CHAPTER 178 ■ MULTILOBAR RESECTIONS AND HEMISPHERECTOMY
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INTRODUCTION
Multilobar resection and hemispherectomy are surgical options available for the treatment of medically intractable seizures arising from a diffuse area of epileptogenicity that remains unilateral but extends beyond one lobe. The nature and extent of the underlying pathologic process, the patient’s neurologic status, and the results of preoperative neurophysiologic and neuroimaging studies together determine which surgical procedure should be recommended.

For clarity, hemispherectomy and multilobar resections are presented separately in this chapter.

HEMISPHERECTOMY

Historical Perspectives

The term hemispherectomy refers to a variety of operations that functionally isolate the cerebral cortex of one hemisphere from the rest of the nervous system. In 1928, Dandy and L’Hermite originally described hemispherectomy for diffuse infiltrative glioma, but in the next decade few other reports of hemispherectomy for tumor were published. Over ten years later, McKenzie performed the first hemispherectomy for epilepsy. In 1950, Krynaus reported on hemispherectomies in 12 children with infantile hemiplegia and intractable epilepsy. In this report, he noted excellent seizure control as well as marked and long-lasting behavioral improvement.

Because of the success of the operation for improving seizure control and behavior, hemispherectomy became widely used in the treatment of seizures associated with infantile hemiplegia. In addition, it came into use for the treatment of other unihemispheric syndromes associated with intractable epilepsy, such as Sturge-Weber disease, Rasmussen encephalitis, and cerebral infarct. By 1961, White reviewed 267 cases from many neurosurgical centers. However, enthusiasm for hemispherectomy waned in the 1960s as more effective anticonvulsants became available and as significant long-term complications became apparent. Many patients developed delayed neurologic and intellectual deterioration postoperatively, and resultant mass effect and cerebral hemosiderosis (SCH), and ultimately it became clear that hemorrhage, superficial vascular insults, and ultimately it became clear that hemorrhage, superficial cerebral hemosiderosis (SCH), and resultant mass effect and hydrocephalus were responsible, and even caused significant mortality. Over the past two decades, modifications to anatomic hemispherectomy were proposed, aimed at eliminating the late complications of SCH. Other approaches removed only the cortex, termed hemidecortication or hemispheric excision.

Over the past 10 to 15 years, there have been further development and modification of functional hemispherectomy techniques, including hemispheric deafferentation, perinsular hemispherectomy, dorsal transcortical subinsular central hemispherectomy, and the transsylvian keyhole functional hemispherectomy (Table 1). All of these procedures are aimed at less resection and more disconnection, increasing the safety of the procedure and reducing postoperative complications. The improved safety of these modifications and the improvements in neuroimaging and neurophysiologic monitoring have made hemispherectomy a more attractive option for patients with intractable unihemispheric epilepsy.

Role of Pathology

Typical pathologies leading to diffuse unihemispheric disease and intractable epilepsy include neonatal injury, vascular insults, hemimegalencephaly, hemispheric cortical dysplasia and other neonatal migration disorders, Sturge-Weber syndrome, and Rasmussen encephalitis. One group of patients has experienced the acute onset of a massive insult resulting in a fixed neurologic deficit. These include those with infantile hemiplegia resulting either from posttraumatic brain injury or vascular insults. A second group of patients has malformations of cortical development (MCD), which vary in severity from focal cortical dysplasias to hemispheric cortical dysplasia and hemimegalencephaly. In these patients, neurologic manifestations are often delayed or progressive. Another group of patients has Sturge-Weber syndrome (encephalotrigeminal angiomatosis). Characterized by a facial port wine stain (nevus flammeus) and pig angiomatosis, patients develop intractable epilepsy associated with impaired cognitive development and hemiparesis. A final group has acquired progressive neurologic deterioration with intractable epilepsy. The prototype for this is Rasmussen encephalitis. In 1938, Rasmussen et al. first described this chronic childhood encephalitis that leads to intractable epilepsy and neurocognitive deficits. In this disease, the presentation may be delayed but the disease course, once established, is inexorably progressive.

Role of Functional Localization

Because no portion of the hemisphere is to be spared functionally, preoperative or functional localization has no specific
role. However, as discussed below, the presence or absence of residual language, motor, and visual function in the affected hemisphere will affect risks of surgery.

