MR imaging of Sturge-Weber syndrome: role of gadopentetate dimeglumine and gradient-echo techniques.

A D Elster and M Y Chen

AJNR Am J Neuroradiol 1990, 11 (4) 685-689
http://www.ajnr.org/content/11/4/685

MR Imaging of Sturge-Weber Syndrome: Role of Gadopentetate Dimeglumine and Gradient-Echo Techniques

Allen D. Elster[1]
Michael Y. M. Chen

Five patients with Sturge-Weber syndrome were evaluated by conventional noncontrast spin-echo MR imaging, a gradient-recalled echo (GRE) technique, and T1-weighted spin-echo imaging after administration of gadopentetate dimeglumine. In four of five cases the full extent of intracranial disease was appreciated only on the postcontrast images. In one patient precontrast and GRE images were entirely normal, while only the postcontrast study demonstrated extensive involvement of both brain and retina. Nevertheless, some abnormal vessels with higher flows were seen better on precontrast T2-weighted images than on postcontrast T1-weighted images. GRE techniques demonstrated calcifications to best advantage, in one case even better than on CT.

Contrast enhancement with gadopentetate dimeglumine is necessary for the complete MR evaluation of patients with suspected Sturge-Weber syndrome. Traditional noncontrast T2-weighted and GRE images may provide additional complementary information.

***AJNR* 11:685–689, July/August 1990**

Sturge-Weber syndrome (encephalotrigeminal angiomatosis) is a phakomatosis characterized by a facial vascular nevus in the territory of the trigeminal nerve together with a variety of CNS manifestations, including seizures, dementia, hemiplegia, hemianopsia, buphthalmos, and glaucoma [1–7]. Cerebral cortical calcifications, atrophy, venous engorgement, choroid plexus enlargement, and leptomeningeal angiomatosis have been well demonstrated by conventional radiologic techniques, including MR imaging [4–7]. However, the precise role of newer MR techniques (gadopentetate dimeglumine enhancement and gradient echo) in these patients has not yet been fully evaluated. We therefore set out to use these new techniques to study prospectively a group of Sturge-Weber patients and to compare these findings with those obtained with the use of conventional spin-echo imaging without contrast.

Subjects and Methods

Five patients with clinically definite Sturge-Weber syndrome were evaluated by MR imaging over a 6-month period (Table 1). The subjects, four males and one female, ranged in age from newborn to 35 years. All patients had cutaneous vascular nevi involving the face and presented with seizures.

MR imaging was performed exclusively at 1.5 T (Picker Vista MR, Highland Heights, OH). For pediatric patients, parental consent was obtained, as was approval of the Institutional Review Board Human Practices Committee. All patients were evaluated by a neurologist before and after contrast administration.

Precontrast spin-echo (SE) and gradient-recalled echo (GRE) sequences were performed. Precontrast T2-weighted images, 2500/60–80/2 (TR/TE/excitations), were done exclusively with SE. Other variables included field of view = 24 cm, slice thickness = 5 mm, slice gap = 1 mm, and matrix = 192 × 256. Precontrast T1-weighted SE images (600/20/2) were obtained with other parameters similar to the T2-weighted sequence. To heighten our sensitivity to calcifications, GRE imaging was also performed [8]. The GRE technique used

Received January 19, 1990; revision requested March 11, 1990; revision received March 20, 1990; accepted March 20, 1990.

[1] Both authors: Department of Radiology, Bowman Gray School of Medicine, Wake Forest University, 300 S. Hawthorne Rd., Winston-Salem, NC 27103. Address reprint requests to A. D. Elster.

0195-6108/90/1104-0685

TABLE 1: Clinical and Radiologic Data on Sturge-Weber Patients

Case No.	Age	Sex	Symptoms and Signs	Radiologic Findings
1	35 yr	F	Seizures, headaches, decreased vision, retinal angioma, nevus in right V_1	Noncontrast CT and MR normal; contrast MR shows right occipital angiomatosis and retinal angioma (Fig. 1)
2	6 yr	M	Seizures, nevus in left V_1 and V_2	Extensive left hemisphere involvement, thick meninges (Fig. 2)
3	8 yr	M	Seizures, decreased vision, glaucoma, nevus in left V_1 and V_2	Right hemisphere atrophy, enhancement bilateral choroid angiomas, disordered veins (Fig. 3)
4	8 days	M	Seizures, right arm hypertrophy, extensive nevus of face, chest, and arms (Klippel-Trenaunay syndrome)	Extensive meningeal and choroid plexus enhancement, right greater than left
5	4 yr	M	Seizures, mental retardation, nevus in bilateral V_1 distribution	Bilateral cerebral calcifications shown best on GRE MR; extensive contrast enhancement shown equally well by CT and MR (Fig. 4)

