Absent Tricuspid Valve with Aortic Atresia in Mixed Levocardia (Atria Situs Solitus, L-Loop)

A Hitherto Undescribed Entity

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SUMMARY Clinical, angiographic and pathologic features are described in a 36-hour-old male infant with mixed (discordant) levocardia, aortic valve atresia, absent tricuspid (left atrioventricular) valve and resultant massive cardiomegaly. This is the first reported case of this type of complex in a heart with discordant chambers. A possible embryologic explanation is offered for the concomitant presence of semilunar valve atresia and absence of the tricuspid valve.

CLINICAL AND PATHOLOGIC REVIEWS of mixed (discordant) levocardia (atrial situs solitus with L-loop with or without corrected transposition of the great arteries) have stressed the high association of this lesion with ventricular septal defect, pulmonic stenosis, left atrioventricular (A-V) valve regurgitation with or without stenosis, and Ebstein's disease of the left A-V valve.1 10 Obstruction to systemic blood flow has been noted infrequently.10, 11 This report presents the clinical, angiographic and pathologic findings in a neonate with mixed levocardia with ventricular inversion associated with aortic valve atresia and almost complete absence of the tricuspid valve.

Case Report

T.W. (UVH# 80-48-89) was the 3.8 kg product of uncomplicated pregnancy, labor, and delivery. Cyanosis and tachypnea prompted referral at 36 hours of age.

Examination on admission revealed a heart rate of 180/minute, respiration rate of 80/minute, and blood pressure of 50 mm Hg in the right upper extremity. Pertinent cardiopulmonary findings included a nonspecific grade I/VI systolic murmur and a single second heart sound. The liver was enlarged to the right iliac crest. The peripheral pulses were intermittently poor in all extremities. An electrocardiogram revealed a frontal plane QRS axis of +210°, absent left ventricular forces, and left atrial enlargement. Plain chest X-ray (fig. 1) demonstrated situs solitus and massive cardiomegaly, without evidence of pulmonary venous congestion. An arterial blood gas while the infant breathed room air revealed a pH of 7.33, PO2 of 33 torr and PCO2 of 36 torr. The infant required assisted ventilation, and immediate cardiac catheterization was performed (table 1).

Biplane angiography in the systemic venous ventricle (right-sided ventricle) revealed this chamber to be displaced rightward and superiorly (fig. 2). The chamber appeared smooth-walled, suggesting left ventricular morphology. The pulmonary artery arose from this chamber and was of normal caliber. Later in the sequence (fig. 3), filling of the descending aorta, brachiocephalic vessels and ascending aorta was noted. The aortic valve appeared anterior, superior, slightly to the left of the pulmonic valve. Retrograde aortography confirmed aortic valve atresia (fig. 4). The pulmonary venous drainage entered a markedly dilated left atrium and then filled the rest of the cardiac silhouette. The left-sided chambers were greatly enlarged with a grossly insufficient left A-V valve noted on left atrial cineangiography. An angiographic diagnosis of aortic atresia with left A-V valve insufficiency and severe cardiomegaly was made.

The infant died several hours after catheterization.

Postmortem Examination

The only pertinent findings12, 13 were in the heart. The heart was immensely enlarged, weighing 32 grams. From its base, two vessels emerged: a minute aorta situated anteriorly and to the left, and a large pulmonary trunk posteriorly and to the right (fig. 5). The atria were in normal position. The right atrium (fig. 6) communicated with the morphologic left ventricle (fig. 7) which was situated anteriorly and to the right. The left atrium (fig. 8) communicated with the morphologic right ventricle (fig. 8) which was situated posteriorly and to the left.

The right atrium (fig. 6) was enlarged, and its wall was of average thickness. What might be considered a limbus lay on the roof of the right atrium and it described a wide arc. The septum primum was poorly approximated to the limbus, producing a large atrial septal defect measuring 2 cm in greatest dimension. This extended in a more downstream direction than the usual fossa ovalis defect. The right A-V orifice (mitral) was enlarged for a mitral or tricuspid orifice. The mitral valve was increased in thickness and was connected to two abnormal papillary muscles, one posterior and

Table 1. Cardiac Catheterization Data

<table>
<thead>
<tr>
<th>Site</th>
<th>O2 Saturation*</th>
<th>Pressures</th>
</tr>
</thead>
<tbody>
<tr>
<td>Inferior vena cava</td>
<td>62%</td>
<td>—</td>
</tr>
<tr>
<td>Right atrium</td>
<td>86%</td>
<td>m = 12</td>
</tr>
<tr>
<td>Left ventricle</td>
<td>85%</td>
<td>50/15</td>
</tr>
<tr>
<td>Left atrium</td>
<td>87%</td>
<td>m = 12</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>87%</td>
<td>m = 12</td>
</tr>
<tr>
<td>Descenting aorta</td>
<td>84%</td>
<td>49/38</td>
</tr>
</tbody>
</table>

*All saturations were obtained with the patient ventilated with 100% oxygen.

