Cardiac Repair in Anatomically Corrected Malposition of the Great Arteries

By John W. Kirklin, M.D., Albert D. Pacifico, M.D., L. M. Barger, Jr., M.D., and Benigno Soto, M.D.

SUMMARY

Anatomically corrected malposition of the great arteries is a congenital cardiac malformation in which the atria and ventricles are in concordant relation, as are the ventricles and great arteries, but the aorta is to the left of the pulmonary artery (1-position). The reported cases and our two patients have had situs solitus of viscera and atria. The aortic and pulmonary circulations are in series. We report successful surgical repair in 2 patients with associated large ventricular septal defects and pulmonary stenosis. In one the defect was posterior and in relation to the pulmonary artery which slightly overrode it. In the other the defect was anterior and the aorta overrode it. The developmental basis of these two examples of the same entity is probably different. An appropriate terminology and categorization that is useful surgically is presented.

Additional Indexing Words: Malpositions of the great arteries 1-position Anatomically corrected transposition

A N AT O M I C A L L Y C O R R E C T E D malposition of the great arteries is the modified phrase suggested by Van Praagh to describe the malformation formerly entitled anatomically corrected transposition of the great arteries. We use this phrase to indicate a malformation consisting of a concordant or normal relation between atria and ventricles and the ventricles and great arteries. Left ventricle ejects into aorta, and right ventricle into pulmonary artery. The pulmonary and systemic circulations are in series. However the great arteries are in abnormal relation one to the other (malposed). In individuals with this malformation and with situs solitus of viscera and atria, the ventricles are in situs solitus but the aorta is to the left (1-position) and the pulmonary artery to the right. Obviously, the phrase “anatomically corrected” is redundant, since the term malposition is a general term used to indicate that the spatial interrelations of aorta and pulmonary artery are abnormal, but the phrase has been retained since it has been in general usage.

When no other defect exists, the circulation of blood proceeds normally. We have cared for two patients who in addition had large ventricular septal defects and pulmonary stenosis.

Case Reports

Case 1 is that of a 31 year old slightly cyanotic man who was active and gainfully employed. Since infancy he had been mildly cyanotic and his exercise tolerance had been limited by dyspnea. One year prior to admission he temporarily had atrial fibrillation. A harsh systolic murmur (grade 4 in intensity on the basis of 1 to 6) was heard over the precordium. The diagnosis was not made until the time of operation. The abdominal viscera and atria were in situs solitus. The heart was to the left. The right ventricle was anterior and to the right and moderately enlarged, and the left ventricle was mildly enlarged and lay posteriorly and to the left. Right atrium drained into right ventricle and left atrium into left ventricle (atria and ventricles were in concordant relation). The aorta received blood only from left ventricle (aorta was above left ventricle) and the pulmonary artery mostly from right ventricle (concordant relation of ventricles and great arteries). However, the positional interrelations of the great arteries were abnormal. The aorta was to the left (1-position) and anterior. The pulmonary artery which was posterior and to the right overrode the large ventricular septal defect. Projecting the plane of the ventricular septum up to the pulmonary valve suggested that about 80% of the valve area was over right ventricle and 20% over left ventricle. A large, well formed muscular band inside the right ventricle swept to the left of and anterior to the pulmonary valve to fuse with anterior right ventricular wall. The anterior leaflet of mitral valve had continuity with the
pulmonary valve across the large ventricular septal defect, which was opposite the pulmonary valve and somewhat posterior. The base of the septal and anterior leaflets of the tricuspid valve was nearly in continuity with the pulmonary valve (as the tricuspid valve relates to the aortic valve in tetralogy of Fallot with severe dextroposition of the aorta). The aorta was just to the left of the large muscular band. Aortic-mitral fibrous continuity was present. Several small muscular ventricular septal defects were present anteriorly and inferiorly.

Right ventricular pressure at preoperative cardiac catheterization was 110/6 mm Hg, that in pulmonary artery was 35/10 mm Hg (subvalvular pulmonary stenosis was present), and that in aorta was 110/65 mm Hg. Systemic blood flow was 5.6 liters/min/m², and pulmonary flow was 3.1 liters/min/m².

The electrocardiogram showed an atrial and ventricular rate of 73. The rhythm was sinus and the P-R interval .18 seconds. The axis of the frontal QRS vector was 100°. The precordial leads showed a normal septal depolarization, an RSR' pattern in V₁ and voltage in V₆ consistent with mild left ventricular hypertrophy. The tracing was thought to be compatible with mild biventricular hypertrophy.

