Aortopulmonary Window and the Interrupted Aortic Arch: Midterm Results With Use of the Single-Patch Technique

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Background. An aortopulmonary window (APW) associated with an interrupted aortic arch (IAA) can be associated with significant rates of perioperative mortality and recurrent arch obstruction. We assessed the outcomes associated with the use of a single pericardial patch technique for primary repair.

Methods. Between 2002 and 2011, 9 neonates and 2 infants with APW and IAA underwent single-stage repair, under a hypothermic (28°C) continuous cardiopulmonary bypass with antegrade selective cerebral perfusion. A single autologous pericardial patch (glutaraldehyde-fixed) was used both to augment the IAA end-to-side anastomosis and to close the APW by use of the "sandwich" technique.

Results. The IAA was type A in 6 patients and type B in 5 patients. The APW morphology was type I in 6 patients, type II in 4 patients, and type III in 1 patient. The median age and weight at operation were 11 days (range, 6 to 180 days) and 2.6 kg (range, 2.2 to 6.5 kg), respectively. The mean cardiopulmonary bypass and aortic cross-clamp times were 108.6 ± 27.5 minutes and 49.3 ± 13.4 minutes, respectively. One patient required additional closure of a ventricular septal defect. Delayed sternal closure was performed in 8 patients. The mean follow-up time was 6 ± 3 years. There were no early and no late deaths. Postoperative morbidity consisted of one postoperative stroke with no late sequelae. There were no reoperations. The last follow-up visits confirmed the absence of recurrent aortic arch obstruction and pulmonary artery branch stenosis in all patients.

Conclusions. Primary anatomic repair of APW associated with IAA can be safely performed. The efficiency of the single-patch technique was confirmed by the restoration of normal functional anatomy of the great arteries and aortic arch during follow-up.

Patients and Methods

Study Population

Approval for this study was granted by the Institutional Ethics Committee, and the need for individual consent from the children’s parents was waived. Eleven consecutive patients (nine neonates and two infants) with APW and IAA, who underwent single-stage repair with the use of a single pericardial patch technique in Marie Lannelongue Hospital between February 2002 and March 2011, were analyzed retrospectively.

Data Collection and Follow-Up

All data (preoperative, operative, postoperative) were collected retrospectively from hospital records between June 2012 and December 2012; data from the last visit (after July 2012) were also considered. The duration of follow-up was defined as the period between the operation and the last clinical visit.

Preoperative Variables

All data were retrieved from medical records. Demographic and anatomic data, past medical history (antenatal diagnosis), preoperative computed
tomographic scan data, and transthoracic echocardiographic results were systematically reviewed from medical records to formulate a database.

The Congenital Heart Surgery Nomenclature and Database Project [5] uses a classification system for APW similar to that proposed by Mori and colleagues [6]: type I is a proximal defect, type II is a more distal defect close to the origin of the right pulmonary artery, type III is complete absence of the aortopulmonary septum, and type IV is an intermediate defect not described by Mori and colleagues. We used this overall classification for APW.

The anatomy was described according to the classification by Cerolia and Patton [7] for IAA.

Operative Technique
Operative reports were recorded. Preoperatively, prostaglandin E1 was systematically administered to improve perfusion of the distal aorta through the ductus arteriosus, except for infants. Neonates and infants with APW and IAA were operated on as soon as a diagnosis was established, except when there was a contraindication. Those patients underwent single-stage repair under hypothermic (28°C) continuous cardiopulmonary bypass (CPB) with antegrade selective cerebral perfusion.

A median sternotomy was performed for all patients, and the aortic arch and cerebral vessels were extensively prepared and dissected. The main pulmonary artery and its branch arteries were mobilized and encircled with a silicone elastomer sling. The CPB was established with the use of bicaval cannulation, and a single aortic cannula was placed in the right side of the distal ascending aorta and advanced into the innominate artery during the aortic arch reconstruction (two infants), or the aortic cannula was placed through a 3.5-mm polytetrafluoroethylene graft (Fig 1) anastomosed to the right innominate artery (nine neonates). As the bypass was started, both pulmonary arteries were snared. Distal perfusion to the lower half of the body was through the APW and then through the ductus arteriosus.

