

SUPERCALIFRAGILISTIC HEMOSIDEROSIS: A RARE AND UNUSUAL COMPLICATION THAT REALLY SOUNDS ATROCIOUS. . .

Anatomical Hemispherectomy for Intractable Seizures: Excellent Seizure Control, Low Morbidity and No Superficial Cerebral Haemosiderosis

O'Brien DE, Basu S, Williams DH, May PL

Childs Nerv Syst 2006;22:489–498.

OBJECTIVE: This current study was performed to evaluate whether superficial cerebral hemosiderosis (SCH) is still a complication of modern day anatomical hemispherectomy.

METHODS: We report a 13-year institutional experience with anatomical hemispherectomy for intractable epilepsy. Seizure control at a mean follow-up interval of 7 years was 83%. Though one patient died post-operatively from

a nonneurosurgical complication, mortality was otherwise zero and morbidity minimal. The much-described complication of SCH following anatomical hemispherectomy was nonexistent. We explain the history of SCH as a complication of anatomical hemispherectomy, and the measures that are presently taken to prevent it.

CONCLUSIONS: We suggest that the importance of SCH in modern epilepsy surgery is probably over-emphasized.

COMMENTARY

The technique of surgically removing an entire hemisphere was originally described by Walter Dandy in 1928 as a treatment for glioma (1). McKenzie later adopted the technique to treat unilateral hemispheric epilepsy (2). This anatomic hemispherectomy involved removal of the brain parenchyma of the entire hemisphere, which assured the absence of any residual epileptogenic tissue. However, the technique soon fell into disfavor as reports emerged of superficial hemosiderosis and hydrocephalus occurring in as many as 33% of patients and as far out as 8 years after surgery (3,4). Superficial hemosiderosis, caused by the formation of a hemorrhagic membrane in communication with the ventricular system, can lead to a variety of symptoms, such as recurrent headaches, cerebellar ataxia, cognitive impairment, urinary incontinence, deafness, and motor abnormalities as well as to death.

Nevertheless, hemispherectomy is an extremely efficacious treatment for unilateral hemispheric epilepsy, with seizure-free rates approaching 70% to 80% in well-selected cases (5–7). For this reason, a variety of alternative surgical techniques have arisen to minimize the risk of superficial hemosiderosis. These approaches include variations in the anatomic hemispherectomy that isolate the ventricular system from the resection cavity (8), hemidecortication (5), and functional hemispherectomy,

which disconnects the ipsilateral abnormal brain tissue from the contralateral hemisphere and descending motor tracts, while leaving significant amounts of brain parenchyma in place (6,7,9,10). In addition to minimizing the risk of superficial hemosiderosis, the functional hemispherectomy has been shown to decrease both the amount of blood loss and the risk of postoperative hydrocephalus. However, the learning curve is steeper with this technique, which has led to an increase in the rate of re-operation to complete a previously inadequate disconnection (6).

In their recent report, O'Brien et al. demonstrate that the anatomic hemispherectomy, performed in the modern era with a few small modifications, does not have the same risk of superficial hemosiderosis as previously thought. Indeed, other authors have noted that anatomic disconnections can be performed safely, with a low or zero rate of superficial hemosiderosis (6). The authors attribute their success to a combination of meticulous hemostasis, aggressive ventriculoperitoneal shunting, when necessary, and anatomic isolation of the ventricular system from the resection cavity. Yet, in spite of these modifications, the morbidity of their procedure remains quite high. Although the authors state that the morbidity was "minimal," they go on to report that among their 19 patients, there was 1 acute subdural hematoma, requiring surgery; 4 infections, 3 of which required craniectomies; 2 cases of hydrocephalus, requiring shunting and then shunt revision; 2 subgaleal collections, requiring aspiration; 1 postoperative tracheostomy; 1 coagulopathy and 1 epidural hematoma, which resolved spontaneously. From this data, one can extrapolate a morbidity rate of 62%, which is hardly minimal. Morbidity rates following functional

hemispherectomy are markedly lower, ranging anywhere from 11% to 35% (6,7).

The question remains as to which technique is preferable to achieve the best outcome, with the least morbidity. Ultimately, seizure control rates are comparable for all techniques, if performed correctly. The greatest determinant of successful seizure control appears to be the pathologic substrate, with lower control rates found in cases of cortical dysplasia, such as hemimegalencephaly (5). With regard to morbidity, although superficial hemosiderosis following anatomic hemispherectomy may not be as common as previously thought, morbidity and blood loss are greater with the anatomic procedure compared with functional disconnection. Hence, all things considered, the latter procedure appears to be preferable. With the proliferation of intra-operative, image-guided stereotactic navigation and the advances in microsurgical technique, the learning curve for the functional hemispherectomy procedures should become less steep—thereby guaranteeing more consistently complete disconnections. Until that time, given the equivalent efficacy of the various techniques and the new understanding of the possibility of low rates of superficial hemosiderosis with anatomic hemispherectomy, surgeons may do well to perform the surgical procedure with which they are most familiar.

by Theodore H. Schwartz, MD, FACS

References

1. Dandy WE. Removal of right cerebral hemisphere for certain tumours with hemiplegia. *J Am Med Assoc* 1928;90:823–825.
2. McKenzie KG. The present status of a patient who had the right cerebral hemisphere removed. *JAMA* 1938;111:168–183.
3. Falconer MA, Wilson PJ. Complications related to delayed hemorrhage after hemispherectomy. *J Neurosurg* 1969;30:413–426.
4. Rasmussen T. Postoperative superficial hemosiderosis of the brain, its diagnosis, treatment and prevention. *Trans Am Neurol Assoc* 1973;98:133–137.
5. Kossoff EH, Vining EPG, Pillas DJ, Pyzik PL, Avellino AM, Carson BS, Freeman JM. Hemispherectomy for intractable unihemispheric epilepsy. Etiology vs outcome. *Neurology* 2003;61:887–890.
6. Cook SW, Nguyen ST, Hu B, Yudovin S, Shields WD, Vinters HV, Van de Wiele BM, Harrison RE, Mathern GW. Cerebral hemispherectomy in pediatric patients with epilepsy: comparison of three techniques by pathological substrate in 115 patients. *J Neurosurg* 2004;100:125–141.
7. Schramm J, Behrens E, Entzian W. Hemispheric deafferentation: an alternative to functional hemispherectomy. *Neurosurgery* 1995;36:509–515.
8. Adams CB. Hemispherectomy—a modification. *J Neurol Neurosurg Psychiatry* 1983;46:617–619.
9. Rasmussen T. Hemispherectomy for seizures revisited. *Can J Neurol Sci* 1983;10:71–78.
10. Villemure J-G, Mascott CR. Per-insular hemispherotomy: surgical principles and anatomy. *Neurosurgery* 1995;37:975–981.