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FIGURE 1. Frontal (left) and lateral (right) roentgenograms, demonstrating gross cardiomegaly. The pulmonary blood flow appears diminished. No pulmonary venous congestion is evident.

FIGURE 2. Anteroposterior (left) and lateral (right) projection of the systemic venous ventricle biplane angiogram. The catheter passes from IVC to RA and through a competent atrioventricular valve (arrows) into a displaced, morphologic left ventricle (LV). Fill of an unobstructed, horizontal pulmonary artery (PA) is noted.

FIGURE 3. Systemic venous biplane angiogram taken 1.5 seconds later in the sequence. Fill of the descending aorta (DAO), brachiocephalic vessels and an atretic ascending aorta (AAO) via a patent ductus arteriosus is seen. The aortic valve appears anterior and superior to the pulmonary artery (PA).
FIGURE 4. Frame from a descending aorta cineangiogram in the anteroposterior projection, confirming retrograde fill of the atretic aorta and coronary vessels and fill of the pulmonary circuit via a patent ductus.

one anterior. Each papillary muscle was thick, and as it approached the valve, it divided into numerous components which were connected by numerous chordae to the leaflets.

The morphologic left ventricle (fig. 7) was enlarged for a right or left ventricle and was distinctly thickened for a pulmonic ventricle. Because of the abnormal papillary muscles previously described, there was gross abnormality in the architecture of this chamber. It was, however, distinctly recognized as morphologic left ventricle by the smoothness of the septum and the absence of the septal and parietal bands. From this chamber emerged the pulmonary trunk; its orifice was enlarged. The ductus arteriosus was widely patent, forming the descending aorta.

The left atrium (fig. 8) was tremendously enlarged, and its

FIGURE 5. Anterior view of the heart with morphologic left ventricle and morphologic right ventricle opened. Note the tremendous size of the left side (morphologic right ventricle). LV = morphologic left ventricle, PT = pulmonary trunk, A = aorta, RV = right ventricle. Arrows point to the aneurysmally dilated morphologic right ventricle.

FIGURE 6. Right atrial and left ventricular view. RA = right atrium, MV = mitral valve, L = limbus, ASD = atrial septal defect.
The slightly ballooned wall was slightly thickened. It was recognized as morphologic left atrium by the presence of the septum primum on this side of the atrial septum and the typical atrial appendage.

The left (tricuspid) A-V orifice was immensely enlarged (fig. 7). It was guarded by a fragment of valvular tissue on the anterior and the inferior wall. This valvular tissue had a tenuous connection, without any papillary muscles, to the wall of the right ventricle. The entire septal wall and most of the inferior wall were devoid of valvular tissue.

The morphologic right ventricle (fig. 8) was markedly enlarged and its wall was paper-thin. Its endocardial aspect on the septal surface revealed a flattened trabecular sinus, and the infundibulum showed flattened septal and parietal bands, including a moderator band. Connected to this chamber was the aorta. The aortic valve, however, was atretic. The aorta consisted of a fine trunk containing a lumen which proceeded to the base of the heart. Here, it ballooned slightly and gave off the two coronary arteries. The coronary artery pattern was inverted. The transverse aorta was larger than the descending aorta, but smaller than normal.

**Discussion**

Absence of the tricuspid valve is a rare lesion. To date, it has been described only in hearts with normally related chambers and vessels. Kugel in 1932 first described absence of the tricuspid valve in pulmonary atresia with intact ventricular septum with aneurysmal dilatation of the right ventricle. Similar cases were described by Green et al., Morgan et al., and Cote et al. Although the latter authors considered their cases to be Uhl's disease, we believe their case belongs in the category of absence of the tricuspid valve. Kanjuh et al. described a case with pulmonary atresia and a small right ventricle. All of the above cases exhibited pulmonary atresia with intact septum and normally related great arteries, but with variation in the size of the right ventricle.

To our knowledge, ours is the first case in which absence of the tricuspid valve was found in a heart with discordant chambers. It is of interest that there is aortic atresia related to the morphologic right ventricle in our case, just as there is pulmonic atresia when absent tricuspid valve occurs in individuals with concordant ventricles and normally related great arteries. It would appear that there is some relationship between atresia of the outflow tract of the morphologic right ventricle and absence of the tricuspid valve.

Although the embryologic origin of the association is unknown, one may speculate as to the mechanism. It is possible that in the absence of a tricuspid valve, the fetal right ventricle is unable to generate sufficient systolic pressures to open its semilunar valve; the latter might be kept closed by pressures transmitted from the systemic circulation via the ductus arteriosus. Persistent semilunar valve closure in early fetal life would result in leaflet fusion and acquired atresia.

Aortic atresia in mixed (discordant) levocardia with ventricular inversion is rare. Out of 48 cases of mixed levocardia with ventricular inversion seen at the Congenital Heart Disease Research and Training Center since September 1, 1957, this is the only case with aortic atresia. Likewise, cases with coarctation are uncommon. It would appear that obstruction to systemic blood flow is uncommon in mixed levocardia with ventricular inversion.
References

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A hitherto undescribed entity.

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