The Spectrum of Transposition of the Great Arteries
With Specific Reference to Developmental Anatomy of the Conus

By DANIEL A. GOOR, M.D., AND JESSE E. EDWARDS, M.D.

SUMMARY
One hundred and sixty five cases of various types of transposition complexes were studied. Based on this and on recent studies of the embryology of the conotruncus, the following observations were made. Regardless of their relations with the ventricles, the semilunar valves may show d-transposition or l-transposition. In the situs solitus heart, the d-type of semilunar transposition is found in the bulboventricular heart, double outlet right ventricle type II, Taussig-Bing malformation, complete transposition of the great arteries, and double outlet left ventricle. This type of transposition of the great arteries results from lack of conotruncal inversion and the differences between the various entities are manifestations of varying degrees of leftward shift of the conoventricular junction and conal absorption. In l-transposition, the conotruncus rotates in the wrong direction. This may occur as an isolated process as in anatomic corrected transposition of the great arteries or with l-looping as in physiologic corrected transposition. Conal absorption and conoventricular shift account for the various entities in each of these two groups of l-transposition. It is proposed that the term transposition of the great arteries should refer only to the mutual interrelationships of the semilunar valves regardless of their ventricular origin.

Additional Indexing Words:
Double outlet right ventricle  Double outlet left ventricle  Bulboventricular heart
Embryology of the heart  Taussig-Bing malformation  Conal embryology and pathology
Origin of both great vessels from the right ventricle

LITERALLY, the composite term “transposition” means “crossed position.” With reference to the great arteries it means that the great arteries reverse their interrelationships. According to most investigators the term “transposition of the great arteries” is linked to two anatomic requirements; the great arteries should reverse their mutual interrelationships and, in addition, they should also reverse their interrelationships with the ventricles. The purpose of this report is to describe the spectrum of transposition complexes and to propose a modification in the interpretation of the term “transposition.” Accordingly, the term “transposition” is restricted only to the mutual interrelationships of the great arteries (or semilunar valves) regardless of their ventricular origin. The embryologic basis of this approach is presented.

Material and Methods
The embryologic information in this report is based on a study of human embryonic hearts carried out in the Department of Embryology of the Carnegie Institution of Washington. Details have been published elsewhere.1, 2
The pathologic material in our study includes the following conditions: (1) Complete transposition of the great arteries (92 specimens). In two of these cases, the transposed aortic valve displayed fibrous continuity with the tricuspid valve. In a third case, a fairly long conus was interposed between the transposed pulmonary valve and the mitral valve. There was no ventricular septal defect in this case. (2) Double outlet right ventricle (26 cases). Six of these were considered typical examples of the Taussig-Bing malformation. In a seventh case, the general anatomy was that of a Taussig-Bing malformation but the aortic valve displayed fibrous continuity with the tricuspid valve. (3) Corrected transposition of the great arteries (40 specimens). (4) Transposition with posterior aorta6 (six specimens). (5) Double outlet right ventricle with l-transposition and d-loop (one case). This case is also to be published in a separate report. The majority of these
specimens were drawn from the files of the Cardiovascular Division of the Miller Division of United Hospitals, formerly Department of Pathology of the Charles T. Miller Hospital. Some of the specimens were studied in the collection of the New York Hospital-Cornell University Medical College.

To present the various types of development encountered in our cases, we have selected specimens from eight cases which we will describe in detail.

**Definition of Terms**

The term "conus," according to Van Praagh and Van Praagh,4 applies to the muscular cardiac segment intervening between the semilunar and atriocentrical valves.

The term "transposition of the great vessels" (TGA) is a nonspecific term which implies that each of the semilunar valves is displaced relative to its own normal position.5 The term transposition, as used here, does not apply to the relationships between the semilunar valves and the ventricles. Hence, the great arteries may be transposed and still arise from either of the ventricles or from both sides of the septum. Each variety of TGA is further signified by a specific title which is defined later in the text. It is our opinion that the ventricles and the great arteries assume their "side" position as a result of rotational processes. Hence, "situs inversus" of the heart as well as its related pathologic conditions presents mirror image situations of situs solitus cases with their related pathologic conditions. In this text only situs solitus conditions are discussed.

