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Choroid Plexus Size in Young Children with Sturge-Weber Syndrome

Paul D. Griffiths, Susan Blaser, Mitra B. Boodram, Derek Armstrong, and Derek Harwood-Nash

PURPOSE: To assess the size of the choroid plexus in young children with unilateral and bilateral Sturge-Weber syndrome. **METHODS:** Subjects included 15 children 4 years old or younger with Sturge-Weber syndrome. Eleven cases were unilateral and four were bilateral. Unilateral or bilateral involvement was determined by the distribution of abnormal leptomeningeal enhancement on MR images. The diameters of the choroid plexus were measured on contrast-enhanced axial MR images. The choroid plexus of the affected and unaffected sides in these cases were compared with those of 15 age-matched children without Sturge-Weber syndrome who were being examined for seizures. **RESULTS:** Our results show a wide variation in the size of the choroid plexus in children with Sturge-Weber syndrome; however, plexus associated with a hemisphere affected by Sturge-Weber syndrome were significantly larger than those on the unaffected side and in the age-matched control group. The size of the choroid plexus was positively correlated with the extent of leptomeningeal involvement as demonstrated by abnormal contrast enhancement. **CONCLUSION:** The choroid plexus is enlarged early in the course of Sturge-Weber syndrome in both unilateral and bilateral cases. There is a positive correlation between choroid plexus size and extent of leptomeningeal involvement in children with Sturge-Weber syndrome.

Index terms: Phakomotoses; Choroid plexus, magnetic resonance; Children, diseases

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Sturge-Weber syndrome is generally diagnosed on clinical grounds by the association of vascular nevi in the territory of the trigeminal nerve and by such signs as seizures, hemiparesis, and hemianopia. Typical imaging features are superficial cerebral calcification, atrophy, and leptomenigeal enhancement (1). Other features include persistence of prominent deep medullary and subependymal veins and enlarged deep venous structures (2).

Several articles have described enlargement and calcification of the choroid plexus occurring on the same side as the hemisphere affected by Sturge-Weber syndrome (3–6). Radiologic reports of MR studies of Sturge-Weber syndrome usually describe only a few cases, and patients are of widely differing ages; moreover, assessments of the size of the choroid plexus are usually subjective and appear without statistical evaluation (6–9). Few authors have compared the size of the choroid plexus in patients with Sturge-Weber syndrome with that in age-matched control groups (4).

The purpose of this study was to assess the size of the choroid plexus in young children with unilateral and bilateral Sturge-Weber syndrome and to compare those sizes with the unaffected side and with the choroid plexus in age-matched patients with seizures but without Sturge-Weber syndrome. In particular, we wished to test our impression, from clinical imaging cases, that the size of the choroid plexus is larger in patients who have more extensive leptomenigeal involvement.

Methods

From 1991 to 1994, 15 children with Sturge-Weber syndrome were examined with the use of contrast-enhanced magnetic resonance (MR) imaging. The children

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were 4 years old or younger at the time of imaging, and four patients had bilateral intracranial involvement. In all cases Sturge-Weber syndrome was diagnosed on clinical grounds by means of criteria described by Roach (10). Computed tomography (CT) was also performed in each case. Sturge-Weber syndrome was determined to be unilateral or bilateral on the basis of the distribution of contrast enhancement on MR images. All children who were classified as having unilateral disease had only contralateral signs and symptoms, and all children with bilateral disease had bilateral neurologic problems. To compare our patients with an age-matched control group, we studied the choroid plexus in 15 children without Sturge-Weber syndrome who were being treated for seizure disorders; these children were subsequently shown to have no anatomic abnormalities.

MR imaging was performed on a 1.5-T Magnetom SP4000 unit (Siemens, Iselin, NJ). Typical examinations and sequences included T1-weighted images obtained at 550/12 (repetition time/echo time) in the three orthogonal planes, axial T2-weighted images at 2800/120, and axial proton density–weighted images at 2800/30. After injection of 0.5 mmol/mL gadopentate dimeglumine (Berlex, Canada) at a dose of 0.2 mL/kg, further axial and sagittal T1-weighted images were obtained.

The imaging studies were reviewed by three neuroradiologists and a consensus opinion was reached as to the extent of leptomenigeal disease and atrophy. The maximum width of the glomus of the choroid plexus on postcontrast T1-weighted axial MR images was measured independently by the three investigators.

