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Why Would You Remove Half a Brain? The Outcome of 58 Children After Hemispherectomy—The Johns Hopkins Experience: 1968 to 1996

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ABSTRACT. *Purpose.* To report the outcomes of the 58 hemispherectomies performed at Johns Hopkins between 1968 and January 1996.

Methods. Charts were reviewed of the 58 hemispherectomies performed at Johns Hopkins Medical Institutions by the Pediatric Epilepsy Group during the years 1968 to 1996. Twenty-seven operations were done for Rasmussen's syndrome, 24 operations for cortical dysplasias/hemimegalencephalies, and 7 for Sturge-Weber syndrome or other congenital vascular problems. Seizure control alone did not seem to adequately describe the outcomes of the procedure. Therefore, a score was constructed that included seizure frequency, motor disability, and intellectual handicap. This burden of illness score better described the child's handicap before and after surgery.

Results. Perioperative death occurred in 4 out of 58 children. Of the 54 surviving children, 54% (29/54) are seizure-free, 24% (13/54) have nonhandicapping seizures, and 23% (12/54) have residual seizures that interfere to some extent with function. Reduction in seizures was related to the etiology of the unilateral epilepsy. Eighty-nine percent of children with Rasmussen's, 67% of those with dysplasias, and 67% of the vascular group are seizure-free, or have occasional, nonhandicapping seizures. All operations were considered by the parents and the physicians to have been successful in decreasing the burden of illness. In 44 the procedure was very successful, in 7 it was moderately successful, and in 3 it was minimally successful. Success was related to the etiology, and early surgery was preferable.

Conclusion. Hemispherectomy can be a valuable procedure for relieving the burden of seizures, the burden of medication, and the general dysfunction in children with severe or progressive unilateral cortical disease. Early hemispherectomy, although increasing the hemiparesis in children with Rasmussen's syndrome, relieves the burden of constant seizures and allows the child to return to a more normal life. In children with dysplasias, early surgery can allow the resumption of more normal development. *Pediatrics* 1997;100:163-171; *hemispherectomy, burden of illness, epilepsy surgery, quality of life.*

ABBREVIATIONS. CVA, cerebrovascular accident; MDQ, motor developmental quotient; IQ, intelligence quotient; DQ, development quotient; EEG, electroencephalogram.

The term hemispherectomy is used throughout the literature for several different neurosurgical procedures designed to eliminate seizures arising diffusely from a single cerebral hemisphere. In experienced hands, there would seem to be little difference in outcome between anatomical hemispherectomies,¹⁻³ functional hemispherectomies,⁴ hemidecortectomies,^{5,6} hemispherotomies,⁷ and other variations. The term hemispherectomy, as used in our series includes both anatomical hemispherectomies and hemidecortectomies. The operation was performed principally by two neurosurgeons (B.S.C. and S.U.). Hemispherectomy, the ultimate focal resection,⁸ is performed when smaller focal resections will not remove all of the epileptic region, or when, as in Rasmussen's syndrome, the progressive involvement of the remaining ipsilateral hemispheric cortex is inevitable.⁹ This article is a report on the outcomes of the hemispherectomies performed by the pediatric epilepsy team of the Johns Hopkins Medical Institutions. The outcome of the procedure is clearly dependent on etiology, not on the type of hemispherectomy surgery performed. Therefore, the

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outcomes in these populations will be discussed based on the underlying pathology—developmental dysplasias, vascular problems, and Rasmussen’s syndrome.

MATERIALS AND METHODS

Fifty-eight hemispherectomies have been performed in children by our group during the years 1968 to 1996. Four anatomical hemispherectomies were performed before 1981. The remaining 54 have been hemidecorticectomies.⁶ The distribution of these operations by underlying disease process and by the year they were performed is seen in Fig 1. There were 24 children operated for extensive cortical dysplasia with or without hemimegalencephaly,¹⁰ 27 children operated because of Rasmussen’s syndrome, and 7 operated because of seizures caused by congenital vascular problems: 3 of these had Sturge-Weber syndrome and 4 had congenital cerebrovascular accidents (CVAs). The ages at surgery, sex, side of hemispherectomy, and length of follow-up are shown Table 1. The female predilection in the population with dysplasias, and the right-sided preponderance in the children with Rasmussen’s syndrome are worthy of note.

Selection for Surgery

Patients were selected for surgery when the burden of seizures was excessive and the course of the condition obvious. The timing of surgery was determined by the burden of the child’s condition as defined by the family, in consultation with the physician, and the family must have been willing to trade the burden of a permanent loss of use of the hand for the burden of relief from the seizures and their treatment.

