D-Bulboventricular Loop with L-Transposition in Situs Inversus

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SUMMARY
In the case described anatomically corrected transposition of the great vessels is associated with situs inversus. Anatomically corrected transposition is extremely rare, and has not been previously reported with situs inversus. This type of transposition should not be confused with classical corrected transposition. Thus the case examined exhibited a D-bulboventricular loop with L-transposition and atrial inversion, so that blood flow was physiologically incorrect as in classical complete transposition. In the case examined, it was also found that bulbar musculature was present between the aorta and the mitral valve.

Additional study of the conducting tissue revealed inversion of the sinoatrial node with the atria, and also of the atrial portion of the atroventricular node. The atrio-ventricular bundle and its branches were in expected positions in relation to a large ventricular septal defect. These results are discussed with regard to previous reports and to the embryology of the conducting tissue.

Additional Indexing Words:
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Bulboventricular loop

THE CONCEPT of anatomically corrected transposition as opposed to physiologically corrected transposition was first referred to by Harris and Farber,¹ who did not, however, personally present any cases of the anomaly. The first adequate documentation of this form of transposition was presented by Van Praagh and Van Praagh.² In their report, the Van Praaghs documented three cases of anatomically corrected transposition in situs solitus but stated that such cases had not been previously described in association with situs inversus. We have recently studied such a case which fulfilled the criteria described by these authors for anatomic correction. In addition we have studied the specialized conducting tissue of this heart and the results are described and compared with previous descriptions of the specialized tissue in transposition complexes.

Report of Cases

Clinical History
A male infant who was admitted to the hospital in 1963 first became ill at the age of 4 weeks, presenting with dyspnea and cyanosis. His condition deteriorated and at the age of 9 weeks he was transferred to our hospital. The initial clinical diagnosis was dextrocardia and ventricular septal defect, and the infant was in severe cardiac failure. Angiography was not performed. Despite treatment his cardiac failure progressed, and he died at the age of 12 weeks. A limited necropsy was performed, and this revealed dextrocardia. The lungs showed evidence of congestion consistent with cardiac failure. Examination of the abdominal organs through the diaphragm showed complete situs inversus. The spleen was of normal dimensions and was situated on the right side.

Results

Gross Cardiac Anatomy
In describing the cardiac chambers in this report, they will be named according to their

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ANDERSON ET AL.

Figure 1

(A) Posterior view of heart. The venous atrium (VA) or anatomic right atrium situated to the left receives the superior vena cava and inferior vena cava. The pulmonary veins drain to the anatomic left atrium (AA) situated on the right. The anatomic left atrial appendage (arrowed) is in an artifactually produced position. (B) Anterior view of the heart. The aorta (AO) arises anteriorly and to the left of the pulmonary artery (PA), the typical configuration of L-transposition. The internal morphology of the ventricular chambers (not visible) indicated that this heart possessed a D-bulboventricular loop.

morphology rather than according to their position. Thus the venous atrium, situated on the left, is termed the "anatomic right atrium." Similarly the arterial atrium on the right is termed the "anatomic left atrium." The anatomic right and left ventricles are on their appropriately correct sides.

The morphology of the anatomic right atrium was typical of the chamber encountered in situs solitus hearts. It contained a well-formed crista terminalis and received the terminations of the superior and inferior cavae (fig. 1A) and the coronary sinus (fig 2B). The anatomic left atrium received arterial blood via four pulmonary veins (fig. 1A), and its morphology was also typical.

The ventricles were situated in the positions usually associated with situs solitus, that is they exhibited a D-bulboventricular loop. The anatomic right ventricle possessed typical trabeculations and normal conal and bulbar musculature. The valve guarding the anatomic right atrioventricular (A-V) orifice was tricuspid. The anatomic left ventricle was also typical, and its inflow tract was guarded by a bicuspid valve. However, bulbar musculature was also present separating the left A-V valve from the anteriorly situated aortic valve (fig. 2). The great vessels exhibited L-transposition with the aorta arising anteriorly and to the left of the pulmonary artery (fig. 1B). The aorta was continuous with the anatomic left ventricle, while the pulmonary artery arose from the anatomic right ventricle. A large subcristal ventricular septal defect was present (fig. 2B).