At 28°C, the ductus arteriosus was ligated, the aorta was cross-clamped, and cold blood cardioplegia was instilled into the aortic root; then antegrade selective cerebral perfusion was instituted (we have previously reported on our continuous cerebral perfusion management for aortic arch operations) [8]. The APW was approached directly through an incision made along the anterior part of the APW itself, just between the great arteries; the incision was then extended proximally, transecting the superior part of the window (Fig 2). We identified the positions of the origins of the coronary artery, the pulmonary and aortic valve leaflets, and the right and left pulmonary arteries. An incision was then made from the anterosuperior part of the APW and extended distally into the ascending aorta and aortic arch concavity. All ductal tissues were resected, and the descending aorta was fully mobilized (if an aberrant right subclavian artery was present, it was divided to ensure proper mobilization). The descending aorta was then anastomosed to the ascending aorta and aortic arch by a partial end-to-side anastomosis (Fig 3). A single autologous pericardial patch (glutaraldehyde-fixed for 6 minutes) was used to both augment the aortic arch anastomosis and close the defect on the lateral aspect of the aorta by use of the “sandwich” technique (Fig 4). Subsequently, the eventual associated intracardiac lesion was repaired after return to full-flow CPB.

Postoperative Variables
The total lengths of stay in hospital and intensive care, duration of mechanical ventilation, and time with open chest were recorded. Reoperation was defined as any operation carried out later than 24 h after the initial repair. After the operation, patients were seen at least once a year for follow-up visits in Marie Lannelongue Hospital or in another hospital by the referring pediatric cardiologists. All patients were assessed by transthoracic echocardiography. Functional status was determined according to the New York Heart Association (NYHA) functional class.

Statistical Analysis
Statistical analyses were carried out with NCSS 2000 for Windows (NCSS, Kaysville, UT). Descriptive data for continuous variables are reported as medians, ranges, and means ± standard deviations; categoric variables are presented as relative frequencies. The probability of freedom from events (deaths) was estimated by the Kaplan-Meier method.

Results
Eleven consecutive patients (nine neonates and two infants) with APW and IAA underwent single-stage repair. There were six boys and five girls. Three patients had received an antenatal diagnosis. Preoperatively, the complete diagnosis was made with transthoracic echocardiography (n = 7) and confirmed by computed tomographic scan to define the aortic arch morphology (n = 4). None of the patients underwent diagnostic cardiac catheterization.

All patients underwent urgent single-stage repair except for 1 patient whose diagnosis was established later and who experienced a postnatal hemorrhagic stroke; for this patient, the operation was thus delayed for 3 weeks but had a favorable postoperative outcome.

The median age and median weight at operation were 11 days (range, 6 to 180 days) and 2.6 kg (range, 2.2 to 6.5 kg), respectively. The demographic data and diagnostic characteristics are summarized in Table 1. None of the patients had DiGeorge syndrome.

The mean CPB and aortic cross-clamp times were 108.6 ± 27.5 minutes and 49.3 ± 13.4 minutes, respectively. The mean period of selective antegrade cerebral perfusion was 42 ± 12 minutes.

Associated procedures were closure of an atrial septal defect in 3 patients and closure of a perimembranous ventricular septal defect in 1 patient. Restrictive muscular ventricular septal defects were left untouched.

There were no perioperative deaths. No patients were lost to follow-up, and the mean follow-up time was 6 ± 3
years. There were no later deaths, and the actuarial survival rate was 100% after 1, 3, and 5 years.

**Early Morbidity**

The chest was left open for 2 days in 5 patients and for 3 days in 3 patients. The mean duration of mechanical ventilation after operation was 167 ± 111 hours, with 2 patients receiving ventilation for 380 and 384 hours, respectively; 1 of these patients had a respiratory infection with favorable outcome, and the other had a perioperative hemorrhagic stroke without late sequelae.

The times in the intensive-care unit ranged from 4 to 18 days (mean, 8.7 ± 4.9 days), and the lengths of hospital stay ranged from 11 to 34 days (mean, 16 ± 9 days).

One patient with esophageal atresia underwent surgical repair 1 week after complete repair of APW associated with IAA. No other significant perioperative complications were observed.

**Late Morbidity**

No patient required reoperation (surgical procedure or use of a catheterization laboratory) for pulmonary artery stenosis or recurrent aortic arch obstruction.

There was no echocardiographic evidence of narrowing at the site of the aortic arch anastomosis. Peak acceleration in the aortic arch ranged from 1.4 to 1.9 m/s. There was no evidence of right or left pulmonary artery stenosis. No patients had significant residual APW or recurrent lesions.

