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AJNR Am J Neuroradiol 1982, 3 (4) 435-439

<http://www.ajnr.org/content/3/4/435.citation>

This information is current as
of June 21, 2025.

Sonographic Demonstration of Galenic Arteriovenous Malformations in the Neonate

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Cerebral arteriovenous malformation with aneurysmal dilatation of the vein of Galen often presents in the neonatal period with congestive heart failure. Five cases of galenic arteriovenous malformations diagnosed primarily by sonography are presented together with their angiographic and computed tomographic findings. A midline cystic structure posterior to the third ventricle with dilated straight sinus and torcular Herophili is typical of this entity.

The vein of Galen may become aneurysmally dilated by increased flow from a deep cerebral arteriovenous malformation or by direct arterial fistulization. Clinical, radiographic, and operative reports of galenic arteriovenous malformations are well documented [1-6]. Cerebral arteriovenous malformations presenting in the neonatal period are usually associated with life-threatening congestive heart failure. Cronquist et al. [6] reported that about 80% of intracranial arteriovenous malformations causing congestive heart failure in the infant involve the galenic system. The mortality in these patients ranges from 66% [3] to 100% [2, 5, 7].

Real-time sonography is of increasing interest as a primary means of investigating neonatal and infant intracranial pathology [8-10]. Two recent reports [11, 12] described the utility of cerebral sonography in neonates in identifying galenic arteriovenous malformation. All of the patients in these reports, however, underwent angiography before sonography and three of four patients had had operations previously. At Primary Children's Medical Center, there have been five patients with neonatal vein of Galen arteriovenous malformations in the previous 6 months in whom the diagnosis was primarily established by real-time sonography and supported by angiography or computed tomography (CT) [13].

Subjects and Methods

Significant clinical data of the five cases are presented in table 1. All patients were studied with an ATL 5 MHz real-time sector scanner (Advanced Technology Laboratories, Bellevue, WA 98005) with simultaneous real-time Doppler capability ("Duplex" system) [14]. Subjects were examined in the coronal and sagittal planes through the anterior fontanelle and in the axial plane through the posterior fontanelle (case 5). Sonography was the first diagnostic method used to demonstrate intracranial pathology in all patients, and CT or angiography was subsequently used to confirm the initial impression of galenic arteriovenous malformation.

Discussion

Clinical findings of a vein of Galen arteriovenous malformation in a neonate or infant include congestive heart failure, cranial bruit, and enlarged head [4]. Often, however, the head enlargement and bruit may be inconspicuous, and these

Received July 6, 1981; accepted after revision January 7, 1982.

Presented at the annual meeting of the Society for Pediatric Radiology, New Orleans, May 1982.

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AJNR 3:435-439, July/August 1982
0195-6108/82/0304-0435 \$00.00
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TABLE 1: Findings with Galenic Arteriovenous Malformations (AVMs)

Case No.	Presenting Signs and Symptoms	Sonographic Findings	Method of Confirmation	Outcome
1	Cyanosis, cardiomegaly, heart murmur	Galenic AVM, mild hydrocephalus	Doppler sonography, angiography	Died at angiography
2	Cyanosis, cardiomegaly with chronic heart failure, heart murmur	Galenic AVM, severe hydrocephalus	CT	Died on medical management
3	Poor feeding, vomiting, cardiomegaly	Galenic AVM, mild hydrocephalus, pulsations of galenic AVM	Doppler sonography, CT, angiography, surgery	Died at surgery
4	Cyanosis, cardiomegaly, heart murmur, cranial bruit	Galenic AVM	Doppler sonography, CT, angiography with embolization	Improved after embolization
5	Apnea, cyanosis, cardiomegaly, cranial bruit	Eccentric galenic AVM, mild hydrocephalus	Doppler sonography, CT, angiography with embolization, surgery	Improved after embolization ×2 and surgery ×2

Note—Presentation was at birth in cases 1, 2, 4, and 5 and at age 7 days in case 3.