We have been advocates of early surgery in children with Rasmussen’s syndrome, who will inevitably have a progressive worsening of their seizures, progressive intellectual deterioration, and a progressive hemiparesis. Many children were operated early in their course, often when only a mild hemiparesis was present and we are willing to operate even when surgery will create a hemiplegia or worsen a hemiparesis.¹¹ We have not required that seizures be deemed intractable before surgery, implying that all available antiepileptic medications have been tried singly and in combination. We have not usually preceded our hemispherectomies with focal resections when such surgery is unlikely to be successful.¹² However, before decisions were made regarding the surgery, the progressive nature of the disease must have become obvious and the absence of other causes must have become clear.

The children with dysplasias were selected for surgery when the continuous seizures were incapacitating and seemed to be unilateral based on clinical observation, EEG and imaging, and when the likelihood of seizure control with further medication trial was low.^{5,8,13} A hemiparesis may or may not have been obvious preoperatively, and the side of the dysgenesis was not an

important consideration. These children were operated as early as three months of age.

Most children with vascular problems had a dense hemiplegia before the operation and their preoperative seizures were less severe than the other groups. They were often operated later in life when ongoing seizures and the effects of medications became too great a burden.

Burden of Illness

Success of epilepsy surgery in children cannot be measured solely by the freedom from seizures.^{14–16} These children with their multiple daily seizures are further impaired by intellectual and motor disabilities, by medications, and by the limitations placed upon them by family and environment. We term the totality of these consequences the burden of illness.¹⁷ A scale was constructed to assess this burden of illness (Table 2). The burden of each component was given a score of 0 to 3, ranging from no burden (0) to a severe burden (3). These components interact so that while a few generalized tonic seizures a week might be a substantial handicap for an otherwise normal child, they might be only a mild additional burden for one who was severely motor handicapped and retarded. The assessments of the burden were made by two of the authors (E.P.G.V. and J.M.F.) and represents a first attempt to capture the totality of the impact of the illness on the children and the changes in this burden which occur as a consequence of the surgery. Success of surgery was determined both by the changes in seizure frequency and by the change in the burden of illness on the child.

Assessment of Motor and Intellectual Function

Motor function was assessed as a mild, moderate, or severe handicap in the context of the child’s intellectual capacity. In all of our children, the hemiparetic hand can only function as a helper hand, with virtually no individual finger function and a major loss of cortical sensation. The arm can be raised at the shoulder, the forearm flexed, and the hand used to help hold a piece of paper or an object. All children with sufficient mental development are walking, most are running, and although paretic, have little functional impairment of the lower extremity. After hemispherectomy, no child is without some motor burden.

In the young child, motor impairment was an approximation of the motor developmental quotient (MDQ). A mild handicap (score = 1) was equated with an MDQ of 50 to 75 or, when motor function could be more accurately assessed was equivalent to a nonhandicapping hemiparesis. For example, in a child with substantial intellectual delay, a nonfunctional hand may be a mild additional handicap. A moderate impairment (score 2), was a handicapping hemiparesis substantially affecting one extremity, usually due to a substantial impairment of the use of the upper extremity, or in the young child equivalent to a MDQ of 25 to 50. Virtually all children in this moderately handicapped category are walking and running with or without braces. Children with severe impairment (score = 3), have a significant handicapping hemiplegia with little use of the upper extremity, are not ambulating independently, and may be wheelchair bound or bedridden. Children had varying degrees of motor handicap before operation, and the motor disability was not always worsened by the surgery.

Intellectual function was assessed as normal (score = 0); mild intellectual impairment, equivalent to an intelligence quotient/developmental quotient (IQ/DQ) of 70 to 85 (score = 1); moderate impairment, an IQ/DQ of 50 to 70 (score = 2); or severe impairment, an IQ/DQ of less than 50 (score = 3).

RESULTS

Population

The three populations are described in Table 1. The mean age at surgery for Rasmussen’s syndrome was 9.7 (3.8 to 20.6) years; for the cortical dysplasias it was 3.0 (0.2 to 17.8) years. The vascular group was small and heterogeneous in age at surgery. As of January 1, 1996, the 55 children who survived the immediate postoperative period have been followed for a mean of 6.2 years (minimum, 0.5 years; maximum, 27.3 years). The mean length of follow-up for

HEMISPHERECTOMY POPULATION JOHNS HOPKINS:1968-1996

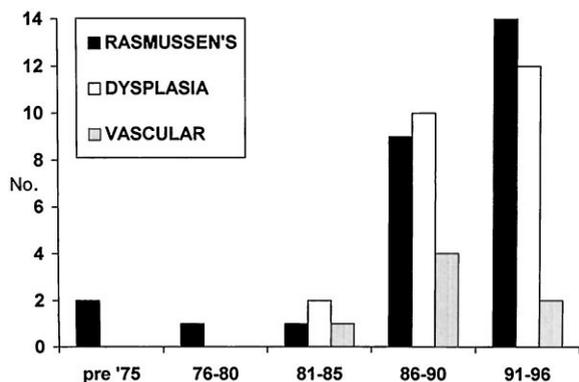


Fig 1. Hemispherectomy population—1996. The number of patients undergoing hemispherectomy during the indicated 5-year time periods is shown according to the three main categories.

