

Pial arteriovenous fistulas: dilemmas in prenatal diagnosis, counseling and postnatal treatment. Report of three cases

C. GAREL*, M. AZARIAN†, P. LASJAUNIAS‡ and D. LUTON†

Departments of *Paediatric Imaging and †Gynaecology and Obstetrics, Hôpital Robert Debré, Paris and ‡Department of Diagnostic and Therapeutic Neuroradiology, CHU de Bicêtre, Le Kremlin Bicêtre, France

KEYWORDS: arteriovenous malformation; brain; fetus; heart failure; infant; magnetic resonance imaging; newborn

ABSTRACT

We report three cases of the antenatal appearance of a pial arteriovenous fistula (AVF). In Case 1, the diagnosis of pial AVF was made by ultrasound examination at 32 + 3 weeks of gestation and confirmed by magnetic resonance imaging (MRI) at 34 + 3 weeks of gestation. After birth, the neonate had cardiac insufficiency and an embolization was performed 8 days postpartum which was followed by extended cerebral ischemia and death. In Cases 2 and 3 pial fistulae were diagnosed at birth. In Case 2, the patient was referred following the diagnosis of an aneurysmal malformation of the vein of Galen at 38 weeks of gestation. Owing to immediate delivery following her transfer, no ultrasound examination was performed and the diagnosis of pial AVF was established on the first postnatal day. An embolization was performed and the child is doing well. Case 3 presented with cardiomegaly at 30 weeks of gestation and cerebral ischemic lesions after birth.

These cases highlight the difficulties encountered in the prenatal diagnosis of pial AVFs. The diagnosis should be considered in cases of unexplained cardiomegaly and it is also important to make sure that AVFs draining into the vein of Galen are not misdiagnosed as aneurysmal malformations of this vein. The complications of pial AVFs are cardiac failure and cerebral ischemia. Embolization is the treatment of choice; however it is a risky procedure in neonates. Copyright © 2005 ISUOG. Published by John Wiley & Sons, Ltd.

CASE REPORTS

Case 1

Routine fetal ultrasound examination in a 35-year-old woman revealed at 30 weeks of gestation a posterior

midline anechoic intracerebral lesion in which blood flow was seen by color Doppler (pulsed Doppler was not performed). The first diagnostic impression was that of a vein of Galen aneurysm. The patient was referred to our institution for ultrasound examination at 32 + 3 weeks of gestation. This showed cardiomegaly and a pial arteriovenous fistula (AVF) located in the right hippocampal fissure draining into the vein of Galen (Figure 1). This diagnosis was confirmed by fetal magnetic resonance imaging (MRI) performed at 34 + 3 weeks (Figure 2). Because of the association with a small vermis and gyration abnormalities of the right temporal lobe, termination of pregnancy was proposed but rejected by the parents for religious reasons. The karyotype was normal. There were no signs of cardiac failure in the antenatal period.

At 37 weeks of gestation, a 2970-g male newborn was delivered vaginally. The neonate rapidly developed cardiorespiratory distress associated with cardiomegaly, high-output cardiac failure and pulmonary hypertension.

On the first day of postnatal life, ultrasonography (Figure 3) and MRI confirmed the antenatal diagnosis. On the 8th day, transfontanellar Doppler ultrasonography showed a diastolic steal phenomenon. Arteriography was performed and showed a pial right temporal AVF mainly supplied by the anterior temporal branch of the right posterior cerebral artery and draining mainly into the vein of Galen and the straight sinus. An embolization was performed and this led to a partial occlusion of the AVF. An erratic embolization of a temporal division of the right Sylvian artery occurred during withdrawal of the catheter. Immediate clinical evolution showed normalization of the central and peripheral hemodynamic parameters with persistence of a small diastolic steal in the isthmus aorta. MRI follow-up at 3 weeks showed a dramatic decrease in the size of the AVF and ischemic damage of both the right Sylvian and posterior cerebral territories. The neonate died shortly after this procedure.

