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Huh? Hemispherectomy in adults?

An exercise in risk-benefit analysis

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Decision-making in complex clinical situations is part of all specialty centers in medicine, and usually this means examining risks *vs* benefits. For surgical epilepsy centers, discussing risk *vs* benefit of various treatment approaches is the purpose of the indispensable multidisciplinary conference. A treatment option can have high benefit or low benefit, and high risk or low risk. A basic and common situation in medically intractable epilepsy is the choice between focal resection and vagus nerve stimulation (VNS) (Benbadis *et al.*, 2000), and patients and families frequently ask for guidance. While the goal of seizure-freedom after focal resection is considered more ambitious and more invasive, VNS is less ambitious (aiming at 50% reduction) and less risky. In a patient with straightforward unilateral mesiotemporal epilepsy, this risk-benefit ratio clearly favors resective surgery, but may favor VNS as a first treatment in extratemporal non-lesional epilepsy. It must also be acknowledged that the risk-benefit ratio can easily be biased by non-rational factors in patients desperate for complete seizure-freedom.

Patients with bitemporal independent seizures present a different set of issues. With > 80% predominance, resective surgery is often considered worthwhile, but is discouraged when the distribution is 50/50. But would a 50% seizure reduction be worthwhile?

In patients with symptomatic generalized epilepsies of the Lennox-Gastaut type, focal resections are futile and not generally recommended unless there is a predominant focus that would benefit from a resective procedure. Is focal resection then warranted?

In hemispheric syndromes such as Rasmussen's encephalitis, HHE and hemimegalencephaly, functional hemispherectomy is often considered the treatment of choice for seizure control. Here the usual dilemma pits seizure control against hemiplegia and hemianopia. Conventional thinking suggests that hemispheric surgery should only be considered in early life in patients with pre-existing hemispheric syndromes. The small series presented in this issue of *Epileptic Disorders* (Steinhoff *et al.*, 2009) within the series of "Anatomo-electro-clinical correlations", challenges this thinking by presenting a series of relatively intact adult patients undergoing hemispheric ablation.

The conventional wisdom in epilepsy surgery is to remove the smallest amount of brain in order to preserve function. However, this does not seem to work in patients with extensive epileptogenic zones or lesions. In Dr. Rasmussen's words, possibly based on his experience with soldiers injured during the World War II, "no brain is better then bad brain."

Generally and traditionally, clinicians and experts "shy away" from such an invasive procedure because there is not enough experience to make appropriate decisions.

From this small series, it would appear that FH *can* be considered an option in some catastrophic epilepsies even in adults. The main problem is the lack of criteria to come to the conclusion that a functional hemispherectomy and the possible deficits associated with it are "worth it." As in children, catastrophic epilepsies in adults have a poor long-term outcome that can be, in some cases at least, well defined. For example, adults with Rasmussen's encephalitis or unilateral hemispheric atrophy (HHE syndrome) are prone to prolonged hemiconvulsions, falls, and recurrent status epilepticus, with all the morbidity related to these severe and potentially lethal conditions. This is also probably true for post-traumatic cases in which extensive unilateral damage is found, and maybe in some cases of post-encephalitic epilepsy. Thus, etiology is a key factor to define prognosis and in particular the severity of the ensuing epilepsy, the risk of seizure-related accidents and death (SUDEP) and of further neurological deterioration due to the ongoing epileptic activity.

This small series suggests that, in some circumstances, newly acquired neurological deficits may be an acceptable price in exchange for seizure-freedom. Is such a decision acceptable? It is difficult to know which of the

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two evils is more disabling. Ultimately each patient and the caregivers must decide. As the authors point out, it is critical that the patient, family, and other decision-makers be adequately informed of the risks and benefits. The caveat is that it is difficult to predict the impact of additional neurological deficits on these patients. Deficits may not be limited to hand function, and can include lower extremity (ambulation) and visual fields. In some cases, language function may also be compromised. Thus, there should be clear discussions between the patient and caregivers and the treating team, including neuropsychologist, psychiatrist and occupational therapist.

This series raises some difficult ethical issues. Hemispheric surgery has traditionally been offered as a treatment in infants and young children for several reasons. Functional reorganization of language cortex and to a lesser degree motor function after hemispherectomy is superior for surgery in the developing nervous system. Furthermore, there is a greater tendency for acquired deficits are more likely to be tolerated when damage occurs early. In fact, this raises the question of why these patients were not referred for surgery earlier, but this delay is not new and is also seen in focal resective surgery (Benbadis *et al.*, 2003; Berg, 2004).

In contrast, acquired deficits in adulthood are much less likely to be tolerated emotionally and may have lead to severe clinical depression. For this reason, data from an adult cohort undergoing FH would be helpful.

In the absence of data to serve as guidelines, appropriateness boils down to informed consent (Vale and Benbadis) and the ability of the patient and family to make the decision. The concern here centers around the adequacy of explanation of the consequences of hemispheric destruction by caregivers. These effects are not technically "risks" of surgery (*i.e.* hemorrhage, infection) but rather are guaranteed known post-operative deficits. Patients and their caregivers often do not have a realistic understanding of these consequences unless given the opportunity to view them first-hand in others. Similar concerns apply to children but are less resonant given the greater potential for neurologic recovery in young patients and concerns over future seizure-induced neuro-cognitive deterioration.

Because this type of risk-benefit decisions may require a level of expertise, compassion and communication heretofore unknown in epilepsy surgery decision-making, we would emphasize the importance of involving multidisciplinary bioethic committees, including psychiatrists, psychologists, and neuropsychologists. The decision on if and when in the course of the disease surgery should be undertaken depends as mentioned on the certainty of the diagnosis, the severity and frequency of the seizures, and on the impact on the psychosocial situation of the patient. Understanding the natural evolution of the disease and the severity of the epilepsy may help them to take a decision and to justify the intervention, even with the possibility of additional deficits. The decision about such a radical procedure requires considerable time and thought, and the psychological preparation of patients and their families is essential. □

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Epileptic Disorders Case Records, published under the heading "Anatomo-electro-clinical correlations" are expected to provide to the reader a comprehensive approach of pre-surgicalevaluation and epilepsy surgery strategies. Authors are expected to provide supplemental data for publication on the DVD to allow further discussion on the surgical approach chosen.

Epileptic Disorders will published all documented comments, critics and suggestions discussing the approach taken by the authors. The readers are invited to submit their eventual comments in the online submission system as "Letter to the Editor" with reference to the Case Records' number.

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