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The Cognitive Outcome of Hemispherectomy in 71 Children

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Summary: *Purpose:* Long-term neuropsychological outcome was studied in 71 patients who underwent hemispherectomy for severe and intractable seizures at The Johns Hopkins Hospital between 1968 and 1997 and who agreed to participate. Seizures were due to cortical dysplasias (n = 27), Rasmussen syndrome (n = 37), or vascular malformations or strokes (n = 7). Both presurgical and follow-up results are available and reported for 53 patients.

Methods: Patients and caretakers were interviewed, and patients were administered standard measures of intelligence, receptive and expressive language, visual-motor skills, adaptive/developmental functioning, and behavior.

Results: Mean age at surgery was 7.2 years. At follow-up, on average 5.4 years after surgery, 65% are seizure free, 49% are medication free, and, of those responding, none rated quality of life as worse than before surgery. Mean IQ was in the 70s for Rasmussen and vascular patients and in the 30s for cortical dysplasia patients. Language and visual-motor skills were consistent with IQ. For Rasmussen patients only, language was significantly more impaired for left than for right hemispherectomy, both before surgery and at follow-up. Adaptive skills were mildly impaired, with greatest impairment in the physical domain. Cognitive measures typically changed little between surgery and follow-up, with IQ change <15 points for 34 of 53 patients; of the remainder, 11 declined and eight improved. Behavior was free of major problems, but social interactions and activities were limited.

Conclusions: The most significant predictor of cognitive skills at follow-up was etiology, with dysplasia patients scoring lowest in intelligence and language but not in visual-motor skills. Regardless of etiology, most patients showed only moderate change in cognitive performance at follow-up. **Key Words:** Hemispherectomy—Neuropsychology.

Cerebral hemispherectomy has been available as a treatment for severe, intractable unilateral epilepsy in children since at least 1945 (1). Whereas many studies have shown improved control of seizures (2,3) and general improvement, or at least no significant decline, in overall intellectual performance after hemispherectomy (4–10), the major studies have rarely gone beyond measurement of overall intelligence (IQ).

Studies in adults suggest that left hemispherectomy leads to differential impairment in language, and right hemispherectomy, to particular loss of spatial skills; in children, however, this pattern has not generally been reported (8–12), at least not in a straightforward way (13,14). Several studies have reported a general trend toward more functional independence (12) and a sometimes marked reduction in disruptive behavior (3,10,13) in children after hemispherectomy. However, such improvement has been hard to quantify, because the studies have generally been based on anecdotal reports or individual clinical observations rather than standardized measures or structured interviews.

At the Johns Hopkins Hospital, hemispherectomy has been performed for children with severe intractable epilepsy since 1968. Earlier reports described the clinical procedure and documented the neurologic outcomes of some of these children (2,16–19). Here we present the results of comprehensive cognitive assessments of all of those children who underwent hemispherectomy at Johns Hopkins between 1968 and 1997 and who could be followed up.

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METHODS

Patients

Seventy-nine children underwent either hemidecortication (n = 77) or anatomic hemispherectomy (n = 2) (2,15) at the Johns Hopkins Hospital between 1968 and 1997. At follow-up in 1996–1998, four were dead (18,19), two could not be located, and two declined to be evaluated. The

TABLE 1. *Cognitive test battery*

Area of functioning	Age	Release date	Measure
Intelligence	1–30 mo	1969	Bayley Scales of Infant Development[23]
	2.5–3 yr	1986	Stanford-Binet Intelligence Scale-Fourth Edition[24]
	4–5 yr	1989	Wechsler Preschool and Primary Scale of Intelligence-Revised (WPPSI-R),[25] short form[26]
	6–16 yr	1974	Wechsler Intelligence Scale for Children-Revised (WISC-R),[27] short form[28]
	17+ yr	1981	Wechsler Adult Intelligence Scale-Revised (WAIS-R),[29] short form[30]
Receptive Language	2+ yr	1981	Peabody Picture Vocabulary Test Revised (PPVT-R)[31]
Expressive Language	2+ yr	1979	Expressive One Word Picture Vocabulary Test (EOWPVT)[32]
Visual-Motor	4+ yr	1982	Developmental Test of Visual-Motor Integration (VMI)[33]
Developmental/adaptive	1 mo–9.5 yr	1992	Developmental Profile-Second Edition (DP-II)[34]
Psychosocial/behavioral	2–18 yr	1991/1992	Child Behavior Checklist (CBCL)[35,36]

remaining 71 patients or their caregivers (as appropriate for patient age and level of competence) gave informed consent to participate in this study in compliance with the requirements of the Johns Hopkins Joint Committee on Clinical Investigation.

Procedure

A comprehensive set of cognitive tests (Table 1) was administered to patients both before surgery and at follow-up, as appropriate for the child's chronologic age and level of functioning. The test battery assessed six areas of functioning, including general intelligence, developmental/adaptive skills, and behavior, as well as three tests sensitive to left- and right-hemispheric function (language and visuospatial tests). However, some patients were not administered all measures, because their chronologic or functional age was outside the test limits (shown in Table 1), because of high seizure frequency, or, in some cases, because surgery predated publication of the test. Patients were located in 18 states and were tested either at the Johns Hopkins Hospital or at their homes.

Information about postsurgical quality of life was obtained by means of a structured interview with the caregiver and, if appropriate, with the patient. Such information included seizure frequency and medication use, most recent level of educational services, and current living arrangements. Seizure outcome was reported by using a 4-point scale based on that of Engel (20). General health was rated on a 4-point scale, and satisfaction with surgery, on a 3-point scale. Patients were administered an overall quality-of-life measure analogous to the Wong-Baker Faces Scale (21), a nonverbal measure with demonstrated validity for assessment of pain in children age 3 years and older (22). Referring to a visual analog (smiley faces expressing different degrees of happiness), respondents rated their current level of happiness on a scale from 0 ("very unhappy") to 10 ("most happy"). Patients also were asked to state what aspect of life was most difficult for them at present.

Data analysis

Cognitive tests were used to assess functioning at the follow-up evaluation for all 71 patients and to examine postsurgical change in functioning for those patients with both pre- and postoperative test results. Patients were grouped by the side of surgery and also by the disorder giving rise to their seizures: Rasmussen syndrome, cortical dysplasia, or congenital vascular problems (19). The cortical dysplasia group included children with a variety of developmental malformations, including hemimegalencephaly. The vascular group included children with Sturge-Weber syndrome and those who had experienced congenital cerebrovascular accidents. The relation between etiology or side and test results was examined by using χ^2 analysis and one-way analysis of variance (ANOVA). Tukey's HDS post hoc analyses were performed on any dependent measure for which ANOVA showed a significant result. For each of the three disorders, the differences in patient clinical characteristics by side of hemispherectomy and changes in characteristics between presurgical and follow-up evaluations were examined by using χ^2 analysis and one-way ANOVA. For each of the disorders, the relation between side of surgery and pre- and postoperative performance on the six cognitive outcome measures (intelligence, receptive language, expressive language, visual-motor skills, developmental/adaptive functioning, and behavior) was examined by using 2×2 repeated-measures ANOVA, with side of surgery as the between-subjects factor and time of evaluation (preoperative/postoperative) as the within-subjects, repeated-measures factor. Statistical analyses were performed by using SPSS version 10. All reported p values used two-tailed tests of significance with α (alpha) set at 0.05.

RESULTS

At least one follow-up evaluation was conducted for all 71 patients; in addition, presurgical test results were available for a subset of 53 patients. In the following, results are presented in two sections: first, follow-up data for all subjects, called the cross-sectional group ($n = 71$), and then cognitive test results for the longitudinal group ($n = 53$).

