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Hemispheric Surgery in Children with Refractory Epilepsy: Seizure Outcome, Complications, and Adaptive Function

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Summary: *Purpose:* To describe seizure control, complications, adaptive function and language skills following hemispheric surgery for epilepsy.

Methods: Retrospective chart review of patients who underwent hemispheric surgery from July 1993 to June 2004 with a minimum follow-up of 12 months.

Results: The study population comprised 24 children, median age at seizure onset six months and median age at surgery 41 months. Etiology included malformations of cortical development (7), infarction (7), Sturge-Weber Syndrome (6), and Rasmussen's encephalitis (4). The most frequent complication was intraoperative bleeding (17 transfused). Age <2 yr, weight <11 kg, and hemidecortication were risk factors for transfusion. Postoperative complications included aseptic meningitis (6), and hydrocephalus (3). At median follow-up of 7 yr, 79% of patients are seizure free. Children with malformations of cortical development and Rasmussen's encephalitis were more likely to have ongoing seizures. Overall adaptive function scores were low, but relative strengths in verbal abilities were observed. Shorter duration of epilepsy prior to surgery was related significantly to better adaptive functioning.

Conclusions: Hemispheric surgery is an effective therapy for refractory epilepsy in children. The most common complication was bleeding. Duration of epilepsy prior to surgery is an important factor in determining adaptive outcome. **Key Words:** Hemispheric disease—Refractory epilepsy—Peri-insular hemispherotomy—Hemidecortication—Postoperative complications—Blood loss—Seizure outcome—Adaptive function—SIB-R—PPVT-III.

Hemispheric surgery is an established treatment for medically refractory epilepsy due to diffuse hemispheric disease. Krynauw (1950) reported the first series of 12 patients with infantile hemiplegia who underwent anatomic hemispherectomy for hemispheric disease and epilepsy. In 1961, White, in a review of the literature on hemispherectomy for infantile hemiplegia, reported 6.6% postoperative mortality in a total of 267 patients. In the 1960s, concerns about late complications of anatomic hemispherectomy were raised. Oppenheimer and Griffith (1966) described three of 17 patients with recurrent episodes of encephalopathy, obstructive hydrocephalus and superficial cerebral hemosiderosis 3–11 yr following hemispherectomy leading to death. Rasmussen (1983) also described intracranial hypertension due to hydrocephalus in 11 of 31 patients, 4.5–24 yr following surgery. He attributed this to the extent of resection of the hemisphere and its coverings. To reduce complications, changes in surgical technique have evolved to include subtotal resection, functional hemispherectomy, and hemidecortication (Carson et al., 1996).

Villemure and Mascott in 1995 described 'peri-insular hemispherotomy' which, in their hands, resulted in shorter operative times and fewer early postoperative complications and bleeding. Seizure control was comparable to anatomic hemispherectomy but morbidity and mortality rates were lower (Villemure and Mascott, 1995; Kestle et al., 2000). There have been numerous reports on the outcomes following hemispheric surgery in children with medically refractory epilepsy (Carson et al., 1996; Vining et al., 1997; Battaglia et al., 1999; Boatman et al., 1999; Carreno et al., 2001; Kossoff et al., 2002; Devlin et al., 2003; Daniel and Villemure, 2003; Kossoff et al., 2003; van Empelen et al., 2004; Jonas et al., 2004; Piastra et al., 2004; Pulsifer et al., 2004; Gonzalez-Martinez et al.,

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2005; Shimizu, 2005). These studies have examined either aspects of cognitive, behavioral and motor outcomes, seizure control or postoperative complications in terms of etiology and outcome predictors. However, there is limited published data reporting both medical and surgical morbidity in a continuous series, which highlights all risk factors and details outcomes.

The objective of this study is to report on seizure control, morbidity, language skills and adaptive functions in a consecutive series of children who underwent hemispheric surgery at our center over an 11 year period and to identify specific risk factors affecting surgical, medical and functional outcomes.

METHODS

Patient population

The medical records and operative notes of children who underwent hemispheric surgery for medically refractory epilepsy between July 1993 and June 2004 at British Columbia Children's Hospital and who had a minimum follow-up of 1 yr were reviewed. Patient characteristics, seizure types, etiology, early and late postoperative complications, seizure control, motor function, and adaptive and cognitive function were recorded.

