The obstructive subaortic conus

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ABSTRACT Eleven children are reported who had stenosis under a malposed aorta with gradients of 20 to 76 mm Hg between the right ventricle and aorta. The subaortic obstruction was caused by hypertrophy of the foreshortened infundibulum and malalignment of the infundibular septum relative to the remainder of the ventricular septum. Of these 11 patients, nine had a ventricular septal defect and seven had coarctation of the aorta. Rightward deviation of the infundibulum and aorta produced an unusually long left main coronary artery that was compressed by the stent of a bioprosthetic conduit valve in one patient. Serial cardiac catheterization studies in four patients showed progressive stenosis in each. Subaortic stenosis can develop in patients with malposition of the aorta and the frequency may be greater than 5% since milder forms are likely to occur. The obstruction can be progressive. The left coronary artery may be particularly vulnerable to compression after operative repair with an extracardiac conduit. Circulation 70, No. 3, 339-344, 1984.

EXTENSIVE experience has been reported regarding most of the cardiac anomalies associated with transposition of the great arteries (TGA), including ventricular septal defect,1-3 patent ductus arteriosus,4 and pulmonary stenosis.5-7 Cases of subaortic stenosis have previously been reported,8 but this anomaly has not been as well characterized. Here we report our findings in 11 patients with subaortic obstruction caused by a hypertrophied and malaligned conus.

Subjects and methods

Eleven patients were identified who each had (1) two ventricles, (2) aorta related to the right ventricle, and (3) a right ventricular-to-aortic gradient of over 19 mm Hg. Ten of the 11 patients had d-TGA and were selected from a total group of 214 patients with TGA; one of the 11 had double-outlet right ventricle (DORV) and was selected from a group of 34 patients with DORV. Clinical data on the 11 patients are listed in table 1. No patient had valvar aortic stenosis. Postmortem examination was possible in three of the eight children who died.

Results

Hemodynamic findings. Gradients across the right ventricular outflow tract ranged from 20 to 76 mm Hg. In some patients with widely patent ductus arteriosus, a gradient was not detected until the ductus was ligated (table 1, patients 1 and 3). Four patients who had no appreciable gradient at initial cardiac catheterization developed obstruction with time (23, 35, 65, and 76 mm Hg, respectively); three of these four (table 1, patients 2 to 4) had not undergone intervening surgery. Eight patients with large ventricular septal defects had equalization of pressure between the two ventricles.

Angiographic findings. Angiographically, the subaortic obstructions appeared similar but varied in degree among the 11 patients. Dynamic subaortic obstruction resulted from circumferential infundibular hypertrophy with both septal and free wall involvement (figure 1). In most cases, the infundibulum appeared foreshortened and deviated to the right. The aortic valve cusps were normal in 10 patients and were irregularly thickened but normally mobile in one.

Among eight patients with large ventricular septal defects, seven had malalignment of the sinus septum with the parietal band. The one child with DORV (table 1, patient 4) had superoinferior ventricles and multiple defects in the interventricular septum.

Cocartation of the aorta was discrete in five and associated with segmental hypoplasia in two patients.
Table 1
Clinical and hemodynamic data in 11 children with malposition of the aorta and subaortic stenosis