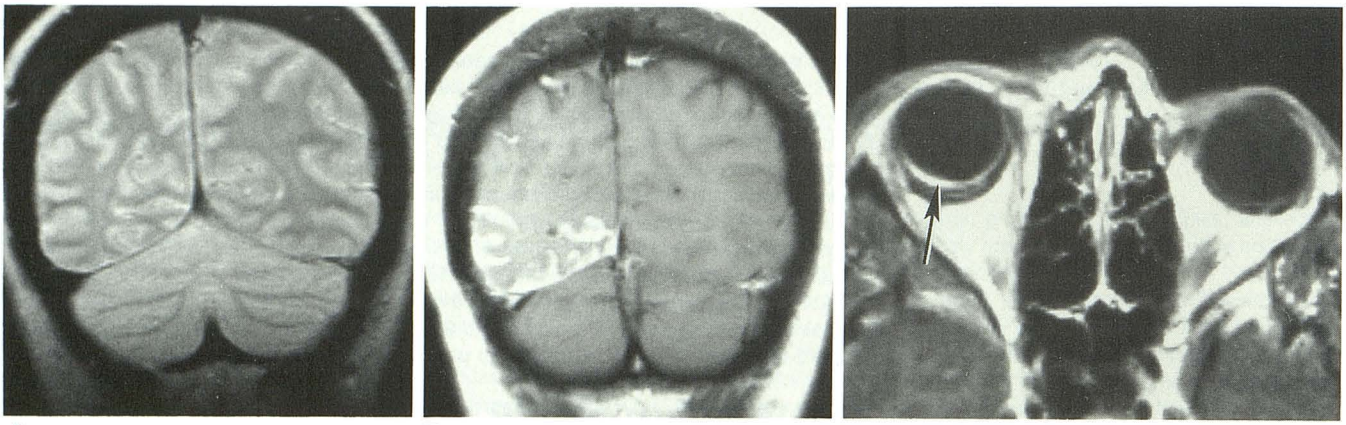


Fig. 1.—Value of gadopentetate dimeglumine enhancement in delineating brain disease is most evident in case 1.
***A*, Precontrast MR study is entirely normal, including this representative T2-weighted coronal image.**
***B*, Postcontrast MR image shows extensive leptomeningeal enhancement in posterior right cerebral hemisphere.**
***C*, Also noted is ipsilateral retinal angioma, confirmed funduscopically (*arrow*).**

was a multisection T2*-weighted sequence (300/18/4) with flip angle = 30°. Gradient-moment nulling (MAST) was also used on the T2-weighted SE and GRE sequences to reduce motion artifacts and improve image quality [9].

Gadopentetate dimeglumine (Magnevist, Berlex, Cedar Knolls, NJ) was administered by slow IV infusion at a dose of 0.1 mmol/kg. T1-weighted coronal and axial images were obtained beginning approximately 3–5 minutes after contrast infusion. The pre- and postcontrast studies were then critically evaluated by an experienced neuroradiologist to assess the role of each MR technique in defining and characterizing the full extent of intracranial disease in patients with Sturge-Weber syndrome. In four cases CT scans were also available for comparison, but in only two of these were both pre- and postcontrast studies obtained.