TABLE 2. *Total sample characteristics by etiology*

	Rasmussen (n = 37)	Dysplasia (n = 27)	Vascular (n = 7)
Gender (male/female)	13/24	7/20	5/2
Side of surgery (left/right)	16/21	15/12	2/5
Age at seizure onset (yr)[a]	6.0 (3.2)	0.7 (2.1)	2.4 (2.6)
Age at surgery (yr)[b]	9.2 (4.0)	4.6 (5.3)	6.8 (5.9)
Duration of seizures (yr)	3.2 (3.0)	3.9 (4.8)	4.4 (4.5)
Age at follow-up (yr)[c]	14.9 (7.6)	11.0 (9.3)	10.9 (9.3)
Interval, surgery to post-op evaluation (yr)	5.7 (6.1)	5.3 (3.5)	4.1 (4.5)

Values expressed as mean (SD); n = 71.
[a] $p < 0.001$.
[b] $p < 0.01$.
[c] $p < 0.05$.

TABLE 3. *Total sample characteristics at follow-up evaluation*

	Rasmussen (n = 37)	Dysplasia (n = 27)	Vascular (n = 7)
Seizure outcome classification[a]			
I. Seizure free	27 (73%)	12 (44%)	7 (100%)
II. Rare disabling seizures	8 (22%)	8 (30%)	—
III. ≥75% reduction in seizures	2 (5%)	4 (15%)	—
IV. <75% reduction in seizures	0 (0)	3 (11%)	—
Medication free			
Yes	18 (49%)	11 (41%)	6 (86%)
No	19 (51%)	16 (59%)	1 (14%)
Satisfaction with surgery			
Pleased	34 (92%)	24 (89%)	7 (100%)
Satisfied with reservations	2 (5%)	2 (7%)	0 (0)
Dissatisfied	1 (3%)	1 (4%)	0 (0)
General health, parental report			
Excellent	10 (27%)	4 (15%)	2 (29%)
Good	26 (70%)	16 (59%)	5 (71%)
Fair	1 (3%)	7 (26%)	0 (0)
Poor	0 (0)	0 (0)	0 (0)
Current educational services			
Mainstream, no services	11 (29%)	0 (0)	3 (42%)
Mainstream with support	15 (41%)	6 (22%)	2 (29%)
Full-day special education	10 (27%)	18 (67%)	2 (29%)
Home schooling	1 (3%)	3 (11%)	0 (0)

[a] Based on Engel's Classification of Postoperative Outcome (1993); $p < 0.05$.

Cross-sectional results (n = 71)

As shown in Table 2, the total sample is predominantly (65%) female and almost evenly divided between left- and right-side hemispherectomy. For just over half of patients (52%), the cause of seizures was Rasmussen encephalopathy; for the remainder, the cause was either cortical dysplasia (38%) or a vascular etiology (10%). No significant difference by etiology was found in the likelihood of left- or right-sided hemispherectomy. As expected, one-way ANOVA found significant differences by etiology in age at seizure onset and age at surgery, both of which were on average significantly later in the Rasmussen than in the vascular and cortical dysplasia groups. The Rasmussen group also was significantly older at follow-up than the vascular or cortical dysplasia groups. Seizure onset was before the first year of life for 24 of 27 dysplasia patients, three of seven vascular patients, and one of 37 Rasmussen patients; latest onset was for a Rasmussen patient, at age 12 years. Age at surgery was between 0.25 and 20.6 years, and age at follow-up ranged from 2.4 to 37.5 years. No significant difference by etiology was found in the duration of seizures or in the time interval between surgery and the follow-up evaluation.

Overall status of patients at follow-up is given in Table 3. Almost all patients experienced a substantial reduction in seizure frequency: disabling seizures were eliminated in 46 (65%) of 71 patients, whereas only three (4.2%) reported no meaningful seizure reduction. Seizures were eliminated in all patients with vascular etiology, in 73% of those with Rasmussen, and in 44% of those with cortical dysplasia, a statistically significant difference by etiology [$\chi^2(2, 71) = 9.79$, $p < 0.01$]. The three patients for whom handicapping seizures persisted at follow-up were all in the cortical dysplasia group. The need for anticonvulsant medications (AEDs) also was greatly reduced at follow-up, particularly in the vascular group, although the differences by etiology were not statistically significant. Overall, 35 (49%) of 71 patients were no longer taking AEDs at follow-up.

Most respondents reported being satisfied at follow-up with the surgery, with only six (8.5%) expressing any reservations. (The respondents were 63 caregivers and eight patients.) All etiology groups reported comparable levels of satisfaction. General health was usually reported to be good; the most common physical complaints were headache and fatigue. Although no patients reported "poor" health at follow-up, the cortical dysplasia group was much more likely to report health problems.

A variety of educational settings were reported. More than half of all children were "mainstreamed" in school, with or without support; however, the likelihood of mainstream education was lower for the cortical dysplasia group than for the other etiologies. Almost all patients (94%) lived with a parent or with similar support. Because of the young age of the sample (83% were younger than 17 years), no conclusion can be drawn regarding employment prospects or capacity for living independently.

The quality-of-life measure was completed by 41 of the 71 patients. Responses were not available for patients with very low verbal skills or for those who were otherwise unable to comprehend the task. The respondent group consisted of 27 (66%) Rasmussen, 10 (24%) dysplasia, and four (10%) vascular etiology, a notably higher proportion of Rasmussen and lower proportion of dysplasia etiology than that in the overall sample. Compared with the total sample, the respondent group was significantly older at seizure onset (4.82 ± 3.72 years) and at surgery (8.69 ± 5.18 years), and experienced a significantly longer interval between surgery and follow-up evaluation

(6.48 ± 5.84 years). Overall cognitive functioning was much higher for respondents (mean IQ, 71.4 ± 20.0) than for nonrespondents (mean IQ, 43.7 ± 25.8). Of those patients responding, 20 (49%) of 41 rated their life within the past month as "most happy" (a score of 10), and none rated their current life as "unhappy" (score, <5). The likelihood of a "most happy" rating was independent of etiology, side of surgery, degree of seizure reduction, or postsurgical need for AEDs. The most common quality-of-life complaint concerned physical handicaps related to hemiplegia—for example, the inability to ride a bicycle or to use scissors with ease.

Patient characteristics were generally not significantly different by side of hemispherectomy across the three etiologies; except in one instance, one-way ANOVA showed no significant difference by side for each etiology group in gender composition, age at seizure onset, age at surgery, duration of seizures, age at follow-up evaluation, and the interval between surgery and most recent follow-up evaluation. The exception is in the dysplasia group, in which the interval between surgery and follow-up evaluation was significantly longer for right-hemispherectomy patients (mean, 7.26 ± 2.89 years) than for left-hemispherectomy patients (mean, 3.74 ± 3.20 years). Neither persistence of seizures nor need for AEDs after surgery was significantly related to side of surgery for the total sample or within any of the etiology groups. However, meaningful statistical comparisons by side are difficult for the vascular group, which consisted of only two left-hemispherectomy and five right-hemispherectomy patients.

Cognitive outcome by etiology

Results of the cognitive assessment at follow-up for the total sample are shown in Table 4. Significant differences between etiology groups were found, using one-way ANOVA, in general intelligence, receptive language, and expressive language, but not in visual-motor skills. Post hoc analyses indicated that no significant difference was present between the Rasmussen and vascular groups in intelligence or language skills. Both Rasmussen and vascular groups scored significantly higher than the cortical dysplasia group in general intelligence and receptive language. In expressive language, the vascular group scored significantly higher than the cortical dysplasia group, whereas the Rasmussen and dysplasia groups were not significantly different.

Several of the patients in the dysplasia group were unable to complete one or more of the language and visual-motor tests, so the results in Table 4 overstate that group's functioning in those areas relative to the other etiology groups. Nonetheless, an analysis conducted for only the 56 patients who completed the full cognitive test battery at follow-up found the same relations between the etiology groups as just reported. Of those patients who obtained an IQ but not an expressive-language score, nine, all in the dysplasia group, were at a preverbal age (18 months or younger) at surgery. The remainder—two in the dysplasia group and one each in the Rasmussen and vascular groups—were functionally preverbal at follow-up assessment.