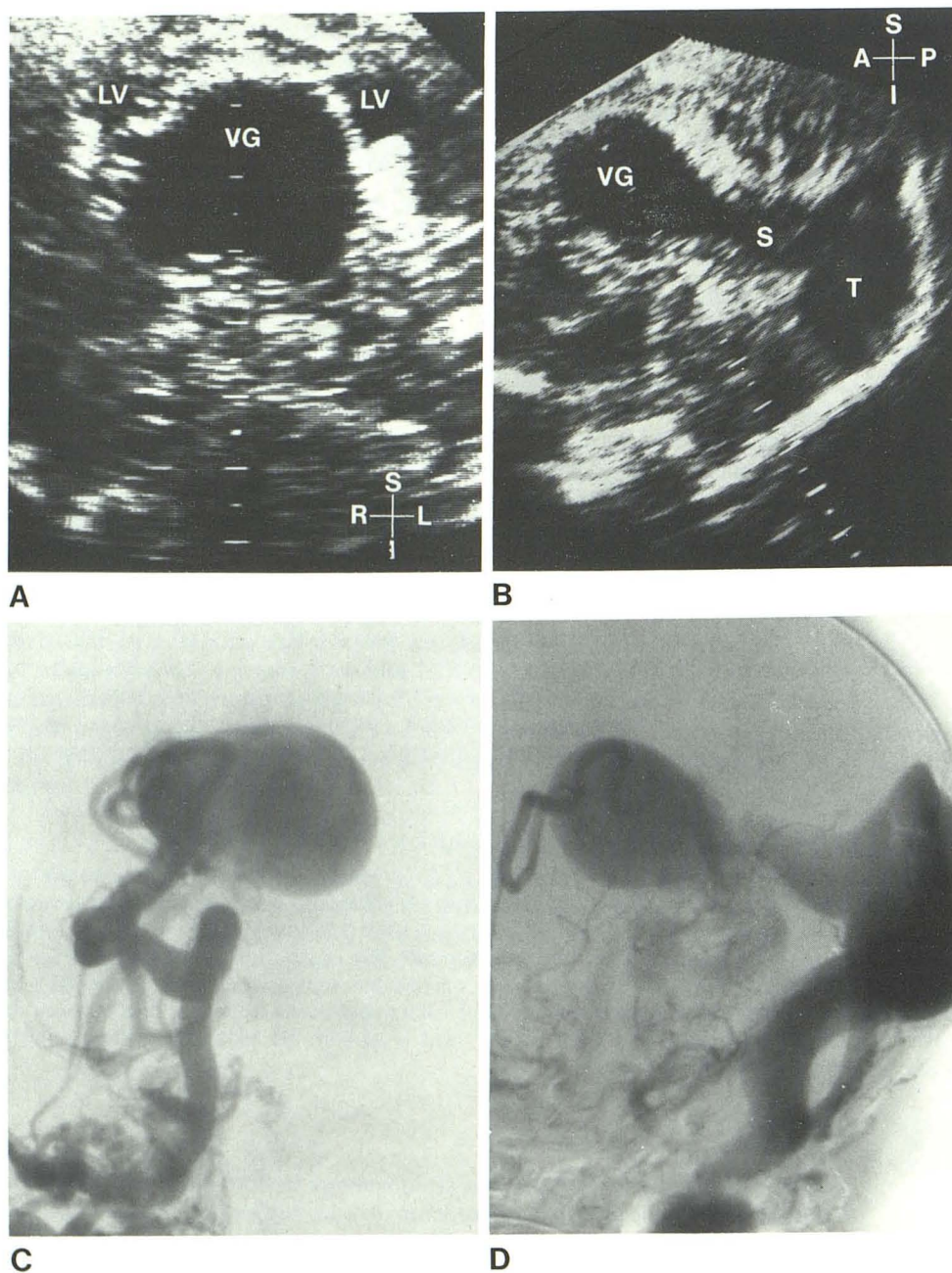


Fig. 1.—Case 1. Typical sonographic appearance of galenic arteriovenous malformation. **A**, Coronal real-time sonogram through anterior fontanelle and directed posteriorly. Dilated vein of Galen (VG) with sharp margins and no internal echoes. Mildly dilated lateral ventricles (LV) on either side of galenic vein. **B**, Midline sagittal scan. Aneurysmal dilatation of vein of Galen (VG), connected with dilated straight sinus (S) and torcular Herophili (T). Small cystic structures inferior to galenic vein pulsated on real-time and represented dilated feeding arteries, but are not well illustrated on stop-frame. **C**, Anteroposterior projection of early arterial phase of right internal carotid injection. Dilated feeding vessels and prompt filling of galenic vein. **D**, Lateral projection of right internal carotid injection at midarterial phase. Vein of Galen shown with rapid filling of dilated straight sinus and torcular Herophili. Appearance closely resembles sagittal sonogram in **B**. S = superior; I = inferior; R = right; L = left; A = anterior; P = posterior.

