Surgical Repair of Aortopulmonary Window in Infants

Chieh-Jen Wu, Bor-Yen Lin, Tung-Ho Wu, Anna Lo, Jun-Yen Pan, Pei-Luen Kang, Kai-Sheng Hsieh and Shu-Min Kuo

Background: Aortopulmonary window is a rare congenital heart defect resulting from a communication between the main pulmonary artery and the ascending aorta. Surgical closure is indicated in all patients with aortopulmonary window and should be performed at the time of diagnosis to prevent irreversible pulmonary vascular disease.

Methods: From 1990 to 2002, 4 infants with aortopulmonary window underwent surgical correction. The defects ranged from 0.4 to 0.6 cm in size and they were all proximal type. Associated cardiac lesions were present in 2 cases, including 1 case with coarctation of the aorta and patent ductus arteriosus, and the other with atrial septal defect. These procedures were performed under the cardiopulmonary bypass and the associated cardiac lesions were corrected simultaneously.

Results: There was neither early nor late death. During the period of follow-up (10 months to 12 years), no residual defect was noted and neither distortion nor stenosis of great artery was found.

Conclusions: Early correction of aortopulmonary window at the time of diagnosis is advised. Repair of aortopulmonary window can be performed in neonates and infants with excellent results.

Key Words: Aortopulmonary window • Aortopulmonary septal defect • Congenital heart disease

INTRODUCTION

Aortopulmonary window (APW), also known as aortopulmonary septal defect, is a communication between the ascending aorta and the main pulmonary artery in the presence of two normally formed semilunar valves, separate aortic valve and pulmonary valve. APW is a rare cardiac lesion representing approximately 0.1~0.2% of all congenital heart disease. Elliotson first described this cardiac anomaly in 1830. APW usually have symptoms of congestive heart failure and pulmonary hypertension. Irreversible pulmonary vascular disease, which is highly lethal, can occur as early as the first year of life. If untreated surgically, 40% of affected patients will die of intractable heart failure during the first year of life, and a large number of survivors will succumb to sequelae of congestive heart failure or pulmonary vascular disease in childhood. Anatomically, Mori and associates classified APW into 3 types. Proximal defects, the most common, are located just above the sinus of Valsalva; distal defects are located in the upper portion of the ascending aorta; total defects involve the majority of the ascending aorta. Different classifications of the APW have been described. Surgical closure is indicated in all patients with APW and should be performed at the time of diagnosis. In 1948, Gross first performed surgical ligation of an APW and closure of a patent ductus arteriosus (PDA) through left thoracotomy. In 1953, Scott and Sabiston described a closed method for division and suture of an APW.
between clamps.\textsuperscript{10} In 1957, Cooley reported the first successful repair of an APW utilizing cardiopulmonary bypass.\textsuperscript{11} The introduction of cardiopulmonary bypass allowed safer and more reliable different techniques to be applied, including the first transaortic closure of an APW reported by Wright et al. in 1968,\textsuperscript{12} and the first use of a patch to close an APW described by Deverall et al. in 1969.\textsuperscript{13} We report our experience of 4 cases with APW and describe the diagnostic methods, surgical techniques, and the results in these patients.

**MATERIAL AND METHODS**

Between October 1990 and May 2002, we performed 2200 cases of congenital heart surgery in our institution. Four patients presented with aortopulmonary window and underwent repair. It accounts for approximately 0.18% of all cardiac defects receiving surgical repair. There were 2 male and 2 female infants ranging in age between 18 days and 6 months (Table 1).

The first patient, a 6-month-old girl, 5.4 kg, had a history of recurrent upper respiratory infections, and obvious chest retraction at rest had been noted for 1 month before referred to our hospital. Cardiomegaly was noted by chest roentgenography, and the cardiothoracic ratio was near 60%. Two-dimensional echocardiography revealed a large aortopulmonary window, enlarged left atrium and left ventricle with moderate degree of mitral regurgitation. The findings of cardiac catheterization were APW with left to right shunting and severe pulmonary hypertension, while the pulmonary artery pressure to systemic pressure ratio was greater than 70%.

Patient 2, a 49-day-old boy, 3.5 kg, he suffered from progressive dyspnea and poor feeding since 2 weeks before admission to our hospital. Chest roentgenography showed cardiomegaly with cardiothoracic ratio of 60% and pulmonary hypervascularity. Two-dimensional echocardiography revealed an APW and mild dilatation of the left atrium and left ventricle. Cardiac catheterization established the definitive diagnosis and moderate pulmonary artery hypertension with a peak pulmonary artery systolic to aortic systolic pressure ratio greater than 0.5.

