Congenital Pulmonary Valvular and Infundibular Stenosis Complicating Ebstein's Anomaly of the Tricuspid Valve

A Case Report

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SUMMARY

Clinical, hemodynamic, angiographic, and echocardiographic as well as operative findings of a 19 years old girl are presented who had the unique combination of severe pulmonary valvular and infundibular stenosis with Ebstein's anomaly of the tricuspid valve. Pulmonary stenosis was relieved by infundibular resection and valvotomy. The atrial septal defect was closed with plication of atrialized ventricle and insertion of a porcine tricuspid prosthesis. Recatheterization 2 months postoperatively revealed disappearance of characteristic angiographic findings and slight residual gradient.

Additional Indexing Words:
Ebstein’s anomaly  Pulmonary stenosis

THE Ebstein's anomaly is characterized by downward displacement of redundant septal and posterior leaflets of the tricuspid valve into the right ventricle, the large anterior leaflet is normally attached to the annulus fibrosus. This results to the incorporation of a part of right ventricle into the right atrium which is called atrialized right ventricle. Different pathologic features and spectrum of abnormalities of this anomaly is discussed in the past1) and has been reviewed recently.2)

Valvular pulmonic stenosis commonly occurs as an isolated, uncomplicated congenital anomaly. Subvalvular pulmonic stenosis, whether infundibular or subinfundibular is generally associated with ventricular septal defect and is uncommon as an isolated anomaly.3)

Isolated pulmonic stenosis with Ebstein's anomaly are previously reported4) but combined valvular and infundibular stenosis of right ventricular outflow tract which was successfully corrected in our case with Ebstein's malformation has to the best of our knowledge not been observed. We are

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reporting the pre and postoperative findings of such a unique example of Ebstein's anomaly who was treated in our institution.

**CASE REPORT**

*Case history:*

The patient was a 19-year-old girl with chief complaint of cyanosis, shortness of breath, and easy fatigability with ordinary physical activities since early childhood. There was a history of occasional episodes of squatting in the past. Her condition deteriorated mostly in the last 2 years. There was no history of maternal rubella. Her mental and physical development was relatively normal.

*Physical examination:*

Patient was a rather well developed young girl with moderate muco-cutaneous cyanosis and digital clubbing, violaceous facial discoloration and rubor of terminal phalanges.

The blood pressure was 110/70 mmHg in both arms. Arterial pulses were normal and jugular venous pulse was striking with a prominent A wave, a normal X descent and a small V wave. Right ventricular lift was felt at left parasternal border. First heart sound was normal at the apex followed by a click. A grade III/VI ejection systolic murmur was heard at pulmouary artery area, radiating towards the lower parasternal as well as left supraclavicular regions accentuating during inspiration. A grade II/VI soft pansystolic murmur was detected at the right lower sternal edge which increased by inspiration. The second heart sound was widely split with a decreased intensity of the pulmonic component. The edge of the liver was hardly palpable and transmitted the presystolic pulsation.

*Laboratory data:*

The hematocrit was 65%. The electrocardiogram revealed the prominent P waves with marked right axis deviation of the QRS and anteriorly directed forces (Fig. 1).

![Fig. 1. Normal sinus rhythm with prominent P waves, marked right axis deviation and anteriorly directed forces.](image-url)
Echocardiogram:
Mitral and tricuspid valve echograms were simultaneously recorded (Fig. 2). The amplitude and the velocity of movement of the tricuspid valve were increased, but its systolic closure was not abnormally delayed compared to mitral valve motion. Diastolic closure slope of the tricuspid valve was also rapid. Interventricular septal motion was abnormal (type A).

Chest rontgenogram:
Cardiothoracic ratio was about normal with slight prominence of the right atrium. Pulmonary vascularity was within normal limits (Fig. 3).

Fig. 2. Simultaneous recordings of mitral and tricuspid valves. T=Tricuspid; M=mitral; RV=right ventricle (see text for details).

Fig. 3. Normal heart size and pulmonary vascularity with slight prominence of the right atrium.
Cardiac catheterization and cineangiography:

Right and left side cardiac catheterization were performed through the right femoral artery and vein. The electrode catheter could enter the left atrium. The pulmonary valve was entered by a balloon-tipped catheter with some difficulty by reducing the size of the balloon just beneath the semilunar valve. Intracardiac pressures were recorded (Table I). Prominent A waves were seen in the right atrium as is shown in Fig. 4.

The left ventricular cineangiogram was normal. Right ventricular angiocardioigraphy revealed a smaller than normal right ventricle, a relatively large right atrium and a cavity in between separated by a cone-shaped density from the right ventricle proper. A trace of dye visualized the left atrium (Fig. 5).

Rocking motion of the whole right heart was quite characteristic with the dye moving back and forth revealing a small amount of tricuspid regurgitation. Paradoxical distension of the middle compartment was apparent. Right ventricular outflow tract showed infundibular stenosis as well as pulmonary valvular stenosis with slight dilatation of the left branch of the pulmonary artery. Discordant intracavitary electrocardiogram and pressure relationship was demonstrated.

Diagnostic conclusion:

Ebstein's anomaly of the tricuspid valve with pulmonic infundibular and valvular stenosis and a right to left shunt through either a patent foramen ovale or an atrial septal defect.

Operative data and surgical method:

After a median sternotomy characteristic rocking motion of the heart could be recognized with paradoxical distension of the atrialized ventricle during systole.

