Repair of Aortico-Left Ventricular Tunnel in the Neonate: Surgical, Anatomic and Echocardiographic Considerations

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SUMMARY Repair of aortico-left ventricular tunnel was accomplished for the first time in the neonatal period. The diagnosis was made with two-dimensional echocardiography. Repair was accomplished using an open-patch aortoplasty technique, which prevented valvular insufficiency, a significant problem in older patients in whom the repair has been attempted.

AORTICO-LEFT VENTRICULAR TUNNEL, first described by Levy et al.,1 is a rare congenital malformation that results in an equally rare condition, aortic insufficiency in the neonate. Although recognized since 1963, no previous repair of this anomaly has been accomplished in the neonatal period. The introduction of two-dimensional echocardiography for its diagnosis and patch aortoplasty for repair in the neonate will facilitate therapy.2

Case Report

A 2800-g male infant was born at 38 weeks of gestation after a normal pregnancy, labor and delivery. He was cyanotic, with a PaO₂ of 52 mm Hg, and had a cardiac murmur soon after birth. The infant was transferred to the University of California, San Francisco, for cardiac evaluation.

Physical examination at 18 hours of age showed a cyanotic infant in mild respiratory distress. Peripheral pulses were described as "full" and equal in all extremities, and there was no radial femoral delay. A thrill and a right ventricular heave were palpable at the lower left sternal border. A left ventricular lift was present in the sixth left intercostal space at the anterior axillary line. The first heart sound was normal; the second heart sound was single and accentuated. A systolic ejection click was heard in the third left intercostal space. A grade IV/VI harsh, ejection systolic murmur and a grade II/VI decrescendo diastolic murmur were heard along the left sternal border. The liver was enlarged and palpable 4 cm below the right costal margin.

The ECG demonstrated right atrial enlargement. Chest x-ray showed generalized cardiomegaly with decreased pulmonary vascular markings. He was suspected of having critical pulmonary stenosis. At 24 hours of age, the infant underwent right- and left-heart catheterization using an umbilical vein approach. Retrograde catheterization through the umbilical artery could not be achieved. Angiography revealed severe pulmonary valvular stenosis with tricuspid insufficiency. The right ventricular outflow tract appeared to be compressed by a dilated anterior sinus of Valsalva (fig. 1). The patient had right-to-left shunting through a patent foramen ovale, a patent ductus arteriosus, and no ventricular septal defect. The left ventricular outflow tract was considered abnormal and was enlarged (fig. 2). Catheterization data are listed in table 1.

Because of the critical pulmonic stenosis, the infant was referred for emergency pulmonary valvulotomy.

On the third day of life, he underwent median sternotomy. Exploration revealed an aneurysmally dilated anterior sinus of Valsalva compressing the right ventricular outflow tract, with enlargement of both the ascending aorta and pulmonary artery. A large ductus arteriosus was identified. The patient was heparinized, the ductus was ligated, and single atrial-to-ascending aortic cardiopulmonary bypass was instituted at 37°C. The heart was fibrillated and no left ventricular dilatation was observed. A domed, gelatinous-appearing pulmonic valve with three fused commissures and a minuscule central orifice were identified through a longitudinal pulmonary arteriotomy. The raphes were incised. No infundibular muscular obstruction was identified. The pulmonary arteriotomy was closed. The patient spontaneously defibrillated, was weaned from cardiopulmonary support without the need for inotropic agents and the incision was closed.

Postoperatively, the physical examination was unchanged except for resolution of the cyanosis, and although successfully removed from respiratory support, congestive heart failure and both systolic and diastolic murmurs persisted. Echocardiography was performed to evaluate the clinical problem. Because of the echocardiographic findings described below, the infant underwent retrograde left-heart catheterization at age 2 weeks (table 1). The aortic pulse pressure was markedly widened to 90/35 mm Hg, and there was free regurgitation of contrast material from the aorta into the left ventricle. Left ventricular end-diastolic pressure was moderately elevated, to 18 mm Hg, and there was no peak systolic pressure difference on withdrawal of the catheter from the left ventricle into the ascending aorta. Angiography demonstrated two channels, as noted on echocardiographic examination leading from the left ventricular chamber (fig. 3).

