EPILEPSY SURGERY FOR TUBEROUS SCLEROSIS PATIENTS

Seizures or epilepsy are common manifestations of tuberous sclerosis. Some 80-90% of individuals with TS have seizure activity during their lifetime, often with onset in childhood. Fortunately, many children and adults with TS have seizure activity that is controlled by medication. After 1-2 years of being seizure free, it is often possible to decrease the use of medications and continue to be seizure free.

Unfortunately, for some children and adults with TS, the seizure activity proves intractable to medication. This means that despite therapeutic trials of many established anticonvulsant medications, the individual continues to have seizure activity. If medications are not successful in controlling seizure activity, alternative methods of controlling the seizures should be considered. Currently, the ketogenic diet, the vagal nerve stimulator and epilepsy surgery are the major alternative treatments for epilepsy management, and each of these should be considered in a child or adult with TS and intractable epilepsy.

Several reports in the medical literature suggest that epilepsy surgery can be a successful treatment for individuals with TS. This has been and is still debated, primarily because individuals with TS often have more than one area of unusual brain formation or tuber location. The ideal epilepsy surgery candidate is considered a person with one seizure type, a clear correlation between clinical seizure activity and EEG abnormalities (i.e. EEG showing focal onset of seizure activity concurrent with or preceding onset of clinical seizures), and evidence of abnormality on neuroimaging that correlates with EEG abnormality. Therefore, children with TS are not considered "ideal" surgical candidates due to the typical multiple tubers in their brain. One concern is that it is not possible to predict the "natural history" of all of the other tubers, i.e. will any become "epileptogenic" in the future, and will the child develop a different seizure focus related to another tuber. However, if the current seizure activity is significantly impacting the child's development or quality of life and is not responding to medications, the possibility of surgery should be considered.

If surgery is to be considered, you should be referred to a major medical center that has a well-established epilepsy surgery program. The NTSA can provide you a list of these centers. In order to determine if you or your child is a surgical candidate, a presurgical or Phase 1 evaluation is performed. This typically consists of a 3-5 day hospitalization at the center, during which time continuous video EEG monitoring is performed with the goal of observing several typical seizure episodes to determine where in the brain the seizures are starting. The duration of the hospital stay depends on the seizure frequency, because most centers prefer to monitor several "typical" episodes to ensure that the localization of EEG abnormalities is consistent with each seizure. Several other tests are also performed, and may vary depending on center. Typically a magnetic resonance image (MRI) of the brain is obtained, and most centers have special protocols for evaluating the epilepsy surgery patients. Additional neuroimaging studies such as PET (positron emission testing), SPECT, MEG (magnetoencephalography), and fMRI (functional MRI) may be used to identify more precisely the "epileptogenic zone", and also to help determine possible deficits that may result from a resective surgery. Most centers also include neuropsychological evaluation as part of the presurgical evaluation, to determine if the child's cognitive deficits match the EEG abnormalities and to help determine whether language or memory would likely be affected by resective surgery.

Following the Phase 1 evaluation, the epilepsy surgery team (consisting of the neurologist/epileptologist, neurosurgeon, neuroradiologists and neuropsychologists) reviews all of the data collected, and decides if epilepsy surgery would likely lead to a reduction in seizure frequency without significant side effects. In children with TS, this evaluation frequently identifies a tuber or tubers thought to be involved in the child's epilepsy that could be resected.

In many cases, particularly children with TS and multiple tubers, further monitoring is recommended to better delineate the epileptogenic regions. This typically involves invasive monitoring, or monitoring with subdural grids rather than surface EEG. If this is done, the child has neurosurgery to expose the region of the brain thought to be involved in the generation of seizure activity, and grids of closely spaced electrodes are placed on the surface of the brain. The piece of skull is placed back over this region, and continuous video EEG monitoring is performed for several days again to monitor several "typical" seizures in order to identify the region of brain in which the seizures appear to start. This type of monitoring allows more accurate localization of the "irritable" region, and...
often helps reduce or limit the size of subsequent brain resection.

As above, there have been several reports in the medical literature of individuals, both children and adults with TS and intractable epilepsy who successfully undergo epilepsy surgery. For some children, it provides a "cure". For other children it reduces the seizure frequency, and allows the number of medications to be reduced. Therefore, for any child or adult with TS who develops a seizure disorder that is not controlled by medication, the possibility of epilepsy surgery should be considered.

* Tuberous Sclerosis Alliance Fact Sheets are intended to provide basic information about TS. They are not intended to, nor do they, constitute medical or other advice. Readers are warned not to take any action with regard to medical treatment without first consulting a physician. The TS Alliance does not promote or recommend any treatment, therapy, institution or health care plan.