The Differential Diagnosis of Levo-Transposed or Malposed Aorta

An Angiocardiographic Study

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SUMMARY

The levo-transposed aorta has previously been considered diagnostic of corrected transposition of the great arteries and hence ventricular inversion in situs solitus. Increasing experience with conotruncal abnormalities has shown that an abnormally leftward aorta may be found in patients with normally related ventricles (D-ventricular loop) and hemodynamically complete transposition of the great arteries; anatomically corrected L-malposition of the great arteries; and rarely in patients with D-ventricular loop, double outlet right and left ventricles. Thus the levo-positioned aorta should suggest a wider spectrum of cardiovascular abnormalities than previously thought. The specific angiocardiographic features of levo-transposition, anatomically corrected malpositions, and D-ventricular loop with levo-transposed aorta are presented in detail; and their differential diagnosis from the very uncommon D-loop, double outlet right and left ventricles with an L-malposed aorta is considered. The role of selective biplane angiocardiography in the assessment of these conotruncal abnormalities is stressed.

Additional Indexing Words:
Anatomically corrected malpositions
Corrected transposition
Double outlet left ventricle

Levo-transposed Aorta has previously been considered diagnostic of corrected (levo-transposition, L-TGA) transposition of the great arteries.1-3

However, increasing experience with conotruncal abnormalities has shown that an abnormally leftward aorta may be found in patients with normally related ventricles (D-ventricular loop) and hemodynamically complete transposition of the great arteries.4-6

Similarly, the angiographic and anatomic features of the levo-transposed (malposed) aorta in anatomically corrected malpositions have been presented elsewhere.6,7 The uncommon finding of a double outlet right ventricle, D-ventricular loop, L-transposed aorta, bilateral conus, and pulmonic stenosis has also been documented.8 This patient subsequently underwent surgical repair. And finally, we have been able to find one patient recorded in the literature with a D-ventricular loop, double outlet left ventricle, probable valvular and subvalvular pulmonic stenosis, and L-malposition of the aorta.9

Obviously then, the levo-transposed or malposed aorta embraces a wider range of cardiovascular abnormalities than previously thought. Because of increasing surgical expertise in dealing with uncommon conotruncal malformations, and the obvious relevance of precise anatomic diagnosis, we wish to present the differential angiocardiographic diagnosis of the levo-transposed and malposed aorta and to comment on the associated cardiovascular lesions.

Definitions
1. D-ventricular loop (D-loop): The primitive cardiac loop
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has looped to the right, resulting in a right-sided morphologic right ventricle, and left-sided morphologic left ventricle. This is the normal ventricular loop in situs solitus (abdominal and thoracic viscera in normal position). L-ventricular loop (L-loop): The primitive cardiac tube has looped to the left, resulting in a left-sided morphologic right ventricle, and right-sided morphologic left ventricle.

3. Transposition: Transposition will be defined according to the criterion of Van Praagh: The aorta originates above the morphologic right ventricle and the pulmonary artery originates above the morphologic left ventricle. Implicit to this definition are a ventricular septum and two ventricles.

4. D-transposition: This implies a D-ventricular loop. The transposed aorta originates above the right-sided morphologic right ventricle and pulmonary artery above the morphologic left ventricle. The aorta is to the right of the pulmonary artery. In situs solitus, this is simple, complete transposition.

5. L-transposition (corrected transposition): There is an L-ventricular loop. The aorta is transposed, originating above the inverted, morphologic right ventricle, and the pulmonary artery originates above the morphologic left ventricle. The aorta is anterior and to the left of the transposed pulmonary artery.

6. Malposition of the great arteries: An abnormal relationship between the great arteries. A positional anomaly of the great arteries, other than transposition. When the malpositioned aorta is to the right of the pulmonary artery, this is D-malposition; when to the left, L-malposition.

Levo-Transposition of the Great Arteries (L-transposition, corrected transposition, fig. 1)

In its classic form this implies:
1. L-ventricular loop
2. L-transposed aorta
3. Subaortic conus only, preventing aortic-left atrioventricular (tricuspid) valve fibrous continuity.

Extensive reviews of the angiographic features of L-transposition have appeared elsewhere. There are two ventricles in the L-loop position (morphologic right ventricle is left-sided), a subaortic conus and no subpulmonic conus, and an anterior, levo-transposed aorta with aortic-left atrioventricular valve fibrous discontinuity.

It must be mentioned that commonly associated anatomic lesions include atrial and/or ventricular septal defects; single left ventricle with an outlet chamber with or without pulmonic stenosis; left atrioventricular valve regurgitation, usually because of an Ebstein’s malformation of the left-sided tricuspid valve; ventricular septal defect and pulmonic stenosis; and dextrocardia. In the patient with visceral heterotaxia, levo-transposition may have an associated endocardial cushion defect and abnormal connections of systemic and pulmonary veins. Origin of both great arteries from the left-sided, morphologic right ventricle has also been seen. Rarely, pulmonary atresia and an intact ventricular septum may occur with levo-transposition of the great arteries.

