An incidental diagnosis of a vein of Galen aneurysmal malformation at 38+5 weeks gestation

Introduction
Vein of Galen aneurysmal malformations (VGAM) are abnormalities of the cerebral vascular circulation. These arteriovenous malformations can have serious cerebral and cardiac consequences. They are associated with a high level of mortality and morbidity and typically present either antenatally or postnatally with cardiac failure. If allowed to progress untreated, multiorgan failure and death are observed.[1]
This case report describes the incidental diagnosis of a VGAM at 38 weeks 5 days gestation during a routine fetal growth and wellbeing ultrasound scan. The ultrasound presentation of this condition and the benefit of an antenatal diagnosis will be discussed.

Case Presentation
A 26 year old G2 P0 woman attended the ultrasound department for a growth scan at 38+5 weeks gestation due to a history of reduced fetal movements. She had declined combined screening and her dating and anatomy scans were reported as normal. A growth scan performed at 31+5 had not identified any abnormalities.
Ultrasound examination revealed a single live fetus in a breech presentation. An elongated cystic structure was demonstrated in the midline region of the fetal brain (Fig. 1.). Colour and pulsed wave doppler interrogation of this area showed it to contain high velocity mixed arterial and venous flow (Fig. 2.). No further abnormalities were detected and the growth velocity, liquor volume and umbilical artery doppler traces were all within normal limits. A diagnosis of a VGAM was suspected.
The patient was transferred to the local tertiary referral centre for repeat scan with a fetal medicine consultant. They agreed with the initial ultrasound findings and the patient was transferred out of area for delivery so that the child would be received at a national centre of expertise for further management immediately after birth.
An in utero MRI was performed at 39 weeks 1 day gestation and a live female was delivered by caesarean section the next day. The neonate clinically worsened in the first week of life and at day 25 underwent an embolization procedure which was complicated by an intraventricular haemorrhage. The child is now doing well but does have some right hemiparesis.

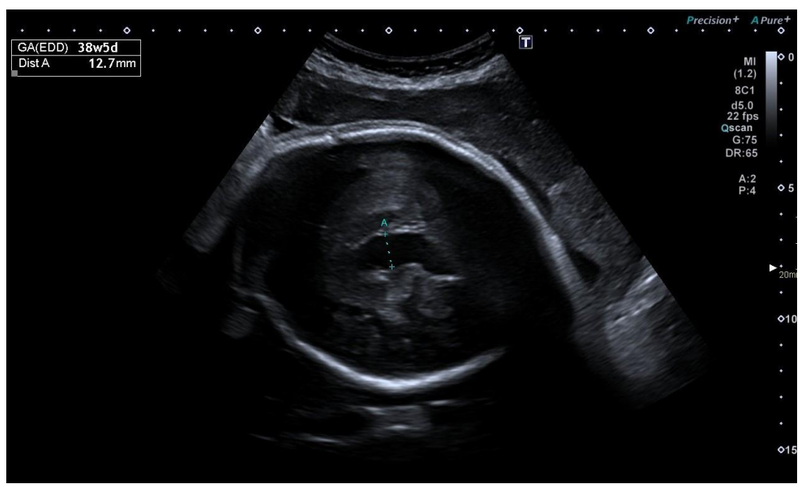

Fig. 1.