In adaptive/developmental functioning, as assessed by the Developmental Profile-II (DP-II), one-way ANOVA found significant differences by etiology. The DP-II is a parent-report inventory that assesses skills in five domains (listed in Table 4) in terms of a developmental quotient (DQ), which is the ratio of developmental age to chronologic age expressed as a percentage. In addition, a DP-II Total DQ score, computed as the mean of the five domain DQs, serves as an overall index of developmental and adaptive level. Post hoc analyses indicated that, with one exception, both the vascular and Rasmussen groups scored significantly higher in all DP-II domains than did the cortical dysplasia group, and no significant difference was noted between the vascular and Rasmussen group scores. The one exception is the DP-II Physical domain, in which the vascular group scored significantly higher than both the Rasmussen and dysplasia groups, and the Rasmussen and dysplasia group scores were not significantly different. The Physical domain measures both gross and fine motor skills, such as the ability to hop on one foot, ride a bicycle, or use scissors.

Overall, little evidence was noted at follow-up of significant behavioral or emotional problems, as measured by the Child Behavior Checklist (CBCL). The CBCL is a parent-report measure that yields area scores that are combined to form a Total Problems T-score. Higher scores indicate greater emotional or behavior problems, with T-scores of >60 denoting clinically significant problems. For ages 4–18 years, the CBCL also yields a Total Competence T-score, a measure of social activity for which lower scores suggest less social interaction in daily living. The mean Total Problems T-scores for each etiology group in Table 4 are within the normal range, but the clinically low mean Total Competence T-scores indicate overall problems engaging in social interactions and activities. One-way ANOVA indicated no evidence of any significant difference in overall emotional/behavioral functioning or in social competence between the etiology groups.

Cognitive outcome by side of hemispherectomy

Results of the cognitive assessment at follow-up are shown in Table 5 within each etiology group by side of hemispherectomy. Significant differences by side of hemispherectomy were found, using one-way ANOVA, only for the Rasmussen group. In both receptive and expressive language, right-hemispherectomy Rasmussen patients scored higher (mean scores in the low-average range) than left-hemispherectomy patients (mean scores in the impaired range). Right-hemispherectomy patients

TABLE 4. *Cognitive test results at follow-up evaluation by etiology for total sample*

	Etiology group				
	1 Rasmussen	2 Dysplasia	3 Vascular	ANOVA *F*	Post hoc
General Intelligence (n = 71)	n = 37	n = 27	n = 7		
IQ/MDI/DQ	72.8 (20.1)	37.0 (19.6)	78.3 (15.4)	29.95[a]	1 > 2; 3 > 2
Receptive Language (n = 64)	n = 37	n = 20	n = 7		
Peabody Picture Vocabulary Test-R SS/DQ	78.9 (21.3)	49.2 (18.5)	71.1 (13.1)	14.74[a]	1 > 2; 3 > 2
Expressive Language (n = 59)	n = 36	n = 16	n = 6		
Expressive One-Word Picture Vocabulary Test SS/DQ	72.2 (24.2)	57.4 (24.0)	88.3 (25.1)	4.32[b]	3 > 2
Visual-Motor (n = 58)	n = 37	n = 15	n = 6		
Developmental Test of Visual-Motor Integration SS/DQ	68.8 (21.3)	59.6 (16.1)	68.7 (19.2)	1.19	—
Developmental Functioning (n = 64)	n = 31	n = 27	n = 6		
Developmental Profile-II					
Total DQ	67.1 (21.0)	40.3 (23.7)	76.5 (8.5)	14.01[a]	1 > 2; 3 > 2
Communication DQ	73.7 (27.1)	41.0 (25.8)	77.8 (8.9)	13.43[a]	1 > 2; 3 > 2
Socialization DQ	73.2 (24.7)	41.9 (25.1)	77.5 (16.6)	13.56[a]	1 > 2; 3 > 2
Academic DQ	74.6 (26.5)	40.4 (24.8)	73.3 (13.1)	14.44[a]	1 > 2; 3 > 2
Physical DQ	47.8 (21.7)	34.8 (21.4)	73.2 (13.6)	8.88[a]	3 > 1; 3 > 2
Self-Help DQ	66.0 (24.4)	43.3 (25.4)	80.5 (8.1)	9.43[a]	1 > 2; 3 > 2
Psychosocial/Behavior (n = 63)	n = 33	n = 24	n = 6		
Child Behavior Checklist (Mean, 50; SD, 10)					
Total Problems T-score	55.12 (13.59)	52.71 (9.56)	53.17 (14.54)	0.28	—
Total Competence T-score	36.54 (12.30)	33.00 (11.92)	40.00 (8.04)	0.54	—

Values expressed as mean (SD).
IQ, Wechsler short form or Stanford-Binet; MDI, Bayley Mental Development Index; DQ, Developmental Quotient; SS, standard score.
[a] $p < 0.01$.
[b] $p < 0.05$.

in the Rasmussen group also tended to score higher in IQ ($p = 0.05$) and in the DP-II Communication domain ($p = 0.05$). No significant difference by side was found in scores for visual-motor skills, total adaptive/developmental functioning, or in the remaining four of the five DP-II domain scores. For the cortical dysplasia and vascular groups, one-way ANOVA found no significant difference by side in any of the cognitive measures.

No evidence was seen of any significant difference in overall emotional/behavioral functioning by side of

TABLE 5. *Cognitive test results at follow-up evaluation by side of surgery*

	Rasmussen (n = 37)		Dysplasia (n = 27)		Vascular (n =7)	
Etiology group Side	Left (n = 16)	Right (n = 21)	Left (n = 15)	Right (n = 12)	Left (n = 2)	Right (n = 5)
General Intelligence (n = 71)	n = 16	n = 21	n = 15	n = 12	n = 2	n = 5
IQ/MDI/DQ	65.6 (13.4)	78.3 (22.7)[a]	35.3 (18.2)	39.0 (21.8)	93.5 (20.5)	72.2 (9.4)
Receptive Language (n = 64)	n = 16	n = 21	n = 11	n = 9	n = 2	n = 5
Peabody Picture Vocabulary Test-R SS/DQ	66.1 (12.6)	88.8 (21.5)[b]	44.0 (14.3)	55.6 (21.9)	76.5 (24.8)	69.0 (9.1)
Expressive Language (n = 59)	n = 16	n = 20	n = 8	n = 8	n = 2	n = 5
Expressive One-Word Picture Vocabulary Test SS/DQ	62.5 (19.3)	80.0 (25.2)[c]	54.4 (19.4)	60.4 (28.9)	70.0 (48.1)	95.6 (11.7)
Visual-Motor (n = 58)	n = 16	n = 21	n = 8	n = 7	n = 2	n = 4
Developmental Test of Visual-Motor Integration SS/DQ	68.6 (18.8)	69.0 (23.5)	62.1 (11.3)	56.7 (20.9)	53.5 (31.8)	76.3 (6.7)
Developmental Functioning (n = 64)	n = 14	n = 17	n = 15	n = 12	n = 1	n = 5
Developmental Profile-II						
Total DQ	62.5 (20.3)	70.8 (21.4)	41.3 (19.8)	39.02 (28.7)	82 (0)	75.4 (9.1)
Communication DQ	63.6 (24.5)	82.1 (26.9)[a]	39.2 (21.1)	43.33 (31.6)	85 (0)	76.4 (9.2)
Socialization DQ	68.9 (23.3)	76.8 (25.9)	44.3 (23.2)	38.92 (28.1)	85 (0)	76.0 (18.1)
Academic DQ	67.4 (23.3)	80.5 (28.2)	40.9 (20.9)	39.92 (29.9)	85 (0)	71.0 (13.2)
Physical DQ	44.4 (18.8)	50.6 (23.9)	37.8 (18.0)	31.00 (25.3)	85 (0)	76.4 (9.2)
Self-Help DQ	68.2 (22.0)	64.2 (26.8)	44.4 (20.5)	41.92 (31.5)	70 (0)	73.8 (15.1)
Psychosocial/Behavior (n = 63)	n =15	n = 18	n = 14	n = 10	n = 1	n = 5
Child Behavior Checklist						
Total Problems T-score	56.3 (14.1)	54.1 (13.4)	50.6 (11.0)	55.60 (6.7)	46 (0)	54.6 (15.8)
Total Competence T-score	36.1 (12.9)	36.9 (12.2)	32.6 (9.9)	33.50 (15.7)	46 (0)	36.3 (4.0)

Values expressed as mean (SD).
IQ, Wechsler short form or Stanford-Binet; MDI, Bayley Mental Development Index; DQ, Developmental Quotient; SS, standard score.
[a] $p = 0.05$.
[b] $p < 0.01$.
[c] $p < 0.05$.

