Aneurysm of the Vein of Galen in Neonates: Report of Four Cases

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In neonates, aneurysm of the vein of Galen often masquerades as cyanotic congenital heart disease. We report 4 cases of neonates presenting with malformation of the vein of Galen at our institution. An increased awareness of this entity seems warranted. (Indian Heart J 2001; 53: 499-502)

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Heart failure and cyanosis in infants almost always results from congenital heart disease. Systemic arteriovenous malformations (AVMs) are a rare cause of such a presentation and the diagnosis is often missed. We report 4 cases of AVMs related to malformation of the vein of Galen who presented to us in the past five years. The purpose of this report is to highlight the clinical presentation and increase awareness of this entity. The clinical diagnosis was missed in the first case but correctly made in the 3 subsequent cases.

Case Reports

Case 1: A full-term male child presented at 5 days of age with severe congestive heart failure (CHF) and mild cyanosis. The peripheral pulses were bounding and there was no radiofemoral delay. The first heart sound was normal while the second had a loud pulmonic component. There was a grade 3/6 systolic murmur heard best in the pulmonary area. The electrocardiogram (ECG) showed evidence of biventricular hypertrophy while the chest X-ray showed cardiomegaly. Echocardiography did not reveal any structural anomaly except that the pulmonary venous drainage was not clearly visualized. On cardiac catheterization, the aorti root angiogram showed markedly enlarged and tortuous neck vessels, suggesting an intracardiac AVM. The diagnosis was confirmed on imaging the cranium (Fig. 1). On detailed clinical examination, a grade 2/6 systolic bruit that had been missed in the initial clinical evaluation was heard over the cranium.

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Fig. 1. Digital subtraction angiogram showing vein of Galen malformation (arrows), dilated carotid artery and jugular vein (arrowheads). AO denotes aortic arch.

Case 2: A full-term male child presented at 2 days of age with severe CHF and mild cyanosis. His peripheral pulses were bounding in all four extremities. The first heart sound was normal while the second had an accentuated pulmonic component. There was a grade 3/6 systolic murmur heard best in the pulmonary area. Due to our experience with the previous patient who had a similar presentation, a cranial bruit was sought and found. The ECG showed evidence of biventricular hypertrophy while the chest X-ray showed cardiomegaly with increased pulmonary vascularity.
Echocardiography revealed dilated right heart chambers, a patent foramen ovale, mild tricuspid regurgitation and an enlarged superior vena cava (SVC) and arch of the aorta (Fig. 2). Pulse Doppler study of the SVC showed increased forward flow with normal flow in the inferior vena cava (IVC). Doppler study of the ascending aorta and carotid arteries showed continuous forward flow (Fig. 3) while that of the descending aorta suggested diastolic run-off. Real-time ultrasound examination of the cranium suggested malformation of the vein of Galen and a contrast-enhanced computed tomographic (CT) scan of the head revealed an aneurysm of the vein of Galen.

**Case 3:** A 6-day-old male child had a history of CHF and mild cyanosis from the first day of life. His clinical findings were similar to those of the previous patients. The ECG was normal for his age while the chest X-ray showed cardiomegaly with increased pulmonary blood flow. Echocardiography, transcranial ultrasound and contrast-enhanced CT scan of the brain identified a malformation of the vein of Galen.

All three neonates succumbed to their illness within a week of being diagnosed.

**Case 4:** A 5-day-old child presented with findings similar to those of the 3 previous patients. Computed tomographic scan of the head showed a large malformation of the vein of Galen (Fig. 4). The parents were keen on aggressive treatment despite the poor prognosis. The patient was taken up for embolization of the feeding vessels using N-butyl cyanoacrylate. There was a decrease in the size of the aneurysm after embolization (Figs 5a and b) along with a marked improvement in CHF, and a significant reduction in cardiac size was seen in the chest X-ray (Figs 6a and b). However, the child succumbed to his illness the next day.

