The Conduction System in Tricuspid Atresia With and Without Regular (d-) Transposition

SAROJA BHARATI, M.D., AND MAURICE LEV, M.D.

SUMMARY This is a serial section examination of conduction system in six hearts with tricuspid atresia. Four had regular (d-) transposition and two did not have transposition. The conduction system was more or less the same in all the hearts. The atrioventricular (A-V) node was in the normal position posteriorly and was short. The A-V bundle was situated in the left ventricular aspect of the subendocardium and passed posteriorly to the ventricular septum. It entered the posterior wall of the coronary sinus, taking in the adjacent part of the right and left atrial walls and the proximal (upstream) part of the ventricular septum and the adjacent posterior wall of the left ventricle and the entire right ventricle. These blocks were fashioned and serially sectioned at various angles to the pulmonary valve anulus more or less in the Mahaim fashion. In case 2 it was completely serially sectioned and all sections were retained. In case 4 every seventh, in case 5 and 6 every fifth, in case 4 and 5 every tenth section was retained. Cases 1 and 2 were alternately stained with hematoxylin-eosin and Weigert-van Gieson stains, while cases 3-6 were stained consecutively with hematoxylin-eosin, Weigert-van Gieson and Gomori trichrome stains. In this manner 670 sections were examined in case 1, 3019 in case 2, 572 in case 3, 273 in case 4, 324 in case 5 and 466 in case 6.

Findings

Case 1

Gross Diagnosis

This patient had tricuspid atresia without transposition (fig. 1). Other defects included atrial and ventricular septal defects, a small right ventricle, and hypertrophy of both atria and left ventricle. The ventricular septal defect was situated beneath the right and noncoronary cusps. It measured 0.8 cm at its greatest dimension. It was not confluent with the aorta. It entered the right ventricle beneath the arch (crista) formed by the septal and parietal bands.
Microscopic Examination

The central fibrous body was formed by fibrous projections from the mitral and aortic anuli. This was joined by a fibrous prong coming from the puckered area in the right atrium. The central fibrous body extended onto a fibrous area beneath the aorta (fig. 2a). More anteriorly it extended onto a longitudinal ridge of fibrous tissue covered by ventricular muscle on the left side. The approaches to the A-V node were normal. The atrioventricular (A-V) node originated adjacent to and on the right side of the central fibrous body and its fibrous extension (fig. 2a). It entered the lower part of the central fibrous body to form the A-V bundle. The bundle thus lay on the left side of the ventricular septum a considerable distance from the aorta (fig. 2b). It traveled on the left side of the summit of the ventricular septum posterior to the defect. The posterior fibers of the left bundle branch were given off continuously (fig. 2b). The bundle then bifurcated into an anterior portion of the left and right bundle branch in the distal portion of the defect (fig. 2c). The right bundle branch went through the septum below the distal part of the defect to become subendocardial on the right side. It terminated in the trabeculated portion of the right ventricle. The fibers of the left bundle branch were not followed to their termination.

Case 2

Gross Diagnosis

Tricuspid atresia without transposition was present. Atrial and ventricular septal defects, a small right ventricle, and hypertrophy of left atrium and left ventricle were associated anomalies. In addition, a Blalock-Taussig procedure was noted. The ventricular septal defect measured 0.1-0.2 cm in its greatest length. It lay posteriorly in the septum adjacent to the mitral anulus and below the noncoronary aortic cusp, a small distance from the aortic anulus. It entered the right ventricle below the arch in the abbreviated sinus.

Microscopic Examination

The central fibrous body consisted of the fibrous junction of the anulus of the mitral valve with a fibrous extension from the aortic valve with a small contribution from the atrophic remnant of the anulus of the tricuspid valve. The SA node showed no changes. The approaches to the A-V node were normal. The A-V node arose from the right atrium at its junction, with the right side of the posterior part of the central fibrous body some distance from the fibrous prong contributed by the aortic anulus. The A-V node was partly enclosed in the central fibrous body. It penetrated the