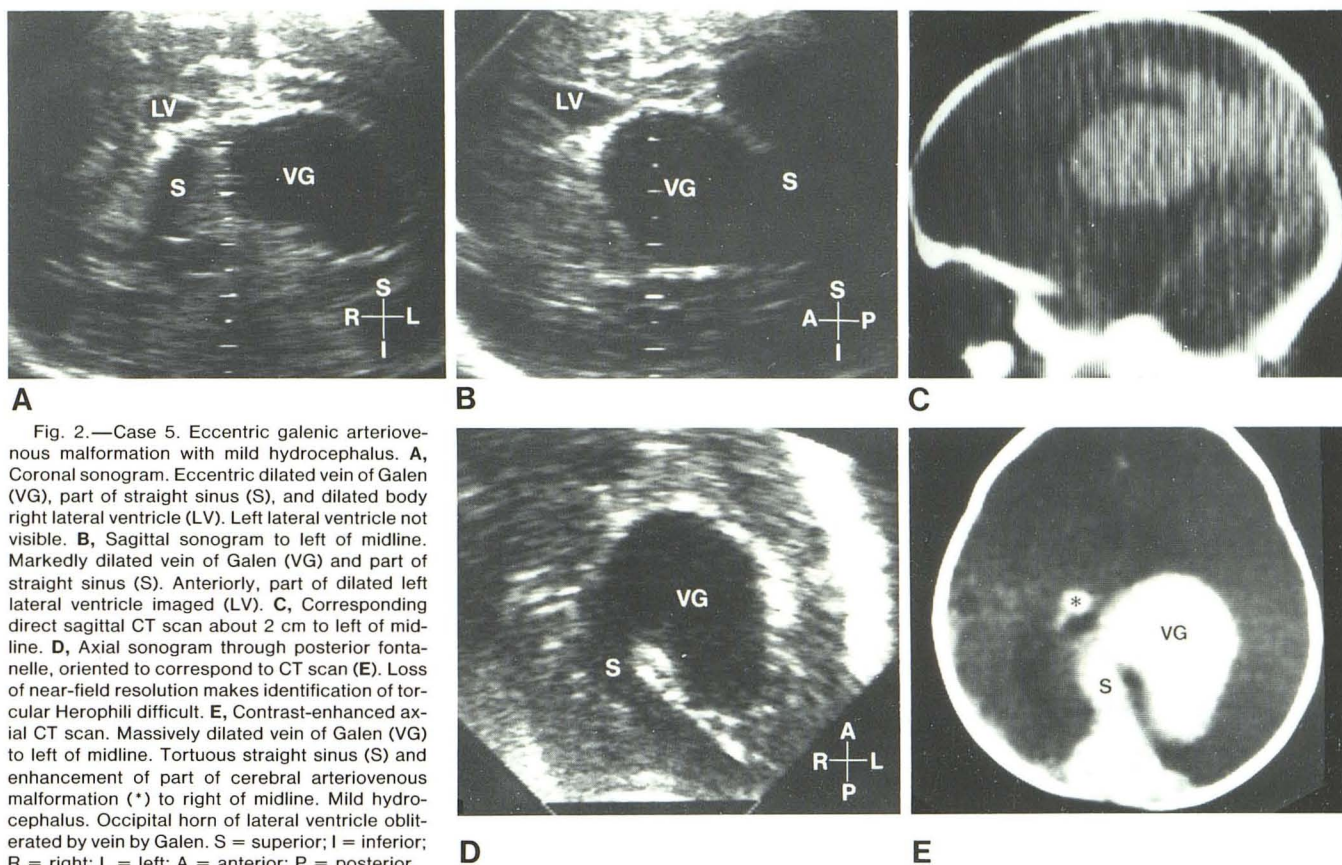


Fig. 2.—Case 5. Eccentric galenic arteriovenous malformation with mild hydrocephalus. **A**, Coronal sonogram. Eccentric dilated vein of Galen (VG), part of straight sinus (S), and dilated body right lateral ventricle (LV). Left lateral ventricle not visible. **B**, Sagittal sonogram to left of midline. Markedly dilated vein of Galen (VG) and part of straight sinus (S). Anteriorly, part of dilated left lateral ventricle imaged (LV). **C**, Corresponding direct sagittal CT scan about 2 cm to left of midline. **D**, Axial sonogram through posterior fontanelle, oriented to correspond to CT scan (**E**). Loss of near-field resolution makes identification of torcular Herophili difficult. **E**, Contrast-enhanced axial CT scan. Massively dilated vein of Galen (VG) to left of midline. Tortuous straight sinus (S) and enhancement of part of cerebral arteriovenous malformation (*) to right of midline. Mild hydrocephalus. Occipital horn of lateral ventricle obliterated by vein by Galen. S = superior; I = inferior; R = right; L = left; A = anterior; P = posterior.

patients are considered to have cardiac failure based on congenital heart disease. The radiologist may be the first to recognize the possibility of a peripheral arteriovenous malformation because of features of congestive heart failure with enlarged aorta on chest radiographs.

The sonographic appearance of the vein of Galen arteriovenous malformation in our five cases was variable, but all had important similarities that allowed correct diagnosis. In the coronal plane through the anterior fontanelle, the cerebral arteriovenous malformation was poorly identified, but the dilated galenic vein was readily seen as an anechoic structure, free of internal echoes (fig. 1). The vein arises posterior to the foramen of Monro and superior to the third ventricle (structures easily identified on most neonatal sonograms). Although the dilated vein of Galen may be considerably off midline (case 5, fig. 2), it invariably lies between the bodies of the lateral ventricles. Hydrocephalus of some proportion was found in four of our cases (fig. 3). Significant hydrocephalus, while common in infants with this condition, is not common in the neonatal period [2, 3].