**Discussion**

Arteriovenous malformations are an uncommon cause of CHF in the neonatal period. Intracerebral AVMs being the most common amongst them. Although malformations of the vein of Galen constitute only 1% of all cerebral vascular
Diagnostic confusion in the neonate arises from the combination of severe CHF and cyanosis, pointing towards a cardiac disorder. Cyanosis results from the torrential venous return crossing the patent foramen ovale and a diagnosis of persistent fetal circulation may be considered by the unwary. Detailed clinical examination, however, reveals evidence of high-output heart failure and a cranial bruit (up to 80% of the cardiac output can be directed towards the cerebrovascular bed due to low resistance within the vascular malformation). Brisk upper limb pulses in the presence of severe heart failure should suggest the possibility of an AVM. Lower limb pulses may be relatively feeble due to steal by the cerebral fistula. In the extremely sick child, however, all pulses may be feeble. Although a benign systolic bruit is described in up to 15% of normal neonates and children, the presence of a systolic bruit over the cranium in a child with CHF should strongly suggest this disorder. All 4 of our patients had a systolic bruit over the cranium.

Two-dimensional echocardiography usually reveals normal cardiac anatomy with dilated right heart chambers and an enlarged SVC and arch vessels. Uncommonly, structural heart disease has been reported in these infants including coarctation of the aorta, atrial septal defect, ventricular septal defect and partial anomalous pulmonary venous drainage. Pulse Doppler echocardiography of the arch vessels demonstrates a continuous forward flow of high velocity which reflects the low peripheral resistance bed introduced by the AVM. High-velocity forward flow in the SVC with normal flow in the IVC are also seen. These features, in the absence of aortic regurgitation and a patent ductus arteriosus, suggest the extracardiac nature of the lesion and its intracerebral location. Transcranial ultrasonography is extremely useful in demonstrating the AVM while Doppler studies aid in demonstrating the flow in it. A semi-quantitative measurement of flow in the feeding and draining vessels can also be obtained. Color Doppler echocardiography helps to demonstrate the turbulent flow within the aneurysm. In addition, contrast venous echocardiography has been shown to be of use as the rapid return of microbubbles into the SVC is a pathognomonic sign of an intracerebral AVM in the absence of another peripheral left-to-right shunt in the upper part of the body. The contrast can be injected into a peripheral leg vein (which then crosses over from the right to the left across a patent foramen ovale) or directly into the aorta through an umbilical artery catheter.

Although CT scan of the brain demonstrates the AVM and visualizes the feeding vessels, the details thus obtained may not be sufficient to guide surgical treatment.
Computed tomography also depicts associated brain lesions such as calcification and anoxic brain damage. Magnetic resonance imaging (MRI) is superior to CT in its ability to demonstrate vascular anatomy, though it is unlikely to supplant angiography in the immediate future since sufficient details of arterial and venous anatomy which permit precise endovascular/surgical therapeutic decisions are usually not obtained. Prenatal diagnosis of this disorder is also possible and one can precisely delineate the vascular malformation.

Embolization of the feeding arteries of the AVM (in a single or staged sitting) is the preferred therapeutic modality for a patient in severe CHF. Transarterial embolization is the preferred route although transvenous and transcardiac routes have also been used. Solid materials, including microcoils (mostly fibered or unfibered platinum coils), microballoons, and silk sutures have been used to embolize these vessels, with variable success. Liquid adhesives that have been used for embolization include cyanoacrylate monomers such as 1-butyl cyanoacrylate and N-butyl cyanoacrylate, and polymers such as ethylenevinyl alcohol copolymer. Even partial reduction in cerebral flow after embolization is sufficient for controlling CHF in the neonate as it permits retrograde thrombosis of the AVM. The mortality rate in neonates remains extremely high (up to 55%) even after embolization, partly due to the large extent of the malformation. Some recent reports have, however, indicated better immediate neonatal outcome. Embolization promptly alleviates the massive intracranial arteriovenous shunting and improves CHF. In infants in whom endovascular therapy is delayed, clinical results may be poor despite obliteration of the aneurysmal malformation. This is believed to be the result of an acquired occlusive venopathy affecting the dural venous sinuses due to high flow. Gamma knife surgery is not a viable option in the size of the AVM. Hence, it is not useful for treating the neonate in intractable CHF but is more appropriate for the clinically stable infant or for managing a residual AVM after endovascular therapy. Surgical clipping of these aneurysms as the primary procedure is no longer done due to the high procedural mortality (up to 90% in neonates and 55% across all age groups) and postoperative morbidity. However, surgical clipping may be performed if CHF persists after endovascular therapy. In conclusion, aneurysm of the vein of Galen, a rare cause of cyanosis and heart failure in infants, can be diagnosed clinically in the appropriate setting. The extensive distribution of the aneurysm usually precludes surgical management/endovascular therapy.

References