The presurgical evaluation comprised high resolution 1.5 Tesla magnetic resonance imaging, interictal and video-electroencephalographic telemetry and, where appropriate, neuropsychological testing. Interictal and ictal technetium-99 m-labeled hexamethyl propylene-amine-oxime single-photon emission computed tomography and interictal 18-fluoro-2-deoxyglucose positron emission computed tomography were used in select patients.

After surgery, all patients were followed at 3, 6, and 12 months, yearly for 5 yr and every 2 yr from then on if stable. Patients were seen more frequently if problems occurred. No patients were lost to follow-up.

Outcome measures

The major outcomes assessed were seizure control (based on patient and family reporting) classified according to Engel's classification (Engel et al., 1993), volume of blood products transfused and adaptive function. In this paper, we report long-term outcome as observed at most recent follow-up visit.

Blood product transfusions were recorded as milliliters per kilogram (ml/kg). If the recorded values were documented as units, the mean volume (ml) per unit was calculated. The mean volume per unit of packed red cells depended on the type of anticoagulant used. Prior to 1998, citrate phosphate dextrose adenine was used, with an estimated volume of 250 ml per unit of red cells and subsequently Nutricel has been used with an estimated volume of 290 ml per unit. The mean volumes per unit of cryoprecipitate, platelet, and plasma infusions were 10, 50, and 175 ml, respectively.

Level of adaptive functioning was assessed with the Scales of Independent Behavior-Revised (Bruininks et al., 1997) (SIB-R), a parent-report rating scale assessing function in four domains: motor skills, social interaction and communication skills, personal living skills, and community living skills. The overall Broad Independence score was used in this study, in standard score format (mean of 100, standard deviation of 15), with higher scores indicating better functional independence. Reliability and validity for this scale have been reported as adequate to high (Sattler, 2002). Tests of cognitive function included the Peabody Picture Vocabulary Test — Third Edition (Dunn and Dunn, 1997) (PPVT-III), a measure of receptive vocabulary, and the Beery Developmental Test of Visual Motor Integration (Beery and Beery, 2004) (VMI).

Data analysis

Outcomes were assessed in terms of underlying pathology (malformations of brain development versus acquired lesions), sex, weight at time of surgery, age at seizure onset, history of infantile spasms, duration of epilepsy prior to surgery, side of surgery, and age at surgery. Preoperative formal IQ testing was not possible in all patients due to young age at the time of surgery or low level of function. Similarly, preoperative and serial postoperative adaptive and cognitive assessments were not available in a sufficient number of cases to conduct meaningful analyses. In those cases with multiple postoperative adaptive and/or cognitive assessments the most recent test scores were included in the analyses.

For four patients, the SIB-R computer scoring program was unable to yield one or more subscale standard scores owing to extremely low raw scores for those subscales. To avoid the loss of these patients' data, standard scores of 19 (i.e., well below the first percentile) were assigned as it was felt that this was sufficiently low to reflect their level of functional limitations.

Approval from the University of British Columbia Clinical Research Ethics Board and Children's & Women's Health Centre of British Columbia Research Review Committee was obtained.

RESULTS

Preoperative details

Twenty-four children, 15 girls and nine boys, underwent hemispheric surgery for refractory epilepsy. Table 1 summarizes patient characteristics. The first five children had hemidecortication which involved temporal lobectomy and removal of the cortex over the remainder of the hemisphere and the remainder periinsular hemispherotomy as described by Villemure and Mascott (1995). Several patients were described in a previous report (Kestle et al., 2000). Seizures began at a median age of 6 months (range 1 day to 7 year). Seventeen children had seizure onset at less than 1 year of age, seven in the neonatal

TABLE 1. *Patient demographics*

Etiology (n)	CD (4)	HME (3)	I (7)	RE (4)	SWS (6)
Mean (± SD) weight at surgery (kg)	29 (± 22)	20 (± 9)	27 (± 19)	33 (± 17)	10 (± 2)
Mean (± SD) age at surgery (months)	75 (± 72)	54 (± 43)	86 (± 61)	97 (± 54)	19 (± 6)
Left-sided surgery	1	1	6	2	3
Surgery type					
Periinsular hemipsherotomy	4	3	7	1	4
Hemidecortication	0	0	0	3	2

SD, standard deviation; CD, cortical dysplasia; HME, hemimegalencephaly; I, infarction; RE, Rasmussen's encephalitis; SWS, Sturge-Weber syndrome.

period. Nine children had infantile spasms, 13 had a history of status epilepticus (recurrent in eight), four had epilepsia partialis continua and 16 had secondarily generalized seizures. Seizures were refractory to a median of seven antiepileptic drugs (range 2–14).