Anatomically Corrected L-Malposition of the Great Arteries (fig. 2)

This implies:
1. D-ventricular loop
2. Bilateral (subaortic and subpulmonic) conus
3. Aortic-mitral fibrous discontinuity and pulmonary-tricuspid valve fibrous discontinuity

4. L-malposition of the aorta, with the aorta originating above morphologic left ventricle, and pulmonary artery above the morphologic right ventricle; the aorta is to the left of the pulmonary artery.

In our recent review of all known patients with anatomically corrected malpositions, we found an unusually high incidence with left juxtaposition of the atrial appendages (7 of 13); tricuspid valve atresia or hypoplasia (6 of 13); and hypoplasia of the morphologic right ventricle (8 of 13). All 13 patients had a ventricular septal defect; 9 had valvar and subvalvar pulmonic obstruction, and three had subaortic obstruction of varying severity. Dextrocardia was present in 7 of 13. Twelve of 13 patients had bilateral conal myocardium which separates semilunar valve from A-V valve. The normal individual has only subpulmonic conus and aortic-mitral continuity.

It should be emphasized, however, that some patients with anatomically corrected malpositions have a normal tricuspid valve and a right ventricle of normal size.

It is also worth noting that anatomically corrected L-malposition of the great arteries with pulmonary-mitral fibrous continuity has been described.

D-Ventricular Loop with L-Transposition of the Great Arteries (fig. 3)

This implies:
1. D-ventricular loop
2. Subaortic conus
3. Oblique relationship between the great arteries, with aorta anterior and to the left of the pulmonary artery. The aorta originates above the right-sided, morphologic right ventricle, and the pulmonary artery above the morphologic left ventricle.

In the two patients studied at our institution with D-loop, L-TGA, one had an infracristal VSD and pulmonic stenosis and the other had an infracristal VSD, but no pulmonary obstruction.

Discussion

The levo-transposed or malposed aorta as now seen includes a rather wide variety of congenital cardiac defects. Levo- or corrected transposition has a distinct clinical, electrovectorcardiographic, and angiocardiographic profile. In this entity, the effect of transposition of the great arteries has been “corrected” by inversion of the ventricles. Selective right-sided and left-sided ventriculograms will delineate the precise ventricular anatomy. Indeed, this is essential in the complete study of the complex conotruncal malformations characterized by L-transposition or malposition. In the “pure” L-transposition, the aortic valve is anterior and to the left of the pulmonic valve. However, in many patients with L-transposition, associated ventricular septal defect and pulmonic stenosis, there is bilateral conal myocardium, and the valve heights will be at the same level.

The transposition portion of Van Praagh’s loop rule has proved most helpful in ventricular localization. Briefly this rule states that since the transposed aorta originates exclusively from the right ventricular infun-
dibulum, the transposed aorta, D- or L- serves as a reliable angiocardiographic marker of the right or left-sided location of the right ventricle. As pointed out by Van Praagh, and as illustrated in the present review, anatomically corrected L-malpositions and D-loop, L-transpositions are exceptions to the loop rule.6, 10 Only selective ventriculography will properly sort out these discordant relationships.

It is unclear how frequently these loop rule exceptions occur. The study of Guerin has shown that in patients with D-ventricular loop and hemodynamically complete transposition of the great arteries, 13% (5 of 38) have a left-sided and anterior aorta.4 It would thus appear that this type of loop rule exception is not as uncommon as previously thought.

The majority of patients with D-loop, complete transpositions studied angiocardiographically at the Johns Hopkins Hospital are positioned in the left anterior oblique view in order that we might visualize the status of ventricular septum as well as the

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**Figure 1**

Levo-transposition of the great arteries (corrected transposition) in a 4-year-old boy. Top left: AP ventriculogram in left-sided ventricle. There is mesocardia. The morphologic right ventricle (RV) is left-sided, indicating an L-ventricular loop (inverted ventricles in situ solitus). The aorta (AO) originates above the morphologic RV and is thus transposed. The aorta is in the classic levo-transposition position. Top right: Lateral ventriculogram in left-sided ventricle. (Same frame as top left). The aorta (AO) originates anteriorly above the morphologic right ventricle (RV). Bottom left: AP ventriculogram in right-sided morphologic left ventricle (LV). The transposed pulmonary artery (PA) arises from this ventricle. The ventricular septum appears intact. Pulmonary valve thickening is also evident. The aorta (top left) is to the left of the pulmonary artery. Note that the ventricular septum in the L-ventricular loop is best visualized in the AP views. Bottom right: Lateral ventriculogram in right-sided ventricle. (Same frame as bottom left). The pulmonary artery is posterior to the aorta. Supravalvar narrowing is seen.
morphology of the ventricles and the relationship between the great arteries. Hence it is difficult for us to estimate the frequency of D-loop, L-transpositions in our material.