TABLE 1. Description of the Patient Population of 58 Children Undergoing Hemispherectomy at Johns Hopkins 1968–1996

Etiology		Mean Age at Surgery (Range)	Sex Male:Female	Side Right:Left	Mean Length Follow-up (Range)
Rasmussen's	(n = 27)	9.7 y (3.8 to 20.6)	10:17	16:11	6.7 y (0.5 to 27.3)
Dysplasia	(n = 24)	3.0 y (0.2 to 17.8)	3:21	13:11	5.5 y (0.5 to 11.9)
Vascular	(n = 7)	8.6 y (1.2 to 19.4)	5:2	3:4	6.2 y (1.1 to 12.4)

TABLE 2. Scoring System for Burden of Illness Scale

Score	Seizures	Motor Dysfunction	Intellectual Dysfunction
0	None	None	IQ/DQ >85
1	Infrequent, insignificant nonhandicapping	Mild, nonhandicapping; motor DQ 50 to 75	IQ/DQ 70 to 85
2	Frequent, moderately severe	Handicapping hemiplegia; motor DQ 25 to 50	IQ/DQ 50 to 75
3	Severe, frequent, handicapping	Severe impairment; motor DQ 25 or less	IQ/DQ <50

DQ = Developmental quotient.

Rasmussen's syndrome is 6.7 years (minimum, 0.5; maximum, 27.3 years); for the dysplasias 5.5 years (minimum, 0.5 years; maximum, 11.9 years); and for the vascular group is 6.2 years (minimum, 1.1 years; maximum, 12.4 years).

Outcomes

Death

Death, related to the hemispherectomy procedure, occurred in 3 of the 58 patients. One child with Sturge-Weber syndrome had massive bleeding from diploic veins when the burr holes were initially made. Inability to stop the bleeding led to completion of the hemispherectomy under hypothermic cardiac arrest and the patient died during surgery. Two other young children (1 and 2 years of age) with dysplasia died within several hours of completion of the procedure. Despite autopsy, the causes of their deaths were not determined, but are believed to be related to autonomic instability and changes in vascular resistance.¹⁸ A fourth child with cortical developmental dysplasia, predominantly affecting one hemisphere, continued to have intractable seizures from the remaining hemisphere after her hemispherectomy and died nine months later of causes related to her cerebral dysgenesis and continued seizures, but unrelated to the surgery.

Perioperative Morbidity

Bleeding

Severe intraoperative bleeding, often difficult-to-control oozing from all cut surfaces requiring 3 to 4 blood volumes of replacement, occurred in 4 out of 24 of the infants with cortical dysplasias. Three were under 1 year of age, and 1 was age 7. Significant bleeding was noted in 3 out of 27 of the children with Rasmussen's syndrome (ages 4.5, 5.3, and 13 years), and in 1 out of 7 children with vascular problems. The oozing (bleeding) began 3 to 4 hours into the procedure, and occurred despite both preoperative elimination of antiepileptic medications that might possibly affect clotting, and careful preoperative evaluation of bleeding and clotting function. Liberal and prophylactic use of fresh frozen plasma and fresh platelets, as well as fresh blood, had no consistent effect. The intra- and perioperative treatment of these patients has been the subject of a prior incom-

plete report¹⁹ and abstract,¹⁸ and will be the subject of a more complete paper.

Fluid Management

During the early part of our series, significant problems with vascular resistance and with fluid and volume management occurred in four infants and young children with cortical dysplasias (ages .75, 1, 2, and 7 years).¹⁹ However, with improved vascular access, and with meticulous monitoring of central venous pressure and vascular resistance by a single pediatric anesthesiologist/intensivist, these problems seem to have disappeared.¹⁸

Postoperative Coma

Three patients have lapsed into coma after operation and two recovered completely after 2 and 6 weeks of coma. It is hypothesized that in these two, coma occurred secondary to torsion of the brainstem during the physical transfer from the operating table to the gurney and to the intensive care unit bed, resulting in magnetic resonance imaging/computed tomography-demonstrable peribrainstem hemorrhages on the unoperated side. With change in the transfer technique and postoperative immobilization of the head (remaining hemisphere down) for several days after the operation, we have had no recurrence. The third child, who had had a prior functional hemispherectomy at another epilepsy center, was reoperated at Hopkins for removal of the residual insular tissue which was producing documented, intractable, insular seizures.²⁰ She remained comatose after surgery and 2 years later continues to have a locked-in syndrome whose cause remains unclear.

