




Hypoxia following warden procedure: evaluation and percutaneous treatment

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Original Article

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Abstract

Partial anomalous venous connection with sinus venosus atrial septal defect is repaired with different approaches including the Warden procedure. Complications include stenosis of the superior caval vein and pulmonary venous baffle; however, cyanosis is rarely seen post-operatively. We report a patient presenting with cyanosis 5 years after a Warden, which was treated with a transcatheter approach.

The Warden procedure is one type of surgical repair for partial anomalous venous connection with sinus venosus atrial septal defect. Complications include stenosis of the superior caval vein and pulmonary venous baffle; however, cyanosis has rarely been reported. We report a patient who presented with cyanosis following a Warden procedure, which was treated with a transcatheter approach.

Case presentation

A 10-year-old female patient, weighing 21 kg, who underwent a Warden procedure 5 years prior, presented to the emergency room with new onset chest pain for 1 day, along with headaches and decreased exercise tolerance for 1 year. The chest pain was felt to be musculoskeletal; however, saturations were 85–90% in room air. A transthoracic echocardiogram showed severe stenosis of the superior caval vein (SVC). An agitated saline contrast study was performed with a left upper extremity injection, which demonstrated opacification of the left heart indicating a right to left shunt (Fig. 1a). A computerized tomography scan (CTA) revealed contrast filling of a dilated hemiazygos vein from a left upper extremity injection, delayed opacification of the left innominate vein and SVC, and a dilated azygos vein draining into the left atrium (Fig. 1b).

She was referred to the cardiac catheterization lab for potential intervention. Haemodynamic assessment demonstrated a Qp:Qs of 0.8 and indexed pulmonary vascular resistance of 1.6. There was severe stenosis at the anastomosis of the SVC to the right atrial appendage with a gradient of 12 mm Hg. Contrast injection through the sheath in the left internal jugular vein (LIJV) confirmed a dilated hemi-azygos with dilated connection to the azygos vein, which had been incorporated into the pulmonary venous baffle (Fig. 2a). A 6-Fr Mullins Transseptal Sheath (Cook, Inc., Bloomington, IN) was advanced over a Super Stiff guidewire (Boston Scientific, Marlboro, MA) from the left internal jugular vein, into the hemiazygos vein, and then across to the azygos vein. A 14-mm Amplatzer Vascular Plug II was advanced through the sheath and deployed in the azygos vein to occlude it just proximal to where the right upper pulmonary vein (RUPV) returned. There was a rapid improvement in oxygen saturation from 90% to 98–100%. Angiography performed in the azygos vein proximal to the device post-occlusion demonstrated that the device was well positioned with no residual flow through the device and pulmonary artery wedge angiography showed no obstruction of pulmonary venous return to the left atrium (Fig. 2b).

Balloon angioplasty of the SVC was performed, and post-intervention angiography revealed significant improvement in the calibre with 1 mm Hg residual mean gradient from the superior caval vein to the right atrium. We opted not to place a stent at this juncture given this would obligate her to subsequent procedures to further dilate the stent for growth, although recognizing there is a chance of restenosis which may require stent implantation at a later date. She was discharged home the following morning with saturations of 98%. At 18 months post-procedure, the patient is doing well with mild residual SVC stenosis (mean gradient by Doppler echo 6 mm Hg), normal saturations, improved activity tolerance, and resolution of her headaches.

Discussion

Surgical repair of partial anomalous venous connection is the definitive treatment and is specific to the individual anatomy of the anomalous connection. There are several surgical approaches including the Warden procedure. Short- and long-term complications include stenosis of

