

# Prenatal diagnosis of thrombosed aneurysm of vein of Galen

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## ABSTRACT

*Aneurysm of the vein of Galen in the fetus is a rare malformation that has been described in a small number of case reports. Thrombosis of aneurysm of the vein of Galen is a rare occurrence reported in postnatal cases. We report a case of thrombosis of vein of Galen aneurysm occurring in a fetus of 21 weeks' gestation. The fetus showed an echogenic mass posterior and superior to the thalamus. It also showed dilated dural venous sinuses, dilated neck vessels, cardiomegaly and minimal ascites. The pregnancy was terminated and autopsy confirmed the diagnosis. The present case is the first report of the prenatal sonographic features of this condition in the literature. Copyright © 2005 ISUOG. Published by John Wiley & Sons, Ltd.*

## CASE REPORT

A 30-year-old woman, gravida 2 para 1, was referred for prenatal ultrasound assessment following identification of cardiomegaly at 21 weeks' gestation. The course of the pregnancy had been uneventful prior to referral and fetal biometry was normal for gestational age. The amniotic fluid volume was normal. There was an echogenic mass of 20 × 17 mm posterior and superior to the thalamus (Figure 1), with no flow seen on color Doppler examination. The sagittal and transverse venous sinuses were dilated, with flow detected when examined by color Doppler (Figure 2). Dilated neck vessels were detected (Figure 3). Cardiomegaly and minimal ascites were also present. A diagnosis of thrombosed aneurysm of the vein of Galen was suggested. The parents were informed of the condition and they chose to terminate the pregnancy. Fetal autopsy confirmed a large thrombus filling the aneurysm of the vein of Galen (Figure 4).

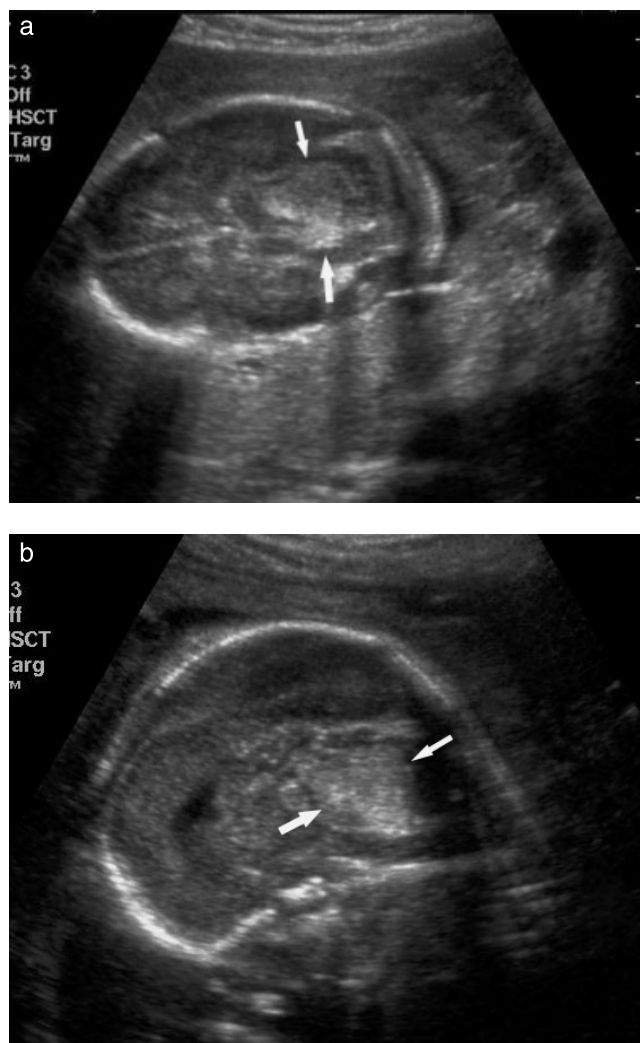
## DISCUSSION

Aneurysm of the vein of Galen is a complex arteriovenous malformation consisting of multiple communications between the system of the vein of Galen and the cerebral arteries (carotid and/or vertebrobasilar systems)<sup>1</sup>. Raybaud *et al.*<sup>2</sup> have concluded that anatomical analysis of cases with this condition and correlation with known embryological data indicate that the venous sac most probably represents persistence of the embryonic median prosencephalic vein of Marknowski, not the vein of Galen *per se*. It may result in hemodynamic and central nervous system disturbances in affected neonates, even during fetal life. Severe high-output cardiac failure is caused by a marked increase of cardiac preload from venous return of the brain due to the 'steal' phenomenon. Hydrocephalus may develop secondary to the obstruction of the aqueduct of Sylvius by the dilated aneurysm. The 'steal' phenomenon, with diversion of blood from the parenchyma to the aneurysm, may further result in brain infarcts and periventricular white matter lesions.