Specialized Conducting Tissue

Two blocks were removed for histologic examination. They consisted of (1) the junction of the superior vena cava, anatomic right atrium, and atrial appendage and (2)
ANATOMICALLY CORRECTED TRANSPOSITION

Figure 2

(A) This view shows the outer side of the right side of the heart, viewed from the septal aspect, following removal of the block for histologic study. Note the bulbar musculature (arrow) which separates the pulmonary valve (PUV) from the tricuspid valve (TV). The morphologic left atrium (AA) which receives the pulmonary veins (PV) opens into the morphologic right ventricle (RV). The aorta (AO), which arises above the morphologic left ventricle (not seen) is also related to bulbar musculature. (B) Block removed for histology. The rod is in the coronary sinus, which opens into the morphologic right atrium. Note the bulbar musculature (arrow) which separates the mitral valve (MV) from the aorta. The subcristal ventricular defect (VSD) is also seen.

the septum of the heart between the coronary sinus and the bulbar musculature (fig. 2B). Each of these blocks was embedded in paraffin and serially sectioned at 10-μ thickness. One section in 50 was mounted and stained with Masson's trichrome technic, and following microscopic examination intermediate sections were mounted and stained as required.

Sinoatrial Node

This structure was situated as a crescent of cells round the anterolateral quadrant of the superior vena caval-atrial junction. The cells of the node were smaller than atrial cells and were surrounded by connective tissue. Two arteries entered the node, one from each end, and the nodal cells were grouped around these vessels. In the middle of the node, however, the vessels ramified and the circumarterial arrangement was not present. Many large nerves entered the node from its epicardial aspect, and several large ganglia were also identified adjacent to the node.

Atrioventricular Specialized Tissue

Three cell types were identified within the atrioventricular node. In the region of the coronary sinus the atrial cells became attenuated and surrounded by much connective tissue. These transitional cells joined together to form columns of cells beneath the endocardium of the anatomic right atrium. These columns formed an oval against the central fibrous body, and extended toward the origin of the mitral valve (fig. 3A). As the node was traced forward, these cells were found to abut against longitudinally running cells which veered toward the right. These longitudinal cells were identical and continuous with the cells of the atrioventricular bundle, which passed through the fibrous body to the right side of the septum (fig. 3B and C). Some
Figure 3

(A) Cells of atrioventricular node (AVN) beneath endocardium of venous atrium (LA). Note transitional cells above nodal cells (TC). CFB = central fibrous body. (B) The bundle of nodal cells (AVN) lie above the atrial extension of the atrioventricular bundle (AVB), which is to the right side of the septum. (C) Nodal cells are to the left (AVN) and bundle cells to the right.
transitional cells passed circumferentially around the upper layer of nodal cells to merge directly with the cells of the bundle (fig. 3C).

The bundle ran an extensive course within the ventricles. It coursed to the left side of the posterior rim of the septal defect and passed to its inferior rim before bifurcating (fig. 4A). The left branch of the bifurcation was a widespread thin sheet of cells, while the right branch was a single fascicle which was traced down the septum to the moderator band (fig. 4B). The cells of the bundle branches were of similar size to ventricular myocardial cells.

Discussion

To produce anatomically corrected transposition, it is necessary for the great vessels to rotate in an opposite direction from the bulboventricular loop. Thus in Van Praagh and Van Praagh's series two cases exhibited a ventricular D-loop with L-transposition of the vessels, while the third case showed complete D-transposition of the vessels with a L-bulboventricular loop. All these cases showed the following features, considered essential by the authors for the pathologic diagnosis of transposed vessels: (1) presence of a combined abnormal conus; nontransposed arteries hardly ever have an aortic conus; (2) an abnormal relationship between the aorta and the atrioventricular ring with absence of mitral-aortic fibrous continuity; (3) abnormal relationship between the great vessels; and (4) although arising above the correct chamber, the arteries are not normally related to their respective chamber.

