Subpulmonic Obstruction in Complete (d) Transposition Produced by Redundant Tricuspid Tissue

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SUMMARY
Two cases are reported in which complete transposition of the great vessels was associated with an unusual form of subpulmonic obstruction. In each case, redundant tricuspid valvular tissue protruded through a ventricular septal defect into the left ventricular outflow tract, producing severe obstruction to the outflow of blood from the left ventricle.

The clinical course suggested the presence of left ventricular outflow obstruction. Both patients demonstrated progressive clinical deterioration characterized by increasing cyanosis, respiratory distress, and decrease in intensity of the cardiac murmur. Serial laboratory determinations revealed increasing hemoglobin and decreasing systemic oxygen saturation. The clinical deterioration was relieved only temporarily by atrial balloon septostomy.

The angiographic findings appear to be specific for this type of subpulmonic obstruction. In each case, a large asymmetric filling defect was demonstrated at the anterior border of the left ventricular outflow tract below the pulmonic valve.

When the clinical and laboratory findings suggest pulmonary or subpulmonary obstruction, careful evaluation of the outflow tract is indicated. If diagnosed clinically, redundant tricuspid tissue could perhaps be resected during a definitive operation, thus relieving the outflow obstruction.

Additional Indexing Words:
Left ventricular outflow obstruction
Ventricular septal defect
Complete transposition
Atrial balloon septostomy

RECENTLY developed technics of atrial septostomy permit most patients with complete transposition of the great vessels to survive the initial months of life. Total correction of this malformation at an appropriate age is now possible. The presence of significant obstruction of the left ventricular outflow tract complicates complete repair and seriously affects ultimate prognosis. Recent reports have described various types of subpulmonic obstruction and emphasized their characteristic angiographic findings.

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The purpose of this report is to document the clinical and pathological findings in two cases of subpulmonic obstruction produced by redundant tricuspid valvular tissue. Analysis of these two cases suggests that this malformation may be identified by angiography and removed at operation. To our knowledge, only a single case with similar pathological findings has previously been reported.

Report of Cases

Case 1
This 3-day-old female infant was admitted to UCLA Medical Center on January 18, 1967, for

*Previously included (case 7) in a report which described the association of complete transposition with hypoplasia of the right ventricle.
evaluation of a cardiac murmur and cyanosis. A grade III/VI ejection murmur was audible at the lower left sternal border. Thoracic roentgenography revealed a normal-sized heart with slightly decreased pulmonary vascularity. The electrocardiogram and Frank vectorcardiogram were normal for age. Hemoglobin was 19.9 g/100 ml. Cardiac catheterization and angiography demonstrated complete transposition of the great vessels with hypoplasia of the right ventricle and a ventricular septal defect (table 1). The lateral projection of the biplane venous angiogram demonstrated a large filling defect at the anterior border of the left ventricular outflow tract. The aortic and pulmonic valves were situated at approximately the same level (fig. 1). An adequate atrial balloon septostomy was performed, and right atrial saturation increased from 47% to 72%. The patient later developed tachycardia and marked dyspnea, which were controlled with digoxin and mercurial diuretics. The electrocardiogram 4 days after ballooning demonstrated right ventricular hypertrophy.

The patient was readmitted at age 3% months because of frequent cyanotic episodes. The systolic ejection murmur had decreased in intensity since the previous admission. Several cyanotic spells were observed during hospitalization, consisting of severe cyanosis, gasping respirations with the head extended, and extreme agitation. Each episode lasted for about 30 min and was variably responsive to morphine, oxygen, and intravenously administered sodium bicarbonate. The electrocardiogram again demonstrated right ventricular hypertrophy. Hemoglobin was 18.3 g/100 ml. Repeat cardiac catheterization demonstrated elevated left ventricular pressure (table 1). Right atrial saturation had decreased since the previous catheterization. Balloon atrial septostomy was again performed without improvement in clinical condition.

Two weeks later atrial septostomy was performed with a 1-cm biconical punch.10 During the procedure, the patient sustained a cardiac arrest. Pulmonary valvulotomy was immediately performed in an attempt to improve pulmonary blood flow. The patient did not respond to this or other therapeutic measures.

Necropsy examination of the heart revealed complete transposition of the great vessels and a 2 by 2-cm, surgically created atrial septal defect. The right ventricular chamber was hypoplastic; this was primarily due to lack of development of the inflow portion of the chamber. The pulmonary artery was smaller in diameter than the aorta, and the pulmonary and aortic valves were situated at the same level. The pulmonary valve was bicuspid, but the leaflets were thin and normally pliable. The tricuspid valve ring was hypoplastic, although the leaflets appeared normal. A 0.5 by 0.7-cm mass of redundant gelatinous valvular tissue originated from the ventricular surface of the septal tricuspid leaflet. The tissue protruded through a muscular septal defect into the left
ventricle, resulting in almost complete obstruction of the outflow tract (fig. 2A). Aberrant chordae tendineae originating from the papillary muscle of the conus passed through the septal defect to attach to the free edge of the redundant tissue.

