The Conduction System in Hypoplasia of the Aortic Tract Complex

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SUMMARY This is a study of the course of the conduction system in two cases of hypoplasia of the aortic tract complex, one with mitral stenosis and the other with mitral atresia. In both there was a posterior atrioventricular (AV) node which formed the AV bundle. In case 1 the bundle was short and bifurcated early. The branching bundle gave off a large left bundle branch (LBB), many Mahaim fibers to the septum and a small right bundle branch (RBB). In case 2 the AV node was divided into two parts which formed two posterior bundles which joined together to form a short branching bundle. Instead of a LBB there were profuse Mahaim fibers passing from the branching bundle to the depths of the ventricular septum. The RBB was large. The abnormalities seen in the conduction system particularly in the LBB are discussed from the embryologic standpoint.

THE CONDUCTION SYSTEM in hypoplasia of the aortic tract complex\(^1,2\) has, to our knowledge, not been described. We studied the conduction system in this complex to ascertain what happens to the left bundle branch when the left ventricle is small, thick-walled and accompanied by fibroelastosis, as in this complex with aortic atresia and mitral stenosis, or markedly atrophic, as in this complex with aortic and mitral atresia.

Case 1

This was a 4-day-old infant. We took no ECG, but there was no clinical evidence of atrioventricular block. The anatomic diagnosis was hypoplasia of the aortic tract complex with aortic atresia and mitral stenosis (fig. 1) with hypertrophy and enlargement of the right atrium and ventricle, a small left atrium with thick wall, a small left ventricle, thick wall and fibroelastosis, an atrial septal defect, fossa ovalis type, and a widely patent ductus arteriosus.

Histologic Examination

Conduction System

Methods. The sinoatrial (SA) node and its approaches were serially sectioned as previously described, and all sections were retained. The posterior and anterior walls of the atria were then removed and the remainder of the heart was serially sectioned as previously described, and all sections were retained. Alternate sections were stained with hematoxylin and eosin and Weigert-van Gieson stains. In this manner 5343 sections were examined. The findings were compared with those found in normal newborn hearts.\(^5,6\)

Findings

SA node, and its approaches, atrial preferential pathways and approaches to the atrioventricular (AV) node. These were in normal position but markedly infiltrated with lymphoid cells.

AV node. This structure was in normal position.
There were no pathologic changes and Mahaim fibers were abundant.

AV bundle, penetrating portion. The bundle was in normal position in the central fibrous body and in the lower confines of the left ventricular-right atrial part of the pars membranacea. Fibrosis and vacuolization of cells were present in the distal part of the penetrating bundle. Copious Mahaim fibers were present.

AV bundle, branching portion. This structure lay on the left side of the summit of the ventricular septum beneath a thick mass of fibroelastic endocardium which extended from the short, thickened interventricular part of the pars membranacea. The bundle was short and the bifurcation occurred early (fig. 2). The size of the cells was normal for this age; however, there was marked vacuolization of cells at the bifurcation. Copious Mahaim fibers proceeded downwards (apically) into the septum from the branching bundle at the bifurcation.

Right bundle branch. The right bundle branch was small compared with the right ventricular mass, intramuscular in its entire course, and terminated as Purkinje cells. The size of the cells was normal for this age. Slight fibrosis was present in the periphery.

**Figure 1.** Case 1. A) Anterior view of the heart. B) Left ventricular view. A = ascending aorta; PT = pulmonary trunk; RA = right atrium; RV = right ventricle; SVC = superior vena cava; LV = left ventricle. Note the arrows pointing to the demarcation of the left ventricle by the anterior and posterior descending coronary arteries.

**Figure 2.** Case 1. Atrioventricular bundle at bifurcation. Weigert-van Gieson stain × 30. LBB = left bundle branch; RBB = right bundle branch; M = Mahaim fibers; F = endocardial fibroelastosis; V = ventricular septal myocardium.
Left bundle branch. The main left bundle was large (fig. 3) and consisted of Purkinje cells. The space around this structure was continuous with the greatly widened longitudinal spaces in the central area of the myocardium of the left ventricular portion of the septum. Within this combined space abundant Mahaim fibers streamed into the septum (fig. 3). The main left bundle then fanned out anteriorly and posteriorly to end in a thick layer of Purkinje cells (fig. 4) beneath the fibroelastic intima of the left ventricle.