Impact of Electrophysiologic Studies

The primary goal of electrophysiologic studies is to lateralize the disease to the radiographically and clinically abnormal hemisphere. Typical detailed investigation with both ictal and interictal recordings is performed. Most often, interictal surface electroencephalogram (EEG) shows a clearly asymmetric tracing with abnormal slowing, low background voltage, and multifocal independent sharp waves and spikes over the affected hemisphere. In specific orders, there may be characteristic EEG abnormalities; for example, in hemimegalencephaly, the EEG may demonstrate hemihyparrhythmia.

The other important goal is to determine whether there are abnormalities in the “good” hemisphere. Bilateral independent epileptogenic foci are associated with a poor prognosis for seizure control. In a study of 12 patients, Carmant et al. demonstrated that a good outcome from hemispherectomy was associated with interictal EEG suppression over the abnormal hemisphere, absence of contralateral slowing, absence of generalized discharges, and absence of bilateral independent spiking. In a study of 28 patients, Doring et al. found that 75% had bilateral EEG abnormalities preoperatively, which was more common in patients with MCD than with acquired lesions. Notably, 77% of patients with acquired lesions became seizure free, compared with only 47% of patients with MCD. Thus, the presence of bilateral EEG abnormalities alone does not preclude consideration for hemispherectomy.

In general, the EEG will lateralize well with structural imaging abnormalities. For hemispherectomy, accurate lateralization and not intrahemispheric localization is most important. In cases without radiographic lateralization, of course, both surface EEG and, when necessary, invasive EEG monitoring play a critical role in both lateralization and localization.

Impact of Neuroimaging

Structural neuroimaging plays an integral role in the preoperative evaluation of the hemispherectomy candidate. The advent of computed tomography (CT) imaging in the 1970s permitted direct visualization of the “good” and “abnormal” hemispheres. While CT can still provide important information (e.g., calcification in Sturge-Weber syndrome), magnetic resonance imaging (MRI) has become the study of choice, as it provides excellent visualization of structural abnormalities in the cerebral cortex and subcortical white matter. It also provides a high-resolution assessment of the “good” hemisphere.

Structural imaging may reveal the severity of disease in vascular or posttraumatic hemispheric injury, from atrophy to pachygyric cortices. Atrophy, characterized by loss of gray and white matter and the presence of an enlarged ventricle, is encountered in most conditions and may be of varying degree, depending on the severity or chronicity of the underlying disease process. In Sturge-Weber syndrome, CT and plain films may show “tram-track” hemispheric calcification, and Gd-DTPA-enhanced MRI demonstrates pial angiomatosis. In hemimegalencephaly, structural imaging shows marked enlargement of the affected hemisphere with abnormal thickening of the cortical mantle. High-resolution structural MRI showing both large and subtle cortical abnormalities has also revolutionized the diagnosis of MCD.

Early MRI may be useful in the diagnosis of Rasmussen encephalitis, but definitive diagnosis requires the appropriate clinical picture together with biopsy demonstrating perivascular lymphocytic cuffing and gliosis.

Functional imaging may also be incorporated into the evaluation of the hemispherectomy candidate. For example, positron emission tomography (18F-FDG-PET) can be useful to follow disease progression, either with hemimegalencephaly, or Sturge-Weber syndrome, or Rasmussen encephalitis. Diffuse unilateral interictal hypometabolism of the affected hemisphere is most commonly found. Single photon emission computed tomography (SPECT) scanning, which provides information regarding cerebral blood flow, may show hyperfusion in the affected hemisphere. In a study of seven patients undergoing hemispherectomy, the six patients with unilateral preoperative SPECT findings all had a favorable outcome regardless of the surface EEG. Functional studies are most important in cases without lateralizing findings on structural imaging studies.

Frequency of Use

Hemispherectomy is primarily a pediatric operation, as perinatal, congenital, and early developmental pathologies account for the majority of conditions leading to intractable unihemispheric epilepsy. In 1993, of 47 surveyed epilepsy surgery centers, 29 were performing hemispherectomies; hemidecortication was advocated in three, the anatomic technique in four, and functional hemispherectomy in 22. In an extensive multicenter study of 333 hemispherectomies from 11 centers published by Holthausen et al. in 1994, distribution of etiologies was as follows: 31% hemispheric dysplasia, 25% Rasmussen encephalitis, 14% vascular, 13% hemimegalencephaly, 8% Sturge-Weber, and 8% other. Surgical techniques included functional hemispherectomy (33%), “Adams modification” (18%), hemidecortication (18%), hemispherectomy (17%), and anatomic hemispherectomy (13%). With the introduction of modern functional hemispherectomy techniques, controversy regarding risk and postoperative complications has decreased.

