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The Conduction System in Double Outlet Right Ventricle with Subpulmonic Ventricular Septal Defect and Related Hearts (The Taussig-Bing Group)

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SUMMARY The Taussig-Bing group of hearts is that group in which the aorta emerges completely from the right ventricle, unrelated to the position of the ventricular septal defect (VSD), while the pulmonary trunk emerges at varying degrees from the right ventricle related to the location of the VSD. Several types of surgical procedures involve the VSD in these hearts. The conduction systems in six of these hearts were therefore studied.

The atrioventricular (A-V) bundle lies in the inferior (posterior) wall of the defect on the left side beneath the summit of the ventricular septum. In some cases, the bundle may be proximal to the defect, thus lying intramuscularly, and only the right and bundle branches are related to the defect.

The pattern of development of the atrioventricular (A-V) node, bundle and bundle branches is related primarily the development of the endocardial cushions and posterior ventricular septum, and not to that of the absorption of the bulbus.

THERE IS A GROUP OF HEARTS in which the aorta emerges completely from the right ventricle, while the pulmonary trunk emerges either entirely from the right ventricle adjacent to a ventricular septal defect (VSD), or overrides a VSD arising in varying proportion from both ventricles. These have been called by us the Taussig-Bing group. The importance of this group lies in the observation that the surgical treatment of all its members may be similar — that is the Hightower-Kirklin procedure or some other type of internal conduit procedure. In the former procedure the pulmonary trunk is made to emerge from the left ventricle utilizing the VSD, thereby creating a complete transposition. This is then followed by the Mustard procedure. In the latter procedures the aorta is made to emerge from the left ventricle by means of a conduit.

It is self evident that to utilize these procedures a knowledge of the course of the conduction system is useful. An anatomic study of this system constitutes the essence of this paper.

Materials and Methods

In six Taussig-Bing hearts the sinoatrial (S-A) node, the approaches to the atrioventricular (A-V) node, the A-V node, the A-V bundle, and the bundle branches up to the level of the moderator band were serially sectioned in a manner previously described, with the exception of case 3. In the latter case, the sectioning was executed from the superior to the inferior wall of the defect. Every tenth section of the S-A node was retained, and every 5th section of the approaches to the A-V node, the A-V node, bundle and bundle branches. Consecutive sections were stained with hema-toxylin-eosin, Weigert-van Gieson and Golgi trichrome stains. In such a manner a total of 442, 1452, 414, 508, 556, and 478 sections were studied in cases 1-6, respectively.

The electrocardiogram of none of the cases revealed evidence of second or third degree A-V block. Cases 2 and 3 revealed first degree A-V block and case 1, complete right bundle branch block (RBBB).

Findings

Case 1

The patient was a 26-day-old male. Anatomic diagnosis was double outlet right ventricle with subpulmonic VSD, or right-sided Taussig-Bing heart with mild overriding (fig. 1).
Associated cardiac anomalies included atrial septal defect (ASD), fossa ovalis type, fetal coarctation, and patent ductus arteriosus.

Microscopic Examination

The central fibrous body was formed by the junction of the mitral and tricuspid anuli. There was no fibrous connection between the central fibrous body and the pulmonic or aortic valves. The pulmonic valve anulus had a wide fibrous connection with the mitral anulus, the aortic valve had a brief but thick fibrous connection with the tricuspid anulus.

Conduction System

The S-A node was in normal position. The A-V node arose in its normal position, but it was short. It penetrated the right side of the central fibrous body, and traveled through it to the left side, becoming progressively engulfed by ventricular musculature. It thus came to lie on the left side of the septum below the VSD. The penetrating bundle showed moderate fibrosis. The A-V bundle flattened out on the left side of the septum, and then gave off the left bundle branch (LBB). The right bundle branch (RBB) was absent.

Case 2

Anatomic diagnosis of this six and one-half year old female was double outlet right ventricle with subpulmonic VSD — Taussig-Bing complex, right ventricular type with mild overriding (fig. 2). Other anomalies included an abnormal mitral valve and abnormal architecture of the left ventricle.

Microscopic Examination

The central fibrous body was formed by the junction of the mitral and tricuspid anuli. The pulmonic valve anulus was connected to the tricuspid anulus and to the mitral anulus by separate connective tissue prongs.

