The Course of the Conduction System in Single Ventricle with Inverted (L-) Loop and Inverted (L-) Transposition

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SUMMARY

The conduction systems of three hearts with single ventricle and inverted (L-) loop and inverted (L-) transposition were studied by serial section. The course of the conduction system was similar in all three hearts. The regular (posterior) atrioventricular (A-V) node was rudimentary and did not form the bundle of His. An accessory (anterior) A-V node was present in the roof of the right atrium. This node pierced the connective tissue adjacent to the right side of the pulmonary annulus to form the bundle of His. The bundle proceeded into the main ventricular chamber lying on the septum between this chamber and the small outlet chamber. It bifurcated a varying distance from the defect in this septum. The right bundle branch proceeded through the septal wall to the trabeculae of the outlet chamber. The left bundle branch proceeded caudad on the anterior wall of the main ventricular chamber.

An unusual feature in one case was the splitting of the anterior node into two parts, one forming the bundle of His, and the other giving off Mahaim fibers to the ventricular myocardium.

The surgical importance of avoiding the right side of the pulmonary annulus and the region adjoining the defect is stressed.

Additional Indexing Words:
L-transposition
His bundle
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THE COURSE OF THE CONDUCTION SYSTEM in single ventricle has taken on importance today because of the possibility of surgical intervention.1-4 We therefore studied this system in three hearts with inverted (L-) loop and inverted (L-) transposition by serial section techniques.

Single ventricle in this paper is meant to be that condition in which both atrioventricular (A-V) orifices or a common A-V orifice enter or enters a single ventricular sinus which is associated with two coni leading into the two efferent vessels.6 The ventricular mass may have originated from a D or L loop. Neither conus may be said to be related directly to an individual A-V orifice. The efferent vessels may be transposed or

normally related. If transposed, the type of transposition usually follows the type of bulboventricular loop and so is either a regular (D-) or inverted (L-) transposition (fig. 1). Common ventricle, on the other hand, is that condition in which both A-V orifices or a common orifice enter or enters two sinuses slightly subdivided by a remnant of posterior ventricular septum. Each of these two sinuses is related to an individual conus. A portion of anterior ventricular septum is usually present. Thus, we are basically dealing with a large ventricular septal defect. The arterial trunks arise again either in normal or in transposed positions (fig. 1).

This paper deals only with single ventricle with inverted (L-) loop and inverted (L-) transposition.

Materials and Methods

The electrocardiogram showed no evidence of A-V block in any of the three cases.

The conduction systems of the three cases were studied as follows: the sinoatrial (SA) node was serially sectioned and every 10th section was retained. The distal half of the atrial septum and the proximal half of the ventricular septum containing the A-V conduction system were serially sectioned at right angles to the posterior wall of the heart. In two cases all sections, and in one case every 10th section, were retained. Where complete serial sectioning was done, every 5th section was stained with Weigert-van Gieson, and the remainder with hematoxylin-eosin stains. In the case where every 10th section was retained, alternate sections were
stained with hematoxylin-cosin and Weigert-van Gieson stains. In this manner, 7118 sections were examined in case 1, 2800 in case 2, and 650 in case 3.

Results

Case 1

Case 1 (3869) was a 21-day-old white female. Anatomic diagnoses were: (1) single ventricle with inverted loop and inverted (L-) transposition, (a) left A-V valve stenosis, (b) fetal coarctation, (c) patent ductus arteriosus, (d) hypertrophy of the ventricular mass, (e) bialtrial hypertrophy.

Course of the Conduction System

The SA node was in normal position. The regular (posterior) A-V node consisted only of a fragment (fig. 2), and did not form a bundle of His. The entire roof of the right atrium adjacent to the atrial septum consisted of loose, lightly staining cells resembling A-V nodal cells (fig. 3 A). Some distance from the septum, this formed a distinct accessory nodal structure (figs. 3 A and B) adjacent to the right side of the pulmonary annulus. The accessory A-V node penetrated the right side of the dense connective tissue adjacent to the pulmonic annulus to become the bundle of His (fig. 4). We could not tell whether the bundle pierced the annulus itself. It then passed into the main ventricular chamber in the connective tissue adjacent to the pulmonary annulus (fig. 5). Here it lay in the septum between the main ventricular chamber and the outflow chamber. About 0.5 to 1 cm caudal to the right

side of the defect in this septum it bifurcated into right and left bundle branches (fig. 6). The right bundle branch (RBB) penetrated the wall of the septum (fig. 7) to reach the subendocardial lining of the outflow chamber where it passed distally, ending in the trabeculae of the chamber. The left bundle branch (LBB) passed vertically caudal on the anterior wall of the main ventricular cavity (fig. 8) and some fibers anchored on the trabeculae of the main chamber. The presumed further progress of the fibers of the LBB to the papillary muscles was not followed.

Case 2

Case 2 (3813) was a 4-day-old white male. Anatomic diagnoses were: (1) single ventricle with inverted loop with inverted (L-) transposition, (a) subaortic stenosis with hypoplasia of the ascending aorta, (b) widely patent ductus arteriosus, (c) hypertrophy of the ventricular mass, (d) bialtrial hypertrophy, (e) aneurysm of an aortic sinus of Valsalva.