Data Handling and Statistics

Interobserver variability in determinations of choroid plexus size was tested for by using the method for continuous measurements described by Brennan and Silman (11). By plotting the difference between two observers' readings against the mean size of the choroid plexus measurements, we could show that the level of precision was not related to the size of the choroid plexus. Therefore, all the measurements could be assessed together. Each observer measured 60 choroid plexus (11 patients with unilateral Sturge-Weber syndrome, 4 patients with bilateral Sturge-Weber syndrome, and 15 control subjects). The mean difference between the measurements of two observers and the standard deviation (SD) of the differences were calculated. The 95% confidence interval was calculated from the difference plus or minus 2 SD. The procedure was repeated for all three combinations of observers.

The diameters of the choroid plexus in the patients with unilateral Sturge-Weber syndrome were used as paired case measurements because of the availability of an internal control; namely, the opposite side. The mean difference in size was calculated along with 95% confidence intervals and the *t*-statistic. The size of the choroid plexus in the control subjects was compared with that on the unaffected side in the patients with unilateral Sturge-Weber syndrome. The size of the choroid plexus in the patients with bilateral disease was compared with that on the affected side in the patients with unilateral disease. The size of the choroid plexus on the affected side in patients with unilateral disease and that on both sides in patients with bilateral disease were correlated with the degree of cortical involvement as determined by the number of anatomic lobes that showed leptomeningeal enhancement on MR images within the abnormal hemisphere.

Results

Summaries of the clinical and MR findings are given in Tables 1A and 1B. The median age of the children with Sturge-Weber syndrome at the time of imaging was 9 months (range, 2 to 48 months). The median age at the time of the first seizure for patients with unilateral Sturge-Weber syndrome was 9 months (range, 1.5 to 13 months); for the patients with bilateral disease, the median age was 2.5 months (range, 2 to 6 months). Two children with unilateral disease had no facial nevi and two other children with unilateral brain involvement and signs had bilateral facial nevi. Cases of Sturge-Weber syndrome such as these have been described by Roach and are well recognized (10). The 11 unilateral and 4 bilateral cases of Sturge-Weber syndrome provided information on 19 affected hemispheres and the associated choroid plexus. The extent of leptomenigeal involvement was assessed by using contrast-enhanced MR imaging, which is considered the most sensitive imaging technique for this disease (4). Of the 19 affected hemispheres 3 had one lobe involved, 3 had two lobes involved, 8 had three lobes involved, and 5 had all 4 lobes involved. The occipital lobes were most commonly affected (18/19), followed by the parietal (14/19), frontal (9/19), and temporal (12/19) lobes.

The level of interobserver agreement for choroid plexus measurement was good. For example, when the measurements of two of the observers were compared, the mean difference was 0.4 mm (SD = 0.3 mm). This produced a 95% confidence interval of 1.0 mm to −0.2 mm, which indicates good agreement. Similar results were obtained for the other observer combinations, with zero included in both 95% confidence intervals.

The choroid plexus of the children without Sturge-Weber syndrome showed no significant size variation between sides and had a mean diameter of 4.0 mm. This was comparable to the size of the choroid plexus in the unaffected hemisphere of children with unilateral Sturge-

TABLE 1A: Clinical and MR imaging summary of 11 cases of unilateral Sturge-Weber syndrome

Case	Age at First Seizure, mo	Age at CT, mo	Facial Nevus Distribution	Other Clinical Findings	Cortical Involvement
1	13	13	Left V_1, V_2	Right homonymous hemianopia	PTO
2	9	46	Bilateral V_1, V_2, V_3; Left > Right	Right hemiparesis and hemiatrophy	P
3	13	18	Right V_1	. . .	TO
4	6	17	Left V_1	Right homonymous hemianopia	TO
5	4	12	Right V_1, V_2, V_3	Left hemiparesis and hemianopia	FPTO
6	10	11	None	. . .	O
7	7	10	Bilateral V_1; Left = Right	Left hemiparesis	FPO
8	12	39	Left V_1	Right homonymous hemianopia	PTO
9	4	7	Right V_1	Left hemiparesis	FPTO
10	12	48	Right V_1, V_2	Left hemiparesis, speech difficulties	PTO
11	1.5	12	None	Right homonymous hemianopia	FPO

Note.—P = parietal, T = temporal, O = occipital, F = frontal.