Figure 1. Case 1. Tricuspid atresia without transposition. A) Right atrial view showing the position of the A-V node. B) right ventricular view showing the course of the right bundle branch. C) left ventricular view showing the course of the A-V bundle, the left bundle branch and the beginning of the right bundle branch. Abbreviations: ASD = atrial septal defect; RA = right atrium; AVN = A-V node; PT = pulmonary trunk; RV = right ventricle; D = ventricular septal defect; PM = papillary muscle; AVB = A-V bundle; LBB = left bundle branch; RBB = right bundle branch; LV = left ventricle; AO = aorta.
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Bharati, Lev

posterior part of the latter and was not related to the aortic anulus. There was no pars membranacea. The ventricular septal defect was an irregular channel lined by elastofibrous tissue which narrowed the lumen. The bundle lay here at the junction of the posterior and septal walls, to the left and posterior but close to the defect. At the distal wall of the defect it divided into right and left bundle branches. The right bundle branch traveled in the distal wall of the defect to

Figure 2. Case 1. Photomicrographs depicting the A-V node, A-V bundle and bundle branches. Weigert-van Gieson stain × 7.5. A) This shows the A-V node in a posterior position close to the tendon of Todaro. B) This shows the A-V bundle removed some distance from the aorta and giving off some fibers of the left bundle branch. C) This shows the right and left bundle branch in the region of the ventricular septal defect. RA = right atrium; LA = left atrium; AO = aorta; CFB = central fibrous body; Tend Tod = tendon of Todaro; AVN = A-V node; RV = right ventricle; AVB = atrioventricular bundle; LBB = left bundle branch; D = ventricular septal defect; RBB = right bundle branch.
reach the endocardial surface in the trabecular region of the right ventricle distal to the mouth of the defect on the right side. There it enlarged somewhat and its cells became Purkinje cells. The left bundle branch became Purkinje cells which reached the myocardium concentrated at the base of the anterior and posterior papillary muscles.

Case 3

Gross Diagnosis (fig. 3)

This patient had tricuspid atresia with regular (d-) transposition associated with bialtrial and left ventricular hypertrophy, ventricular septal defect, atrial septal defect (fossa ovalis type), patent ductus arteriosus, and abnormal architecture of the left ventricle. The ventricular septal defect measured 0.6 cm at its greatest dimension and was situated anteriorly, a slight distance from the left anterior pulmonary cusp. It entered the right ventricle beneath the arch formed by the septal and parietal bands.

Microscopic Examination

The SA node was in normal position. The approaches to the A-V node were normal. The A-V node originated from the right atrial musculature and to the right side of the central fibrous body formed by connective tissue from the mitral valve anulus and a prong of connective tissue from the pulmonary trunk (fig. 4a). The node was short. It pierced the central fibrous body to become the A-V bundle. The latter lay on the left side of the ventricular septum some distance from the pulmonary trunk. The left bundle branch was given off in small groups of fibers. The bundle lay posterior and to the left of the defect (fig. 4b). At the distal wall of the defect the bundle gave off the remaining fibers of the left bundle branch and the right bundle branch (fig. 4c). The latter went to the trabecular area of the right ventricle which extended far down the wall of the septum (fig. 4b).

Case 4

Gross Diagnosis

This heart represented tricuspid atresia with regular (d-) transposition with absence of the transverse arch. Bialtrial and left ventricular hypertrophy, atrial septal defect (fossa ovalis type), a ventricular septal defect, and a patent ductus arteriosus were also present. The greatest dimension of the ventricular septal defect was 0.5 cm. It was situated anterior, some distance from the pulmonary anulus beneath the junction of the left and right anterior cusps. It entered the right ventricle beneath the arch or crista.

Microscopic Examination

The SA node was normal in position. The central fibrous body was made up of the junction of the mitral anulus, a small spur representing the tricuspid anulus and a prong of

