A 1-year-old boy weighing 10.4 kg underwent successful biventricular repair for transposition of the great arteries, a ventricular septal defect, and a left ventricular outflow tract (LVOT) obstruction with moderate pulmonary stenosis of the bicuspid pulmonary valve (z score $-4.4$ for the pulmonary valve) by means of a modified Nikaidoh procedure with double root translocation by use of a valve-spared pulmonary root. The postoperative echocardiogram showed no LVOT obstruction, no aortic valve regurgitation, and mild pulmonary stenosis and pulmonary valve regurgitation. No reintervention has been required during the 6-year follow-up, with annular growth of the pulmonary valve.


Rastelli and réparation à l’etage ventriculaire (REV) procedures are the most common surgical procedures for transposition of the great arteries (TGA), ventricular septal defects (VSD), and left ventricular outflow tract (LVOT) obstructions with or without more than mild pulmonary stenosis. However, more aggressive procedures, including aortic root translocation (eg, the original Nikaidoh procedure) and double root translocation (DRT), have recently become important treatments of choice [1, 2]. We report our experience with a modified Nikaidoh procedure with DRT using a valve-spared pulmonary root.

A boy 1 year and 8 months old, weighing 10.4 kg, was referred to our institution for surgical repair after undergoing a left modified Blalock-Taussig shunt at 2 months of age. The preoperative examinations revealed a borderline cardiac anomaly between the TGA and a double-outlet right ventricle, VSD with an inlet extension considered to require repair for a remote-type VSD, and an LVOT obstruction with a 65 mm Hg pressure gradient, involving a bicuspid and tethered pulmonary valve (diameter, 7.4 mm; z score, $-4.4$). The aortic valve also had a 7.4-mm diameter (z score, $-4.6$).

The patient underwent a modified Nikaidoh procedure with aortic root translocation with use of a pivotal rotation technique, pulmonary root translocation with a valve-spared pulmonary root, and an arterial switch procedure with the Lecompte maneuver in 2006.

After cardiac arrest was induced in the usual manner, the pulmonary valve was checked through a longitudinal incision on the main pulmonary artery. This examination confirmed that a neoaortic valve would be difficult to use because of the size and severely fused bicuspid condition. We performed a modified Nikaidoh procedure with pulmonary root translocation (Fig 1). The cardiopulmonary bypass time and aortic clamping time were 323 and 171 minutes, respectively. The patient had a smooth postoperative course and was extubated the next day. Cardiac catheterization 1 year later confirmed a good LVOT without aortic insufficiency and an acceptable right ventricular outflow tract (Fig 2). Moreover, it revealed minor pulmonary regurgitation and 20 mm Hg pressure gradient at his bicuspid pulmonary valve, amorphous background of fibrin with degeneration and focal inflammation [3].

Calciﬁed amorphous tumors can present with symptoms of obstruction or embolization [6, 7]. The presence of MAC was noted to double the risk of stroke or transient ischemic attack in the Framingham cohort [8]. Embolization of fronds of MAC associated CAT may contribute to such an observation, although such link has not been conclusively determined.

Our case illustrates an atypical location of this nonmalignant, rare and unusual intracardiac tumor. Such tumors have a potential for embolization leading to stroke or transient ischemic attack, and surgical removal is usually curative. Our case was facilitated by use of intraoperative angioscopy, which resulted in complete removal of the mass reducing the likelihood of potential cardioembolic events in this patient.

References


Modified Nikaidoh Procedure With Double-Root Translocation in a 1-Year-Old Boy

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which had grown to 9.7 mm in diameter. He is now 8 years old, weighs 25 kg, takes no medication, and has no limits on exercise.

**Comment**

The Rastelli procedure and additional modified procedures show disappointing late outcomes for LVOT and a right ventricular outflow tract. For improving these results, several approaches have been proposed [1, 3], and in 2007 Hu and colleagues [2] reported the DRT technique, which comprises aortic root translocation and pulmonary root translocation with use of the pulmonary root enlarged by a transannular patch of bovine jugular vein. They subsequently reported good early results (5% deaths) and normal heart function with physiologic LVOT flow in 40 consecutive cases [4], concluding that the DRT technique was feasible and effective in treating anomalies of the ventriculoarterial connection, VSD, and pulmonary stenosis. However, 13 patients (~30%) had moderate pulmonary valve regurgitation.

Da Silva and coworkers [5] proposed pulmonary root translocation in a modified Rastelli-type repair. They

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**Fig 1. Details of surgical procedure. (A, B)** The entire pulmonary root, including the valve, was carefully removed. The ascending aorta was transected, and the aortic root was partially resected from the right anterior area to the left posterior area where the pulmonary root was removed. The coronary artery arising from the left anterior-facing sinus was temporarily resected. (C) The coronary artery in the right posterior-facing sinus and the surrounding myocardium were left intact to enable use of this area as a pivot in the rotation of the aortic root. The aortic root was translocated to the left ventricular outflow tract by use of a one-third pivot rotation. (D) A Lecompte maneuver was performed. The neoaorta was reconstructed after the form was adjusted to prevent too much pressure to the neopulmonary artery by partial resection of the neoaortic wall. (E) The pulmonary annulus was sutured to the periannular myocardium of the neoaorta. Aggressive commissurotomy was performed, and an acceptable level of function (100% of that of a normal pulmonary annulus) was achieved. Hence, a transannular patch was not added. The pulmonary artery was enlarged by a Y-shaped autologous pericardial patch.