Repair was accomplished on August 7, 1968, during 124 min of cardiopulmonary bypass in five periods of aortic cross-clamping of 10 to 15 min each. A transverse right ventriculotomy was made. A portion of the heavy muscular band was excised to improve exposure. The ventricular septal defect was repaired by sewing into place a piece of knitted Dacron, placing it around the base of the pulmonary valve so that pulmonary artery now arose entirely from right ventricle. The muscular ventricular septal defects were closed with mattress sutures brought to the outside of the heart and tied over pledgets of Dacron. After repair, peak pressure in right ventricle was 60 mm Hg, and that in left ventricle 105 mm Hg. The cardiac rhythm was sinus. Convalescence was uneventful and he remains well four years later.

Case 2 is that of an eight year old girl who had been active throughout childhood, but had been mildly limited by dyspnea and cyanosis. On examination she was cyanotic, and had clubbing of the fingers and toes. A loud systolic murmur (grade 5) was heard over the precordium. The chest roentgenogram was abnormal (fig. 1).

The electrocardiogram showed an atrial and ventricular rate of 100. The rhythm was sinus and the P-R interval was .16 sec. The axis of the frontal QRS vector

Figure 1
Poster-anterior (A) and lateral (B) roentgenogram of the chest of Case 2. The gas bubble of the stomach is beneath the left hemidiaphragm (situs solitus of the viscera). The ascending aorta is a border-forming structure in the upper left of the cardiac silhouette (l-position). Assuming a concordant viscero-atrial relation, the patient could have a concordant atrioventricular relation and the diagnostic possibilities would include anatomically corrected malposition of the great arteries, double outlet left ventricle, and double outlet right ventricle. The patient could have a discordant atrioventricular relation (the most common situation in l-position of the aorta), in which case the diagnosis could be corrected transposition of the great arteries (discordant ventriculo-arterial relation), double outlet right ventricle, or single ventricle (double inlet ventricle).

Circulation, Volume XLVIII, July 1973
was 120°. The precordial leads showed normal septal depolarization and were suggestive of moderately severe right ventricular hypertrophy.

Preoperatively, biplane angiocardiology allowed the diagnosis of anatomically corrected malposition of the great arteries (figs. 2 and 3). The angiographic findings were confirmed at operation. The abdominal viscera and atria were in situs solitus. The heart was left-sided. The right ventricle was anterior and to the right, and the left ventricle posterior and to the left. The right atrium emptied into right ventricle and the left atrium into left ventricle (concordant ativoventricular relation). The pulmonary artery received blood from right ventricle and the aorta primarily from left ventricle (concordant ventriculoarterial relation). However, the aortic valve and aortic root were to the left (1-position) and anterior to the pulmonary valve and first portion of the pulmonary artery, which were to the right and posterior. The pulmonary valve was entirely over right ventricle. The aorta was in relation to (overrode) the large anteriorly placed ventricular septal defect. Projection of the plane of the septum to this semilunar valve angiocardiographically and surgically led to the conclusion that about 60% of the valve area was over left ventricle and 40% over right ventricle. A heavy band of right ventricular musculature swept up from the base of the ventricular septal defect to separate the aortic orifice from pulmonary orifice. This contributed to the subvalvar pulmonary stenosis, and the pulmonary valve also was stenotic. A subpulmonary conus existed. Peak pressures in right and left ventricles were the same. The atrial appendages were juxtaposed to the left of the great arteries.

Repair was effected on July 25, 1972, during cardiopulmonary bypass of 88 min and aortic cross-clamping of 55 min after profound cardiac cooling. Through a transverse right ventriculotomy the heavy muscular band was mobilized and that part under the pulmonary valve excised. Pulmonary valvotomy was done. The ventricular septal defect was closed by suturing into place a piece of knitted Dacron, placing it somewhat in the fashion used to repair double outlet right ventricle with subaortic ventricular septal defect* and avoiding the production of subaortic stenosis. After repair, peak pressure in left ventricle and ascending aorta was 105 mm Hg and in right ventricle 75 mm Hg. Pulmonary artery peak pressure was 35 mm Hg. Sinus rhythm was present. Convalescence was uneventful and she is well six months postoperatively.

Morphology

Clearly, there is a form of congenital heart disease in which the great arteries are in abnormal spatial relation one to the other (malposed) but in which the circulatory pathway is normal since pulmonary venous (arterial) blood drains from the morphologically left atrium to morphologically left ventricle and is ejected into aorta, and systemic venous blood passes from the morphologically right atrium to morphologically right ventricle to pulmonary artery. Van Praagh and Van Praagh, in reporting on this entity, published two cases (Cases 1 and 2) with similarities to the two cases reported by us. The categorization of these cases requires exact definition of terms. A terminology and categorization that has been consistent and useful surgically has been evolved by us (table 1); its development has been aided greatly by Van Praagh (personal communication).

