CLINICOPATHOLOGIC CORRELATIONS

Anatomic Causes of Pulmonary Stenosis in Complete Transposition

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SUMMARY Among 166 specimens with classical complete transposition, pulmonary or subpulmonary stenosis was present in 22 cases. The bases for obstruction were pulmonary valvular stenosis (one case), membranous subpulmonary stenosis (nine cases), anomalous attachment of the mitral valve to the ventricular septum (five cases) and combinations of causes (seven cases). Conditions contributing to combinations of causes included the above, in addition to accessory mitral or pulmonary valvular tissue and herniation of a tricuspid valvular pouch through a related ventricular septal defect.

In addition to the 166 cases mentioned, our collection contained six cases of atypical complete transposition characterized by a conus being present in each ventricle. In three of these six, subpulmonary stenosis was present and caused by a narrow inlet to the left ventricular conus.

PULMONARY STENOSIS associated with complete transposition of the great vessels may take several forms. Usually, this condition results from an abnormality additional to the basic malformation. Less commonly, the obstruction to pulmonary flow is part of an anatomic variant of complete transposition characterized by the presence of a left ventricular conus.

It is the purpose of this presentation to review the anatomic causes of pulmonary or subpulmonary stenosis in complete transposition. Among the 172 specimens with complete transposition in our collection, there are six cases of atypical complete transposition characterized by the presence of a conus in each ventricle (bilateral conus). In view of the unusual nature of bilateral conus, each of these six cases will be described, although subpulmonary stenosis was present in only three. The anatomic types of pulmonary or subpulmonary stenosis observed in classical complete transposition also will be considered.

Atypical Complete Transposition (Bilateral Conus)

According to current usage, a conus of a ventricle is a muscular walled tube that leads to either the aorta or the pulmonary trunk. The muscular tube is interposed between the semilunar valve, on one hand, and the related atrioventricular valve, on the other. In the normal heart, a conus is present in the right ventricle, while on the left side, as there is fibrous continuity between the aortic and mitral valves, no conus is present. In so-called double outlet right ventricle, the right ventricle possesses two coni, one subaortic and one subpulmonary. In classical complete transposition a conus leads from the right ventricle to the aorta, while no conus is present in the left ventricle as the pulmonary and mitral valves are in fibrous continuity. Under unusual circumstances, complete transposition may deviate from the classical, in that each ventricle has a conus, the right conus leading to the aorta and the left one to the pulmonary trunk.1-3

In the latter situation, the inlet to the left ventricular conus may be stenotic and be the basis for associated subpulmonary stenosis. In three of our six cases with bilateral conus, subpulmonary stenosis was present; in three there was no obstruction to pulmonary flow.

The critical distinction between the presence or absence of a conus depends upon whether there is or is not fibrous continuity between a semilunar valve and the related atrioventricular valve. This is usually easily determined by gross examination. In some instances histologic examination is necessary to make the distinction. Figure 1 shows both situations in complete transposition. When continuity is present (absent conus), a section taken of the pulmonary valve and anterior mitral leaflet shows interposition of fibrous tissue (the intervalvarular fibrosa) between the two valves. When continuity is not present, conus muscle intervenes between the two valves.

In each of the six cases herein described, the latter feature was present establishing the fact that a left ventricular conus, according to definition, was present. In each, a right ventricular conus was also present. The aortic and pulmonary valves lay at about the same plane.

In each case with a left ventricular conus, the clinical features were consistent with complete transposition. Situs solitus was present in each case and none showed splenic anomalies. In each specimen the aorta was more anteriorly positioned than the pulmonary trunk and the ascending aorta curved to the right, its position being that of d-transposition. As reference will be made to certain specific features of individual cases, the cases are summarized in Table I.

In each of the six cases, at least one ventricular septal defect (VSD) was present and in three of the cases additional deficiencies of the ventricular septum were found. The latter were considered as additional anomalies to the one that appeared to be part of the basic anomaly. In such cases the seemingly specific defect will be referred to as the VSD.

