Aortico–Left Ventricular Tunnel: 35-Year Experience

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OBJECTIVES
The purpose of this study was to review our 35 years of experience with aortico–left ventricular tunnel (ALVT), with emphasis on diagnosis, surgical details, and follow-up.

BACKGROUND
Aortico–left ventricular tunnel is a rare congenital anomaly. Neonatal surgery has been advocated in all due to long-term concern of valvar aortic regurgitation (AR).

METHODS
We identified 11 patients from 1963 to August 2002. Clinical, echocardiographic, catheterization, and surgical details were reviewed.

RESULTS
Eight of 11 patients presented at less than six months old (six with congestive heart failure) and three later with a murmur, all with clinical evidence of AR. Associated lesions, most commonly aortic valve and coronary artery anomalies, were present in 45%. Catheter occlusion was considered but not performed in five. Spontaneous occlusion was documented in one. Ten had surgery (nine in our institution), seven with direct suture and two by patch closure of the aortic end of the AVLT. At follow-up (median, 5 years; 1 month to 35 years), all were asymptomatic; three had residual ALVT (one moderate, two small/trivial), with at most mild AR.

CONCLUSIONS
Aortico–left ventricular tunnel is a rare cardiac malformation with a good post-operative long-term outcome. Associated lesions occurred in 45%. Catheterization should be reserved for patients with unclear non-invasive findings or transcatheter closure. We recommend surgery for most patients. We report spontaneous closure in one patient, prompting consideration of conservative follow-up in rare small, asymptomatic AVLT. (J Am Coll Cardiol 2004;44:446–50) © 2004 by the American College of Cardiology Foundation

RESULTS
Clinical presentation. Seven of 11 patients were male. Eight presented less than six months old, six with congestive heart failure (CHF), and three presented later in childhood with a murmur (Table 1). All had clinical findings consistent with aortic regurgitation (AR).

Echocardiography. Pre-operative echocardiograms were available in 9 of 11 patients; transthoracic 9 of 9, transesophageal 4 of 9. Aortico–left ventricular tunnel was diagnosed on the initial echocardiogram in 8 of 9. The ALVT originated superior to the right sinus of Valsalva. The sinotubular junction was difficult to assess anteriorly due to the tunnel. The tunnel coursed inferiorly and leftward, anterior to the aortic annulus (Fig. 1), posterior to the pulmonary trunk and infundibulum, and entered the LV superior to the ventricular septum, immediately below the aortic valve (AoV). In the long axis, clockwise rotation of the probe was sometimes necessary to demonstrate the entire length of the tunnel (Fig. 2A). Color Doppler demonstrated to-and-fro flow (Fig. 2B). The smallest diameter of the regurgitant jet was always at the ventricular end and measured 2 to 8 mm. Tunnel regurgitation was moderate (three patients) or severe (six patients). Aortic valve regurgitation was trivial or absent (six patients) or mild (three patients).

Catheterization. Catheterization was performed in nine patients. The ALVT was visualized in eight of nine (Fig. 3). Careful camera positioning was necessary to separate the AVLT from valvar AR, temporary balloon occlusion of the ALVT facilitating differentiation in two patients. There was a single left coronary artery with the right coronary artery.
(RCA) arising from the left coronary system in two patients. A normal conus coronary artery was seen in one patient. Transcatheter closure was considered, but not performed, in five patients.

**Associated lesions.** Associated cardiac anomalies (one or more) were present in five patients (45%) (Table 1), including AoV (three patients) and coronary artery (three patients) anomalies.

**Treatment.** Ten of 11 patients underwent surgery, nine in our institution, at median age 12 months (1 day to 24.4 years) (Table 2). The ALVT was clearly visible as an out-pouching in the anterior aspect of the aortic wall. The orifice was located above the right sinus of Valsalva or directly above the right-left commissure, close to the RCA ostium if present. The ALVT opened into the LV immediately below the right coronary cusp.

The aortic end of the tunnel was closed in nine patients, the tunnel plicated in two, and the ventricular end was closed in one (Table 2). One patient had closure of the aortic end of the ALVT at 21 months, followed by two AoV replacements, before referral to our institution, age 24 years. An RCA to residual ALVT fistula was identified and surgically closed.

