For children with rare genetic disorder, more extensive epilepsy surgery yields better seizure control

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Children with the genetic disorder tuberous sclerosis complex (TSC) often need epilepsy surgery for severe, uncontrollable seizures. A new study finds that seizure control is improved for patients undergoing more extensive surgery, reports the October issue of *Neurosurgery*, official journal of the Congress of Neurological Surgeons.

Seizures occurring in TSC are related to development of brain tumors, known as "tubers," that develop in this disorder. But the new study by Dr. Aria Fallah of Miami Children's Hospital and colleagues finds better outcomes when surgery includes the entire "epileptogenic zone" from which seizures are originating—not just the tuber itself.

Epilepsy Surgery for Tuberous Sclerosis—Study from Six Specialty Centers

The study included data on a total of 74 children with TSC who underwent epilepsy surgery at six leading pediatric epilepsy centers from 2005 through 2013. Although TSC is rare, occurring in about 1 in 10,000 children, it is one of the most common genetic causes of epilepsy.

Tubers are non-cancerous (benign) tumors that develop in the brain (and other organs) of patients with TSC. Nearly 90 percent of affected children have seizures related to brain tubers; in most cases, these

seizures can't be controlled by antiepileptic medications.

Epilepsy surgery is an effective treatment, but many children continue to have seizures after surgery. Because TSC is rare and only some patients undergo surgery, it's difficult to predict the outcomes and best approaches to surgery.

Using a combined experience of six US and Canadian hospitals, Dr. Fallah and colleagues analyzed factors affecting the outcomes of "curative" epilepsy surgery for children with TSC. The patients were 41 males and 33 females. The median age was ten years, with a range from three months to 18 years.

One year after epilepsy surgery, 65 percent of patients were free of seizures; by four years, that figure had decreased to 43 percent. Initial analysis suggested a lower rate of recurrent seizures for younger patients, for those with a larger "predominant" tuber, and in resections larger than the tuber alone. However, after adjustment for other factors, age and tuber size were no longer significant predictors.

In the adjusted analysis, only one factor predicted a better chance of freedom from seizures: more extensive surgery. Children who had resection of the tuber and surrounding area (lobectomy) were about three times more likely to remain seizure-free longer, compared to those undergoing resection of the tuber only.

Epilepsy surgery is appropriate only for a subgroup of children with TSC: those whose seizures are mapped to an identifiable epileptogenic zone, which can be removed or disconnected without causing new neurological abnormalities. But even for this group of patients, there are no established data on which factors predict the success of curative epilepsy surgery.

The new results suggest that simply resecting the tuber may not be enough—more extensive surgery is needed to achieve the best chances of longer-term seizure-freedom. The study confirms previous results suggesting that "the malformed cortex surrounding the tuber may contribute more to the epileptogenicity as opposed to the tuber itself." Dr. Fallah and coauthors add, "This limits the role of tuberectomies."

The researchers note some important limitations of their study: it evaluated a relatively small number of potential predictors and was limited to children treated at a handful of specialized centers. Dr. Fallah and colleagues call for long-term follow-up to clarify the factors associated with better or worse outcomes of epilepsy surgery in TSC, along with efforts to standardize the criteria for determining which children are eligible for this procedure.

More information: Aria Fallah et al. Resective Epilepsy Surgery for Tuberous Sclerosis in Children, *Neurosurgery* (2015). DOI: 10.1227/NEU.0000000000000875

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