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Hemispherectomy for intractable seizures in children: a report of 58 cases

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Abstract Fifty-eight children who underwent anatomical, functional, or modified anatomical hemispherectomy for intractable seizures from 1986 to 1995 were evaluated for seizure control, motor function, and complications. Age at surgery ranged from 0.3 to 17.3 years (median 2.8 years). Twenty-seven anatomical, 27 functional, and 4 modified anatomical hemispherectomies were performed. Seizure control and motor function in the 50 patients with more than 1 year follow-up revealed a 90% or better reduction in seizure frequency in 44/50 (88%) overall: 19/22 (86%) anatomical, 23/26 (89%) functional, and 2/2 modified anatomical. Motor function of the preoperatively hemiparetic extrem-

ities was improved or unchanged postoperatively in 38/50 (76%) of the patients. Complications included one intraoperative death, one late death from shunt obstruction managed elsewhere, late postoperative seizure breakthrough requiring reoperation and further disconnection in 5/27 functional hemispherectomy patients, mild cerebrospinal fluid infections in 3/27 anatomical hemispherectomy patients, and hydrocephalus requiring shunting in 3/27 functional hemispherectomy patients. A review of the literature and comparison of techniques is presented.

Key words Hemispherectomy · Epilepsy · Infantile hemiplegia · Surgery · Children

Introduction

Cerebral hemispherectomy is one of the most successful surgical procedures used in the treatment of intractable epilepsy. Walter Dandy introduced this procedure in 1928 as a radical attempt to treat adults with diffuse nondominant hemispheric gliomas [11]. Although the surgery failed to cure malignancy, he was surprised by the degree of functional recovery made by the patients. Hemispherectomy was first utilized in the treatment of intractable epilepsy in 1938 by Kenneth McKenzie, who found the procedure to be effective in controlling seizures in a hemiplegic woman [31]. From 1945 to 1950, Roland Krynauw performed 12 hemispherectomies in children with intractable epilepsy

and infantile hemiplegia [30]. His short-term results showed marked improvement in seizure control as well as motor function, personality, behavior, and mentation. Subsequently, hemispherectomy was adopted by neurosurgical centers worldwide as a successful means of controlling refractory seizures in children.

This initial enthusiasm for hemispherectomy began to fade when long-term follow-up revealed late postoperative complications. In 1966, Oppenheimer and Griffith reported a syndrome of repeated episodes of intracranial bleeding occurring in 18% (3/17) of their patients [33]. Clinically, these children had an initial trouble-free period, followed by a gradual deterioration to death 7.5–11 years postoperatively. Autopsies showed evidence of multiple bleeding points in the subdural membrane lining the hemispherectomy

tomy cavity resulting in granular ependymitis, obstructive hydrocephalus, and superficial hemosiderosis of the central nervous system. Wilson in 1970 also reported delayed intracranial hemorrhage in 30% (15/50) of his patients, stating the causal lesion to be a chronic subdural hematoma of the hemicranial cavity [53]. Rasmussen found that early hydrocephalus developed without evidence of hemorrhage in 10% of 31 patients following anatomical hemispherectomy, which he attributed to extensive removal of the arachnoidal absorbing surface [36].

In an effort to eliminate recurrent intracranial hemorrhage and its potentially devastating complications, multiple modifications were proposed. Rasmussen reasoned that the total removal of one hemisphere left the other unsupported, predisposing to hemorrhagic complications [35]. He suggested that the classical anatomical hemispherectomy be abandoned in favor of a functionally complete but anatomically subtotal hemispherectomy. He devised a procedure which entailed the removal of the central cortical area and the temporal lobe. The anterior frontal, posterior parietal, and occipital lobes were preserved but disconnected from the remainder of the brain by dividing the corpus callosum and the internal capsule, leaving their blood supply intact. Rasmussen compared the outcome of his series of anatomical hemispherectomy to functional hemispherectomy patients and reported comparable success rates of 83% and 85% near or complete seizure control [37]. Long-term follow-up ranging from 2 to 33 years indicated that 35% (11/31) of the anatomical hemispherectomy patients developed late pressure complications, compared to only 7% (1/14) of the functional hemispherectomy patients.

Hemicortectomy or hemidecortication was proposed independently by Winston in 1992 and Hoffman in 1993 as another alternative to anatomical hemispherectomy [49, 54]. This technique involves removal of the cortical gray matter, which is the origin of epileptogenic activity, with partial preservation of the underlying white matter. The goal is to preserve the integrity of the ventricular system by leaving the ependyma and overlying white matter undisturbed. Winston reported that 91% (10/11) of his patients had near or complete seizure control, with only one requiring shunting for early postoperative hydrocephalus. However, as Villemure commented, hemidecortication was an option only in candidates with a cortical mantle thick enough to allow its removal without damaging the underlying ventricular system [47].

Because of the highly effective seizure control achieved with anatomical hemispherectomy, some neurosurgeons continue to employ the classical procedure with partial modifications. In 1983, Adams described dural plication to reduce the size of the subdural cavity in exchange for a large epidural cavity [1]. He also isolated the subdural space from the ventricular system by plugging the ipsilateral foramen of Monro with muscle. In 1990, Peacock at the University of California at Los Angeles reported

the use of routine early post-operative shunting in anatomical hemispherectomy [34]. Following immaculate hemostasis, the subdural cavity was allowed to drain externally for 3–5 days prior to placement of a subdural-peritoneal shunt. This allowed removal of blood products (most importantly thrombin) that are known to induce subdural membrane formation and its resultant hemorrhagic complications.

In addition to anatomical hemispherectomy, Peacock also utilized the techniques of functional hemispherectomy and modified anatomical hemispherectomy (hemidecortication) at UCLA. The present paper reports the results in a series of 58 patients who underwent anatomical hemispherectomy, functional hemispherectomy, or modified anatomical hemispherectomy.

Patients and methods

Patient population

From January 1986 to August 1995, 58 children with intractable epilepsy underwent hemispherectomy at UCLA Medical Center. Twenty-seven anatomical, 27 functional, and 4 modified anatomical hemispherectomies were performed.

Table 1 summarizes general patient characteristics. There were 31 male patients and 27 females. The left hemisphere was involved in 33 patients and the right in 25 patients. Seizure onset ranged from the day of birth to age 11.4 years, with a median age at onset 0.1 year. Age at operation ranged from 0.3 to 17.3 years with a median of 3.3 years. The interval between onset of seizures and surgery ranged from 0.3 to 14.1 years with a median of 2.8 years. Seizure types were partial with or without secondary generalization and infantile spasms. Daily seizure frequency before surgery ranged from 3 to 250 with an approximate mean of 50 per day. Table 2 lists the distribution of patient age at the time of surgery.

Table 1 General characteristics of patient group ($n=58$) undergoing hemispherectomy

Gender distribution (M/F)	31/27
Hemisphere involved (L/R)	33/25
Age at onset of seizures	
Range	0.0–11.4 years
Median	0.1 year
Mean	0.8 year
Age at operation	
Range	0.3–17.3 years
Median	3.3 years
Mean	4.8 years
Interval from seizure onset to surgery	
Range	0.3–14.1 years
Median	2.8 years
Mean	3.9 years
Preoperative seizure frequency	
Range	3–250 per day
Mean	50 per day