Presentation, imaging features, and endovascular treatment of vein of Galen aneurysmal malformations in the neonatal period and early infancy.

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Learning objectives

To describe the clinical presentation, imaging features, and endovascular management of vein of Galen aneurysmal malformation (VGAM), with reference to five cases treated with transarterial embolisation in our institution.
Background

VGAM is a severe paediatric neurovascular developmental abnormality. It is characterized by arteriovenous shunts draining into a dilated median prosencephalic vein of Markowski\(^1\). The name "vein of Galen malformation" is therefore a misnomer, as the dilated vein is actually the median prosencephalic vein of Markowski, the embryonic precursor to the vein of Galen\(^2\).

Vascular anatomy

The exact aetiology is unclear, but arteriovenous shunts are believed to develop between 6 and 11 weeks gestation.\(^2\) Two distinct types are recognized, based on vascular anatomy: the choroidal type in which multiple feeding choroidal arteries empty into a nidus adjacent to the dilated vein; and the mural type in which direct arteriovenous fistulae are seen in the wall of the dilated vein.\(^3\)

Clinical manifestation

The most common clinical manifestations of VGAM are high-output cardiac failure and neurological abnormalities.

High-output cardiac failure results from high volume flow through low resistance arteriovenous shunts, which in turn causes volume overload on the right atrium, increased pulmonary blood flow, and pulmonary hypertension. Symptoms can vary from tachycardia and poor feeding, to cardiogenic shock and respiratory distress syndrome. The severity depends on the degree of shunting and the age presentation, with neonates more severely affected than those who present in infancy or early childhood.\(^4\)

VGAM can cause multiple secondary neurological abnormalities. The venous aneurysm can cause obstructive hydrocephalus by compression of the aqueduct of Sylvius or third ventricle. Increasing head circumference is a common presenting complaint for infants with the malformation. High volume flow through the aneurysm may divert blood flow away from brain parenchyma, causing ischemia. Reduced venous drainage may cause cerebral oedema and venous hypertension. Reduced venous drainage with resultant impaired cerebrospinal fluid resorbtion may also lead to hydrocephalus.\(^4\)

Background to endovascular treatment
Before the development of endovascular treatment techniques, prognosis for VGAM case was extremely poor, with reported mortality rates of 100% for neonates.\textsuperscript{5}

The development of endovascular techniques began in the early 1980s and has significantly improved clinical outcomes. A review of the 337 reported cases of VGAM treated with endovascular embolization between 2001 and 2010 reported a "good" outcome (normal development or mild developmental delay) in 60.8\% of patients; a "fair" (moderate or severe developmental delay) in 23.4\%; and death in 15.7\%.\textsuperscript{4}
Imaging findings OR Procedure details

Imaging features of vein of Galen malformations

The imaging features of VGAM are relatively straightforward. Cranial Doppler ultrasound is a safe and easy screening test, which will clearly demonstrate the malformation and provide information on the size of the ventricles (Fig. 1 on page 12).

MR confirms the diagnosis and provides further information about the effect of the lesion on the brain parenchyma and ventricular system. In addition, MR angiography and venography often provides useful information on the specific vascular anatomy of the VGAM which can aid treatment planning (Fig. 2 on page 12).

Digital subtraction angiography (DSA) is not required for diagnosis, and is only used in cases when endovascular treatment is to be performed. It provides detailed anatomical information to allow planning for treatment. Post treatment DSA allows assessment of the degree of occlusion of the malformation (Fig. 3 on page 12).

Some cases are diagnosed incidentally on prenatal anatomical survey in the second trimester (Fig. 4 on page 13).

Local endovascular treatment technique

Endovascular embolisation of VGAM is reserved for patients who have congestive heart failure (CHF) refractory to medical treatment or active hydrocephalus.

We operate a predominantly adult neurovascular service within an adult tertiary referral hospital. Paediatric patients are transferred intubated from specialist paediatric hospitals, accompanied by paediatric anaesthetists, and return immediately after treatment.