In the present case bulbar musculature is identifiable on both sides of the ventricular

\[ \text{(AVB)} \text{ Note transitional cells impinging directly on the bundle cells (††)}. \text{(D) Atrioventricular bundle (AVB) passing down the right side of the septum beneath the tricuspid valve (TV). Atrioventricular node (AVN) is related to the mitral valve (MV).} \]
Although the present case had been referred to as a D-bulboventricular loop, associated with L-transposition and atrial inversion.

Examination of the conducting tissue revealed many interesting features. The sinoatrial node is in its usual position in relation to the superior vena cava, and has undergone rotation with the atria. Its morphology is in accordance with earlier reports on the human node. The atroventricular node in the present case extends throughout the septum, but its atrial portion is beneath the endocardium of the anatomic right atrium. Its cellular morphology is similar to the arrangement described recently in infant hearts. The distal portion of the node is located on the right side of the septum, as is the atroventricular bundle. In previous descriptions of the conducting tissue in L-transposition with L-bulboventricular loop, the atroventricular node and bundle were described on the left side of the septum, suggesting that the entire node had been inverted with the ventricular loop. We have recently studied a case with L-transposition with complete A-V block and fragmentation of the conducting tissue, however, in which the atroventricular node was hypoplastic, isolated, and situated in the right atrium associated with the coronary sinus. We have also examined two additional hearts which show isolation of the node while possessing a well-developed ventricular conducting network. These cases support the concept summed up by James that node and bundle develop independently of each other rather than the concept that the bundle develops by migration from the node. Regarding the development of the node itself these cases are in contrast to the previous reports mentioned.

Patten has suggested that the node is developed from left sinus horn musculature while others consider it to be formed from the A-V canal musculature. More recently it has been suggested that the node may develop in different portions. The atrial portion may well develop from the left

Figure 5
A composite view of the sections shown in figures 3 and 4, with the upper end of the septum being posterior in relation to the VSD. It is constructed as though several transparencies had been superimposed on each other.

The part on the left forms an aortic conus which interposes between the aortic and mitral valves, preventing fibrous continuity. The aorta is also situated anteriorly and to the left of the pulmonary conus, and both vessels are abnormally related to their respective chambers. Thus all the criteria listed by Van Praagh and Van Praagh are satisfied in the present case. Since the bulboventricular loop is opposite to the looping of the vessels, an anatomically corrected transposition is present. In an earlier work Van Praagh and associates pointed out that visceral situs was invariably indicated by atrial situs. This was borne out by the present case. Van Praagh and associates have also indicated the danger of using the term "corrected transposition." Thus, although the present case is anatomically "correct," it is clearly physiologically incorrect, since it yields a functional situation identical with that in classical complete transposition of the great vessels. If used, the term "corrected transposition" requires qualification as to physiologic or anatomic considerations. It is preferable to adopt the classification suggested by Van Praagh and associates, and describe the position of the variable components of the cardiac tube. The present case is therefore described as a D-bulboventricular loop, associated with L-transposition and atrial inversion.
sinus horn, while the portion continuous with the A-V bundle, but an integral part of the node, could develop from the A-V canal musculature. This concept is supported by the present findings, since the sinus horn would be inverted with the atria and this would account for the left-sided situation of the atrial node. The bundle, however, would develop in relation to the ventricles and would rotate with the bulboventricular loop. This mode of development would also explain the morphologic arrangement of the atrioventricular node recently reported by Anderson and Latham.

If this concept of nodal development is substantiated by further studies in progress at present, it indicates that the position of the conducting tissue can be forecast at operation. Thus the sinoatrial (SA) node and atrial portion of the A-V node will be situated in the venous atrium, in relation to the right and left superior caval openings. The A-V bundle and branches, however, will rotate with the bulboventricular loop.

The arrangement of the bundle in the present case is further complicated by the presence of a large ventricular septal defect. The relationship of the bundle to the posteroinferior rim of such defects has been well documented, and it is well recognized that this area is "at risk" during repair operations.

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