

Prenatal depiction of angioarchitecture of an aneurysm of the vein of Galen with three-dimensional color power angiography

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ABSTRACT

A fetus with a large supratentorial cyst and cardiomegaly was encountered at 33 weeks of gestation. The cyst appeared as an aneurysmal, fluid-filled structure extending posteriorly with a finger-like appendage. Using color flow mapping, we disclosed rapid in-and-out blood flow with marked turbulence within the cyst. For evaluation of its blood supply and venous drainage of the vascular malformation, a three-dimensional reconstruction of the power Doppler image was conducted. The results revealed that the vascular malformation was supplied by a small contralateral aneurysm from the branches of Willis' circle, draining posteriorly into an abnormal falcine sinus and then into the superior sagittal sinus. No other fetal abnormality such as hydrocephalus or hydrops was discovered. The prenatal diagnosis of an aneurysm of the vein of Galen was made on the basis of the gray-scale, color Doppler findings, and also the angioarchitecture obtained by three-dimensional power Doppler imaging. The woman was admitted at 37 weeks of gestation due to labor onset and delivered the baby via the vaginal route without complication. Postnatal angiography and magnetic resonance imaging confirmed the diagnosis of an aneurysm of the vein of Galen, and the angioarchitecture depicted it before birth. We suggest that three-dimensional power Doppler ultrasonography may assist in the diagnosis of an aneurysm of the vein of Galen, and precisely delineate the complicated corresponding vasculature. This may guide postnatal management and predict the prognosis more accurately.

INTRODUCTION

Conventional color Doppler imaging is a widely used diagnostic tool which is based on mean Doppler frequency

shift. A refinement, the power Doppler image, also known as color power angio (CPA), color Doppler energy (CDE), or amplitude-based Doppler ultrasonography, has been used in clinical practice for many years¹. It has the apparent benefits that it can detect low flow state, thereby obviating aliasing, angle-dependence, and overwhelming noise that are usually encountered in conventional color Doppler images. Blood flow in a confined area or continued vessel can be rendered in an impression of angiography. In some studies the authors suggested that power Doppler ultrasound is superior to conventional color Doppler imaging in depiction of normal intrarenal and intracranial vascular anatomy^{2,3}. Recently, three-dimensional (3D) rendering of power Doppler images has been introduced in the fields of obstetrics and gynecology^{4–6}.

We described the application of 3D CPA as an adjunct in the diagnosis of an aneurysm of the vein of Galen and visualization of its angioarchitecture *in utero*.

CASE REPORT

A 28-year-old, gravida-3-para-0 female was referred for evaluation of a large brain cyst detected at 33 weeks of gestation. She had received left nephrectomy due to a benign nephrogenic tumor 5 years previously. The course of pregnancy and other medical, genetic, and toxicologic histories were otherwise unremarkable. Our initial scanning at 33 weeks revealed a large biparietal diameter (92 mm, larger than 95th centile for the gestational age). Other fetal biometries were within normal limits. The brain ventricle appeared normal in size. However, we depicted a 38 × 30 mm anechoic cyst located superior-posterior to the thalamus, mainly over the right hemisphere, at a higher axial plane of fetal head (Figure 1a). Color Doppler interrogation depicted an abundant,

