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Abnormal Ocular Enhancement in Sturge-Weber Syndrome: Correlation of Ocular MR and CT Findings with Clinical and Intracranial Imaging Findings

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PURPOSE: To estimate the prevalence of abnormal ocular enhancement in children with Sturge-Weber syndrome as detected with MR imaging and CT and to correlate this with the clinical, fundoscopic, and intracranial imaging findings. **METHODS:** Fifteen children, 4 years old or younger, with Sturge-Weber syndrome were examined with enhanced CT and MR imaging. Eleven children had unilateral intracranial involvement and 4 had bilateral involvement, for a total of 19 abnormal hemispheres and related orbits. The presence of ocular enhancement was compared with the fundoscopic findings independently. Ocular enhancement was correlated with the extent of leptomeningeal disease, the severity of the cutaneous lesion, and the presence of glaucoma by the calculation of likelihood ratios and 95% confidence limits. **RESULTS:** Seven of the 15 patients had abnormal ocular enhancement, which was present in 10 (53%) of the eyes associated with the 19 abnormal hemispheres. MR imaging showed choroidal hemangioma in 7 of 8 patients in whom hemangiomas were shown at fundoscopy. The likelihood of ocular enhancement was increased with the presence of bilateral disease, extensive facial nevi, and glaucoma; there was no significant correlation with the extent of hemispheric involvement. **CONCLUSION:** Both enhanced MR imaging and CT can show diffuse choroidal hemangioma in patients with Sturge-Weber syndrome. However, MR imaging is more sensitive and is recommended to aid in the detection of abnormalities with preventable late complications.

Index terms: Eyes, computed tomography; Eyes, magnetic resonance; Phakomatoses

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Sturge-Weber syndrome is usually diagnosed on clinical grounds by the combination of typical symptoms and nevus flammeus. The cutaneous abnormality is commonly present in the distribution of the trigeminal nerve, usually ophthalmic, but is not essential for diagnosis (1). Computed tomography (CT) and magnetic resonance (MR) imaging features have been reported extensively and reviewed recently by Braffman et al (2). Abnormalities of the ipsilateral eye may occur in Sturge-Weber syndrome. This disease is a cause of buphthalmos, but glaucoma is the most common clinical finding. Retinal and choroidal detachments have been reported (3, 4). Ocular hemangiomas involving the choroid are estimated to occur in approximately one third of cases (5) and these may be seen at fundoscopy. The purpose of this study was to determine whether MR and CT can show this entity reliably and to evaluate the correlation between imaging findings and the fundoscopic diagnosis of diffuse choroidal hemangioma. The practical consequences of the findings are discussed.

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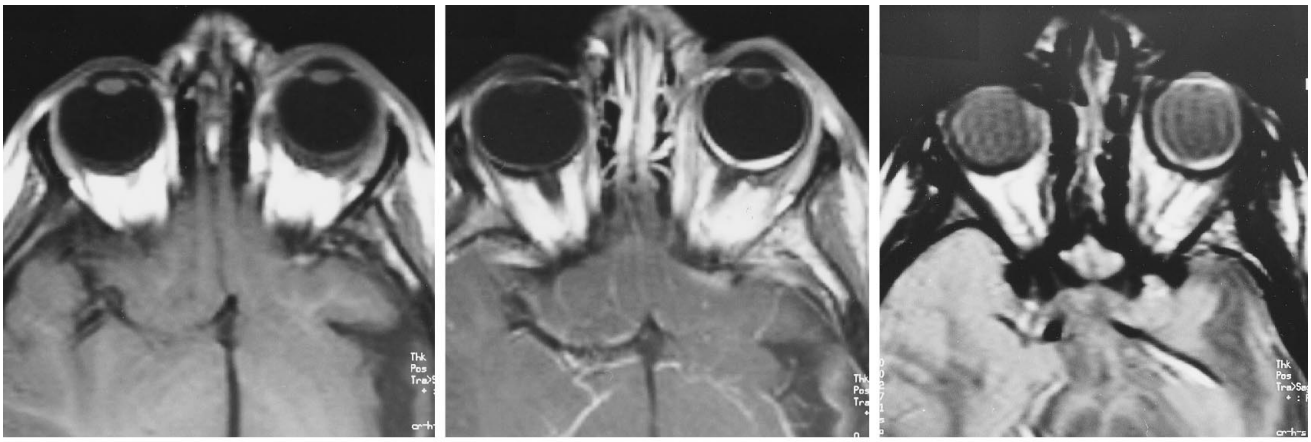