The term "d-transposition" means that the aorta is to the right of the pulmonary artery6 (relative to the ventricular septum), while the term "l-transposition" implies that the aorta is to the left of the pulmonary artery. d-loop means that the right ventricle is in its appropriate right side and l-loop indicates that the anatomic right ventricle is on the left side, being inverted.6

The term "double outlet right ventricle"7 indicates that one of the two great arteries (or their conuses) arises exclusively from the right ventricle and the other great artery (or its conus) arises either partially or exclusively from the right ventricle.

**The Embryology of the Conotruncus**

The definitive shape of the outflow tracts of the ventricles is primarily due to three developmental processes:2 conotruncal (bulbar) inversion, leftward shift of the conoventricular junction, and absorption of the conus.

**Conotruncal Inversion**

In the primitive cardiac tube the predecessor of the aortic pathway is on the right side and in line with the right half of the right ventricle. The pulmonary pathway is on the left and in line with the left half of the right ventricle. The inversion of the arterial pathways, or conotrunci, so that the aorta will become relatively left-sided while the pulmonary artery becomes relatively right-sided, is a process that occurs in two stages. The first one takes place during the d-looping, and it is described as rotation of the conoventricular junction of 110° in the counterclockwise direction8,9 (that is, looking downstream). Consequently, the proximal parts of the coni reverse their relationships so that the aortic conus is transferred to the back and the left side of the right ventricle. In the second stage, which occurs later, the truncus (semilunar valves) rotates in the same manner as the proximal parts of the coni did,2,9 thus completing the inversion of the conotruncus. As a result of the truncal rotation, the spiral course of the conotruncus is reduced, while the great arteries become entwined.

**Leftward Shift of the Conoventricular Junction**8,9-11

In this process, which occurs during horizon XVI, the conus which is on the left side (normally the aortic conus) moves leftward to spatially override the left ventricle. Consequently, flow concordance is achieved between the ventricles and the proximal coni. During that shift some reverse rotation of the ostium bulbi occurs.9,11

**Absorption of the Conus**

Absorption reduces the length of the aortic conus from about 400 μm in horizon XV to 20 μm in horizon XIX.2 Consequently, in the definitive heart there is an aorticomitral fibrous continuity.4,12,13 The proximal half of the pulmonary conus is also absorbed and its length is reduced from about 400 microns to about 200 microns. As a result of the absorption of the proximal conus (underneath the aorta and the pulmonary artery) the distal conus with its septum is conveyed toward the ventricles.2 In the definitive heart the conal septum appears as the subpulmonary portion of the ventricular septum.

**Description of Specimens**

In situs solitus, there are two spectra of transpositions of the semilunar valves, namely, d and l. In each of these two lines, the various entities differ by the spatial position of the transposed semilunar valves (and their conuses) and by the length of the aortic and/or pulmonary conuses.

**Spectrum of d-Transpositions**

Case 1. Bulboventricular Heart,14 Type II: In this most primitive form of double outlet right ventricle the semilunar valves display d-transposition. There is in toto persistent bilateral conus and both conuses arise exclusively from the right ventricle.

_Circulation, Volume XLVIII, August 1973_
Case 2. True Double Outlet Right Ventricle, Type II (Double Outlet Right Ventricle with d-Transposition) (fig. 1a): As in the previous case, in this one the semilunar valves display d-transposition. Usually these valves are interrelated side-to-side, but oblique relationships, namely aorta in front and to the right of the pulmonary artery, were recently recorded.\(^\text{15}^{,}\text{16}\) The proximal ends of the aortic and pulmonary pathways arise exclusively from the right ventricle. The aortic conus is continuous with the tricuspid valve and the base of the right ventricle. The pulmonary conus arises in front of, and to the left of, the aortic conus and exclusively from the right ventricle. The bilateral conus is short, being approximately one-third of the length of the right ventricle, and the distal conus septum is close to the base of the ventricular septum.

Case 3. Double Outlet Right Ventricle with Mild Levoposition of the Pulmonary Conus (fig. 1b): The situation in this case differs from that in Case 2 in the relationship between the pulmonary conus and the left ventricle. This relationship is displayed by anatomic continuity between the pulmonary conus and the mitral valve, and by the spatial overriding of the left ventricle by the pulmonary conus (PC) is exclusively from the right ventricle. The bilaterial conus is short, being approximately one-third of the length of the right ventricle, and the distal conus septum is close to the base of the ventricular septum.