Table I. Intracardiac Pressures

<table>
<thead>
<tr>
<th>Location</th>
<th>Preoperative pressure (mmHg)</th>
<th>Postoperative pressure (mmHg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right atrium</td>
<td></td>
<td></td>
</tr>
<tr>
<td>A wave</td>
<td>13</td>
<td>7</td>
</tr>
<tr>
<td>V wave</td>
<td>7</td>
<td>5</td>
</tr>
<tr>
<td>Mean</td>
<td>6</td>
<td>4</td>
</tr>
<tr>
<td>Right ventricle</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Systolic</td>
<td>100</td>
<td>60</td>
</tr>
<tr>
<td>Diastolic</td>
<td>2-6</td>
<td>0-4</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Systolic</td>
<td>18</td>
<td>30</td>
</tr>
<tr>
<td>Diastolic</td>
<td>8</td>
<td>16</td>
</tr>
<tr>
<td>Mean</td>
<td>11</td>
<td>20</td>
</tr>
<tr>
<td>Left ventricle</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Systolic</td>
<td>110</td>
<td></td>
</tr>
<tr>
<td>Diastolic</td>
<td>0-3</td>
<td></td>
</tr>
<tr>
<td>Aorta</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Systolic</td>
<td>110</td>
<td></td>
</tr>
<tr>
<td>Diastolic</td>
<td>60</td>
<td></td>
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<tr>
<td>Mean</td>
<td>75</td>
<td></td>
</tr>
</tbody>
</table>
Fig. 4. Right atrial pressure recording, revealing prominent A waves (13 mmHg).

Fig. 5. Right ventricular angiogram in late diastole revealing the right ventricular double density sign. Small arrow = true annulus of the tricuspid valve; large arrow = displaced tricuspid leaflets or false annulus; double arrow = infundibulum in diastole.
Cardiopulmonary bypass was utilized. The atrialized portion of the right ventricle collapsed after emptying the heart revealing the border between the atrialized segment and the right ventricle proper. Right atriotomy revealed a small atrial septal defect which was closed. Anterior leaflet of the tricuspid valve was large, thick, and with a whitish rough surface and a few fenestration. Besides ordinary chordae tendinae, there were a few abnormal chordae inserted directly to the right ventricular wall. The posterior and septal leaflets were atrophic with down and leftward displacement adhering directly to the border of the atrialized portion and right ventricle proper. A portion of the right ventricular infundibulum was resected and dome shaped pulmonic stenosis was corrected by valvotomy. A porcine heterograft valve was inserted at the position of the true annulus, distal to the coronary sinus with obliteration of the atrialized ventricle.

Surgical result:

This patient did very well after operation and was in class I New York Heart Association classification 2 months after surgery. Recatheterization revealed residual right ventricular outflow tract stenosis with a 30 mmHg gradient (Table I). In right ventricular cineangiography, rocking motion was almost abolished and paradoxical distension of the atrialized ventricle was not present.

Discussion

Although reparative surgery for Ebstein’s anomaly has been performed since 1962, the indication for surgery and the surgical procedure of choice for its correction has not been clearly defined.5) Despite the controversy regarding the optimal surgical approach in typical cases, there is no doubt in need for surgical repair in a patient who is symptomatic because of additional severe pulmonic stenosis.

Right ventricular and pulmonary arterial pressures are usually normal in patients with Ebstein’s anomaly but slight elevation of right ventricular pressure have been observed with parallel rise in pulmonary artery pressure or with right ventriculo-pulmonary arterial gradients.6),7) Two out of the 13 patients with Ebstein’s anomaly who underwent surgical repair in the Mayo Clinic had mild pulmonary stenosis.8) One of these patients was in functional class I but was operated on because of 2 episodes of transient hemiparesis attributed to paradoxic emboli through an atrial septal defect.8)

The variable clinical course is not always well explained by the wide range of pathological anatomy. The extent to which right ventricular outflow obstruction contributes to a patient’s symptoms is difficult to ascertain but it can increase the amount of right-to-left shunt, tricuspid regurgitation and probably the hemodynamic disturbance created by poor and often paradoxically contracting atrialized ventricle. There is no proof that combined pulmonic stenosis and atrial septal defect can increase the incidence of para-
doxic emboli but this possibility exists.

In our patient, although the anterior leaflet was dysplastic and other leaflets were markedly atrophic and deformed, atrialized portion of the RV was small.

Electrocardiographic pattern was indicative of right ventricular hypertrophy, not showing a long P-R interval, right bundle branch block, wide and polyphasic QRS complexes which are usually seen in Ebstein’s malformation.\(^7\),\(^9\),\(^10\)

Heart size was normal in contrast to the typical balloon-shaped enlarged heart seen in Ebstein’s malformation. Echocardiographic features were also different from what usually are seen in typical cases of Ebstein’s malformation.\(^11\) Lack of delayed tricuspid closure in systole and rapid diastolic closure slope of this valve as well as type A paradoxical interventricular septal motion were unusual, because delayed systolic tricuspid closure, decreased diastolic closure slope, and type B septal motion (flattened septal echoes during systole, instead of anterior motion in type A) are usually considered specific echocardiographic features of this anomaly.

Cineangiography is the most definitive way of diagnosis demonstrating the typical rocking motion of the right heart, and left and downward displacement of the septal and posterior tricuspid leaflets creating a right ventricular crest or double density sign which may take a cone-shaped appearance mostly in diastole. Right atrium and atrialized right ventricle were relatively small compared to other cases of this entity. In contrast to the dilated right ventricular infundibulum which is seen in typical cases of Ebstein’s anomaly, the infundibulum was narrowed and hypertrophied in our patient which needed resection. Also a dome-shaped pulmonic stenosis was present which was relieved by valvotomy. Despite infundibular resection and pulmonary valvotomy, 30 mmHg residual gradient was detected on recatheterization 2 months after surgery.

We conclude that additional lesions complicating Ebstein’s anomaly changes the typical diagnostic features of this entity and these patients may require earlier corrective surgery despite normal heart size and small atrialized ventricle.

**References**