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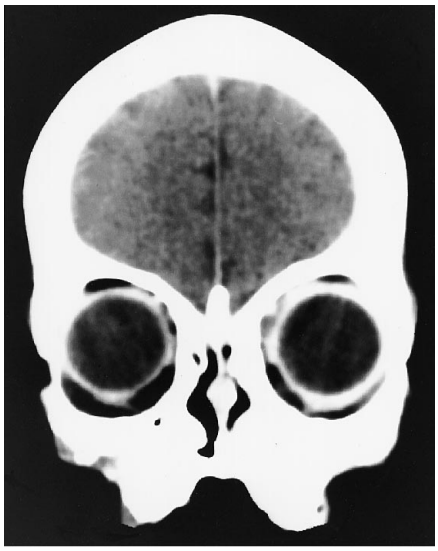

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Fig. 1 Unilateral Sturge-Weber syndrome with left-sided facial nevus, left-sided intracranial involvement, and glaucoma of the left eye. Unenhanced axial T1-weighted image (*A*) shows thickening of the posterior wall of the globe, which has an abnormal high signal on the proton density–weighted image (*C*). Postcontrast image (*B*) shows marked enhancement of the posterior wall of the globe. Coronal postcontrast CT scan (*D*) also shows abnormal thickening and enhancement of the globe. A diffuse choroidal hemangioma was confirmed at fundoscopy.

Materials and Methods

From 1991 to 1994, 15 children, 4 years old or younger, with Sturge-Weber syndrome were examined with contrast-enhanced CT and MR imaging with adequate visualization of the orbits. Eleven children had unilateral intracranial involvement and 4 had bilateral involvement. Among children with unilateral brain involvement and symptoms, 2 had no facial nevi and 2 had bilateral facial nevi. The median age at the time of imaging was 9 months (range, 2 to 48 months). The median age at the time of the first seizure for the children with unilateral involvement was 9 months (range, 1.5 to 13 months); for the children with bilateral involvement, the median age was 2.5 months (range, 2 to 6 months). In all cases, the diagnosis of Sturge-Weber syndrome was made on clinical grounds, with typical cutaneous manifestations together with seizures and neurologic deficits in the majority.

The CT and MR examinations were performed within 3 months of each other in all cases. Axial CT scans (GE 9800 Quick, General Electric, Milwaukee, Wis) were acquired before and after injection of iohexol 300 mg/mL (Winthrop, Aurora, Ontario, Canada) at a dose of 2.5 mL/kg. The orbits were imaged totally in the axial plane and in four cases direct coronal images were also obtained. MR imaging was done on a 1.5-T Magnetom SP4000 unit (Siemens, Iselin, NJ). Typical examinations and sequences included T1-weighted images in the axial and sagittal planes, and T2-weighted and proton density–weighted images in the axial plane. After injection of 0.5 mmol/mL gadopentetate dimeglumine (Berlex, Canada) at a dose of 0.2 mL/kg, further axial and sagittal T1-weighted images were obtained. In one case, postcontrast T1-weighted fat-suppression imaging of the orbits was performed (500/15 [repetition time/echo time], frequency = 300 Hz). The imaging findings were assessed independently by three neuroradiologists. Specific attention was paid to the extent of leptomeningeal enhancement and the presence of ocular enhancement.

Correlation was tested between abnormal ocular enhancement and leptomeningeal involvement, extent of fa-