Spontaneous closure, confirmed echocardiographically, occurred in one asymptomatic patient. At presentation with critical AoV and subaortic stenosis, the AVLT was mistaken echocardiographically and angiographically for valvar AR. Successful balloon aortic valvuloplasty was performed. The subaortic gradient resolved without intervention. At AVLT diagnosis, the LV end measured 2 mm, with mild-moderate regurgitation and normal LV dimensions. Tunnel anatomy was similar to other patients, but smaller, with less regurgitation. Conservative management pending possible further AoV interventions was undertaken. No further intervention has been required. Between age 2 and 3 years, the tunnel spontaneously closed.

**Follow-up.** Our nine surgical patients have been followed for 1 month to 35 years, median 5 years (Table 3). All are asymptomatic. Residual ALVT were excluded echocardiographically in five of eight patients, including one spontaneous closure. Residual ALVT was seen in three patients (trivial/small [two patients], moderate [one patient]). Additionally, residual ALVT was excluded angiographically six years post-operatively in one, and the other has no clinical evidence of residual shunt eight years after surgery. Coil occlusion of the moderate residual ALVT is planned.

**DISCUSSION**

**Etiology.** The embryological basis for ALVT remains unknown. Speculation has included an anomalous coronary artery, possibly the conal vessel, opening in the LV and rupture of a sinus of Valsalva aneurysm (5). However, we identified a normal conal vessel in one patient, and sinuses...
rupture generally occurs inferior to the RCA ostium (1,6). Other theories included an anterior aortic wall abnormality with communication into the LV (7), defective incorporation of the distal end of the bulbus cordis (8), and persistence of embryonic crests of the fifth aortic arch (9).

Clinical/diagnosis. Aortico–left ventricular tunnel is extremely rare (0.001% patients with congenital heart disease [10]). There were 9 patients with AVLT among 17,381 patients (0.0005%) catheterized in 35 years at our institution. As previously reported, we found a male predominance (6,11), often early presentation with CHF (6,11,12), and a significant incidence (45%) of associated cardiac defects (6,11,13,14). All our patients with associated defects had AoV (27%) and/or coronary artery (27%) anomalies.

The AVLT anatomy in our patients was similar to that previously described (7). Aortico–left ventricular tunnel can be diagnosed by transthoracic (5,15,16), transesophageal (17), and fetal (12) echocardiography and by magnetic resonance imaging (16). A significant amount of “aortic regurgitation” in infants should raise the possibility of this lesion (14). In the apical four-chamber view, the tunnel and valvar AR jets may be superimposed. Clinical findings, regurgitant jet width, LV dilation, and retrograde flow in the descending aorta are useful in assessing the severity of tunnel regurgitation. In our view, transesophageal echocardiography is helpful during surgery and interventional catheterization.

Management. Surgical closure has been recommended at the time of diagnosis, including asymptomatic patients (3,5,6,11,12), due to inadequacy of medical management (6,11,13), risk of developing severe AR in patients repaired later (2,3), and satisfactory surgical results in neonates and infants (12,18). We support this approach, but would consider close medical follow-up of small, asymptomatic ALVT, as we have documented spontaneous closure in one such patient.

Surgical closure techniques have included combinations of suture or patch closure of the aortic orifice of the ALVT, obliteration of the tunnel, and occlusion of the ventricular orifice, all with comparable long-term survival (6,11). There has been concern about possible AoV leaflet distortion after direct suture closure of the aortic orifice compared with
patch closure (13). Some investigators have favored patch closure in neonates and infants (12) and those with a larger aortic orifice (11), to avoid valvar distortion. However, seven of nine of our patients had suture closure of the aortic end and, at follow-up, valvar AR was, at most, mild.

Valvar AR is a major long-term concern, requiring valve replacement in up to half of reported patients (2,3,8). Aortic regurgitation may be secondary to turbulence-related damage to the leaflets or progressive aneurysmal dilation of the aortic root in patients surgically repaired at an older age (18), lack of support of the right aortic sinus (2,6), concomitant congenital AoV abnormalities, or iatrogenic surgical distortion of the valve (6). In our surgically repaired patients, including four operated on in infancy, AR is, at most, mild at follow-up. However, continued long-term follow-up is essential.

Successful percutaneous closure of an ALVT has been reported (19). We considered device closure in five patients, but desisted due to significant distensibility of the tunnel and/or proximity to the AoV leaflets and annulus or RCA ostium.

