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**Progressive venous occlusion in a neonate with Sturge-Weber syndrome: demonstration with MR venography.**

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# Progressive Venous Occlusion in a Neonate with Sturge-Weber Syndrome: Demonstration with MR Venography

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Summary: Progressive cerebral sinovenous occlusion in a neonate with Sturge-Weber syndrome was documented by using two-dimensional time-of-flight MR venography. There was no evidence of intraluminal thrombus on routine spin-echo images obtained either before or after the onset of seizures, despite MR venographic evidence in both studies of venous abnormalities.

Index terms: Phakomatoses; Venography; Magnetic resonance angiography; Veins, stenosis and occlusion

Sturge-Weber syndrome is a phakomatosis that is associated with a leptomeningeal venous malformation and a paucity of normal cortical veins ipsilateral to the malformation. Absence of various deep veins and dural venous sinuses has also been documented with both conventional and magnetic resonance (MR) angiography. It is not known whether the nonvisualized veins are thrombosed or aplastic. Venous thrombosis has been suggested as a cause for the clinical deterioration often observed in these patients, and antiplatelet therapy has been advocated. However, neither acute venous thrombosis nor progressive venous occlusion has been demonstrated radiographically in patients with Sturge-Weber syndrome. We present a case of progressive venous occlusion in a neonate with Sturge-Weber syndrome that was documented with MR venography.

## Case Report

A newborn black boy was examined because of bilateral facial port-wine nevi, left-sided glaucoma, and a cloudy cornea. The neurologic examination was normal. Sturge-Weber syndrome was suspected clinically. Computed tomography showed mild right cerebral hemiatrophy but no cortical calcifications. The mild right cerebral atrophy was confirmed with routine fast spin-echo MR images. There was subtle diffuse bilateral leptomeningeal enhancement after intravenous administration of gadopentetate dimeglumine. The glomus of the choroid plexus was enlarged and prominently enhanced bilaterally. A large vascular flow void was noted extending inferiorly from the tip of the right anterior horn to the right sylvian cistern. A two-dimensional time-of-flight MR venogram, 45/8.7/1 (repetition time/echo time/excitations), showed a paucity of cortical veins on the right. The straight sinus was very small and there was a prominent falcine sinus (Fig 1A and B). The internal cerebral veins and vein of Galen were not visualized. Deep vessels drained from the right basal ganglia region into a large right sphenopetrosal vein. There was asymmetric visualization of the right superior ophthalmic vein. The patient received no antiplatelet or anticonvulsant therapy.

Complex partial seizures commenced at approximately 8 months of age. A repeat CT study showed progressive bilateral cerebral atrophy and new subtle gyral calcifications in the left parietal lobe. A follow-up MR venogram with technical factors identical to those of the initial examination showed increased prominence of the deep vein draining from the region of the right frontal horn and of the sphenopetrosal vein into which it drained (Fig 1C). The straight sinus was no longer visualized and the falcine sinus appeared more obvious. The proximal right transverse sinus appeared stenotic (Fig 1D), and the right occipital and marginal sinuses were more apparent. No intraluminal venous thrombus was identified on routine T1-weighted spin-echo images or on proton density– or T2-weighted fast spin-echo images. Enhanced T1-weighted images (Fig 2) showed more pronounced leptomeningeal enhancement in the left temporal, parietal, and occipital regions and in the right frontal and parietal regions. Prominently enhancing deep medullary veins were noted traversing the right cerebral hemispheric white matter, converging at the lateral wall of the body of the lateral ventricle into a longitudinal caudate vein. It appeared that the latter then drained via the anomalous vein extending inferiorly from the right anterior horn, rather than into the apparently occluded deep venous system.

Fig 1. Sturge-Weber syndrome, progressive venous occlusion.

*A*, Lateral view of MR venogram in a 3-day-old boy. Note small straight sinus (*medium-sized arrowheads*), persistent falcine sinus (*largest black arrowhead*), and anomalous deep veins draining into the transverse sinus by way of a sphenopetrosal vein (*smallest arrowheads*). The internal cerebral veins are not visible.

*B*, Anteroposterior view. Right transverse sinus (*arrowheads*) is patent.

*C*, Lateral view at follow-up study performed after the onset of seizures shows occlusion of the straight sinus and increased prominence of the falcine sinus (*large arrowhead*) and of the anomalous deep vein and sphenopetrosal vein (*small arrowheads*).