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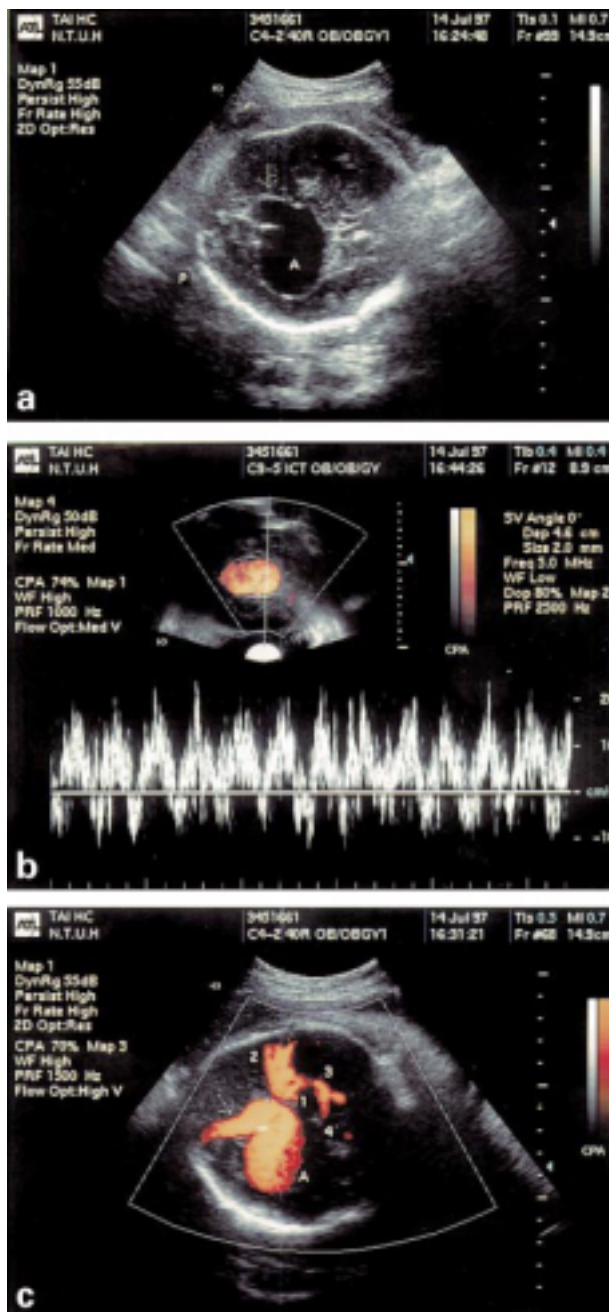


Figure 1 (a) Fetus at 33 weeks of gestation: gray-scale ultrasound revealed an anechoic cyst (A) located superior-posterior to the thalamus. The arrow indicates a dilated tail of the cyst. p = posterior skull. (b) Doppler waveform analysis showed a bidirectional flow with extreme turbulence within the cystic malformation shown in (a). The 'chaotic' waveform indicates the mixture of various velocity vectors aimed in all directions within the aneurysmal malformation. (c) Power Doppler image of the same area shown in (a). The blood supply of the aneurysm (A) is mainly from a small contralateral aneurysm (1) over the location of the choroidal branches of the posterior cerebral artery. The small aneurysm (1) also receives blood from branches of the middle cerebral artery (2), and the anterior cerebral artery (3). The anterior communicating artery (4) is also shown. The venous drainage is directed toward the appendage of the aneurysm (arrow).

bidirectional blood flow with extreme turbulence within the cyst (Figure 1b).

In order to understand the angioarchitecture of the vascular lesion, the fetus was further investigated with

color power angio (CPA) and 3D CPA using the built-in program of the ultrasound machine (HDI-3000, Advanced Technology Laboratories, Bothell, WA, USA). The vascular anatomy, including the supply and drainage, were subsequently clearly depicted. A small contralateral aneurysm over the choroidal branches of the posterior cerebral artery, also connected to the anterior and middle cerebral arteries, was found to feed the aneurysm (Figure 1c). The venous drainage, including an aneurysmal falcine sinus, superior sagittal sinus, transverse sinus, and occipital sinus, were all clearly depicted by 3D CPA (Figure 2a). The diagnosis of an aneurysm of the vein of Galen was made based on the findings of B-mode, color Doppler interrogation, and also the angioarchitecture depicted by 3D CPA.

We underwent a systemic evaluation of the fetus. Cardiomegaly with minimal pericardial effusion was disclosed, yet no other cardiovascular pathology was detected. The resistance of the middle cerebral artery was significantly reduced (systolic/diastolic ratio 3.1), representing the 'steal' phenomenon of Galen's aneurysm, which leads a large proportion of brain circulation being shunted directly into the aneurysm. We did not discover any other structural anomalies such as hydrocephalus, leucomalacia, or other signs of hydrops fetalis. Parents were counseled about the prognosis and opted for a close follow-up at our clinic. The Galen's aneurysm remained stable, but cardiomegaly of the fetus spontaneously resolved 3 weeks after the first visit. The mother was admitted to our ward due to onset of labor at 37 weeks of gestation. A female baby weighing 3144 g was spontaneously delivered with an Apgar score of 6 and 8 at 1 and 5 min, respectively. Placenta abruptio was noted after delivery by disclosing the bloody amniotic fluid and retroplacental hematoma.