<table>
<thead>
<tr>
<th>Patient No./sex</th>
<th>Age</th>
<th>Diagnosis</th>
<th>RV</th>
<th>AAo</th>
<th>RV-Ao gradient</th>
<th>DAo</th>
<th>LV</th>
<th>MPA</th>
<th>Sat. (%)</th>
<th>Surgical procedures</th>
<th>Outcome; notes</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/M</td>
<td>1 day</td>
<td>d-TGA, VSD, CoA, PDA</td>
<td>65</td>
<td>65</td>
<td>0</td>
<td>50</td>
<td>65</td>
<td>55</td>
<td>88</td>
<td>BAS, CoA, PDA lig, PAB</td>
<td>Died; LCA compression</td>
</tr>
<tr>
<td>8 mo</td>
<td>17 mo</td>
<td>d-TGA, VSD, CoA</td>
<td>105</td>
<td>90</td>
<td>15</td>
<td>80</td>
<td>105</td>
<td>30</td>
<td>78</td>
<td>Damus and CoA</td>
<td>Died</td>
</tr>
<tr>
<td>2/M</td>
<td>3 mo</td>
<td>d-TGA, VSD, CoA</td>
<td>127</td>
<td>104</td>
<td>23</td>
<td>75</td>
<td>120</td>
<td>16</td>
<td>64</td>
<td>Damus and CoA</td>
<td>Died</td>
</tr>
<tr>
<td>7 mo</td>
<td>8 mo</td>
<td>d-TGA, VSD, CoA</td>
<td>140</td>
<td>105</td>
<td>35</td>
<td>35</td>
<td>85</td>
<td>85</td>
<td>67</td>
<td>B-H, PAB, CoA</td>
<td>Died</td>
</tr>
<tr>
<td>3/M</td>
<td>7 days</td>
<td>d-TGA, VSD, CoA, PDA</td>
<td>60</td>
<td>—</td>
<td>—</td>
<td>55</td>
<td>60</td>
<td>60</td>
<td>85</td>
<td>BAS, PDA lig, PAB</td>
<td>Died</td>
</tr>
<tr>
<td>5 mo</td>
<td>12 mo</td>
<td>d-TGA, VSD, CoA</td>
<td>115</td>
<td>90</td>
<td>25</td>
<td>68</td>
<td>110</td>
<td>33</td>
<td>70</td>
<td>Damus and Fontan</td>
<td>Alive and well</td>
</tr>
<tr>
<td>4/M</td>
<td>2 wk</td>
<td>l-MGA, DORV</td>
<td>90</td>
<td>90</td>
<td>0</td>
<td>50</td>
<td>90</td>
<td>90</td>
<td>86</td>
<td>CoA, PAB</td>
<td>Died</td>
</tr>
<tr>
<td>4 mo</td>
<td>18 mo</td>
<td>l-MGA, DORV</td>
<td>87</td>
<td>77</td>
<td>10</td>
<td>80</td>
<td>87</td>
<td>35</td>
<td>91</td>
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<td></td>
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<tr>
<td>4 yr</td>
<td>110</td>
<td>90</td>
<td>20</td>
<td>—</td>
<td>110</td>
<td>30</td>
<td>90</td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>5/M</td>
<td>2 days</td>
<td>d-TGA, small VSD, CoA</td>
<td>110</td>
<td>80</td>
<td>30</td>
<td>—</td>
<td>100</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
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<tr>
<td>6/M</td>
<td>3 mo</td>
<td>d-TGA, VSD</td>
<td>170</td>
<td>130</td>
<td>40</td>
<td>—</td>
<td>155</td>
<td>—</td>
<td>—</td>
<td>BAS</td>
<td>Died</td>
</tr>
<tr>
<td>7/M</td>
<td>2 mo</td>
<td>d-TGA</td>
<td>140</td>
<td>85</td>
<td>55</td>
<td>—</td>
<td>135</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>Died; total dynamic obstruction</td>
</tr>
<tr>
<td>8/M</td>
<td>3 days</td>
<td>d-TGA, VSD</td>
<td>130</td>
<td>100</td>
<td>30</td>
<td>—</td>
<td>130</td>
<td>—</td>
<td>—</td>
<td>BAS</td>
<td>Died at home</td>
</tr>
<tr>
<td>9/M</td>
<td>3 days</td>
<td>d-TGA, VSD, PS</td>
<td>105</td>
<td>70</td>
<td>35</td>
<td>—</td>
<td>100</td>
<td>—</td>
<td>—</td>
<td>BAS</td>
<td>Awaiting surgery</td>
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<tr>
<td>10/F</td>
<td>14 days</td>
<td>d-TGA, PS</td>
<td>130</td>
<td>100</td>
<td>30</td>
<td>—</td>
<td>130</td>
<td>—</td>
<td>—</td>
<td>BAS</td>
<td>Lost to follow-up</td>
</tr>
<tr>
<td>11/F</td>
<td>7 mo</td>
<td>d-TGA, CoA</td>
<td>120</td>
<td>100</td>
<td>20</td>
<td>—</td>
<td>65</td>
<td>—</td>
<td>—</td>
<td>Mustard</td>
<td>Alive and well</td>
</tr>
</tbody>
</table>