Conduction System

The S-A node was in normal position. The A-V node commenced on the right side of the distal part of the atrial septum and was enveloped by the central fibrous body. It however maintained its connections with the atrial septum. It was short, and considerably infiltrated with mononuclear cells. It penetrated the right side of the central fibrous body to form the bundle of His. The latter was infiltrated with mononuclear cells and showed slight fibrosis. It then proceeded through the central fibrous body to the left and gradually became covered by ventricular musculature. It thus reached the level of the defect, with the mitral and tricuspid valves bridging the defect on both sides. At the level of the defect, the penetrating portion of the bundle lay on the left side of the septum below the defect, some distance from the summit. It now flattened out and moved toward the summit (fig. 3). The bifurcation was reached rapidly at the distal end of the defect. The RBB lay within the summit of the inferior wall of the VSD. It moved to the right and climbed to the lower part of the septal band where it continued to the moderator band. The RBB was infiltrated with mononuclear cells. The LBB was not followed to its termination.

Case 3

The anatomic diagnosis of this male infant, aged one month, five days, was double outlet right ventricle with subpulmonic VSD (Taussig-Bing complex, right ventricular type, with slight overriding). In addition, the heart showed

Figure 1. Case 1. Left panel] Right ventricular view. Outflow tract into aorta. Right panel] Right ventricular view. Outflow tract into pulmonary trunk. P1 = first parietal band; P2 = second parietal band; A = aorta; PT = pulmonary trunk; D = ventricular septal defect.
an abnormal mitral valve, a patent ductus arteriosus, surgically closed, coarctation, surgically corrected, and a banding procedure.

Microscopic Examination

The central fibrous body was formed by the junction of the mitral and tricuspid anuli aided by a prong of connective tissue from the pulmonary anulus. There was also pulmonic-mitral continuity.

Conduction System

The S-A node was in normal position. The A-V node arose in its usual posterior position in the right atrium. It entered the right side of the central fibrous body to become the penetrating bundle. It traveled through the central fibrous body to the left side. Here it came to lie on the left side of the posterior wall of the defect (fig. 4). At the distal wall of the defect it bifurcated into right and left bundle branches. The RBB passed through the distal portion of the inferior wall of the defect and went its usual course along the septal band. The LBB was not followed to its termination.

Case 4

This four and one-half month old female had the Taussig-Bing complex, intermediate type (fig. 5). In addition, ASD, fossa ovalis type, fetal coarctation, patent ductus arteriosus, surgically closed, and banding procedure were found in this heart.

Microscopic Examination

The mitral and tricuspid anuli formed the central fibrous body, with a prong of connective tissue from the pulmonary anulus contributing to the body.

Conduction System

The S-A node was in normal position but showed considerable hemorrhage and an infiltration of mononuclear cells. The A-V node arose in its usual position posteriorly and showed marked hemorrhage. This entered the right side of the central fibrous body where it showed zones of hemorrhage. As it passed through the central fibrous body to the left, it became covered with ventricular musculature. It thus reached the myocardium on the left side where it traveled along the left side of the inferior wall of the defect (fig. 6). The branching portion was very short and showed hemorrhage. It thus reached the bifurcation at the level of the distal part of the defect. Here the RBB passed through the infero-distal wall of the defect and proceeded in its usual way along the septal band. The LBB was very large. It showed marked hemorrhage. This bundle was not followed to its termination.

Case 5

This was a two month, 14 day old male with an anatomic diagnosis of Taussig-Bing complex, intermediate type. The heart also showed patent ductus arteriosus, fetal coarctation, and ASD, fossa ovalis type.
FIGURE 3. Case 2. Relationship of the A-V bundle to the ventricular septal defect. Weigert-van Gieson stain $\times$ 9.8. PT = pulmonary trunk; MV = mitral valve; V = ventricular septal defect; TV = tricuspid valve.

FIGURE 4. Case 3. Horizontal section of inferior wall of ventricular septal defect. Hematoxylin-eosin stain $\times$ 7.7. AS = atrial septum; VS = ventricular septum; N = A-V node; B = A-V bundle; B$_1$ = bifurcation; LBB = left bundle branch; RBB = right bundle branch; MV = mitral valve; TV = tricuspid valve.
Microscopic Examination

The central fibrous body was formed by a union of the mitral and tricuspid anuli aided by a prong of connective tissue from the pulmonic anulus. There was a small area of pulmonic-mitral fibrous continuity.

Conduction System

The S-A node was in normal position. The A-V node was in normal position posteriorly. It penetrated the right side of the central fibrous body to become the A-V bundle. It passed to the left side, lying in the myocardium before it became subendocardial. It thus came to lie on the left side of the posterior wall of the ventricular septum, proximal to the defect. The branching bundle was short. It then bifurcated into left and right bundle branches. The RBB moved through the septum to the right. Thus the left and right bundle branches came to lie on each side of the ventricular septum, some distance from the summit of the posterior wall of the defect (fig. 7). The RBB then proceeded along the septal band in its usual manner. The LBB was not followed to its termination.