Shunts

Shunts have not been placed prophylactically in these patients, but rather only when symptoms of increased pressure and clear evidence of progressive ventricular enlargement become present. A ventricular drain is left in postoperatively for 2 to 4 days, and when necessary, subsequent daily spinal taps are done to remove the high protein, viscous fluid, and blood which seems to transiently clog the pachionian granulations. Five children with Rasmussen's syndrome, 10 children with dysplasias and 1 child with Sturge-Weber syndrome have required

ventriculo-peritoneal shunts within several weeks of surgery. The reason for the disproportionate number of young children needing shunts is unclear. Only two of our patients have ever needed shunt revisions.

Other Perioperative Complications

Clinically significant septic meningitis occurred in only two patients, and a significant bone infection occurred in one. Fever, white cells in the cerebrospinal fluid (CSF), and increased CSF protein occurring 3 to 10 days after surgery were common. Deep venous thrombosis in the leg has occurred in two older children.

Late Complications

One child (see above) died 9 months after operation. Another continues with a locked-in syndrome. One child, with Rasmussen's syndrome, developed signs and symptoms of hydrocephalus due to aqueductal stenosis 7 years after his initial surgery. The symptoms were relieved with shunting.

The Effect of Hemispherectomy on Seizure Frequency

Transient seizures occurred in a few of the 54 children during the first postoperative weeks attributable to fluid or electrolyte imbalance, or to rapid changes in anticonvulsant levels. In the total population of 54 survivors (Table 3), 29 (54%) currently have no seizures. Only infrequent, nonhandicapping seizures occur in 13 out of 54 (24%) of children. These include three children who experienced what they termed ghosts, feelings as if they were having a seizure without a change in the clinical state apparent to an observer, and in one case without change in concurrent electroencephalogram (EEG). The episodic nature of these ghost-like events, and the fact that two of the children seemed to respond to anticonvulsants made us believe that the ghosts were nonhandicapping auras or mild partial seizures. A moderate to significant seizure disorder, with seizures that were more frequent, more severe, and which interfered to some extent with function, occurred in 9 out of 54 children (17%). Thus, 78% of the total population that previously had severe handicapping epilepsy no longer has a significant seizure disorder.

All children who have had hemispherectomies have similar degrees of hemiplegia. The intellectual impairment and residual seizures in these children, and thus the residual burden, varies with the underlying etiology. We have, therefore, chosen to discuss outcome by the individual etiologies.

Outcomes: Rasmussen's Syndrome

Twenty-seven children with Rasmussen's syndrome have had a hemispherectomy (Table 1). Seventeen have been female, 10 are male; 16 had right hemispherectomies and 11 were left.

No perioperative deaths occurred. Perioperative complications included intraoperative, intracranial bleeding in two younger children and one older child, and transient postoperative coma lasting 2 to 6 weeks in two. One additional child developed a locked-in state as discussed above. Five children required ventriculo-peritoneal shunting in the perioperative period and one child has required shunting 7 years after the original surgery. Perioperative infection occurred in one child, and two others had a deep vein thrombosis in the leg.

Seizure Outcome

Eighteen of the 27 children (67%) with Rasmussen's syndrome who had previously been severely handicapped by multiple, daily seizures and their treatment now have no seizures. Six children (22%) have nonhandicapping seizures and 3 of these have had ghost-like spells as described above. Thus, 89% (24/27) of children with Rasmussen's are no longer handicapped by significant seizures. Three children (11%) continue to have moderately severe seizures, although substantially less frequent and severe than before hemispherectomy (Table 3). Four of these 9 children's seizures are believed to arise from small amounts of residual mesial frontal or mesial temporal tissue inadvertently left behind at the time of surgery. These remaining seizures have not been sufficiently handicapping to the children to warrant reoperation. The fifth child's seizures come from the right (unoperated) hemisphere where EEG abnormalities appeared before left hemispherectomy during the 3 months of status epilepticus at another hospital.

Burden of Illness

Because we have advocated early surgery in children with Rasmussen's syndrome, many had a relatively mild hemiparesis before surgery and most had only mild to moderate intellectual impairment related to the disease and the medications. Thus, their preoperative disability was primarily due to the seizures. Despite the fact that in many of these children surgery increased their hemiparesis, the overall burden of their condition was lessened due to the marked decrease in both seizure frequency and severity, and to some extent to improvement in intelligence. (Table 4). The change in the total burden of

TABLE 3. Outcome of Seizures After Hemispherectomy as Shown by Etiology of the Seizures

Etiology	Frequency/Severity of Seizures			
	None	Insignificant	Moderate	Severe
Rasmussen's (n = 27)	18 (67%)	6 (22%)	3 (11%)	None
Dysplasias (n = 21) (3 died)	8 (38%)	6 (29%)	6 (29%)	1 (4%)
Vascular (n = 6) (1 died)	3 (50%)	1 (17%)	—	2 (33%)
Overall (n = 54)	29 (54%)	13 (24%)	9 (17%)	3 (6%)