RV = right ventricle; AAo = ascending aorta; DAo = descending aorta; LV = left ventricle; MPA = main pulmonary artery; LCA = left coronary artery; VSD = ventricular septal defect; CoA = coarctation of the aorta; PDA = patent ductus arteriosus; l-MGA = l-malposition of the great arteries; DORV = double outlet right ventricle; PS = pulmonary stenosis. Surgical procedures: BAS = balloon atrial septostomy; CoA = repair of coarctation; PDA lig = ligation of PDA; PAB = pulmonary artery band; Damus = MPA-to-aortic anastomosis; B-H = Blalock-Hanlon operation; Mustard = venous switch with pericardium; Fontan = right atrial-to-pulmonary arterial anastomosis.

(figure 2). The coronary ostia arose in a manner consistent with d-TGA in nine patients, while the right coronary artery originated from the anterior aortic sinus in two.

Findings at autopsy. The three cardiac specimens obtained were remarkably similar, and their anatomic features confirmed the clinical observations (table 1, patients 1 to 3; figures 3 through 5). In each, the right ventricular free wall and ventricular septum were greatly hypertrophied. Malalignment of the hypertrophied infundibular septum produced not only pulmonary valvular overriding but also narrowing and rightward displacement of the subaortic right ventricular outflow tract.

The ventricular septal defects in the three specimens were of the malalignment infundibular type, bordered by the septal band, the infundibular septum, and the overriding pulmonary valve. Several tricuspid chordae inserted into the infundibular septum. Although the pulmonary artery arose approximately equally from each ventricle, the valve shared fibrous continuity with the mitral valve in all three cases.

The aorta was unusually rightward for transposition specimens, resulting in a long main left coronary artery. In one patient (table 1, patient 1) in whom an external conduit was inserted, the stent of the valve compressed the left coronary artery (figure 5, B).

Discussion

d-TGA may be complicated by associated anomalies such as ventricular septal defect and pulmonary stenosis. Subaortic stenosis has not been previously recognized as a potentially important and relatively common associated anomaly. We identified 10 of 214 of our d-TGA patients as having this condition and the incidence would have been higher if we had included patients with subaortic gradients less than 20 mm Hg.

From an anatomic perspective, subaortic stenosis is
subaortic stenosis was also observed even when the ventricular septum was intact. Foreshortening and lateral deviation of the infundibulum also contributed to obstruction. It may be tempting to interpret the conditions of our patients as tetralogy with transposition, but the geometry of the infundibular septal malalignment was clearly different in our patients than in classic tetralogy of Fallot. 

During embryogenesis in the patient with DORV, the subpulmonary conus was resorbed but the subaortic stenosis was also observed even when the ventricular septum was intact. Foreshortening and lateral deviation of the infundibulum also contributed to obstruction. It may be tempting to interpret the conditions of our patients as tetralogy with transposition, but the geometry of the infundibular septal malalignment was clearly different in our patients than in classic tetralogy of Fallot. 

Perhaps best considered simply a variant in the spectrum of infundibular pathology that may accompany conotruncal malformations. The cause of obstruction in our patients appeared to be circumferential hypertrophy of the subaortic infundibulum and malalignment of the infundibular septum relative to the remainder of the ventricular septum. While this malalignment usually resulted in a large ventricular septal defect,
tic conus was not. This rare variant of DORV dramatically illustrates the fact that the primary disorder resides in the conus, whether deviated rightward as in the patients with TGA (figure 1), or leftward as in the patient with DORV (figure 1, D).

