

Taussig-Bing Heart with Mitral Valve Straddling

Case Reports and Postmortem Study

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SUMMARY

Seven autopsied cases of the Taussig-Bing Heart diagnosed at the Heart Institute of Japan were reexamined for this report. Three cases had been correctly classified according to the original description of the Taussig-Bing Heart. The remaining four cases provided examples of a rare variation of this malformation. Characteristic features included the following: the mitral valve straddled the ventricular septum in all cases; the chordae of the anterior leaflet crossed over the septum into the right ventricle through a large subpulmonary ventricular septal defect; and an abnormal muscle ridge, extending from the caudal part of the parietal band, covered the right ventricular side of the ventricular septal defect. Although these four cases differ anatomically from the original Taussig-Bing Heart, their hemodynamics are similar. We have called this anomaly the Taussig-Bing Heart with mitral valve straddling.

Additional Indexing Words:

Congenital heart disease Double outlet right ventricle Ventricular septal defect

VARIOUS TYPES of cardiac malformations are known to develop due to an abnormality in the origin of the two great arteries. Included in these is a type in which the aorta is transposed to the right, originating entirely from the right ventricle, while the origin of the pulmonary trunk is normal. This type is known as a double outlet right ventricle. A severe heart malformation known as the Taussig-Bing Heart¹ falls into this category.²⁻⁴ Before diagnostic techniques such as angiocardiology were widely available, these cases were only rarely diagnosed during the patient's lifetime and frequently only attracted the attention of highly specialized anatomists and pathologists. With recent developments in cardiovascular surgery, cases of severe heart malformations have begun to draw the clinician's attention and radical surgery can now be performed in patients with the Taussig-Bing malformation. The disease is now being extensively studied by pathologists, and successful operations have been reported by Hightower et al.,⁵ Thomson,⁶ Wedemeyer,⁷ and Kawashima.⁸

To further clarify the anatomy, seven autopsied

cases, which had been classified as the Taussig-Bing Heart at the Heart Institute of Japan, were reexamined. Three cases⁹ fit the original description of the Taussig-Bing Heart. The remaining four cases were found to represent a distinct although similar heart malformation. While the clinical symptoms and hemodynamics were similar to the findings in Taussig-Bing Heart, significant differences were found. Detailed pathological studies were made of these four cases and they have been classified as the Taussig-Bing Heart with mitral valve straddling.

Pathological Findings

All four patients died following surgery and postmortem examination was performed at the Institute. The surgical procedures included banding of the pulmonary artery in one patient. Two patients had undergone the Blalock-Hanlon procedure. A staged operation was planned in the remaining case, but the patient died following the initial Mustard procedure.

Case 1

Examination of the closed heart showed the aorta located to the right of and slightly anterior to the pulmonary trunk. The diameter ratio of the aorta to the pulmonary trunk was 2 to 3. The free wall of the right ventricle was noted to be markedly hypertrophied but its cavity was well developed.

The aorta, transposed to the right from its normal position, arose completely from the right ventricle (fig. 1). A well developed parietal band extended forward and backward between the aortic valve and the pulmonary valve. The pulmonary valve was located

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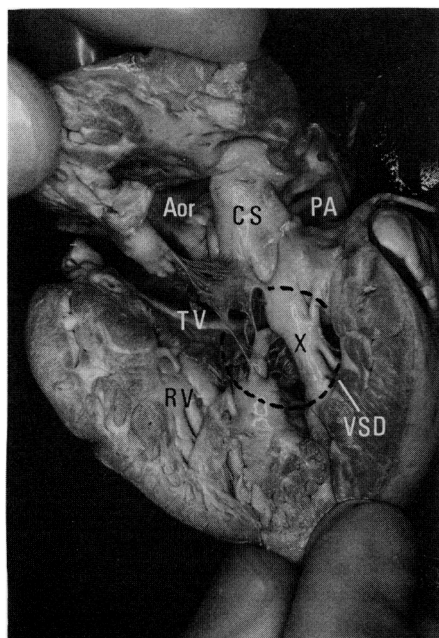