Case 6

Left-sided Taussig-Bing was the anatomical diagnosis of this three month, 15 day old female (fig. 8). Her heart also revealed fetal coarctation, patent ductus arteriosus, surgically closed, banding procedure, and atrial septostomy (balloon).

Microscopic Examination

There was no connection between pulmonic anulus and the central fibrous body and no pulmonic-mitral continuity.
Conduction System

The S-A node was in normal position. There was an adjacent pericarditis. The A-V node originated in its usual posterior position. It entered the right side of the central fibrous body and became covered by musculature. As the bundle moved toward the left it came to lie within the myocardium (fig. 9). It was longer than usual. As it reached the left side it lay on the left side of the posterior wall of the defect, some distance from the latter. It then became the branching bundle, giving off fibers of the LBB. It then moved closer to the defect where the bifurcation took place. Near the distal wall of the defect the RBB passed through the myocardium to the right side (fig. 10). It proceeded in its usual manner along the septal band. The LBB was not followed to its termination.

Discussion

We have previously delineated a group of hearts which may be called the Taussig-Bing group. In these hearts, the aorta emerges completely from the right ventricle unrelated to the VSD with no aortic-mitral fibrous continuity, while the pulmonary trunk is related to the defect. The pulmonic trunk may emerge completely from the right ventricle (right-sided Taussig-Bing with no overriding), also called double outlet right ventricle with subpulmonic VSD. In the second type the pulmonic trunk arises mostly from the right ventricle with minimal overriding (right-sided Taussig-Bing with overriding), also called double outlet right ventricle with subpulmonic VSD. In the third type the pulmonic trunk may straddle the ventricular septum about 50% over the defect, (Taussig-Bing, intermediate type). A fourth variation is described by the pulmonic trunk emerging almost entirely from the left ventricle (left-sided Taussig-Bing).

In these hearts the central fibrous body is abnormally
formed. It may consist only of the junction of the mitral and tricuspid anuli, with no pulmonic or aortic contribution to this junction, or a pulmonic component may be present as a prong of tissue to help form the central fibrous body. On gross inspection the presence or absence of pulmonic-mitral fibrous continuity varies from case to case. In some cases in which no continuity was observed in the gross specimen, a varying area of such continuity is apparent on histological examination.

This concept of the Taussig-Bing group, regardless of varying terminology, has been upheld by recent workers. It has been objected to by Van Praagh on semantic grounds. According to Van Praagh the term Taussig-Bing should be applied only to those hearts in which the pulmonic trunk emerges from the right ventricle without pulmonic mitral fibrous continuity.

The course of the conduction system in all these types is similar (figs. 2, 8). The A-V node originates in its usual position between the coronary sinus and medial leaflet of the tricuspid valve. However, it may be deformed or engulfed by the central fibrous body. The node is often short. It penetrates the central fibrous body to form the penetrating portion of the A-V bundle. This may be unusually long, traveling through the central fibrous body and often through the ventricular septum to the left side where it comes to lie on the left side of the posterior wall of the VSD, at varying distances from the edge. At the distal part of the inferior wall the A-V bundle divides into right and left bundle branches, very often with little or no branching portion for the more posterior fibers of the LBB. In some cases the bifurcation occurs at the proximal portion of the inferior wall of the defect. At the distal portion of the inferior wall the RBB may come close to the defect, as it passes through the septum to the right side. The RBB then passes along the inferior part of the septal band to the moderator band. The LBB fans out as it proceeds downward but this path was not followed to its termination in this work.

All of the above types of hearts may be amenable to the Hightower-Kirklin or other procedure. In the former procedure the pulmonary trunk is transferred to the left ventricle by way of the VSD thus producing complete transposition. A Mustard procedure is then performed. In the former part of the procedure it is apparent that the right side of the defect is less vulnerable from the standpoint of the conduction system than is the left, with the exception of the RBB.

In the other surgical procedures, a tunnel is fashioned between the aorta and the VSD. Here there is a distinct danger of damaging the conduction system on the left side. Perhaps under these circumstances it might be useful to electrophysiologically locate the bundle. It is also clear that if it is necessary to enlarge the defect in any of these procedures this may be done at the expense of the distal (more apically situated) wall of the defect.

