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Aortopulmonary Window with Subaortic Fibrous Stenosis and Septal Defect: Surgery through a Minimal Right Vertical Infra-Axillary Thoracotomy

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ABSTRACT

Aortopulmonary window with subaortic stenosis and ventricular septal defect is an uncommon congenital cardiac malformation that is repaired using cardiopulmonary bypass. The authors describe a 3-year-old patient on whom we performed surgery through a minimal right vertical infra-axillary thoracotomy. This minimally invasive surgery is likely to be applicable in a few cases.

INTRODUCTION

Aortopulmonary window (APW) accounts for 0.1% to 0.2% of all congenital heart disease. Other heart defects are found in half of all cases, and in 25% of cases these are complex, such as interrupted aortic arch, anomalous origin of the coronary arteries, transposition of the great arteries, and tetralogy of Fallot [Santos 2008]. APW with subaortic stenosis and ventricular septal defect is a rare case. In our department, minimal right vertical infra-axillary thoracotomy has been used for repair of atrial septal defect, ventricular septal defect, and mitral valve replacement and has proved to be a safe and cosmetic alternative to median sternotomy [Yang 2001; Yang 2003; Wang 2009; Wang 2010]. With the accumulated surgical experience, we did the surgery using the minimal right vertical infra-axillary thoracotomy. We did not find any similar cases in the literature, so we report the surgical treatment of this rare abnormality.

CASE REPORT

A 3-year-old female was admitted with exertional dyspnea. Past medical history was remarkable for recurrent pulmonary infection. The blood pressure was measured at 90/60 mmHg on the right arm and 95/55 mmHg on the left arm; blood pressure was 110/60 mmHg on the right and 100/69 mmHg on the left lower extremities. On physical examination, all peripheral arteries were palpable. Cardiac auscultation revealed a 2 over 6 soft systolic ejection murmur heard over the mesocardiac area. Oxygen saturation in the upper and lower extremities was 98%.

Chest X-ray showed increased cardiothoracic index and pulmonary vascular markings. Transthoracic echocardiography (TTE) demonstrated concentric hypertrophy of the left ventricle with an ejection fraction of 63%. A peak Doppler pressure gradient, which was caused by the subaortic stenosis, was 79 mmHg. Short-axis view at the level of great vessels revealed a large defect between the ascending aorta and the main pulmonary artery, termed as an intermediate defect. The multiple views showed subaortic stenosis and the ventricular septal defect.

The operation was performed through a minimal right vertical infra-axillary thoracotomy. The thoracic cavity was entered through the fourth intercostal space. The lung was retracted posteriorly using wet sponges to expose the pericardium. The pericardium was opened 2 cm anterior to the phrenic nerve, superiorly to the pericardial reflection and inferiorly to the diaphragm, to provide enough exposure of the ascending aorta and inferior vena cava. The superior pericardial stay stitches were placed on partial pleura of ribs to elevate the aorta into the operative field. The cannulation of the inferior vena cava, superior vena cava, and ascending aorta were performed as described previously [Yang 2001; Yang 2003]. A distal APW located in the middle of the ascending aorta (Figure 1), termed as a type IV defect [Backer 2002]. Through an aortic incision, the subaortic stenosis and the APW could get a good exposure (Figures 2 and 3). The APW was closed with a Dacron patch (Figure 4) with running polypropylene suture. The subaortic stenosis was a fibrous ridge and was removed from the endocardium directly. Simultaneously, the ventricular septal defect was closed through the right atrium incision [Wang 2010]. The aortic clamp time was 56 minutes.

The patient was discharged 7 days after surgery. The 6-month follow-up echocardiography showed complete occlusion of the defect. There was no evidence of the subaortic stenosis, and chest X-ray revealed a decrease in cardiac size with an absence of pulmonary plethora.

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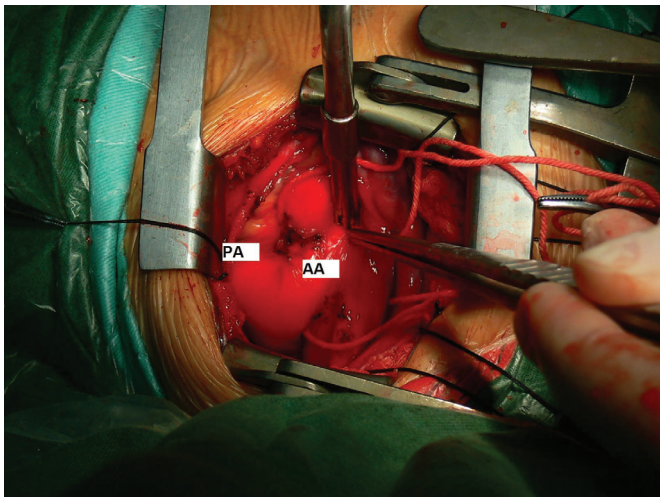

Figure 1. External view of the ascending aorta (AA) and the pulmonary artery (PA) through the minimal right vertical infra-axillary thoracotomy.