TABLE 4. Burden of Illness Scores*

Etiology	Seizures Pre/Post	Motor Pre/Post	IQ/DQ Pre/Post	Total Pre/Post
Rasmussen's	2.6/0.4	1.7/1.6	1.7/1.4	6.0/3.5
Dysplasia	3.0/1.0	2.3/1.9	2.3/1.8	7.5/4.7
Vascular	3.0/1.2	1.8/2.0	1.5/1.3	6.3/4.5
Overall	2.8/0.7	2.0/1.8	1.9/1.5	6.7/4.0

* The average preoperative and postoperative burden of illness scores are shown for each etiology and for the overall population.

illness after the surgery is shown in Fig 2. No child in this group has been worsened by the surgery, and despite the permanent dysfunction of the upper extremity in all of the children after hemispherectomy, the burden of their disability has been lessened from an average score of 6.0 preoperatively to an average score of 3.5 postoperatively. The changes in the individual components of the burden of illness in this population is shown in Fig 3. The burden of the seizures in each child was moderate to severe (scored 2 or 3) before surgery and 22 of the children have shown reduction in burden of two or more levels. The children's current burden of illness is shown in Table 5.

Motor impairment was worsened in seven of the children who were operated early in the course of their disease (Fig 3). Thirteen of the 27 children had no change in their motor impairment, and 7 children had less motor impairment after surgery.

Despite removal of one hemisphere, the intellect of all but one of the children seems either unchanged or improved. The improvements are the result of the lessening of the impairments in function from the seizures, and from the decrease or, in most cases, elimination of the anticonvulsants. Intellect was worsened in the one child who has remained in a coma vigil-like state attributable to perioperative complications. Although there have been major concerns about loss of language after left hemispherectomy, all eleven of these children have regained virtually normal language.

**CHANGE IN OVERALL BURDEN OF ILLNESS
Rasmussen's Syndrome**

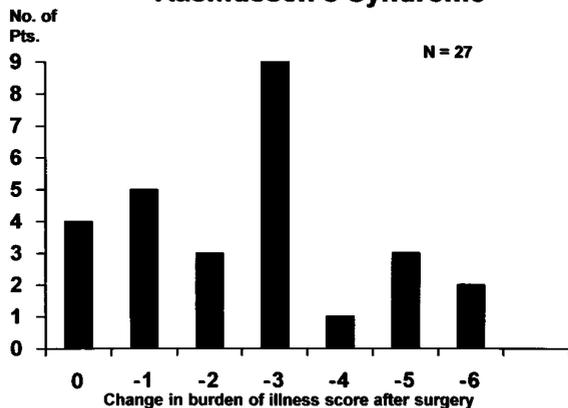


Fig 2. Change in overall burden of illness: Rasmussen's syndrome. The postoperative score was subtracted from the preoperative burden of illness score for each patient. The number of patients showing changes (0 to -6) is shown. No child had an increased burden postoperatively.

**CHANGE IN BURDEN OF ILLNESS
Rasmussen's Syndrome**

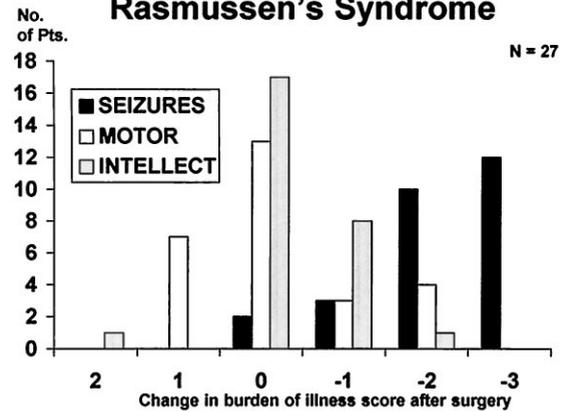


Fig 3. Change in burden of illness (individual factors): Rasmussen's syndrome. The postoperative score for each of the three factors was subtracted from the preoperative score. In the majority of instances, the burden lessened (negative change).

Cortical Dysplasias

Twenty-four children with unilateral, or predominantly unilateral cortical dysplasias underwent hemispherectomy (Table 1). There were 3 males and 21 females. Eleven had left hemispherectomies and thirteen had right hemispherectomies. Seizures in these children were predominantly unilateral, but at times the seizures generalized or were infantile spasm-like. At the time of the procedure, the children were all substantially delayed in development and further handicapped by the frequent daily seizures and the medications necessary to treat them. The operations were done at a far younger age than the other groups, some infants were as young as 3 months of age and 9 were under 1 year of age. The mean age at surgery was 3.0 years. As indicated previously, operations in this group were more difficult due to bleeding and problems related to fluid volumes and to vascular resistance. There were 2 perioperative deaths and 6 children who had substantial intraoperative and/or perioperative complications (discussed above).