Conclusions. Aortico–left ventricular tunnel is a rare cardiac malformation with a good long-term outcome after surgery. Most patients present early in life with CHF and many have associated lesions. The diagnosis should be considered in infants with clinical signs of AR. Echocardiography can identify the ALVT and associated lesions. Catheterization should be reserved for patients with unclear non-invasive findings or transcatheter closure. We recommend surgery soon after diagnosis in symptomatic patients. Long-term review for AR is essential. Conservative management may be considered in rare, asymptomatic, small AVLT as we observed spontaneous closure in one such patient.

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REFERENCES

Table 2. Management of AVLT and Associated Defects

<table>
<thead>
<tr>
<th>Pt.</th>
<th>Age at Surgery</th>
<th>Tunnel Closure</th>
<th>Other Procedures</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1 day</td>
<td>Direct suture aortic end</td>
<td>Patch ventricular septal defect, Aortic valvotomy</td>
</tr>
<tr>
<td>2</td>
<td>15.6 yrs</td>
<td>Direct suture aortic end</td>
<td>Plication residual tunnel</td>
</tr>
<tr>
<td>3</td>
<td>24.4 yrs</td>
<td>Plication residual tunnel</td>
<td>(previous surgeries, another institution, see text)</td>
</tr>
<tr>
<td>4</td>
<td>32 months</td>
<td>Direct suture aortic end, patch closure ventricular end, plication ascending aorta</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>12 months</td>
<td>Patch closure aortic end</td>
<td>Aortic valvotomy</td>
</tr>
<tr>
<td>6</td>
<td>3 months</td>
<td>Direct suture aortic end</td>
<td>Balloon aortic valvuloplasty</td>
</tr>
<tr>
<td>7</td>
<td>4 months</td>
<td>Direct suture aortic end, plication tunnel</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>Surgery another institution</td>
<td></td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>Spontaneous closure</td>
<td></td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>3 yrs</td>
<td>Direct suture aortic end, plication ascending aorta</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>1 month</td>
<td>Patch closure aortic end, plication ascending aorta</td>
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AVLT = aortico–left ventricular tunnel.

Table 3. Follow-Up

<table>
<thead>
<tr>
<th>Pt.</th>
<th>Follow-Up*</th>
<th>AR†</th>
<th>Tunnel†</th>
<th>LVEDD</th>
<th>LV Function†</th>
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<tr>
<td>1</td>
<td>4.8 yrs</td>
<td>Mild</td>
<td>Moderate</td>
<td>Normal</td>
<td>Good</td>
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<tr>
<td>2</td>
<td>7 months</td>
<td>Trivial</td>
<td>Mild</td>
<td>Normal</td>
<td>Good</td>
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<tr>
<td>3</td>
<td>8.3 yrs</td>
<td>Mild‡</td>
<td>No</td>
<td>n/a</td>
<td>Good</td>
</tr>
<tr>
<td>4</td>
<td>33 yrs</td>
<td>Mild</td>
<td>No</td>
<td>n/a</td>
<td>Good</td>
</tr>
<tr>
<td>5</td>
<td>18.4 yrs</td>
<td>Trivial</td>
<td>No</td>
<td>n/a</td>
<td>n/a</td>
</tr>
<tr>
<td>6</td>
<td>14.7 yrs</td>
<td>Mild</td>
<td>No</td>
<td>Normal</td>
<td>Good</td>
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<tr>
<td>7</td>
<td>14.8 yrs</td>
<td>Trivial</td>
<td>No</td>
<td>Normal</td>
<td>Good</td>
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<tr>
<td>8</td>
<td>None‡</td>
<td>No</td>
<td>Trivial</td>
<td>No</td>
<td>Normal</td>
</tr>
<tr>
<td>9</td>
<td>6.8 yrs</td>
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<td>Normal</td>
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<td>Good</td>
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<tr>
<td>10</td>
<td>2 months</td>
<td>Mild</td>
<td>No</td>
<td>n/a</td>
<td>Good</td>
</tr>
<tr>
<td>11</td>
<td>1 month</td>
<td>Trivial</td>
<td>Mild increase</td>
<td>Good</td>
<td>Good</td>
</tr>
</tbody>
</table>

*From surgery to 8/02, except nine (no surgery) from first visit; †Echocardiography 1, 2, 3, 6, 7, 9, 10, 11; angiography 4; clinical examination 5; ‡Surgery, follow-up another institution; §Aortic valve replacement.

AR = aortic regurgitation; LVEDD = left ventricular end-diastolic dimension; LV = left ventricle; n/a = not available.