TABLE 6. *Rasmussen syndrome: longitudinal sample characteristics by side of surgery*

	Total n = 31	Left n = 11	Right n = 20
Age at seizure onset (yr)	6.4 (3.3)	5.8 (3.1)	6.7 (3.4)
Age at surgery (yr)	9.7 (4.1)	8.9 (2.9)	10.1 (4.6)
Duration of seizures (yr)	3.3 (3.2)	3.1 (1.6)	3.4 (3.9)
Age at follow-up evaluation (yr)	14.7 (7.2)	12.3 (3.1)	16.1 (8.4)
Interval, surgery to post-op evaluation (yr)	5.1 (4.9)	3.5 (2.8)	5.9 (5.6)

Values expressed as mean (SD).

hemispherectomy across the three groups. The CBCL T-scores were in the normal range for all groups, and the social competence T-scores were equally low for the right- and left-hemispherectomy patients in all groups.

Longitudinal sample (n = 53)

Longitudinal data for those patients with pre- and post-operative evaluations are presented here separately by the three etiology groups. General intelligence (IQ) was assessed for all 53 patients in the longitudinal group; results of the other tests were available for a subset of this group.

Rasmussen syndrome

Longitudinal data were available for 31 of the 37 Rasmussen patients. The characteristics of this subgroup, given in Table 6, are not substantially different from those given in Table 2 for the larger group. One-way ANOVA showed no significant difference by side of surgery in terms of age at seizure onset, age at surgery, duration of seizures, age at follow-up testing, or time interval between surgery and follow-up evaluation.

Results of cognitive tests for the Rasmussen group are shown in Table 7 and illustrated in Fig. 1 by side of surgery. For patients with Rasmussen syndrome, the left-hemispherectomy group scored consistently lower than the right-hemispherectomy group in general intelligence (IQ), receptive language (PPVT), and expressive language (EOWPVT); repeated-measures ANOVA found these group differences to be statistically significant. No significant effect of side of surgery was found on visual-motor abilities or behavior. In terms of overall adaptive/developmental functioning, the left-hemispherectomy group obtained lower scores, but the group differences fell short of statistical significance ($p = 0.07$). A significant effect of side of surgery occurred on two DP-II domains, academic and communication skills, in which the left-hemispherectomy group obtained lower scores, but not in physical, socialization, and self-help adaptive skills.

A statistically significant effect of time of evaluation (presurgical/follow-up) across the side-of-surgery groups was found only on expressive language scores, which were lower on average at follow-up than at presurgical evaluation. Repeated-measures ANOVA found no significant change at follow-up in general intelligence, receptive language, visual-motor skills, behavior, or adaptive/developmental functioning. No significant interaction was found between side of surgery and time of evaluation for any of the six outcome measures; that is, any change in test scores at follow-up was independent of side of surgery.

Cortical dysplasia

Longitudinal data were available for 15 of the 27 dysplasia patients. As can be seen in Table 8, patients in

TABLE 7. *Rasmussen syndrome: cognitive test results at preoperative and follow-up evaluation by side of surgery*

	Total		Left		Right	
	Pre-Op	Post-Op	Pre-Op	Post-Op	Pre-Op	Post-Op
General Intelligence (n = 31)	n = 31	n = 31	n = 11	n = 11	n = 20	n = 20
IQ/MDI/DQ	80.5 (24.1)	75.7 (20.1)	71.5 (28.4)	67.5 (14.8)	85.6 (20.5)	80.3 (21.5)
Receptive Language (n = 26)	n = 26	n = 26	n = 10	n = 10	n = 16	n = 16
Peabody Picture Vocabulary Test-R SS/DQ	79.0 (21.8)	82.1 (18.4)	68.9 (19.1)	68.7 (11.7)	85.4 (21.5)	90.4 (17.0)
Expressive Language (n = 20)	n = 20	n = 20	n = 8	n = 8	n = 12	n = 12
Expressive One-Word Picture Vocabulary Test SS/DQ	89.3 (21.7)	82.0 (21.1)	76.5 (22.3)	68.1 (17.1)	97.8 (17.4)	91.3 (18.6)
Visual-Motor (n = 23)	n = 23	n = 23	n = 9	n = 9	n = 14	n = 14
Developmental Test of Visual-Motor Integration SS/DQ	76.8 (17.5)	77.0 (11.5)	72.1 (10.4)	76.9 (8.7)	79.9 (20.6)	77.2 (13.4)
Developmental Functioning (n = 16)	n = 16	n = 16	n = 10	n = 10	n = 6	n = 6
Developmental Profile-II						
Total DQ	72.6 (22.1)	72.4 (14.6)	67.6 (24.1)	65.6 (12.8)	80.9 (17.1)	83.5 (10.2)
Communication DQ	77.0 (31.2)	76.0 (21.3)	68.7 (35.7)	65.5 (19.9)	90.8 (15.8)	93.5 (7.3)
Socialization DQ	71.6 (22.4)	77.2 (16.6)	65.9 (25.5)	72.1 (17.2)	81.0 (12.7)	85.7 (12.6)
Academic DQ	78.2 (21.9)	79.2 (18.5)	71.9 (24.8)	71.0 (17.9)	88.7 (10.9)	92.8 (9.5)
Physical DQ	62.8 (23.3)	54.2 (17.5)	60.4 (17.4)	48.8 (12.9)	66.8 (32.4)	63.2 (21.5)
Self-Help DQ	73.3 (25.2)	75.2 (15.5)	71.0 (25.9)	70.9 (13.7)	77.0 (25.7)	82.3 (17.0)
Psychosocial/Behavior (n = 19)	n = 19	n = 19	n = 9	n = 9	n = 10	n = 10
Child Behavior Checklist						
Total Problems T-score	55.9 (9.9)	53.8 (13.8)	56.6 (8.4)	55.6 (16.0)	55.4 (11.5)	52.2 (12.1)

Values expressed as mean (SD).
IQ, Wechsler short form or Stanford-Binet; MDI, Bayley Mental Development Index; DQ, Developmental Quotient; SS, standard score.