Imaging studies, including angiography, ultrasonography, computed tomography (CT) and magnetic resonance imaging (MRI), provide the basis for postnatal diagnosis of aneurysm of the vein of Galen. Thus far, prenatal diagnosis of such an aneurysm has been made in several cases, essentially by pulsed Doppler ultrasonography and color flow mapping<sup>3–6</sup>. The most striking prenatal finding is the presence of a posterior, midline or slightly lateral supratentorial cystic structure in the fetal brain<sup>6</sup>. The sagittal sinus is reported as dilated in most cases. The turbulent vascular flow, diagnostic of an aneurysm, is detected by pulsed and color Doppler ultrasonography. Demonstration of blood flow in the cystic structure enables the diagnosis of an aneurysm of the vein of Galen to be made, as opposed to that of an abnormality of other intracranial midline structures, for example arachnoid or porencephalic cyst, Dandy–Walker malformation or intracerebral hematoma. Sepulveda *et al.* described

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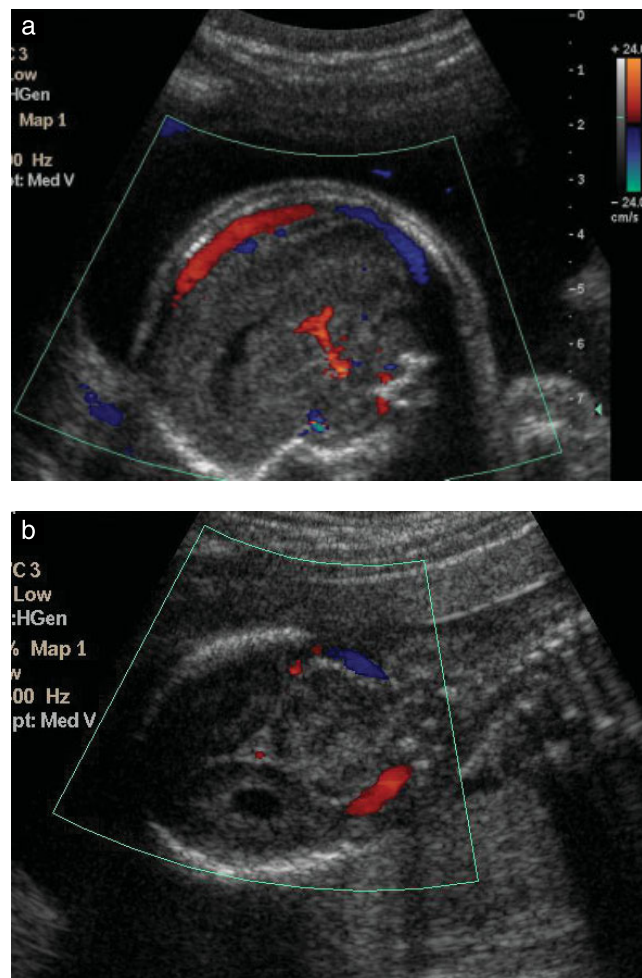
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**Figure 1** Axial (a) and sagittal (b) scan of the fetal skull showing the echogenic mass (arrows) posterior and superior to the thalamus.

ventriculomegaly, cardiomegaly and dilated neck vessels as commonly present associated findings<sup>3</sup>, while Pilu *et al.* reported distortion of the cerebral architecture in cases with aneurysm of the vein of Galen as a negative sign<sup>6</sup>. There are recent reports of the use of three-dimensional color power angiography to depict the angioarchitecture of this malformation<sup>7–10</sup>. Prenatal MRI is useful in assessing the vascular connection of this condition as well as the degree of cerebral damage<sup>10</sup>.