Figure 1

Right views of various types of double outlet right ventricle with d-transposition (type II). Lack of ostium bulbar and truncal incision is displayed by the insertion of conus ridge 1 (the septal end of the conal septum) into the back wall of the conus, interrupting the continuity between the aortic conus (AC) and the left ventricle. The feature is common to all varieties of noninverted conotruncus. A varying degree of levoposition of the pulmonary conus (PC), however, accounts for the differences in the three specimens. In the photographs, conus ridge 1 is represented by the dorsal half of the conal septum (CS). The anterior half of the septum (conus ridge 3) has been removed, together with the anterior wall of the infundibulum. In the diagrammatic illustrations (lower row) both conus ridges are shown. (a) A "true" double outlet right ventricle, type II, in which both coni arise exclusively from the right ventricle. (b) A mild degree of pulmonary levoposition is apparent. M = anterior mitral leaflet; T = anterior tricuspid leaflet. (c) Taussig-Bing malformation. The pulmonary conus (PC) is in levoposition presumably as a result of complete leftward shift of the conoventricular flange. In all three varieties, the pulmonary and the aortic coni are of equal length so that the semilunar valves are at the same level (PA and Ao).
conus. The ostium which allows communication between the pulmonary conus and the left ventricle is termed “pulmonarycono-left ventricular ostium.” This ostium is more commonly known by the inaccurate term, “subpulmonary ventricular septal defect” (VSD). The aortic conus and the valve show the same anatomy as in Case 2 (fig. 1a).

Case 4. Taussig-Bing Malformation (figs. 1c, 2a, c): The aortic valve and conus show the same structure as in the preceding two cases. The pulmonary conus, however, in this case is pulled farther to the left and its posterior wall grossly arises from the anterior mitral leaflet. As in Case 3, the pulmonary conus communicates with the left ventricle via the pulmonarycono-left ventricular ostium (subpulmonary VSD) and with the right ventricle via the pulmonarycono-right ventricular junction or ostium. The pulmonary and aortic coni, as in Case 3, are approximately but not necessarily the same length and both semilunar valves are usually at the same level. In two of the three cases in which the lungs were present and the semilunar interrelationships could be accurately determined, the aorta was to the right and in front of the pul-

![Figure 2](image)

Transitional patterns between true double outlet right ventricle and complete transposition of great arteries. (a) Right view. The presence of a rudimentary pulmonary conus and the minimal rightward displacement of the conal septum differentiate this specimen from complete TGA. (b) Left view of complete TGA with biventricular pulmonary trunk. Pulmonary conus is absorbed but obvious deviation of the anterior end of the conal septum (CS) toward the right is displayed. Compare with (d) (c). Left view of Taussig-Bing malformation (same specimen as figure 1c). Supramitral pulmonary conus (PC) lies between the pulmonary and mitral valves. (d) Left view of complete TGA with VSD. Despite lack of fusion of conal and ventricular septa, both are perfectly in line. Ao = aorta; CS = conal septum; PA = pulmonary artery; PC = pulmonary conus; M = anterior mitral leaflet.
monary artery, and in one case it was in front of the pulmonary artery. Others have recorded similar oblique and anteroposterior semilunar interrelationships in Taussig-Bing malformation.\textsuperscript{17, 18} Since the subpulmonary outflow tract (conus) is not recognized by some investigators as a developmental segment but rather as a part of the right ventricle, the condition is commonly known as a variation of double outlet right ventricle.\textsuperscript{9}

\textbf{Case 5. Complete Transposition of the Great Vessels with Biventricular Pulmonary Trunk}\textsuperscript{19} (fig. 2b): In this case all the features of complete TGA are present except that the conal septum and the ventricular septum are not in line. As in complete TGA, the pulmonary artery is in fibrous continuity with the anterior mitral leaflet. The anterior end of the conal septum, however, is deviated toward the right, an orientation characteristic of Cases 2, 3 and 4. Consequently, the pulmonary trunk has a biventricular origin. In Taussig-Bing malformation (Case 4) the pulmonary artery arises exclusively from a conus (fig. 2c), the posterior wall of which in turn arises from the mitral valve. In a similar case that was recorded previously,\textsuperscript{19} a hemodynamic similarity to Taussig-Bing malformation was observed.