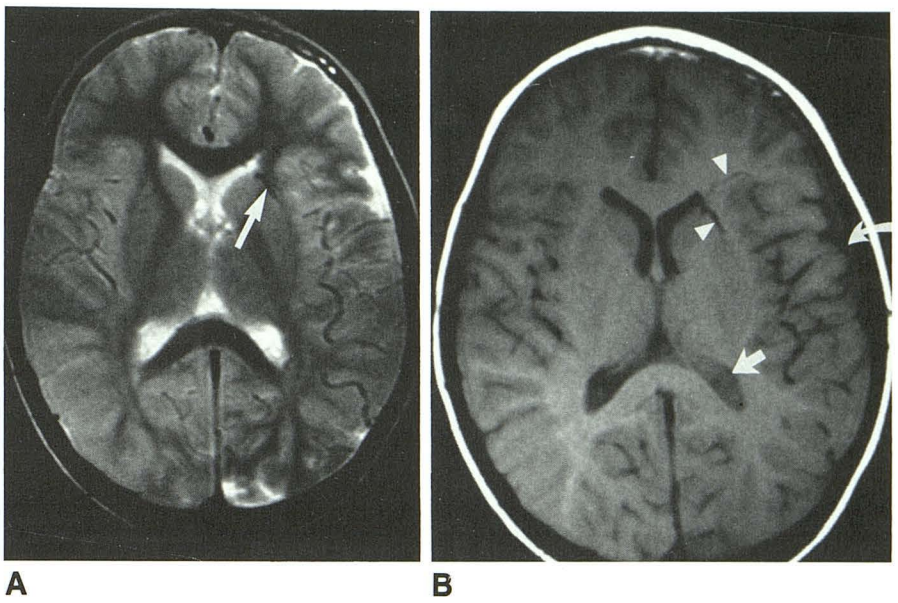
Results

The clinical history, physical signs, and radiologic findings for the five patients are recorded in Table 1. Illustrative images are presented in Figures 1–4.

Clinical findings were typical of those reported in other series of Sturge-Weber patients [3–7]. All five of our patients presented with seizures, either generalized (n = 2) or partial (n = 3). Two patients had decreased visual acuity, one with glaucoma and one with a retinal angioma. All patients had cutaneous vascular nevi involving the ophthalmic division (V_1) of the trigeminal nerve. Two patients also had involvement of the maxillary division (V_2), while two had bilateral facial nevi. One patient who had an extensive nevus involving the head, trunk, and arms, with hypertrophy of the right upper extremity, had coexistent Sturge-Weber and Klippel-Trenaunay syndromes [10, 11].

Intracranially, we saw a spectrum of findings as classically described in the CT and precontrast MR literature. These included calcifications (n = 3) detected best by GRE, hemiatrophy (n = 3), gyriform enhancement (n = 5), choroid plexus enlargement (n = 4), meningeal thickening (n = 2), and disordered venous drainage (n = 3). Bilateral intracranial involvement was noted in both patients with bilateral nevi; a third patient with a unilateral nevus (case 3) also had bilateral intracranial involvement.

Contrast enhancement was believed to be necessary to characterize completely the intracranial involvement in all cases. The most striking example can be seen in case 1, in which the precontrast MR study was completely normal (Fig.



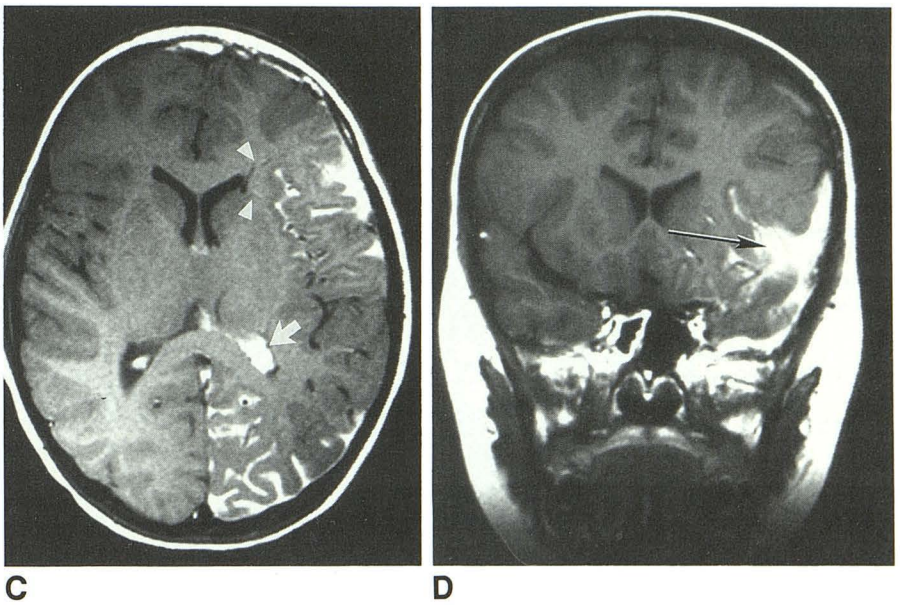


Fig. 2.—Case 2.

A, Precontrast T2-weighted MR image shows focal atrophic changes in left frontal and occipital lobes. At least one abnormal deep vessel is identified (*arrow*).