Correspondence to: Dr C. Garel, Department of Paediatric Imaging, Hôpital Robert Debré, 48 Boulevard Sérurier 75019 Paris, France (e-mail: catherine.garel@rdb.ap-hop-paris.fr)

Accepted: 14 January 2005

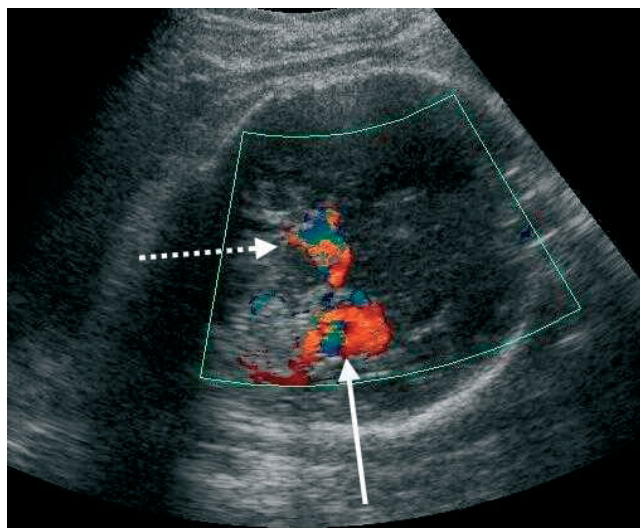


Figure 1 Case 1: Ultrasonography performed at 32 + 3 weeks. The pial AVF is visible (arrow) in the right hippocampal fissure. More medially, enlarged feeding arteries are also depicted (broken arrow).

Case 2

A 23-year-old woman was referred to our institution at 38 weeks of gestation because of the detection by ultrasonography of a posterior midline anechoic intracerebral lesion diagnosed as a vein of Galen aneurysm. Ultrasound examination performed at 22 and 33 weeks of gestation was normal. No further fetal examination could be performed because the patient delivered a 4070-g male newborn vaginally, immediately after having been transferred.

On the first day of postnatal life, cardiac ultrasonography was normal and transfontanellar ultrasonography showed a left medial frontal pial AVF located in the interhemispheric fissure, which drained into a pericallosal vein and then into the vein of Galen. MRI was performed on the same day and confirmed this diagnosis without evidence of brain damage. Partial embolization of the AVF was performed.

At the time of writing the child was 22 months of age and had normal neurological and psychomotor development.

Case 3

Details of Case 3 have previously been reported; a routine fetal ultrasound examination in a 30-year-old woman was performed at 30 weeks of gestation and revealed unexplained cardiomegaly¹. Cerebral ultrasonography (without Doppler) was normal. At 37 weeks, a 3490-g female neonate was delivered vaginally. Physical examination revealed a transfontanellar murmur. Transfontanellar Doppler ultrasonography showed a right parietal pial AVF. MRI confirmed the presence of a pial AVF located in the right Sylvian fissure which was associated with an enlarged right Sylvian artery. The right cerebral hemisphere was atrophic and presented MRI signal abnormalities. The neonate rapidly developed signs of congestive

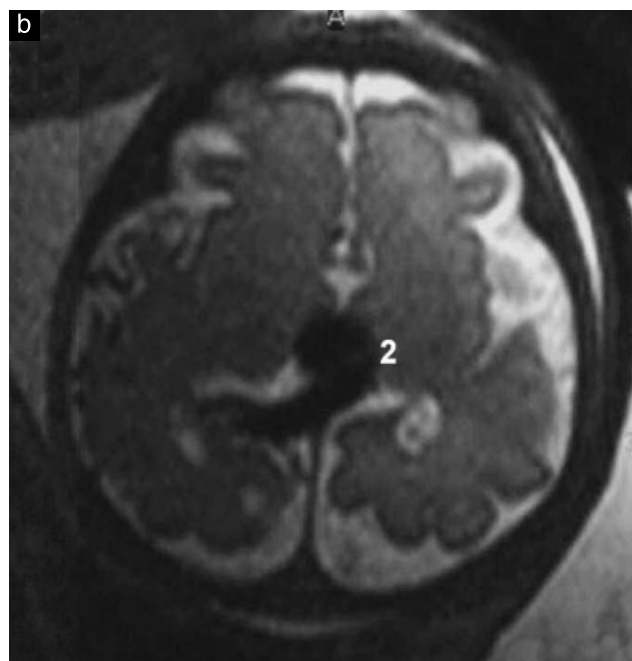
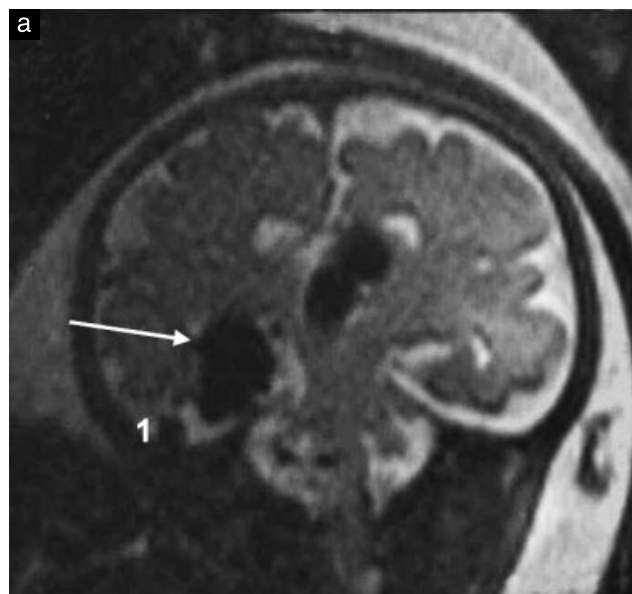