The cases could be subdivided into two groups based upon the characteristics of the VSD. Thus, Cases 1–3 were placed into Group I and Cases 4–6 into Group II.
In Group I, from the right ventricular aspect, the VSD lay immediately subjacent to the aortic valve. It had no upper border and intervened between the parietal and the septal limbs of the crista supraventricularis. From the left ventricular aspect, the VSD lay anterior to the base of the mitral valve (fig. 2). In two of the cases (Cases 1 and 2) the VSD lay proximal to the left ventricular conus. In Case 3 the upper aspect of the defect communicated with this subdivision of the left ventricle. In the cases of Group I the left ventricular conus was a distinct subdivision of the left ventricle which led to the pulmonary valve. The inlet to the left ventricular conus was walled by a column of muscle which extended from the septal to the posterior walls of the left ventricle. In Cases 1 and 2 the column of muscle formed the posterior wall of the inlet to the conus, while in Case 3 the column of muscle lay anterior to the inlet of the conus so that the inlet occupied a posterior position. In each case, the inlet of the left ventricular conus was narrow, yielding a severe degree of subpulmonary stenosis in Case 1 and moderate stenosis in Cases 2 and 3.

Regarding associated anomalies of significant nature, in Group I there was an additional VSD in Case 3 and double inlet left ventricle in Case 2.

In Group II, from the right ventricular aspect the VSD was remote from the parietal limb of the crista. In one case (Case 4) the defect lay above the septal limb of the crista, as seen from the right, while from the left ventricular aspect the defect lay at the level of the inlet of the left ventricular conus, lying below the muscle which was interposed between the pulmonary and mitral valves (fig. 3).

In Case 5, the right ventricular aspect of the VSD was at the junction of the sinus and infundibular segments of this chamber, the anterior edge of the defect lying along the posterior aspect of the lower part of the septal limb. From the left ventricular aspect the defect extended from the pulmonary valve to the base of the anterior lateral papillary muscle (fig. 4a). An additional VSD occupied the inferior aspect of the ventricular septum.

In Case 6, the VSD was similar to that in Case 5 and, as in Case 5, there was a second VSD, posteriorly. In Case 6 the myocardium of both ventricles was markedly hypertrophied with prominent trabeculations (fig. 4b).

In Group I the left ventricular conus formed a distinct basal subdivision of the left ventricle, while in Group II there was no such distinct subdivision. Except for muscular interposition between the mitral and pulmonary valves, the general configuration was similar to that of classical complete transposition. No pulmonary or subpulmonary stenosis was present in the specimens of Group II.

Table 1. Summary of Cases

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age</th>
<th>Sex</th>
<th>Position VSD</th>
<th>Pulmonary stenosis</th>
<th>Associated conditions</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>11 years</td>
<td>M</td>
<td>Between parietal and septal limbs</td>
<td>Proximal to conus</td>
<td>Severe Aberrant right subclavian, bicuspid pulmonary valve</td>
</tr>
<tr>
<td>2</td>
<td>1 month</td>
<td>F</td>
<td>Between parietal and septal limbs</td>
<td>Proximal to conus</td>
<td>Moderate Double inlet left ventricle, juxtaposition of atrial appendages, bicuspid pulmonary valve, hypoplastic pulmonary trunk, right aortic arch</td>
</tr>
<tr>
<td>3</td>
<td>16 years</td>
<td>M</td>
<td>Between parietal and septal limbs</td>
<td>Proximal to conus</td>
<td>Moderate Ventricular septal defect</td>
</tr>
<tr>
<td>4</td>
<td>Infant</td>
<td>F</td>
<td>Below septal limbs</td>
<td>At LV conus</td>
<td>None</td>
</tr>
<tr>
<td>5</td>
<td>9 days</td>
<td>F</td>
<td>Below septal limb</td>
<td>At LV conus</td>
<td>None Parachute mitral valve, subtotal, multiple muscular VSDs</td>
</tr>
<tr>
<td>6</td>
<td>3 months</td>
<td>M</td>
<td>Below septal limb</td>
<td>At LV conus</td>
<td>None Right ventricle anterior and to left, left ventricle posterior and to right, double inlet left ventricle, mild coarctation, VSD posterior basal</td>
</tr>
</tbody>
</table>
Classical Complete Transposition

Among the 166 specimens with classical complete transposition, there were 22 cases in which pulmonary or subpulmonary stenosis was present. In four additional cases lesions were present which, although not causing significant obstruction to pulmonary flow, appeared to have this potential were they more fully developed. In view of this potential, these four cases will be described separately in a subsequent section.

