Criss-Cross Atrioventricular Relationships
Producing Paradoxical Atrioventricular
Concordance or Discordance
Their Significance to Nomenclature of Congenital Heart Disease

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SUMMARY
Two cases are described in which the systemic and pulmonary blood streams cross at atrioventricular level. One was examined pathologically, the other was diagnosed at cardiac catheterization by angiography. Both possessed situs solitus, but in one heart the right atrium was connected to a morphologic right ventricle on the left, whereas in the other it was connected to a left-sided morphologic left ventricle. In both cases transposition of the great arteries was present. The cases have been interpreted as representing rotation of embryonic d- and l-bulboventricular loops following septation.

In attempting to describe these heart difficulties in terminology were encountered which were partially overcome by combining the concepts of concordant and discordant atrioventricular relationships with those of transposition and malposition. However, to give an accurate interpretation of the hearts, it proved necessary to include a term indicating rotation of the bulboventricular loop following septation. Thus the hearts were described as situs solitus, discordant (d-rotated) loop with d-transposition, and situs solitus, concordant (l-rotated) loop with l-transposition.

Additional Indexing Words:
Morphology
Malposition
Embryology
"Corrected" transposition
Transposition
Bulboventricular looping

An ANGIOGRAPHIC TECHNIQUES now allow accurate localization of the positions of the cardiac chambers and great arteries in congenitally malformed hearts. Van Praagh has for some time1-4 promoted a segmental approach to the nomenclature of such anomalies, believing that description of the position of the "embryonic variables" of the primitive heart tube enables subjective visualization to be made of the anomalous hearts. In this system, atrial chambers are described as solitus, inversus, or ambiguous,5 the bulboventricular loop is considered to be in dextro- or levo-position, and the great arteries are considered to be nontransposed, transposed, or malposed, the latter two being possible in both dextro- or levo-positions. Kirklin et al.6 modified this system by including the terms concordant (RA to RV, LA to LV) and discordant (RA to LV, LA to RV) atrioventricular relationships, and by describing only "positions" of the great arteries. However, it was recognized by these authors that exceptions to their systems exist. One exceptional situation would be produced by rotation of part of a malformed heart following septation. An example of this is a levo-transposed aorta arising from the right ventricle of a d-bulboventricular loop.4,7 A less common but equally important rotational variable would produce apparent concordance of a discordant atrioventricular (A-V) relationship. This situation was demonstrated by two of the specimens of mixed levocardia reported by Lev and Rowntree8 where the solitus atrial chambers communicated with inappropriate ventricles arranged in normal fashion. A right-sided right atrium therefore drained to a left-sided left ventricle. A similar "criss-cross" was reported but not illustrated by Van Praagh.4 This heart exhibited apparent atrioventricular discordance in that the atria were in solitus position and the right ventricle was left sided. However, this left-sided right ventricle received right atrial blood so that concordance was present. To the
best of our knowledge these are the only reported “criss-cross” hearts. We have recently studied an example of both these types of “criss-cross,” one pathologically and one angiographically. In this presentation we describe the anatomic features of these hearts, consider their possible embryogenesis and discuss their significance to the nomenclature of congenital cardiac disease.

Case Reports

Case 1

CR was a two-weeks postmature infant, delivered normally. She was immediately noted to be of poor color and rapidly became cyanosed although no intracardiac murmur was detected. Despite supportive therapy she died 23 hours after birth. Necropsy examination revealed no abnormalities in the extracardiac organs apart from a uniloculated cyst, 2.5 cm in diameter, attached to the second and third right ribs.

The heart was of normal size with its apex directed to the right (fig. 1a). The atria were in solitus position, and systemic and pulmonary veins drained to right and left atrial chambers respectively (fig. 1b). Internal appearances of the atria were normal and a patent foramen ovale was present. The right atrium drained via a tricuspid valve into a morphological right ventricle. However, although the inflow portion of the ventricle was situated anteriorly, its trabeculated portion extended posteriorly and to the left (fig. 1c). Its septal surface extended from the right anterior to left posterior quadrants of the ventricles, and was covered by a well formed trabecula septomarginalis, from which arose the conal papillary muscle (fig. 1c). The ventricle drained anteriorly through the aorta. The aortic valve arose from a completely muscular conus. The right portion of the conus was formed by the infolded right anterior atrioventricular groove. Posteriorly the conus septum separated aortic and atretic pulmonary valves (q.v.) and a small ventricular septal defect was present beneath this structure. To the left the inner surface of the left anterior A-V groove supported the aortic valve and anteriorly the aortic conus was completed by the parietal wall of the right ventricle (fig. 1c). The left atrium drained through a mitral valve to a morphologic left ventricle situated on the right side of the heart (fig. 1d). A ventricular septal defect was present in the left anterior quadrant of this ventricle, which communicated with the right ventricle beneath the conus septum (fig. 1d). The atretic pulmonary valve ring was sandwiched between the conus septum and the anterior cusp of the mitral valve. The aorta communicated with a right-sided aortic arch, and a patent ductus connected the left subclavian artery to the left pulmonary artery (fig. 1b). The main pulmonary artery was poorly formed and was situated posteriorly and to the right of the aorta (fig. 1a).