Despite treatment with hydralazine, digoxin and furosemide, the patient continued in severe congestive heart failure and, by 3 weeks of age, was 200 g below

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TABLE 1. Catheterization Data

<table>
<thead>
<tr>
<th></th>
<th>Pressures (mm Hg)</th>
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<tbody>
<tr>
<td>Right atrium</td>
<td>12 (a-wave) 6</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>68/16</td>
</tr>
<tr>
<td>Left atrium</td>
<td>16 (a-wave) 8</td>
</tr>
<tr>
<td>Left ventricle</td>
<td>70/10</td>
</tr>
<tr>
<td>Aorta</td>
<td>95-118/30-35 54</td>
</tr>
</tbody>
</table>

birth weight. At age 25 days, through the previous median sternotomy incision, exploration again revealed a markedly dilated anterior sinus of Valsalva, from which the right coronary artery arose (fig. 4A). The patient was heparinized, and single atrial-to-ascending aortic cardiopulmonary bypass was instituted with a 4°C prime. The patient was cooled to 22°C and the aorta was cross clamped. Through a longitudinal aortotomy, exploration revealed an 8-mm bicuspid aortic valve without commissural fusion and a markedly dilated anterior sinus of Valsalva with a 5 × 5-mm

aortico-left ventricular tunnel (fig. 4B). Closure of the aortico-left ventricular tunnel was accomplished using a 7 × 8-mm oval Teflon-felt patch and interrupted mattress 5-0 Tycron sutures (fig. 4C). Two sutures were placed through the free aortic wall, taking care to avoid injury to the right coronary artery; two were placed at the juncture of the annulus and the aortic wall, and one through the annulus itself. The aortotomy was closed, air was evacuated and the cross clamp removed (cross-clamp time 20 minutes). The patient was rewarmed, defibrillated at 32°C using 10 watt-sec into sinus rhythm, weaned from cardiopulmonary support at 37°C without the need for inotropic support, and closed.

Postoperatively, no evidence of aortic insufficiency was noted. Blood pressure was 70/40 mm Hg. Postoperatively, the echocardiogram showed slightly improved left ventricular function, and the mitral valve flutter found with aortic insufficiency was no longer present. There was persistence of the left ventricular outflow channel. Left ventricular hypertrophy persisted, and the aortic valve retained the appearance of a bicuspid valve. The patient was weaned from respirator support on the second postoperative day and was discharged on the tenth postoperative day. Follow-up at 5 months revealed normal growth and

Figure 1. Right ventriculogram demonstrating the domed valve (white arrow) with compression of the right ventricular outflow tract by large extrinsic filling defect, the sinus of Valsalva aneurysm and aortico-left ventricular tunnel vessel defect (black arrows).

Figure 2. Left ventriculogram demonstrating an anterior aneurysmal dilatation of the sinus of Valsalva (white arrows) and an abnormal left ventricular outflow tract (black arrows).

Figure 3. Retrograde left ventriculogram demonstrating two channels of egress from the left ventricular outflow tract: valve (small arrows) and tunnel (large arrows).
A development, with height and weight within the normal range. The child is thriving and demonstrates no evidence of aortic regurgitation. There is still a residual ejection systolic murmur and click in the aortic area consistent with mild aortic stenosis. The ECG shows findings of left ventricular hypertrophy.

**Echocardiographic Findings**

The two-dimensional echocardiogram was obtained after pulmonary valvulotomy. In multiple views, the right ventricular anterior wall, septum and left ventricular posterior wall were markedly thickened (fig. 5 and 6). The left ventricular end-diastolic dimensions and fractional shortening were increased, suggesting acute left ventricular volume overload. The M-mode echocardiogram showed diastolic flutter and early closure of the mitral valve leaflets suggesting aortic insufficiency as the source of left ventricular volume overload. The findings of exaggerated descending aortic pulsations in the subcostal long-axis view was further evidence of significant aortic insufficiency.

In the parasternal and apical long-axis views and the suprasternal notch short-axis view, there appeared to be two passageways for blood exit from the left ventricle (figs. 5 and 7). The anterior passageway led from the left ventricle to the anterior sinus of Valsalva. This passageway in the long-axis view resembled a large ventricular septal defect; however, we knew this was not the case from the angiogram. The posterior passageway led from the left ventricle to a thickened, domed aortic valve. The aortic annulus was approximately 7 mm in diameter, and the ascending aorta above the valve was markedly dilated. In the parasternal short-axis view, the aortic valve was bicuspid. Inspection of the suprasternal short-axis plane (fig. 5) showed that the passageway anterior and to the right led from the left ventricle to an enlarged sinus of Valsalva. This communication between the left ventricle and ascending aorta through the right sinus of Valsalva represents an aortico-left ventricular tunnel. Although the left ventricular end of the tunnel was visualized well from multiple echocardiographic
planes, it was extremely difficult to image the aortic end of the communication.