As with all surgical procedures, the benefit of the operation earlier in life is expected that the number of centers and hemispherectomy cases will increase.
Indications
The indications for hemispherectomy include damage to one hemisphere accompanied by medically intractable epilepsy and accompanying neurologic deficits. As described above, etiologies include extensive hemispheric dysplasia, hemimegalencephaly, Sturge-Weber disease, Rasmussen encephalitis, and perinatal infarction. One group of patients presents with a maximal hemispheric deficit, typically hemiparesis and hemianopsia, usually due to a fixed perinatal insult. The other group presents with intractable epilepsy associated with progressive neurologic deterioration, most often due to a pathologic condition that will evolve ultimately into a maximal hemispheric deficit. Hemispherectomy is contraindicated if presurgical evaluation fails to demonstrate that ictal activity arises from the affected hemisphere. Usually, the history of medically intractable epilepsy often with lateralizing semiology is combined with evidence of extensive hemispheric involvement on high-quality MRI and other lateralization and localization with inpatient video-EEG seizure monitoring. Furthermore, hemispherectomy is not employed if less extensive surgical therapy (cortisectomy or focal or multilobar resection) may potentially be effective.

Language Evaluation
Preoperative language function depends on whether the dominant hemisphere is affected and the severity of disease progression. The younger the age of injury, the more likely language transfer to the healthy hemisphere will occur. Thus, the potential for postoperative language deficits depends on the timing of transfer of language function and in which hemisphere transfer of language function is a relative contraindication to hemispherectomy. Older children with dominant hemisphere disease or some language function should undergo assessment of laterализation of language (e.g., with the intracarotid amobarbital [Wada] test). The “worst-case scenario” is a late appearance of progressive disease (e.g., Rasmussen encephalitis) in the dominant hemisphere, in which patients may have severe language deficits following dominant hemispherectomy.

Electroencephalography and Imaging Findings
As described above, there is usually a concordance between EEG abnormalities and a structurally abnormal hemisphere as seen on preoperative imaging studies. In a significant number of cases, however, EEG abnormalities may be recorded from the good hemisphere, either secondary to or independent of the abnormalities of the diseased hemisphere. Bilateral abnormal EEG findings do not represent an absolute contraindication to hemispherectomy, as they may reflect secondary epileptogenesis and disappear after surgery. An explanation for the presence of independent epileptic activity recorded from the good hemisphere should be taken into account during the decision-making process for hemispherectomy.

Etiology, Natural History, and Timing of Surgery
Three aspects of the etiology and natural history of the condition may affect timing of surgery: Whether the condition is congenital or acquired, whether it is strictly unilateral or possibly bilateral, and whether it is static or progressive. Congenital pathologies such as large porencephaly resulting from intrapartum or perinatal ischemia or trauma, or lesions which are usually strictly unilateral, have a better prognosis with surgery than a congenital lesion such as hemimegalencephaly or hemispheric dysplasia, which may be associated with some degree of contralateral involvement. Acquired unilateral pathology, such as Rasmussen encephalitis, has a better prognosis than infectious processes, which usually have bilateral involvement.

Timing of surgery is determined by the severity of epilepsy, the age of the patient, the natural history of the disease, and the adequacy of therapeutic trials of anticonvulsant medications. Hemispherectomy before the second or third year of life carries no risk of increased deficit and is therefore ideal in those cases that present early for diagnosis and evaluation. In later-onset cases (e.g., Rasmussen encephalitis in older children), timing is controversial, while complete transfer of language and motor function to the healthy hemisphere is less likely to occur in older children, nevertheless, the intractable seizures can produce psychosocial deterioration and may prompt earlier surgery. Evidence is accumulating that seizures themselves as opposed to the pathologic substrate may significantly decrease cognitive and psychosocial development. In certain conditions (e.g., inborn brain malformations), late-stage nonprogressive status epilepticus, very early surgery may be optimal; in our experience, hemispherectomy can be performed safely at 4 months of age.