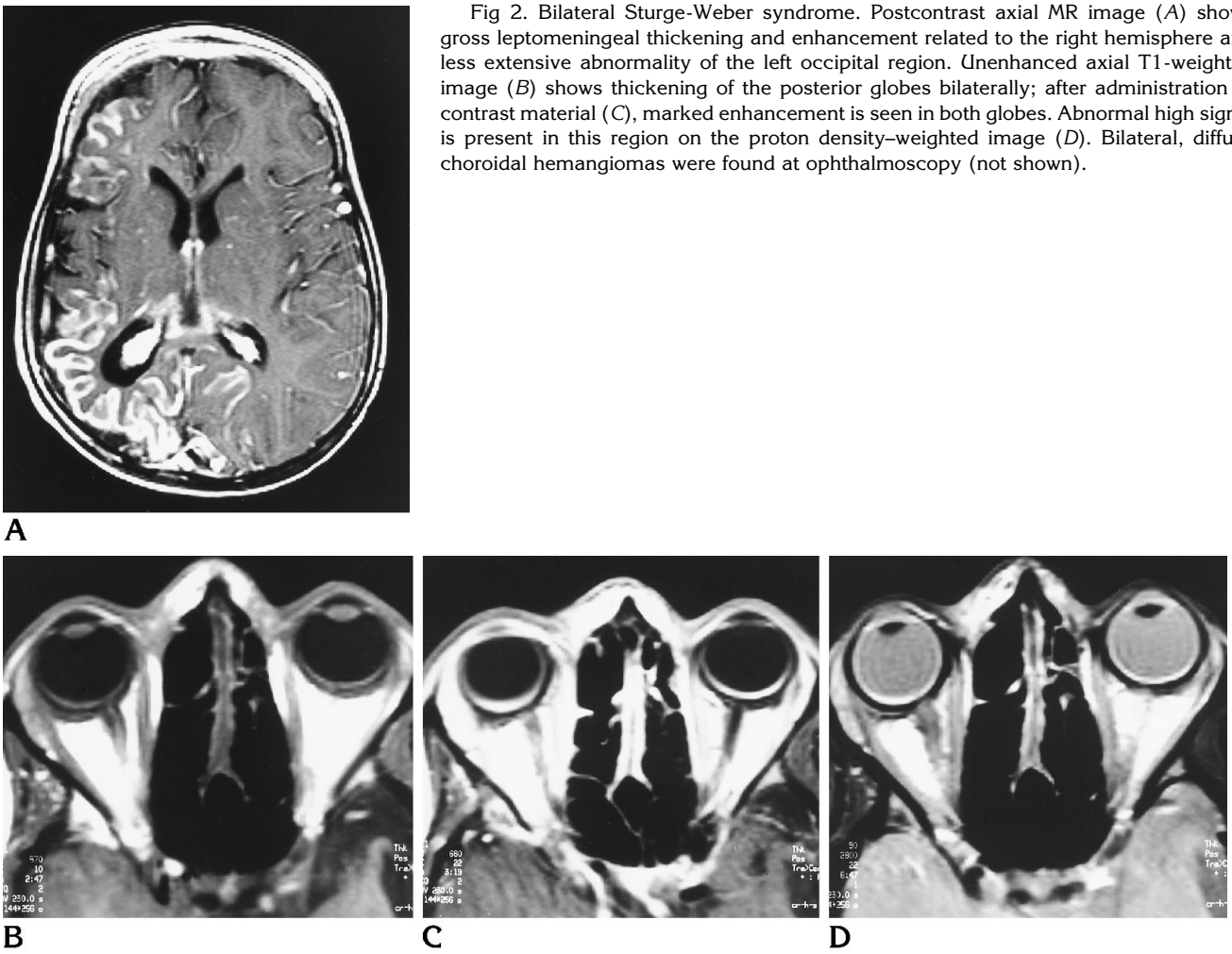

Fig 2. Bilateral Sturge-Weber syndrome. Postcontrast axial MR image (*A*) shows gross leptomeningeal thickening and enhancement related to the right hemisphere and less extensive abnormality of the left occipital region. Unenhanced axial T1-weighted image (*B*) shows thickening of the posterior globes bilaterally; after administration of contrast material (*C*), marked enhancement is seen in both globes. Abnormal high signal is present in this region on the proton density–weighted image (*D*). Bilateral, diffuse choroidal hemangiomas were found at ophthalmoscopy (not shown).

cial nevus, and presence of glaucoma. For this purpose, cortical involvement was categorized as unilateral or bilateral and as mild or severe in the individual hemispheres. MR enhancement of one or two cortical lobes was taken to represent mild intracranial involvement; enhancement of three or four lobes implied severe disease. Similarly, no facial nevus or nevus involving only one trigeminal division was considered to be mild cutaneous involvement, whereas involvement of two or three trigeminal divisions was considered severe. Likelihood (risk) ratios were calculated, and confidence intervals for the population likelihood ratios were constructed through a logarithmic transformation (6).