Course of the Conduction System

The SA node was in normal position. The regular A-V node was well developed but abnormal in shape and it ended blindly without forming an A-V bundle. The roof of the right atrium from the atrial septum to the trabeculae of the right atrial appendage was occupied by nodal tissue. More distally this nodal tissue divided into two parts (fig. 9). The part adjacent to the pulmonary artery formed a more distinct nodal...
structure (the anterior accessory A-V node) (fig. 10). Many of the cells of this structure were vacuolated and in the structure there was a small zone of cartilage. This node gave rise to the A-V bundle. The part close to the right atrial appendage resembled the junctional tissue between A-V node and A-V bundle. This gave rise to Mahaim fibers which joined (fig. 11) the main ventricular mass, and then this structure terminated. The blood supply to the accessory A-V nodal region came from an anterior artery, which was not followed to its origin. The bundle passed horizontally to the left in the dense connective tissue adjacent to the pulmonary annulus. It thus lay subendocardially in the main ventricular chamber on the septal wall between the main ventricular cavity and the outflow chamber. It increased in size, and adjacent to the defect it bifurcated. The RBB journeyed through the septum to the trabecular portion of the outflow chamber. The larger LBB passed caudal on the anterior wall of the main ventricular chamber.

Case 3

Case 3 (2029) was a 10-week-old white male. Anatomic diagnoses were: (1) single ventricle with inverted loop and inverted (L-) transposition, (a) left A-V valve stenosis, (b) coarctation of the aorta, (c) hypertrophy of the ventricular mass, (d) bialtrial hypertrophy, (e) patent ductus arteriosus.

Course of the Conduction System

The SA node was in normal position. No regular A-V node could be found. An accessory (anterior) A-V node arose in the roof of the right atrium adjacent to the septum and close to the pulmonic annulus. The blood supply to the A-V node came from an anterior vessel, which was not followed to its origin. The node pierced the annulus of the pulmonic valve on the right side, thus becoming the A-V bundle. The A-V bundle proceeded to the left, in the connective tissue adjacent to the pulmonic annulus. Distally it came to lie on the wall between the outflow chamber and main ventricular chamber. Adjacent to the defect between these two chambers on the anterior wall of the main ventricular chamber it divided into right and left bundle branches. The RBB passed horizontally through the septum adjacent to the defect to reach the endocardial surface of the outflow chamber. Here it descended to the trabecular area of the outflow chamber.
chamber where it terminated. The LBB descended caudad on the anterior wall of the main ventricular chamber. It was not followed to its termination.

Discussion

Mönckeberg, who pioneered in the studies of the conduction system in congenital heart disease, stated that in “cor biaatriatum triloculare” the A-V node originates in its usual position, and then forms a bundle of His which passes along a ridge on the posterior wall of the main ventricular mass where it divides into right and left branches. We are unable to tell from his description whether he refers to single ventricle, common ventricle or both.

While this work was in progress, Anderson et al. reported the course of the conduction system in 14 cases of single ventricle, seven with regular (D-) transposition and seven with inverted (L-) transposition. In both types, they found that the regular (posterior) A-V node was rudimentary, and did not

Figure 5

Case 1. Bundle of His (arrows) in main ventricular chamber adjacent to pulmonary valve. Weigert-can Gieson stain × 7.5. PV = pulmonary valve, MVC = main ventricular chamber.
Conduction System in Single Ventricle

Figure 6
Case 1. Bifurcation. Hematoxylin-eosin stain × 26. RBB = right bundle branch, LBB = left bundle branch, S = septum between main ventricular chamber and outflow chamber, MVC = main ventricular chamber.

Form a bundle of His. Instead, an accessory (anterior) A-V nodal structure was present at the junction of the atrial septum and the right atrial appendage, adjacent to the pulmonary trunk. This pierced the fibrous ring alongside the pulmonary valve ring to form the bundle of His. This entered the main ventricular chamber and came into contact with the right rim of the foramen between the main and small outflow chamber. Here it bifurcated into right and left bundle branches.

Our findings confirm and extend those of Anderson et al. in single ventricle with inverted loop and inverted transposition. In addition to their findings, in two of our cases the roof of the right atrium was

Figure 7
Case 1. Right bundle branch (arrows) passing through the septum. Hematoxylin-eosin stain × 30.

Figure 8
Case 1. Left bundle branch (arrows) passing caudal in the main ventricular chamber. Weigert-van Gieson stain × 5. MVC = main ventricular chamber, SOC = small outlet chamber, S = septum between both chambers, RBB = right bundle branch.
diffusely altered to form nodal-like tissue. In this tissue, in one case the accessory node formed. In another, this accessory (anterior) node divided into two parts — one part forming the A-V bundle, while the other gave off Mahaim fibers to the ventricular myocardium. This might prove of importance in pre-excitation. Furthermore, the bifurcation may be close to the defect in the septum between the main chamber and the outlet chamber (cases 2 and 3) or removed from it as much as .5–1.0 cm (case 1).