All patients had hemiparesis and only two had useful hand function prior to surgery. Homonomous hemianopia could be demonstrated in 19 children before surgery. In five children no visual field abnormalities could be identified, although their young age made clinical assessment less reliable. Two patients had ventriculo-peritoneal (VP) shunts placed in the neonatal period for posthemorrhagic hydrocephalus. Twenty patients had strictly unilateral MR abnormalities and four patients had bilateral MRI abnormalities, often subtle signal change or volume loss and three had cerebral infarcts, two had perinatal encephalopathy attributed to a hypoxic ischemic insult, one had herpes encephalitis and the fourth patient had Rasmussen's encephalitis. Contralateral interictal EEG abnormalities were observed in 14 patients, slowing in 12 and epileptiform discharges in 11. A seizure was recorded from the contralateral hemisphere in one patient who had Rasmussen's encephalitis and bilateral MRI abnormalities.

Patients with Rasmussen's encephalitis underwent focal cortical resections (frontal, frontotemporal and temporo occipital resections) in order to confirm the diagnosis before proceeding to hemispheric surgery. Focal frontal resections were performed prior to hemispheric surgery in two children with malformations of cortical development, which were initially felt to involve primarily the frontal lobe, but were unsuccessful in controlling seizures.

Perioperative details

The median age at the time of surgery was 41 months (range 3 months to 14.8 year). Three children were less than one year at surgery and two children weighing less than 10 kg had surgery planned in two stages over three months to reduce morbidity. Seven patients weighed less than 11 kg at the time of surgery. Surgery was performed in 13 who were younger than 4 year of age, eight of whom were under 2 year. The duration of epilepsy prior to surgery was 2 year or less in 12 of the patients. Four patients who underwent left hemispheric operations were 4 yr of age or older at the time. In one patient, hemispheric surgery was performed urgently because of refractory status epilepticus. The median length of hospital stay was 9 days (range 6–66 days). Table 2 describes blood product usage according to etiology, weight and age at the time of surgery and type of surgery. Intraoperative bleeding resulting in hypotension occurred in three patients. One patient had a 15 min cardiac arrest intraoperatively due to volume depletion. The mean volume of red cells transfused was 33 ml/kg (range 5–88 ml/kg; standard deviation 20 ml/kg); total blood products transfused (packed red cells, fresh frozen plasma, cryoprecipitate and platelets) was 39 ml/kg (range 5–109 ml/kg; standard deviation 28 ml/kg). Autologous blood collected and stored preoperatively was used in three patients. One patient received recombinant human erythropoietin (rhEPO) preoperatively and did not receive blood products. Patients with an ischemic/hemorrhagic cerebral infarction were the least likely to be transfused. Patients who underwent hemidecortication required a greater volume of blood product transfusion than the periinsular hemispherotomy group.

Early postoperative seizures occurred in six patients. One patient (aged 18 months at the time of surgery) developed symptomatic seizures secondary to hyponatremia

TABLE 2. *Operative and postoperative blood product usage*

	No. transfused	Mean volume transfused (ml/kg)	No. requiring > 40 ml/kg
Etiology (n)			
Cortical dysplasia (4)	3	47	-
Hemimegalencephaly (3)	2	47	1
Infarction (7)	3	14	-
Rasmussen's encephalitis (4)	3	56	2
Sturge-Weber syndrome (6)	6	51	3
Patient weight (n)			
<11kg (6)	6	52	3
≥11kg (18)	11	32	3
Patient age (n)			
≤2 yr (8)	7	48	3
> 2 yr (16)	10	33	3
Surgery type (n)			
Hemidecortication (5)	4	65	4
Periinsular hemispherotomy (19)	12	28	2

due to the syndrome of inappropriate secretion of antidiuretic hormone (SIADH). Following hemispheric surgery, VP shunts were placed in two patients and in one patient a preexisting shunt was revised. Aseptic meningitis occurred in five patients. Extraxial fluid collections were observed in two patients but did not require surgical intervention. Transient sixth cranial nerve paresis occurred in one child. One patient developed an infected central venous catheter and another had a decubitus ulcer. Reoperation for ongoing seizures was performed in two patients with Rasmussen's encephalitis, a right frontal resection two months after right hemidecortication, and, a right frontal and insular cortical resection 5 months after right periinsular hemispherotomy and both are seizure free.

