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 multisystem 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 falxine 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.

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worsening cardiac failure leading on to multiorgan failure. A
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VGAMs, of 21 babies diagnosed antenatally, 14 were consid-
ered fit for embolisation and 12 of these had normal
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Although hydrocephalus may be well tolerated (with open
fontanelles), overt cardiac failure has a very poor prognosis. This
fetus died in utero.

More commonly VGAMs are diagnosed after birth. Often
delivery and the first 24 hours are unremarkable. Larger
shunts may then show rapid deterioration with progressively
worsening cardiac failure leading on to multiorgan failure. A
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TREATMENT AND OUTCOME IN VGAMS

Ideally the initial treatment of VGAM is conservative. Embolisa-
tion of a neonate is a high risk procedure and where possible we
treat the child medically (for cardiac failure) until aged 5 or 6
months with regular outpatient assessment. Elective embolisa-
tion can be scheduled for this time with the aim of closing the
AVS with cyanoacrylate glue by the arterial route. If the infant
deteriorates (seizures, failure to thrive, worsening cardiac

failure, etc) treatment is performed earlier. Surgery has little
role in the modern treatment of VGAM. Surgical attempts at
closure of the shunt have a very high mortality or severe
morbidity, and shunting of the ventricles before embolisation
may accelerate progressive atrophy (“melting brain”).

Embolisation in a 5–6 month old baby is technically easier
than in a neonate (fig 4). Navigation of microcatheters to the
site of the shunt can still be difficult though, because of the
often extreme tortuosity of the arterial system. Embolisation is
best performed with liquid embolic agents, usually a pure
cyanoacrylate glue such as Histoacryl (Braun, Melsungen, Ger-
man) opacified with tantalum powder. We have found newer
cyanoacrylates such as Glubran-2 polymerise too slowly for safe
use in this high flow situation. The alternative transtorial
venous approach (although technically easier) is probably asso-
ciated with a higher risk of haemorrhagic complications (and
lower success rate) and we have normally avoided this route.

Restraints on the amount of parenteral fluid that can be
given to a neonate or infant (contrast medium, flush
solutions, and so on) impose limits on the duration of the pro-
cedure. Often a large shunt with many feeding vessels will
need several embolisation sessions. We have varied from com-
plete occlusion of small lesions in a single session, to a maxi-
um of six sessions for complete occlusion.

Progressive stenosis of the jugular bulbs complicates the
course of many VGAM babies. In the neonate, cortical veins do
not yet communicate with the cavernous sinus and there is thus
no other route for venous outflow. Once cavernous capture, by
the cortical venous system, occurs (during the first year) alter-
native pathways become available (via the ophthalmic and
pterygoid veins). Stenting of the jugular bulb in VGAM babies
was first described by Dr Wendy Taylor’s group at Great Ormond
Street Hospital (now Cromwell Hospital) in London.7 Although
short term results have been promising, the long term patency
of stents in the venous system is unknown.

Where treatment is performed before significant brain
damage has occurred, a good outcome is anticipated. In
Lasjaunias’s series,7 the largest discussing the management of
VGAMs, of 21 babies diagnosed antenatally, 14 were consid-
ered fit for embolisation and 12 of these had normal
neurological development. In the neonatal diagnosis group of
50 babies, 19 had pre-existing irreversible damage, and 29
were considered fit for embolisation, 50% of these showing
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.

References