In the 5 cases treated in our institution we have used only a transarterial approach, without prior venous occlusion. In all cases, access was established via the common femoral artery. A 4 or 5 Fr guide catheter was passed to the internal carotid or vertebral artery. A flow directed catheter (Magic 1.8, Balt, Montmorency, France) was used to access arterial feeders to the VGAM, in most cases without use of a wire. Acrylic glue (Glubran 2, GEM, Viareggio, Italy) at a concentration of 80% was used to embolise the feeding arteries.
The duration of the procedure is limited by patient tolerance of iodinated contrast. No more than 5 ml per kg body weight of contrast is used in a single procedure.

The goal of treatment to improve the clinical symptoms. If this is not achieved after the first treatment, it may be repeated until improvement is attained. Complete obliteration of the malformation is not necessarily essential for symptom improvement. We have seen spontaneous resolution of the VGAM following partial embolisation in some of our cases.

**Case series**

Over the last 7 years, 5 patients with VGAM have been treated with endovascular embolisation at our institution. No patient referred during that time was refused treatment. The features of each case are summarised in Table 1 on page 14, and described in more detail below.

**Case 1**

This female neonate was immediately tachypnoeic after birth by caesarean section. Her chest radiograph demonstrated cardiomegaly and echocardiogram showed severe congestive heart failure (CHF). A VGAM was diagnosed by cranial ultrasound and MR on day 1.

Her CHF deteriorated despite maximal medical treatment, and she received endovascular treatment on day 5. Three injections were performed: two into left thalamoperforators and one into a branch of the left anterior cerebral artery (ACA). There was marked reduction in flow but the aneurysm was not completely occluded. Her CHF began to improve immediately after treatment. Cranial ultrasound the following day showed reduction in flow though the VGAM.

Her cardiac function continued to improve and she was discharged home 8 days after endovascular treatment. She has remained well, displayed normal head growth, and developed normally. Follow up MR studies have shown decreased in size and eventual spontaneous obliteration of the VGAM.
Fig. 5: Case 1: (a) T2 weighted MR sequence shows VGAM. Lateral projection of left internal carotid artery angiogram before (b) and after (c) endovascular embolisation: flow through the malformation is slowed and flow is diverted to normal arterial branches after treatment. Follow up T2 weighted MR demonstrates reduction in size of VGAM at 10 weeks (d) and complete obliteration at 6 years (e).

References: Radiology, Beaumont Hospital - Dublin/IE

Case 2

This male neonate was born prematurely at 35 weeks gestation. He developed CHF within a few hours of birth. A VGAM had been diagnosed antenatally by on routine Doppler ultrasound. Decision for endovascular embolisation was made after failure of maximal medical treatment for CHF. Endovascular treatment was performed on day 9. Six separate glue injections were performed. There was a large decrease in flow through malformation on post-precedure angiogram.

His cardiac function improved, but he developed active hydrocephalus. A ventriculo-peritoneal shunt was placed at 6 weeks. At age 2 he developed seizures, which are well controlled with anti-epileptic medication. He is otherwise developing normally. Follow up cranial ultrasound shows no residual flow through VGAM.
Fig. 6: Case 2: (a) Frontal projection of left internal carotid digital subtraction angiogram showing vein of Galen malformation with supply from the anterior circulation. (b) Mask from post embolisation DSA run, frontal projection, showing radiopaque glue cast in the midline.

References: Radiology, Beaumont Hospital - Dublin/IE

Case 3

This male infant was referred by the public health nurse with irritability and inability to lift his head. At 16 weeks of age his head circumference was 50.5 cm (above the 99th centile). MR demonstrated a large VGAM causing obstructive hydrocephalus. His cardiac function was normal at echocardiography.

On the basis of active hydrocephalus with deteriorating neurological function, the decision was made to treat with endovascular embolisation. He received 2 treatments at 16 weeks of age: 3 glue injections were performed on the first treatment (2 into right ACA branches and 1 into right MCA branch) and one on the second treatment (into right MCA branch). The VGAM was almost completely occluded after the second treatment. He was discharged home well.