Figure 2 Case 1: Fetal MRI performed at 34 + 3 weeks. The pial AVF is visible in the right hippocampal fissure (arrow) as is the right petrous vein (1) (a), while the vein of Galen (2) is visible in (b). Notice the asymmetry of gyration of both temporal lobes and the dilated vessel in the pericerebral space on the right.

heart failure. A partial embolization was performed at 6 days of age. The child was 6 years old at the time of writing and had left hemiparesis.

DISCUSSION

The vast majority of cerebral vascular malformations reported in the antenatal literature are aneurysmal malformations of the vein of Galen (VGAM)^{2,3}. This anomaly consists of dilatation of the median prosencephalic vein associated with AVFs within the wall of the vein or multiple shunts communicating with the anterior

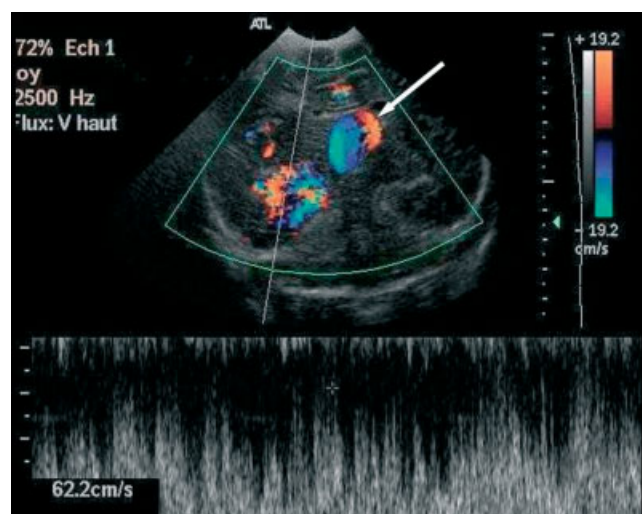


Figure 3 Case 1: Transfontanellar ultrasonography performed on the first day of postnatal life. Color Doppler shows aliasing phenomenon within the AVF. Spectral analysis demonstrates turbulent flow. Conversely, color Doppler within the vein of Galen (arrow) does not show any aliasing phenomena. The spectral analysis (not shown) revealed normal venous-type flow.

part of the vein. Other types of intracranial vascular malformations are exceptional in fetuses. Dural sinus malformations often present in the antenatal period⁴. These anomalies are caused by the persistence of an embryonic sinus or persistent enlargement of a sinus with secondary arteriovenous shunt within the wall of the dural lake.

Pial AVFs correspond to an abnormal communication between arteries and veins. Most of the arteries feeding the pial AVF open into a single ectatic draining vein^{5,6}. They may be located in the posterior fossa or more commonly in the supratentorial space^{6,7}. Supratentorial AVFs are always superficial and are fed by pial (cortical) arteries. As for dural sinus malformations, there is a male dominance. To our knowledge, pial AVFs revealed in the antenatal period have been reported only once⁸. Yet AVFs account for 17% of all intracranial arteriovenous malformations in children⁷. Pial AVFs are rarely diagnosed *in utero*, and this may be due to a lack of understanding of the condition and because AVFs may be peripheral and hard to visualize (e.g. Case 3).

The main vascular and clinical features of VGAM and pial AVFs are summarized in Table 1. As illustrated by our three cases, the following ultrasound signs should lead to the suspicion of cerebral pial AVF:

- Unexplained cardiomegaly (with increased velocities and without cardiac malformation) (Case 3). AVFs (and VGAM) may be responsible for high-output congestive cardiac failure.
- Dilation of the vein of Galen (Cases 1 and 2) may result from drainage of pial AVFs into the deep venous system, and should not be misdiagnosed as a VGAM^{1,9}. Two of the three patients (Cases 2 and 3) reported here were referred to our institution following the diagnosis of VGAM. In order to differentiate these two entities, it is essential when performing ultrasonography to use Doppler imaging (color and pulsed Doppler). In the case of a VGAM, Doppler will show mixed arterial and venous flow within the VGAM and venous-type flow in the draining vessels². In the case of a pial AVF, however, there will be a venous-type flow both in the dilated vein of Galen and in the vessels draining into this vein. Moreover, the use of Doppler imaging is indispensable in searching for the pial AVF draining into the vein of Galen.
- Visualization of the site of the pial AVF is the main diagnostic feature of this condition. AVFs must be searched for carefully, bearing in mind that they may be located close to the midline (Case 2) or more laterally (Cases 1 and 3). The advantage of fetal MRI over ultrasonography in the depiction of the pial AVF resides in the possibility of examining the pericerebral space without being hindered by the cranial vault, and whatever the position of the fetus's head. The feeding and draining vessels are more clearly visible than with ultrasound (Case 1). Fetal MRI may also be helpful in the diagnosis of associated brain anomalies as demonstrated in Cases 1 and 3.

