

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

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ABSTRACT

The prenatal diagnosis of an aneurysm of the vein of Galen has been reported on only a few occasions. This rare arterio-venous fistula is suspected when an intracerebral hypoechoic cyst is found in which blood flow can be demonstrated by Doppler ultrasound. This is one of the few conditions where Doppler ultrasound is critical for the diagnosis. Three-dimensional color power Doppler (3D-CPA) is a recent ultrasound modality which enables the three-dimensional visualization of vessels and which is more readily available than magnetic resonance imaging (MRI). We report a fetus in which an aneurysm of the vein of Galen was detected at 33 weeks' gestation and where the application of 3D-CPA enabled a better understanding of the spatial orientation and course of the dilated vessels. The neonate was successfully treated with coil angiography but subsequently died at 5 months of age from cardiac decompensation. Where fetal malformations involve the vascular system, 3D-CPA appears to be a promising technique.

INTRODUCTION

Aneurysms of the vein of Galen in the fetus are rare malformations and have been described in only a small number of case reports^{1–7}. The malformation is suspected on real-time imaging by detection of an intracranial cystic structure of which arachnoid, porencephalic or choroid plexus cysts, pineal tumours, intracerebral haematoma or choroid papilloma are the main differential diagnoses. In cases of aneurysm of the vein of Galen signs of volume overload, i.e. cardiomegaly, dilated neck venous vessels and hydrops will also be evident. Previous authors have reported confirming this diagnosis by identifying blood flow within the cyst using either spectral^{1–4} or color Doppler^{5,8–10}. It is of interest that this is the first

malformation where the application of Doppler ultrasound has confirmed the prenatal diagnosis¹.

Three-dimensional color power angiography (3D-CPA) is a new diagnostic technique allowing the spatial visualization of flow¹¹. We report on the prenatal application of 3D-CPA (HDI 5000, Advanced Technology Laboratories, Seattle, USA) in the diagnosis of an aneurysm of the vein of Galen and compare it with antenatal MRI images and postnatal X-ray angiography.

TECHNICAL ASPECTS OF CPA AND 3D-CPA

In CPA, the information acquired from Doppler signals is not analyzed by assessing frequency shift and direction of flow but by employing signal amplitude. The signal amplitude is proportional to the power inherent in blood cell motion and thus the method is three to five times more sensitive than conventional color Doppler. This enables small vessels and lower flows to be visualized. The image display is similar to that of color Doppler, with an overlay of a color image onto a gray scale background. This new technique has many advantages¹²:

- The side-by-side visualization of small and large vessels is possible due to perfusion rather than velocity analysis.
- The information from the amplitude from both signals tends to be additive, resulting in a powerful signal. This allows an angle-independent visualization of vessels. The uniform color display, independent of the direction of flow or transducer position, makes the method more suitable than color Doppler for the 3D reconstruction of vessels. This feature is one of the main advantages of the new technique, especially in fetal medicine.
- There is better edge definition in displaying flow, because the signals from the vessel wall and surrounding tissue have low amplitude.
- The color assigned to noise is uniform and is distinguishable from true flow signals, whereas in color Doppler noise is coded in the same colors as vessel flow.

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The main basic factors that have to be considered in evaluating a 3D imaging system are volume data acquisition, image rendering and image display^{11,12}.

Volumetric data acquisition is the assessment of information and parameters within a volume of interest. The acquired volume is generally a cine loop sequence of a defined number of 2D slices. The system must assess the spatial relationship of the structures to each other in the different 2D images. The data acquisition performed by a free hand sweep achieves a linear translation through the volume of interest. Because the transducer is moved manually the data collected allow only a qualitative 3D description of vessels. There is no calculation of angles or distances. This acquisition can allow a accurate spatial registration and is good enough to be used if certain assumptions are made, namely: (1) the volume sweep is in one direction only; (2) the data are acquired by translation; and (3) the interslice thickness is fixed and known.

To achieve a reliable fixed interslice thickness, the examiner should maintain a constant velocity of sliding or tilting of the probe during data acquisition. As most of the fetal vessels are at a similar depth and have a relatively short course a fanning acquisition movement gives the same information as a sliding one. This technique is highly operator-dependent and needs some training to obtain satisfactory results.

Image rendering is the process of creating a visual representation of the parameters of interest. In general, the principle used is the planar geometric projection of the 3D image, i.e. a 2D image representing the 3D data. The impression of the third dimension is generally given by the on-line rotation of the image on the screen and by different shadowing of anterior and posterior structures. The gray scale information is subtracted and only the image of the vessels is visualized three-dimensionally on a black background. The sharp edge definition characteristic of color power imaging plays an important role in the spatial representation, especially when gray scale information is omitted. As several vessels generally need to be visualized within the region of interest, the 3D appearance is supported by the rendering of the topology.

Image display is the visualization of the rendered representation for viewing. After the 3D images are reconstructed they are directly displayed on the ultrasound screen (image rendering). The colors used for vessel visualization are the same as those used in power Doppler, i.e. red, orange, blue, yellow and gray. The background noise and low amplitude signals are displayed using darker colors or, in one setting, with blue rendering. The image rotates on a vertical axis with a horizontal visualization angle of $\leq 45^\circ$ from the middle image. The vertical axis follows the same direction as the power Doppler ultrasound beam and is virtually positioned in the centre of the CPA box.

