

# Large vein of Galen malformation associated with an apical ventricular septal defect and a patent ductus arteriosus in an infant - a case report

**P Szkup**  
MD

**AM Cilliers**  
MBBCh, FCPaed (SA)

Divisions of Radiology and Paediatric  
Cardiology, Baragwanath Hospital,  
Johannesburg.

## Abstract

A four month old male infant who had a transarterial embolization of a large vein of Galen malformation attempted is reported. The infant presented in cardiac failure and a hydrocephalus. In addition the rare association of an apical ventricular septal defect and a patent ductus arteriosus with pulmonary hypertension was diagnosed. A staged transarterial coil embolisation using

microcoils was commenced. The first attempt was uneventful. This was followed by the unfortunate demise of the patient from an intercurrent nosocomial pneumonia.

## Introduction

Vein of Galen malformations (VGM) are rare vascular anomalies in which a primary feature is the dilatation of the vein of Galen which becomes dilated as a result of direct cerebral arterial connections bypassing the capillary network.<sup>1</sup> The cardiac output and venous return to the heart increases resulting in progressive heart failure and pulmonary hypertension. Hydrocephalus may occur as a result of direct obstruction of the Sylvian aqueduct by the malformation or a resorptive block of cerebrospinal fluid due to increased pressure within the sagittal sinus.<sup>2</sup> Cranial bruits are audible in only a third of cases.<sup>1</sup> Fewer than 300 cases of this anomaly have been reported between 1937 and 1994.<sup>2</sup>

## Case report

A male infant presented to our institution at the age of four months with a history of cough and shortness of breath for eight days duration. Clinical examination revealed a wasted infant in cardiac failure. In addition the patient had a large head (beyond the 97th centile for age) and a loud cranial bruit. The blood pressure was 70/50. Biventricular hypertrophy was present clinically and a 3/6 pansystolic

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murmur over the apex was audible. The liver measured 4 cm below the right costal margin and the pulmonary component of the 2nd sound was loud indicating the presence of pulmonary hypertension. The chest radiograph showed a cardiothoracic ratio of 70% with biventricular enlargement and plethoric lung fields. Right ventricular hypertrophy was the main feature electrographically. Two dimensional echocardiography showed a 3 mm apical ventricular septal defect (VSD), a 3 mm patent ductus arteriosus (PDA) and severe pulmonary hypertension with an estimated pulmonary artery pressure of 70 mmHg. Cardiac catheterization confirmed balanced pulmonary and systemic pressures with poor response of the pulmonary vasculature to the administration of oxygen.

Cranial ultrasonography (Figure 1) and Doppler flow studies revealed



Figure 1: Sagittal view of the cranial ultrasound showing the vein of Galen malformation and a dilated straight sinus.

a VGM measuring 2.5 x 2.8 cm draining into a dilated straight sinus. Dilatation of the lateral and third ventricles was evident. Pre- and post-contrast axial CT (Figure 2) confirmed a VGM and mild hydrocephalus.

Cerebral angiography (Figures 3 and 4) was performed via the transfemoral arterial route. There were major feeder arteries arising

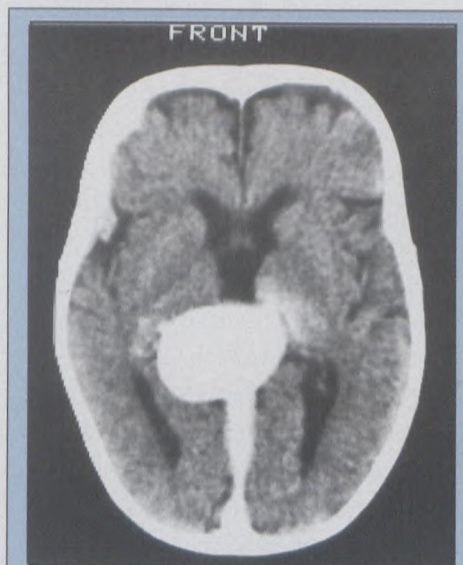


Figure 2: Contrast enhanced axial computerised tomography scan of the head showing the large vein of Galen malformation, dilated straight sinus and mild hydrocephalus.

from both the posterior cerebral arteries, both left and right middle cerebral arteries and the right anterior cerebral artery. Other lesser arterial feeder arteries were apparent.

