

Epilepsy in Sturge-Weber syndrome

Few studies have reported the clinical presentation, surgical treatment, outcomes, and influential factors for patients with [epilepsy](#) and [Sturge-Weber syndrome](#). This large-scale retrospective study continuously enrolled 132 patients with Sturge-Weber syndrome and epilepsy from January 2008 to December 2018 at [SanBo Brain Hospital, Beijing](#) to analyze their characteristics. Among these patients, 90 underwent [epilepsy surgery](#), and their postoperative 2-year follow-up [seizure](#), cognitive, and motor functional outcomes were assessed and analyzed. Univariable and multivariable logistic analyses were conducted to explore the influential factors. Among the Sturge-Weber syndrome patients for whom characteristics were analyzed ($n = 132$), 76.52% of patients had their first epileptic seizures within their first year of life. The risk factors for cognitive decline were seizure history ≥ 2 years (adjusted odds ratio [aOR] = 3.829, 95% confidence interval [CI]: 1.810-9.021, $p = 0.008$), bilateral leptomeningeal angiomas (aOR = 3.173, 95% CI: 1.970-48.194, $p = 0.013$), age at onset < 1 year (aOR = 2.903, 95% CI: 1.230-6.514, $p = 0.013$), brain calcification (aOR = 2.375, 95% CI: 1.396-5.201, $p = 0.021$) and left leptomeningeal angiomas (aOR = 2.228, 95% CI: 1.351-32.571, $p = 0.030$). Of the patients who underwent epilepsy surgery ($n = 90$), 44 were subject to focal resection, and 46 underwent hemisphere surgery (19 anatomical hemispherectomies and 27 modified hemispherotomies). A postoperative seizure-free status, favorable cognitive outcomes, and favorable motor outcomes were achieved in 83.33%, 44.44%, and 43.33% of surgical patients, respectively. The modified [hemispherotomy](#) group had similar surgical outcomes, less intraoperative blood loss and shorter postoperative hospital stays than the anatomical [hemispherectomy](#) group. Regarding seizure outcomes, full resection (aOR = 11.115, 95% CI: 1.260-98.067, $p = 0.020$) and age at surgery < 2 years (aOR = 6.040, 95% CI: 1.444-73.367, $p = 0.031$) were positive influential factors for focal resection. Age at surgery < 2 years (aOR = 15.053, 95% CI: 1.050-215.899, $p = 0.036$) and infrequent seizures (aOR = 8.426, 95% CI: 1.086-87.442, $p = 0.042$; monthly vs. weekly) were positive influential factors for hemisphere surgery. In conclusion, epilepsy surgery resulted in a good postoperative seizure-free rate and favorable cognitive and motor functional outcomes and showed acceptable safety for patients with epilepsy and Sturge-Weber syndrome. Modified hemispherotomy is a less invasive and safer type of hemisphere surgery than traditional anatomic hemispherectomy with similar surgical outcomes. Early surgery may be helpful to achieve better seizure outcomes and cognitive protection, while the risk of surgery for young children should also be considered ¹⁾.

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Wang S, Pan J, Zhao M, Wang X, Zhang C, Li T, Wang M, Wang J, Zhou J, Liu C, Sun Y, Zhu M, Qi X, Luan G, Guan Y. Characteristics, surgical outcomes, and influential factors of epilepsy in Sturge-Weber syndrome. *Brain*. 2021 Dec 21:awab470. doi: 10.1093/brain/awab470. Epub ahead of print. PMID: 34932802.

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