\textbf{Case 6. Complete Transposition of Great Arteries with Ventricular Septal Defect} (fig. 2d): In this case all the features of complete transposition are present: (1) there is a d-transposition of the semilunar valves; (2) each artery arises from the wrong ventricle; (3) the ventricular and conal septa are in line; and (4) pulmonary-mitral fibrous continuity is present. The VSD is a junctional defect along the fusion line of the conal and ventricular septa. Both septa, however, are perfectly in line.

\textbf{Case 7. d-Transposition of the Great Arteries with Posterior Aorta:} This entity was recently defined by Van Praagh et al.\textsuperscript{8} As in other varieties of d-transposition, the semilunar valves display d-type of transposition. The aortic conus, however, straddles the ventricular septum, being in continuity with both atrioventricular valves (fig. 3a). The conotruncal septum is characteristically in transverse orientation. The septal end of the conus septum (conus ridge 1) joins either the left wall of the left ventricular outflow tract or the supramitral parietal wall of the conus. The latter occurred in one of our six specimens of TGA with posterior aorta. In two of our six cases, no conal septum was present, the transverse ridge being the truncal septum.

\textbf{Spectrum of l-Transpositions}

\textbf{Case 8. Double Outlet Right Ventricle with l-Transposition, and d-Loop\textsuperscript{20, 21} (DORV Type III)} (fig. 3b): As in true double outlet right ventricle, both coni arise in this situs solitus specimen exclusively from the right-sided right ventricle. The difference, however, between this and the ordinary DORV type II is that the aorta is anterior to and to the left of the pulmonary artery, a situation which is seen in corrected transposition and which is defined as l-transposition. The pulmonary (rather than the aortic) conus is continuous with the tricuspid valve (fig. 3b). A similar case in which the pulmonary conus was absorbed and pulmonary-tricuspid fibrous continuity was present was reported by Shafer and associates.\textsuperscript{22}

\textbf{Discussion}

In the normal heart the definitive shaping of the ventricular outflow tracts depends upon complete conotruncal inversion in the counterclockwise direction, leftward shift of the conoventricular junction, total absorption of the aortic conus and proximal absorption of the pulmonary conus. In transposition complexes there is invariably faulty inversion of the conotruncus. In addition, greater or lesser degrees of faulty development in absorption of the conus and in the leftward shift of the conoventricular junction account for the variability of transposition complexes.

The common denominator of Cases 1–7 is d-transposition of the semilunar valves in situs solitus. Except for Cases 1 and 8 which are rare, the typical side-to-side, oblique and anteroposterior intersemilunar relationships which are characteristic for complete TGA were observed in each of the malformations in that spectrum.\textsuperscript{18–19} In addition, in each of the varieties of conditions in Cases 1–7, the conus septum is also inverted (developmentally non-inverted). One arrives at the conclusion that the conus septum is pathologically inverted because the septal end of the conal septum (conal ridge 1), which is normally anterior, is invariably inserted into the posterior parietal wall of the conus above the junction of the mitral and tricuspid valves. In the normal heart it is the parietal end of the conal septum (conal ridge 3) which is inserted at this site. Another relatively consistent feature in d-transposition (in situs solitus) is the characteristic location of the aortic valve with reference to the tricuspid valve and the persistence of the supratricuspid aortic conus. Rarely, however, fibrous continuity of the aorta and tricuspid valve may be
encountered (three specimens in this series). The location of the aortic valve and the aortic conus on the relative right side and the inversion of the distal conus septum indicate that in the spectrum of d-transposition (in situs solitus) conotruncal inversion failed to occur. The anatomic as well as the developmental variables in this spectrum of d-transposition are the length of the conus and the spatial position of the pulmonary conus and/or valve. Accordingly, in the bulboventricular heart the conus is as long as the right ventricle and it exclusively arises from the right ventricle. In DORV type II, the conus is short (proximal conus bilaterally absorbed, distal conus bilaterally persists) and it arises exclusively from the right ventricle. The term “true” DORV type II is used in cases in which the d-transposed semilunar valves (and conuses) exclusively arise from the right ventricle.