Figure 1

Right ventricular view of the Taussig-Bing Heart with mitral valve straddling (case 1). Aor = aorta; PA = pulmonary artery; CS = crista supraventricularis (parietal band); TV = tricuspid valve; VSD = ventricular septal defect; RV = right ventricle; X = muscle band extending from the tail of the parietal band (CS).

above the ventricular septal defect (VSD) and arose to the right of the septum without overriding. The VSD was situated in front of and above the crista supraventricularis and its diameter was as large as 35 mm. The right side of the ventricular septal defect was covered by the projection of a thick muscle band and thick anterior papillary muscle. The muscle band extended from the tail of the parietal band while the anterior papillary muscle originated from an area close to the apex. Because of this anatomic relationship, it was impossible to obtain a clear outline of the VSD from the right ventricular aspect.

Figure 2 illustrates the heart viewed from the left ventricle. The left ventricular cavity was found to be extremely underdeveloped, and the large VSD was readily visualized. All chordae of the anterior leaflet of the mitral valve ran through the VSD and were attached to a thick anterior papillary muscle within the right ventricle. Also, there was no fibrous continuity between the anterior mitral leaflet and the pulmonary valve.

Other associated anomalies included a small patent ductus arteriosus (PDA), patent foramen ovale of about 5 mm in diameter, and mild coarctation of the aorta. There was also a right aortic arch with a right descending aorta.

Case 2

When the heart was viewed from the outside, both great vessels were parallel to a line drawn between the

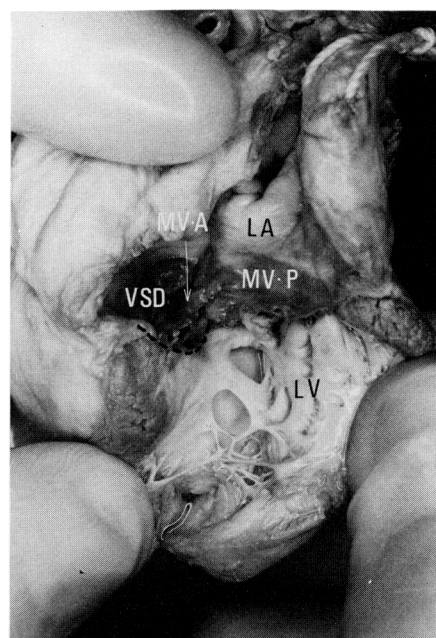


Figure 2

Left ventricular view of case 1. The left ventricle is hypoplastic. LA = left atrium; LV = left ventricle; MV-P = posterior leaflet of the mitral valve; MV-A = anterior leaflet of mitral valve straddling the ventricular septum.

atria. The aorta traversed to the right of the pulmonary trunk and had a diameter ratio of 2 to 3. The wall of the right ventricle showed marked hypertrophy, but the development of the ventricular cavity was normal. Both great vessels had subvalvular conuses and arose entirely from the top of the right ventricle. A well developed, thick parietal band was noted between the two vessels (fig. 3). The pulmonary valve was situated above the VSD and to the right of the ventricular septum without overriding observed. The VSD, which was large (30 × 20 mm), was located in front of the upper part of the crista supraventricularis. The right ventricular side of the defect was covered with three thick muscle bands which branched out from the tail of the parietal band, a formation which obscured the VSD in the right ventricular perspective. The cavity of the left ventricle was underdeveloped (fig. 4) and the mitral valve was seen to be normal. However, all chordae of its anterior leaflet and some chordae of its posterior leaflet entered the right ventricle through the VSD. These chordae were attached to a short papillary muscle extending from the right and upper part of the septum.