The sagittal image clearly demonstrates the enlarged vein of Galen draining posteriorly into the dilated straight sinus and torcular Herophili. The straight sinus may become tortuous with increased flow, making display of the complete sinus difficult in a single field of view. However, minor adjustments of the transducer at real-time imaging should trace the cystic structure into the torcular. We found these

features in all our cases to be corroborated with CT or angiography.

Pulsations of the vein of Galen were observed in one case, and in all cases the carotid arteries at their origin in the thorax were enlarged and hyperdynamic when imaged from the suprasternal notch.

Differential considerations are few and should be distinguishable by sonography. Peripineal teratomas or lipoma of the corpus callosum are solid lesions that lack the characteristic features of cystic structures. An arachnoid cyst of the quadrigeminal plate, partial agenesis of the corpus callosum with interhemispheric cyst, or a recently described [15] pulsion diverticulum of the atrium of the lateral ventricle with marked hydrocephalus may all cause confusion, particularly in coronal sections. However, sagittal sonography will usually identify the cystic galenic vein by its connection posteriorly into the dilated straight sinus. In one case of an intraventricular cyst (fig. 4), a posteriorly displaced third ventricle simulated the straight sinus, and only careful attention to anatomy allowed this distinction. Care must also be taken to avoid mistaking a dilated sylvian aqueduct and fourth ventricle for the straight sinus and torcular Herophili, respectively.

A Doppler flow meter offers simple confirmation when a vein of Galen aneurysm is suspected. The duplex system allows simultaneous visualization of the dilated vascular structures and measurement of blood flow within them

