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Editorial

Hemispherectomy for epilepsy

When is one half better than two?

Michael Duchowny, MD

Cerebral hemispherectomy, the surgical removal of one cerebral hemisphere leaving the basal ganglia undisturbed, was introduced by Walter Dandy in 1928 as a radical treatment for diffuse malignant gliomas of the nondominant hemisphere.[1] Although largely unsuccessful as a therapy for this malignancy, the degree of postoperative functional recovery was notable; 10 years later, this inspired the Canadian neurosurgeon Kenneth McKenzie to successfully employ hemispherectomy in a hemiplegic adult with medically resistant partial epilepsy.[2] In 1950, Roland Krynauw, a South African neurosurgeon, reported beneficial effects of hemispherectomy on seizures, motor functioning, and development in a small cohort of children with congenital hemiparesis and epilepsy.[3]

After gaining widespread endorsement as a useful technique for seizures in patients with partial epilepsy and cerebral hemispheric dysfunction, the procedure was nearly abandoned when late-developing complications were recognized. Following an initially benign course of seven to ten years, some hemispherectomized patients gradually deteriorated and died from complications of obstructive hydrocephalus. The cause was traced to recurrent subdural bleeding from the vascular pedicle of the operative cavity, leading to granular ependymitis and superficial hemosiderosis. A smaller number of patients developed late intracranial hemorrhage attributed to chronic subdural hematoma formation within the cavity.

In an effort to reduce morbidity and mortality associated with the classic complete or "anatomic" hemispherectomy, Theodore Rasmussen proposed a modified "functional" procedure in which only the central portion of the cerebral hemisphere was removed to allow exposure and disconnection of the anterior frontal and posterior parieto-occipital poles and the corpus callosum. This technique rendered the remaining cerebral tissue intact but nonfunctional, while preserving blood supply and mechanically stabilizing the intact hemisphere. The complication rate of the functional procedure was drastically reduced while rates of seizure freedom remained comparable to the anatomic procedure.[4] Postoperative improvement in language, motor skills and development has been documented repeatedly.[5,6]

Recent innovations in hemispherectomy technique have shortened operative time and reduced blood loss. These innovations all involve initial tissue removal and disconnection of remaining structures, and include cerebral hemicorticectomy, hemidecortication, trans-sylvian and transventricular functional hemispherectomy, peri-insular and central vertical hemispherotomy. Continued efforts to improve morbidity attest to the clinical importance of hemispheric surgery as a treatment for intractable seizures.

As is often the case with successful clinical and scientific endeavors, the establishment of a new therapeutic procedure typically raises more questions than it answers—and hemispherectomy is no exception. The ability to study the consequences of removing one cerebral hemisphere has provided a wealth of data for investigators interested in lateralized cognitive functioning and plasticity. Which patients stand to benefit and when is the optimal time to perform the procedure? Can we predict seizure outcome and do we know how such a radical procedure, often performed early in life, affects future brain functioning? What is the role of neural plasticity in postoperative recovery?

Obtaining answers to these and other questions is a slow process. Hemispherectomy is currently performed at selected tertiary epilepsy centers that treat a high volume of children. The overall number of procedures performed annually is small, and many hemispherectomy candidates are either too young or too disabled to complete standardized neurocognitive testing protocols. Follow-up may also be problematic as some patients end up requiring institutionalized care, even if they become seizure-free.

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From the Comprehensive Epilepsy Program, Miami Children's Hospital; and Department of Clinical Neurology, University of Miami School of Medicine, Miami, FL

Address correspondence and reprint requests to Dr. Michael Duchowny, Comprehensive Epilepsy Program, Miami Children's Hospital, 3200 S.W. 60th Court, Miami, FL 33155; e-mail: michael.duchowny@mch.com

Recent large childhood hemispherectomy series suggest that hemispherectomy is beneficial in highly selected cases, and is the only effective treatment for disorders such as Rasmussen's encephalitis.[7,8] The presence of pre-existing neurologic deficits such as hemiplegia or homonymous visual field deficit greatly simplifies candidate selection, but the absence of lateralized neurologic dysfunction may not be a strict surgical contraindication if the potential benefits of hemispherectomy outweigh the risks. Approximately 50 to 80% of candidates are seizure-free or significantly improved, an outcome that is especially welcome for patients who are typically resistant to antiepileptic drugs and who are deteriorating clinically. While a radical procedure that sacrifices an entire cerebral hemisphere might intuitively be thought to result in seizure freedom in all or most patients, this has not been the case. Unappreciated pathology in the non-operated hemisphere and failure to completely remove or disconnect the operative cerebral hemisphere are two important reasons.

In this issue of *Neurology*, Jonas et al.[9] analyzed the effect of critical variables on postoperative outcome in the largest pediatric hemispherectomy series reported to date. This unique single-procedure surgical population is particularly notable because it permits meaningful analyses of post-surgery seizure, motor, developmental and language status as a function of preoperative risk factors and pathological substrate. Fifty-eight percent of the patients were seizure-free at five years, a finding consistent with other hemispherectomy series. As a group, hemispherectomized children in this study and in others[10] achieved very modest developmental gains. This finding is not entirely unexpected because the extent of pre-existent brain damage is often severe and may involve the non-operated hemisphere. Although not statistically significant, patients with hemimegalencephaly experienced the lowest incidence of seizure freedom. From the surgical standpoint, hemimegalencephaly patients were also the most challenging because they required greater operative time and experienced higher blood loss. Hemimegalencephaly patients also had the poorest prognosis for postoperative language and motor status compared to other pathological groups.

More important is the finding that irrespective of underlying pathological substrate, the greatest gains in postoperative development occurred in patients with higher preoperative developmental attainment, shorter preoperative seizure duration, and better postoperative seizure control. Thus, an epilepsy patient's pathological substrate alone does not determine ultimate neurodevelopmental outcome. Eliminating seizures is also critical. This finding provides direct support for promptly referring children with severe and medically resistant seizures for surgical evaluation; delay in referral could further worsen long-term neurodevelopmental status. Hemispherectomy, once regarded as a radical intervention of last resort, may become a standard of early intervention.

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