Results

Leptomeningeal enhancement on MR images was taken to be the radiologic sign of Sturge-Weber syndrome in this study. Of the 19 abnormal hemispheres, 3 had leptomeningeal enhancement in 1 anatomic lobe, 3 had enhancement in 2 lobes, 8 had enhancement in 3 lobes, and 5 had enhancement in all 4 lobes. The occipital lobe was affected in 18 of 19 hemispheres, the parietal lobe in 14 of 19, the temporal lobe in 11 of 19, and the frontal lobe in 10 of 19. In 1 case of unilateral Sturge-Weber syndrome, glaucoma and asymmetry of the globes were sufficient to make the diagnosis of buphthalmos. Abnormal ocular enhancement was seen in 7 patients, in 4 cases unilateral (ipsilateral to the intracranial disease) and 3 bilateral (both sides in each case), and therefore was associated with 10 (53%) of the 19 abnormal hemispheres. On MR images, the enhancement had a constant appearance of a sickle-shaped region, thickest over the posterior portion of the globe and thinning toward the ciliary body (Fig 1A and B). A corresponding area of high signal abnormality was present on proton density–weighted images (Fig 1C) but was not seen on the T2-weighted images, pre-

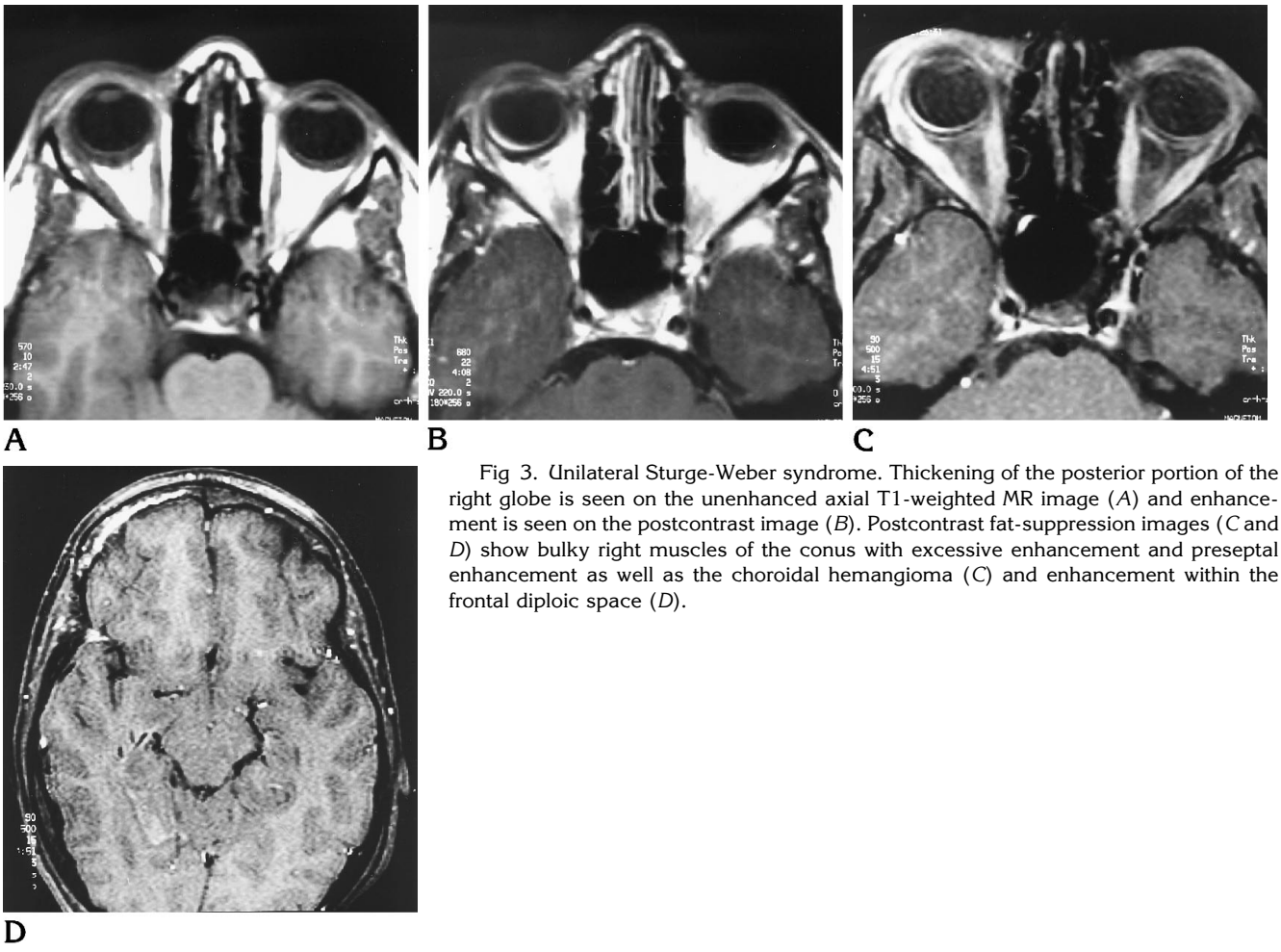

Fig 3. Unilateral Sturge-Weber syndrome. Thickening of the posterior portion of the right globe is seen on the unenhanced axial T1-weighted MR image (*A*) and enhancement is seen on the postcontrast image (*B*). Postcontrast fat-suppression images (*C* and *D*) show bulky right muscles of the conus with excessive enhancement and preseptal enhancement as well as the choroidal hemangioma (*C*) and enhancement within the frontal diploic space (*D*).

sumably because of the high signal from the adjacent vitreous. These findings are consistent with diffuse choroidal hemangioma.