Central fibrous body and pars membranacea. The tricuspid and mitral annuli joined to form the central fibrous body, which was markedly thickened by connective tissue. This was normally joined by the tendon of Todaro. The left ventricular-right atrial part of the pars membranacea was also thickened and shortened. The short, thick, fibrous prong from the proximal portion of the aorta joined this part of the pars membranacea.

Case 2

This was a 2-day-old male infant. The ECG showed right heart strain. The anatomic diagnosis was hypoplasia of the aortic tract complex with aortic and mitral atresia with hypertrophy and enlargement of the right atrium and ventricle, atrophy of the left atrium and minute left ventricle, an atrial septal
defect, fossa ovalis type, and a widely patent ductus arteriosus.

Histologic Examination

Conduction System

Methods. The whole heart was serially sectioned from the posterior wall anteriorly and all sections were retained. Alternate sections were stained with hematoxylin and eosin and Weigert-van Gieson stains. In this manner 2710 sections were examined. The findings were compared with the conduction system of normal newborn hearts, as in the previous case.

Findings

SA node and its approaches, atrial preferential pathways and approaches to the AV node. These structures were in normal position and there were no pathologic findings.

AV node. The AV node lay proximal to and to the right of the central fibrous body. In the beginning it showed slight fibrosis. It then divided into two parts (fig. 5), right and left, the right lying more proximally (upstream) than the left. The left part of the node formed Mahaim fibers which entered the left side of the ventricular septum.

AV bundle, penetrating portion. Each of these parts of the AV node formed a portion of the bundle of His in the central fibrous body (fig. 6). The two portions were then joined together by a bridge of bundle tissue and eventually fused into one bundle.

AV bundle, branching portion. This structure lay in the abbreviated and thickened left ventricular-right atrial portion of the pars membranacea. Posteriorly,
the branching bundle formed a continuous sheet of fibers passing down into the septum (fig. 7). These could be considered to be the posterior fibers of the left bundle branch, but they had the form of Mahaim fibers because they proceeded into the depths of the septum. The branching bundle then reached the bifurcation (fig. 8). The size of the cells was normal for this age.

Right bundle branch. This structure was broader in diameter (fig. 8) than normal for a heart of this age; the size of the cells, however, was normal. The course of the right bundle branch through the right side of the septum was also normal.

Left bundle branch. As mentioned above, the posterior part of the main left bundle branch consisted of fibers which proceeded downward from the branching bundle (fig. 7). In the beginning these fibers lay adjacent to the endocardium of the left ventricle, and then dipped into the wall of the septum. The cells of these fibers in the beginning were small like the AV bundle cells. Further down they became somewhat larger, with pale cytoplasm, but they were not typical of the large Purkinje cells. They became continuous with the cells of the left ventricle. In the middle of the septum, the fibers of the left bundle branch showed degenerative changes with large spaces and enlarged vascular channels. The innermost lining of the myocardial cells of the left ventricle was small and contained hemosiderin pigment. The anterior fibers of the left bundle branch were also small, more subendocardial in the beginning, and then became intramyocardial. None of these cells could be called Purkinje cells.

Central fibrous body and pars membranacea. The central fibrous body was formed by the tricuspid and mitral anuli. The central fibrous body became continuous with thick fibrous tissue which projected into the left atrium in cyst-like formations containing
blood. These formations became continuous with the small lumen of the left ventricle. There was no mitral orifice. The central fibrous body continued as a thick left ventricular portion of pars membranacea. The aorta sent a long prong of collagenous tissue to join this part of the pars membranacea. There was no interventricular part of pars membranacea.

Discussion

In 1952 and 1953 we described the entity “hypoplasia of the aortic tract complex,” and divided this group of hearts into 1) isolated hypoplasia of the aorta, 2) hypoplasia of the aorta with ventricular septal defect, and 3) hypoplasia of the aorta with stenosis or atresia with or without mitral atresia. In 1966 we modified our concept, excluding the first two types so that the entity consisted only of the type with aortic atresia or severe aortic stenosis with mitral stenosis or atresia with intact ventricular septum. Many cases have been reported by other authors before and after our work. Case 1 in this paper had aortic atresia and mitral stenosis and case 2 had aortic and mitral atresia; both had intact ventricular septum.