He re-presented 9 days after treatment with drowsiness and poor feeding. MR brain demonstrated thrombosis of straight sinus with left thalamic haemorrhage. His ventricle size had increased and a ventriculoperitoneal shunt was placed.
He now has a residual partial right hemiparesis which is resolving. Otherwise neurological development and head growth are satisfactory. Follow up MR studies have shown decrease and eventual obliteration of the VGAM, and changes secondary to thalamic infarct.

Fig. 7: Case 3: Frontal projection of right internal carotid artery angiogram before (a) and after (b) endovascular embolisation, and lateral projection or right internal carotid artery before (c) and after (d). There is minimal flow to the malformation post-treatment. T2 weighted MR 9 days after treatment (e) demonstrates partial occlusion of VGAM (containing thrombus) and signal abnormality centred on left thalamus consistent with haemorrhage. Follow up MR at 8 months (f) showing decrease in size of VGAM, and at 14 months (g) showing obliteration of malformation, with encephalomalacia and volume loss in the left hemisphere consistent with old thalamic haemorrhage.

References: Radiology, Beaumont Hospital - Dublin/IE

Case 4

This male neonate was born at 39 weeks via elective cesaerean section. He was born with severe CHF, which by day 6 could no longer be managed by maximal medical therapy. Cranial ultrasound and MRI demonstrated a large VGAM.

Pre-treatment angiography showed a large choroidal-type VGAM with multiple tortuous feeders from both internal carotid arteries and the posterior circulation. Over the course of three treatments, 4 glue embolisations in posterior choroidal arteries and 5 embolisations in right ACA branches were preformed. In spite of the high flow through the VGAM there was difficulty negotiating the tortuous feeding vessels, and a wire was employed within the flow directed catheter during the final treatment. This probably contributed to the
eventual rupture of the catheter on glue injection. Pulling the catheter resulted in trauma to vessel and intracranial haemorrhage. The procedure was therefore abandoned.

Despite embolisation of multiple feeders to the VGAM, the patient's cardiac function continued to deteriorate. He developed severe pulmonary hypertension despite full medical treatment. The decision was made to withdraw intensive care unit support, and the patient died.

Fig. 8: Case 4: Chest radiograph (a) demonstrates cardiomegaly. Colour Doppler cranial ultrasound (b) and time-of-flight MR angiogram demonstrate large VGAM with multiple tortuous feeding vessels. Angiography of right internal carotid artery (lateral projection) (d) and left vertebral artery (lateral projection) (e) demonstrating choroidal type VGAM with multiple abnormal feeding vessels from anterior and posterior circulation. Non-contrast CT (f) showing intraventricular haemorrhage complicating final treatment.

References: Radiology, Beaumont Hospital - Dublin/IE

Case 5

This male infant was a twin, born via elective caesarean section at 36 weeks. VGAM was diagnosed antenatally. He developed CHF immediately after birth. Cranial ultrasound
and MRI on day 1 showed a large VGAM with feeders from both anterior and posterior circulation. He improved with diuretics and was discharged home day 22.

Follow up ultrasound and MR at 4 months showed increasing size of VGAM and developing hydrocephalus. For this reason, the decision was made to proceed with endovascular embolisation. He received a single treatment: two glue injections via right posterior communicating (PCOM) artery, and three via the left PCOM artery. There was an overall large reduction in flow through the aneurysm. The procedure was uncomplicated.

A follow up MRI at 7 months showed significant decrease in size of VGAM, and decrease in ventricular size. He continues to do well: cardiac function, neurological development, and head growth are all normal.

**Fig. 9:** Case 5: (a) T2 weighted MR at 4 months demonstrating large VGAM. Left internal carotid angiogram before (b) and after (c) embolisation. Right internal carotid angiogram before (c) and after (d) transarterial embolisation, showing decrease in flow through the malformation. (e) Post-treatment T2 weighted MR showing significant reduction in the size of the VGAM.

**References:** Radiology, Beaumont Hospital - Dublin/IE
Fig. 1: Cranial ultrasound performed on 4 month old patient. There is a well circumscribed anechoic lesion in the midline (a), which demonstrates turbulent flow on colour Doppler imaging. VGAM is the only diagnosis which consistent with these findings.