The baby was transferred to the neonatal intensive care unit for further care. Both magnetic resonance imaging (MRI) and digital subtraction angiography (DSA) confirmed the diagnosis of aneurysm of the vein of Galen, as well as the angioarchitecture depicted by antenatal 3D CPA (Figure 2b). Transarterial embolization of the aneurysm was attempted on day 11 but failed due to difficult approach through the small intracranial vessels. However, the hemodynamics remained stationary. Cardiomegaly was not seen after birth and the aneurysm decreased to 2.5×2.2 cm in size. The pediatricians suggested a conservative management and the baby was followed regularly at our pediatric department. The aneurysm decreased to 1.2×1.2 cm in size 1 year later and no other brain pathology was noted. There has been no growth and developmental delay. Neurologic development is within normal limits thus far (1 year and 3 months old), except for mild dynamic ataxia whilst walking.

DISCUSSION

Aneurysm of the vein of Galen is a complex arteriovenous malformation (AVM) consisting of multiple communications between the system of the vein of Galen and the cerebral arteries (carotid or vertebrobasilar systems)⁷. It may result in hemodynamic and central nervous system

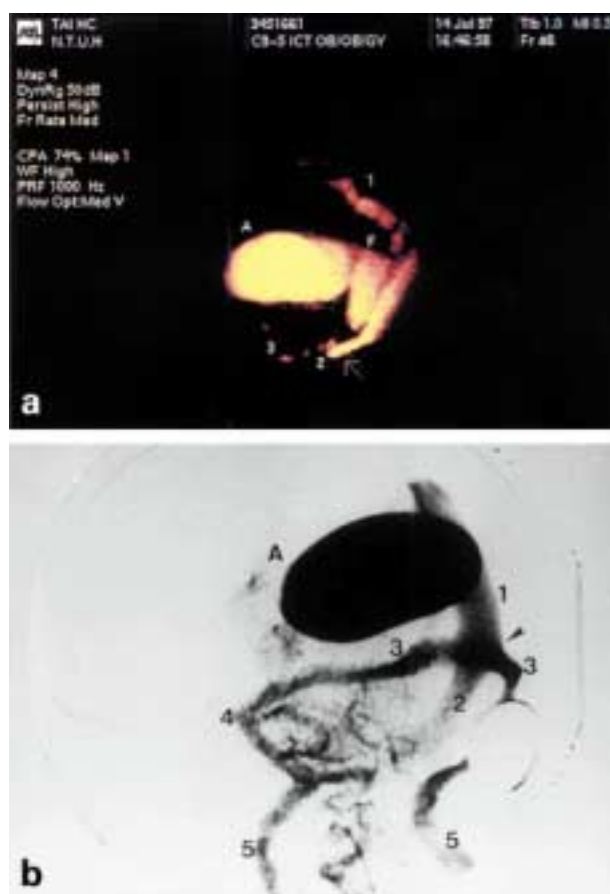


Figure 2 (a) The aneurysm of the vein of Galen assessed by transvaginal 3D CPA. The venous drainage of the aneurysm (A), including the aneurysmal falcine sinus (F), superior sagittal sinus (1), occipital sinus (2) and transverse sinus (3) and confluence of sinuses (arrow) are clearly depicted. (b) Postnatal angiography injected via the internal jugular artery (in venous phase) shows serial drainage sinuses of the Galen's malformation. 1 = superior sagittal sinus, 2 = occipital sinus, 3 = transverse sinuses, 4 = sigmoid sinus, 5 = internal jugular veins. A = aneurysm of the vein of Galen. The arrowhead indicates the confluence of sinuses. The falcine sinus is shaded by the Galen's aneurysm in this image and hence cannot be seen. The findings are consistent with the 3D CPA image shown in (a).

disturbances in the affected neonates *in utero*. Severe high-output cardiac failure is caused by marked increase of cardiac preload from venous return of brain due to the 'steal' phenomenon. Hydrocephalus may develop secondary to obstruction of the aqueduct of Sylvius by the dilated aneurysm. Compression of the cerebral cortex by the dilated ventricle and the 'steal' phenomenon, with diversion of blood from the parenchyma to the aneurysm, may further result in brain infarcts and leucomalacia⁸.