Seizure Outcome

One patient with bilateral preoperative cortical problems had severe, continuing postoperative seizures and died 9 months after surgery. Eight of the surviving 21 children (38%) currently have no seizures, (Table 3), 6 additional children (29%) have mild (nonhandicapping) seizures, another 6 (29%) have moderately severe seizures, and only 1 child continues to have severely handicapping seizures. Although 1 child's current seizures may be related to

CHANGE IN OVERALL BURDEN OF ILLNESS DYSPLASIAS

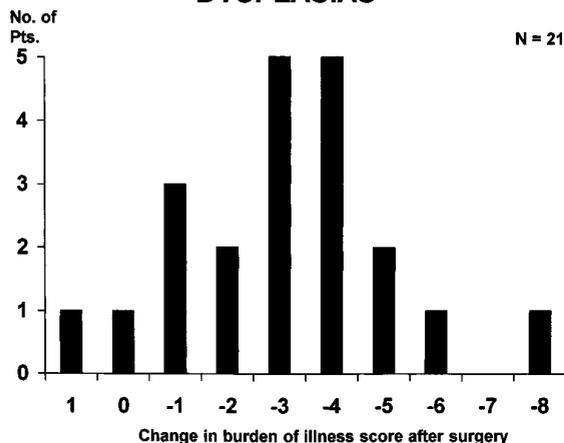


Fig 4. Change in overall burden of illness: dysplasias. The postoperative score was subtracted from the preoperative burden of illness score for each patient. The number of patients showing changes (+1 to -8) is shown. Only one child had an increased burden of illness as a result of the surgery.

perioperative problems, the others seem to have underlying abnormalities in the remaining hemisphere.

Burden of Illness

The overall change in the burden of illness in these children with cortical dysplasias is shown in Fig 4. Perioperative complications increased 1 child's burden and another was left unimproved. The average score was 7.5 preoperatively and was reduced to 4.7 postoperatively. The current burden of illness for these children is shown in Table 5.

The most dramatic improvement was seen in relief of the burden of seizures (Fig 5). One-third became seizure free (a decrease of 3 points). One-third experienced a moderate improvement (2 points), and one-third of the children with dysplasias experienced a mild improvement in seizure control (a decrease of 1 point).

Motor impairment was worsened by perioperative problems in 1 child. Eleven were unchanged, and nine have improved motor function after hemispherectomy.

The intellectual/developmental burden was judged to have improved to some degree in 12 children and stayed the same in 8. Six of the children with dysplasias are of normal intelligence or are only mildly retarded. Only the 1 child with major perioperative problems had worsening of motor and intellectual function. Language developed in the 11 chil-

dren with left hemispherectomy commensurate with the child's intellectual ability.

Vascular Problems

Seven children with vascular problems of multiple etiologies underwent hemispherectomy. These children varied in age, and in the degree of motor and mental handicap. The only commonality was the burden of seizures. Of the 3 children with Sturge-Weber syndrome, 1 died intraoperatively due to complications of the cardiac by-pass needed to control the bleeding from diploic veins (see above). A second child suffered major postoperative morbidity in the intensive care unit due to bleeding and fluid and electrolyte problems. The third child with Sturge-Weber syndrome, and 4 individuals with congenital CVAs (ages 2, 15, 18, and 19) were successfully operated without significant complications. These 5 children are walking and talking. Two remain significantly mentally retarded.

Seizure Outcome

Two-thirds of the surviving vascular patients (4/6) have few or no seizures (Table 3). One child with infrequent, nonhandicapping seizures has residual brain tissue on the operated side. In 1 of the 2 with significant seizures, the seizures are secondary to perioperative complications; the other's seizures are probably attributable to abnormalities in the remaining hemisphere.

Burden of Illness

The overall change in the burden of illness in this heterogeneous population was from a mean preoperative score of 6.3 to a postoperative score of 4.5 (Table 4). Most of the improvement occurred in the decrease in seizures. Motor burden was minimally worsened in this group and IQ/DQ possibly improved. The current burden of illness for these patients is shown in Table 5. One patient had an increased burden of illness because of perioperative complications and the other child whose burden has not changed substantially has continued seizures and mental retardation.

DISCUSSION

A decrease in the number of seizures was, and still is considered by many to be the best measure of effectiveness of the epilepsy surgery. However, for children with severe epilepsy, even complete seizure control may be an inadequate measurement of the

TABLE 5. Current Burden of Illness of Children Who Have Undergone Hemispherectomy

Etiology	Normalcy Burden = 0	Mild Burden <4*	Moderate Burden 4 to 8	Major Burden >8†
Rasmussen's n = 27	0	15	12	0
Dysplasias n = 21	0	6	12	3
Vascular n = 6	0	2	3	1
Overall n = 54	0	23	27	4

All children have at least a hemiplegic arm although walking, running and in school.