The aberrant, anterior type of conduction system, with an anterior node forming a bundle of His passing into the anterior part of the septum and dividing here into right and left bundle branches was described by Lev et al. in a case of mixed levocardia with ventricular inversion with inverted (corrected) transposition and with ventricular septal defect and fetal coarctation. This type was again found in a number of cases of corrected transposition by Anderson et al. Somewhat similar but not identical types of conduction systems were found by Mönckeberg in two cases. In one case of cor biloculare the main A-V bundle was interrupted from atrium to ventricle. An accessory bundle arose in the left atrial wall adjacent to

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to the left A-V orifice near the aorta. It passed along the left side of the aorta to join the subendocardial portion of the left ventricle where it divided into two branches. In a case of mixed levocardia with ventricular inversion, with both arterial trunks arising from the inverted right ventricle, with a rudimentary left ventricle and conus stenosis, he also found an anterior accessory connection, which however joined the main posterior connection to form a structure which then proceeded downward into the ventricles.

To understand aberrant types of conduction systems, the development of the normal A-V conduction system should be considered. There is considerable controversy as to this development. Mackenzie\(^1\) believed the A-V node to be derived from the sinus venosus, while the A-V bundle arose from the atrial canal. Koch\(^12\) and Aschoff\(^13\) thought that only the posterior part of the A-V node arose from the sinus, while the anterior part of the node and the bundle were derived from the atrial canal. Patten\(^14\) suggested that the A-V node is developed from the myocardium of the left sinus horn. Recently Anderson\(^11\) suggested that the proximal part of the A-V node arises from the sinus venosus, while the distal part comes from the atrial canal.

Most other authors,\(^15\)\(^-\)\(^20\) however, believe that the A-V node and bundle originate from the atrial canal musculature. According to these authors, they originate from the posterior part of the canal which lies behind the posterior endocardial cushion at a time when the musculature of this canal is still unbroken. There is a difference in opinion as to whether both the A-V node and bundle originate \textit{in situ},\(^18\)\(^,\)\(^20\) or whether the bundle originates from a proliferation of A-V nodal tissue.\(^19\) There is also a difference of opinion as to whether the bundle branches originate \textit{in situ} from ventricular trabeculae,\(^18\)\(^,\)\(^20\) or whether they originate from a proliferation of the tissue of the bundle of His.\(^19\) Anderson\(^16\) has recently voiced the opinion that they originate \textit{in situ} from the anterior (bulboventricular) part of the ventricular septum. Anderson has also advanced the concept that the entire atrial part of the atroventricular junction in the normal heart may be considered as part of the conduction system.\(^15\)

With this embryologic development of the conduction system in mind, regardless of the controversy as to details, it is not surprising that under certain abnormal circumstances, an anterior type of conduction system as the one found in single ventricle might develop. This might become possible when there is no posterior ventricular septum or it is displaced, or the atria and ventricles are discordant. Under these circumstances the musculature of the anterior part of the atrial canal has the potential to elaborate an A-V nodal structure, which in turn stimulates the production of an aberrant A-V bundle and bundle branches.

In previous work, Lev et al.\(^5\) and Van Praagh et al.\(^21\) suggested that the elaboration of the course of the conduction system in single ventricle might help to delineate the nature of this anomaly. This has proven not to be the case, as seen by our work and that of Anderson et al.\(^7\) In single ventricle, in one view,\(^21,\)\(^22\) the main ventricular chamber is a true left ventricle. This means that the septum between this chamber and the outflow chamber is the true ventricular septum. A second view\(^9\) is that the main ventricular chamber is the primitive descending limb of the bulboventricular loop reinforced by a part of the ascending limb, with the outflow chamber representing only the remainder of the ascending limb. Under these circumstances, the septum would then be an anterior (bulbar and metaampullar) septum and the posterior septum would be absent. As pointed out by Anderson et al.,\(^7\) from the findings in the conduction system one may interpret the septum either way.

\begin{figure}
\centering
\includegraphics[width=0.5\textwidth]{conduction-system.png}
\caption{Diagrammatic sketch of the course of part of the conduction system in case 2. Black line indicates course of bundle of His and left bundle branch. Dotted line indicates right bundle branch passing through wall between main and outflow chambers. RAV = right atroventricular valve, LAV = left atroventricular valve, A = anterior wall, P = posterior wall.}
\end{figure}

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There may be a true ventricular septum which is pushed aside, and because of this deviation an aberrant conduction system is formed. Or there may be an absence of the posterior portion of the ventricular septum and because of this, an anterior node makes connection with a bundle and bundle branches on the anterior septum. Thus, the conduction system does not prove either supposition as to the nature of the ventricular septum.

The surgical implications of this type of conduction system in single ventricle are clear (fig. 12). The surgeon must try to avoid the right side of the pulmonary valve annulus where the penetrating A-V bundle lies. In addition the area from the above point on the anterior wall to the area near the defect in the septum should be handled carefully as the remainder of the bundle and bifurcation are situated here. Recording from the His bundle as done by some surgeons might be helpful."

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