The abnormalities could be seen on CT scans with confidence in the patients with unilateral disease, where the opposite side could be used for comparison. The coronal plane was often useful for this purpose (Fig 1D). In bilateral cases, CT was less useful because it was difficult to distinguish between enhancement and beam-hardening effects, although abnormal choroidal thickening was often present. MR images of a patient with bilateral disease are shown in Figure 2. The likelihood ratios (r) for ocular enhancement were significantly increased for bilateral involvement ($r = 2.09$; 95% confidence interval, 1.95 to 2.21), extensive facial nevi ($r = 2.05$; 95% confidence interval, 1.87 to 2.20), and the presence of glaucoma ($r = 1.93$; 95% confidence interval, 1.79 to 2.05); there was no significant correlation with the extent of hemispheric involvement ($r = 1.24$; 95% confidence interval, 0.92 to 1.48).

In 14 of the 15 children, the imaging and fundoscopic findings matched perfectly. In the 7 children with ocular enhancement detected with MR imaging, a choroidal abnormality was also found at ophthalmoscopy. In 6 children, both MR imaging and ophthalmoscopy showed no ocular abnormality. In 1 child with bilateral Sturge-Weber disease, ophthalmoscopy showed bilateral choroidal hemangioma but no imaging abnormality was seen. In 1 child with unilateral involvement, a choroidal hemangioma was suggested on the first ophthalmoscopic examination, but this was not confirmed by subsequent fundoscopic review or by MR imaging. In the 1 case in which postcontrast T1-weighted fat-suppression imaging was performed, the choroidal hemangioma was shown adequately but in addition there was excessive enhancement of the orbital muscles, which were

also bulky. On higher sections, prominent enhancement of the frontal diploic space was seen. Neither of these findings was seen on the standard postcontrast T1-weighted images (Fig 3).

