsilateral hemispheric syndromes associated with medically refractory epilepsy
- Common diagnoses
  - Hemimegalencephaly
  - Hemispheric or extensive focal cortical dysplasia
  - Sturge-Weber syndrome
  - Ipsilateral recurrence of seizures after functional hemispherectomy

Timing of Surgery

- The appropriate timing for surgical intervention is controversial. Many experienced epilepsy centers recommend early intervention to stop seizures and maximize chances for normal neurodevelopment. Despite this, there is little evidence supporting early surgery and the risks related to the surgical procedure (blood loss, hypothermia) in younger patients need to be considered. In general, for less severe epilepsy, we consider a body weight of 10 kg or above acceptable. All patients and/or families are asked to donate blood prior to the operative procedure. For catastrophic hemispheric epilepsy (frequent and severe seizures), surgery is performed earlier with appropriate informed consent on the risks of excessive blood loss and mortality.

Preoperative

History and Physical Examination

- Detailed history including prenatal events, birth and developmental history, and possible epilepsy risk factors are obtained.
- Neurologic examination focuses on sensorimotor, language, and visual function.
- Cognitive function should be generally assessed.
- Degree of motor impairment needs to be accurately documented to help counsel parents on what to expect postoperatively. Similarly, presence or absence of hemianopsia should be assessed and parents need to be counseled about the presence of a contralateral hemianopsia postoperatively.

Clinical Semiology and Video Electroencephalography

- Perform preoperative video electroencephalography monitoring to document seizure semiology and interictal/ictal electroencephalographic (EEG) data. Seizure type(s) and location of epileptic events are documented and characterized.