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)].

Fourteen children with unilateral SWS were categorized according to age, i.e., ≤ 2 years (group A, n = 5, mean age 1.1 years, 3 males) and > 2 years (group B, n = 9, mean age 3.9 years, 4 males). All children underwent two-dimensional synthetic qMRI. The myelin volume in the cerebral hemisphere and white matter (WM) myelin volume fraction (MVF), proton density (PD), R1 and R2 relaxation rates ipsilateral to the leptomeningeal enhancement, and/or a port-wine birthmark were compared with the corresponding values in the contralateral hemisphere.

In group A, 3 patients had a higher myelin volume in the ipsilateral hemisphere and a higher MVF, R1, and R2 and lower PD in the ipsilateral WM than on the contralateral side; the findings were the opposite in the remaining two patients. All patients in group B had a significantly lower myelin volume in the ipsilateral hemisphere ($P < 0.05$) and a lower MVF and R1 and higher PD in the ipsilateral WM than on the contralateral side ($P < 0.0125$).

Higher estimated myelin was observed on the ipsilateral side in some patients aged ≤ 2 years and lower myelin on the ipsilateral side in all older patients. Synthetic qMRI might be useful for showing myelin-related abnormalities in SWS [2)].

1)

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2)

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