Discussion

A review of the literature reveals one case of a 35-year-old patient with Sturge-Weber syndrome in whom ocular enhancement was seen on MR images (7) and one other case of a patient of unspecified age in whom enhancement was also shown (8). In both these reports the finding was interpreted as showing "retinal angioma." We think the enhancement represents choroidal hemangiomas, which are known to occur in Sturge-Weber syndrome; this has been corroborated with ophthalmoscopy in our study. The ocular findings in Sturge-Weber syndrome include glaucoma, choroidal hemangioma, and dilatation and tortuosity of the vessels of the conjunctiva, episclera, iris, and retina. Conjunctival or episcleral vascular malformations and elevated episcleral venous pressure have also been described (9–11). Glaucoma occurs in approximately 30% of patients with Sturge-Weber syndrome (9), although one study produced a prevalence of 76% (12). Glaucoma is most often congenital and ipsilateral to the facial nevus, especially if it involves the upper eyelid or conjunctiva (3). Some patients have anomalous anterior chamber angles (10). Surgical treatment is eventually required because medical treatment alone is typically unsuccessful. In other children with Sturge-Weber syndrome, glaucoma does not develop until the second half of the first decade or even later. Choroidal hemangioma in patients with Sturge-Weber syndrome is usually diffuse, primarily related to the posterior pole, and often seen as red, flat to moderately elevated masses. At histopathology, the vessel size is variable, with mixed capillary and cavernous morphology in all 17 cases of diffuse hemangiomas in one study (12). The diffuse lesions show engorgement of the choroidal vasculature, which is intermixed with abnormal vascular channels.

All four of the patients with bilateral Sturge-Weber syndrome in this study had bilateral choroidal hemangiomas recognized at fundoscopy, and in three cases, a bilateral abnormality was shown on MR images. In children with Sturge-Weber syndrome, the choroidal abnormality may be discernible only by its production of a bright red fundus, which has been labeled the "tomato catsup" fundus (13), as compared with the choroidal pattern in the uninvolved eye. Nevertheless, diffuse choroidal hemangioma may be a difficult diagnosis to make by fundoscopy in children. Although not routinely performed, fluorescein angiography shows early hyperfluorescence of choroidal hemangiomas, late staining with or without dye leakage. Choroidal hemangiomas may occur in early adulthood, accompanied by visual loss or visual field defects related to compression of melanocytes and hyperplasia of retinal pigment epithelium or retinal degeneration overlying the choroidal tumor. Continuing exudation of the hemangioma may lead to retinal or choroidal detachment, which may require surgery (3). Choroidal hemorrhage with overlying serous or exudative retinal detachment may result from hypotony during or after intraocular surgery. Preoperative recognition of choroidal hemangioma is therefore essential.

The MR features of the choroidal hemangiomas in our patients were constant, showing thickening of the posterior wall of the globe on unenhanced T1-weighted images and abnormal signal on proton density–weighted images. After injection of contrast material, crescentic enhancement was noted, thickest posteriorly, extending to the anterior portion of the globe. These features are similar to those reported in a study of the MR appearance of diffuse choroidal hemangioma not associated with Sturge-Weber syndrome (14). Abnormal enhancement was common, occurring in 7 of 15 children, 4 of 11 unilateral cases and 3 of 4 bilateral cases. These figures are comparable to those cited in ophthalmologic and neurologic studies (5).

We have shown that ocular enhancement was positively correlated with bilateral disease, with the extent of facial involvement, and with glaucoma, but not with the degree of hemispheric involvement as defined by abnormal enhancement. We have also described findings in one child in whom enhanced fat-suppression images showed irregular, bulky muscles of the conus with excessive enhancement on the ipsilateral side. This presumably represents hypervascularity of the muscles, possibly due to angiomatosis. This child also had enhancement within the frontal diploic space adjacent to the cutaneous flammeus nevus, indicative of more extensive extraaxial involvement than de-

scribed previously. Because of this finding, we consider enhanced fat-suppression MR imaging of the orbits to be useful in the full examination of these children.

In summary, we consider contrast-enhanced MR imaging of the brain to be the most important imaging method in the examination of children with Sturge-Weber syndrome, and we believe that this examination should include thorough investigation of the eyes. Abnormal ocular enhancement is common in children with Sturge-Weber syndrome, and the enhancement represents diffuse choroidal hemangioma. This is an important diagnosis to make because of the increased risk of glaucoma, as shown in this study. In addition, choroidal hemangioma predisposes to retinal and choroidal detachment, and may produce choroidal hemorrhage during surgery for glaucoma. Thorough fundoscopic examination is essential in all cases of Sturge-Weber syndrome, and in our institution fundoscopy is done before imaging.

Because the fundoscopic diagnosis of diffuse choroidal hemangioma can be difficult, particularly in young children, we recommend that enhanced MR imaging of the brain include thorough investigation of the eyes because of the importance of preventable late complications. Postcontrast fat-suppression techniques may provide additional information.

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