B, Precontrast T1-weighted MR image shows abnormal deep vessels to greater advantage (*arrowheads*). Note also the slightly higher signal intensity in left atrium (*straight arrow*) and in left sylvian fissure (*curved arrow*).

C, Postcontrast T1-weighted axial MR image shows extensive involvement of left hemisphere, including left choroid plexus (*arrow*). Note that visualization of abnormal deep vessels (*arrowheads*) has not been aided by contrast administration.

D, Coronal postcontrast MR image shows extensive leptomeningeal thickening (*arrow*), which accounted for the abnormal signal noted previously in sylvian fissure.

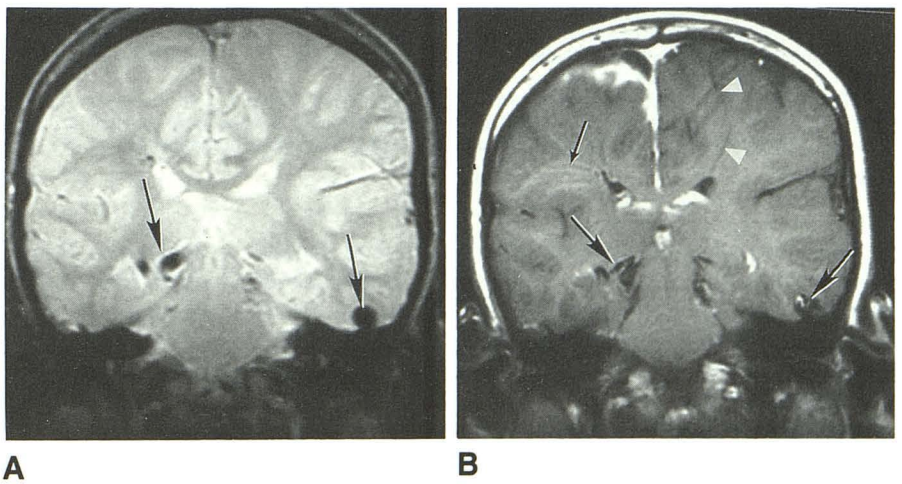


Fig. 3.—Pre- and postcontrast MR studies may provide complementary information (case 3).

A, Precontrast T2-weighted image shows right cerebral hemiatrophy, despite the fact that the bulk of the cutaneous nevus was on left side of face. Several enlarged veins are noted (*arrows*).

B, Postcontrast image shows leptomeningeal thickening and enhancement over right convexity. Abnormal enhancement in tiny deep medullary veins on the right is now appreciated (*small arrow*). Because of higher flow rates the previously noted large veins enhance inconsistently (*large arrows*). Another anomalous vein ascending from left ventricle is not opacified with contrast (*arrowheads*).

