

Superior Sinus venosus atrial septal defect (SVASD) and PAPVD: Post-operative Complications in Warden procedure

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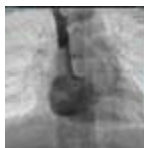
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Background. The Warden procedure is a surgical correction for superior sinus venosus atrial septal defect (SVASD) associated with partial anomalous pulmonary venous drainage (PAPVD). It restores normal pulmonary venous return by redirecting anomalous veins into the left atrium (LA). The technique involves transecting the superior vena cava (SVC) above the entry of the anomalous veins, anastomosing the cephalic SVC to the right atrial appendage (RAA), and using the caudal SVC to construct a baffle (often with a patch) to direct pulmonary venous flow into the LA. The atrial septal defect is usually closed during the same procedure. While generally successful, a potential complication is stenosis at the SVC-RAA anastomosis, which requires close monitoring.

Clinical Case. We report the case of A., a female child born at term by cesarean section, with normal early growth. At 2.5 years, a heart murmur led to cardiologic evaluation, revealing a large SVASD with PAPVD of the right upper pulmonary vein to the SVC, and moderate right-sided volume overload. She was then monitored regularly. At 4 years, a cardiac MRI better defined the anatomy, and corrective surgery using the Warden procedure was performed at a specialized cardiac surgery center, with uneventful recovery. Subsequent echocardiographic follow-up showed a gradual increase in the trans-anastomotic gradient, suggesting progressive stenosis at the cephalic SVC-RAA junction, while flow through the pulmonary venous baffle remained laminar. This condition remained clinically stable until November 2024, when for the first time, a superficial venous network with a mantle-like distribution appeared on clinical examination. At that time, echocardiography showed a significantly increased gradient at the anastomosis level (mean 13 mmHg vs. 4 mmHg the previous year; peak 22 mmHg vs. 11 mmHg), confirming hemodynamically relevant stenosis. After multidisciplinary evaluation, the patient was referred for cardiac catheterization. Under general anesthesia and via percutaneous access through the right jugular and femoral veins, angiography with invasive pressure measurements confirmed severe stenosis at the SVC-RAA anastomosis. The stent segment was successfully crossed with a guidewire, which was retrieved through the jugular sheath to create a venovenous loop for stability. After appropriate sizing, percutaneous transluminal balloon dilation was performed, followed by the placement of a covered CP stent to minimize the risk of restenosis due to surrounding atrial muscle tissue.



Pre-cath



Post-cath

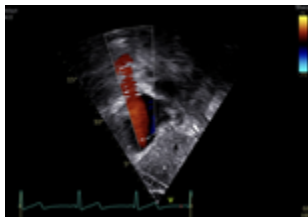


Figure 3. Echocardiography post-cath

The procedure was completed without complications and resulted in resolution of the obstruction. Three months later, echocardiography (figure 3) follow-up showed laminar flow through the stent and complete resolution of the superficial venous network, confirming clinical and hemodynamic improvement.

Discussion and Conclusions. The appearance of the venous mantle suggests that, over time, the azygos system alone was insufficient to manage upper body venous return, prompting the formation of collateral pathways. Reestablishing unobstructed flow via stenting was essential to restore normal hemodynamics. Stenosis at the SVC-RAA anastomosis is a recognized late complication of the Warden procedure, occurring in 5-15% of cases. In this patient, the progression of the stenosis was initially slow but steady, as evidenced by a gradual increase in Doppler-derived pressure gradients. Starting from November 2024, however, a sudden worsening was observed, marked by a sharp rise in the trans-anastomotic gradient and the clinical appearance of superficial venous collateralization. This finding prompted an urgent re-evaluation and the subsequent interventional treatment. This case underscores the critical importance of ongoing, long-term surveillance in patients undergoing corrective surgery for congenital heart defects. Continuous follow-up allows for the timely identification and management of both early and late complications, ensuring optimal long-term outcomes.