TABLE 1B: Clinical and MR imaging summary of four cases of bilateral Sturge-Weber syndrome

Case	Age at First Seizure, mo	Age at MR Imaging, mo	Facial Nevus Distribution	Other Clinical Findings	Cortical Involvement
12	1.5	4	Right V_1, V_2; Left V_1, V_2	Right hemiparesis Severe developmental delay	Right: FO Left: FPTO
13	2	5	Bilateral V_1, V_2, V_3	Left hemiparesis Poor language development	Right: FPTO Left: O
14	2	2	Bilateral V_1, V_2, V_3	Severe developmental delay	Right: FPTO Left: PTO
15	5	8	Right V_1; Left V_1, V_2, V_3	Severe developmental delay	Right: FPO Left: PTO

Note.—F = frontal, P = parietal, T = temporal, O = occipital.

Weber syndrome (Table 2). Comparison of the choroid plexus sizes between the normal and abnormal sides in the children with unilateral Sturge-Weber syndrome (Fig 1) showed a significant enlargement on the affected side (mean difference in size = 4.4 mm, 95% confidence interval = 2.8 to 6.0 mm, *t*-statistic <.01).

Similar changes were seen in the choroid plexus of the children with bilateral disease (Fig 2). Figure 3 shows a trend toward larger choroid plexus with increasing cortical involvement and a similar correlation was found with the extent of atrophy. Cysts (defined as centrally situated, unenhancing, rounded areas) within the choroid plexus were also common, occurring in 11 (58%) of the 19 affected hemispheres (Fig 1

TABLE 2: Mean measurements of the transverse diameters of the glomi of the choroid plexus in patients with Sturge-Weber syndrome and in age-matched patients with seizures only

	No. of Choroid Plexus Measured	Mean Size, mm	Standard Deviation, mm
Patients with seizures only	30	4.0	0.5
Unilateral Sturge-Weber syndrome, unaffected side	11	5.3	0.5
Unilateral Sturge-Weber syndrome, affected side	11	9.7	2.5
Bilateral Sturge-Weber syndrome	8	10.8	2.6

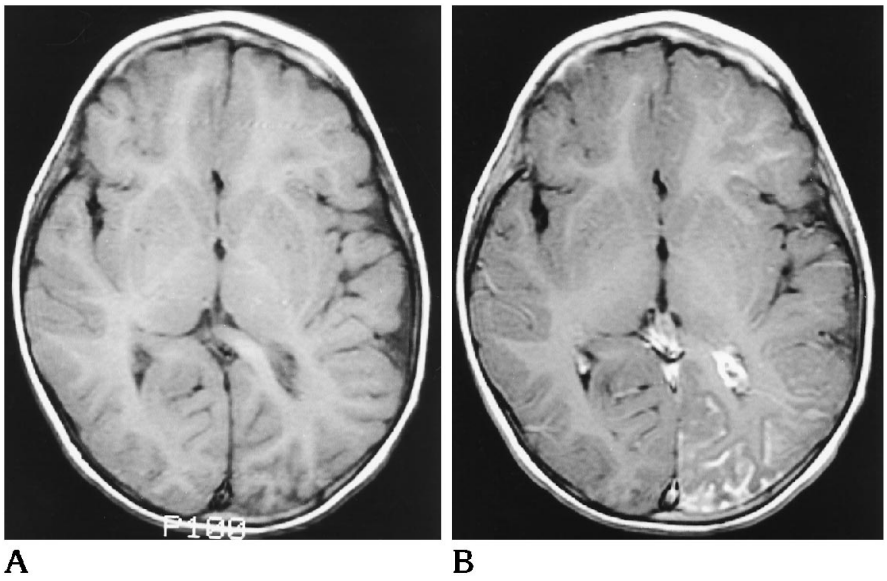

Fig 1. *A* and *B*, Unenhanced (*A*) and enhanced (*B*) T1-weighted axial MR images of a patient with unilateral Sturge-Weber syndrome with a small volume of leptomenigeal involvement. Enhancement is present in the meninges related to the occipital and parietal lobes (not shown). The ipsilateral choroid plexus shows modest enlargement compared with the normal side. Note the small central cyst in the left choroid plexus.

and 4). All the children in this study also had had a recent unenhanced CT examination. Calcification was not present in the choroid plexus in any of these cases. Similar appearances have been described in other MR imaging reports (8, 9).