**Fig 2. Results of cardiac catheterization 1 year after the procedure. The straight left ventricular outflow tract is shown, with no pressure gradient, no aortic regurgitation, and no defect in the coronary arterial perfusion. (AR = aortic regurgitation; LVOTO = left ventricular outflow tract obstruction; PCW = pulmonary capillary wedge pressure; PD = perfusion defect; PR = pulmonary regurgitation; PS = pulmonary stenosis; PV = pulmonary valve.)**

<table>
<thead>
<tr>
<th>Aorta</th>
<th>101/56</th>
<th>Right ventricle</th>
<th>47/10</th>
</tr>
</thead>
<tbody>
<tr>
<td>PCWP</td>
<td>9</td>
<td>Main pulmonary artery</td>
<td>24/11 mean 16</td>
</tr>
<tr>
<td>Right atrium</td>
<td>8</td>
<td>rt. &amp; lt. pulmonary artery</td>
<td>20/10 mean 15 (mmHg)</td>
</tr>
</tbody>
</table>
removed the pulmonary root with the valve and translocated it with or without a transannular patch enlargement, depending on a z score greater than −3 for the pulmonary valve. Most consecutive patients had acceptable results on follow-up; however, the pulmonary root translocation without the patch enlargement showed better late outcomes than did that with the patch enlargement for pulmonary valve growth. Those authors suggested that pulmonary root translocation might offer permanent resolution for complex congenital heart defects with malposition of the great arteries.

Some authors recommend performing an arterial switch operation and subpulmonary conal resection for TGA and LVOT obstruction with mild pulmonary stenosis [3]. Park and coworkers [6] reported acceptable results in 8 patients with TGA, VSD, and pulmonary stenosis using this approach. The aortic z score of the pulmonary annulus was −1.50 ± 1.13 (−3.42 to −0.35) preoperatively and increased to 1.10 ± 1.15 (−0.8 to 2.10) postoperatively [6]. They concluded that the indication for arterial switch operation could be extended to patients with a pulmonary annulus having an aortic z score of −3, despite a bicuspid pulmonary valve.

The valve-sparing approach has been proposed and adopted for patients with tetralogy of Fallot [7]. We have performed this approach for tetralogy of Fallot from 2005 and have been able to spare valves in more than 80% of consecutive patients. Moreover, more than 50% of patients with z scores ranging from −5 to −4 could be treated without transannular patches [8]. Thus, we aggressively chose DRT and used a valve-spared pulmonary root on this patient and observed an acceptable intermediate outcome. In our limited experience, valve-spared pulmonary root translocation may be feasible and effective for patients with a pulmonary annulus having a z score of approximately −4, even when DRT is performed.

**References**


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**Slide Tracheoplasty With Concomitant Aortic Arch Repair in a Low-Weight Neonate**

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We report the successful surgical management of a hypoplastic aortic arch, severe tracheal stenosis, and acute respiratory failure in a low-birth-weight neonate. The technical details that facilitated concomitant slide tracheoplasty and aortic arch reconstruction are discussed. (Ann Thorac Surg 2014;97:1057–9) © 2014 by The Society of Thoracic Surgeons

Although the outcomes of tracheoplasty have improved during the last decade [1, 2], surgical procedures on the trachea remain difficult, especially when associated with simultaneous repair of coexisting cardiovascular anomalies in low-birth-weight neonates. Herein, we describe the successful surgical treatment in a neonate with a hypoplastic aortic arch, severe tracheal stenosis, and acute respiratory failure that required emergent extracorporeal membrane oxygenation (ECMO) before repair.

A 5-day-old girl weighing 2.2 kg born at 37 weeks of gestation with an antenatal diagnosis of aortic coarctation and duodenal atresia underwent a duodenoduodenostomy and experienced severe respiratory acidosis despite high-frequency oscillatory ventilation. Emergency ECMO was initiated in the intensive care unit through a midline sternotomy with cannulation of the aorta and right atrium. The echocardiogram revealed a hypoplastic transverse aortic arch, coarctation, and a large patent ductus arteriosis. The proximal and distal aortic arch had z score values of −2.0 and −1.6, respectively. The z score of the aortic isthmus was −2.8. Computed tomography confirmed the hypoplastic aortic arch and a distal tracheal stenosis with complete tracheal rings (Fig 1). ECMO was converted to cardiopulmonary bypass (CPB) with bicaval cannulation and cooling to 26°C. The aortic arch and

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