Goals of Surgery
The primary goal of hemispherectomy is to achieve seizure control via complete disconnection of the epileptogenic abnormal
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Hemispherectomy – Change in Paradigm

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<th>Type of Removal</th>
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<td>large tissue removal</td>
<td>anatomic hemispherectomy</td>
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<td>partial removal</td>
<td>functional hemispherectomy (Rasmussen)</td>
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<td>small removal</td>
<td>perisylvian hemispherectomy (Sbumain, Vitalaro, Shinnick + Neurons)</td>
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<td>minimal removal</td>
<td>transsylvian hemispherectomy (Blaine + others)</td>
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FIGURE 1. Hemispherectomy techniques. Over time, there has been a change in paradigm to more minimal resections combined with more extensive disconnection.

hemisphere from the “good” hemisphere. In anatomic hemispherectomies, this was accomplished by extirpation of the affected hemisphere. In functional hemispherectomies, the emphasis is on complete disconnection rather than resection, although some tissue is resected in each of the current variations of functional hemispherectomy.

The second major goal is to improve psychosocial and cognitive development. Adequate seizure control can be expected to lead to better psychosocial and cognitive development and improved quality of life.30,49,58

Surgical Approaches (Table 1, Fig. 1)

The classic anatomic hemispherectomy involved a large E-shaped skin incision along the midline down to the temporal base; hemicraniotomy; occlusion of the anterior and middle cerebral artery and parasagittal veins; interhemispheric calloso-tomy and frontobasal disconnection; disconnection of insular cortex and temporal stem; and removal of the hemisphere en bloc or in lobes. Total anatomic hemispherectomy was associated with severe intraoperative hemorrhage, hypotension, early and late hydrocephalus, and SCH.25,33,42,44,74,83,88 Development of hydrocephalus necessitated shunting in ~50% of patients. SCH typically developed after 8 to 15 years and led to progressive neurologic deterioration, increased intracranial pressure, and death in many cases.33,42,74,83

The Oxford-Adams modification was an attempt to avoid the complications of SCH and hydrocephalus by reducing the subdural space and insulating the subdural cavity from the ventricular system. In this technique, the classic anatomic hemispherectomy is supplanted by occlusion of the foramen of Monro with a muscle plug and occlusion of the subdural space by folding down the convexity dura onto the tentorium, falx, and floor of the middle fossa, thus creating a large extradural space.1,7 The shunted hemispherectomy used by Peacock at UCLA supplemented the classic anatomic hemispherectomy with routine subdural drainage for 3 days postoperatively to evacuate the debris resulting from the operation followed by placement of a shunt in all cases.31,42,74,85

Hemidecortication or hemicorticectomy was developed based on the principle that only the ictogenic cortex needs to be removed. This procedure, developed by Ignelzi and Bucy in 1968,48 involves a large craniotomy; removal of all cortical gray matter while the white matter is left intact, without callosotomy; and no opening into the lateral ventricle except at the temporal tip required to remove the hippocampi. Several modifications of hemidecortication have been proposed.44,52,115

In response to the problems associated with classic anatomic hemispherectomy, Rasmussen introduced the functional hemispherectomy, which involved a slightly smaller craniotomy, temporal lobectomy, large central hemispheric resection, callosotomy, and fronto- and parieto-occipital disconnection82 (Fig. 2). This procedure improved seizure control while maintaining a low incidence of SCH.

Over the past 15 to 15 years, there have been further development and modification of functional hemispherectomy techniques, including hemispheric deafferentation, 1–7 peri-insular hemispherotomy, 8–10 transsylvian subinsular hemispherotomy, 11–13 the Japanese peri-insular modification, 14,15 and the transsylvian keyhole functional hemispherectomy. 16–20 (Fig. 3). While in the literature these are sometimes called “hemispherectomies,” there is a degree of brain resection in each case as “functional hemispherectomies.” All of these procedures have the common anatomic goal of complete hemispheric disconnection and involve smaller brain resections with disconnections and callosotomy that are essential variations of the Rasmussen functional hemispherectomy technique (Villemure et al. 26–44, 105). One specific question that arises in these functional hemispherectomy modifications is whether insular cortex should be removed. The concern is that residual insular cortex may be a generator of persistent postoperative seizures. Villemure et al. report using intraoperative electrocorticography from the insular cortex following hemispherectomy and proceeded to remove the structure if abnormal spiking is recorded. 45–50 This question has also been addressed by other investigators. 51–53 However, in a large multicenter study and in a recently reported series, the presence of residual insular cortex was not statistically associated with poor seizure control. 54–56 Perhaps the most prudent approach is to always opt for removal of insular cortex during the initial surgery to exclude this possible cause of persistent seizures. 57–60