* Mild burden includes those children who may have some seizures or mental impairment.

† A major burden describes children with substantial impairment in two or more spheres.

CHANGE IN BURDEN OF ILLNESS DYSPLASIAS

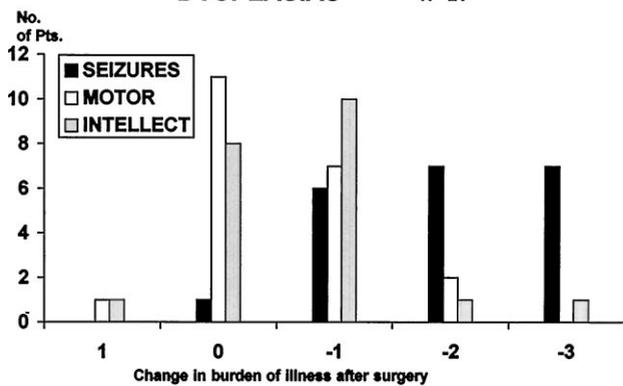


Fig 5. Change in burden of illness (individual factors): dysplasias. The postoperative score for each of the three factors was subtracted from the preoperative score. Almost half had a diminished motor burden, and more than half had improved from an intellectual standpoint.

success or failure of surgery.^{14,21,22} Our least successful patient, who remains in coma-vigil is seizure-free.

If one measures the success of surgery solely by the total relief from seizures, 54% of our hemispherectomy series are seizure free and an additional 24% have insignificant residual seizures (Table 3). Only 23% of the total population have continuing problems with seizures. However, residual seizure frequency is not independent of the underlying etiology. In our series, children with Rasmussen's syndrome have a 67% chance of becoming seizure-free; those with unilateral dysplasias have a 38% chance of being seizure-free, and 50% of children with vascular problems are seizure-free.

If we consider success to be a substantial decrease in seizures, then 89% of those with Rasmussen's syndrome, two-thirds of infants with dysplasias, and two-thirds of those with vascular problems are now either seizure-free or have only nonhandicapping seizures and would be considered to have a successful outcome. We believe that these impressive results can be made even better as perioperative problems are brought under control, and as even more careful attention is paid to removal of residual gray matter along the mesial temporal lobe and in the insula.

Seizure control, however, is not itself an adequate measure of the success of epilepsy surgery,²² and quality of life has become a fashionable supplementary measure.²³ When one attempts to apply the concept of quality of life to a heterogeneous population of children, of various ages and with varying disabilities, who have either progressive or static diseases, measurements of quality of life become increasingly elusive. Our populations consist of children with multiple interacting handicaps. None of these children were normal at the time of surgery and none will ever have normal motor function. All have, at a minimum, a permanent hemiplegic arm. However, many of our children have excellent qualities of life with variable burdens of illness. Both before surgery and at follow-up the children with dysplasias have had combinations of mental retardation and motor

dysfunction as well as seizures. The children with Rasmussen's syndrome were handicapped by seizures, medications, and increasing motor and mental dysfunction. They seem to have a static encephalopathy with less impairment than at the time of surgery.

We have attempted to construct a simple assessment of the burden of these handicaps that will provide a clearer picture of the changes in burden after surgery. This first attempt at the construction of a burden of illness scale is imperfect and unvalidated. Our scale starts with the child's mental and motor disability and then superimposes the additional disability due to the seizures, their treatment, and the family's response and overprotection. In the future, as we assess the total effect of the disorder we must also assess the burden of illness which also falls on the parents, the siblings, and the community. What is needed is a scale which is simple, validated and reproducible and which would give a more complete assessment of the benefits and costs of surgery and other interventions.

Defining the Success of Hemispherectomy

Success of surgery must be measured in terms of the child's preoperative condition and the change in that condition after surgery. For the vast majority of children, despite substantial postoperative dysfunction of the paretic arm, the burden of seizures, and the overall residual dysfunction has been lessened. Some authors²¹ have been concerned about increasing the motor dysfunction in children, such as those with Rasmussen's syndrome, who preoperatively have less than a total hemiplegia. Some have recommended deferring hemispherectomy until the hemiplegia is complete. Because a hemiparesis is inevitable in this disorder, we have been willing to cause earlier onset of the motor dysfunction in order to prevent the inevitable intellectual deterioration of Rasmussen's syndrome.

If the success of hemispherectomy is defined only as the return of the child to normalcy, then none of our children are successes, but only 4 of the 54 children have a major burden. Burden of illness will vary with the underlying etiology.