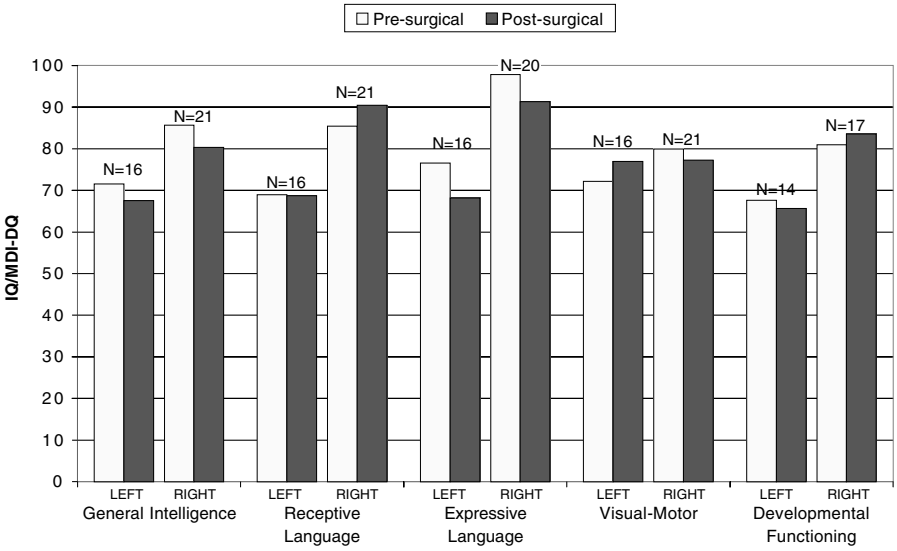

FIG. 1. Cognitive test results by side for the Rasmussen group. Number of subjects in each category is given above the bars.

this subgroup had on average a later age at seizure onset, were older at surgery, had longer duration of seizures, and were assessed at a shorter interval after surgery than were those in the larger, cross-sectional group (Table 2). One-way ANOVA showed no significant difference by side of surgery in terms of age at seizure onset, age at surgery, duration of seizures, age at postoperative testing, or time interval between surgery and the most recent evaluation.

Results of cognitive tests for the cortical dysplasia group are shown in Table 9 and illustrated in Fig. 2 by side of surgery. For the cortical dysplasia patients, repeated-measures ANOVA revealed no significant effect of side of surgery on general intelligence (IQ), receptive language (PPVT), expressive language (EOWPVT), or visual-motor (VMI) scores, with both groups earning consistently low scores (the apparent dramatic improvement in right-side language scores represents the performance of only a single individual). No significant effect of side of surgery was found on overall adaptive/developmental functioning or on any of the five adaptive domains.

For the cortical dysplasia group as a whole, no significant effect of time of evaluation (presurgical/follow-up) was found on intelligence, receptive language, or visual-motor functioning. Repeated-measures ANOVA found a relation approaching statistical significance ($p = 0.05$) between time and overall adaptive/developmental scores, which were somewhat lower at follow-up. In particular, significant changes were seen at follow-up in self-help and academic skills. Clearly (see Fig. 2), the overall change is due to the large decline in adaptive skills in the right-hemispherectomy group. A significant side-of-surgery group by time interaction was found for overall adaptive/developmental functioning and for self-help skills; in these two areas, the left-hemispherectomy group improved slightly, whereas the right-hemispherectomy group sharply declined. No other interactions were found for the cognitive test scores.

TABLE 8. *Dysplasia etiology: longitudinal sample characteristics by side of surgery*

	Total n = 15	Left n = 10	Right n = 5
Age at seizure onset (yr)	1.0 (3.8)	1.3 (3.4)	0.7 (0.9)
Age at surgery (yr)	6.7 (6.2)	7.2 (5.5)	5.6 (7.9)
Duration of seizures (yr)	5.6 (5.8)	5.9 (5.0)	4.9 (7.7)
Age at follow-up evaluation (yr)	10.5 (6.4)	10.2 (5.9)	10.9 (8.0)
Interval, surgery to post-op evaluation (yr)	3.8 (2.4)	3.0 (2.4)	5.3 (1.5)

Values expressed as mean (SD).

Vascular etiology

Longitudinal data were available for all seven of the patients with vascular etiology. As shown in Table 10, the vascular group included two left-side and five right-side hemispherectomy patients. One-way ANOVA showed significant differences by side of surgery in terms of age at seizure onset, age at surgery, and age at follow-up testing. However, the small size of the left-side group makes it hard to generalize from such observations. No significant differences were found by side of surgery in duration of seizures or in time interval between surgery and follow-up evaluation.

Results of cognitive tests for the vascular group are shown in Table 11 and illustrated in Fig. 3 by side of surgery. Repeated-measures ANOVA revealed no significant effect of side of surgery on IQ scores and no significant effect of time of evaluation on IQ scores for the total vascular group. However, the small size of the vascular group makes such comparisons statistically suspect.

Overall cognitive change

Regardless of etiology group, most patients showed only moderate change in cognitive performance at

TABLE 9. *Dysplasia etiology: cognitive test results at preoperative and follow-up evaluation by side of surgery*

	Total		Left		Right	
	Pre-Op	Post-Op	Pre-Op	Post-Op	Pre-Op	Post-Op
General Intelligence (n = 15)	n = 15	n = 15	n = 10	n = 10	n = 5	n = 5
IQ/MDI/DQ	40.6 (18.4)	40.1 (21.4)	38.7 (18.5)	36.3 (20.0)	44.4 (19.7)	47.6 (24.4)
Receptive Language (n = 7)	n = 7	n = 7	n = 4	n = 4	n = 3	n = 3
Peabody Picture Vocabulary Test-R SS/DQ	43.0 (8.5)	51.7 (22.2)	40.0 (2.0)	45.5 (7.9)	47.0 (13.0)	60.0 (34.6)
Expressive Language (n = 5)	n = 5	n = 5	n = 4	n = 4	n = 1	n = 1
Expressive One-Word Picture Vocabulary Test SS/DQ	55.8 (3.5)	57.2 (25.1)	56.0 (4.0)	50.2 (22.8)	55 (0)	85 (0)
Visual-Motor (n = 5)	n = 5	n = 5	n = 4	n = 4	n = 1	n = 1
Developmental Test of Visual-Motor Integration SS/DQ	66.8 (9.6)	69.8 (11.2)	69.8 (7.8)	69.3 (12.8)	79 (0)	72 (0)
Developmental Functioning (n = 10)	n = 10	n = 10	n = 7	n = 7	n = 3	n = 3
Developmental Profile-II						
Total DQ	59.7 (25.4)	53.3 (19.5)	51.1 (16.7)	51.9 (14.3)	79.6 (34.7)	56.5 (32.9)
Communication DQ	56.9 (26.8)	52.8 (22.4)	48.1 (15.0)	48.1 (14.6)	77.3 (40.7)	63.7 (36.8)
Socialization DQ	58.3 (30.4)	57.7 (20.3)	50.4 (15.3)	56.9 (18.3)	76.6 (52.2)	59.7 (29.1)
Academic DQ	69.9 (26.0)	54.3 (23.1)	57.6 (20.9)	51.3 (17.5)	85.3 (30.7)	61.3 (36.9)
Physical DQ	54.6 (26.6)	47.6 (20.0)	49.1 (25.5)	47.4 (13.8)	67.3 (29.8)	48.0 (35.0)
Self-Help DQ	63.1 (28.6)	54.1 (18.2)	50.1 (19.6)	55.9 (15.3)	93.3 (23.7)	50.0 (27.5)
Psychosocial/Behavior (n = 6)	n = 6	n = 6	n = 6	n = 6	n = 0	n = 6
Child Behavior Checklist						
Total Problems T-score	54.5 (9.3)	45.8 (9.1)	54.5 (9.3)	45.8 (9.1)	—	—

Values expressed as mean (SD).
IQ, Wechsler short form or Stanford-Binet; MDI, Bayley Mental Development Index; DQ, Developmental Quotient; SS, standard score.

follow-up. As shown in Fig. 4, a change in IQ of magnitude <15 points was the most common outcome. A substantial decline in IQ was most likely in the initially high-functioning Rasmussen group, whereas such a decline was not seen in the vascular group. However, the group differences were not statistically significant.

One-way ANOVA indicated that postsurgical persistence and severity of seizures was significantly related to follow-up IQ for the dysplasia group only (no such comparison was possible for the vascular group, all of whose members were seizure free at follow-up). The mean follow-up IQ for the seven dysplasia patients in the longitudinal group with continuing seizures, either mild or severe, was 29.3, compared with a mean IQ of 49.5 for the eight seizure-free dysplasia patients. However, persistence of seizures was not related to change in IQ for any etiology group or for the total sample.