Anatomically corrected malpositions, specifically, D-ventricular loop with an L-malposed aorta are uncommon conotruncal-ventricular disturbances which have been confused with levo-transposition of the great arteries. In this malposition according to Van Praagh, the great arteries and ventricles have apparently twisted in opposite directions, resulting in a severe conoventricular malalignment, with a ventricular defect characteristically at the conoventricular junction. Bilateral conal myocardium was evident in 12 of the 13 patients with anatomically corrected malpositions reviewed by Freedom and Harrington. As illustrated in figure 2 top, the aortic valve is clearly to the left of the pulmonic valve in this malposition. If selective ventriculography had not been performed, the levo-position of the aorta would have been presumptive evidence for ventricular inversion. Yet selective ventriculography clearly demonstrated a normal ventricular loop (D-loop) with bilateral conal myocardium and semilunar valve-atrioventricular valve fibrous discontinuity.

Implicit in the recognition of anatomically corrected malpositions is awareness of the high association with left juxtaposition of the atrial appendages, tricuspid valve atresia or hypoplasia, hypoplasia of the right ventricle, subvalvar pulmonic and aortic obstruction because of poorly expanded conal myocardium, and dextrocardia. As mentioned earlier, some patients with anatomically corrected transpositions will have a normal tricuspid valve and right ventricular cavity, and that the associated intracardiac defects in these patients may be amenable to open heart surgery as recently reported by Sunada and Kirklin.

Double outlet right ventricle can occur with a levo-transposed aorta. Usually, though, there is associated ventricular inversion (L-loop) with both great arteries originating from the inverted, left-sided morphologic right ventricle. Recently, Lincoln has described a patient with a D-ventricular loop, double outlet right ventricle, bilateral conus, L-transposition (more properly L-malposition)
and pulmonic stenosis. The angiocardiograms (Lincoln, fig. 4a) demonstrate an L-malposed aorta and his figure 5b suggests that both great arteries originate above the anterior, morphologic right ventricle. This patient underwent successful operative correction of her defects and was apparently doing well six months postoperatively. Although D-loop, double outlet right ventricle with L-malposition of the aorta is uncommon, excellent angiocardiograms of this malposition have recently been presented (reference 22, fig. 7; reference 23, fig. 1). These cases suggest that adequate angiographic study requires selective ven-
triculography. That this type of double outlet right ventricle with an L-malposed aorta is rare is illustrated by the observation that Lev in his recent extensive review of double outlet right ventricle does not mention it.26

Double outlet left ventricle with origin of both great arteries from the left-sided, morphologic left ventricle is most uncommon and is usually confused with tetralogy of Fallot. Recently Pacifico et al. have reported the surgical correction of four patients with this type of malposition.9 One of their patients, a 5½-year-old (their case 4), had a D-ventricular loop, double outlet left ventricle with L-malposition of the aorta, a large anterior ventricular septal defect, and probable valvar and subvalvar pulmonic stenosis. The left ventriculogram in this patient (their fig. 2) nicely demonstrates the double outlet left ventricle with L-malposition, and reinforces the need for selective ventriculography.

In summary, a levo-transposed or malposed aorta should suggest a broader spectrum of congenital cardiac anomalies than previously thought. Specifically, a levo-transposed or malposed aorta may occur in levo-transpositions, anatomically corrected malpositions, D-loop, complete transpositions, and double outlet right and left ventricles. Each has associated intracardiac defects. The precise conotruncal-ventricular relationships can be determined by selective ventriculography.

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Figure 3
D-ventricular loop. L-transposition of the great arteries in a one-year-old girl. Top left: Selective ventriculogram in AP view. There is opacification of a morphologic RV. The aorta appears in the levo-transposed position and arches to the right. Top right: Lateral frame from selective ventriculogram. (Same frame as top left.) The catheter is in an anterior chamber, the morphologic RV. The transposed aorta originates above this chamber. The ventricular septum is well seen in this projection, indicative of a D-ventricular loop. The posterior ventricle is well-opacified. Bottom left: Selective ventriculogram in AP view. A morphologic LV is opacified. The relatively small pulmonary artery (PA) is noted to be to the right of the aorta (AO). The aorta appears to be levo-transposed. The pulmonic and aortic calvices appear to be at the same level. This projection erroneously suggests that both great arteries originate from the morphologic LV. Pulmonary calve thickening can be seen. Bottom right: Selective ventriculogram in lateral projection (Same frame as bottom left.) The posterior, morphologic LV is clearly seen. The posterior pulmonary artery originates above LV. The aorta (AO) is noted anterior to the pulmonary artery. Subpulmonic obstruction is demonstrated (arrow) and is possibly due to accessory endocardial cushion tissue. The anterior, morphologic RV is minimally opacified.

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