Long-term follow-up

At a median follow-up of 7 yr (mean 6.6 yr, range 1–12.25 yr), there has been no mortality. As expected, all have hemiparesis, with no useful hand function (four patients have worsening of their preoperative hemiparesis and homonymous hemianopia). However, all patients are ambulatory and have useful proximal upper limb function despite loss of fine finger function. Recurrent severe headaches occurred in five patients, four of whom had migraine. The fifth patient, who had Rasmussen's encephalitis, developed intracranial hypertension without hydrocephalus, 71 months following hemispherotomy and responded to acetazolamide. The children with migraine had frequent severe headaches and all required prophylaxis. Glaucoma requiring intervention occurred in three patients with Sturge-Weber syndrome.

Table 3 summarizes seizure outcome according to etiology. Complete seizure control occurred in 19 (79%), 18 of whom are no longer on antiepileptic drugs. Four are on monotherapy and one patient with Rasmussen's encephalitis is on two antiepileptic drugs. In four patients with bilateral MRI abnormalities, three remain on antiepileptic drugs (range 1–3), two are currently seizure-free but had seizures during medication reduction, one with Rasmussen's encephalitis has ongoing complex partial seizures. Preoperatively, she also had independent seizures recorded from both hemispheres. The fourth patient has been seizure-free since surgery. Twelve of 14 patients with preoperative slowing or epileptiform activity in the contralateral hemisphere are seizure-free at most recent follow-up.

TABLE 3. *Postoperative seizure outcome at most recent follow-up and etiology*

	Engel's classifiaction				Follow-up in years	
Pathology (n)	Ia	II	III	IVa	Median	Range
Cortical dysplasia (4)	3	1	–	–	7.50	7.00–9.50
Hemimegalencephaly (3)	2	1	–	–	7.00	2.50–10.0
Infarction (7)	6	1	–	–	3.00	1.75–10.25
Rasmussen's encephalitis (4)	2	1	–	1	11.50	5.00–11.75
Sturge-Weber syndrome (6)	6	–	–	–	9.50	1.75–12.25
Surgery type (n)						
Hemidecortication (5)	3	1	–	1	11.75	10.75–12.25
Periinsular hemispherotomy (19)	16	3	–	–	4.50	1.00–10.25

TABLE 4. *Descriptive statistics for adaptive and cognitive measures*

	Mean	Standard deviation	Median	Minimum	Maximum
SIB-R					
Broad independence	45.5	25.2	42.5	4	93
Motor	42.5	28.4	32.5	19	127
Social/communication	69.7	27.7	76	15	113
Personal living	52.9	23.5	54	6	94
Community living	55.8	27.2	63	11	94
Support	58.2	16.1	62	24	83
PPVT	74.6	17.5	77	46	100
VMI	63.4	14.9	63	45	88

PPVT, peabody picture vocabulary test; VMI, visual-motor integration.

The mean age at the most recent postoperative adaptive/cognitive assessment was 10.5 yr (standard deviation 5.0, range 2.5–20.3 yr). The median time to the most recent follow-up was 5.5 yr (mean 5.1 yr, range 0.3–10.3 yr). Descriptive statistics for postoperative scores on cognitive and adaptive measures are shown in Table 4. The mean SIB-R Broad Independence score was very low (45.5), indicating severe impairment in carrying out age-appropriate activities of daily living. However, there was a broad range of functioning; approximately 14% of the sample scoring within broadly normal limits ($\geq$ standard score of 80). From a normative standpoint, the participants scored highest on the PPVT-III (47% scored $\geq$ standard score of 80) and the Social/Communication scale (43% of the children scored $\geq$ standard score of 80) of the SIB-R; on these scales, mean scores were commensurate with borderline to mild impairment in language skills, while the SIB-R Motor scale was the lowest overall score (82% of the children scored $<$ 1st percentile), commensurate with severe impairment in functional motor skills.