Case 2

This patient was first admitted to the UCLA Medical Center at 12 hours of age because of marked cyanosis since birth. Physical examination revealed a grade II systolic ejection murmur at the left sternal border. Hemoglobin was 18.2 g/100 ml. The electrocardiogram was normal for age. Thoracic roentgenogram disclosed slight cardiomegaly and normal pulmonary vascularity. Umbilical cardiac catheterization revealed complete transposition of the great vessels (table 1). Atrial septostomy was performed using a 6.5F balloon catheter. Aortic oxygen saturation increased from 44% to 73% following the procedure, and the systolic murmur increased in intensity.

The patient was readmitted at age 3 months because of recent onset of dyspnea and grunting respirations and progressive cyanosis. No murmur was audible. Hemoglobin was 16.2 g/100 ml. Electrocardiography demonstrated left atrial enlargement and right ventricular hypertrophy. Repeat balloon atrial septostomy resulted in significant clinical improvement. Right atrial saturation increased from 33% to 64%. A grade III ejection murmur was again audible at the left sternal border and was thought to be due to pulmonic stenosis.

At 5½ months of age the patient was readmitted because of increasing cyanosis and respiratory distress. Hemoglobin was 20.4 g/100 ml. Electrocardiography again demonstrated left atrial enlargement and right ventricular hypertrophy. Cardiac catheterization revealed a decrease in right atrial oxygen saturation and elevated left ventricular pressure (table 1). Left ventricular biplane cineangiography demonstrated complete transposition of the great vessels and a ventricular septal defect. The left anterior oblique projection revealed a large filling defect in the left ventricular outflow tract which appeared to originate from the ventricular septum (fig. 3). The pulmonary valve could be visualized superior to the filling defect and the leaflets appeared limited in their motion.

The final hospital admission was at 8 months of age because of progressive dyspnea and severe cyanosis. Physical examination demonstrated a grade III ejection murmur at the third left intercostal space and hepatomegaly. Hemoglobin had increased to 23.6 g/100 ml. Electrocardiography demonstrated combined atrial enlargement and right ventricular hypertrophy. Congestive cardiac failure was treated with digoxin and mercurial diuretics. The patient was taken to the operating room where atrial septostomy was performed with a biconical punch, and a right Blalock-Taussig anastomosis was constructed. The patient seemed much improved in the early postoperative period. Systemic oxygen saturation increased from 47% to 85%. Eighteen hours after surgery the heart rate became irregular, blood pressure decreased, cardiac arrest ensued, and the patient could not be resuscitated.

Necropsy examination of the heart demonstrated complete transposition of the great vessels with a 5 by 10-mm iatrogenic atrial septal defect. The pulmonary artery was smaller than the aorta, and the pulmonary and aortic valves were situated at the same level. The pulmonary valve was bicuspid with thin leaflets of normal consistency. The tricuspid valve appeared normal when viewed from the right atrium. A large 4 by 8-mm firm nodular mass of tissue originated from the inferior surface of the septal leaflet (fig. 4) and was also attached to the posterior superior margin of a ventricular septal defect. A smaller 2 by 5-mm nodular mass of redundant tissue

Figure 1

Case 1. Forward angiogram (lateral view) demonstrating the right atrium (RA) and right ventricle (RV). The anterior aorta (Ao) arises from the right ventricle. Contrast material passing through a ventricular septal defect opacifies the posterior pulmonary artery (PA). The arrow points to a large asymmetric filling defect in the subpulmonic region.

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RIEMENSCHNEIDER ET AL.

Figure 2

(A) Case 1. (B) Case 2. Each panel displays a lateral view of the opened left ventricle (LV). Redundant tricuspid tissue (TT) protrudes through a ventricular septal defect into the outflow tract. The pulmonary trunk (PT) has been opened, while the bicuspid pulmonary valve is intact (opposing arrows). (A) The redundant tricuspid tissue (TT) extends from the tip of the arrow to the pulmonary valve ring. (B) A small probe has been placed in the outflow tract and passes through the bicuspid valve into the pulmonary trunk. The valve leaflets are thin and pliable and attach eccentrically to the wall of the pulmonary trunk by means of a raphé. There is no continuity between the redundant tissue and the mitral valve (MV).

originated from both the larger mass and the inferior edge of the ventricular septal defect. Chrodae tendineae from the papillary muscle of the conus and the septal wall attached to the redundant tissue. Both nodular masses protruded through a 1 by 0.5-cm muscular ventricular septal defect which was situated below the crista supraventricularis. The left ventricular outflow tract was almost totally obstructed by the redundant tricuspid tissue which was situated below the pulmonary valve (fig. 2B). Microscopic examination of the redundant tissue revealed highly cellular avascular primitive connective tissue (fig. 5).

