

## HEMISPHERECTOMY OUTCOMES FAVORABLE IN TWO REPORTS

### Clinical Outcomes of Hemispherectomy for Epilepsy in Childhood and Adolescence

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Hemispherectomy has been performed in the treatment of epilepsy in association with hemiplegia for >50 years. However, the optimal timing of surgery with respect to age at presentation and the influence of underlying pathology on outcome is only slowly emerging. This study reports on the clinical course and outcomes of 33 children who underwent hemispherectomy at Great Ormond Street Hospital, London, between 1991 and 1997. Age at surgery was 0.33–17 years (median, 4.25) with 1–8 years' follow-up (median, 3.4 years). The underlying pathology was developmental in 16 (10 hemimegalencephaly, two polymicrogyria, two focal cortical dysplasia, one diffuse cortical dysplasia, and one microdysgenesis), acquired in 11 (six middle cerebral artery infarct, three postencephalitis/trauma, and one each of hemiconvulsion–hemiplegia epilepsy and perinatal ischaemic insult) and progressive in six children (four Rasmussen encephalitis, two Sturge–Weber syndrome). At follow-up, 52% were seizure free, 9% experienced rare seizures, 30% showed >75% reduction in seizures, and 9% showed < 75% seizure reduction or no improvement. Seizure freedom was highest in those with acquired pathology (82%), followed by those with progressive pathology (50%) and those with developmental pathology (31%). However, seizure freedom, rare seizures, or >75% reduction in seizures occurred in 100% of those with progressive pathology, 91% of those with acquired, and 88% of those with developmental pathology, indicating a worthwhile seizure outcome in all groups. Hemiplegia remained unchanged after surgery in 22 of 33 children, improved in five, and was worse in six. No significant cognitive deterioration or loss of language occurred, and four children showed significant cognitive improve-

ment. Behavioral improvement was reported in 92% of those who had behavior problems before surgery.

### Outcomes of 32 Hemispherectomies for Sturge–Weber Syndrome Worldwide

Kossoff EH, Buck C, Freeman JM

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**BACKGROUND:** Epilepsy affects 80% of patients with Sturge–Weber syndrome; the majority of seizures begin before age 1 year. When seizures are intractable to medications and unihemispheric, hemispherectomy is often advised.

**PURPOSE:** To examine the natural history of patients who underwent hemispherectomy and identify the outcomes in terms of seizure reduction, cognition, and motor deficits.

**METHODS:** A questionnaire was mailed to the parents of patients identified by the Sturge–Weber Foundation as having had a hemispherectomy between 1979 and 2001. Thirty-two (46%) of 70 of the parents responded. **RESULTS:** The mean age at onset of seizures was 4 months, and the median age at surgery was 1.2 years. Children had failed to respond to 3.7 anti-convulsants (AEDs) before surgery and averaged 387 seizures/month. Forty-seven percent had complications (e.g., hemorrhage and hypertension) in the perioperative period; however, 81% are currently seizure free, with 53% off AEDs. Hemispherectomy type (anatomic vs. functional vs. hemidecortication) did not influence outcome. Age at onset of seizures did not predict seizure freedom; however, an older age at hemispherectomy was positively correlated. Postoperative hemiparesis was not more severe than that before surgery. Cognitive outcome was not related to the age at operation, side of operation, or seizure freedom.

**CONCLUSIONS:** Children undergoing hemispherectomy presented at a young age and had frequent seizures for ~1 year but are now mostly seizure free. Age at surgery did not have an adverse effect on either seizure or cognitive outcomes.

## COMMENTARY

For children with unihemispheric seizures and associated pathology of a variety of congenital and acquired causes producing neurologic deficit, hemispherectomy is used in an attempt to abolish seizures and to restore cognitive, behavioral, and motor function to the greatest extent possible. These companion reports assess clinical outcomes after hemispherectomy in a total of 65 patients: The report by Devlin et al. offers a retrospective medical records–based review of 33 children and adolescents who underwent hemispherectomy between 1991 and 1997 at a single institution, whereas the series by Kossoff et al. provides a retrospective questionnaire-based analysis focused on children with Sturge–Weber syndrome treated between 1979 and 2001 at many centers throughout the world.

Devlin et al. report on patients whose epilepsy was due to a spectrum of pathology (16 with disorders of cerebral development, 11 with acquired insults, and six with progressive syndromes), with a median follow-up time of 3.4 years (range, 1–8 years). Seizure outcomes were largely favorable, with 52% of patients seizure free, 9% experiencing rare seizures, and 30% demonstrating a >75% reduction in their preoperative seizure rate; just 9% had little or no improvement in seizure control. Patients with an acquired cause for their seizures were most likely to achieve seizure freedom after hemispherectomy, followed by those with a progressive pathology; those with developmental pathology were least likely to achieve complete seizure control with surgery. Severity of hemiparesis was unchanged postoperatively in the majority of patients (22 of 33), although it improved in five patients and worsened in six. Behavioral and cognitive outcomes were largely favorable. Life-threatening operative complications related to hemorrhage occurred in three children, all younger than 18 months at the time of surgery. Delayed complications occurred in five patients (three required ventriculoperitoneal shunting, one developed a bone flap infection, and one had bilateral iliac vein occlusion related to central venous access requirements for the surgery).

The report by Kossoff et al. relies on responses from a questionnaire mailed to families contained in a voluntary registry of Sturge–Weber patients as the basis for their analysis.

The anonymous questionnaire, completed by 32 respondents (about half of those to whom it was sent), extracted information related to preoperative status, type of surgical procedure and related complications, and the patient's neurologic status at the time of written response to the survey. In this population, mean age at seizure onset was 4 months (range, 2 days to 11 months), and mean age at which hemispherectomy was performed was 2.7 years (range, 3 months to 17.5 years, with all but three patients undergoing surgery before age 9 years). Most patients (17) appeared to have had complex partial seizures, but many had mixed seizure types. Based on family description, surgical procedures included 16 anatomic hemispherectomies, 14 functional hemispherectomies, and two hemidecortications performed at 18 different centers throughout the world. Clinical outcomes were favorable in the majority of patients, with 81% described as seizure free and 53% of patients completely off antiepileptic medication; older age at time of surgery statistically correlated with a more favorable outcome, but other factors (e.g., age at epilepsy onset, type of procedure) did not. A large number of patients experienced perioperative complications, including hemorrhage in four, infection in four, severe headache in two, need for immediate reoperation due to ongoing seizures in one, shunting in one, and hypertension in one; complications did not correlate with any particular type of procedure. Postoperative motor deficits were reported as not significantly changed compared with preoperative status. Based on descriptive responses by the patients' families cognitive status was judged by the authors to be similar to that of the general Sturge–Weber population.

Although both of these studies are retrospective in design and one relies purely on information ascertained by family questionnaire, they provide a relatively large series of cases from which to understand better, and evaluate the risks and benefits of hemispherectomy for children with intractable epilepsy. The analysis by Devlin et al. emphasizes that worthwhile outcomes occur after hemispherectomy in patients with intractable unihemispheric epilepsy of diverse pathology, and the report from Kossoff et al. highlights the benefit of hemispherectomy in patients with Sturge–Weber syndrome.

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