Figure 3. Case 3. Tricuspid atresia with regular (d-) transposition. A) This is a left ventricular view showing the course of the bundle and the left bundle branch. B) This is a right ventricular view showing the course of the right bundle branch. Abbreviations: PT = pulmonary trunk; D = ventricular septal defect; MV = mitral valve; B = A-V bundle; LBB = left bundle branch; AO = aorta; RV = right ventricle; RBB = right bundle branch.
FIGURE 4. Case 3. Tricuspid atresia with regular (d-) transposition. Photomicrographs showing the A-V node, A-V bundle and bundle branches. Weigert-van Gieson stain (A) × 7.5. This shows the position of the A-V node adjacent to the central fibrous body and close to the tendon of Todaro. B) × 7.5. This shows the position of the A-V bundle in relationship to the ventricular septal defect. C) × 15. This shows the right bundle branch passing into the right ventricle and the left bundle branch passing downwards into the left ventricle. Abbreviations: Ao = aorta; PT = pulmonary trunk; Tend Tod = tendon of Todaro; CFB = central fibrous body; MV = mitral valve; AVN = A-V node; AVB = A-V bundle; D = ventricular septal defect; RVS = sinus of right ventricle; LBB = left bundle branch; RBB = right bundle branch; RV = right ventricle.
connective tissue from the pulmonic anulus. The approaches to the A-V node, which originated from the right atrium close to the coronary sinus and the tendon of Todaro, were normal. The node pierced the junction of the spur of the tricuspid anulus with the central fibrous body, entering the myocardium of the left ventricle to become the A-V bundle. This bundle then descended into the ventricular septum on the left side. The bundle entered the subendocardium posterior to the defect and continued at this level between the defect and the mitral anulus. It then turned toward the apex, posterior to the defect. The right bundle branch passed subendocardially in the left ventricle near the distal wall of the defect and entered the right ventricle where it became very large and broke into numerous cords of Purkinje cells in the trabecular portion of the right ventricle. The left bundle branch proceeded toward the apex and was not followed to its termination.

Case 5

Gross Diagnosis

This was a case of tricuspid atresia with regular (d-) transposition. The heart also showed right aortic arch, fetal (predual) coarctation, bialtral and left ventricular hypertrophy, atrial septal defect (fossa ovalis type), ventricular septal defect, patent ductus arteriosus (right-sided), and juxtaposed atrial appendages. The ventricular septal defect measured 1 cm in greatest dimension. It lay anteriorly, beneath the right and posterior pulmonary cusps, a small distance from the pulmonary valve. It entered the right ventricle beneath the arch or crista formed by the septal and parietal bands.

Microscopic Examination

The central fibrous body was formed by an extension of the mitral anulus with a portion of tricuspid anulus going to the epicardium on the right side, and a fibrous extension of the pulmonic anulus. The SA node was in the normal position. The approaches to the A-V node were normal. The A-V node originated in the right atrium on the right side of the central fibrous body. It penetrated the posterior part of the central fibrous body to become the bundle of His, which now lay on the left side of the ventricular septum. The bundle migrated anteriorly and now lay posterior to the ventricular septal defect. Along its journey it constantly gave off fibers of the left bundle branch. It then bifurcated into a left anterior portion and the right bundle branch. The latter lay within the myocardium, then entered the subendocardium. It passed through the inferior part of the distal wall of the defect and into the septal band on the right side. It ended in the trabecular portion of the right ventricle. The fibers of the left bundle branch were not followed to their termination.

Case 6

Gross Diagnosis

The basic diagnosis of this case was tricuspid atresia with regular (d-) transposition. The heart also showed bialtral and left ventricular hypertrophy, ventricular septal defect, fetal coarctation, banding procedure, atrial septal defect (fossa ovalis type), and aneurysm of the fossa ovalis. The ventricular septal defect measured 0.5 cm in greatest dimension. This was situated far anteriorly, entering the right ventricle just below a shallow arch formed by the septal and parietal bands.

Microscopic Examination

The SA node was in the normal position. The central fibrous body consisted of the fibrous junction of the mitral and pulmonic anuli. The approaches to the A-V node were normal. A short A-V node emerged from the right atrium adjacent to the coronary sinus and the tendon of Todaro. It pierced the right side of the mitral anulus just before it formed the central fibrous body to become the bundle of His. The bundle then entered the left side of the myocardium of the septum and was intramyocardial. When it reached the ventricular septal defect, it lay posterior to the defect, where it gave off fibers of the left bundle branch. The right bundle branch was given off in the posterior wall of the defect and journeyed to the trabeculae of the right ventricle. The fibers of the left bundle branch were not followed to their destination.

Discussion

Our early morphologic studies have indicated that tricuspid atresia without or with regular (d-) transposition is not a form of single ventricle, because in tricuspid atresia the right-sided ventricular mass may be considered to be a true right ventricle. We define a right ventricle as that chamber having both an infundibulum with septal and parietal bands, and a trabeculated area beneath these muscles which we call sinus. If we accept this definition then there is both a sinus and infundibulum in this chamber in tricuspid atresia in almost all cases, although the sinus portion may not be apparent on gross examination in some cases with transposition. Usually the sinus portion can be identified in these cases microscopically (fig. 4b). Even though this area may not have a tricuspid tensor apparatus, its trabecular nature and its position warrant its being labeled a sinus of the right ventricle. Furthermore, occasional papillary muscles are sometimes found in the sinus (fig. 1). The left-sided ventricular mass resembles the true left ventricle, although there are some abnormalities in its structure, especially with transposition.

