Vein of Galen abnormality

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Abstract
Vein of Galen abnormality is a rare congenital malformation of blood vessels of the brain and while often referred to as “Vein of Galen aneurysm” its is really not an aneurysm but usually an arteriovenous (AV) malformation. The following case report will be of interest to sonographers as it demonstrates early prenatal diagnosis of this intracranial vascular abnormality causing cardiac overload and subsequent cardiac failure. Diagnosis of the AV malformation can be graphically demonstrated using colour Doppler or 3D colour power angiography.

Introduction
The anterior and central regions of the brain including the choroid drain into the internal cerebral veins (vein of Galen) and then the great cerebral vein of Galen before continuing into the inferior sagittal sinus, internal jugular vein and right side of the heart.

Vein of Galen abnormality is a rare sporadic abnormality of unknown origin and results from an aneurysmal malformation with an arteriovenous shunting of blood. The congenital malformation occurs during weeks 6–11 of fetal development and whilst the actual incidence of vein of Galen abnormality is unknown, it is rarely diagnosed in early pregnancy. While several references to vein of Galen abnormality (thrombosis/fistular) at 21–22 weeks have been reported, most prenatal diagnosis has been at 32–34 weeks gestation. It is however the most common cerebral arteriovenous (AV) malformation and is usually associated with high cardiac output leading to cardiac failure in approximately 95% of cases. It may also be associated with hydrocephalus, cerebral haemorrhage or cerebral ischaemia and infarction.

In the majority of cases, the diagnosis is made in early childhood usually following the development of cardiac failure or, less often, subsequent to the onset of neurological signs following cerebral haemorrhage or ischaemia. While associated with high infant mortality, successful treatment of less serious cases has been reported.

Fig. 1: Cardiomegaly.

Fig. 2: Dilated internal jugular vein.

Case history
The patient was a 36-year-old primigravida. Down syndrome screening was considered low-risk and an 18-week morphology scan had been performed elsewhere by an experienced sonographer; no abnormality was reported.

Ultrasound review of the baby at 22 weeks was performed by her obstetrician as a routine office procedure and the heart was noted to be abnormal.

The patient was referred for cardiac review and was scanned twice over the next two days with a GE Voluson 730 Expert (GE Healthcare, Wakesha, Wisconsin, USA) revealing cardiomegaly with the heart lying horizontally rather than obliquely in the chest and with a cardio-thoracic ratio of 0.82.

It was considered that there was slightly greater enlargement of the right heart, especially of the right atrium.

Both the mitral and tricuspid valves functioned normally and more specifically there was no evidence of Tricuspid regurgitation noted on colour or spectral imaging. However the pulmonary trunk was dilated and there was a dilated vessel in the neck which was considered to be the internal jugular vein draining into the superior vena cava and dilated right atrium. A significant hydrothorax and small pericardial effusion was also noted confirming evidence of early foetal cardiac failure.

In the absence of an obvious cardiac cause for the
heart failure, colour Doppler examination of the head was performed which demonstrated a massive arteriovenous abnormality (fistula) of the vein of Galen with excessive blood draining from the internal and great cerebral veins into the inferior sagittal sinus and right side of the heart.

**Discussion**

Vein of Galen abnormalities are rare and can be associated with massive AV malformations resulting in excessive venous drainage into the heart and subsequent cardiac failure. In most reported cases, the abnormality has been diagnosed following delivery but our case is interesting in that the diagnosis was made at 22 weeks gestation following the unexpected identification of cardiomegaly. Probably the take-home message is that, in cases of intrapartum cardiomegaly and cardiac failure, it is essential that intracranial AV malformation (vein of Galen aneurysm) is excluded as a cause. While standard 2D imaging will reveal the vascular abnormality, colour and power Doppler with spectral imaging will provide a more accurate and graphic display of the arteriovenous malformation and identify the reason for the cardiac overload and subsequent heart failure.

The differential diagnosis includes arachnoid cyst, porencephalic cysts, cavum septum pellucidum and quadrigeminal cistern but none of these will demonstrate flow on Doppler evaluation.

The prognosis in this case was, unfortunately, poor and as we expected intrauterine foetal death occurred several weeks after our initial ultrasound diagnosis.

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**Further reading**

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