Results and Complications

Potential operative and postoperative complications include hemorrhage, infection, hydrocephalus, SCH, and cerebral hemorrhages with enlarged ventricles and is especially suitable for cases of perinatal infarction and cystic encephalomalacia. A key important advantage over the “classic” Rasmussen functional hemispherectomy is in the smaller exposure, shorter operative time, and lower blood loss. 61–65 However, it is not ideal for hemimegalencephaly, in which there is an enlarged and dysplastic hemisphere. Instead, for hemimegalencephaly, a hemispherotomy technique is preferred to allow space for postoperative swelling. 66–70


Fig. 3

FIGURE 4. Sturge-Weber disease in a 16-month-old male. Axial (left) and coronal (right) magnetic resonance images with contrast (upper) show the pathologic vascularization and the enlarged insular cortex and atrophy of angle gyri and the whole hemisphere. Whitelines indicate the short distances between circular sulci and ventricles. This case was done with the transsylvian keyhole functional hemispherectomy technique. (Reproduced from Schramm J. Hemispherectomy techniques. Neurosurg Clin N Am 2002;13(1):113–134, with permission.)

Fig. 4
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FIGURE 5. Right-sided hemimegalencephaly in a 7-month-old male. Preoperative T2-weighted magnetic resonance images (top) demonstrate an enlarged right hemisphere with impaired sulcation, especially posteriorly. Corresponding axial, coronal, and sagittal postoperative images (bottom) demonstrate functional hemispheric disconnection. This case was done with a perisylvian window technique.

edema. In early series, these problems led to unacceptably high perioperative mortality (6% to 8%). Modern series are associated with lower mortality: In the recent multicenter series reported by Holthausen et al. mortality was 1.5%. The original Montreal series of anatomic hemispherectomy had a 52% hydrocephalus rate and a 30% SCH rate. Fortunately, there have been few reports of SCH since the 1970s, and the hydrocephalus rate is much lower in modern procedures. The Oxford-Adams modification was associated with a reduced rate of hydrocephalus (<10%). However, modern hemispherectomy approaches are still associated with an appreciable rate of hydrocephalus requiring shunting: 10 of 53 patients with the Delalande technique, 5 of 32 cases with Shimizu and Machara’s technique, and 2 of 49 cases with Schramm’s transsylvian keyhole technique.

All of the modern hemispherectomy techniques have as their primary goal the elimination of seizures via total hemispheric disconnection. Approximately 70% of patients will be seizure free following surgery, with anticonvulsant requirement either eliminated or simplified. Comparisons of seizure outcome between series are limited by lack of standardized outcome measures in some studies. Most useful is the extensive review of 333 patients from 11 centers reported by Holthausen et al. In that study, overall 70.4% of patients became seizure free (Engel class I). Interestingly, there appeared to be significant differences between operative techniques, with better results from modern functional hemispherectomy techniques (85.7% class I) compared with the Rasmussen technique (66.1% class I) or the hemidecortication techniques (60.7% class I). In Binder and Schramm’s series of 49 patients, with a median follow-up of 52 months (range 12 to 146 months), 44 patients were seizure free (Engel class I, 90%), three patients experienced seizure reduction of more than 75% (Engel class III, 6%), and two patients had no seizure relief (Engel class IV, 4%). In the Holthausen et al. series, there was also a difference in outcome by pathology, with the best results in Sturge-Weber syndrome (Engel class I, 82.1%) and the worst with dysplastic lesions (Engel class I, 56.6%). No significant relationship was found between seizure outcome and preoperative EEG, seizure characteristics, or age at surgery.

Clearly, longer follow-up is necessary in all modern series, both with respect to late development of complications and whether the seizure
control is persistent. For example, in a large recently re-
ported series from UCLA, 78.6% of patients were seizure
free at 6 months, 76.3% at 1 year, 70.4% at 2 years, but
only 58.0% at 5 years.46 In addition, longitudinal studies of
these patient populations will be critical to determine long-
term postoperative cognitive,5,27,65 language,7,27,28,95,100 and
development.15,21,45,61,89 outcomes.