Discussion

Although typically unilateral, Sturge-Weber syndrome occurs bilaterally in a significant portion of cases, and children with bilateral involvement tend to have severe developmental delay as well as focal neurologic deficits. Our study population of 15 children included four cases of bilateral disease (27%), and a recent review of 40 cases of Sturge-Weber syndrome described bilateral involvement in 33% of the subjects (12). Enlargement of the choroid plexus is a recognized feature of Sturge-Weber syndrome and has been attributed to angiomatosis (3). Choroid plexus enlargement in patients with Sturge-Weber syndrome was confirmed in vivo soon after the advent of CT (13, 14). These early CT reports described enlarged, enhancing choroid plexus that were often calcified and were thought to be consistent with choroid angiomas. Stimac et al (4) produced data of choroid plexus size in control subjects of different

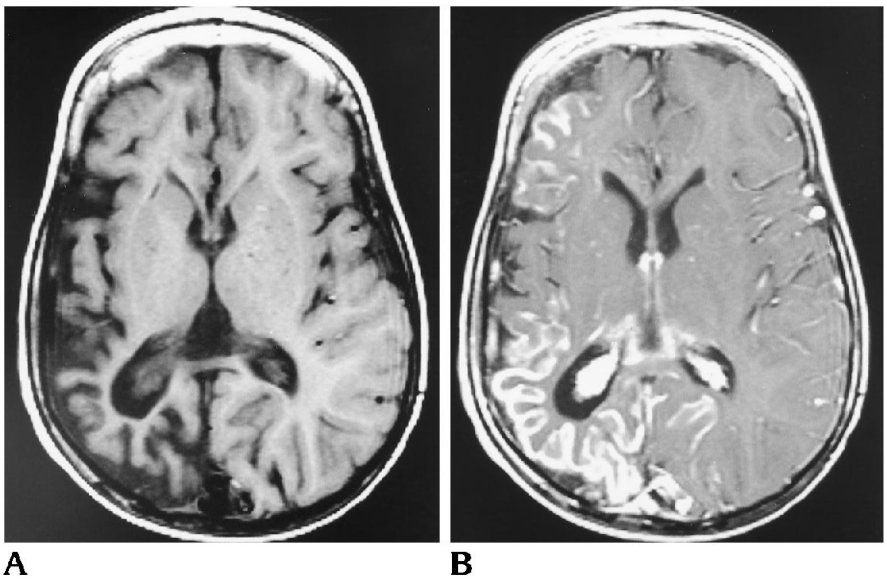

Fig 2. *A* and *B*, Unenhanced (*A*) and enhanced (*B*) T1-weighted axial MR images in a patient with bilateral Sturge-Weber syndrome with extensive leptomeningeal involvement of the right hemisphere and only occipital involvement on the left. The right choroid plexus is considerably larger than the left.

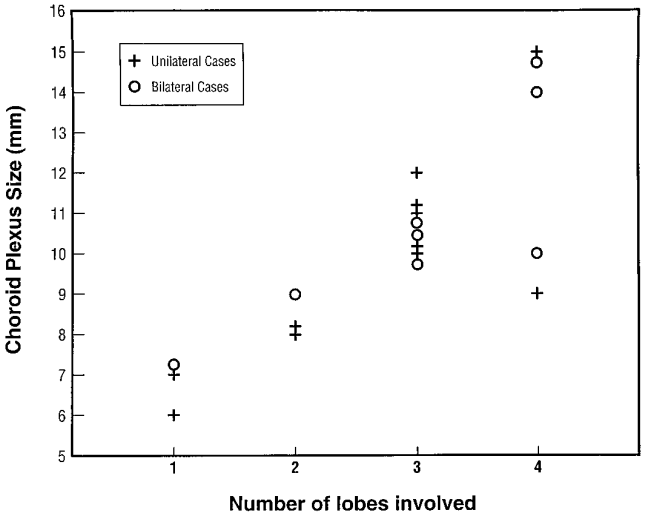

Fig 3. Graph shows the relationship between choroid plexus size and extent of cortical involvement represented by abnormal contrast enhancement on MR images. Unilateral (+) and bilateral (*O*) cases are shown. There is a trend toward larger choroid plexus with increasing cortical involvement.

ages as measured with the use of CT. In that study, none of the 10 patients younger than 5 years old received contrast material, so the dimensions of their choroid plexus were not measured. Fourteen children ages 6 to 10 years old had a mean choroid plexus diameter of 2.5 mm, with no significant side-to-side variation. Calcification was seen in only one case in this age group. In four patients with unilateral Sturge-Weber syndrome who were younger than 10 years old, the difference between the two sides was 4.0, 7.5, 0, and 0 mm, respectively. These authors suggested that enlargement of the choroid plexus may be the only sign of intracranial involvement of Sturge-Weber syndrome.