Patient 3, an 18-day-old boy, 2.2 kg, was referred to our hospital after birth immediately due to intrauterine growth retardation with low birth body weight of 2250 gm and imperforated anus. Surgical intervention of colostomy was carried out on the second day. Chest roentgenography showed cardiomegaly with cardiothoracic ratio of 60% and pulmonary hypervascularity. Two-dimensional echocardiography revealed an APW and atrial septal defect (ASD). Cardiac catheterization (Figure 2) confirmed the diagnosis, with

![Figure 1. Preoperative echocardiogram of case 3, high parasternal short-axis view showing the aortopulmonary window (arrow) between the ascending aorta (Ao) and main pulmonary artery (PA).](image)

<table>
<thead>
<tr>
<th>Case</th>
<th>Sex</th>
<th>Age</th>
<th>Weight (Kg)</th>
<th>Major symptoms</th>
<th>Associated anomalies</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>6 months</td>
<td>5.4</td>
<td>Recurrent pulmonary infection</td>
<td>None</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>49 days</td>
<td>3.5</td>
<td>Failure to thrive</td>
<td>None</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>18 days</td>
<td>2.2</td>
<td>Congestive heart failure</td>
<td>PDA, Coarctation</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>29 days</td>
<td>3.0</td>
<td>Congestive heart failure</td>
<td>ASD</td>
</tr>
</tbody>
</table>
Pulmonary artery hypertension at the ratio 0.7 of systemic pressure.

**Surgical technique**

In cases 1, 2, and 4, through a median sternotomy, cardiopulmonary bypass was instituted with bicaval and aortic cannulation. The right and left pulmonary arteries were temporarily snared on the commencement of bypass. Under moderate hypothermia, the heart was arrested with cold crystalloid cardioplegic solution injecting into the aortic root. In cases 1 and 2, a vertical incision was made over the main pulmonary artery. The defects were seen to be about 6 mm and 5 mm in diameter, respectively, and they were proximal type (Type I). The pulmonary artery branches were assessed carefully. Teflon patches were used to close the windows. The aortic cross-clamping times were 28 and 26 minutes, respectively. The pulmonary bypass times were 49 and 54 minutes, respectively. In case 4, a vertical incision was made over the anterior wall of the defect lesion. One 4-mm proximal defect (Type I) was inspected. The patch was sewn to the circumference of the defect, and the anterior edge of the patch was then sandwiched between the incision edges of the anterior aspect of the window by continuous 6-0 polypropylene running suture. The ASD was repaired simultaneously. The aortic cross-clamping time was 30 minutes, and the cardiopulmonary bypass time was 64 minutes.

In case 3, who suffered from APW, PDA and coarctation, cardiopulmonary bypass was established with the ordinary bicaval cannulation and arterial cannula connecting to innominate artery via a 4-mm Gore-Tex vascular graft and an additional descending aorta cannulation. The PDA was divided. The ductus arteriosus tissue and the aortic coarctation lesion were resected. The aorta was end-to-end anastomosed directly. Cardiac arrest was then achieved as per the procedures described above. A 4-mm proximal defect (Type I) was identified through an incision over the anterior wall of the defect lesion. It was closed with a glutaraldehyde-treated pericardium patch. The aortic cross-clamping time was 27 minutes, and the cardiopulmonary bypass time was 126 minutes.

**RESULTS**

All 4 patients had good recovery from operation (Table 2). They survived well, without any cardiac events. They are followed up regularly by echocardiography. These studies have documented normal left ventricular function, no residual aortopulmonary defect, no evidence of narrowing or stenosis for great arteries, and no evidence of semilunar valve distortion. The first case, who presented with moderate mitral regurgitation (MR) pre-operation, his heart function improved post-operation.

**Table 2. Investigative data, surgical intervention and outcomes**

<table>
<thead>
<tr>
<th>Case</th>
<th>Pulmonary/Systemic pressure ratio</th>
<th>Pulmonary/Systemic Flow ratio</th>
<th>Location of defect</th>
<th>Technique (approach)</th>
<th>Type of repair</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>0.7 (65/90)</td>
<td>3</td>
<td>Proximal</td>
<td>Transpulmonary</td>
<td>Teflon patch</td>
<td>Good</td>
</tr>
<tr>
<td>2</td>
<td>0.5 (50/100)</td>
<td>2.5</td>
<td>Proximal</td>
<td>Transpulmonary</td>
<td>Teflon patch</td>
<td>Good</td>
</tr>
<tr>
<td>3</td>
<td>None</td>
<td>2.5</td>
<td>Proximal</td>
<td>Transwindow</td>
<td>Pericardial patch</td>
<td>Good</td>
</tr>
<tr>
<td>4</td>
<td>0.7 (55/80)</td>
<td>3</td>
<td>Proximal</td>
<td>Transwindow</td>
<td>Teflon patch</td>
<td>Good</td>
</tr>
</tbody>
</table>
with trivial to mild MR. The third patient, who required repair of aortic coarctation, showed no evidence of stenosis. They are all active and asymptomatic and in New York Heart Association class I at follow-up ranging from 10 months to 12 years (mean 5.5 years).

