Case Report

Aortopulmonary Window Associated with an Ascending Aorta Aneurysm in an Adult

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Abstract

Aortopulmonary window (APW) is a rare congenital malformation. It results from an incomplete division between the ascending aorta and the pulmonary artery. We describe a 26-year-old male, who presented with a grade II exertional dyspnea and palpitations. Echocardiography revealed an APW with an ascending aorta aneurysm. He underwent surgery under cardiopulmonary bypass without aortic cross-clamping. The APW was closed via the pulmonary artery flap technique using an autologous pericardial patch, and the aneurysm was repaired through the reduction aortoplasty technique. The patient was discharged on the 4th postoperative day. At 2 years' follow-up, he had remained asymptomatic and echocardiography showed aortic valve competence, ascending aorta diameter of 38 mm, and no residual shunt.

Keywords: Aortopulmonary septal defect • Aneurysm • Aorta • Heart defects, congenital

Introduction

The aortopulmonary window (APW) is a rare congenital malformation and represents only 0.1% of all congenital heart diseases.1 It results from an incomplete division between the ascending aorta and the pulmonary artery. The natural history of this anomaly is characterized by the early manifestations of congestive heart failure, gradual development of irreversible pulmonary hypertension occurring early in life, and death if not treated.2,3,4 Surgical correction is, therefore, advised as early as possible. Although there are various reports of an uncorrected APW in adulthood,2,4 there are very few reports of operated cases. We describe a 26-year-old male patient suffering from an APW associated with an ascending aorta aneurysm who underwent successful surgery and discuss the clinical and surgical features of this condition.

Case Report

A 26-year-old man was admitted to our institution for the assessment of a grade II exertional dyspnea and palpitations. The patient was born from a first-degree consanguineous marriage. He had no history of recurrent respiratory infections. On physical examination, he had normal vital signs: blood pressure at 140/50 mmHg with a bounding pulse and saturation at 96% on room air. He had pectus carinatum (Figure 1). Auscultation found a 4/6 left laterosternal systolic murmur. There were no signs of heart failure. Electrocardiography showed regular sinus rhythm at 80 beats/minute with left heart axis deviation and negative anteroseptal T waves. Chest X-ray demonstrated marked pulmonary vascularity and cardiomegaly. Echocardiography revealed a dilated left ventricle with a good ejection fraction.
The ascending aorta was enlarged above the sinotubular junction measuring 48 mm, with trivial aortic regurgitation. There was no abnormality in the coronary arteries. A 12-mm APW was identified with a left-to-right shunt. The surgical closure of the APW in conjunction with reduction aortoplasty was indicated.

The patient underwent surgery through median sternotomy. The dilatation of the ascending aorta and the APW were plainly evident. Surgery was conducted under normothermic cardiopulmonary bypass between bicaval cannulas and an aortic cannula without aortic-cross clamping. After a lateral clamping of the ascending aorta excluding the APW, without compromising the aortic flow, the pulmonary artery was discharged and then incised a few millimeters to the left of the APW to create a flap (Figure 2). This flap was thereafter used to reconstruct the ascending aorta with a continuous 5-0 polypropylene suture. The main pulmonary artery defect was closed using glutaraldehyde-treated autologous pericardium. The aortic dilatation was repaired after repositioning of the clamp to the anterior side of the aorta. A longitudinal oval resection of the aortic wall was subsequently performed. The aortotomy was closed in two rows using polypropylene 4-0 suture (Figure 3).

The patient’s postoperative course was uneventful, and he was discharged on the fourth postoperative day. At 2 years’ follow-up, he had remained asymptomatic and echocardiography showed aortic valve competence, ascending aorta diameter at 38 mm, and no residual shunt.

**Discussion**

The APW is a rare congenital heart anomaly and appears in approximately 0.1% of all congenital heart diseases. In this anomaly, a communication exists between the ascending aorta and the pulmonary trunk. The APW may be located anywhere from above the semilunar valves to the more distal ascending aorta. Despite the rarity of the APW, several classifications have been proposed. The latest and the most commonly used one was described by Jacobs et al., who classified the condition as proximal, distal, total, and intermediate, depending on the site in relation to the pulmonary artery. Most commonly, the APW is an isolated single defect. Anomalies such as interrupted aortic arch, coartation, transposition of the great arteries, and tetralogy of Fallot have been reported in association with the APW. Our patient had an intermediate type of the APW; and to our knowledge, it is the first reported case of the APW with an ascending aorta aneurysm.