The term concordant relation (between atria and ventricles and ventricles and great arteries) means that right atrium empties into right ventricle and this into pulmonary artery, and left atrium into left ventricle and this into aorta. The terms left and right atrium and ventricle of course refer to the morphologically left and right atria and ventricles. Most classifications have some impurity, and so does this one. When atria and ventricles are in situs solitus and in concordant arrangement, and left ventricle ejects into aorta and right ventricle into pulmonary artery, the aorta is usually to the right (d-position). Rarely, it is to the left (1-position), as

<table>
<thead>
<tr>
<th>Table 1</th>
</tr>
</thead>
<tbody>
<tr>
<td>Surgical Classification of Basic Cardiac Malformations*†</td>
</tr>
</tbody>
</table>

| Concordant ativoventricular relation |
| Concordant ventriculo-arterial relation |
| d—position of aorta (normal) |
| l—position of aorta (anatomically corrected malposition of the great arteries) |
| Discordant ventriculo-arterial relation (transposition of the great arteries) |
| Double outlet right ventricle |
| d—position of aorta |
| l—position of aorta |
| Double outlet left ventricle |
| d—position of aorta |
| l—position of aorta |
| Common arterial trunk |

| Discordant ativoventricular relation |
| Discordant ventriculo-arterial relation (isolated ventricular inversion; isolated atrial inversion) |
| l—position of aorta |
| d—position of aorta |
| Discordant ventriculo-arterial relation (corrected transposition of the great arteries) |
| Double outlet right ventricle |
| l—position of aorta |
| d—position of aorta |
| Double outlet left ventricle |
| Common arterial trunk |
| Single ventricle |
| Common atrium |
| Cor bilocular, etc. |

*The heart may be in levoposition or dextroversion (or their analogues in situs inversus of the viscera).
†These may occur with situs solitus of viscera (and usually the atria) or situs inversus.

Circulation, Volume XLVIII, July 1973
Figure 2

Antero-posterior (A) and lateral (B) projections of left ventriculogram. The left ventricle (LV) ejects through a subaortic conus (C) into the aorta (AO), which is to the left of the pulmonary artery (PA) in l-position. A later sequence (C and D) shows the right ventricle (RV) opacified via the ventricular septal defect. The conus (C) and conal narrowing beneath the pulmonary artery are shown. The aortic conus overrides the ventricular septal defect, but about 60% of it appears to be over left ventricle.
MALPOSITION OF THE GREAT ARTERIES

in the cases described here. Nonetheless, the term concordant ventriculo-arterial relation, as defined above, still applies. As indicated in the table, rarely the aorta may be to the left (1-position) in other malformations in which the morphologic right ventricle is to the right and anterior.

By angiocardiology and inspection at operation one determines chamber and arterial morphology and position. The surgeon, in planning the needed operative repair, must think morphologically but also functionally. Functional thinking is aided by identifying early in the analysis concordance or discordance in atioventricular relations and ventriculo-arterial relations. When both are concordant (as in the normal) or both are discordant (as in corrected transposition of the great arteries) the pulmonary and systemic circulations are in series. When either atioventricular relations (such as in isolated atrial5 or ventricular inversion as defined in the table6) or ventriculo-

arterial relations (such as in transposition of the great arteries) are alone discordant, the two circulations are in parallel. When both atria empty into one ventricle, the phrases concordant or discordant atio-ventricular relations are not applicable. When both great arteries come off the same ventricle, or there is a common arterial trunk, the phrase concordant or discordant ventriculo-arterial relation is obviously not applicable.

Transposition of the great arteries is defined variably, but we use it to refer to a specific malformation in which the ventriculo-arterial relations are discordant. That is, the aorta arises from right ventricle and pulmonary artery from left ventricle. The phrase connotes that the atioventricular relations are concordant. When they are discordant in this situation, the descriptive phrase is corrected transposition of the great arteries. Malposition of the great arteries is a general phrase indicating an abnormal spatial relation between the

Figure 3

Antero-posterior (A) and lateral (B) projections of right ventriculogram. Right ventricle is anterior and to the right of left ventricle. The pulmonary artery arises solely from right ventricle, and there is subpulmonary stenosis.

Circulation, Volume XLVIII, July 1973
aortic and pulmonary valves. The great arteries can be abnormal in their spatial relations one to the other (such as being side-by-side in double outlet right ventricle, or parallel in transposition of the great arteries) with the aorta still to the right of the pulmonary artery (d-position), or the aorta may be to the left (l-position).

In view of the great variation in conal anatomy and semilunar-atroventricular valve continuity or discontinuity within the accepted categories of congenital heart disease, these are not useful as criteria for diagnosing a given entity. This implies that the entity of transposition of the great arteries or double outlet right or left ventricle may result from several embryologic aberrations.