Case 2

AS is a 9-year-old girl in whom a murmur was first noted at three years when there was minimal effort intolerance. Aged six years, at thoracotomy elsewhere, the patient had a persistent ductus arteriosus ligated, but the cardiac anatomy was not elucidated. Physical examination revealed a well-nourished, acyanotic girl. Pulses were normal as was the cardiac impulse. The first heart sound was normal, the second single, and there was a grade 4/6 long ejection systolic murmur maximal in the second left intercostal space. Apart from a thoracotomy scar the rest of the physical examination was normal.

The electrocardiogram showed sinus rhythm and a mean frontal QRS axis of $-140^\circ$. A peaked P wave in lead II in-

![Figure 1](http://circ.ahajournals.org/)

**Figure 1**

*Case 1. Photographs of the heart. A. Anterior view from right side. The cut is into the morphologic right ventricle, from which the aorta (Ao) arises. The pulmonary artery (PA) is small and is posterior and to the right of the aorta. The right atrium (RA) is retracted to reveal the anterior atrioventricular groove (RAVG). B. Posterior view from left side. The atria are in solitus position, with systemic veins (SVC, IVC) draining to right atrium and pulmonary veins (PV) to left atrium. The cut is into the morphologic left ventricle. The ductus (PDA) connects the left subclavian artery to the left pulmonary artery. A typical left atrium is visible (LA). C. Heart opened through the anterior cut. The tricuspid valve (TV) drains from right atrium to morphologic right ventricle (MRV) situated on the left. The aortic valve is separated from the tricuspid valve by the conus septum (CS) and the internal part of the anterior A-V groove (RAVG). The trabecula septomarginalis (TSM) is posterior. D. Heart opened through the posterior cut. The mitral valve (MV) connects the left atrium (LA) to the morphologic left ventricle (MLV) which is to the right of the morphologic right ventricle (MRV). The rod is in the septal defect which connects beneath the conus septum with the right ventricle.*

*Circulation, Volume 50, July 1974*
dedicated right atrial hypertrophy and the P wave vector was +50°. Right ventricular hypertrophy was present as indicated by an rSr' pattern in V1, R, and V1 with an R' of 17 mm and 30 mm respectively. Left-sided q waves were absent. Chest X-ray showed slight cardiomegaly and normal pulmonary vascular markings. There was a straight left upper heart border.

Cardiac catheterization showed no evidence of intracardiac shunting. Systolic pressure in the systemic ventricle was 125 mm Hg, in the pulmonary ventricle 82 mm Hg, and in the pulmonary artery 25 mm Hg indicating pulmonary stenosis. Arterial blood was fully saturated.

Angiography

The catheter was passed from a normally situated right atrium into a posterior left-sided ventricle. This had the angiographic characteristics of a left ventricle (figs. 2a and 2b) in that it was globular and relatively smooth walled. From it arose a posterior left-sided pulmonary artery. The pulmonary valve leaflets were normal but there was evidence of subvalvar obstruction. In the levophase contrast passed from a posterior left-sided left atrium into an anterior relatively right-sided ventricle from which arose an anterior aorta (fig. 2c). This ventricle had the characteristics of a right ventricle with well marked inflow and outflow portions. A distinct shelf of tissue separated aortic and atrioventricular valves. The aortic valve was above and to the right of the pulmonary valve (figs. 2a, c, d).