Coarctation of the aorta was present in seven of 11 patients; in six of these seven, a large ventricular septal defect was also present. We believe that lateral deviation of the parietal band is related to all of these TGA-associated defects. Although the lateral deviation was rightward in most of our patients, Quero Jimenez and Perez Martinez\textsuperscript{11} have reported subaortic stenosis in a patient with D-TGA and leftward displacement of the infundibulum. Van Praagh et al.\textsuperscript{12} have previously reported that rightward deviation of the parietal band is found in patients with D-TGA and interrupted aortic arch. It is likely that the parietal band anomaly (which produces the subaortic stenosis and the ventricular septal defect) decreases intrauterine transascending aortic blood flow so that the stimulus to promote aortic growth is diminished.\textsuperscript{13, 14}

While the severity of the obstruction was often reflected in the magnitude of the subaortic pressure gradient, some patients had mild gradients when the obstruction appeared severe angiographically. In such patients, one must be aware of the associated conditions that affect the gradient: a large ventricular septal defect, patent ductus arteriosus, or coarctation of the aorta may decrease the subaortic gradient while converse conditions — intact interventricular septum, pulmonary stenosis, or pulmonary vascular obstruction — can enhance the gradient.

With time, progressive infundibular hypertrophy may cause increasing subaortic narrowing. Further-
more, pulmonary artery banding may induce infundibular hypertrophy, exacerbating the obstruction. Since clinical and electrocardiographic signs may not accurately reflect the severity of stenosis, repeated echocardiographic imaging or cardiac catheterization is necessary to follow the patient with subaortic obstruction.

When considering options for surgical repair of subaortic stenosis, excision of portions of the infundibular septum and free wall may be indicated; if the right ventricle is to remain the systemic ventricle, the resection should be accomplished through the aortic and tricuspid valves. The tissue to be resected is well away from the conduction system, and the development of surgically induced heart block would therefore be very unlikely. However, excessive resection of the infundibular septum could weaken the muscular support of

**FIGURE 4.** Pathologic examination of patient 3. Abbreviations are as in figure 3. A, View of the right ventricle demonstrating the cristal architecture, the ventricular septal defect, and the TV. B, The VS is thickened and a large defect is seen immediately caudad to the PV.

**FIGURE 5.** Postmortem photographs from patient 1. A, Right ventricular (RV) view shows tricuspid valve (TV) and patch (P) closing the ventricular septal defect. The infundibular septum (IS) is deviated to the right and there is severe subaortic stenosis (white arrow). B, View of anterior surfaces of the heart and the external cardiac conduit containing the bioprosthetic valve (valve), the stent of which is impacting the main left coronary artery (LCA).
the aortic valve, causing aortic regurgitation, and excessive excavation of the infundibular free wall could eventually produce a right ventricular aneurysm.

If relief of obstruction is accomplished directly, surgical options for restoration of normal physiology include a venous switch, the arterial switch, or the Rastelli repair in which an intraventricular baffle is used to connect the left ventricle to the aorta, creating right ventricular–to–pulmonary arterial continuity with an extracardiac conduit. When a valve-containing conduit is used, the prosthetic valve stent should be positioned to avoid compression of the left coronary artery16 (figure 5, B). The anteroposterior relationship of the great arteries appears to make the arterial switch procedure technically more feasible; most patients with TGA and subaortic obstruction have lateral deviation of the conus, creating a side-by-side relationship of the great arteries.

If the subaortic obstruction is not relieved directly, the Damus procedure17-19 can bypass the narrowing; in our patient (No. 4) with multiple ventricular septal defects and DORV, the Damus approach was combined with tricuspid valve closure and right atrial–to–pulmonary arterial anastomosis.

References