There was no echocardiographic evidence of stenosis of the ascending aorta or pulmonary artery trunk, and no stenosis or insufficiency on the semilunar aortic or pulmonary valve.

**Functional Status**

At the final follow-up visits, all patients were within New York Heart Association class I.

**Comment**

An antenatal diagnosis was rare in our study, as in other reports, partly because equal pressure in the ascending aorta and pulmonary root in the fetus results in minimal detectable flow through the defect [1]. Furthermore, APW associated with IAA lacks the characteristic posterior deviation of the infundibular septum, which may prompt further interrogation of the aortic arch. As a consequence, antenatal diagnosis of APW with IAA has been only recently reported [9]. All patients received correct diagnoses and were assessed preoperatively with echocardiography and without catheterization; indeed, all patients underwent repair when they were younger than 6 months old. Catheterization in patients with APW...
should be reserved for the evaluation of pulmonary vasculature in patients older than 6 months [3].

We found, like others, that APW associated with IAA was not associated with DiGeorge syndrome, suggesting that the aortopulmonary window is a distinct malformation not related to abnormalities of the conal septum (ie, there is an intact ventricular septum [10]. The sole ventricular septal defect, which was surgically closed, was perimembranous. Posterior malalignment of the conal septum has been rarely described in APW with IAA, and this explains why left ventricular outflow tract obstruction is less common in this combination of defects than in isolated IAA [3]. We did not observe any coronary anomalies in our cohort of patients, as is sometimes described by other authors [11]. Because of the rarity of this entity, reports on treatments and outcomes are limited to relatively small groups of patients, most as case reports [12]. This study represents the largest unicentric series of patients with APW associated with IAA repair in neonates and young infants. A summary of the outcomes in patients from four series (reporting more than eight cases) is shown in Table 2. Our results compare favorably with other published series (Table 2), with no mortality and no need for reintervention for recurrent aortic arch stenosis. Early mortality had been reported to vary from 0% to 22% and aortic arch reintervention from 0% to 51%. Contrary to other series, extending over 23 years and starting in the early 1970s [3], our series is recent and homogenous, with patients treated within a short time period with the latest advances in pediatric intensive care and pediatric cardiac surgical procedures. This could explain our good results concerning mortality. Furthermore, single-stage repair is currently the rule but was not always used in other studies [10] in patients who underwent staged repair for this combination of defects.

We acknowledge that our technique is very similar to that already described by Barnes and colleagues [1]; unlike them, we always used selective cerebral perfusion and treated autologous pericardium, and we systematically enlarged the first centimeter of the descending aorta.

We used a unique surgical technique throughout our study, different from methods described in other reports [10, 13]. A variety of techniques has been used to repair APW: a transwindow approach with or without great vessel separation, transaortic closure, simple APW ligation, and single-patch or double-patch APW closure [10]. Furthermore, a variety of techniques has been used to repair an IAA: direct anastomosis with or without various patch augmentation, and end-to-end repair with use of a left subclavian arterial flap [10]. Our single-patch technique showed excellent results with no need for an aortic arch reintervention. As in other published studies, extensive mobilization of the descending aorta and arch vessels and meticulous resection of all ductal tissue are key elements of a good long-term outcome, and patch...
enlargement of the aortic arch anastomosis ensures optimal survival that does not require reintervention (aortic arch stenosis and compression of the bronchi) [14].

In contrast to the largest (multiinstitutional) study, which included 20 patients operated on between 1987 and 1997, at a time when enlargement of patch anastomosis was rarely used [10], we did not find that APW and type B IAA were risk factors for aortic arch reintervention. Other surgical techniques have been described for the management of APW combined with IAA, such as direct anastomosis of the descending aorta to the aortic window, but this has the same issue of aortic arch obstruction in the follow-up period [15].

Our single-patch technique for APW closure and aortic arch augmentation allowed closure of the APW by use of the “sandwich” technique [16]. The optimal approach for APW repair may depend on the size and specific location of the defect in any given patient, thus placing an emphasis on preventing narrowing of the great arteries or distortion of the aortic and pulmonary roots [3]. The “sandwich” technique provides good exposure and ensures that the patch does not interfere with the aortic or pulmonary valve, the coronary artery ostia, or the origin of the right pulmonary artery. In our study, there was no right pulmonary artery stenosis or recurrence of APW, no evidence of narrowing or stenosis of the ascending aorta, and no distortion of the semilunar valve. However, this procedure does carry the potential risk of residual or recurrence of the defect because there is no definite separation of the two great arteries. Our single-patch technique enables treatment of the majority of cases except those in which there is a rare aortic origin of the right pulmonary artery associated with IAA, which is then classified as a separate defect and is not considered a type of APW [5].