Cases 3 and 4 display the same length of bilateral conus as Case 2 (persistence of only distal bilateral conus), but the pulmonary coni respectively show moderate and significant degrees of levoposition. This levoposition is due to a leftward shift of the pulmonary conoventricular junction.

Case 4 is a typical example of Taussig-Bing malformation, whereas Case 3 is a transitional situation between a “true” DORV type II and DORV type II with pulmonary levoposition (Taussig-Bing malformation).

There is some controversy about the spatial position of the pulmonary valve in Taussig-Bing malformation. This subject was recently studied by Hinkes, Rosenquist and White who observed in the original specimen described by Taussig and Bing that the pulmonary valve spatially overrides the left ventricle. A more important anatomic landmark for the leftward shift of the conoventricular junction is the presence of continuity between the pulmonary conus and the mitral valve.

Figure 3

(a) Left view of TGA with posterior aorta. The drawing at the bottom illustrates the right view of the muscular skeleton of the anomaly. The conus septum (CS) shows a transverse orientation. Arrow leads into aortic pathway. (b) Double outlet right ventricle type III (DORV with l-transposition and d-loop). This situation is identical to “true” double outlet right ventricle type II, except for the reverse interrelationships of the great arteries (aorta in the front and to the left). CS = conus septum; AC = aortic conus; PC = pulmonary conus; M = anterior mitral leaflet; PA = pulmonary artery.
The transition from Case 4 to Case 5 is due to absorption of the pulmonary conus. This is displayed by the fibrous continuity between the pulmonary and mitral valves. As a result of the absence of the pulmonary conus, the pulmonary valve is found in a lower level than the aortic valve. In this particular case, however, the conal septum is not fully aligned with the ventricular septum; this is a retained feature of double outlet right ventricle. This condition was defined in the past as complete TGA with biventricular pulmonary trunk and it represents a stage in the transition from Taussig-Bing malformation to complete TGA.

In Case 6 (fig. 2d) the features are the same as in Case 5 except that the conal and ventricular septa, although not fused, are nevertheless perfectly aligned in the same plane. If the ventricular septal defect closes, complete and perfect d-TGA is present. Hence the line of production of complete d-TGA starts in the primitive formation of the bulboventricular heart (table 1). Absorption of the proximal conus transforms this malformation to "true" DORV type II. Leftward shift of the conoventricular junction transforms "true" DORV type II to DORV type II of the Taussig-Bing variety. Absorption of the pulmonary conus transforms Taussig-Bing malformation to complete TGA.

The rare conditions, TGA with posterior aorta and double outlet left ventricle, also display the basic d-type of semilunar interrelationships described by Elliott et al. in complete transposition. The transverse orientation of the conotruncal septum in these two conditions, however, indicates that some degree of conotruncal inversion does occur but this inversion is incomplete. As postulated by Paul et al., the origin of the d-transposed great arteries from the left ventricle is due to overshift of the conoventricular junction to the left.

Strong support for the theory of lack of conotruncal inversion in d-transposition of TGA is found in Shaner's description of an 18 mm pig embryo, where noninverted coni and straight noninverted conotruncal ridges present typical features of DORV with d-transposition.

In l-transposition in the situs solitus condition, the aortic valve, instead of being transferred behind to the left of the pulmonary artery, is in front and to the left of the pulmonary artery. In other words, in this group conotruncal inversion occurs in the wrong direction (clockwise instead of counterclockwise). This theory was first postulated by Lochte in relation to physiologic corrected TGA (l-TGA with l-loop) and later confirmed in embryos by Lewis and Abbott and Shaner.