Existing Pulmonary Stenosis

Among the 22 cases of classical complete transposition with pulmonary or subpulmonary stenosis, there were 15 cases in which the obstruction was caused by one condition: pulmonary valvular stenosis or membranous subpulmonary stenosis or anomalous attachment of the mitral valve to the ventricular septum. In seven cases, obstruction to pulmonary flow was caused by a combination of two or more conditions.

Pulmonary Valvular Stenosis (One Case)

Congenital stenosis of the pulmonary valve was observed in one case, the valve exhibiting a unicommisural, unicuspid deformity. The ventricular septum showed a VSD.

Membranous Subpulmonary Stenosis (Nine Cases)

In nine cases, the obstruction was located in the outflow tract of the left ventricle and was characterized by the presence of a fibrous encircling membrane similar to that observed in membranous subaortic stenosis (fig. 5a). In this group of cases, the ventricular septum was intact in five cases and showed a VSD in four.

Anomalous Attachment of the Mitral Valve (Five Cases)

Subpulmonary stenosis caused by anomalous attachment of the mitral valve was observed in five cases. This was characterized either by direct or indirect (through chordae) attachment of the anterior mitral leaflet to the ventricular septum at the left ventricular outlet (fig. 5b). Such attachment prevented normal movement of the involved mitral leaflet from the left ventricular outflow tract during ventricular systole and obstructed the outflow tract. In one of these cases the mitral valve also showed a parachute deformity. In the latter case, the subpulmonary stenosis caused by the anomalous attachment of the anterior mitral leaflet to the ventricular septum was severe. It was associated with hypoplasia of the left ventricular cavity and major hypertrophy and endocardial fibroelastosis of the left ventricular wall (fig. 6). The appearance of the left ventricle was like that.
seen in congenital aortic stenosis with the so-called hypoplastic left heart syndrome. The ventricular septum was intact. Of the remaining four cases with anomalous attachment of the mitral valve, a VSD was present in three.

**Combinations of Causes**

Among the 22 cases of classical complete transposition with pulmonary or subpulmonary stenosis, there were seven cases, in each of which the obstruction resulted from two or more conditions. In two of these, there were membranous subpulmonary stenosis and a herniation of a pouch of the tricuspid valve through a VSD (fig. 7a and b). In one case a stenotic unicuspid pulmonary valve was associated with membranous subpulmonary stenosis.

In the fourth case anomalous attachment of the mitral valve was associated with accessory tissue of the mitral valve, the latter presenting into the outflow tract of the left ventricle (fig. 7c). In the fifth case, anomalous mitral valvular attachment was associated with herniation of a pouch of the tricuspid valve into the left ventricular outflow tract through a VSD.

Each of the remaining two cases showed a combination of three abnormalities contributing to the obstruction of pulmonary flow. In one of these, there were herniating septal leaflet of the tricuspid valve through a VSD and accessory tissue of both the pulmonary and mitral valves. The second case showed herniation of the tricuspid valve, membranous subpulmonary stenosis and accessory mitral tissue.

**Potential for Subpulmonary Stenosis**

We observed two conditions which, while not causing obstruction to pulmonary flow, nevertheless had this potential were the lesions of greater magnitude.

The first of these conditions was a pouch-like protrusion of the septal leaflet of the tricuspid valve, the pouch protruding through a related VSD and its fundus presenting in the left ventricular outflow tract. Ten examples of such pouches were observed in our entire series. In five cases described, such pouches contributed to the pulmonary stenosis caused in part by other conditions.

In five other cases such pouches, although present, did not appear either to cause or to contribute to pulmonary stenosis. In two of these, subpulmonary stenosis was caused by another condition and in three the pouches were isolated conditions and did not cause obstruction to pulmonary flow.

Accessory tissue either at or below the pulmonary valve was observed in two cases. In one this took the form of papillary-like excrescences attached to the contact surfaces of the pulmonary cusps and was associated with obstruction contributed by a herniated tricuspid valve and accessory.

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**Figure 4.** Two cases of complete transposition with left ventricular coni and without pulmonary stenosis. a. Case 5. Left ventricle (LV) and pulmonary trunk (PT). The pulmonary valve is separated from the mitral valve by a column of muscle. Two VSDs are present. b. Case 6. Left ventricle (LV) and pulmonary trunk (PT). The mitral valve is separated from the pulmonary valve by a column of muscle. The left ventricular muscle is dysplastic and hypertrophied. A VSD is present.