*D*, Anteroposterior view now shows narrowing of the right transverse sinus (*large arrowheads*). Note anomalous deep veins and sphenopetrosal vein (*small arrowheads*). The occipital and right marginal sinuses (*arrows*) have become larger, presumably reflecting rerouting of venous drainage because of transverse sinus stenosis.



A    B

C    D
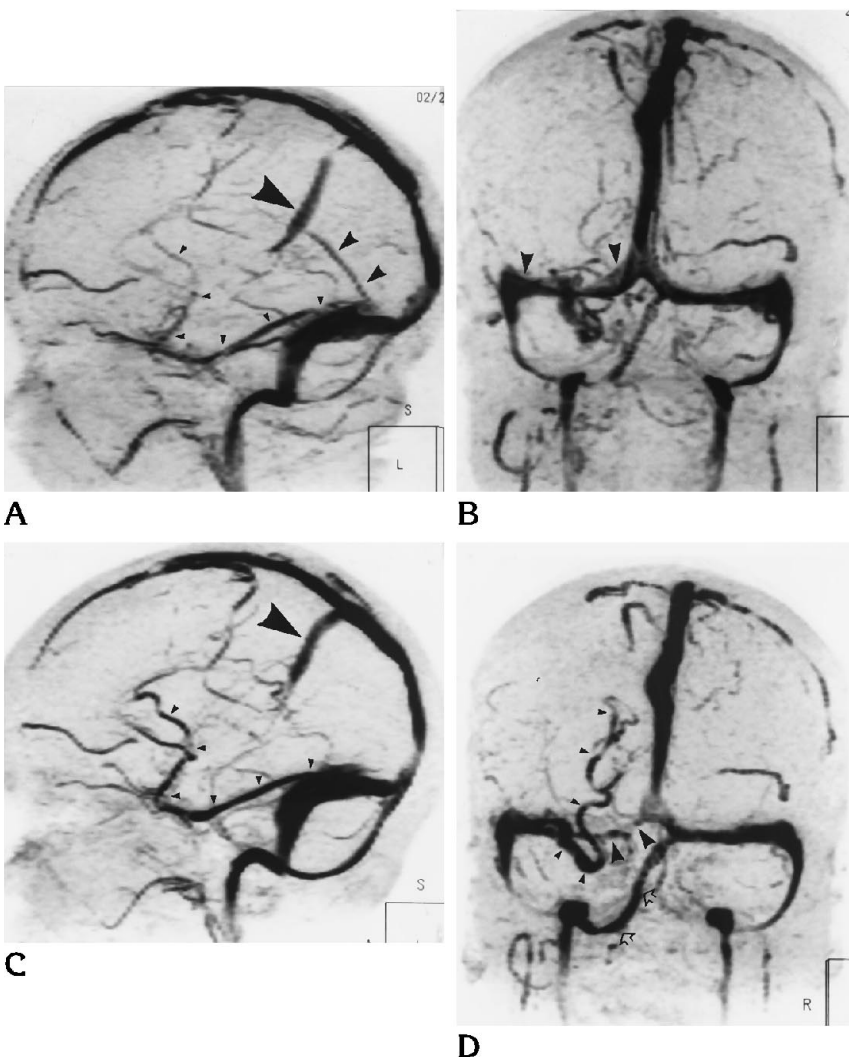
## Discussion

Sturge-Weber syndrome is a neurocutaneous syndrome (phakomatosis) characterized by a facial port-wine nevus (a capillary malformation), seizures, and mental retardation (1). Central nervous system manifestations include a leptomeningeal venous malformation ("angioma") and cerebral hemiatrophy, which are typically, though not always, ipsilateral to the facial nevus. Calcifications are seen in the atrophic cortex deep to the venous malformation. Absence of cortical veins with centripetal venous drainage into enlarged medullary veins or anomalous deep veins in Sturge-Weber syndrome has been described in the angiographic, computed tomographic, and MR literature (2–10). These enlarged deep veins typically drain into the galenic venous system and

likely represent collateral veins rather than primary vascular abnormalities. Dural venous sinus and galenic venous occlusions have also been documented with conventional and MR angiography in patients with Sturge-Weber syndrome (3, 6–10), and they may modify the pattern of centripetal venous drainage.