MULTILOBAR RESECTIONS

Indications and Role of Pathology

Multilobar resections are indicated for the control of phar-
macologically refractory seizures in the presence of epilep-
togenic zones that affect more than one lobe of the brain in
patients with some preserved neurologic function.13,31 As for
hemispherectomy, multilobar resections are considered with
pathologies that lead to unihemispheric disease and intractable
epilepsy, including neonatal injury, vascular insults, tumors,
glioneuronal cortical dysplasia and other neuronal mi-
gation disorders, and Sturge-Weber syndrome.34 The extent
of the pathologic process responsible for the seizures or the
wish to spare some function guides the extent of the resec-
tion and the decision between multilobar resection and hemi-
spherectomy. Seizures related to large porencephalic cysts re-
sulting from prenatal vascular occlusion of either the anterior
or posterior main branch of the middle cerebral artery may be
successfully treated by bilobar or trilobar resection. A signifi-
cant percentage of patients with Sturge-Weber syndrome do not
have complete hemispheric involvement, and if the sensorimotor
cortex is not involved, a lobar or multilobar resection can be
performed. Extensive dysplastic lesions may also be associated
with multilobar epileptogenicity. In general, if the degree of cor-
tical dysplasia is less than that seen in hemimegalencephaly
and the patient has no evidence of a hemiplegia, multilobar resec-
tion is an option considered instead of hemispherectomy. In contrast,
Rasmussen encephalitis, epileptogenicity may appear partial
at the onset but the disease is known to be progressive and ul-
timately involve the whole hemisphere; in this disease, early
multilobar resection is unlikely to provide permanent seizure
control.

Role of Functional Localization

In multilobar resections, surgery aims at excising the epilep-
togenic zone as completely as possible while sparing function
and avoiding the creation of a new deficit or the exacerbation
of an existing one. Preoperative or intraoperative functional
localization may be obtained to map out the anatomic region
responsible for speech as well as sensorimotor cortical areas.
Functional mapping before resection is obtained either through
deep electrodes or subdural grids for localization of epilepto-
genic sites and motor, sensory, or speech areas. Intraoperative
mapping by cortical electrical stimulation or the use of sensory-
evoked potentials may help to indicate the exact location of
motor and sensory functional areas, so that damage to them
during resection can be avoided and the extent of removal of
the epileptogenic zone optimized. The widespread extent of
brain pathology in these patients (e.g., large atrophic lesions
or extensive dysplasia) may distort the normal anatomy and
therefore functional mapping may be helpful.

Impact of Electrophysiologic Studies

Electrophysiologic investigations follow the usual approach of
intertial surface EEG recordings, including sleep recordings,
drug tapering, and long-term video-EEG monitoring of ictal
and interictal states. Typical findings with multilobar epilepto-
genic regions are widespread interictal abnormalities with in-
dependent multifocal foci. It may be difficult to determine
the exact lobar zone of seizure onset. As usual, electrophysi-
ologic findings must be correlated with seizure semiology and
imaging findings.

In some patients, there is also a role for invasive intracra-
nial monitoring (9% in our series in Bonn). Placement of depth
electrodes, subdural strips, or grids can help to delineate the
predominant epileptogenic zone11 and also to map functional
areas. However, Rasmussen observed that in these patients even
the full battery of recording techniques frequently failed to
demonstrate a focal seizure onset in the damaged hemisphere.86
In cases in which the imaging studies demonstrate a widespread
lesion corresponding anatomically to diffuse electrophysiologic
abnormalities, there is probably no need for invasive recording,
and it can be assumed that the epileptogenic zone corresponds
with the zone of anatomic abnormality. In some cases (34% of the
Bonn series), after an intermediate degree of resection
and/or disconnection, intraoperative electrocorticography may
help to determine the final extent of resection, sparing certain
areas if no spikes can be recorded there following the nearby
resection.

Impact of Neuroimaging

Imaging studies are important to localize the hemispheric
pathology. However, it is important to keep in mind that the
epileptogenic zone may extend outside the range of the obvious
anatomic lesion. CT is useful, especially in processes of atro-
phy, showing enlarged sulci and ventricles. MRI is superior in
defining the anatomy and adds higher resolution in delineating
the gyral pattern and in demonstrating parenchymal lesions or
signal abnormalities. Recent improvements in MRI led to
much greater recognition and delineation of varieties of cortical
dysplasia.37,63 If the pathology on imaging is focal and the epileptic ac-
tivity widespread, metabolic imaging studies may help in the
localization of the epileptogenic zone. For example, Chugani
et al. demonstrated the value of PET when combined with in-
traoperative electrocorticography in defining the boundaries
for multilobar resections.20 Hypometabolism on interictal 18
F-fluorodeoxyglucose (FDG-PET) was found in the parieto-
ocipitotemporal cortices of patients who eventually under-
went multilobar resections with successful control of their
seizures.