From the embryologic standpoint, the minor changes in the course of the conduction system in the Taussig-Bing group of hearts as compared to the normal is at least theoretically explainable. Normally the A-V node in man develops from the left side of the sinus venosus, according to some, or from the posterior part of the atrial canal, according to others, or from both, according to still others, at about 8 mm of fetal length. Hence, its definitive position is in the lower distal part of the atrial septum between the coronary sinus and the medial leaflet of the tricuspid valve. The A-V bundle (of His) develops either as an offshoot from the A-V node according to some or as an independent structure from atrial canal musculature behind the posterior endocardial cushion according to others. Hence its definitive position is in the lower confines of the central fibrous body and the pars membranacea, which originate from the endocardial cushions joined by connective tissue beneath the aorta after the absorption of the bulbus.

The bundle branches originate from the ventricular trabeculae in situ according to some or from a proliferation of tissue from the bundle of His according to others. This latter development differs on both sides. On the left side

FIGURE 8. Case 6. Left panel) Right ventricular view at outflow tract of aorta. Right panel) Left ventricular view. A = aorta; P1 = parietal band 1; P2 = parietal band 2; PT = pulmonary trunk; RBB = course of right bundle branch; LBB = course of left bundle branch; B = course of A-V bundle; D = ventricular septal defect.

it apparently takes place in the sinus musculature of the septum, while on the right side it develops on the summit of the posterior part of the ventricular septum and the sinus musculature of the right side of the septum. Hence the position of the main part of the left bundle branch is in the sinus of the left ventricular aspect of the septum, and that of the right bundle branch between sinus and conus musculature of the right ventricle. The Purkinje nets are in the sinuses of the two chambers and there are practically no Purkinje fibers in the coni of these chambers.

This difference in position and architecture in the right and left bundle branches, although they have never been fully explained, is, we believe, in some way related to the difference in the absorption of the bulbus on the left and right sides. On the left side the more or less total absorption of the bulbus into the ventricular myocardium results in a flattening out of the LBB over the sinus portion of the septum. On the right side however the sharp demarcation of the conus and sinus results in a RBB proceeding between sinus and conus and then terminating in the Purkinje cells of the sinus musculature.

In the Taussig-Bing group of hearts, we may postulate an abnormality in the absorption of the bulbus. One of its effects is the malformation of the central fibrous body. In this group of hearts, in some cases the pulmonary trunk sends a prong of connective tissue to the central fibrous body while the aorta is moved away from the central fibrous body. In others no connective tissue prong goes from either the aorta or pulmonary trunk to the central fibrous body. The pars membranacea in the Taussig-Bing group of hearts is almost completely replaced by muscle. Thus the penetrating bundle, after passing through the central fibrous body, lies encased in muscle on the left side of the septum instead of passing through the pars membranacea. This apparently makes the development of the branching bundle, which normally gives off the posterior LBB fibers, impossible since the latter are endocardial. Thus the bifurcation begins after the penetrating portion, and often after passage of the penetrating bundle through the myocardium.

This is only a minor variation from normal and points to the fact that the conduction system is related more to the development of the endocardial cushions and the posterior ventricular septum than to the absorption of the bulbus.

We have found the A-V bundle to be situated more to the left of the summit of the ventricular septum than normal in cases in which the aorta is shifted to the right as in tetralogy of Fallot, double outlet right ventricle with subaortic VSD, and with subpulmonic VSD and complete transposition. This conception has been denied by Titus et al. and by Visioli et al.

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Nitroprusside after Open-Heart Surgery

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SUMMARY The effects of intravenous infusion of sodium nitroprusside were studied in 11 children immediately after open-heart surgery for congenital heart disease. The patients were selected because, following bypass, their cardiac index was below 2.0 L/min/m² and their systemic vascular resistance exceeded 30 units.

INTRAOPERATIVE MYOCARDIAL DEPRESSION may occur following repair of congenital cardiac defects, especially after procedures that require significant intervals of interruption of coronary flow. The usual response to an inadequate cardiac output secondary to myocardial insufficiency is an increase in systemic vascular resistance.1 Vascular resistance is also frequently increased following the use of hypothermia during perfusion.2 Reduction of systemic vascular resistance by intravenous

infusion of sodium nitroprusside has been reported3 to improve the hemodynamic status of patients with congestive cardiac failure and with acute myocardial infarction. In response to nitroprusside infusion, Guiha et al.4 observed a prompt decrease in left ventricular filling pressure, an increase in cardiac output, and a fall in systemic vascular resistance in a group of patients with intractable cardiac failure caused by cardiomyopathy or ischemic heart disease. Chatterjee et al.5 noted an improved cardiovascular performance of patients who had an acute myocardial infarction and a left ventricular filling pressure of greater than 15 mm Hg during vasodilator therapy.

Palmer and Lasseter6 suggest sodium nitroprusside is an ideal agent to lower blood pressure because of its rapid onset of action, direct effect to relax vascular smooth muscle, few side effects, relative absence of toxicity in the usual thera-

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