Correlations were computed to determine risk factors for poor adaptive functioning outcome. Correlations between age of onset of epilepsy, age at surgery, duration of epilepsy, gender, weight at surgery, side of surgery, etiology, history of infantile spasm, and status epilepticus adaptive, and cognitive scores are presented in Table 5. Duration of epilepsy was significantly related to the SIB-R Broad Independence score and all four SIB-R scale scores (i.e., Motor, Social/Communication, Personal Living, and Community Living scores), with shorter duration associated with higher scores in each case. Similarly, age at surgery was significantly related to the SIB-R Broad Independence and the Social/Communication scale

TABLE 5. *Correlations between clinical variables and cognitive/adaptive measures. The values in the table indicate the correlation coefficients*

	Seizure onset	Age at surgery	Duration	Sex	Weight	Side of surgery	Etiology	Spasms	Status epilepticus
SIB-R									
Broad independence	.1	−.44*	−.51*	−.23	−.25	.10	−.37	−.34	.08
Motor	−.07	−.42	−.46*	.00	−.40	.16	−.56*	−.30	−.12
Social/communication	.15	−.43*	−.52*	−.35	−.15	.03	−.17	−.25	−.20
Personal living	.18	−.34	−.43*	−.25	−.16	−.08	−.34	−.46*	.12
Community living	.13	−.41	−.49*	−.27	−.15	.20	−.35	−.27	−.20
PPVT	.24	−.02	−.12	−.50*	.02	.09	−.52	−.12	.11
VMI	.31	.13	−.02	−.24	.20	−.07	−.53	−.30	.20

[a] $p < .05$.
PPVT, Peabody picture vocabulary test; VMI, visual-motor integration.

scores, with younger age associated with better functioning. Female gender was significantly related to higher PPVT-III scores. At follow-up, seven of the female patients had a PPVT-III score of 80 or higher and only one had a PPVT-III score below the 5th percentile. In contrast, only one male patient scored above the 3rd percentile. SIB-R Motor scale score was significantly related to etiology, with acquired pathologies associated with a better motor function. A history of infantile spasms was significantly related to lower scores on the SIB-R Personal Living scale.

DISCUSSION

Although this is a relatively small series of patients, no children were lost to follow-up. In previously reported series, there was either lack of or shorter follow-up periods of all patients (Devlin et al., 2003; Kossoff et al., 2003; van Empelen et al., 2004; Jonas et al., 2004). In this series, 83% of patients had onset of epilepsy in the first year of life, similar to other reports (Devlin et al., 2003; Jonas et al., 2004; Gonzalez-Martinez et al., 2005). Neonatal onset epilepsy and West syndrome were common, 42% of patients.

Complete seizure control in 79% compares favorably with published data. Krynauw (1950) reported complete seizure control in all but one patient, and 67% of patients were seizure free in White's (1961) review of anatomic hemispherectomy. Following functional hemispherectomy, seizure-free outcomes have ranged from 52 to 75% (Devlin et al., 2003; van Empelen et al., 2004; Jonas et al., 2004; Shimizu, 2005; Gonzalez-Martinez et al., 2005). Patients with epilepsy due to acquired vascular pathologies appear to have the best outcome, and malformations of cortical development, especially hemimegalencephaly, are associated with the worst outcome (Devlin et al., 2003; Kossoff et al., 2003; Jonas et al., 2004; Gonzalez-Martinez et al., 2005; Shimizu, 2005). Van Empelen et al. (2004), reported a less favorable seizure outcome in patients with Rasmussen's encephalitis. However, in the various series in the literature, there are differences in etiology, which may have influenced seizure outcome. Cerebral infarction and Sturge-Weber syndrome were more common in our series than in the series reported by Devlin et al. (2003) and Jonas et al. (2004) where malformations of cortical developmental were the most common. Rasmussen's encephalitis was more common in the reports of Kossoff et al. (2003) and Van Empelen et al. (2004).

