A girl with symmetrical intrauterine growth retardation and a birth weight of 2240 g was delivered at term by primary cesarean section to a 27-year-old primiparous woman. Fetal echocardiography had shown a nearly atretic tricuspid valve. Postnatally, the child stabilized rapidly but showed signs of heart failure with tachypnea and general weakness, as well as cyanosis (transcutaneous oxygen saturation, 85%).

The presence of an aorto–right ventricular (RV) tunnel (ARVT) was confirmed by transthoracic echocardiography,
which showed a 5 to 6 × 15-mm structure extending from the proximal ascending aorta to the outflow tract of the RV, with the left coronary artery originating from the aortic end of the tunnel. The ARVT caused a strong left-to-right shunt during diastole (Figure 1 and Movies I and II in the online-only Data Supplement).

Both ventricles were well developed, with the RV showing distinct hypertrophy. Tricuspid valve diameter was 10 mm, with <3 mm of maximum diastolic opening and fibrotic, misshapen chorda. Blood flow across the tricuspid valve was minimal and mainly regurgitant, giving the appearance of nearly tricuspid atresia. There was a 5-mm-wide right-to-left shunt across a secundum-type atrial septal defect.

The mitral and semilunar valves were intact; the aortic and pulmonary valve annular diameters were 8 and 10 mm, respectively; and the ascending aorta was dilated to 16 mm. Chest x-ray revealed severe cardiomegaly and pulmonary hyperperfusion (Figure 2).

Retrograde catheterization of the ascending aorta and contrast injection proximal to the orifice of the tunnel confirmed a tunnel length of 15 mm, width of up to 6 to 7 mm, and normal origin of the right coronary artery from the right coronary sinus. The left coronary artery could not be visualized (Figure 3 and Movie III in the online-only Data Supplement).

After retrogradely passing the ARVT, the RV showed systemic pressure. Injection of contrast into the RV revealed mild tricuspid regurgitation and normal antegrade emptying into the pulmonary artery.

Surgery was carried out by opening the ascending aorta. Because of the intratunnular origin of the left coronary artery, the aortic orifice was left open, and a right ventriculotomy and patch closure of the right ventricular orifice of the tunnel were performed.

The hypertrophied RV showed marked endocardial fibroelastosis. Muscular resection was necessary to improve RV

Figure 2. Preoperative (A) chest x-ray showing severe cardiomegaly and 3 weeks postoperatively (B).

Figure 3. Still images from angiography. A, Contrast injection into the left ventricle (LV) shows the tunnel originating from the ascending aorta (Ao) and leading to the right ventricle (RV). B, Contrast drains into the RV from the ascending aorta. C, A catheter is passed from the descending aorta through the tunnel into the RV, thus demarcating the position of the tunnel.
function. The tricuspid valve appeared to open enough to allow adequate RV filling after tunnel closure.

Postoperatively, antegrade flow across the tricuspid valve was not accelerated above 1 m/s. RV systolic pressure was echocardiographically estimated at half the systemic blood pressure.

Delayed sternal closure was performed on the third postoperative day, and the patient’s recovery was uneventful. She receives acetylsalicylic acid for anticoagulation.

Histology of the right ventricular part of the tunnel revealed an elastic artery with smooth muscle cells and well-organized elastin lamellae (Figure 4).

ARVT is a rare entity thought to represent abnormal formation of the arterial valvar sinuses and leaflets during embryonic development of the outflow tracts,1 with only 16 patients reported in the English-language literature to date (Table).2–4 We demonstrate a neonate with ARVT in whom both preoperative echocardiography and angiography were normal.