In situs solitus, there are two subspectra of l-transposition (table 1): (A) l-transposition with d-loop. This subspectrum includes two reported varieties: (1) Double outlet right ventricle with l-transposition and d-loop (Case 8). In this malformation, the semilunar valves display l-type of TGA both, however, arising from the anatomical right ventricle which is located in its normal right side. (2) Isolated bulbar inversion (anatomically corrected transposition). In this malformation, as in the previous one, the semilunar valves display l-type transposition and the ventricles are in their appropriate sides (d-loop). The aortic valve, however, arises from the left ventricle and the pulmonary valve arises from the right ventricle. From a developmental viewpoint, both conditions of subspectrum A belong in the same category, displaying isolated inversion of the conotruncus. The difference between the two is that in DORV with l-TGA and d-loop (type III), leftward shift of

---

Table 1

<table>
<thead>
<tr>
<th>Developmental Classification of the Various Types of Transpositions of the Great Arteries</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
</tr>
<tr>
<td><strong>Table 1</strong></td>
</tr>
<tr>
<td><strong>Anatomic types of transposition of the great arteries (situs solitus)</strong></td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td>d-transposition with d-loop (conotruncus fails to invert)</td>
</tr>
<tr>
<td>l-transposition with d-loop (isolated bulbar inversion)</td>
</tr>
<tr>
<td>l-transposition with l-loop (physiologic corrected transposition)</td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td><strong>Lack of conoventricular shift</strong></td>
</tr>
<tr>
<td>1. Bulboventricular heart</td>
</tr>
<tr>
<td>2. Double outlet right ventricle with d-transposition (type II)</td>
</tr>
<tr>
<td>3. Taussig-Bing malformation</td>
</tr>
<tr>
<td>4. Complete TGA</td>
</tr>
<tr>
<td>5. Double outlet left ventricle</td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td><strong>Correct degree of conoventricular shift</strong></td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td></td>
</tr>
<tr>
<td><strong>Over shift</strong></td>
</tr>
<tr>
<td></td>
</tr>
</tbody>
</table>
the conoventricular junction does not occur, while in anatomically corrected transposition this shift takes a full span. That conus absorption is not a factor in the pathogenesis of this group of malformations is evidenced in Shafer et al.'s case\textsuperscript{22} where pulmonary tricuspid fibrous continuity was present. (B) \textit{l}-transposition with \textit{l}-loop. This group of malformations is commonly known as (physiologically) corrected transposition. This subspectrum includes the three following varieties: (1) Double outlet (left-sided) right ventricle in corrected transposition.\textsuperscript{22-24} In this group, the semilunar valves display \textit{l}-type of transposition with both arising from a left-sided anatomic right ventricle. The situs of the atria is normal. (2) Physiologic corrected transposition with biventricular origin of the pulmonary valve (and conus).\textsuperscript{33} This group is similar to the preceding one except for the biventricular origin of the pulmonary valve. (3) Classical physiologic corrected TGA. This group is similar to the preceding two varieties except for the origin of the pulmonary artery (and conus) from the right-sided left ventricle. Pulmonary and/or aortic conuses may or may not be present in each of these three varieties. Developmentally, hence, the common denominator of subspectrum B is the counternormal inversion of the conotruncus as well as the counternormal inversion of the ventricles (\textit{l}-looping instead of \textit{d}-looping in situs solitus). The variable anatomic (and embryologic) factor is the rightward shift of the conoventricular junction. In the first variety (DORV with R-TGA and \textit{l}-loop), no shift occurs. In the second variety moderate shift results in biventricular origin of the pulmonary trunk and in the third variety (\textit{l}-TGA, \textit{l}-loop) there is a correct degree of conoventricular shift. The shift in subspectrum B is to the right (instead of left) because of the reverse direction of the \textit{l}-looping.

Other rare conditions of physiologic transposition in situs solitus, such as isolated atrial inversion\textsuperscript{35} and isolated ventricular inversion\textsuperscript{4, 36, 37} may be explained on the basis of segmental rotation, conoventricular shift and conal absorption.

The segmental and spectral nature of the pathology as well as embryology of transposition complexes require that the terminology used for description of these entities will be segmental. It is therefore suggested that the terms \textit{d}- and \textit{l}-transposition will merely indicate the nature of the mutual intersemilunar (intertruncal) relationships. Additional terms are required for specifying the semilunar-ventricular relationships. Since, however, the same conditions may appear in situs solitus and in situs inversus, nondescriptive terms such as Taussig-Bing malformation, DORV types I-III, etc. cetera, are very useful, particularly once the anatomical and developmental aspects of the malformation are understood.