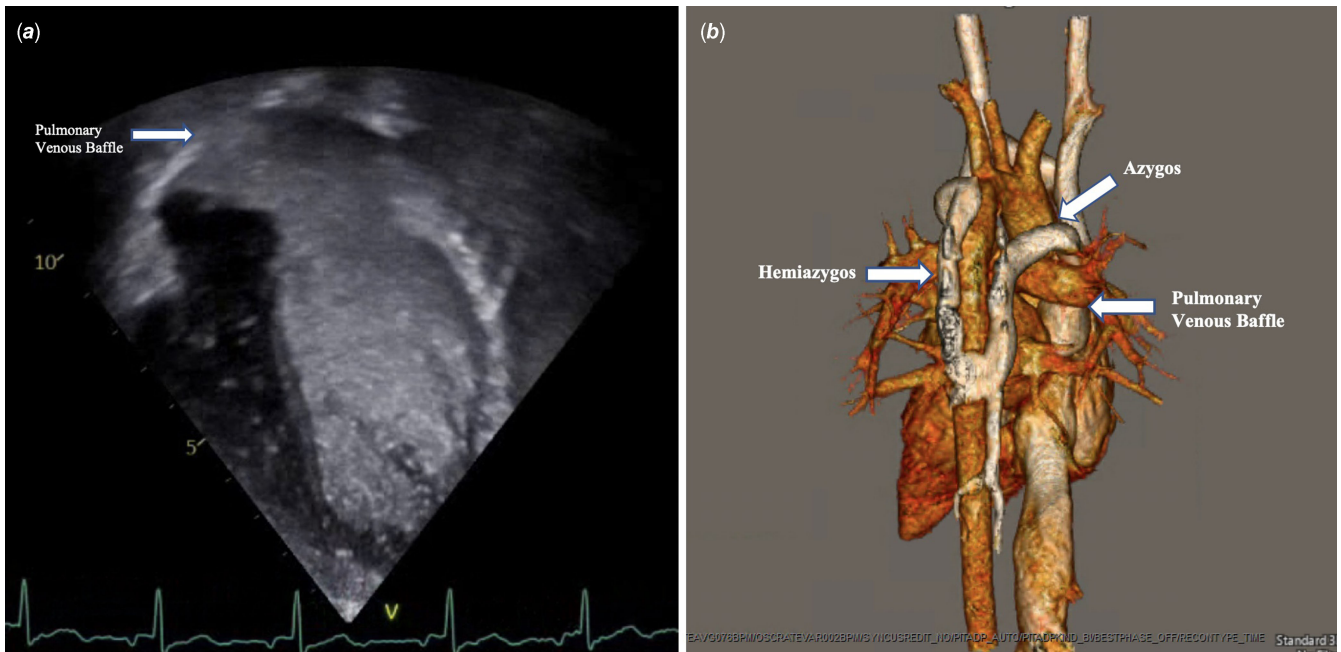


Figure 1. *a*) Apical four-chamber view during agitated saline injection demonstrating opacification of the pulmonary venous baffle, left atrium, and left ventricle *b*) CTA 3D reconstruction demonstrating hemiazygos vein connecting to dilated azygos vein which is incorporation into pulmonary venous baffle.

