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Vein of Galen Malformation and High-output Cardiac Failure

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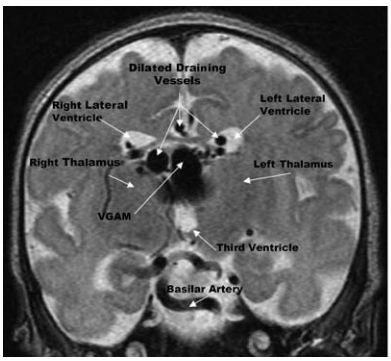

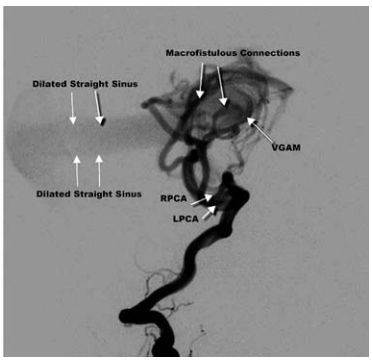

A NEWBORN male born at 40 weeks presented with acute tachypnea with feeding. Marked cardiomegaly was noted on chest radiography, and echocardiogram revealed dilated cardiac chambers and systolic failure.

Coronal brain magnetic resonance image consistent with Vein of Galen aneurysmal malformation (VGAM) demonstrates an enlarged midline vascular flow void in the pineal region associated with numerous ectatic vessels in the lateral ventricles and roof of the third ventricle, without evidence of parenchymal hemorrhage, hydrocephalus, or cerebral infarction (fig.). A sagittal left vertebral artery angiogram in the early arterial phase shows rapid arteriovenous shunting from choroidal branches of the right and left posterior cerebral arteries (RPCA and LPCA, respectively) to VGAM. This flow is unloaded into a massively enlarged straight sinus through macrofistulous connections between the choroidal arteries and the wall of the VGAM.

Vein of Galen aneurysmal malformations are rare congenital anomalies (less than 1/25,000 deliveries) of intracranial circulation that constitute 1% of all intracranial vascular malformations.[1] These malformations are made from multiple arterial feeders establishing direct or indirect shunts with a large median venous collector. The origin of VGAM is from a persistent embryonic median vein of prosencephalon. *In utero*, cardiac failure secondary to VGAM is rare because the placental circulation provides a low-resistance path preventing damage from fluid overload. With loss of the placenta at birth, up to 70% of cardiac output is directed to VGAM's low-resistance arteriovenous shunt that allows direct return of large flow volume to the right heart.[1] In one series, 46% of VGAM patients presented with high-output cardiac failure.[2] Cardiac failure occurs as the large flow volume of the VGAM shunt is unloaded into the right atrium and pulmonary circulation, with pulmonary vasoconstriction and pulmonary hypertension resulting in right ventricle failure. Early identification and staged embolization of both the feeding arteries and draining veins can result in reduction of blood flow and a greater survival rate in these patients.[3] VGAM patients may require anesthesia for preoperative investigation (computed tomography or angiography), endovascular embolization, or surgical resection of the anomaly. Anesthetic management of these patients poses multiple challenges to the anesthesiologist in view of the complex pathophysiology of VGAM and must be tailored to address heart failure and pulmonary hypertension.[4]

Competing Interests

The authors declare no competing interests.

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