In a recent study of 14 children with Sturge-Weber syndrome who were examined with contrast-enhanced CT, the choroid plexus were considered to be enlarged in 50% of the cases (5). Wasenko et al (6) used CT to examine four children with Sturge-Weber syndrome who were 7 to 16 years old. The affected glomera of the choroid plexus measured 7, 8, 9 and 10 mm, respectively, and three of four choroid plexus were considered to be enlarged. Choroid plexus enlargement has also been seen on contrast-enhanced MR images. Most of the MR studies involved a small number of subjects, but among them, enlargement was found in four of five cases (7), in five of seven cases (8), and in two of four cases (6, 9). These reports did not present any measurements or statistics and did not include age-matched control groups.

Our results confirm that the choroid plexus of hemispheres affected by Sturge-Weber syndrome are enlarged at an early age, and this finding is substantiated by statistical analysis. We have also shown that the size of the choroid plexus in the uninvolved hemisphere in patients with unilateral Sturge-Weber syndrome is not significantly different from that in the hemispheres of age-matched control subjects. There is a positive correlation between the extent of leptomeningeal involvement and the size of the choroid plexus. This may be interpreted as se-

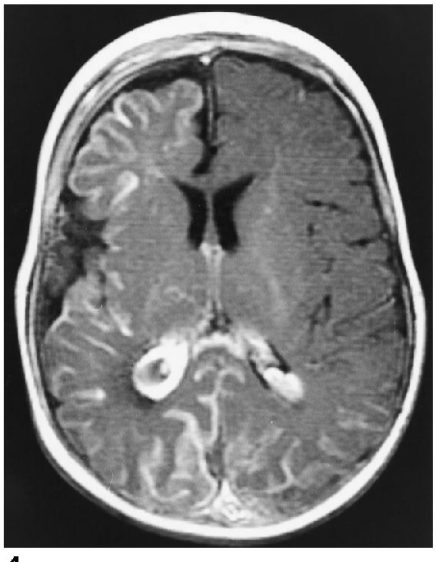

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Fig 4. Enhanced T1-weighted axial MR image of a patient with bilateral Sturge-Weber syndrome. Note the large central cyst in the right choroid plexus.

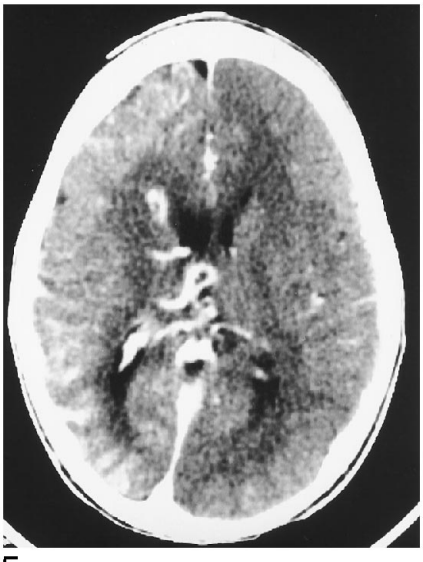

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Fig 5. Enhanced axial CT scan of a patient with unilateral Sturge-Weber syndrome shows enlarged deep venous structures including the atrial vein draining the right choroid plexus.

vere choroidal angiomatosis, as has been suggested by some authors (3).

We speculate on another possible cause for the choroid plexus enlargement; namely, that the extent of leptomeningeal angiomatosis is most likely related to the lack of superficial cortical venous drainage. In cases of widespread hemispheric disease, there may be greater deep venous diversion, which will raise the deep venous pressure on the affected side. The choroid plexus is drained by the atrial vein and dilatation of this vessel is frequently seen in cases of severe hemispheric Sturge-Weber syndrome. An example of the marked deep vascular engorgement can be seen on contrast-enhanced CT scans (Fig 5). It is possible that vascular engorgement of the choroid plexus contributes to the increase in size of the choroid plexus in Sturge-Weber syndrome. This would explain the positive correlation between choroid plexus size and the extent of leptomeningeal disease found in our study. This explanation has been suggested previously (8) on the basis of vascular engorgement seen on imaging studies. However, deep venous flow in the affected and unaffected hemispheres in patients with Sturge-Weber syndrome remains to be evaluated.

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