Overall psychosocial and behavioral functioning for the cross-sectional group before surgery and at follow-up is indicated by the CBCL scores given by etiology in Tables 7, 9, and 11. Behavior scores were not significantly different by etiology or side of surgery, and 2 × 2 repeated-measures ANOVA found no interaction between etiology or side and change in score at follow-up. Overall, no significant behavioral or emotional problems were seen either before surgery or at follow-up. However, elevated mean scores indicated possible problems before surgery in two of the CBCL subscales: attention (T-score, 65.3 ± 9.5;

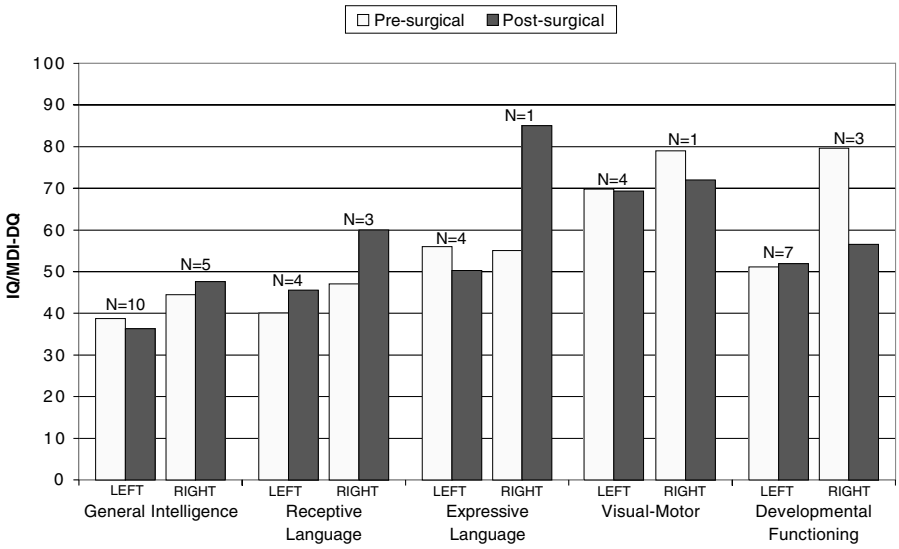

FIG. 2. Cognitive test results by side in the Dysplasia group. Number of subjects in each category is given above the bars.

TABLE 10. *Vascular etiology: longitudinal sample characteristics by side of surgery*

	Total n = 7	Left n = 2	Right n = 5
Age at seizure onset (yr)	2.4 (2.6)	5.5 (2.1)	1.2 (1.6)
Age at surgery (yr)	6.8 (5.9)	14.8 (4.8)	3.6 (1.5)
Duration of seizures (yr)	4.4 (4.5)	9.3 (6.9)	2.4 (0.9)
Age at follow-up evaluation (yr)	10.9 (9.3)	21.8 (13.2)	6.6 (2.0)
Interval, surgery to post-op evaluation (yr)	4.1 (4.5)	7.0 (8.3)	3.0 (2.9)

Values expressed as mean (SD).

n = 15) and thought problems (T-score, 60.9 ± 6.7; n = 15). Scores in these areas improved significantly subsequent to surgery (paired *t* test).

DISCUSSION

Epilepsy of sufficient severity to prompt consideration of hemispherectomy has devastating consequences for a child's functioning and development. The problems associated with such severe and frequent seizures extend far beyond the disruption caused by the seizures themselves. Children with severe epilepsy often have physical disability that can include hemiparesis (8,16), and their cognitive and adaptive development is usually substantially below age norms (11), with mean presurgical IQ in large hemispherectomy studies typically in the 50–69 range (4–7). Developmental and behavioral problems associated with severe, intractable epilepsy can pose a major barrier to independent living when the children reach adulthood. These kinds of preexisting physical and cognitive impairments have been reported in previous studies from Johns Hopkins that include subsets of the present sample, and they are important factors in the difficult decision of whether to embark on this radical surgery (2,17).

Given this background, the outcome of hemispherectomy documented here is remarkably good. Roughly two thirds of all subjects are seizure free, and nearly half are medication free. This outcome compares favorably with other studies, most recently that of Devlin et al. (10), in which overall, 17 (52%) of 33 subjects were seizure free, and 16 (48%) of 33 were no longer taking AEDs at follow-up. It appears that, for most children, surgery has not imposed a significant additional burden and, in their judgment, has actually improved their quality of life. The families (as well as the children, if old enough) are satisfied with the surgery. At follow-up, on average about 5 years after surgery, intelligence, language, visual-motor, and adaptive/developmental skills for most children were impaired, but abilities in these areas were generally no worse than before surgery and were consistent with those that have been reported elsewhere for children with severe intractable epilepsy. These results are particularly notable for the patients with Rasmussen encephalopathy, because they indicate a halt in the precipitate cognitive de-cline associated with that disorder (17).

The nature of the brain disorder giving rise to the seizures appeared to affect outcome far more than the procedure itself. Patients with Rasmussen encephalopathy, cortical dysplasias, and vascular problems differed significantly in terms of subject characteristics before surgery and test performance after surgery. Children with Rasmussen encephalopathy averaged age 6 years at seizure onset and 9 years at the time of surgery. At

TABLE 11. *Vascular etiology: cognitive test results at preoperative and follow-up evaluation by side of surgery*

	Total		Left		Right	
	Pre-Op	Post-Op	Pre-Op	Post-Op	Pre-Op	Post-Op
General Intelligence (n = 7)	n = 7	n = 7	n = 2	n = 2	n = 5	n = 5
IQ/MDI/DQ	72.7 (11.1)	78.7 (11.1)	80.0 (18.4)	93.5 (20.5)	69.8 (7.9)	72.2 (11.1)
Receptive Language (n = 3)	n = 3	n = 3	n = 1	n = 1	n = 2	n = 2
Peabody Picture Vocabulary Test-R SS/DQ	92.3 (15.9)	76.7 (15.0)	103 (0)	94 (0)	87.0 (18.4)	68.0 (1.4)
Expressive Language (n = 3)	n = 3	n = 3	n = 1	n = 1	n = 2	n = 2
Expressive One-Word Picture Vocabulary Test SS/DQ	90.3 (8.6)	105.0 (5.6)	92 (0)	104 (0)	89.5 (12.0)	105.5 (7.8)
Visual-Motor (n = 3)	n = 3	n = 3	n = 1	n = 1	n = 2	n = 2
Developmental Test of Visual-Motor Integration SS/DQ	79.3 (11.8)	76.7 (3.1)	73 (0)	76 (0)	82.5 (14.8)	77.0 (4.2)
Developmental Functioning (n = 5)	n = 5	n = 5	n = 1	n = 1	n = 4	n = 4
Developmental Profile-II						
Total DQ	73.7 (14.2)	75.6 (9.3)	72 (0)	82 (0)	74.1 (16.4)	74.0 (9.9)
Communication DQ	76.8 (13.7)	75.2 (7.0)	77 (0)	85 (0)	76.8 (15.9)	72.8 (5.0)
Socialization DQ	72.6 (24.1)	75.6 (17.8)	75 (0)	85 (0)	72.0 (27.7)	73.3 (19.7)
Academic DQ	78.6 (13.4)	72.2 (14.3)	79 (0)	85 (0)	78.5 (15.5)	69.0 (14.3)
Physical DQ	71.0 (10.8)	74.4 (14.8)	66 (0)	70 (0)	72.2 (12.0)	75.5 (16.8)
Self-Help DQ	69.4 (18.7)	80.6 (9.1)	64 (0)	85 (0)	75.8 (21.4)	79.5 (10.1)
Psychosocial/Behavior (n = 4)	n = 4	n = 4	n = 1	n = 1	n = 3	n = 3
Child Behavior Checklist						
Total Problems T-score	60.0 (8.9)	54.3 (17.7)	61 (0)	46 (0)	59.7 (9.0)	57.0 (21.0)

Values expressed as mean (SD).
IQ, Wechsler short form or Stanford-Binet; MDI, Bayley Mental Development Index; DQ, Developmental Quotient; SS, standard score.