In published series, blood loss and the requirement for blood transfusion are frequently encountered and expected complications of hemispheric surgery, and may be associated with hypovolemia and death (Devlin et al., 2003; Jonas et al., 2004; Piastra et al., 2004; Gonzalez-Martinez et al., 2005). In our series, there was a wide variation in blood loss. Hemidecortication, was associated with more severe bleeding requiring replacement. Young age and low weight were also important factors in predicting severe blood loss. Carson et al. (1996) and Vining et al. (1997) reported, in large series of patients who underwent hemidecortication, significant intraoperative bleeding and volume depletion in 12/58, most frequently seen in patients with malformations of cortical development (age less than 1 yr in all but one patient), followed by Rasmussen's encephalitis (older age group) and Sturge-Weber syndrome (where the only death related to bleeding occurred). Similarly, Piastra et al. (2004) noted that hemimegalencephaly was associated with greater risk of bleeding but this may be related to the younger age at surgery (less than 12 months). Kossoff et al. (2002) reported a series of 32 patients with Sturge-Weber syndrome undergoing hemispheric surgery, with four patients having significant intraoperative bleeding. Gonzalez-Martinez et al. (2005) reported no risk factors associated with degree of blood loss although the mean weight at surgery was 9.3 kg, all patients were less than 2 yr at time of surgery and all but one patient had abnormalities of cortical development. We report the first use of autologous blood in hemispheric surgery and in the patients where this technique was employed the use of allogenic blood was avoided. Other strategies employed to reduce transfusions and its complications included staging

of the hemispheric surgery in very young patients and those of low body weight, and the use of rhEPO but this is not an approved indication for widespread use of rhEPO.

Hydrocephalus requiring the insertion of a shunt occurs less frequently with functional hemispherectomies than hemidecortication (Carson et al., 1996) and anatomic hemispherectomies (Rasmussen, 1983). However, it remains a risk of surgery, occurring in 9–11% of cases (Devlin et al., 2003; Gonzalez-Martinez et al., 2005; Shimizu, 2005). Hemimegalencephaly has been more frequently associated than other pathologies with the need for shunting posthemispheric surgery (Vining et al., 1997; Devlin et al., 2003; Shimizu, 2005).

The 25% rate of early postoperative seizures in our series is similar to 30% reported by Devlin et al. (2003). The risk of recurrent seizures requiring reoperation has been documented as 6–23% (Carreno et al., 2001; Kossoff et al., 2003; Jonas et al., 2004; Gonzalez-Martinez et al., 2005; Shimizu, 2005). Contralateral MRI abnormalities were associated with a risk of ongoing seizures and need for antiepileptic drugs but contralateral interictal EEG abnormalities were not associated with poorer seizure outcome long-term.

In this series, two patients (8%) required reoperation because of incomplete disconnection for persistent seizures and are now seizure-free. Both had Rasmussen's encephalitis; one had a hemidecortication and the other periinsular hemispherotomy. Gonzalez-Martinez et al. (2005) report reoperation for persistent seizures related to incomplete disconnection in three (16%). Symptomatic hyponatremia and SIADH (Jonas et al., 2004) has infrequently been reported as a postoperative complication despite the nature of the surgery and the young population. We encountered this problem in only one patient. The other complications observed in our patients were self-limiting, required no specific therapy, and had no long term consequences for the patients.

The expected postoperative motor impairment in our patients is comparable to that described in reports by Vining et al. (1997) and Devlin et al. (2003). However, we did not document improvement postoperatively in preexisting hemiplegia which has been previously reported (Krynauw, 1950; White, 1961; Devlin et al., 2003) although all patients continued to develop gross motor function or maintain ambulatory abilities. We observed worsening of preoperative hemiplegia in 17% of patients, which contrasts with White's (1961) report of only 6%. This may be explained in part by the younger age at surgery in our series at which time the full extent of the hemiplegia had not yet become apparent. Homonomous hemianopia is an expected complication following hemispheric surgery in all patients, as is loss of useful hand function.