Discussion

The frequent association of valvular anomalies in complete transposition of the great vessels has been emphasized by Layman and Edwards. Abnormalities of one or more valves were encountered in 26% of the pathological specimens examined by these authors. Of particular importance in terms of management and prognosis is the presence of pulmonary valvular and subvalvular types of left ventricular outflow tract obstruction.

Pulmonary valvular obstruction is often due to a bicuspid valve but may be the result of fusion of the commissures of a valve with three cusps.

Subpulmonic obstruction may be caused by fibromuscular abnormalities of the outflow tract or by anomalies of the ativoventricular valves (table 2). Three types of fibromuscular abnormalities have been described: (1) fibromuscular tunnel, (2) fibrous diaphragm, and (3) hypertrophy of the ventricular septum. Anomalous insertion of the anterior leaflet of the mitral valve into the upper portion of the ventricular septum has been reported as an uncommon cause of subpulmonic obstruction.

In addition to the two cases in this report, one other case has been documented in which redundant tricuspid valvular tissue caused subpulmonic obstruction. A nonobstructive
SUBPULMONIC OBSTRUCTION IN TRANSPOSITION

bicuspid pulmonary valve was also present in our cases, while in the previously documented case a subpulmonary diaphragm produced additional obstruction.

Redundant tissue of the atrioventricular valves, similar to that encountered in both of our cases, occurs in association with various cardiac malformations. Levy and associates described accessory tissue of the mitral (venous) valve which produced obstruction to blood flow from the venous ventricle to the pulmonary artery in three cases of corrected transposition of the great vessels. Redundant tricuspid valvular tissue obstructed the ventricular septal defect in three cases of tetralogy of Fallot described by Neufeld and co-workers. In three cases reported by Sellers and his group anomalous atrioven-

Figure 3
Case 2. Left ventricular (LV) cineangiogram (left oblique view) demonstrates a posterior pulmonary artery (PA) originating from the left ventricle. The arrow points to an asymmetric filling defect in the left ventricular outflow tract.

Table 2
Classification of Types of Left Ventricular Outflow Tract Obstruction in d-Transposition

<table>
<thead>
<tr>
<th>I. Pulmonary valvular</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Bicuspid valve</td>
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<tr>
<td>B. Commissural fusion</td>
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</table>

<table>
<thead>
<tr>
<th>II. Subpulmonary</th>
</tr>
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<tbody>
<tr>
<td>A. Fibromuscular</td>
</tr>
<tr>
<td>1. Fibromuscular narrowing</td>
</tr>
<tr>
<td>2. Fibrous diaphragm</td>
</tr>
<tr>
<td>3. Septal hypertrophy</td>
</tr>
<tr>
<td>B. A-V valvular</td>
</tr>
<tr>
<td>1. Redundant tricuspid tissue</td>
</tr>
<tr>
<td>2. Anomalous insertion of mitral valve</td>
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</table>
situated at the same level. Shafer and associates have emphasized that these findings are diagnostic of left ventricular outflow tract obstruction. When the clinical course and laboratory data suggest obstruction of the pulmonary or subpulmonary regions, and the above angiographic findings are noted, careful evaluation of the outflow region is indicated. The specific type and extent of the obstruction may be demonstrated by left ventricular angiography. Shafer and associates have described the angiographic findings in three forms of subpulmonic obstruction, including the long, fixed narrowing of a fibromuscular tunnel, the ringlike filling defect of a fibrous diaphragm, and the prominent muscular convexity due to hypertrophy of the ventricular septum. When subpulmonic obstruction is caused by abnormal insertion of the anterior mitral leaflet, angiography might be expected to demonstrate an asymmetric posterior narrowing of the outflow tract with limitation of movement of the anterior mitral leaflet.

In contrast to earlier descriptions of subpulmonic obstruction, the angiographic findings produced by redundant tricuspid tissue appear to comprise a unique pattern which may be diagnostic of this malformation. The redundant tissue is best visualized in the lateral or left oblique views. The asymmetric filling defect is situated adjacent to the ventricular septum and below the pulmonary valve. The abnormal tissue may also be demonstrated by right ventricular angiography if contrast material streams through the ventricular septal defect into the outflow tract. The septal defect may appear to be small because of partial obstruction of the lumen by the redundant tissue.

Subpulmonic obstruction due to redundant tricuspid tissue appears to be a surgically correctable malformation. The redundant tissue could theoretically be resected from the ventricular surface of the septal tricuspid leaflet without damaging the leaflet, thus relieving the subpulmonic obstruction and
Figure 5

Microscopic slide of redundant tricuspid tissue, demonstrating a highly cellular primitive connective tissue which is relatively avascular. Reduced from ×100.

permitting a significant increase in pulmonary blood flow.

Acknowledgment

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Addendum

Since the preparation of this manuscript, a difference, although closely related form of subpulmonic obstruction caused by redundant mitral valvular tissue has been reported by Rastelli and associates (Circulation 39: 83, 1969) in patients with complete transposition of the great vessels.

References

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