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Aortopulmonary Window in Adult Patient Operated on With Echocardiography Assessment

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Adult aortopulmonary window is a rare presentation of a rare disease; only a few cases are reported to have undergone successful surgical closure without development of Eisenmenger syndrome. We describe the second oldest patient, a 25-year-old woman, who underwent successful surgical repair of aortopulmonary window after favorable indirect measures on echocardiography without the "gold standard" preoperative cardiac catheterization study. At 2 months after the operation, the patient remains in New York Heart Association class II.

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Aortopulmonary window (APW), a rare congenital heart disease with occurrence of 0.2% to 0.6%, is managed by either surgical or interventional device closure on diagnosis.[1] Untreated large APW has a fatal outcome even during infancy, with mortality as high as 40%.[2] Because of an earlier tendency for development of Eisenmenger syndrome in patients with APW, when it is manifested in the adult, cardiac catheterization to study pulmonary vascular resistance and its response to vasodilators is the "gold standard" for assessment of operability. There are few case reports in the English literature of adult patients with APW (from 18 years up to 31 years) who successfully underwent surgery after right-sided heart catheterization favored intervention.[3] We describe the second oldest adult patient who underwent surgical repair of APW after echocardiographic diagnosis and assessment without right-sided heart catheterization for pulmonary vascular resistance to determine operability.

A 25-year-old woman (39 kg, 161 cm, and body surface area of 1.36 m^2) presented with worsening dyspnea and fatigue. Despite that the patient was diagnosed with APW at the age of 9 years, she was lost to follow-up and was subjected to delayed presentation while waiting for free cardiac surgery service by an international humanitarian charity. On physical examination, oxygen saturation was 94% on room air; on auscultation, there was a continuous murmur at the left lower sternal border.

On the chest radiograph, the cardiothoracic ratio was 67%, with a prominent main pulmonary artery and hypervascularized bilateral lung field. Left ventricular hypertrophy was demonstrated on the electrocardiogram. The preoperative hemoglobin level was 12.4 mg/dL. All other blood test results were unremarkable.

During transthoracic echocardiography assessment, the defect of the APW was 19 mm wide and close to the right pulmonary artery takeoff, making it type II APW (Figure 1). The shunt was bidirectional, predominantly left to right (Video), with flow reversal in the aortic arch and descending aorta. There was no tricuspid or pulmonary valve regurgitation, which made it difficult to measure the pulmonary artery hypertension. The left-sided chambers were more dilated than the right ones. The left atrium measured 38 mm and the right atrium 32 mm in diameter; the basal right ventricle measured 23 mm, and the basal left ventricle measured 53 mm. There was no other associated congenital abnormality like interrupted arch, bicuspid aortic valve, or any other intracardiac defect on echocardiography assessment. Despite that right-sided heart catheterization services are not readily available in our setting and we could not assess for

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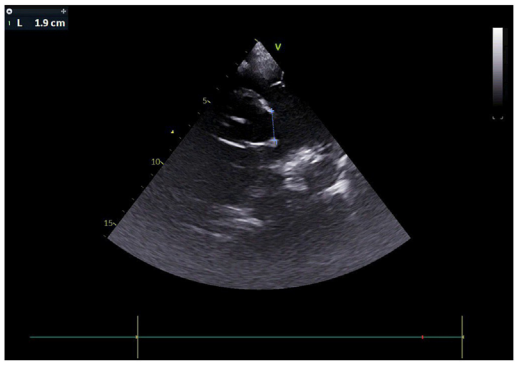

FIGURE 1 Preoperative transthoracic echocardiography parasternal short-axis view showing a large 19-mm type II aortopulmonary window.

pulmonary vascular resistance, we decided to repair the defect not to delay further in the face of favorable findings on echocardiography.

