Management of children with high flow arteriovenous shunts of the brain is among the most challenging areas in modern medicine. Intracranial arteriovenous shunts (AVS) in children differ considerably from those seen in adults, in whom brain arteriovenous malformations (AVMs) and acquired dural arteriovenous fistulae predominate. These differences are seen both in the types of lesion and in their effects. In the neonatal and infantile age groups, the most common type of AVS is the vein of Galen aneurysmal malformation (VGAM), which has a male-to-female ratio of 3:1. Progressing further into childhood, dural malformations and brain arteriovenous malformations become more common.

The consequences of an AVS in the developing brain are different from those in an adult, principally because of the immature cerebral venous system: the arachnoid granulations by which cerebrospinal fluid will be returned to the cerebral venous sinuses are not fully matured until 16–18 months of age.

In infancy, cerebrospinal fluid is reabsorbed across the ventricular ependyma and brain parenchyma into the medullary veins. The presence of a large AVS such as a VGAM may raise venous sinus pressure, which is transmitted in turn to the cortical and finally the medullary veins. This will result in water congestion of the brain parenchyma, and impaired oxygenation leading to subependymal atrophy and in severe cases a progressive “melting brain syndrome”.

The most common presentation of VGAM results from the size of the shunt itself, imposing elevated preload on the right side of the heart leading to cardiac failure. This may progress to multi-system failure. Haemorrhage in children with VGAMs is rare.

These are rare lesions and experience in their management has been restricted generally to large paediatric centres where a close collaboration between neuroradiologists, neonatologists, paediatric cardiologists, and neurologists has been achieved. Foremost among these centres has been Bicetre Hospital in Paris where Professor Pierre Lasjaunias’ group has done much to clarify the nature of the disease and its appropriate management. A typical neurosciences unit serving a population of about three million could expect approximately one new VGAM patient a year. The aetiology of VGAMs is unknown; however, an early insult, perhaps resulting in a somatic mutation in neural crest and/or adjacent cephalic mesoderm in the early embryo, could be expected to cause such vascular abnormalities.

ANATOMY OF VGAM

The VGAM is a rare and dramatic form of embryonic arteriovenous shunt located in the midline in the choroidal fissure. It consists of multiple feeding arteries, principally the anterior and posterior choroidal arteries and the anterior cerebral artery (following the cingulate gyrus and representing the embryonic limbic arterial arch), draining directly into an enlarged venous pouch (fig 1). This usually grossly dilated vein was recognised by Raybaud and colleagues to be the embryonic precursor of the vein of Galen—the median vein of the prosencephalon. This malformation develops before the formation of the vein of Galen and straight sinus, and the pouch drains via a falci- nal sinus to the superior sagittal sinus. The deep venous system of the brain does not usually communicate with the venous pouch, but rather drains by mesencephalic collateral veins often visible with an epsilon shape (fig 2). The VGAM should be distinguished from aneurysmal dilatation of the true vein of Galen caused by an adjacent brain AVM (vein of Galen aneurysmal dilatation or VGAD) which has a higher risk of haemorrhage.

PRESENTATION AND DIAGNOSIS OF VGAMS

VGAMs are occasionally detected on antenatal ultrasound scans (from about 25 weeks gestation) as apparently cystic midline brain lesions, colour flow Doppler then suggesting a VGAM. Antenatal magnetic resonance imaging will confirm the diagnosis and allow assessment of any pre-existing damage to the brain (fig 3). It will also allow treatment planning with delivery at a centre with the appropriate facilities and expertise, principally fetal medicine, neonatology, paediatric cardiology and intensive care, and interventional neuroradiology. A team approach is critical to successful management.
Angiography is best performed at the time of embolisation. Be by magnetic resonance imaging or computed tomography.

Initial assessment of the AV shunt should be normal; transfontanelle ultrasound will detect the cardiac cause for the high output failure. When cardiac examination is normal, transfontanelle ultrasound will detect the cardiac cause for the high output failure. When cardiac examination is normal, transfontanelle ultrasound will detect the cardiac cause for the high output failure. When cardiac examination is normal, transfontanelle ultrasound will detect the cardiac cause for the high output failure. When cardiac examination is normal, transfontanelle ultrasound will detect the cardiac cause for the high output failure. When cardiac examination is normal, transfontanelle ultrasound will detect the cardiac cause for the high output failure. When cardiac examination is normal, transfontanelle ultrasound will detect the cardiac cause for the high output failure. When cardiac examination is normal, transfontanelle ultrasound will detect the cardiac cause for the high output failure. 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permanent neurological deficits. In the infant presentation group of 35 babies, embolisation was performed in 26 with 22 having a good neurological outcome. Twelve children presented later, nine requiring embolisation and six of these having a normal course. The worst prognosis was thus seen, as expected, in the babies with the largest shunts, presenting as neonates with severe cardiac failure.

Untreated VGAMs have a poor prognosis and are almost always fatal, but it is equally important to recognise that treatment may be inappropriate in some cases. Where there is evidence of pre-existing brain damage (progressive atrophy or “melting brain syndrome”, parenchymal calcification, and so on) or severe multiorgan failure, a poor outcome (death or survival with severe brain damage) is inevitable.

CONCLUSIONS

High flow arteriovenous shunts of the brain in young children are among the most challenging conditions treated in modern medicine. Early and appropriate diagnostic tests (mainly ultrasound and magnetic resonance imaging) permit the proper management by a team that should include interventional neuroradiologists, neonatologists, paediatric cardiologists, and neurologists. Where possible, VGAMs are treated medically until 5–6 months of age, while unstable babies require early treatment, principally with cyanoacrylate embolisation.

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