The diagnosis of the aneurysm of the vein of Galen mainly depends on imaging studies, including angiography⁹, ultrasound¹⁰, computed tomography¹¹ and MRI¹². Thus far, prenatal diagnosis of aneurysm of the vein of Galen has been made in several cases, essentially by pulsed Doppler and color flow mapping^{13–15}. Gray-scale ultrasound can detect anatomic changes (such as cardiomegaly or hydrocephalus), and color Doppler interrogation is helpful to assess the hemodynamics of the brain cyst. However, it is still difficult to delineate precisely the

feeding arteries and venous drainage, and to differentiate pure Galenic anomalies from parenchymal AVM draining into the vein of Galen merely by color Doppler sonography. Antenatal MRI was reported to be useful in making a definite diagnosis in this situation⁶.

This report demonstrated that 3D CPA is useful in prenatal detection and differential diagnosis of the vein of Galen anomaly by precise location of the aneurysm and depicting the corresponding angioarchitecture. The examining field of endovaginal probe is usually too limited to illustrate the detailed vascular anatomy of the target, especially when the vasculature is very complicated. Therefore, examiners can only mentally assemble and visualize the sectional vascular images in three dimensions. However, 3D CPA is advantageous in that it can allow us not only to display the 3D vascular image on the screen, but also to interactively assess the lesion from different perspectives (with $\pm 50^\circ$ horizontal rotation of the target).

The aneurysm of the vein of Galen represents an embryonic persistence of the median prosencephalic vein and not the vein of Galen *per se*¹⁶. The vascular anatomy of the aneurysm has wide variations of arteriovenous connections. In a review article of a pediatric series, the aneurysm is supplied invariably by choroidal branches from posterior cerebral artery (as shown in this case) and sometimes also from transmesencephalic arteries. Venous drainage is more complicated. Normally the vein of Galen connects first to a sagittal sinus, which then drains into the confluence of sinuses (the junction of straight, superior sagittal, occiput, and bilateral transverse sinuses)⁷. However, the straight sinus is usually absent or thrombosed in cases of aneurysm of the vein of Galen. Instead, the Galen's aneurysm usually drains into a so-called falcine sinus that extends directly into superior sagittal sinus, as shown in this case, rather than directly into the confluence of sinuses. Judging from the previous antenatal reports, it is difficult to recognize either the straight sinus or the abnormal falcine sinus connecting the Galen's aneurysms. In fact, identification of straight sinus drainage may allow a simple and effective modality—transthoracic embolization¹⁷ of the Galen's aneurysm via the confluence of sinuses after delivery.

The optimal management and prognostic indices for the neonates with Galen's aneurysm remain to be established. Generally, post-delivery 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^{7,18}. It is generally agreed that the presence of cardiac decompensation and gross brain pathology indicate adverse perinatal outcomes and necessitate active intervention. However, there are several reports of spontaneous thrombosis of aneurysm of the vein of Galen and also a case of spontaneous disappearance (without evidence of thrombosis) of the aneurysm^{19,20}. The hemodynamics usually worsens after delivery due to the removal of the low-resistance unit (placenta), which may facilitate more flow across the foramen ovale. In our case, however, the aneurysm spontaneously decreased in size before delivery

and regressed continuously after birth. Therefore, we believe that the natural process of the vein of Galen's malformation is more complicated than our previous understanding. Further investigation is necessary to reanalyse the prognostic factors of the affected fetuses.

In a recent article, the author reviewed the literature and scored the prognostic indices for those neonates complicated with a Galen's aneurysm²¹. Not only the presence of hydrops, brain changes (grave prognosis) and cardiomegaly (moderate risk), but also the angioarchitecture of the aneurysm would determine the perinatal outcome of the affected neonates. Multiple (five or more) feeding vessels and abnormal dilated venous drainage were scored as moderate fetal risk and prone to neonatal heart failure. Therefore, the angioarchitecture depicted by 3D CPA may help us in identifying the neonates prone to poor prognosis. As illustrated in this case, abnormal dilated venous drainage (falcine sinus) indicated the occurrence of heart failure before birth, although recovery followed delivery.

In short, we report an unusual case whose aneurysm of the vein of Galen and associated cardiomegaly spontaneously underwent an involution process after delivery. Prenatal detection of an aneurysm of the vein of Galen has been greatly facilitated by modern color Doppler techniques. This paper illustrates that 3D CPA may further assist the diagnosis of Galen's aneurysm by identification of angioarchitecture such as feeding and drainage vessels. This may then guide the postnatal treatment more precisely and may possibly help in identifying a fetus at risk of neonatal congestive heart failure and poor prognosis.

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