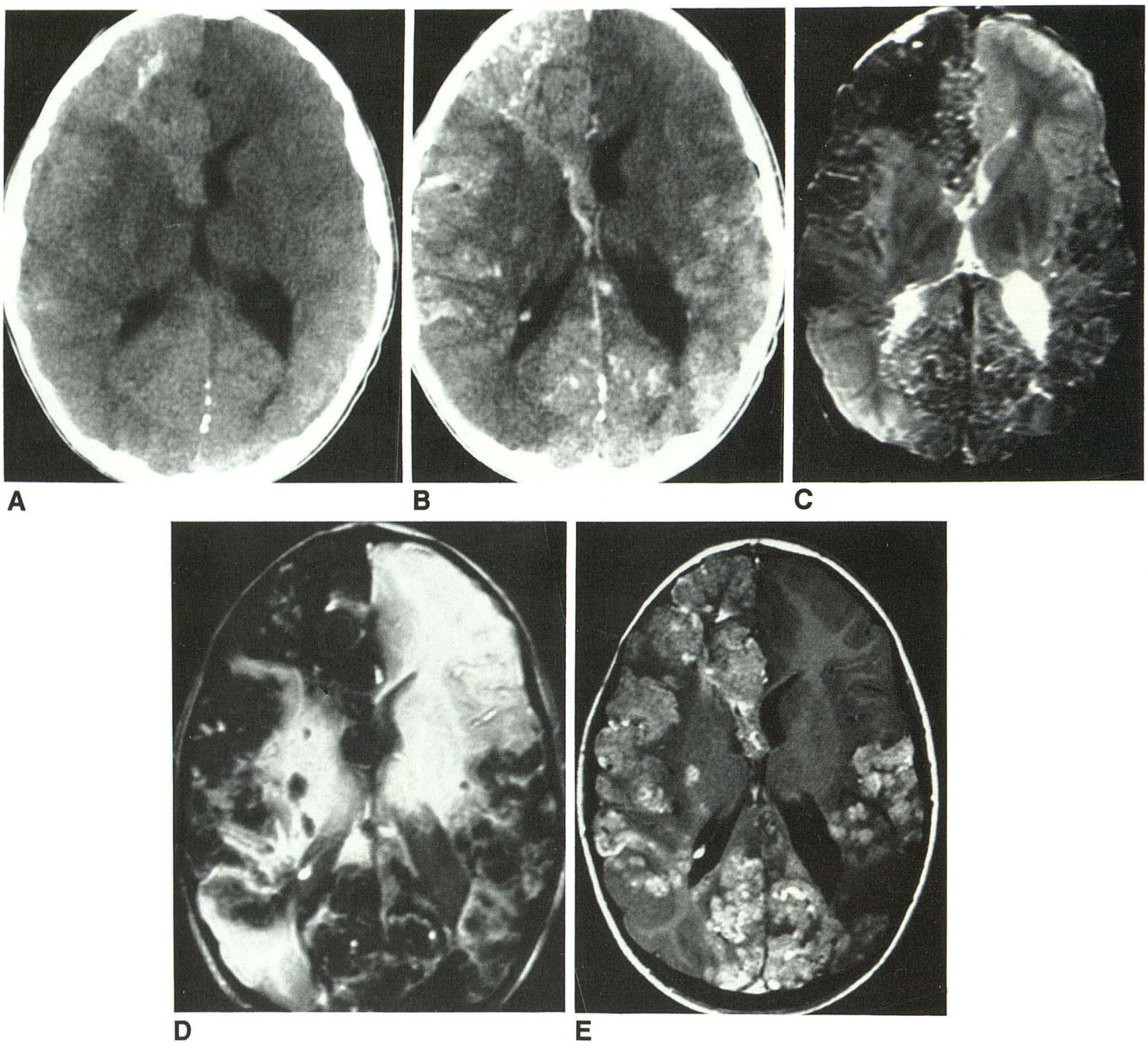


Fig. 4.—Comparative roles of CT, MR, contrast-enhanced MR, and GRE techniques (case 5).

A, Precontrast CT scan shows bilateral cerebral involvement with punctate calcifications as well as diffuse parenchymal regions of subtly higher attenuation. The full extent of disease is difficult to define precisely.

B, Postcontrast CT scan reveals an extensive abnormality.

C, Precontrast T2-weighted MR image.

D, Precontrast GRE MR image shows extensive regions of low signal in both hemispheres, presumably caused by susceptibility effects of tissue microcalcifications. Note that the GRE MR demonstrates the extent of disease more convincingly than does the noncontrast CT scan.

E, Postcontrast MR image shows congruent enhancement in the regions that enhanced on CT.

1). Here, not only were intracranial meningeal and choroid plexus involvement noted, but also occult angiomatosis of the retina. Contrast enhancement was perhaps least helpful in case 5 (Fig. 4), in which precontrast GRE MR images revealed abnormalities in all areas where contrast enhancement was later noted.

Despite the importance of contrast enhancement, not every abnormality was apparent on postcontrast T1-weighted images alone. Some abnormal vessels, with presumably more rapid flow, did not enhance uniformly with gadopentetate dimeglumine (Figs. 2 and 3). Precontrast T2-weighted images were better for defining these abnormal vessels.

Calcification, present in three cases, was most easily identified with a GRE technique. Figure 4 illustrates how GRE may be even more definitive than CT in detecting small parenchymal calcifications in Sturge-Weber syndrome.

Discussion

Sturge-Weber syndrome is a phakomatosis characterized by a cutaneous vascular nevus and angiomatosis of the leptomeninges. The syndrome was initially described by Sturge in 1879 [1], while Weber in 1922 first noted the characteristic intracranial calcifications on skull X ray [2]. Since that time, details of the syndrome and its variants have been elucidated. An overlap with the Klippel-Trenaunay syndrome has been reported [11]. These latter patients frequently have an extensive corporal nevus and hemihypertrophy, as well as the characteristic intracranial involvement.