While the basic theory of the development of \textit{l}-transposition is shared by most investigators,\textsuperscript{6, 27, 28, 38} opinion about \textit{d}-transposition differs. The main difference between the theory which we have presented and other theories is our multifactorial approach as opposed to the single factorial approach of most investigators. According to Shaner\textsuperscript{27} and de la Cruz and associates,\textsuperscript{38} the conal septum rotates as it grows into the ventricles, thus accounting for the spiral shape of the ventricular outflow tracts. In transposition complexes, according to this theory, the conal septum rotates as it grows into the ventricles in a disoriented fashion.\textsuperscript{27, 38} We believe that the investigators favoring this hypothesis interpret the migration of the septum toward the ventricles as growth. It is true that the conal septum migrates into the ventricles along the proximal ridges and therefore it does indeed undergo a slight degree of rotation. We believe, however, that the lines along which the conal septum moves into the ventricles are delineated before the septum ever shows. In other words, transposition is determined prior to the growth of the conal septum.

According to Pernkopf and Wirtinger\textsuperscript{9} TGA results from lack of conotruncal inversion. As shown herein, this mechanism is indeed the basis of the spectrum of \textit{d}-transposition.

According to Van Praagh and associates,\textsuperscript{3} \textit{d}-TGA is due to faulty conotruncal inversion. In their opinion the aortic conus assumes the primitive right position only during \textit{d}-looping, and conotruncal inversion is a result of later growth of the pulmonary conus. Our embryologic studies as well as those of others,\textsuperscript{2, 9, 11} however, indicate that during the \textit{d}-looping the aortic conus is transferred behind the pulmonary conus to the left. Our studies also, we believe, indicate that the presence or absence of a conus is an accompanying rather than a causative factor.

This statement is based on the existence of such cases as complete (\textit{d})-TGA with aortictricuspid fibrous continuity, complete (\textit{d})-TGA with persistent pulmonary conus, Taussig-Bing-like malformation with aortic tricuspid fibrous continuity, DORV type III with pulmonary tricuspid fibrous continuity.
and other varieties of transposition complexes where a conus may or may not be present.

Keith’s and Lev and Saphir’s theories concerning the role of absorption of the aortic conus may explain most situations of TGA but the occasional presence of bilateral coni as well as the existence of absorbed bilateral coni in TGA indicates that the conal absorption is only one factor in the complex development of this anomaly. Van Mierop is of the opinion that truncal ridges 2 and 4 fuse as a septum (instead of truncal ridges 1 and 3). To him, this septum is continuous with a normal conal septum. As described in this report, however, the conal part of the septum is as inverted as the truncal septum.

References
12. KEITH A: The Hunterian lectures on malformations of the heart: Lectures I, II, and III. Lancet 2: 359, 433, 519, 1909
22. SHAFER AB, LOPEZ JF, KLINE IK, LEV M: Truncal inversion with bicuspid pulmonary trunk and aorta from right ventricle (variant of Taussig-Bing). Circulation 36: 783, 1967
23. HINKES P, ROSENQUIST GC, WHITE RI Jr: Roentgenographic reexamination of the internal anatomy of the Taugss-Bing heart. Am Heart J 81: 335, 1971

Circulation, Volume XLVIII, August 1973
27. SHANER RF: Complete and corrected transposition of the aorta, pulmonary artery and ventricles in pig embryos, and a case of corrected transposition in a child. Am J Anat 88: 35, 1951
32. FRAGOYANNIS S, KARDALINOS A: Transposition of the great vessels, both arising from the left ventricle (juxtaposition of pulmonary artery): Tricuspid atresia, atrial septal defect and ventricular septal defect. Am J Cardiol 10: 601, 1962
35. CLARKSON PM, BRANDT PWT, BARBATT-BOYES BG, NEUTZE JM: "Isolated atrial inversion": Visceral situs solitus, visceroatrial discordance, discordant ventricular d loop without transposition, dextrocardia: Diagnosis and surgical correction. Am J Cardiol 29: 877, 1972
The Spectrum of Transposition of the Great Arteries: With Specific Reference to Developmental Anatomy of the Conus

DANIEL A. GOOR and JESSE E. EDWARDS

Circulation. 1973;48:406-415
doi: 10.1161/01.CIR.48.2.406

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1973 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/48/2/406

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation is online at:
http://circ.ahajournals.org//subscriptions/