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Fig. 2: MRI: (a) Maximum intensity projection (MIP) view of time-of-flight (TOF) MR angiogram showing multiple hypertrophied arteries feeding VGAM. (b) MIP view of TOF venogram showing VGAM draining into dilated transverse sinus, sigmoid sinus, and internal jugular veins. (c) T2 weighted axial MR showing large midline flow void characteristic of VGAM.

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**Fig. 3:** DSA: (a) Lateral and (b) frontal views of right internal carotid angiogram demonstrating mural type VGAM with individually identifiable hypertrophied feeding arteries. (c) Lateral and (d) frontal view of left vertebral artery angiogram showing choroidal type VGAM with multiple tortuous feeders draining into a nidus. Note the choroidal type is a much more complex vascular abnormality.

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**Fig. 4:** Antenatal imaging: (a) Antenatal colour Doppler ultrasound performed at 33 weeks demonstrating turbulent flow within an intracranial midline lesion, suggestive of VGAM. (b) Sagittal T2 weighted image from antenatal MRI performed at 34 weeks gestation demonstrates midline flow void, confirming the diagnosis of VGAM.

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<table>
<thead>
<tr>
<th>Case</th>
<th>Gender</th>
<th>Age at diagnosis</th>
<th>Clinical problem prompting treatment</th>
<th>Age at treatment</th>
<th>Treatment number (injections per treatment)</th>
<th>Immediate angiographic outcome</th>
<th>Clinical outcome</th>
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<tbody>
<tr>
<td>Case 1</td>
<td>F</td>
<td>1 day</td>
<td>Congestive heart failure</td>
<td>4 days</td>
<td>1 treatment (3 injections)</td>
<td>Partial occlusion</td>
<td>Well; normal development. Now 6 years old</td>
</tr>
<tr>
<td>Case 2</td>
<td>M</td>
<td>Antenatal diagnosis</td>
<td>Congestive heart failure</td>
<td>11 days</td>
<td>1 treatment (6 injections)</td>
<td>Partial occlusion</td>
<td>Partial seizures began at age 2. Well controlled on anti-epileptics. Normal development. Now 5 years old</td>
</tr>
<tr>
<td>Case 3</td>
<td>M</td>
<td>16 weeks</td>
<td>Hydrocephalus</td>
<td>16 weeks</td>
<td>2 treatments (3+1 injections)</td>
<td>Partial occlusion</td>
<td>Venous thrombosis and thalamic haemorrhage 9 days post treatment. Resolving right hemiparesis. Now 2.5 years old</td>
</tr>
<tr>
<td>Case 4</td>
<td>M</td>
<td>1 day</td>
<td>Congestive heart failure</td>
<td>6 days</td>
<td>3 treatments (4+5+1 injections)</td>
<td>Partial occlusion</td>
<td>Intraventricular haemorrhage during last treatment. CHF worsened despite treatment. Died on day 13</td>
</tr>
<tr>
<td>Case 5</td>
<td>M</td>
<td>Antenatal diagnosis</td>
<td>Hydrocephalus</td>
<td>14 weeks</td>
<td>1 treatment (5 injections)</td>
<td>Partial occlusion</td>
<td>Well; normal development. Now 1 year old</td>
</tr>
</tbody>
</table>

**Table 1:** Table summarising the cases of the 5 patients with VGAM treated with endovascular embolisation at our institution.

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Conclusion

- VGAMs do not pose a diagnostic challenge on cranial ultrasound or MRI. Some cases are diagnosed antenatally.

- Angiography is reserved for cases in which endovascular treatment is planned.

- The indication for treatment in neonates or infants is CHF not responding to medical treatment, or deteriorating neurological function with hydrocephalus.

- Treatment technique in our institution involves trans-arterial glue embolisation. These are high-risk procedures requiring an experienced neurovascular team. The vessels injected are fragile and minimal catheter and wire manipulations are required.

- The goal of treatment is partial occlusion of the malformation and symptom relief, rather than complete occlusion.

- The development of endovascular embolisation has significantly improved the prognosis for children with VGAM. In our series, 4 out of 5 patients treated had good clinical outcomes.
Personal Information

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