CASE REPORT

A 37-year-old, gravida 3, para 2 woman was referred for prenatal ultrasound assessment following the identification



Figure 1 Conventional B-mode image showing a large cystic structure in the fetal brain at 33 weeks' gestation. The atypical localization between thalamic area and sinus sagittalis is clearly demonstrated.

of an intracranial cystic structure at 33 weeks of gestation. The course of the pregnancy had been uneventful prior to referral. A large cystic structure, 16 mm in diameter and located between the thalamic area and the sinus sagittalis, was visualized on B-mode imaging (Figure 1). Blood flow in this mass, and its very tortuous course, were demonstrated using color Doppler and color power angio and led to the tentative diagnosis of an aneurysm of the vein of Galen. The position the aneurysm was atypical but was clearly demonstrated using 3D-CPA (Figure 2). Magnetic



Figure 2 The spatial structure of the vascular malformation (white arrows) is demonstrated by use of 3D-CPA. The aneurysm drains into a vertical vein (V. vert.) connecting to the sagittal sinus (Sinus sag.).



Figure 3 The prenatal MRI of the fetus, showing the same cystic structure as demonstrated on ultrasound (see Figure 1).

resonance imaging was undertaken which confirmed the 3D-CPA findings and the normal appearances on B-mode ultrasound imaging of the remaining intracranial structures (Figure 3). A mild tricuspid regurgitation and dilated neck vessels were the only signs of hemodynamic impairment identified. A cardiomegaly of 60 mm in length and 55 mm in width was noted on gray-scale imaging. Myocardial contractility was normal.

Interdisciplinary consultation with the neonatologists, neuroradiologists and the neurosurgeons confirmed that the chances of successful surgery by primary embolization of the aneurysm were very low.



Figure 4 Angiographic demonstration of the vascular structure, showing an image similar to that seen prenatally using 3D-CPA (see Figure 2).

A Cesarean section was performed at term at a perinatal centre and the prenatal diagnosis of an aneurysm of the vein of Galen was confirmed by X-ray-angiography (Figure 4). The fistulae were closed by coil angiography. After four treatments, the baby was discharged but died at 5 months of age owing to cardiac decompensation.

DISCUSSION

Arterio-venous fistulae including aneurysm of the vein of Galen can affect cardiac function and may lead to nonimmunologic hydrops fetalis^{3,13}. The dilated venous sac of the aneurysm probably represents persistence of the embryonic median prosencephalic vein and represents <1% of all intracranial arteriovenous malformations¹⁰. Aneurysm of the vein of Galen can include different cerebral arteriovenous malformations although the embryological development of these vascular malformations is unknown¹⁴.

The diagnosis is made postnatally by ultrasound and computer tomography. Clinical signs include cyanosis, systolic murmur, cardiomegaly and increased pressure with hydrocephaly. A congestive heart failure is a frequent complication.

The few case reports of the prenatal diagnosis of aneurysm of the vein of Galen have emphasized the importance of pulsed and color doppler^{4-6,8,9,15}. The most striking prenatal finding is the presence of a posterior, midline or slightly lateral supratentorial cystic structure in the fetal brain¹⁵. The sagittal sinus is reported as dilated in most cases^{3,8,10,13}. The turbulent vascular flow, diagnostic of an aneurysm, is detected by pulsed and color Doppler^{1,3-5,7-9}. The 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 described ventriculomegaly, cardiomegaly and dilated neck vessels as commonly found associated findings¹⁰ while Pulu reported the distortion of the cerebral architecture in cases with aneurysm of the vein of Galen as a negative sign¹⁵.

In most reported cases the diagnosis of the arteriovenous malformation has been made during the third trimester, frequently after the 34th week of gestation. The earliest reported diagnosis was made at 25 weeks' gestation⁹.

The introduction of 3D gray scale sonography into prenatal diagnosis has lead to an improvement of our understanding of the spatial appearance of fetal structures and thus to an improvement of diagnostic capability. The reconstruction of planes not usually seen on 2D ultrasound and the visualization of the spatial course of blood vessels are now possible using 3D-CPA. It is important to consider the settings of the CPA mode which will optimize acquiring a 3D image. Pulse repetition frequency should be set at 700 or 1000 Hz to assess small vessels. Where selective visualization of arteries with a higher flow is required, a higher pulse repetition frequency should be chosen. The

gain should be reduced by 30% or 40% as we have found that a lower gain setting gives a better image on 3D reconstruction. Wall filter should be set to 'medium' and the dynamic motion differentiation set to 'on' to avoid signals appearing as background noise on 3D. Sensitivity should be set to 'medium' and the priority to 'high'. We also found that it was important to select a high persistence especially when visualizing arteries in order to avoid pulsations of vessels during data acquisition. This enables a smooth continuity of vessel outline during sweeping of the transducer¹¹.

In the case reported here the 3D-CPA reconstruction revealed an image directly comparable to that of the postnatal angiogram. The 2D-CPA image was also comparable but the visualization of blood flow not sufficiently informative. We think the precise spatial description of vessel malformation achieved with 3D-CPA provides the multidisciplinary team involved in the management of such cases with more detailed information. This new diagnostic method, which is comparable to MRI, could thus improve perinatal counseling and evaluation of prognosis.

Although our case is similar to others reported in the literature concerning time of diagnosis and (poor) outcome, it is the first case reported where the application of 3D-CPA has been made. Our findings suggest that CPA, like MRI, is an informative additional diagnostic technique in fetal imaging which can aid in improving the prenatal diagnosis and subsequent decision making.

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