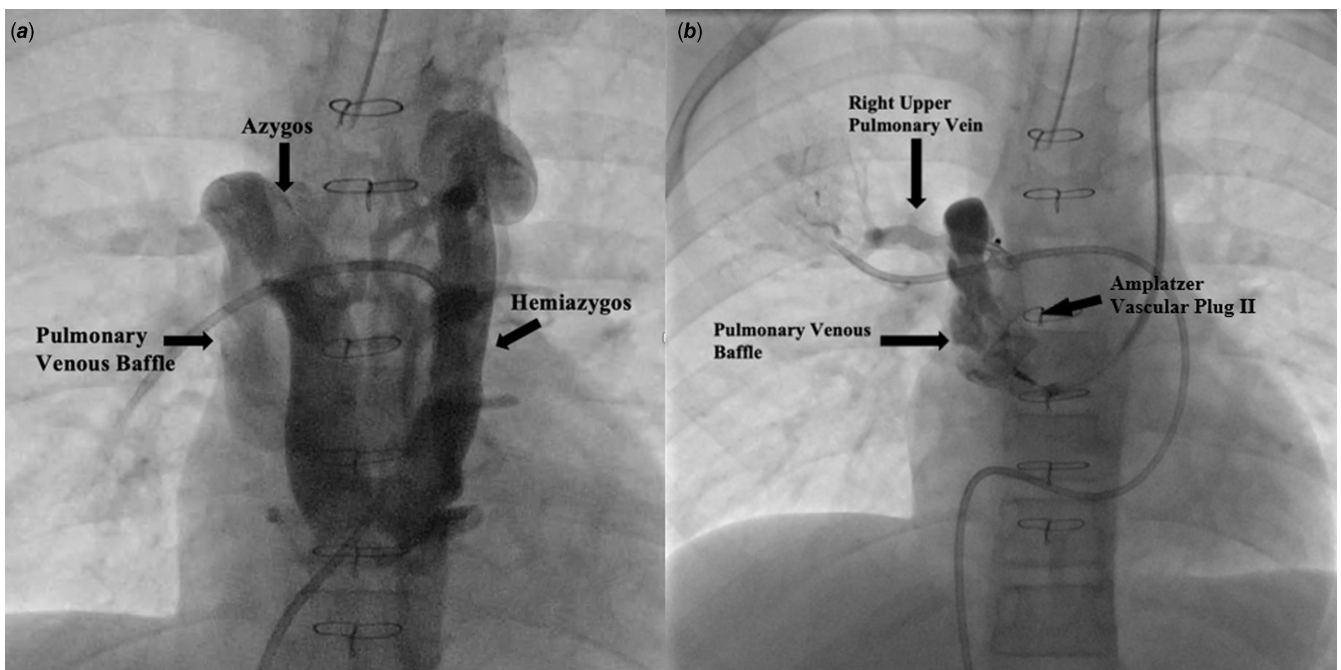


Figure 2. *a*) Anterior posterior (AP) projection of initial angiography through LIJV sheath showing opacification of hemiazygos vein with connections to azygos vein, which ascends along right side of spine, and enters pulmonary venous baffle to the left atrium, and *b*) right upper pulmonary artery wedge angiogram post-device occlusion of the azygos vein demonstrating RUPV returns unobstructed into pulmonary venous baffle.

superior caval vein or pulmonary venous baffle and arrhythmia.¹ The incidence of re-intervention following a Warden procedure has been reported as high as 17%; however, Binsalamah et al found that in their cohort, re-intervention was seen only in patients who underwent repair before 4.9 years of age.² Cyanosis is a rare complication following a Warden procedure. Stenosis of the SVC alone would not cause hypoxia. One case of cyanosis following a Warden procedure has been described in a patient who had

re-opening of a left superior caval vein to the left atrium in the presence of severe right-sided SVC stenosis. The patient underwent treatment of the stenosis of the right SVC, and transcatheter device occlusion of persistent left SVC was performed.³

In our case, the azygos venous system was not identified intra-operatively and likely thought to be an additional right pulmonary vein. It was therefore incorporated into the pulmonary venous baffle. Severe superior caval vein stenosis caused preferential filling

of the azygos system, which drained into the left atrium, causing a significant right to left shunt and hypoxia. We felt treating the caval vein stenosis and addressing the azygos vein flow into the pulmonary venous baffle during the same procedure was warranted. Treatment of the superior caval vein stenosis alone might still allow some degree of right to left shunt through the azygos system with potential for continued exercise intolerance and cyanosis with activities. By closing the azygos vein closer to the pulmonary venous baffle, the hemiazygos vein could still serve as a source of venous collateral blood flow through its normal connection with the azygous vein inferior to the vascular plug without resultant cyanosis in the event of significant recurrent superior caval vein stenosis. This approach led to a favourable outcome with normalization of her saturations and resolution of symptoms. Our patient had signs of SVC stenosis prior to presentation including headaches and fatigue for 1 year. Thus, evaluation for clinical signs of caval vein obstruction including fatigue, activity intolerance, headaches, facial swelling, and engorgement of superficial veins in the upper chest, neck, and extremities is important in all patients following a Warden procedure.

Conclusion

We describe a rare case of cyanosis presenting 5 years after a Warden procedure which was treated percutaneously with

balloon angioplasty of the superior caval vein and device occlusion of the azygous vein which had been incorporated into the pulmonary venous baffle. Awareness of this case should alert physicians in a patient who presents with cyanosis following a Warden procedure to evaluate for possible superior caval vein stenosis and to utilize imaging to assess the systemic and pulmonary venous drainage.

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Competing interests. None.

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