Frequency of Use

Multilobar resections constitute a small proportion of epilepsy
surgery procedures. In the Bonn series of 2,000 epilepsy pro-
cedures, there were 32 multilobar resections (1.6%). How-
ever, the increased diagnosis of hemispheric or subhemispheric
pathologies (such as multilobar cortical dysplasia) with mod-
ern MRI techniques may lead to identification of more patients
who would benefit from multilobar resections, especially in
those with preserved functions that are not good candidates
for hemispherectomy.

Evaluation Criteria

The most important decision with this group of patients is
whether to perform a hemispherectomy or a multilobar re-
section. The presence of only a minimal or moderate hemi-
paresis, the presence of useful vision in the contralateral

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visual field, or the preservation of speech functions in the damaged hemisphere often precludes consideration of hemispherectomy despite the presence of extensive damage and widespread epileptogenesis in the involved hemisphere. In particular, patients with nonprogressive disorders (e.g., atrophy, gliosis, dysplasia) with good contralateral extremity function (e.g., fine finger movements and foot tapping) are potential candidates for multilobar resections instead of hemispherectomy. However, if a hemiplegia is present with loss of digital dexterity, the EEG shows diffuse abnormalities throughout the affected hemisphere, and the MRI reveals widespread hemispheric pathology, hemispherectomy is indicated. If the history is suggestive of Rasmussen encephalitis, and surgery has been declined by the family, it is wise to perform a hemispherectomy as the primary procedure, because any lesser procedure is unlikely to control seizures and the inevitable subsequent surgery is rendered more difficult. In young children with infantile spasms, the clinical appearance of the seizures and the hypsarrhythmic pattern on EEG would suggest a generalized seizure disorder, rendering the patient unsuitable for surgery.

During the preoperative evaluation for multilobar resections, attempts are made to tailor the surgical excision for maximal benefit. To eradicate the epileptogenic tissue, the epileptogenic zone must be defined. Clues are obtained from analysis of the patient's history, which helps determine the possible etiologic factors. These may be congenital (dyplastic) or acquired (infection, head injury), static (vascular), or progressive (Rasmussen encephalitis). The clinical seizure pattern often helps to indicate lateralization and localization of onset. Noninvasive and invasive electrophysiologic studies further pinpoint the epileptogenic zone. These observations should be closely correlated with findings from anatomic (CT and MRI) and functional (PET, ictal SPECT) imaging investigations. In cases in which the imaging abnormalities are minor, invasive monitoring may be indicated or a multistage surgical approach may be considered. In staged multilobar resections, the obvious focal, lobar, or multilobar epileptogenic zone may first be excised, with the plan to proceed to further excision at a later date if seizures are not satisfactorily controlled. Recently, Olivier as well has taken the approach of performing a hemispherectomy as the primary procedure, because any lesser procedure is unlikely to control seizures and the inevitable subsequent surgery is rendered more difficult. In young children with infantile spasms, the clinical appearance of the seizures and the hypsarrhythmic pattern on EEG would suggest a generalized seizure disorder, rendering the patient unsuitable for surgery.

Goals of Surgery

Multilobar resection aims at achieving complete cessation of seizures or significant improvement in seizure frequency without producing neurologic deficits. Most widespread lesions responsible for seizure disorders are static and thus do not require surgery to alter their natural course. Atrophic, porencephalic, and dysplastic lesions are not by definition progressive disorders. However, these static lesions may indirectly interfere with brain function through mechanisms of secondary epileptogenesis. On the basis of these mechanisms, another goal of multilobar resections may be to prevent further brain damage. It is thus important to identify candidates, localize the lesion, and then proceed to surgery early in the evolution of the seizures. Good seizure control in these patients is accompanied by improved psychological performance and social integration, which are often listed as secondary benefits.

The ultimate goal of the surgery should be to improve psychosocial functioning and thereby promote a better quality of life.