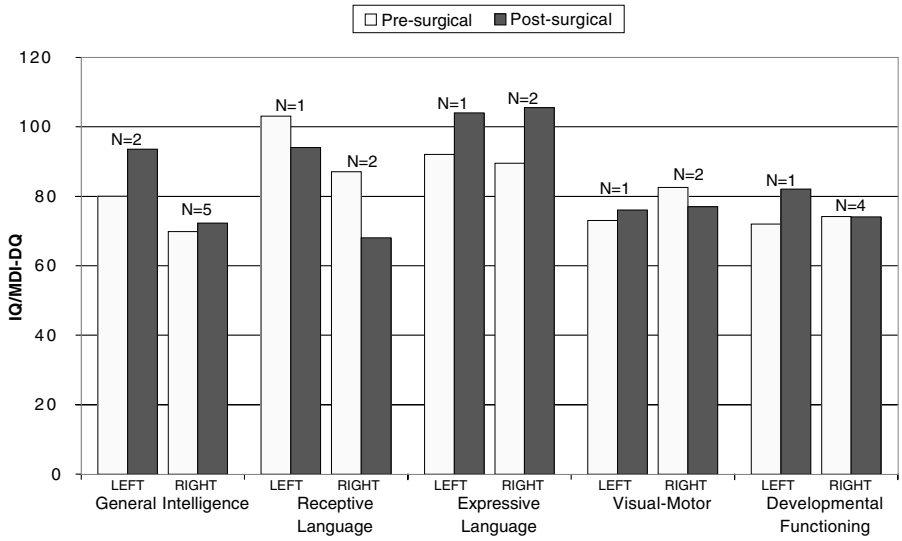

FIG. 3. Cognitive test results by side for the Vascular group. Number of subjects in each category is given above the bars.

follow-up, most had no seizures or only nonhandicapping seizures, which did not significantly affect cognitive outcome. The children were in good health and were mainstreamed in school with or without support. They had IQs in the 70s and were nonaphasic, although IQ and receptive and expressive language skills were significantly worse in those for whom the lesion was in the left hemisphere.

Although the number of children in this series with cerebrovascular disorders and difficult-to-control seizures is too small to make definitive statements, they appear to do as well as those with Rasmussen syndrome, and better on tests of gross and fine motor skills. The apparent lack of physical impairment in the vascular group could be related to either their early age at onset (with greater age-related plasticity), or to their relatively young age at follow-up (less complex physical abilities are assessed).

Children with large unilateral dysplasias such as hemimegalencephaly had the least favorable outcome. For these children, brain dysgenesis has occurred prenatally, and seizure onset was almost always within the first year. Hemispherectomy was typically performed at ages 4 to 5 years. These children are more likely to have continuing seizures and general health problems, and to require full-day special education. IQ and language skills, especially receptive language, were much lower in the dysplasia group, although no significant dependence on side of lesion was seen. These children were also the most delayed in all adaptive/developmental abilities measured except in the physical domain. For the dysplasia group, continuing seizures after hemispherectomy were associated with lower IQ at follow-up but not with a greater or lesser change in IQ subsequent to surgery. This suggests that extremely low cognitive functioning is an indicator of generalized brain dysfunction with a reduced likelihood of seizure elimination, but is not necessarily an indicator of poorer cognitive outcome for hemispherectomy. The

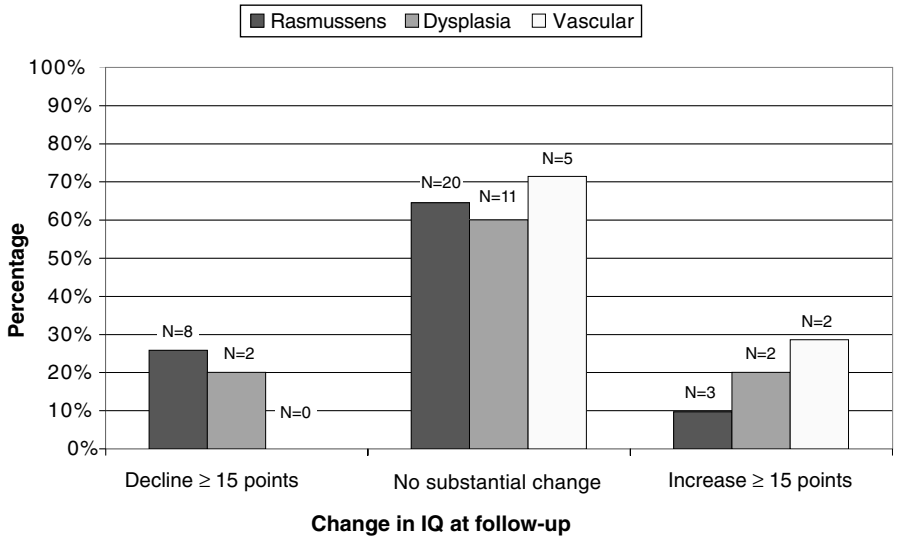

FIG. 4. Longitudinal group, IQ change.

particular problems of children in the dysplasia group have been increasingly recognized (9,10) and could reflect organic impairment of the remaining hemisphere as well as cumulative developmental delay from the very-early-onset severe seizure disorder. Despite their less favorable outcome, children with cortical dysplasias and their families expressed comparable satisfaction with surgery (only 4% were dissatisfied) and rated their relative quality of life after surgery equally high as did those with other etiologies.

Only for Rasmussen etiology was the side of the brain lesion significantly related to cognitive abilities. Rasmussen patients with left-side lesions obtained significantly lower scores on IQ and language tests (both receptive and expressive) and displayed lower academic and communication skills than did those with right-side lesions. Interestingly, these differences by side of injury were observed before surgery as well as at follow-up and so are most likely a consequence of the disease pathology. The only significant change in cognitive abilities observed subsequent to surgery was a roughly 7-point decline in the expressive language standard score for both left- and right-side Rasmussen patients. The particular impairment of language functioning in left-hemispherectomy patients is consistent with models of brain lateralization, at least in adults, but the absence of relative impairment in visual-spatial skills in right-hemispherectomy patients is not consistent with those models. The lack of obvious lateralization in the cortical dysplasia patients may reflect a greater brain plasticity at the very early age at seizure onset and at surgery characteristic of this group. It may also be related to the low overall level of functioning in these individuals. Cortical dysplasia patients with right-side lesions did show a decline at follow-up in adaptive/developmental functioning, particularly self-help and academic skills, whereas left-side patients showed essentially no change at follow-up. In terms of patient clinical characteristics, no significant differences were noted between left-side and right-side hemispherectomy patients in the study within any etiology group.

These results for cognitive functioning have similarities to those of earlier studies. The present study's mean follow-up IQ of 59.7 for the 71 patients is comparable to those of the earliest cognitive studies (4,5) and consistent with those of later ones that also recognize the importance of controlling for etiology. In those later studies, IQ results similar to those presented here have been obtained for developmental and vascular etiologies (9,10). Although initial reports credited hemispherectomy with often-dramatic improvements in IQ, the most common outcome is that of the present study: for most patients a modest change in IQ, <15 points, and large improvements or reductions in IQ for a few.

In terms of behavior, the present study did not find the near-universal and extreme disturbances reported by some earlier studies (1,5,7,10–12), although deficits were seen in social competence, possibly reflecting the socially isolating effects of frequent seizures, physical disability, and intense medical treatment. Some evidence before surgery was found of mild attention problems, which improved at follow-up. The contrast between the present results and earlier reports is notable—for example, Basser (11) cited "serious outbursts of ill temper" as a principal reason for performing the surgery in patients with severe hemiplegia, and Lindsay (15) found "gross behavior disorder," including "murderous aggression," to be "a major clinical handicap." In the present study, neither such a prevalence nor such an intensity of behavior problems were seen.