With cranial angulation from the apical four-chamber view, a dilated structure thought to represent a portion of the tunnel projected toward the right ventricle. This structure was seen intermittently moving into and out of the examining plane because of the cardiac motion. After a peripheral venous injection in this view (fig. 6), contrast echoes passed from right to left at the atrial level and filled all four chambers. Initially, there was an area of nonopacified blood in the left ventricle in the region of the left ventricular end of the tunnel (fig. 6, middle). This may have been caused by regurgitation of blood free of microbubbles from the aortic end of the tunnel into the left ventricle. Within a few beats the entire tunnel was opacified with the microbubbles as they were ejected into the aorta (fig. 6, bottom). Contrast echocardiography in the parasternal and apical long-axis views showed contrast echoes exiting from the left ventricle by way of the tunnel and the aortic valve. Also, because of severe aortic insufficiency, there was delayed clearing of contrast echoes from the left ventricle.

Additional abnormal findings on the two-dimensional echocardiographic examination included a thickened, dysplastic pulmonary valve, small branch pulmonary arteries and a dilated right atrium.

The echocardiogram taken on the eighth postoperative day no longer demonstrated findings consistent with insufficiency of the aortic valve, although left ventricular hypertrophy persisted.

**Discussion**

The diagnosis of aortico-left ventricular tunnel is difficult to establish in the neonate, but should be considered whenever aortic insufficiency presents as an isolated finding. Since the first description by Levy et al. in 1963, 1 32 cases of aortico-left ventricular tunnel in infants and children and five cases of aortico-left ventricular sinus of Valsalva rupture in adults have been reported. 2-4, 7-28 In infants, the tunnel has been proposed to represent an abnormal in utero com-

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**Figure 5.** Suprasternal notch short-axis view obtained with the transducer tilted anteriorly to visualize the left ventricle (LV) and ascending aorta. To the left, the LV communicates with the ascending aorta by way of the aortic valve (AoV). The aortic valve is thickened, and the annulus is narrowed. On the right, the LV communicates with dilated ascending aorta by way of an aortico-left ventricular tunnel (arrow). R = right; S = superior.

**Figure 6.** (top) Apical four-chamber view before a contrast injection. The left ventricle (LV) is thickened and dilated. The right ventricle (RV) is also thickened. The aortico-left ventricular tunnel cannot be seen in this still frame. (middle) After a peripheral venous saline injection, contrast echoes are seen in all four cardiac chambers because of a right-to-left atrial shunt. (Also, because of aortic insufficiency, non-contrast-containing blood from the ascending aorta washes contrast-containing blood out of the tunnel [T] in diastole. Therefore, the tunnel is relatively free of contrast echoes.) (bottom) In a subsequent systole, contrast-containing blood from the left ventricle flows forward into the tunnel and fills the tunnel with contrast echoes. A = apex; LA = left atrium; R = right; RA = right atrium.
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Figure 7. Apical long-axis view in diastole. The left ventricle (LV) is concentrically thickened and has an increased diastolic dimension. There are two passageways exiting from the LV to the ascending aorta (arrows). The posterior passageway leads from the LV to the aortic valve (AoV). The aortic valve is thickened and the aortic annulus is approximately 7 mm in diameter. The ascending aorta above the valve is dilated. The anterior passageway (arrow) leads from the LV to the ascending aorta by way of the right sinus of Valsalva and represents an aortico-left ventricular tunnel. A = anterior; I = inferior; LA = left atrium.

Communication between the aorta and left ventricle. The etiology of this condition is controversial; however, in utero aneurysmal rupture, ectopic coronary artery dissection, and valve development have been proposed.3, 11, 14 Aneurysmal rupture appears the most likely, as seen in the current case, with the defect occurring between the aortic annulus and the left ventricular septum. Our patients also had valvular stenosis as a presenting feature. Other studies have shown an association of pulmonic stenosis with this lesion.3, 11, 12, 16, 19, 20 In our patient, it was severe enough to be life-threatening and an immediate pulmonary valvulotomy was indicated. It was only after valvulotomy that the signs and symptoms of the aortico-left ventricular tunnel became evident. The association of pulmonary stenosis with the aortico-left ventricular tunnel appears to occur more frequently than by chance. Possibly, the dilated tunnel compresses the right ventricular outflow tract in utero, and this may cause secondary pulmonary valvular changes.