It has been suggested that stepwise clinical deterioration in patients with Sturge-Weber syndrome might be caused by cerebral venous hypertension resulting from sluggish venous drainage through the leptomeningeal angioma (9). Probst (9) hypothesized that the latter might be further exacerbated by progressive occlusion of deep veins with resultant centrifugal rerouting of deep cerebral venous drainage into the already overburdened angioma. Slowing and diminution of regional cerebral blood
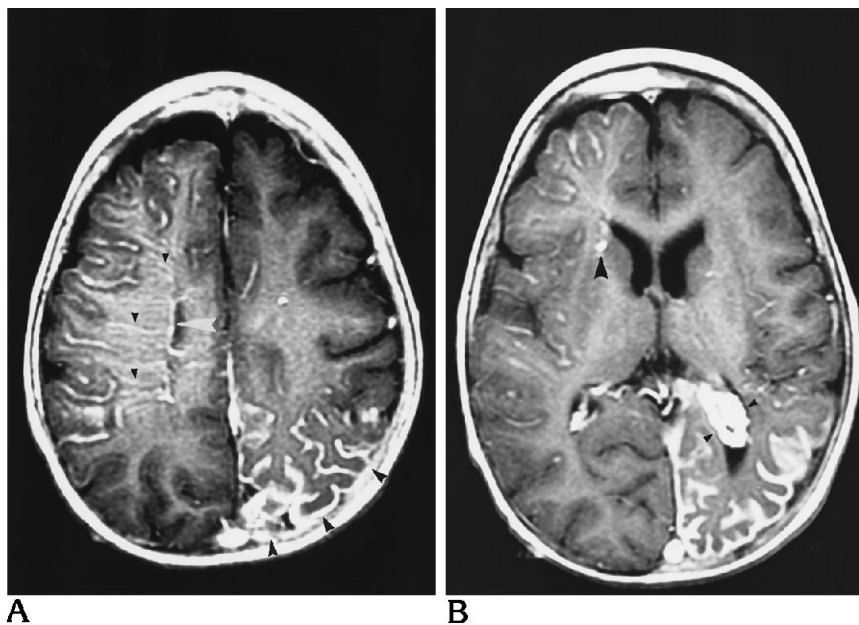
A  B

Fig 2. Sturge-Weber syndrome, venous drainage modified by deep venous occlusion.

A, Axial enhanced T1-weighted MR image shows enlarged deep medullary veins (*small black arrowheads*) draining centripetally from the cortical surface into a longitudinal caudate vein (*white arrowhead*). Note leptomeningeal "angioma" (*large black arrowheads*).

B, More inferiorly at the level of the cavum velum interpositum, the anomalous deep vein can be seen traveling in a vertical orientation (*large arrowhead*). The internal cerebral veins are not visible. Note prominent glomus of the left choroid plexus (*small arrowheads*) and the left occipital leptomeningeal "angioma."

flow has been demonstrated by using radionuclide and xenon inhalation techniques in these patients (9, 11). On the basis of the postulate that these findings might be caused by progressive venous thrombosis, antiplatelet therapy was administered in one series of patients and resulted in stabilization of neurologic function (12).

Isolated nonvisualization of cortical veins, deep veins, and dural venous sinuses on conventional or MR angiography in patients with Sturge-Weber syndrome does not establish whether these veins are aplastic, hypoplastic, thrombosed, or occluded by processes such as intimal hyperplasia or other nonthrombotic diseases. Without follow-up, these findings also do not prove a progressive venoocclusive process in Sturge-Weber syndrome.

The observation of venous abnormalities in this neonate suggests that the venous pathology in Sturge-Weber syndrome might commence in utero and progress postnatally. We could not identify intraluminal venous thrombus as the cause of progressive nonvisualization of veins in this patient, suggesting that *nonthrombotic* venoocclusive processes might play a role in the pathophysiology of Sturge-Weber syndrome. Indeed, abnormal veins with walls thickened by a layer of hyalinized connective tissue or fibrosis have been noted at pathologic examination within the leptomeninges and subcortical white matter of patients with Sturge-Weber syndrome (13).

Given the noninvasive nature of conventional MR and MR venography, it should be feasible to study a larger group of patients with Sturge-Weber syndrome to determine if there is a relationship between clinical deterioration and progressive venous occlusion, and to what extent venous thrombosis contributes to venous occlusion in these patients. If thrombosis is established as a significant contributor to the disease process, clinical trials that use antiplatelet medications could be guided by periodic MR venographic evaluation.

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