Adaptive functioning is generally low in children with severe epilepsy. Acquisition of self-help skills is slowed both by the disruptions from frequent seizures and by general cognitive and physical impairments. The presence of shortcomings in adaptive functioning is one reason that children and adults with severe epilepsy are most often found in supported-living settings. Although we found adaptive functioning to be generally consistent with IQ, both the higher-IQ Rasmussen group and the lower-IQ dysplasia group had particular deficits in the physical domain. These deficits, which relate to gross and fine motor skills applicable to everyday life, were found even when basic visual-motor abilities were relatively spared (as they were for the dysplasia group). The deficits existed before surgery and had, on average, neither improved nor worsened at follow-up, but such physical difficulties as the inability to ride a bicycle or to use scissors easily were cited by patients at follow-up as the most important impediments to their quality of life. Early intervention to develop gross and fine motor skills should be an important part of postsurgical treatment.

One important limitation of this study was the difficulty of reliably assessing extremely low-functioning individuals, where DQs were sometimes used in lieu of standard scores and, in some cases, certain tests could not be given because of the low level of functioning. The exclusion of low-functioning individuals tends to increase mean scores artificially; this was not a problem for intelligence scores, which were available for all patients, but caution should be used in comparing mean intelligence scores with those in other areas such as language, for which smaller numbers of patients were available.

More study is needed of language outcome, particularly the differing relations of expressive and receptive language to factors like presurgical functioning and age at surgery. Furthermore, a more detailed look at complex language than the single-word measures used in this study would be very helpful to gain more insight into hemispheric specialization of language functions. Better understanding of development after hemispherectomy will be very important in improving outcome and guiding postsurgical treatment.

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REFERENCES

1. Krynauw RA. Infantile hemiplegia treated by removing one cerebral hemisphere. *Dev Med Child Neurol* 1950;28:251–8.
2. Vining EPG, Freeman JM, Pillas DJ, et al. Why would you remove half a brain? The outcome of 58 children after hemispherectomy: the Johns Hopkins Experience: 1968–1996. *Pediatrics* 1997;100:163–71.
3. Lassonde M, Sauerwein HC, Jambaque I, et al. Neuropsychology of childhood epilepsy: pre- and post-surgical assessment. *Epileptic Disord* 2000;2:3–13.
4. McFie J. The effects of hemispherectomy on intellectual functioning in cases of infantile hemiplegia. *J Neurol Neurosurg Psychiatry* 1961;24:240–9.
5. Lindsay J, Ounsted C, Richards P. Hemispherectomy for childhood epilepsy: a 36-year study. *Dev Med Child Neurol* 1987;29:592–600.
6. Tinuper P, Andermann F, Villemure J-G, et al. Functional hemispherectomy for treatment of epilepsy associated with hemiplegia: rationale, indications, results, and comparison with callosotomy. *Ann Neurol* 1988;24:27–34.
7. Verity CM, Strauss EH, Moyes PD, et al. Long-term follow-up after cerebral hemispherectomy: neurophysiologic, radiologic, and psychologic findings. *Neurology* 1982;32:629–39.
8. Brandt J, Vining EPG, Stark RE, et al. Hemispherectomy for intractable epilepsy in childhood: preliminary report on neuropsychological and psychosocial sequelae. *J Epilepsy* 1990;3(suppl):261–70.
9. Wyllie E, Comair YG, Kotagal P, et al. Seizure outcome after epilepsy surgery in children and adolescents. *Ann Neurol* 1998;44:740–8.
10. Devlin AM, Cross JH, Harkness W, et al. Clinical outcomes of hemispherectomy for epilepsy in childhood and adolescence. *Brain* 2003;126:556–66.
11. Basser LS. Hemiplegia of early onset and the faculty of speech with special reference to the effects of hemispherectomy. *Brain* 1962;85:427–60.
12. Beardsworth ED, Adams CBT. Modified hemispherectomy for epilepsy: early results in 10 cases. *Br J Neurosurg* 1988;2:73–84.
13. Stark RE, Bleile K, Brandt, J, et al. Speech-language outcomes of hemispherectomy in children and young adults. *Brain Lang* 1995;51:406–21.
14. de Bode S, Curtiss S. Language after hemispherectomy. *Brain Cogn* 2000;43:135–205.
15. Lindsay J, Glaser G, Richards P, et al. Developmental aspects of focal epilepsies of childhood treated by neurosurgery. *Dev Med Child Neurol* 1984;26:574–87.
16. Vining EPG, Freeman J, Carson B, et al. Hemispherectomy in children: the Hopkins experience (1968–1988): a preliminary report. *J Epilepsy* 1990;3(suppl):169–76.
17. Vining EPG, Freeman JM, Brandt J, et al. Progressive unilateral encephalopathy of childhood (Rasmussen's syndrome): a reappraisal. *Epilepsia* 1993;34:639–50.
18. Carson BS, Javedan SP, Freeman JM, et al. Hemispherectomy: a hemidecortication approach and review of 52 cases. *J Neurosurg* 1996;84:903–11.
19. Freeman JM, Vining EP, Pillas DJ, et al. Seizure outcome after hemispherectomy: the Johns Hopkins experience. In: Tuxhorn I, Holthausen H, Boenig KH, et al., eds. *Pediatric epilepsy syndromes and their surgical treatment*. London: John Libbey, 1997:743–8.
20. Engel J Jr, Van Ness PC, Rasmussen TB, et al. Outcome with respect to epileptic seizures. In: Engel J Jr, ed. *Surgical treatment of the epilepsies*. 2nd ed. New York: Raven Press; 1993:609–21.
21. Wong DL, Baker CM. Pain in children: comparison of assessment scales. *Pediatr Nurs* 1998;14:9–17.
22. Luffy R, Grove SK. Examining the validity, reliability, and preference of three pediatric pain measurement tools in African-American children. *Pediatr Nurs* 2003;29:54–9.
23. Bayley N. *Bayley scales of infant development*. New York: Psychological Corporation, 1969.
24. Thorndike RL, Hagen EP, Sattler JM. *The Stanford-Binet Intelligence Scale*. 4th ed. *Guide for administering and scoring the fourth edition*. Chicago: Riverside Publishing, 1986.
25. Wechsler D. *Wechsler Preschool and Primary Scale of Intelligence-Revised*. New York: Psychological Corporation, 1989.
26. Kaufman AS. A short form of the Wechsler Preschool and Primary Scale of Intelligence. *J Consult Clin Psychol* 1972;39:361–9.
27. Wechsler D. *Wechsler Intelligence Scale for Children-Revised*. New York: Psychological Corporation, 1974.
28. Kaufman AS. A four-test short form of the WISC-R. *Contemp Educ Psychol* 1976;1:180–96.
29. Wechsler D. *Wechsler Adult Intelligence Test-Revised*. New York: Psychological Corporation, 1981.
30. Silverstein AB. Two- and four-subtest short forms of the Wechsler Adult Intelligence Scale–Revised. *J Consult Clin Psychol* 1982;50:415–8.
31. Dunn L, Dunn M. *Peabody Picture Vocabulary Test-Revised*. Circle Pines, MN: American Guidance Service, 1981.
32. Gardner MF. *Expressive One Word Vocabulary Test Manual*. Novato, CA: Academic Therapy Publications, 1979.
33. Beery KE. *The VMI Developmental Test of Visual-Motor Integration*. Cleveland, OH: Modern Curriculum Press, 1989.
34. Alpern G, Boll T, Shearer M. *Developmental Profile II, manual*. Los Angeles: Western Psychological Services, 1992.
35. Achenbach TM. *Manual for the Child Behavior Checklist/4–18 and 1991 profile*. Burlington, VT: Department of Psychiatry, University of Vermont, 1991.
36. Achenbach TM. *Manual for the Child Behavior Checklist/2–3 and 1992 profile*. Burlington, *VT*: Department of Psychiatry, University of Vermont, 1992.