Midline sternotomy was performed and a bicaval cannulation established; inferior and superior venae cavae were snared, and right and left pulmonary arteries were mobilized and snared before the cross-clamp was applied. During cardioplegia delivery, the right atrium was opened parallel to the atrioventricular groove, and the left ventricle was vented by creating a fossa in the interatrial septum. After the heart was arrested, a direct incision over the aortopulmonary communication was made, and a glutaraldehyde-treated autologous pericardial patch was wedged in the defect to start closing it from the base to the common wall in both directions with 5-0 Prolene. The patch was sandwiched between the incised anterior walls of the defect to finish closing the APW. The right atrium was closed after removal of air, leaving an estimated 4-mm interatrial communication open. Milrinone was used for weaning and continued for 3 days after the operation. The patient was extubated 6 hours after the operation, and oxygen saturation is maintained above 90% with 4 L/min of intranasal oxygen.

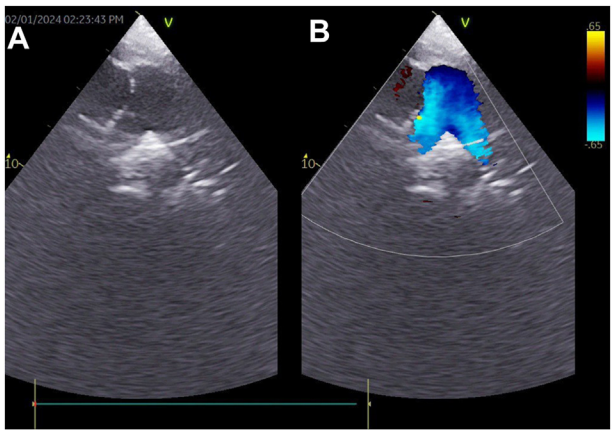

FIGURE 2 Postoperative transthoracic echocardiography parasternal short-axis view of the patient (A) without color flow and (B) with color flow showing intact patch with no residual flow across the aortopulmonary junction.

Postoperative echocardiography showed an intact patch with no residual shunt in the aortopulmonary junction (Figure 2). There was mild tricuspid regurgitation with measured pulmonary arterial hypertension of 34 mm Hg and 16-mm circumferential pericardial effusion. The foramen ovale created was shunting right to left and reduced biventricular function. The postoperative course was uneventful. The patient was discharged from the hospital on postoperative day 7 with sildenafil, furosemide, enalapril, and colchicine.

Two months after the operation, there was no pericardial effusion, and the patient reported improvement of fatigability and dyspnea. Oxygen saturation was 92% on room air, and she was at New York Heart Association functional class II.

COMMENT

A 25-year-old woman underwent successful surgical repair of APW by sandwich technique with glutaraldehyde-treated autologous pericardium without preoperative cardiac catheterization study. Right-sided heart catheterization to study pulmonary vascular resistance is not readily available in our setting, and this forced us to resort to indirect signs of pulmonary vascular resistance by echocardiographic assessment. According to the British Society of Echocardiography guideline protocol, findings of a predominant left-to-right shunt at the APW along with flow reversal at the aortic arch as well as descending aorta, absent tricuspid or pulmonary valve regurgitation, and dilated left atrium and ventricle were used as an indirect indicator of reversible pulmonary vascular resistance.[4] The absence of other associated congenital anomalies despite the large defect might be the reason for favorable echocardiographic findings and the subsequent decision for surgical repair in this patient.

Our patient is the second oldest to undergo successful surgical closure of adult APW. From the few case reports, a patient aged 31 years was the oldest to undergo successful surgical repair.[5,6]

However, it is only 2 months since our patient's index surgery, and it is difficult to compare the results with the other case reports with longer follow-up periods of 13 months to 8 years.[3]

With emerging local cardiac surgery programs elsewhere, we expect more adult patients with the same problem to present where cardiac catheterization is not readily available. Therefore, this approach for operability might be an opportunity not to delay further for adult patients with favorable echocardiography parameters.

In conclusion, we describe the second oldest patient, a 25-year-old woman, who underwent successful surgical repair of a 19-mm type II APW without preoperative cardiac catheterization assessment after favorable indirect measures on echocardiography. Because the outcome of this particular case might have been due to the individual patient's anatomy and physiology, absent associated anomalies, and case selection, we do not advise the routine use of echocardiography without right-sided heart catheterization.

The Video can be viewed in the online version of this article [https://doi.org/10.1016/j.atssr.2024.04.023] on http://www.annalsthoracicsurgery.org.

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DISCLOSURES

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PATIENT CONSENT

Obtained.

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