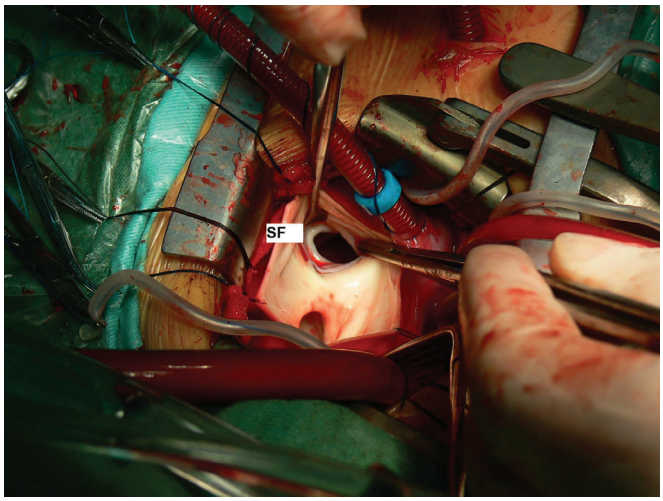

Figure 3. View through the aortic incision showing the subaortic fibrous membrane (SF), which caused the subaortic stenosis.

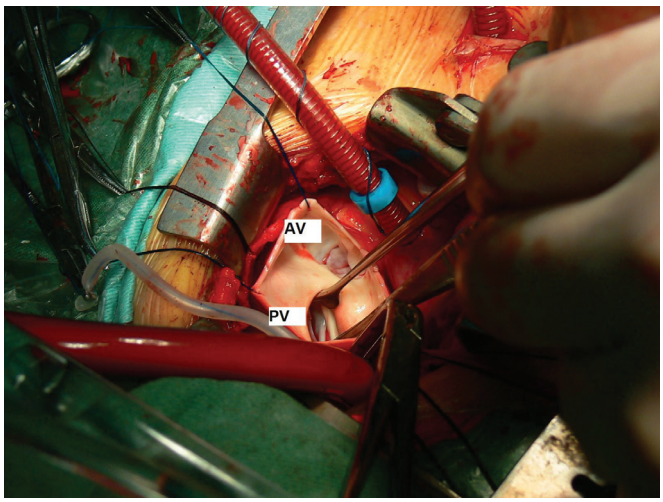

Figure 2. Internal view through the aortic incision showing the aortic valve (AV) and the pulmonary valve (PV). The aortopulmonary window is the intermediate defect near the right pulmonary artery.

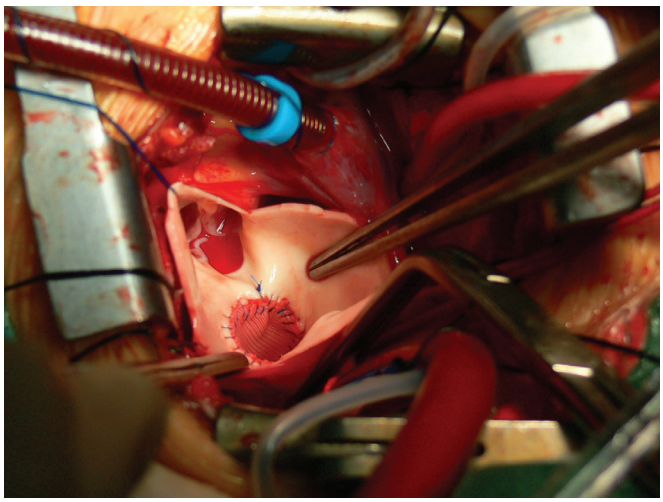

Figure 4. The aortopulmonary window was closed from the aortic side by using a Dacron patch, and the subaortic fibrous stenosis was removed.

DISCUSSION

The reason for reporting this case is based upon the rarity of the association of defects presented herein, and the surgery was finished with minimally invasive incision. The surgical approaches to APW began with ligation without the use of cardiopulmonary bypass (CPB), progressed to division and primary closure with CPB and cross clamp, and transaortic patch closure. Transcatheter closure has also been reported [Stamato 1995], but in that case the APW was a very small defect. Previously in our department, right vertical infra-axillary thoracotomy had been employed to repair congenital heart defects such as atrial septal defects [Yang 2001], ventricular septal defects [Wang 2010], atrioventricular septal defects [Wang 2003], and mitral valve replacement [Wang 2009]. With our accumulated experience and accurate diagnosis, we decided to perform the surgery through the right vertical infra-axillary incision and got a good result.

This case demonstrates the use of a minimal invasive surgery to close the APW. The advantages of this right vertical infra-axillary incision have been reported previously [Wang 2009; Wang 2010]. When APW is associated with interrupted aortic arch, patent ductus arterious, or tetrology of Fallot, the surgery should be performed through the standard median sternotomy. For appropriate patients with the simple APW, the right vertical infra-axillary incision may be a good choice.

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