Both of these cases show changes in the conduction system. The most remarkable are those in the branching portion of the bundle of His and the left bundle branch. In both cases, the branching bundle sends more abundant Mahaim fibers into the ventricular septum than is normal for this age, especially in case 2 (fig. 9). In case 1 the branching bundle gives off a distinct left bundle branch which ends in a wide layer of Purkinje cells. In case 2 the Mahaim fibers and the left bundle branch cannot be differentiated. Both posteriorly and anteriorly in case 2, the fibers proceed downward as a broad sheet from the branching bundle like profuse Mahaim fibers connecting to the ventricular myocardium. Some of these fibers become subendocardial, while others enter the depths of the left ventricular portion of the septum. We found no distinct Purkinje cells in the periphery of the left bundle branch in this case.
There is also a difference in the right bundle branch in both cases. In case 1 it is small in diameter compared with the right ventricular muscle mass. In case 2 the right bundle branch is very large in diameter throughout its course. The size of the cells of these structures, however, is about the same as that of the cells of the right bundle branch for this age.

What is the explanation for the abnormalities of the left bundle branch in these two cases? A possible explanation may be obtained from the work of Viragh and Challice. According to these authors, the AV bundle arises in situ, and is first widely connected to the summit of the ventricular septum. Only secondarily does fibrous tissue grow in and separate the bundle from the developing ventricular septum. Only secondarily does fibrous tissue grow in and separate the bundle from the developing ventricular septum, normally leaving a small amount of fibers as the only connections of the bundle to the summit of the ventricular septum in some cases. These may or may not disappear. These are the fibers originally described by Mahaim and reviewed by Lev and Lerner in human hearts.

Where there is a severe abnormality in the formation of the left ventricle, as in hypoplasia of the aortic tract complex, there may be persistence of the connections of the bundle to the ventricular septal musculature with copious Mahaim fibers remaining (case 1), and complete continuity of the bundle with the septum (case 2) as in fetal life. Where there is no mitral atresia and hence a partially functioning left ventricle in fetal life as in case 1 (albeit with retrograde functioning, i.e., from left ventricle to left atrium), then the left bundle branch may develop a Purkinje system peripherally. Where there is, in addition, mitral atresia and hence a nonfunctioning left ventricle, there may be an inhibition in the formation of the peripheral Purkinje network on the left side, with the Mahaim fibers remaining as the left bundle branch (as in case 2) with no formation of a Purkinje.
network. In the latter case the right bundle branch might be larger, since it would contribute to the conduction of most of the ventricular muscle mass, there being no Purkinje network on the left side. In contrast, in case 1, without mitral atresia, the right bundle branch would be normal in size, since the left bundle branch had developed a Purkinje network. The large size of the right bundle branch in case 2 is not due to an increased size of the cells, but to an increase in the number of cells.

In case 1 there are degenerative changes in the bundle of His. In case 2, the AV node lies proximal to and to the right of the central fibrous body. It divides into two parts, each forming part of a bundle of His. Both parts then coalesce to form one penetrating portion of the AV bundle. These findings are very likely related to the aortic or mitral atresia and the abnormally formed central fibrous body is probably related to the atresias. The changes in the central fibrous body may produce changes in the AV node and bundle of His with fibrosis, fragmentation, and degenerative changes in these structures. This might be enhanced by the coronary ischemia thought to be present in hypoplasia of the aortic tract complex. We do not know the explanation for the lymphoid cell infiltration in the SA node and its approaches and in the atrial preferential pathways in case 1.

The conduction system in this entity is the posterior type seen normally and in most types of congenital heart disease. This is in contrast to the anterior type seen in single ventricle and in some cases of corrected transposition. Thus, from the standpoint of the conduction system hypoplasia of the aortic tract complex is not related to single ventricle type of hearts.

The electrocardiographic abnormalities related to the abnormalities in the conduction system in this entity are only partly known. No ECG was taken in case 1. In case 2, the electrocardiographic diagnosis was marked right heart strain with no evidence of AV block. In other cases various degrees of AV block have been found, which might reflect changes in AV bundle similar to those seen in our case 2. We have seen only one case of Wolff-Parkinson-White syndrome in hypoplasia of the aortic tract complex (unpublished observations) despite the presence of the numerous Mahaim fibers seen in our presently reported cases.

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