The prognosis of this type of cerebral vascular malformation when diagnosed during fetal life, as in cases of postnatal diagnosis, depends on the presence of congestive cardiac failure and associated cerebral ischemic lesions.

It is of paramount importance to differentiate in fetuses a pial AVF from a VGAM. In asymptomatic neonates with a VGAM, the goal is to try to wait until the fifth or sixth month to perform the first diagnostic and therapeutic angiography². Conversely, in asymptomatic neonates with a pial AVF, cerebral tissue injury can develop rapidly, and one should try to significantly reduce shunting through the fistula as soon as possible after delivery in order to avoid loss of brain substance and to allow neurocognitive development and normal brain

Table 1 Vascular and clinical features of vein of Galen aneurysmal malformations (VGAM) and pial arteriovenous fistula (AVFs)

	Vascular features			Clinical features		
	VGD	DSG	CVD	Macrocrania	Cardiac changes	Ischemic cerebral damage
VGAM	+	+	–	+	+	+
Pial AVF	+/-*	+/-*	+	+/-*	+/-	+

*Observed (+) in the cases of a midline drainage. CVD, cerebral venous dilation; DSG, dural sinus dilation; VGD, vein of Galen dilation.

maturation^{3,10}. Embolization is the treatment of choice. Demonstration of the location and nature of the AVF, the arterial supply and the number and location of draining veins can be anticipated by MRI prior to the invasive procedure. Parents should be counseled that this is a high-risk procedure for neonates and that fatal complications may occur⁹. Thus, embolization should be performed in highly specialized centers.

REFERENCES

1. Rey F, Vuillard E, Sibony O, Magnier S, Oury JF, Luton D. Prenatal diagnosis of pial-superficial arteriovenous malformation. *Fetal Diagn Ther* 2002; **17**: 255–256.
2. Lasjaunias P. Vein of Galen aneurysmal malformation. In *Vascular diseases in neonates, infants and children*, Lasjaunias P (ed). Springer: Berlin, 1997; 67–202.
3. Garel C. Intracranial space-occupying lesions. In *MRI of the fetal brain: normal development and cerebral pathologies*, Garel C (ed). Springer: Berlin, 2004; 177–200.
4. Barbosa M, Mahadevan J, Weon YC, Yoshida Y, Ozanne A, Rodesch G, Alvarez H, Lasjaunias P. Dural sinus malformations (DSM) with giant lakes, in neonates and infants. *Intervent Neuroradiol* 2003; **9**: 407–424.
5. Lasjaunias P. Pial arteriovenous malformation. In *Vascular diseases in neonates, infants and children*, Lasjaunias P (ed). Springer: Berlin, 1997; 203–319.
6. Yoshida Y, Weon YC, Sachet M, Mahavedan J, Alvarez H, Rodesch G, Lasjaunias P. Posterior fossa single hole arteriovenous fistulas in children. *Neuroradiology* 2004; **46**: 474–481.
7. Weon YC, Yoshida Y, Sachet M, Mahavedan J, Alvarez H, Rodesch G, Lasjaunias P. Supratentorial cerebral arteriovenous fistulas in children: review of 41 cases with 63 non choroidal single-hole AVFs. *Acta Neurochir (Wien)* 2005; **147**: 17–31.
8. Auyeung KM, Laughlin S, terBrugge KG. Prenatal diagnosis of unusual fetal pial arteriovenous malformation. *Intervent Radiol* 2003; **9**: 163–168.
9. Boet R, Poon WS, Chan MSY, Yu SCH. Childhood posterior fossa pial-dural arteriovenous fistula treated by endovascular occlusion. *Child's Nerv Syst* 2001; **17**: 681–684.
10. Rodesch G, Malherbe V, Alvarez H, Zerah M, de Victor D, Lasjaunias P. Non galenic cerebral arteriovenous malformations in neonates and infants. Review of 26 consecutive cases (1982–1992). *Child's Nerv Syst* 1995; **11**: 231–241.