Anatomically corrected malposition of the great arteries is a malformation in which the atria and ventricles are in concordant relation as are the ventricles and great arteries. Yet in spite of ventriculo-arterial concordance, there is malposition of the great arteries. With situs solitus of viscera, atria, and ventricles, the aorta is in l-position. Two such cases were reported by Van Praagh and Van Praagh (their Cases 1 and 2). In a patient with situs inversus totalis, the phrase “anatomically corrected malposition” would connote atrioventricular concordance, ventriculo-arterial concordance, and d-position of the aorta. In Case 3 of Van Praagh and Van Praagh, the atrioventricular relation was discordant and the ventriculo-arterial relation was concordant (personal communication). The viscera and atria were in situs solitus, the ventricles were in situs inversus (discordant atrioventricular relation), the aorta came off left ventricle and pulmonary artery off right ventricle (concordant ventriculo-arterial relation), but unexpectedly, the aorta was to the right (d-position). We would use the term “isolated” ventricular inversion (which like many old terms is not ideal but is satisfactory if defined clearly by those who use it) with d-position of the aorta. Anatomically corrected malposition has, to our knowledge, been reported to date only in patients with situs solitus of the viscera and atria.

Our two patients had malformations which probably were of differing embryologic origin. In Case 1, the ventricular septal defect was posterior in the septum and related to the pulmonary artery, which overrode it. Fibrous continuity was present between anterior leaflet of mitral valve and both pulmonary and aortic valves. Both conus were probably absent. Durnin et al. have also studied such a case.1 This form of the malformation has anatomic similarities to certain forms of double outlet left ventricle. The ventricular septal defect in Case 2 was anterior and related to the aortic valve. The aorta overrode the defect. Subaortic and subpulmonary conuses both seemed to be present. This type of anatomically corrected malposition is possibly related to double outlet right ventricle with l-position of aorta.8,9

Obviously in a few patients a great artery overrides the ventricular septal defect so that it arises exactly equally from the two ventricles. To avoid still another category of defects with “exactly biventricular origin” of a given vessel, we have arbitrarily assigned the origin of a malposed vessel to one or the other ventricle, using all available facts to make the assignment.

Diagnostic Considerations

The first clue to the diagnosis of anatomically corrected malposition of the great arteries may be in the chest roentgenogram. The gas bubble of the stomach is to the left (situs solitus of the viscera) and the ascending aorta is seen on the left. Most often in this situation, the atria will prove to be in situs solitus, the ventricles inverted, and a discordant relation will exist between atria and ventricles and ventricles and great arteries (corrected transposition of the great arteries) or there will be single ventricle with double inlet left ventricle; rarely, the diagnosis may prove to be discordant atrioventricular relation and double outlet right ventricle. Uncommonly when the aorta is in l-position, the atrioventricular relation is discordant; there may be a discordant ventriculo-arterial relation (anatomically corrected malposition) or double outlet left ventricle or double outlet right ventricle.

The diagnosis can be made by high-quality biplane angiocardiography. Views which profile the ventricular septum precisely are important.

The surgeon must always verify the anatomic relations for himself at operation, since the preoperative diagnosis may not be accurate in all details.

Surgical Implications

The ventricles in a patient with anatomically corrected malposition of the great arteries presumably are essentially normal in their internal anatomy and interrelations to each other, as are the atria. Therefore the usual guides to the location of the bundle of His are applicable in the repair of associated ventricular septal defects.10 Such is not the case when there is a discordant atrioventricular

Circulation, Volume XLVIII, July 1973
MALPOSITION OF THE GREAT ARTERIES

relation, such as in corrected transposition of the great arteries.

The pulmonary stenosis in Case 2 was correctable by valvotomy and resection of the subvalvar narrowing. Because the coronary arteries may be in abnormal position, a valved external conduit between right ventricle and pulmonary artery may be needed when direct relief of the pulmonary stenosis is not possible.

References
3. Harris JS, Farber S: Transposition of the great cardiac vessels, with special reference to the phylogenetic theory of Spitzer. Arch Path 28: 427, 1939
5. Clarkson PM, Brandt PWT, Barratt-Boyes BG, Neutze JM: "Isolated atrial inversion." Visceral situs solitus, visceraloatrial discordance, discordant ventricular d loop without transposition, dextrocardia: Diagnosis and surgical correction. Amer J Cardiol 29: 877, 1972
Cardiac Repair in Anatomically Corrected Malposition of the Great Arteries
JOHN W. KIRKLIN, ALBERT D. PACIFICO, L. M. BARGERON, JR. and BENIGNO SOTO

_Circulation_ 1973;48:153-159
doi: 10.1161/01.CIR.48.1.153
_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/48/1/153

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/