Postoperative severe headache and chronic intracranial hypertension have been reported (Carson et al., 1996). Late intracranial hypertension occurred in one patient in our series, and was likely due to a disturbance of cerebrospinal fluid flow dynamics. This patient responded to acetazolamide and further surgery was not required. Migraine has not been previously reported following hemispheric surgery. The prevalence of migraine requiring prophylactic therapy observed in our study was 17%; most patients had malformations of cortical development and all had undergone periinsular hemispherotomy. A family history of migraine in a first degree relative was observed in all. Bigal et al. (2003) reviewed the relationship between epilepsy and migraine, and reported prevalence of 8–15% in the literature, with an increased incidence of migraine in both idiopathic and symptomatic epilepsy. Yankovsky et al. (2005) recently reported in a series of adults with intractable partial epilepsy undergoing presurgical evaluation a 59% incidence of recurrent headaches; migraine occurring in 27%. Yamane et al. (2004) reported a migraine rate of 20% in a study examining comorbidity of headaches and epilepsy in children. Gee et al. (2003) documented the prevalence of postcraniotomy headache in 19% of cases (11/58), eight of whom had craniotomy as part of surgical management of epilepsy (no hemispheric procedures were performed).

With respect to cognitive and adaptive functioning, the most striking finding was the relationship of duration of epilepsy to SIB-R scores, such that better outcome in terms of overall broad independence and functioning in all four SIB-R domains was associated with a shorter duration of epilepsy. This finding is similar to the results of Jonas et al. (2004), and suggests that prolonged seizure activity may be a risk factor for poor outcomes, possibly by adversely affecting the nonoperated hemisphere. Thus, as suggested by Jonas et al. (2004), early control of seizures, in this case via hemispheric surgery, should be a high priority. Devlin et al. (2003) also reported that duration of epilepsy prior to surgery negatively impacted on behavior in a subgroup of their patients, although details of the specific behavioral/adaptive measures were not described. A history of infantile spasms further negatively impacted functional outcome; impact on the contralateral 'normal' hemisphere may be a particular feature of this type of epileptic encephalopathy, or serve as a marker for global cognitive dysfunction.

Our data also suggest that younger age at surgery is a positive factor with respect to aspects of postsurgical adaptive functioning, with higher SIB-R Broad Independence and Social/Communication scores related to earlier age at surgery. This finding supports the claim made by Devlin et al. (2003) that younger age at surgery maximizes the beneficial effects of surgery on future development, suggesting greater developmental plasticity early in life. However, it is notable that this outcome was not uniform

in our series; specifically, the three children with the earliest age at surgery in our sample had SIB-R Broad Independence scores in the very low range (<1st percentile). Thus, although it appears to be generally related to a better prognosis, younger age at surgery does not guarantee better adaptive functioning at follow-up. It is also important to note that children in this series, and in most series in the literature, are not randomly assigned to early vs. late surgical intervention. Thus, in many cases, age at surgery may be related to specific patient characteristics that lead to early surgical intervention such as seizure severity and etiology.

We observed a relative preservation of verbal abilities, both in terms of functional skills as assessed by the SIB-R, and in terms of standardized assessment via PPVT-III, after hemispheric surgery. Similar findings have been reported by Devlin et al. (2003) and Jonas et al. (2004). Language functioning, as indicated by the PPVT-III and SIB-R Social/Communication scores, was an area of strength overall in our sample at follow-up. A relative preservation of language abilities is well recognized in children following hemispherectomy, which is independent of laterality of disease (Krynauw, 1950; White, 1961). Mariotti et al. (1998) discussed the hypothesis of early hemisphere equipotentiality relating to linguistic abilities and that there may be a functional reorganization in a hierarchical fashion privileging linguistic skills. However, Vanlancker-Sidtis (2004) has reported a case where subtle deficiencies in higher linguistic function after hemispherectomy were demonstrated at follow-up, suggesting an incomplete equipotentiality of the hemispheres or a developmental or age limitation to full preservation of language skills. This is also supported by our findings. Although language was an area of relative strength, standard scores were nevertheless generally below age expectations in the majority of children.

Limitations

Our study is limited by a relatively small sample size and lack of complete preoperative and serial postoperative adaptive functioning and cognitive data on all patients.

CONCLUSION

This study has demonstrated that, etiology influenced motor and seizure control outcome following hemispheric surgery. Blood loss was the most common complication and age, weight and hemidecortication were risk factors. This study reports adaptive function in all children on follow-up. Poor functional outcome was associated with longer duration of epilepsy, older age at time of surgery and a history of infantile spasms. There was a relative preservation of language skills despite laterality of disease. Early age at surgery was associated with better adaptive outcome.

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