A corollary to this concept is the existence of a true ventricular septum consisting of anterior (bulbar-conus) and posterior (main) portions. This is attested to by the fact that the anterior and posterior descending coronary arteries usually meet at the apex, as in the normal heart. The presence of the anterior and posterior descending coronary arteries meeting at the apex usually implies that there is an anterior and posterior ventricular septum. As we have previously demonstrated, in most cases of single ventricle, it is usually difficult to designate any vessel as anterior or posterior descending. The small outlet chamber is usually demarcated by what we have called right and left demarcating arteries which are short and do not join.

Our present work on the conduction system lends credence to the above notions. The peripheral conduction system normally consists of the approaches to the A-V node, the A-V node, the bundle and bundle branches. The approaches to the A-V node are situated in the region of the coronary sinus and
the tendon of Todaro. The A-V node is situated on the right side of the central fibrous body. The A-V node pierces the central fibrous body to become the penetrating portion of the bundle of His. The latter then lies in the lower confines of the pars membranacea and eventually becomes the branching portion which gives off fibers of the left and right bundle branch.

In tricuspid atresia with or without transposition, the central fibrous body is abnormally formed since there is no, or only a remnant of tricuspid anulus and a remnant or no pars membranacea. The fibrous extension anteriorly of the central fibrous body seen in some cases may be considered to be a pars membranacea (figs. 1c and 3a). The A-V node and its approaches, however, are still seen to be in the region of the coronary sinus and tendon of Todaro. The node pierces the abnormal central fibrous body to form the bundle of His. The His bundle may course through the myocardium on the left side or immediately become subendocardial on the left side. This is true whether there is or is not regular (d-) transposition.

In single ventricle with regular (d-) transposition Anderson et al. found an anterior A-V node in the roof of the right atrium with a bundle which passed through the pulmonic anulus.\(^5\) We did not find this in any of our cases in tricuspid atresia, whether or not transposition was present. Our work confirms the findings of Titus and Guller\(^6\) in cases of tricuspid atresia without transposition. Hence our work emphasizes that there are not only gross anatomic differences but also histologic conduction system differences between tricuspid atresia on the one hand, and single primitive ventricle on the other, supporting the concept that tricuspid atresia and single ventricle should continue to be regarded as separate entities.

The course of the bundle of His in tricuspid atresia is to a certain extent related to the location of the ventricular septal defect as is true in any case with ventricular septal defect. The more anterior the defect, the further to the left and away from the defect is the bundle. Usually, the defect is more posterior in cases without transposition than with transposition, although there are variations with and without transposition. Thus the bundles in cases 1 and 2 are closer to the defect in its course than in cases 3–6 (compare figs. 1c with 3a).

We have chosen to designate the junction of the extension of the pulmonic or aortic anulus to the mitral anulus as a central fibrous body in the context of the absence of a well-formed tricuspid anulus. We think this is proper since in some cases of tricuspid atresia a small portion of tricuspid anulus is present (cases 2, 4 and 5). Furthermore, embryologic development theory suggests that the anterior and posterior endocardial cushions do meet and join in the formation of a central fibrous body in tricuspid atresia as they do normally, since there is a more or less normally formed mitral valve.

These findings in tricuspid atresia with transposition may have some surgical importance. In a previous work we have pointed out that a Fontan-like or modified procedure may be of use in patients with this anomaly.\(^1\) In this type of treatment, the pulmonary trunk may be closed and a bypass procedure may be attempted between the right atrium and the pulmonary trunk. Under these circumstances the left and right ventricle would be used to convey blood to the aorta. It may be necessary to enlarge the ventricular septal defect in some of these patients. Since the A-V bundle lies posterior to the defect on the left side and the right bundle branch passes through the distal wall of the defect, injury in either of these regions should be avoided.

We have previously suggested that whenever the aorta is located to the right, and anterior of its normal anatomic position, there is a tendency for the A-V bundle to move further toward the left side of the ventricular septum than normal. This we believe is true of tetralogy of Fallot,\(^9\) double outlet right ventricle,\(^10\) and regular (d-) transposition.\(^11\) The present study shows that this generalization also applies to cases of tricuspid atresia with regular (d-) transposition.

References

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