The development of the aneurysm can be explained in part by the turbulent flow in the ascending aorta; this turbulence is a result of several mechanisms, including the wide pulse pressure (it reached 90 mmHg in our patient), the directional...
Cardiac catheterization is... residual shunt, and aortic valve distortion. Therefore, long-term follow-up is mandatory."

"The surgical treatment of the APW represents a continuous evolution of technique. Since the first successful ligation was performed by Gross in 1948, several surgical techniques have been proposed from simple ligation to closure of the defect with a pulmonary artery flap. The closure of the APW can be performed off-pump, avoiding the morbidity associated with cardiopulmonary bypass, but with greater risk of leaving a residual shunt. Transaortic repair with patch closure is the most widely used technique."

"Surgical repair is indicated immediately after the diagnosis of the APW is established, regardless of the patient’s age. Although early surgical repair is recommended, some patients may survive until adulthood without developing irreversible pulmonary vascular hypertension. These patients could benefit from surgery with good outcomes."

"In the current era, early mortality following the repair of a simple APW approaches zero and the long-term outcome should be excellent. Early morbidity includes pulmonary artery stenosis, residual shunt, and aortic valve distortion. Therefore, long-term follow-up is mandatory."

### References

1. Jacobs JP, Quintessenza JA, Gaynor JW, Burke RP, Mavroudis C. Congenital Heart Surgery Nomenclature and Database Project: aortopulmonary window. Ann Thorac Surg 2000;69(4 Suppl):S44-49.
2. Alkumaim M, Al-Fayez M, Munibari A-N. The surgical treatment of aortopulmonary window in Yemeni adolescents and adults. Heart Mirror J 2010;4:217-219.
3. Backer CL, Mavroudis C. Surgical management of aortopulmonary window: a 40-year experience. Eur J Cardiothorac Surg 2002;21:773-779.
4. Aggarwal SK, Mishra J, Sai V, Iyer VR, Panicker BK. Aortopulmonary window in adults: diagnosis and treatment of late-presenting patients. Congenit Heart Dis 2008;3:341-346.
5. Kervancioglu S, Soydinc S, Davutoglu V, Kervancioglu R, Sirikci A, Bayram M. Aortopulmonary window: a rare adult case demonstration by echocardiography, MRI, CE-MRA and angiography. Cardiovasc Intervent Radiol 2004;27:175-178.
6. Chen CA, Chiu SN, Wu ET, Lin MT, Wang JK, Chang CI, Chiu IS, Wu MH. Surgical outcome of aortopulmonary window repair in early infancy. J Formos Med Assoc 2006;105:813-820.
7. Barnes ME, Mitchell ME, Tweddell JS. Aortopulmonary window. Semin Thorac Cardiovasc Surg Pediatr Card Surg Ann 2011;14:67-74.
8. Moruno Tirado A, Santos De Soto J, Grueso Montero J, Gavilán Camacho JL, Álvarez Madrid A, Gil Fournier M, Descalzo Señorans A. Aortopulmonary window: clinical assessment and surgical results. Rev Esp Cardiol 2002;55:266-270.
9. Wixon CL. Vascular hemodynamics. In: Hallet JW, Mills JL, Earnshaw JJ, eds. Comprehensive Vascular and Endovascular Surgery. 2nd ed. Philadelphia: Mosby Elsevier; 2009. p. 39-52.
10. Tekbucuha T, von Segesser LK, Vogt PR, Bauersfeld U, Jenni R, Künzli A, Lachat M, Turina M. Congenital aortopulmonary window: a 40-year experience. Eur J Cardiothorac Surg 1997;11:293-297.
11. Gross RE. Surgical closure of an aortic septal defect. Circulation 1952;5:858-863.
12. Van Son JA, Hambsch J, Mohr FW. Anatomical reconstruction of aorta and pulmonary trunk in patients with an aortopulmonary window. Ann Thorac Surg 2000;70:674-675.