Tuberous sclerosis complex surgery

A review of the tuberous sclerosis complex surgery literature demonstrates impressive success rates for seizure-free outcomes. These studies describe a number of novel noninvasive methods for seizure localization including PET, SPECT, and magnetoencephalography. Additionally, there is a subset of patients with TSC with bilateral, multifocal, or generalized epileptiform discharges that would have previously been excluded from resection. New developments in neuroimaging and invasive monitoring with intracranial electrodes are useful methods in identifying an epileptogenic tuber in these individuals with refractory epilepsy.

Case series

Liu et al. reported a nationwide multicentre retrospective study and analyse the long-term seizure and neuropsychological outcomes of epilepsy surgery in patients with tuberous sclerosis complex. There were 364 patients who underwent epilepsy surgery in the study. Patients' clinical data, postoperative seizure outcomes at 1-, 4-, and 10-year follow-ups, preoperative and postoperative intelligence quotients, and quality of life at 1-year follow-up were collected. The patients' ages at surgery were 10.35 ± 7.70 years (range: 0.5-47). The percentage of postoperative seizure freedom was 71% (258/364) at 1-year, 60% (118/196) at 4-year, and 51% (36/71) at 10-year follow-up. Influence factors of postoperative seizure freedom were the total removal of epileptogenic tubers and the presence of outstanding tuber on MRI at 1- and 4-year follow-ups. Furthermore, monthly seizure (versus daily seizure) was also a positive influence factor for postoperative seizure freedom at 1-year follow-up. The presence of an outstanding tuber on MRI was the only factor influencing seizure freedom at 10-year follow-up. Postoperative quality of life and intelligence quotient improvements were found in 43% (112/262) and 28% (67/242) of patients, respectively. Influence factors of postoperative quality of life and intelligence quotient improvement were postoperative seizure freedom and preoperative low intelligence quotient. The percentage of seizure freedom in the tuberectomy group was significantly lower compared to the tuberectomy plus and lobectomy groups at 1- and 4-year follow-ups. In conclusion, this study, the largest nationwide multi-centre study on resective epilepsy surgery, resulted in improved seizure outcomes and quality of life and intelligence quotient improvements in patients with tuberous sclerosis complex. Seizure freedom was often achieved in patients with an outstanding tuber on MRI, total removal of epileptogenic tubers, and tuberectomy plus. Quality of life and intelligence quotient improvements were frequently observed in patients with postoperative seizure freedom and preoperative low intelligence quotient [1)].

Epilepsy surgery can provide good seizure outcome in selected children with intractable epilepsy due to tuberous sclerosis complex (TSC) [2)]

Huang et al. demonstrated the efficacy of tuberous sclerosis complex treatment and intractable epilepsy with surgery. Detailed perioperative tests are a reliable predictor of postoperative seizure freedom [3)].

Case reports

Evans et al. offered a survey of the literature and a description of these methods. Additionally, they present an illustrative case of ictal SPECT and intracranial electroencephalography used in the preoperative evaluation of a 10-year-old girl with intractable seizures and TSC. This patient ultimately underwent resection of an epileptogenic region within the occipital lobe [4].

1)
Liu S, Yu T, Guan Y, Zhang K, Ding P, Chen L, Shan Y, Guo Q, Liu Q, Yao Y, Yang M, Zhang S, Lin Y, Zhao R, Mao Z, Zhang J, Zhang C, Zhang R, Yang Z, Qian R, Li Y, Zhang G, Yuan L, Yang W, Tian H, Zhang H, Li W, Zhang X, Yin J, Guo Y, Zou L, Qin J, Fang F, Wang X, Ge M, Liang S. Resective epilepsy surgery in tuberous sclerosis complex: a nationwide multicentre retrospective study from China. Brain. 2020 Feb 1;143(2):570-581. doi: 10.1093/brain/awz411. PMID: 31953931.
2)
Gupta A. Epilepsy Surgery in Tuberous Sclerosis Complex: In Pursuit of the Epileptogenic Center(s). Epilepsy Curr. 2017 May-Jun;17(3):150-152. doi: 10.5698/1535-7511.17.3.150. PMID: 28684945; PMCID: PMC5486420.
3)
Huang Q, Zhou J, Wang X, Li T, Wang M, Wang J, Teng P, Qi X, Zhu M, Luan G, Zhai F. Predictors and Long-term Outcome of Resective Epilepsy Surgery in Patients with Tuberous Sclerosis Complex: A Single-centre Retrospective Cohort Study. Seizure. 2021 Mar 25;88:45-52. doi: 10.1016/j.seizure.2021.03.022. Epub ahead of print. PMID: 33812307.
4)
Evans LT, Morse R, Roberts DW. Epilepsy surgery in tuberous sclerosis: a review. Neurosurg Focus. 2012 Mar;32(3):E5. doi: 10.3171/2012.1.FOCUS11330. PMID: 22380859.