**Limitations**

Our results need to be interpreted with some caution because it included only a small number of cases. The mean follow-up time was quite short, and regular postoperative checks are recommended throughout the growth period of these patients to detect recurrent aortic arch stenosis. Arch reintervention for arch obstruction or bronchial compression can be an issue for the first year after the initial repair; however, recurrent obstruction can occur for as long as 13 years after complete repair [10, 17].

In conclusion, primary anatomic repair of APW associated with IAA can be safely performed. Patients with a primary single-patch repair of APW combined with IAA, at a maximum of 6 years follow-up, are demonstrated by echocardiography to have normal pulmonary arteries and aortic growth with excellent late functional results.

### Table 1. Patient Demographic and Diagnostic Characteristics

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age, Days</th>
<th>APW Type</th>
<th>IAA Type</th>
<th>Associated Cardiac Anomalies</th>
<th>Associated Noncardiac Anomalies</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>28</td>
<td>I</td>
<td>A</td>
<td>ASD</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>12</td>
<td>II</td>
<td>B</td>
<td>...</td>
<td>...</td>
</tr>
<tr>
<td>3</td>
<td>59</td>
<td>II</td>
<td>A</td>
<td>ASD</td>
<td>...</td>
</tr>
<tr>
<td>4</td>
<td>11</td>
<td>I</td>
<td>B</td>
<td>VSD, aRSCA</td>
<td>...</td>
</tr>
<tr>
<td>5</td>
<td>10</td>
<td>I</td>
<td>B</td>
<td>ASD</td>
<td>...</td>
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<td>180</td>
<td>I</td>
<td>A</td>
<td>...</td>
<td>...</td>
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<tr>
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<td>15</td>
<td>II</td>
<td>A</td>
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<td>9</td>
<td>8</td>
<td>II</td>
<td>A</td>
<td>mVSD</td>
<td>...</td>
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<tr>
<td>10</td>
<td>12</td>
<td>I</td>
<td>B</td>
<td>...</td>
<td>...</td>
</tr>
<tr>
<td>11</td>
<td>12</td>
<td>I</td>
<td>B</td>
<td>mVSD</td>
<td>Esophageal atresia</td>
</tr>
</tbody>
</table>

APW = aortopulmonary window; aRSCA = aberrant right subclavian artery (arteria lusoria); ASD = atrial septal defect; IAA = interrupted aortic arch; mVSD = restrictive muscular ventricular septal defect; VSD = perimembranous ventricular septal defect.

### Table 2. Literature Review of Series That Included More Than 8 Patients With Concomitant Repair of Aortopulmonary Window and Interrupted Aortic Arch

<table>
<thead>
<tr>
<th>Study</th>
<th>Follow-Up Time, y</th>
<th>Patients, n</th>
<th>Early Mortality, n</th>
<th>Late Mortality, n</th>
<th>Arch Reintervention, %</th>
</tr>
</thead>
<tbody>
<tr>
<td>Barnes et al [1]</td>
<td>u</td>
<td>9</td>
<td>0</td>
<td>u</td>
<td>u</td>
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<tr>
<td>Murin et al [13]</td>
<td>10</td>
<td>8</td>
<td>1</td>
<td>0</td>
<td>0</td>
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<tr>
<td>McElhinney et al [3]</td>
<td>11</td>
<td>9</td>
<td>2</td>
<td>0</td>
<td>28a</td>
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<tr>
<td>Konstantinov et al [10]</td>
<td>u</td>
<td>20</td>
<td>3</td>
<td>3b</td>
<td>51</td>
</tr>
<tr>
<td>Our study</td>
<td>6</td>
<td>11</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

a 3 patients with evidence of recurrent arch obstruction were monitored. b 3 late deaths at 2, 5, and 72 months postoperatively.

u = unknown.
Addendum

During the writing of this report, another two neonates have undergone the same single-patch repair. A 10-day-old boy with type 1 APW and type A IAA had a favorable early outcome. The other patient was a 12-day-old boy with type 1 APW and coarctation of the aorta. He underwent the same single-patch repair without resection of the coarctation and only patch enlargement of the aortic arch inner curve. Aortic arch angioplasty was performed for recurrent stenosis 2 months postoperatively.

References


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