Since early diagnosis can improve the results of treatment of this condition, as progressive left ventricular hypertrophy, dilation, and failure can occur, a high index of suspicion is mandatory in the infant presenting with aortic insufficiency. In the youngest patient yet treated (unsuccessfully), Giardina et al.7 identified the tunnel at angiography; however, it may be difficult to differentiate the possible causes (i.e., Marfan’s syndrome) of aortic insufficiency.1, 8, 7 Two-dimensional echocardiography can allow clear definition of the abnormal subvalvular pathway in multiple projections, as well as definition of the insufficient diastolic tunnel flow when contrast (microbubbles) is injected. We believe the echocardiographic findings in this case are unique. The echocardiographic findings were typical of acute aortic insufficiency. The left ventricle was dilated and contracted poorly. The entire course of the fistulous tract could not be identified, but the subaortic end was prominent and initially had to be differentiated from a ventricular septal defect. This may be an important echocardiographic indicator of the pathology. We believe that the finding of a fistulous tract from the aorta to multiple echocardiographic windows may be a characteristic finding. Contrast echocardiography was helpful in excluding a ventricular septal defect as there was no ventricular right-to-left shunting, which might be expected with a large ventricular septal defect and residual elevation of right ventricular pressure. Indeed, when this space filled, it did so after the contrast agent passed from the right to the left atrium, and hence to the left ventricle.

Previous studies have noted that aortic valve anomalies can occur with aortico-left ventricular tunnel. The hemodynamic consequences of aortic stenosis may not be evident while the tunnel is still patent. The echocardiogram demonstrated an abnormal aortic valve that was thickened and domed. The annulus was narrowed and the ascending aorta was dilated. After surgery, the finding of a bicuspid valve remained, but without clinical findings of severe aortic stenosis. Repeat hemodynamic studies will be needed to evaluate the development of aortic stenosis.

Repairs have been previously accomplished in 17 patients ages 4 months to 11 years.8 No adult with acute rupture has survived.9 Methods have included direct closure, external plication, pledged closure and, in two prior cases, patch closure.1, 3, 4, 8, 10, 11, 12-26 Among those who were successfully repaired, residual aortic insufficiency has represented a significant problem.8 In only three—a multiple pledgeted repair by Spooner et al.,10 a pericardial patch closure by Somerville et al. and a synthetic patch closure by Bjork et al.—have aorticovalvular insufficiency been documented.4, 8, 10 The etiology of this problem is described by Bjork to be distortion of the aortic annulus produced by tension in the tunnel closure.8 This is best avoided by the intraluminal aortoplasty patch repair (fig. 4C). This approach also allows direct inspection of the aortic valve and subvalvular areas critical in these patients due to the high incidence of associated lesions.

The current case demonstrated that diagnosis of the aortico-left ventricular tunnel, a rare congenital malformation resulting in aortic insufficiency in the neonate, is facilitated by the use of two-dimensional echocardiography as an adjunct to angiography, and with patch aortoplasty, successful repair is feasible in the neonatal period.

References

Computerized Axial Tomography of the Chest for Visualization of "Absent" Pulmonary Arteries

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SUMMARY To expand the search for central pulmonary arteries in six patients with absence of cardiac-pulmonary continuity, computerized axial tomography (CAT) of the chest was performed. The CAT scans were compared with previous arteriograms and pulmonary vein wedge angiograms. Three patients with type IV truncus arteriosus were studied, and none had a central, right or left pulmonary artery on CAT scan. However, two patients with tetralogy of Fallot with pulmonary atresia and a patent ductus arteriosus to the right lung demonstrated the presence of a left pulmonary artery. In addition, one child with truncus arteriosus with "absent" left pulmonary artery demonstrated a left pulmonary artery on the CAT scan. The CAT scan may therefore enhance our ability to search for disconnected pulmonary arteries in children with complex cyanotic congenital heart disease.

THE PRESENCE of bilateral pulmonary arteries is critical in the decision to consider patients with type IV truncus arteriosus, tetralogy of Fallot with pulmonary atresia, or truncus arteriosus with a unilateral pulmonary artery for surgical correction. If a central pulmonary artery can be found in each lung, then appropriate palliative surgery, such as banding of a hypertensive pulmonary artery or shunting to a small, underdeveloped artery, can be used to prepare the artery and lung for open heart surgery. Hypertensive pulmonary arteries are readily defined during diagnostic angiography. The small, underperfused pulmonary artery, however, is difficult to detect. Recent pathologic work suggests that central left and right pulmonary arteries are almost always present and patent, even in patients thought to have type IV truncus arteriosus. Sotomora and Edwards questioned whether truncus type IV even exists.

The standard method of looking for central pulmonary arteries has been aortography, followed by selec-
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