**Figure 5.** Isolated causes of pulmonary stenosis in classical complete transposition. a. Left ventricle (LV) and pulmonary trunk (PT). Membranous subpulmonary stenosis (between arrows) distal to VSD. Alteration of pulmonary trunk secondary to previous pulmonary valvular bacterial endocarditis. b. Anomalous attachment of anterior mitral leaflet into ventricular septum. Probe in VSD, which is obscured by the anomalous mitral attachment (point of arrow).
mitral tissue. In the other case, inverted cusp-like formations were attached near the roots of the pulmonary cusps. No pulmonary stenosis appeared to have been caused by this isolated condition.

Accessory tissue of the anterior leaflet of the mitral valve was observed in three cases. In each, the accessory tissue contributed, along with other conditions, in causing pulmonary stenosis. In no case did we observe accessory mitral tissue as an isolated cause of subpulmonary stenosis, although such a state might be anticipated.

Comment

Left ventricular outflow obstruction has important surgical and prognostic implications in complete transposition. Hence the recognition of an anatomical basis for co-existing or potential subpulmonary stenosis is important. Shaher and associates, in their study of 149 cases of complete transposition, observed left ventricular outflow obstruction in 23. This was caused, solely or in combinations, by pulmonary valvular stenosis, subvalvular fibromuscular or fibrous stenosis and bulging of the ventricular septum into the left ventricular cavity. Elliott and associates, Layman and Edwards and Rosenquist and associates showed that in complete transposition anomalous attachment of the anterior leaflet of the mitral valve to the ventricular septum could also result in obstruction to pulmonary flow. Rastelli and associates, describing their experience with 23 cases of complete transposition with pulmonary stenosis, noted three cases showing redundant tissue in the left ventricular outflow tract, which tissue contributed to obstruction of pulmonary flow. Recent angiographic studies by Silove and Taylor and echocardiographic studies by Nanda and associates have stressed abnormal septal and mitral apposition as causes of subpulmonary stenosis. The present study reveals, as previously described by Riems Schneider and associates, that in addition to previously described causes, tricuspid valvular pouches protruding through a VSD into the left ventricular cavity may form the basis of subpulmonary stenosis in some cases of complete transposition.

Our material emphasizes that in classical complete transposition pulmonary or subpulmonary stenosis may be caused by a combination of associated conditions. Of the 22 cases of classical complete transposition with pulmonary or subpulmonary stenosis, in seven (32%) there was more than one basis for the obstruction.
Six of our cases of complete transposition varied from the classical state in that in each case a conus was present in each ventricle. This is in contrast to the state in classical examples of complete transposition, in which there is absence of fibrous continuity between the pulmonary and mitral valves. Instead, in the six atypical cases a muscular segment lay interposed between these two valves.

Certain practical points are to be considered in relation to this variation. In the first instance, echocardiographically demonstrated lack of fibrous continuity between the semilunar valves, on one hand, and the atrioventricular valves, on the other, is one of the features of double outlet right ventricle. Recognition that such an anatomic process may occur in atypical complete transposition, as described by Van Praagh and associates, by Goor and Edwards, by Angelini and Leachman, and by Jimenez and Martinez, and as demonstrated by this study, broadens the differential diagnosis of the echocardiographic demonstration of lack of continuity between the semilunar and atrioventricular valves.

The two semilunar valves lying at about the same plane are a factor which occurs both in double outlet right ventricle and in complete transposition with left ventricular conus. This anatomic feature must be evaluated against the two conditions named in review of angiocardiograms.

It is recognized that among our six cases with a left ventricular conus, there were two patterns relative to the nature of the left ventricular conus. In one (Group II) this did not form a specific subdivision of the left ventricle, while in the other (Group I), the conus was separated from the left ventricle by a distinct inlet. In the latter, this inlet was stenotic. This pattern is to be included among those of complete transposition with pulmonary stenosis and poses challenges peculiar to its anatomic state in planned correction of obstruction to the pulmonary arterial pathway.

References
Anatomic causes of pulmonary stenosis in complete transposition.
S Shrivastava, S M Tadavarthy, T Fukuda and J E Edwards

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