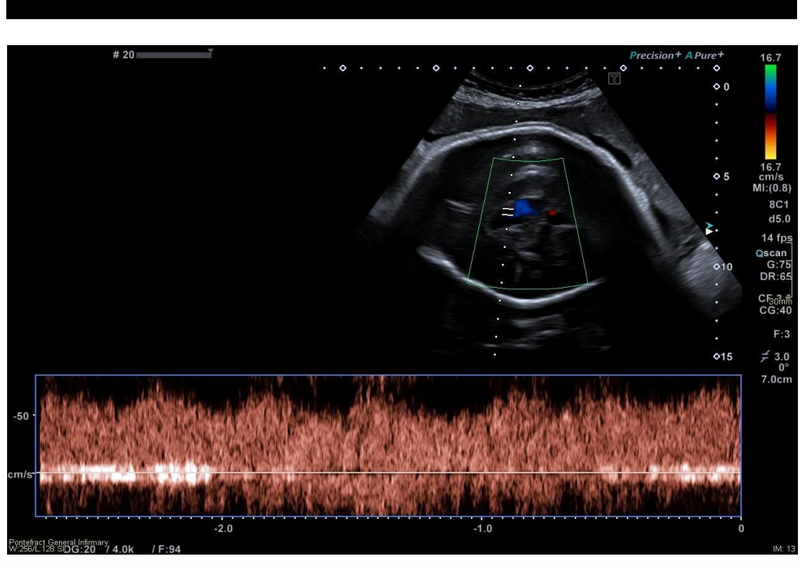

Fig. 2.

Discussion
A VGAM is a rare congenital defect thought to occur between 6 and 11 weeks gestation. These anomalies are the result of an arteriovenous connection between the choroidal arteries and the embryonic medial prosencephalic vein.[2] The incidence of VGAM is less than 1 in 25000 births yet they account for 30% of paediatric vascular malformations.[3] Whilst the cause for VGAM is unknown some recent discoveries about possible genetic origins have been made.[3]
A VGAM does not usually become visible on ultrasound until the third trimester however there have been reports of second trimester diagnoses. Antenatally, ultrasound will usually demonstrate a midline intracranial cystic structure posterior to the third ventricle with turbulent internal blood flow on doppler interrogation as was discovered in this case.[4] The use of colour doppler helps to rule out some other differential diagnoses such as an arachnoid cyst or Dandy Walker malformation.[4] It is also possible for the lesion to appear hyperechoic if there has been spontaneous thrombosis in utero.[3]
The clinical presentation of VGAM varies depending upon the time of diagnosis. Antenatally it can be an isolated finding as in this case. Due to the strain placed upon the right side of the heart as a result of the increased venous return by the lesion there can be indicators of cardiac failure such as cardiomegaly, fetal hydrops and polyhydramnios. Ventriculomegaly can also occur due to both compression from the malformation and the prevention of reabsorption of the cerebrospinal fluid by the increased pressure in the cerebral veins.[3] In the early neonatal period heart failure is the most common presentation whereas older children will usually have neurodevelopmental problems.
Treatment requires embolization of the defect with either glue, or less commonly, coils.[1] These procedures are frequently complicated with the risk of haemorrhage, thought to be due to the immature and fragile nature of the vessels at this young age.[5]
In the United Kingdom, children with VGAM are treated at one of two specialist centres in either London or Glasgow.[1] This centralised management has been shown to improve outcomes, particularly in neonates.[2] An antenatal diagnosis and intrauterine transfer as was achieved in this case, provides optimum management, negating the need for a potential unstable relocation of a critically unwell child.

References
1. Lecce et al. (2018). Cross-Sectional Study of a United Kingdom Cohort of Neonatal Vein of Galen Malformation. Annals of Neurology, 84: 547-555
2. Nunez, F. and Dohna-Schwake, C. (2021). Epidemiology, Diagnostics, and Management of Vein of Galen Malformation. Pediatric Neurology, 119: 50-55
3. Thorey et al. (2022). State of the art of antenatal diagnosis and management of vein of Galen aneurysmal malformations. Prenatal Diagnosis, 42: 1073-1080
4. Deloison et al. (2012). Hidden mortality of prenatally diagnosed vein of Galen aneurysmal malformation: retrospective study and review of the literature. Ultrasound Obstet Gynecol, 40: 652-658
5. Karadenix et al. (2010). Vein of Galen aneurysmal malformation: Prenatal diagnosis and early endovascular management. Journal of the Chinese Medical Association, 74: 134-137

Working together
making a difference

J.Johnson, Advanced Practitioner Sonographer