The optimal management strategies and prognostic indices for neonates with aneurysm of the vein of Galen remain to be established. Generally, postdelivery management of such fetuses requires detailed anatomical delineation with imaging studies, followed by neurosurgical treatment such as total excision, ligation or embolization of the aneurysm<sup>1,11</sup>. At present embolization is the treatment of choice, with good results and a good neurological outcome<sup>11</sup>. It is generally agreed that the presence of cardiac decompensation and gross brain pathology indicates adverse perinatal outcomes and necessitates active intervention. However, there are some reports of spontaneous thrombosis of aneurysm of the

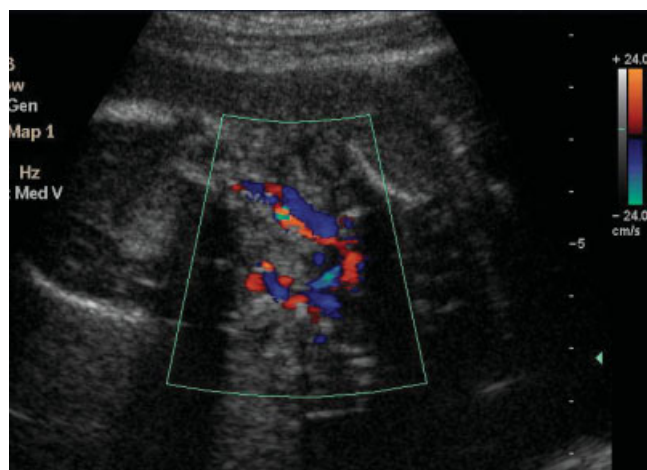


**Figure 2** Color Doppler imaging showing the dilated sagittal venous sinus (a) and transverse sinuses (b).

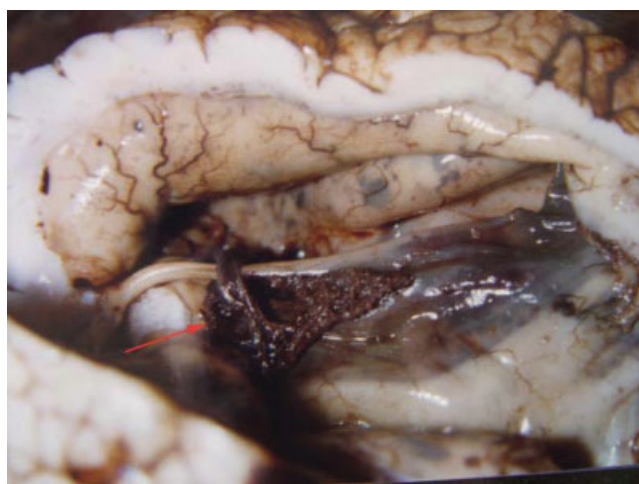
vein of Galen in the postnatal period<sup>12,13</sup>. In these cases, the thrombosed aneurysm of the vein of Galen is seen as a mass of high density/intensity values with varying degrees of calcification on CT and MRI. A completely calcified aneurysm of the vein of Galen has also been reported<sup>14</sup>. All the reports of thrombosis of aneurysm of the vein of Galen have recorded excellent clinical outcomes of the patients. So far, there has been no report of prenatal thrombosis of aneurysm of the vein of Galen; this case report is the first to describe the fetal sonographic features of a thrombosed aneurysm of the vein of Galen.

The thrombosed aneurysm of the vein of Galen in the fetus was seen on prenatal sonography as an echogenic mass in the midline, posterior and superior to the thalamus. Even though the same features may be seen in masses of the pineal gland or third ventricle, the diagnosis of thrombosed aneurysm of the vein of Galen was made because of associated findings of dilated cerebral venous sinuses, dilated neck vessels and cardiomegaly, in the absence of any vascularity of the mass itself.

To conclude, an echogenic mass in the classical location with dilated dural venous sinuses and dilated neck vessels is diagnostic of a thrombosed aneurysm of the vein of Galen.



**Figure 3** Color Doppler examination of the neck showing dilated neck vessels.



**Figure 4** Postmortem image of the incised aneurysm of the vein of Galen showing the thrombus inside (arrow).

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