Discussion

The cases presently described are interpreted as representing crossing of systemic and pulmonary venous blood streams (without mixing) at atrioventricular level, hence our designation as “criss-cross” hearts. We further interpret our Case 1 as representing the situation where an embryonic d-bulboventricular loop has undergone rotation following septation and Case 2 as rotation of an I-loop following septation. These cases are rare and to the best of our knowledge the only previous reports are by Lev and Rowlatt of two cases and a short description of personal experience by Van Praagh. Case 6 of Wagner et al. is similar to these but while exhibiting malrotation it did not show a complete criss-cross. Van Praagh used his case to illustrate difficulties of providing a foolproof, all-encompassing terminology, and this problem is further exemplified by the cases of Lev and Rowlatt. They describe them within the term “mixed levocardia” indicating an apex to the left with atrioventricular discordance. However, within this definition they also describe eight cases of “classical corrected transposition,” two cases of “anatomically corrected transposition” and one case of “classical complete transposition.” Furthermore, one of our cases and the case described by Van Praagh could not be included within the term “mixed levocardia” since the atria and ventricles were, in fact, concordant.

However, we describe our two cases together since we believe that both are criss-cross hearts, and that they represent the criss-cross situation in embryonic d- and I-loops respectively. We would interpret both of the criss-cross cases of Lev and Rowlatt as representing postseptational rotation in an I-loop, or in other words, in the presence of atrioventricular discordance. We consider that this interpretation makes our Case 2 much easier to comprehend, since the heart has the same discordant A-V relationship as the “classically corrected transposition.” It is then clear why the right atrium connects with a left-sided chamber with typical angiographic characteristics of the left ventricle. Similarly subpulmonary obstruction is a common feature of “corrected transposition.” To achieve the demonstrated chamber orientation it is necessary to

Figure 2

Case 2, angiocardiograms. A and B. Postero-anterior and lateral views of contrast injected into a posterior left-sided morphological left ventricle (LV) from which arises a posterior left-sided pulmonary artery (PA). Subvalvar pulmonary obstruction is present (4v). The catheter course is from the superior vena cava to the right atrium and thence to the left ventricle. The catheter does not pass through the left atrium. C and D. Postero-anterior and lateral views of the leوقفة of the injection into the pulmonary ventricle (figs. 2A and B). Contrast is seen passing from left atrium (LA) anteriorly into a right-sided morphological right ventricle (RV) which in turn drains to an anterior right sided aorta (Ao). A distinct shelf separates the aortic and atrioventricular valves (4v).
postulate a clockwise rotation of the ventricles looking toward the cardiac base. Such rotation would turn the originally I-transposed aorta into its demonstrated d-position. We are unable to construct any other embryologic sequence which would produce this demonstrated chamber orientation. The same detailed interpretation applied to Case 1 produces the conclusion that an originally d-transposed heart with atrioventricular concordance was transformed into the described specimen by counter-clockwise rotation around the base — apex axis looking toward the base. It is a possibility that additional conal rotation relative to bulboventricular rotation could have produced the valvar pulmonary atresia present in this specimen.

We consider that the conclusions reached have important diagnostic implications. In Case 2 the great arteries are in the configuration of d-transposition. With solitus atria and normally situated ventricles this usually implies "classical complete transposition." If surgery became necessary at any stage upon this heart it would be necessary to know that atrioventricular discordance was present, since a recent study has demonstrated that conducting tissue is arranged in a different fashion from normal in hearts with "classical corrected transposition." Thus in the heart presently described we believe that the atrioventricular node and bundle would be situated anteriorly rather than posteriorly as in the normal heart, and hearts with isolated VSDs. We are of the opinion that diagnosis of this arrangement becomes more likely if all are aware of the existence of "criss-cross" hearts, a fact which was not apparent to us until we made detailed study of the hearts presently described.

We have illustrated above the difficulties encountered in classifying these hearts, and we believe their main significance is to the nomenclature of congenital heart disease. While the segmental concept of Van Praagh is exceedingly useful, it suffers from the disadvantage that it does not "connect" the cardiac segments. Thus S-D-L in his terminology could describe either "anatomically corrected malposition" or complete transposition with an I-transposed aorta.

This ambiguity is also present in the nomenclature of Kirklin et al. and they themselves indicate that the "I"-positioned aorta arising from a concordant loop with solitus atria is an impurity. Insofar as the use of concordance or discordance "connects" the atrial and ventricular chambers, the terminology of Kirklin et al. represents a refinement of the segmental description of Van Praagh. The cases presently described are accurately classified as discordant relationships in Case 1 and discordant relationships in case 2, both associated with solitus atria. However, such terminology would still be confusing since apparent discordance is present in Case 1 and apparent concordance in Case 2.