Intracranially, patients with Sturge-Weber syndrome consistently have venous angiomatosis of the leptomeninges. The meningeal abnormality is found most commonly in the anterior occipital lobe, with variable extension to the posterior parietal and temporal lobes [3]. The intracranial lesion is usually ipsilateral to the nevus, but contralateral and bilateral lesions have been described [12]. Calcifications are frequently noted in the brain parenchyma adjacent to the leptomeningeal abnormality. Pathologically, these calcifications occur in a pericapillary distribution, usually in the fourth layer of the atrophied cerebral cortex, and are thought to be related to chronic tissue hypoxia [3].

Traditional MR imaging in cases of Sturge-Weber syndrome has been well described [4–7]. Noncontrast MR images may reveal hemiatrophy, angiomatous malformations of the choroid plexuses, and signal changes in the underlying brain to good advantage. Early investigators [6, 7] stressed the continued importance of obtaining pre- and postcontrast CT examinations in Sturge-Weber patients to detect calcifications and leptomeningeal angiomas.

Now, with the availability of GRE techniques and paramagnetic contrast agents, the limitations of MR relative to CT in evaluating Sturge-Weber syndrome raised by previous authors [6, 7] may no longer apply. With such a small number of patients, strong conclusions about the relative roles of CT and MR in evaluating Sturge-Weber syndrome cannot be supported dogmatically. It is clear, however, that GRE may be superior to CT for detecting microcalcifications, illustrated in Figure 4 and demonstrated in other diseases by Atlas et al. [8]. Nevertheless, even our small series has shown that GRE and gadopentetate dimeglumine enhancement add significantly to the precontrast MR diagnosis. Without these techniques, precontrast MR alone would have significantly underestimated the extent of intracranial disease in at least four of our five cases.

Conversely, contrast-enhanced T1-weighted images may also be insufficient by themselves to characterize the full extent of intracranial disease. While leptomeningeal anomalies are clearly demonstrated, not all of the disordered veins may show enhancement because of flow-related signal loss (Figs. 2 and 3). Traditional T2-weighted noncontrast SE images may reveal these vessels to better advantage than either GRE or postcontrast T1-weighted images.

In conclusion, the use of both contrast enhancement and GRE techniques considerably improves the delineation of intracranial abnormalities in patients suspected of having Sturge-Weber syndrome. Nevertheless, noncontrast T2-weighted images still provide complementary information and should continue to be part of the routine MR examination.

REFERENCES

1. Sturge WA. A case of partial epilepsy apparently due to a lesion of one of the vaso-motor centers of the brain. *Trans Clin Soc London* **1879**;12: 162–167
2. Weber FP. Right-sided hemi-hypertrophy resulting from right-sided congenital spastic hemiplegia, with a morbid condition of the left side of the brain, revealed by radiograms. *J Neurol Psychopathol* **1922**;3:134–139
3. Coulam CM, Brown LR, Reese DF. Sturge-Weber syndrome. *Semin Roentgenol* **1976**;11:55–60
4. Braffman BH, Bilaniuk LT, Zimmerman RA. The central nervous system manifestations of the phakomatoses on MR. *Radiol Clin North Am* **1988**;26:773–800
5. Bilaniuk LT, Zimmerman RA, Hochman M, et al. MR of the Sturge-Weber syndrome (abstr). *AJNR* **1987**;8:945
6. Stimac GK, Solomon MA, Newton TH. CT and MR of angiomatous malformations of the choroid plexus in patients with Sturge-Weber disease. *AJNR* **1986**;7:623–627
7. Chamberlain MC, Press GA, Hesselink JR. MR imaging and CT in three cases of Sturge-Weber syndrome: prospective comparison. *AJNR* **1989**; 10:491–496
8. Atlas SW, Grossman RI, Hackney DB, et al. Calcified intracranial lesions: detection with gradient-echo-acquisition rapid MR imaging. *AJNR* **1988**; 9:253–259
9. Elster AD. Motion artifact suppression technique (MAST) for cranial MR imaging: superiority over cardiac gating for reducing phase-shift artifacts. *AJNR* **1988**;9:671–674
10. Phillips GN, Gordon DH, Martin EC, Haller JO, Casarella W. The Klippel-Trenaunay syndrome: clinical and radiological aspects. *Radiology* **1978**; 128:429–434
11. Deutsch J, Weissenbacher G, Widhalm K, et al. Kombination von Sturge-Weber und Klippel-Trenaunay Syndrome. *Klin Paediat* **1976**;188:464–471
12. Boltshauser E, Wilson J, Hoare RD. Sturge-Weber syndrome with bilateral intracranial calcification. *J Neurol Neurosurg Psychiatry* **1976**;39:429–435