

# Repair of Partial Anomalous Pulmonary Venous Connection and Sinus Venosus Defect in a Child with Vein of Galen Malformation

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## Abstract:

**Background:** Sinus venosus defect (SVD) is a deficiency in the sinus venosus portion of the atrium near the superior or inferior venae cavae, with the former being more common. It is often associated with partial anomalous pulmonary venous connection (PAPVC) of one or more of the right pulmonary veins. It is one of the most commonly associated congenital heart defects in patients with Vein of Galen malformation (VGAM), a rare cerebral arteriovenous malformation of the midline venous structure known as the vein of Galen. SVD provides a unique challenge in treating patients with transarterial embolization for the VGAM due to risk of systemic embolization through the intracardiac shunt.

**Case Description:** We report the case of a child who underwent percutaneous device closure of a SVD and PFO at another institution prior to VGAM embolization in an attempt to subvert this issue without the risks associated with cardiac bypass in the setting of a high output cerebral shunt. The patient later had successful transarterial embolization of the VGAM. He was evaluated at our institution 9 years after his device implantation due to progressive right atrial and ventricular enlargement. We surgically removed the septal occluder devices and re-rerouted the anomalous pulmonary venous return to the left atrium via Warden procedure.

**Conclusion:** This case illustrates the difficulty with management of these patients and the need for an individualized approach to staged repair of combined cerebral and cardiac defects. Unfortunately current percutaneous devices do not provide a stable option for occlusion of SVD and would not be recommended for these patients.

## Figures

Figure 1: (a) View through the right atrium of occluder device located in the sinus venosus defect. (b) Explanted PFO and sinus venosus occluder devices demonstrating in vivo configuration.

Figure 1a

Occluder device  
in sinus venosus  
defect

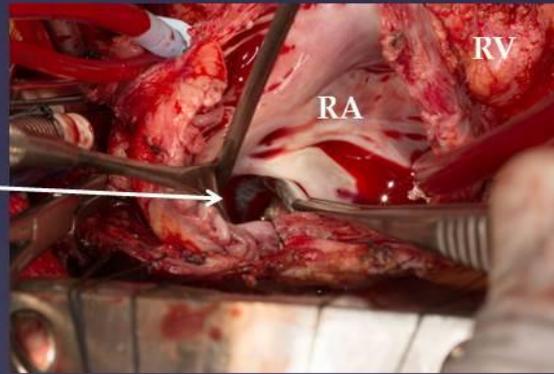


Figure 1b

PFO occluder  
device  
Sinus venosus occluder  
device



