Warden Procedure in a 77-year-old Man

Running Head: Warden Procedure

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Abstract

Partial anomalous pulmonary venous return (PAPVR) is a rare congenital heart defect characterized by one or more but not all of the pulmonary veins draining somewhere other than the left atrium thereby creating a left-to-right shunt. Over time, patients may develop right-sided volume overload and its subsequent complications. We present a case of isolated PAPVR in an older patient who underwent a Warden procedure at age 77 years with rapid improvement in right ventricular size and function.

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Partial anomalous pulmonary venous return (PAPVR) is a rare congenital heart defect (estimated prevalence of 0.4-0.7% in autopsy studies) (1) that is usually associated with other heart defects, most commonly an atrial septal defect (2, 3). In a series of 306 patients with PAPVR, the most common lesion was a right-sided anomalous vein connecting to the superior vena cava (SVC), which occurred in 74% (2).

Patients with PAPVR can present at any time from infancy through adulthood. If symptomatic, patients may experience signs and symptoms of right-sided volume overload, including dyspnea on exertion, palpitations due to atrial arrhythmias, and pulmonary hypertension (4). While there are clear guidelines for surgical correction in most circumstances of PAPVR, they are not always clear in the case of isolated PAPVR (4). Here we present a case of isolated PAPVR that was uncorrected until age 77 years, at which time the patient had developed clinically significant left-to-right shunt with right sided dilation. He underwent a Warden procedure with a good clinical outcome.

The patient was a 77-year-old man who volunteered to undergo an echocardiogram as a 27-year-old medical intern that revealed a suspected atrial septal defect (ASD). This was followed by a right-sided heart catheterization that revealed suspected PAPVR at the level of the high superior vena cava (SVC). At that time, there was an oxygen saturation step-up from 75% in the SVC to 81% in the right atrium (RA) with a calculated Qp:Qs ratio of 1.3:1. No further evaluation or treatment was recommended.

In his early 30s, the patient was diagnosed with hypertension requiring medical therapy. He continued to engage in intense workouts, including 6-10 mile runs. In his 60s, the patient transitioned to indoor activities due to knee issues and worked with a trainer for 90 minutes at a time. He developed some palpitations that were shown to be premature atrial complexes (PACs) on Holter monitor. Although his exercise tolerance was starting to decrease, he attributed this to normal aging and continued exercising without limitations.

At 76 years, the patient noted increased palpitations during exercise. Further testing revealed frequent PACs. Transthoracic echocardiogram demonstrated right atrial and right ventricular (RV) dilatation as shown in Figure 1. On computed tomography scan as seen in Figure 2, the dilated right-sided pulmonary vein drained into the high SVC and measured 1.4 cm in diameter. Cardiopulmonary exercise stress testing revealed maximal oxygen uptake (VO_2 maximum) of 77% predicted. Catheterization revealed that his shunt had increased to a Qp:Qs of 2.1. Coronary arteries were normal. Mean pulmonary artery was 24 mmHg. An elevated left ventricular end-diastolic pressure (LVEDP) of 18 mmHg indicated diastolic dysfunction and an elevated right ventricular end-diastolic pressure (RVEDP) of 9 mmHg indicated only mild right-sided diastolic dysfunction. Mean pulmonary wedge pressure was 15 mmHg.

The patient was subsequently referred for surgical correction, including the Warden procedure in which the proximal SVC was connected to the right atrial appendage. Because of the high location of the anomalous veins, the proximal SVC was divided above the entrance of the anomalous veins and elongated with a 16-mm ringed Gore-Tex graft (Figure 3). The distal SVC containing only the anomalous veins transported the pulmonary venous blood to the RA. A large atrial septal defect was created and a 3 x 6 cm long Gore-Tex patch was used to cover the SVC orifice and baffle the anomalous pulmonary veins to the left atrium across the atrial septectomy. The patient recovered uneventfully and was discharged from the hospital on post-operative day five.

The patient's echocardiograms demonstrated improvements in a variety of RA and RV parameters. In comparison of preoperative echocardiogram to that on postoperative day 17, there was increased RV fractional area change from 25.5 % to 32.8 %, increased tricuspid annular planar excursion from 1.37 cm to 1.79 cm, decreased RV diameter from 5.53 cm to 4.54 cm, decreased RA area from 32.84 cm^2 to 19.54 cm^2, and increased velocity time integral of the TV annulus from 1.44 cm. to 1.61 cm. Four months after surgery, repeat exercise stress testing

was essentially unchanged with exception of fewer ectopic beats. The patient has returned to exercising with a trainer for 90 minute increments without limitation.

Comment

Isolated PAPVR is a rare cause of right sided dilatation and pulmonary hypertension. The decision to operate and its timing can be complicated. In a review of 43 adults aged 20-73-years-old with isolated PAPVR, Majdalany et al. concluded that patients with only one anomalous vein did well with monitoring alone while those with two or more anomalous veins tended to develop RV enlargement as in our patient (4). The patient presented here had known PAPVR with an initially insignificant shunt in his third decade of life; however, over the next 50 years the Qp:Qs increased from 1.3:1 to 2.1:1. Worsening left ventricular diastolic dysfunction with pulmonary venous hypertension was out of proportion to the right-sided dysfunction likely causing increased left to right shunting. The significant shunt was a clear surgical indication to prevent further deterioration according to the 2018 ACC/AHA Adult Congenital Heart Disease Guidelines (5). The patient's overall health and score of "very fit" on the Clinical Frailty Scale (6) made him an excellent surgical candidate.

Of the 28 patients in Majdalany's study who underwent surgical repair for isolated PAPVR, there was no mortality associated with the repair and most had significant physiological improvement (4). A review by Alsoufi et al, of 306 pediatric patients who underwent surgical repair of PAPVR (aged 5 months to 18 years), 175 had right-sided PAPVR into the SVC, similar to our patient. There were no deaths and minimal morbidity in 15 years of follow-up (2). These data were presented to the patient who opted to proceed with the Warden procedure given his good quality of life and likelihood of worsening without PAPVR correction. Similar to younger patients, he demonstrated rapid post-operative improvement in echocardiographic RV parameters and improved ectopy on stress testing, suggesting decreased right heart strain.

In conclusion, when appropriately selected, the Warden procedure can benefit patients with PAPVR, even at advanced ages.

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Figure Legends

Figure 1: Echocardiographic images prior to warden procedure of dilated right atrium and ventricle in systole (A) and diastole (B).

Figure 2: Computed tomography images of dilated right-sided anomalous vein draining into the high superior vena cava in coronal view (A) and sagittal view (B).

Figure 3: An intra-operative photograph of the ringed Gore-Tex graft connecting the superior vena cava with the right atrial appendage.