Rasmussen's Syndrome

All children with Rasmussen's now are in school, some in special classes. Despite their hemipareses, all except the child with the locked-in syndrome, are walking with or without short leg braces; all are functioning independently despite the limitations of their helper hand. Language has not been a problem even for the eight right-handed children with Rasmussen's syndrome who have undergone a left hemispherectomy, the oldest 2 being 12 and 13 years of age at the time of the surgery. While preliminary studies of their language recovery have been reported elsewhere,^{24,25} language has redeveloped in all, although more completely and with a shorter delay in the younger children, and in those who have had a shorter course of the disease. Left hemisphere involvement in Rasmussen's syndrome should not be a reason for delaying or denying the operation,

and indeed may be a reason for operating at an earlier stage.

One child with Rasmussen's has seizures from remaining tissue on the operated side. These seizures interfere with his function, and he continues to have a moderate burden.

Dysplasias

Despite the fact that many of the children with dysplasias had bilateral preoperative EEG abnormalities, the presence of predominantly unilateral abnormalities on magnetic resonance imaging scans and unilateral predominance on the EEG, in the setting of incapacitating, life-threatening seizures, led us to perform the surgery with the parental understanding that we were unlikely to make the child worse.

Motor function seemed to improve after hemispherectomy in nine of these children and seemed to be the result of decrease in spasticity on the paretic side.

Virtually all parents, even those whose children have died, when asked, state that they are pleased that they gave their children the chance at the operation. Those whose children survived, would classify the success of the procedure with varying degrees of enthusiasm. Most children are in school. Most parents are happy with the outcome and the burden of the illness has been decreased in 19 of the 21 children. Three of the surviving children are at home, profoundly retarded, and with substantial seizures and have a major residual burden (Table 5). Although those parents are glad the surgery was performed, the burden of the child's handicap was not substantially lessened.

Vascular Problems

The children with vascular problems are a small and heterogeneous group. Four of the 5 who had no perioperative problems have done well, and 3 of these are without seizures. All 4 have good function. Two of the 6 have continuing seizures and substantial residual mental retardation. In 1 of these the problems are most-likely perioperative.

Conclusion

We have learned that in both the Rasmussen and the dysplasia group, earlier surgery seems preferable. It is not necessary, as was once taught, to wait until the hemiplegia is complete before performing a hemispherectomy. When the course of the condition is clear, and the difficulty in controlling the seizures is obvious, surgery should be performed, regardless of the side of the disease and the availability, or potential availability, of yet another new anticonvulsant.

Once the necessity of sacrificing the motor strip is accepted, leaving involved portions of the frontal lobe or the occipital lobe without complete disconnection inevitably leads to continued seizures. There seems to be little advantage to one type of hemispherectomy over another, if cortical removal and/or disconnection is complete, but complete disconnection is often surgically difficult. The risk of death from superficial cortical hemosiderosis, which

was the motivation for the development of functional hemispherectomy, seems to have disappeared with current surgical techniques, and as newer imaging modalities have permitted the early evaluation of hydrocephalus.

Surgery should never be performed until both parents understand the potential complications of the surgery and the nature of the expected residual handicaps, and can accept any of the potential surgical outcomes. Thus, although we advocate early surgery, such surgery is never an emergency, and is rarely urgent. Time for parental reflection and acceptance is of primary importance. Families require enormous support preoperatively, postoperatively, and throughout the long term. Older children may need counseling and support during adolescence. This maximally invasive surgery should only be performed in a limited number of centers where an experienced team of surgeons, epileptologists, psychologists, and social workers, can be concentrated, and a network of parent support from those who have gone before is in place.

Hemispherectomy is a far more difficult and risky procedure in young infants and in those with developmental and vascular problems, and has far higher intraoperative and perioperative morbidity and mortality in these groups than the same procedure done by the same team in older children. When there are no complications, these children leave the hospital for rehabilitation 10 days after surgery.

Speculation

Why should removal of half a brain be of benefit to a child? Decrease in constant, uncontrollable seizures and the attendant decrease in medications with their side-effects may be part of the reason. However, it is tempting to speculate, that the continuous electrical activity of these severely dysfunctional hemispheres interferes with the function of the other, more normal hemisphere. This might explain why motor function improves after hemispherectomy and why language recovers after removal of the dysfunctional left hemisphere,²⁶ but does not seem to fully transfer before surgery. Perhaps it also partially explains intellectual improvement in these children after removal of half of the cortex. We are awed by the apparent retention of memory after removal of half of the brain, either half, and by the retention of the child's personality and sense of humor. Yet we look forward to the time when there are less mutilating approaches to these problems. Until then it seems that half of a brain is less burdensome to these children than a whole brain where one side is badly misfiring.

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Why Would You Remove Half a Brain? The Outcome of 58 Children After Hemispherectomy —The Johns Hopkins Experience: 1968 to 1996
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