**DISCUSSION**

Aortopulmonary window, a rare congenital heart anomaly, is caused by abnormal development of the embryologic truncoconal septum (aorticopulmonary septum), which normally separates the primitive arterial trunk into the ascending aorta and the pulmonary artery. This lesion represents by underdevelopment of the truncoconal septum, failed fusion or incompletely formation of the conotruncal ridges, resulting in the aortopulmonary window. Jacobs and associates defined aortopulmonary window as a communication between the main pulmonary artery and the ascending aorta in the presence of 2 separate semilunar valves. The presence of 2 separate semilunar valves distinguishes aortopulmonary window from truncus arteriosus. Commonly, the defect is large enough to function as a nonrestrictive left-to-right shunt, resulting in congestive heart failure, pulmonary hypertension and early development of pulmonary vascular disease.

In the past, APW was more frequently reported as an isolated lesion. In our experience, 50% of cases with APW had associated cardiac anomalies, 1 with ASD and the other patient with PDA and coarctation of the aorta. In recent reported series, 1 half to 2-third of patients with APW were associated with major cardiac defects. These cardiac defects include ventricular septal defect, ASD, PDA, interrupted aortic arch, coarctation of the aorta, tetralogy of Fallot, transposition of the great arteries, and coronary anomalies.

Many different classifications of APW have been advocated. Earliest, Mori and associates classified APW into 3 types, proximal, distal, and total. Proximal defects are located just above the sinus of Valsalva, a few millimeters above the semilunar valves, which is the most common type. Distal defects are located in the uppermost portion of the ascending aorta. Total defects consist of the majority of the ascending aorta. Richardson and associates proposed an alternative classification, which included both the APW lesions and anomalous right pulmonary artery origin from the ascending aorta. The Richardson system describes 3 types of anomalies of APW: Type I, typical aortopulmonary septal defect or window, is situated between the ascending aorta and the main pulmonary artery just above the sinus of Valsalva; it is due to incomplete septation of the aortopulmonary trunk. Type II, distal aortopulmonary septal defect, is located more distally, in which the ascending aorta communicated with the origin of the right pulmonary artery and the right pulmonary artery is from the main pulmonary artery. This anomaly is due to abnormal migration of the sixth aortic arch. Type III, consists of anomalous origin of the right pulmonary artery from the ascending aorta. This type is the result of unequal septation of the aortopulmonary trunks or truncus arteriosus. Ho and associates modified Mori’s anatomic classification, which is more useful to cardiologists considering transcatheter device for APW closure. They kept the terminology but added some additional description. Proximal defect are distinguished with little inferior rim separating the APW from the semilunar valves but with good superior rim. Distal defects are noted with a well-formed inferior rim but little superior rim. Total defects, also called confluent defects, are noted with little superior and inferior rims. Intermediate defects are recognized with adequate both superior and inferior rims. This defect is the most suitable group for transcatheter device closure. Our 4 cases all were proximal defects.

The initial diagnostic approach to APW was with cardiac catheterization. Current diagnostic techniques emphasize non-invasive assessment, and 2-dimensional echocardiography is very useful and facilitated in diagnosis of an APW. In transthoracic studies, the communication between the aorta and pulmonary artery can be best visualized from a variety of views, including subcostal coronal view, parasternal short and long-axis views and high parasternal short-axis view. It allows adequate demonstrations of the exact location and morphological features of the defect and associated cardiac anomalies. However, cardiac catheterization is necessary sometimes to provide information about pulmonary artery pressure and pulmonary vascular resistance.

Surgical closure is indicated in all patients with aortopulmonary window, and it should be undertaken as early as possible after diagnosis. Since the first report about successful repair of an APW by Gross in 1952, many authors have reported their results with a variety of
different techniques. In 1957, Cooley reported the first successful repair of an APW utilizing cardiopulmonary bypass. In 1966, Johanson and coworkers described the first transaortic direct closure of an APW. The first use of a patch to close an APW was described by Deverall et al. in 1969. We employed transpulmonary approach for APW in our first two cases. Neither coronary artery injury nor pulmonary stenosis or distortion was found. During the follow-up period (12 years and 8 years, respectively), no reintervention was needed. Currently, most centers recommend repairing defects through transaorta or transwindow with a patch placement. It gives better exposure of the defect, the edge is easily identified, and the orifices of the coronary arteries and aortic valve are noted to carefully avoid injury.

In conclusion, most patients with APW develop severe clinical features in early infancy. Neonatal repair or early surgical closure is indicated as soon as the diagnosis is established due to easily developed congestive heart failure, and to prevent the development of pulmonary hypertension. Associated cardiovascular anomalies should be repaired simultaneously. Repair of APW can achieve good results with low mortality and morbidity, even in early infancy, and late postoperative results are excellent.

REFERENCES