Despite medical treatment in the form of oxygen, digoxin and diuretics the patient remained in cardiac failure and failed to gain weight over the next month. In view of the major contribution of the cerebral pathology to the patient's morbidity it was decided to

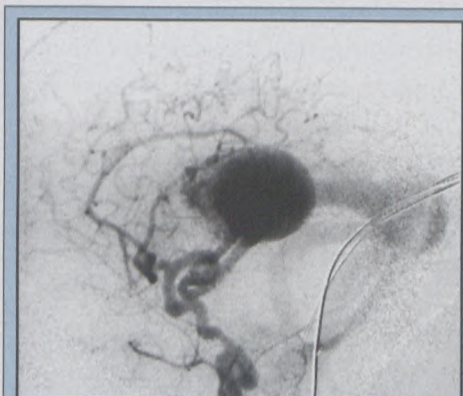


Figure 3: Lateral view of the cerebral angiogram showing multiple feeder vessels to the vein of Galen malformation arising from the right middle cerebral artery and the anterior cerebral artery.

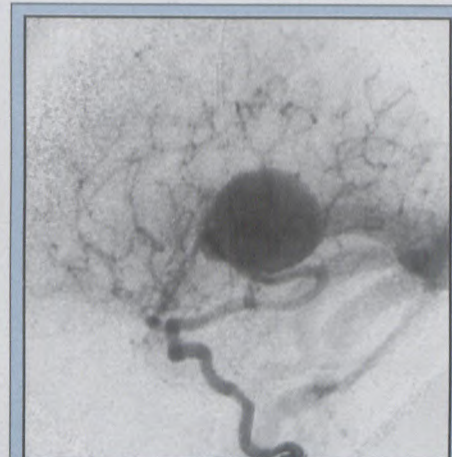


Figure 4: Lateral view of the cerebral angiogram showing multiple feeder vessels to the vein of Galen malformation arising from the left middle cerebral artery.

perform a staged transarterial embolization of the VGM.

The first stage was performed under general anaesthesia and involved selective catheterization of a feeder artery to the malformation originating from the left middle cerebral artery. Access to the feeder artery was accomplished using a FasTracker 18MX microcatheter, together with Dasher 14 and Mach 16FIX Nitinol guidewires. The feeder artery was embolised (Figure 5) with the placement of ten microcoils. The patient tolerated this first procedure well and suffered no complications. Unfortunately before the second staged procedure could be attempted the patient demised from a nosocomially acquired pneumonia in the intensive care unit. Permission for a post mortem examination was not granted.

## Discussion

Congestive cardiac failure unresponsive to medical management in the infant is the primary indication for treatment.<sup>2,3</sup> The severe cardiac failure and failure to thrive in our patient were the major motivational factors in attempting to treat the VGM.



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Figure 5: Lateral view of the left internal carotid artery angiogram following the embolisation procedure showing microcoils occluding the arterial feeder vessels to the vein of Galen malformation.

The cardiac lesions were deemed to be of minor significance not requiring treatment at the time. An endovascular approach was undertaken because of the lower anticipated mortality rate of 13%<sup>4</sup>, compared to a surgical approach.<sup>2,4,5</sup>

A staged transarterial occlusion was initiated to gradually reduce the flow through the fistula to control the cardiac failure. A partial occlusion frequently reverses the cardiovascular symptoms so that a complete occlusion may not be required.<sup>2</sup>

A further rationale for a staged endovascular occlusion is to avoid "normal perfusion pressure breakthrough" which may occur with either a transvenous<sup>2</sup> or transarterial approach in a large VGM i.e. the previously hypoperfused peripheral brain after embolisation of a large VGM

may develop a sudden increase in blood flow associated with regional brain swelling and haemorrhage.<sup>3</sup> Severe neurological damage may result.

Associated structural cardiac defects are rarely reported. PDA,<sup>6,7,8</sup> transposition of the great vessels,<sup>8</sup> a VSD, and coarctation of the aorta<sup>9</sup> have been described. Coarctation of the aorta may have a physiological explanation in that the developing foetus with an intracranial fistula has reduction of blood flow down the descending aorta which induces development of hypoplasia of the isthmus of the aorta.<sup>10</sup>

## Conclusion

Management of vein of Galen aneurysmal malformations is a rare and challenging therapeutic problem for the neuroradiological interventionalist. While there is a higher risk of morbidity and mortality associated with a direct surgical approach, an endovascular approach has potential complications. Cardiac lesions are rare associations and if significant may contribute substantially to the therapeutic dilemma.

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