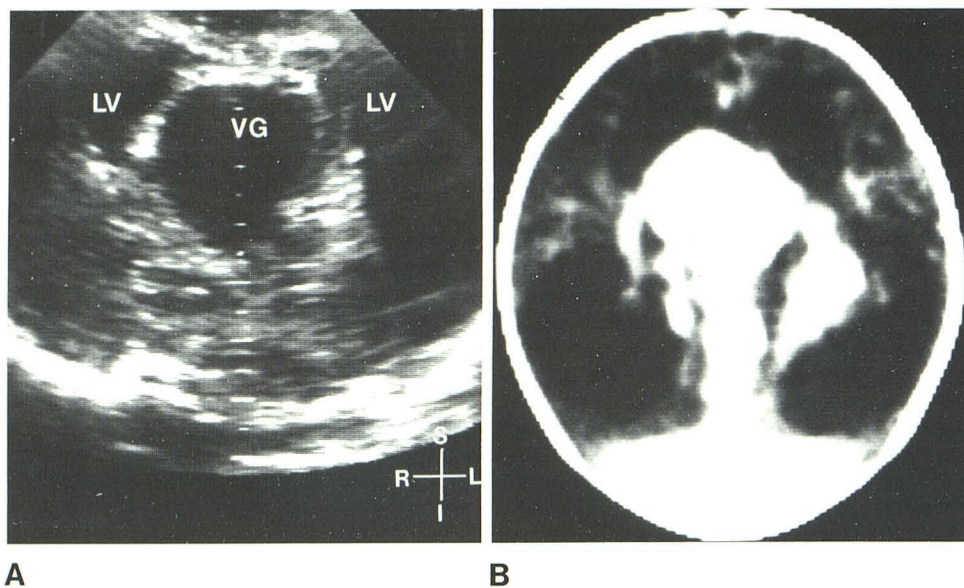


Fig. 3.—Case 2. **A**, Coronal scan. Midline cystic galenic vein (VG). Its more inferior part was confluent with straight sinus as transducer swept posteriorly. Markedly dilated lateral ventricles (LV) on either side of vein of Galen aneurysm. **B**, Axial CT scan. Contrast-enhanced midline galenic arteriovenous malformation and marked hydrocephalus clearly shown. Malformation extends anteriorly and is surrounded by prominent feeding vessels. Posteriorly, parts of straight sinus and torcular Herophili. S = superior; I = inferior; R = right; L = left.

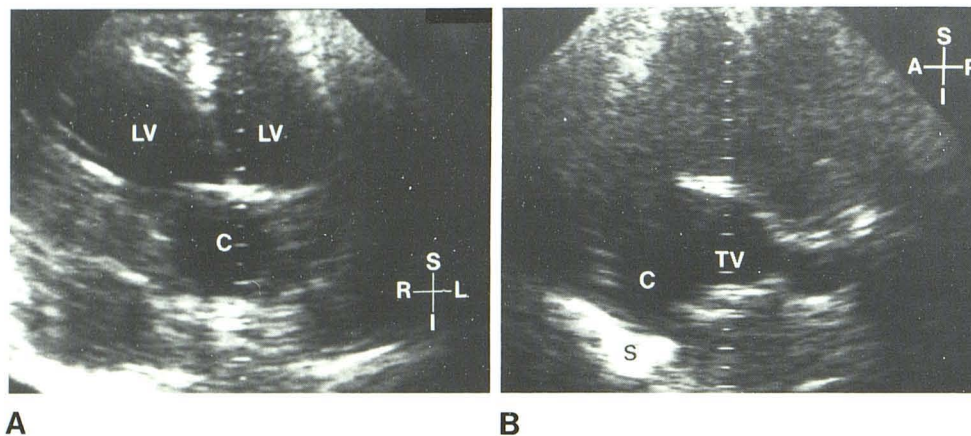


Fig. 4.—**A**, Subarachnoid intraventricular cyst (C). Coronal sonogram. Anterior third ventricle with marked hydrocephalus of lateral ventricles (LV). This appearance may cause some confusion with dilated vein of Galen, but cyst does not separate lateral ventricles as did all our galenic arteriovenous malformations. **B**, Midline sagittal scan, same patient. Intraventricular cyst (C) creates image that might be mistaken for dilated vein of Galen and straight sinus. Careful attention to close relation of cyst to posteriorly displaced third ventricle (TV) and sella (S) permits differentiation. S = superior; I = inferior; R = right; L = left; A = anterior; P = posterior.

[14]. Should real-time and Doppler sonography not yield the necessary diagnostic information, or if the patients have closed sutures, CT may be required. Angiography is not recommended as the primary mode of investigation, but will be needed for evaluation of possible embolic or operative intervention.

Real-time sonography offers the distinct advantages of being a noninvasive investigation with mobile equipment. It is ideally suited to the neonate with congestive heart failure as both the cardiac anatomy and function and intracranial contents can be evaluated. A vein of Galen arteriovenous malformation with associated dilatation of the straight sinus and torcular Herophili causing congestive heart failure may be easily demonstrated. Its sonographic appearance is characteristic and allows primary diagnosis of this life-threatening anomaly.

ACKNOWLEDGMENT

We thank Laura Newren for technical assistance.

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