Table. Reported Cases of Aorto–Right Ventricular Tunnel

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Journal</th>
<th>Age at Diagnosis</th>
<th>Other Features</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bharati et al</td>
<td>1973</td>
<td>Chest</td>
<td>6 h</td>
<td>RCA and left circumflex artery from ARVT</td>
<td>Autopsy case</td>
</tr>
<tr>
<td>Saylam et al</td>
<td>1974</td>
<td>Annals of Thoracic Surgery</td>
<td>10 y</td>
<td>Absent LCA ostium</td>
<td>Survived</td>
</tr>
<tr>
<td>Jureidini et al</td>
<td>1989</td>
<td>Pediatric Cardiology</td>
<td>0 d</td>
<td>Critical PS</td>
<td>Died</td>
</tr>
<tr>
<td>Kleikamp et al</td>
<td>1992</td>
<td>Journal of Thoracic and Cardiovascular Surgery</td>
<td>0 d</td>
<td>LCA from ARVT, rudimentary valve in ARVT, persistent ductus arteriosus, persistent foramen ovale</td>
<td>Died</td>
</tr>
<tr>
<td>Westaby et al</td>
<td>1992</td>
<td>Annals of Thoracic Surgery</td>
<td>3 mo</td>
<td>…</td>
<td>Survived</td>
</tr>
<tr>
<td>Rosengart et al</td>
<td>1993</td>
<td>Annals of Thoracic Surgery</td>
<td>3 mo</td>
<td>Absent RCA ostium, PS</td>
<td>Died</td>
</tr>
<tr>
<td>Van Son et al</td>
<td>1998</td>
<td>European Journal of Cardio-Thoracic Surgery</td>
<td>15 mo</td>
<td>…</td>
<td>Survived</td>
</tr>
<tr>
<td>Vargas et al</td>
<td>1998</td>
<td>Annals of Thoracic Surgery</td>
<td>7 mo</td>
<td>Persistent ductus arteriosus</td>
<td>Survived</td>
</tr>
<tr>
<td>Talwar et al</td>
<td>1999</td>
<td>International Journal of Cardiology</td>
<td>2 y</td>
<td>RCA from ARVT</td>
<td>Died</td>
</tr>
<tr>
<td>Hruda et al</td>
<td>2001</td>
<td>Heart</td>
<td>10 d</td>
<td>PS</td>
<td>Survived</td>
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<tr>
<td>Freund et al</td>
<td>2007</td>
<td>Fetal Diagnosis and Therapy</td>
<td>Prenatal, 36-wk gestation</td>
<td>LCA from ARVT</td>
<td>Survived</td>
</tr>
<tr>
<td>Poptani et al</td>
<td>2010</td>
<td>Journal of Invasive Cardiology</td>
<td>5 y</td>
<td>Single coronary artery</td>
<td>Survived</td>
</tr>
<tr>
<td>Bratsas et al</td>
<td>2010</td>
<td>Journal of the American College of Cardiology</td>
<td>31 y</td>
<td>LCA from ARVT</td>
<td>Adult</td>
</tr>
<tr>
<td>Talwar et al</td>
<td>2011</td>
<td>Journal of Cardiac Surgery</td>
<td>6 wk</td>
<td>RCA from ARVT</td>
<td>Survived</td>
</tr>
<tr>
<td>Singh et al</td>
<td>2012</td>
<td>Annals of Thoracic Surgery</td>
<td>16 y</td>
<td>Single coronary artery</td>
<td>Survived</td>
</tr>
<tr>
<td>Dwivedi et al</td>
<td>2012</td>
<td>Circulation</td>
<td>15 y</td>
<td>Single coronary artery</td>
<td>Survived</td>
</tr>
</tbody>
</table>

RCA indicates right coronary artery; ARVT, aorto–right ventricular tunnel; LCA, left coronary artery; and PS, pulmonary stenosis.
Phy suggested nearly tricuspid atresia, whereas the RV was well developed and hypertrophic as a result of retrograde diastolic filling via the ARVT (Figure 5). It is therefore important to consider that tricuspid valve function cannot be assessed reliably unless blood flow through the tunnel has been eliminated. In this patient, the tricuspid valve opened sufficiently; thus, surgical repair by patch closure of the ARVT and resection of muscular and endocardial fibroelastotic material from the RV was successful. Corrective surgery in the early neonatal period was necessary because of pulmonary hypertension and highly excessive pulmonary blood flow. Clinicians should be certain of the origin and course of the coronary arteries before making surgical decisions.

Disclosures

None.

References

Aorto–Right Ventricular Tunnel Causing Functional Tricuspid Atresia
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