We believe this confusion can be removed by describing rotation of the bulboventricular loop, considering leftward shift of the right ventricle as levoration and rightward shift (of a left sided right ventricle) as dextro-rotation. Thus, incorporating this concept into the nomenclature of Kirklin et al., Case 1 is described as solitus-concordant loop (I-rotated) and Case 2 as solitus-discordant loop (d-rotated).

Figure 3
Diagrammatic representations of hearts to illustrate the concept of malposition and transposition suggested by Van Praagh. The hearts are viewed from above as if they were transversely sectioned at the level of the atrioventricular junction. A represents classically complete transposition; it is a TRANSposition because both arteries are placed across the septum so that they are above inappropriate ventricles. B and C are MALpositions. B is a double outlet right ventricle, and only the aorta is placed across the septum, hence its malposition. C is anatomically corrected malposition, the arteries are related to appropriate ventricles, but are abnormally arranged since the aorta is to the left and possesses a complete muscular conus. Horizontal lines = ventricular septum; diagonal lines = conus septum; vertical lines = conal musculature; MV = mitral valve; Ao = aorta; PA = pulmonary artery.

Figure 4
Diagrams to illustrate the "impurity" in the classifications of Van Praagh and Kirklin et al. The hearts illustrated are A) "classical complete transposition" with leftward rotation of the aorta and B) anatomically corrected malposition. It will be noted that these hearts are described in identical terms in both the above classifications, but can be distinguished if reference is made to TRANSposition and MALposition (see fig. 3). Cross-hatching and abbreviations as for fig. 3.
Diagrammatic representations of the hearts presently described. The hearts are depicted in oblique plane to demonstrate the relationships of atria, ventricles and great arteries. The diagram of Case 1 is constructed from the anatomic data. The exact anatomy depicted in Case 2 represents our interpretation of the angiocraphic data. It is demonstrated how the amended nomenclature can accurately describe these hearts. Cross-hatching and abbreviations as for figure 3: LA = left atrium; RA = right atrium; LV = left ventricle; RV = right ventricle.

Figure 5

However, this still leaves the "impurity" of the great arteries unaccounted for. We believe that this problem is solved by adopting the recent definition of Van Praagh\textsuperscript{12} pertinent to transposition. He now considers that TRANSposition is only present when both arteries are placed across the septum to adopt positions above inappropriate ventricles. When only one artery is placed across the septum, or when both arteries are incorrectly related to their appropriate ventricles, they are said to be MALposed (fig. 3). If one adopts this definition then an aorta arising from a right ventricle in a d-loop can be either d-transposed or l-transposed depending on its position relative to the pulmonary artery. If it arose to the left but from a left ventricle in a d-loop it would be l-malposed. Thus this definition removes the impurity from both terminologies, and "anatomically corrected malposition" can be accurately described as either S-D-L(mal) or solitus-concordant-L(mal). The impure form of "complete" transposition is then fully described as S-D-L(trans) or solitus-concordant-L(trans) (fig. 4). When applied to the hearts presently studied, the full description of Case 1 becomes solitus-concordant(l-rotated)-l(trans), and Case 2 becomes solitus-discordant(d-rotated)-d(trans) (fig. 5). Case 12 of Lev and Rowlatt\textsuperscript{a} is identical to our Case 2, while their Case 13 can be accurately described as solitus-discordant(d-rotated)-DORV with l-malposition. This succinctly indicates that the left atrium connects with a right-sided morphological right ventricle, above which both great arteries take origin with the aorta to the left. We believe that this adaptation of the nomenclature of Kirklin et al.\textsuperscript{8} to include the concept of malposition and transposition\textsuperscript{12} is the shortest and most accurate method of describing hearts distorted by postseptational rotation.

Acknowledgments

We are grateful to Dr. R. Van Praagh, Boston, for helpful discussion pertinent to these cases. The photographs were produced by Mr. K. Walters, Institute of Child Health, University of Liverpool and by the Photographic Department, Royal Marsden Hospital, London. We are also indebted to Miss E. H. W. Thompson for help during preparation of the manuscript.

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Circulation, Volume 50, July 1974
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Circulation. 1974;50:176-180
doi: 10.1161/01.CIR.50.1.176

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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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:
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