Surgical Approaches

Multilobar resections may involve the frontostriatal, frontotemporal, parieto-occipitotemporal, or parieto-occipital lobes (Figs. 6 and 7). The techniques utilized vary from extensive corticectomy, lobar excision, or lobe disconnection to a combination of these. The corticectomy technique involves excision by aspiration of the cortical gray matter felt to correspond to the epileptogenic zone. Lobar excision consists of the anatomic removal of gray and white matter corresponding to the epileptogenic zone, the brain structural abnormalities responsible for the seizures, or both. Disconnection of a lobe
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5. References


SUMMARY AND CONCLUSIONS

In multilobar resections and hemispherectomy, intractable seizures are secondary to widespread, usually unilateral, hemispheric damage resulting from multiple possible etiologies—some static, others progressive. Surgery has to be extensive and should aim at eliminating the influence of the abnormal tissue responsible for epileptogenesis. Seizure characteristics, neurologic status, EEG and imaging findings, and judgment regarding medical intractability all guide the decision regarding timing of surgery and the most appropriate surgical procedure. In the presence of some degree of preserved neurologic function, multilobar resection should aim at eliminating the epileptogenic zone without creating new deficits. In similar pathologies in which the underlying condition has created a complete hemispheric syndrome, the need to preserve function does not limit the surgery and hemispherectomy can be considered. Hemispherectomy under these circumstances is most likely to bring about complete seizure control. With the use of modern functional hemispherectomy techniques, operative morbidity and mortality can be minimized, and seizure control is usually good. The improved safety of these modifications together with improvements in neuroimaging and neurophysiologic monitoring has made hemispherectomy a more attractive option for patients with intractable unihemispheric epilepsy.

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Results and Complications

There have been relatively few studies of outcome in children undergoing extratemporal and multilobar epilepsy surgery. One problem is that this is a very heterogeneous group of patients, both in clinical and pathologic characteristics. In general, seizure-free outcome in patients with multilobar resections is poorer than those with unilobar resections or hemispherectomy, which is not surprising since most unilobar resections are temporal lobe resections, and hemispherectomy provides a complete disconnection of the affected hemisphere. Outcome for hemispheric cortical dysplasia is clearly related to the extent of resection of the dysplastic tissue. For the subset of patients with malformations of cortical development in particular, approximately 50% of patients become seizure free postoperatively. One recent study indicates that the presence of acute postoperative seizures is an independent predictor of poor postoperative seizure outcome at 2 years. Complications are not usually separately reported for multilobar resections. Clearly, neurologic outcome depends on preoperative neurologic status as well as location and extent of multilobar resection. Patients undergoing multilobar resections as a repeat resection for initially failed epilepsy surgery have increased potential for neurologic deficits after the second operation. Long-term follow-up of cognitive outcomes, as for hemispherectomy, will be an important component of future research.

or its isolation from the rest of the brain has been utilized by some authors, thereby avoiding removal of the lobe (frontal or parieto-occipital) but eliminating, by disconnection, the epileptic manifestation. Removal of the epileptogenic tissue by en bloc resection appears to be the preferred technique in multilobar resections. Rasmussen stressed two important technical aspects of the resection strategy: (a) subpial removal along sulci to leave the removal cavity lined by untraumatized pial-covered gyrus and (b) avoidance of movement of the underlying white matter during the cortical excision to minimize risk of injury.

Unlike the case of hemispherectomy, intraoperative attention to preservation of function is essential. When a less than maximal hemispheric resection is performed, the arm and leg area of the sensorimotor cortex should be preserved. If a useful visual field is present, the posterior parietotemporal and occipital regions should also be preserved. During surgery, somatosensory-evoked potentials and motor-evoked potentials can be used to delineate the sensorimotor cortex, which can then be left intact. Preoperative investigations localizing the epileptogenic area should be available in the operating room, so that the intraoperative findings can be correlated with those obtained preoperatively. Some surgeons have also used intraoperative electrocorticography to further define the margins of the multilobar resection.

FIGURE 7. Multilobar resection for Sturge-Weber disease in a 10-year-old female. Axial (top left), coronal (top right), and sagittal (bottom left) magnetic resonance images demonstrate a left parieto-occipital lesion consistent with vascular malformation. Invasive diagnostic monitoring was performed with grid and strip electrodes (bottom right). Subsequently, a left parieto-occipital extended leucotomy with multiple subpial transections was performed. Pathology demonstrated Sturge-Weber disease.


23. Curtiss S, de Bode S, Mathern GW. Spoken language outcomes after hemi- 22. Curtiss S, de Bode S. Age and etiology as predictors of language outcome