As noted in case 1 there was no fibrous continuity between the mitral valve and the pulmonary valve. A patent ductus arteriosus was the only associated anomaly. A defect in the atrial septum, 20 × 10 mm in size, created by the Blalock-Hanlon operation, could be seen.

Case 3

When the heart is viewed from the outside, both great vessels were parallel with a line drawn between

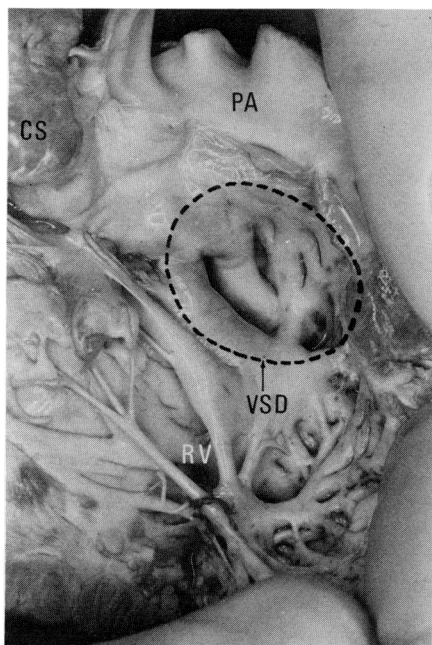


Figure 3

Right ventricular view of case 2. The ventricular septal defect (VSD) is covered with three thick muscle bands which branch out from the tail of the parietal band (CS). RV = right ventricle; PA = pulmonary artery.

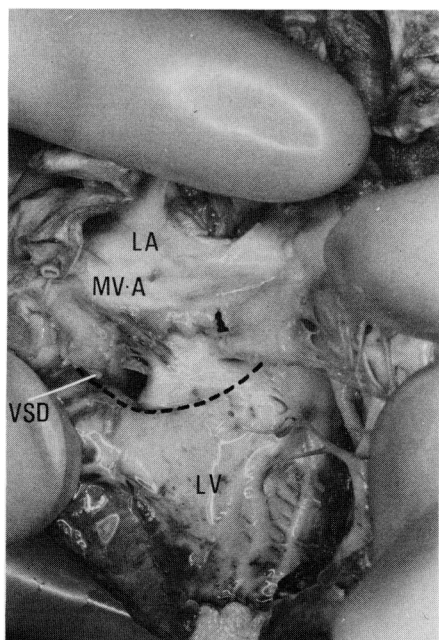


Figure 4

Left ventricular view of case 2. All chordae of MV-A and some of MV-P pass through the VSD to the right ventricle and attach to a short papillary muscle extending from the right and upper part of the septum. LA = left atrium; MV-A = anterior mitral valve leaflet; VSD = ventricular septal defect; LV = left ventricle.

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the atria. The aorta was located to the right of the pulmonary trunk. The ratio of the diameter of the aorta to the pulmonary trunk was 2 to 3. The walls of the right ventricle were hypertrophied, but the cavity was fully developed and normal. The valves of both great vessels were side by side, originating entirely from the right ventricle. Each valve had a subvalvular conus.

A well developed parietal band extended forward and backward between the two vessels (fig. 5). The pulmonary valve was immediately above the VSD and arose completely from the right ventricle without overriding the VSD. The VSD was large (40×20 mm) and was located in front of and above the crista supraventricularis. However, since the anterior leaflet of the mitral valve protruded through the VSD in a waterfall-like aspect, a complete outline of the VSD could not be seen from a right ventricular view. As compared with the right ventricle, the left ventricle was extremely underdeveloped (fig. 6). The entire anterior leaflet of the mitral valve passed through the VSD into the right ventricle, where it was attached to the divided heads of the anterior papillary muscle of the right ventricle. In this case again no continuity existed between the mitral valve and the pulmonary valve. No other anomaly was found. The atrial septum had the pericardial baffle of the Mustard procedure.

Case 4

When the heart was viewed from the outside, the aorta was located to the right of and slightly anterior to

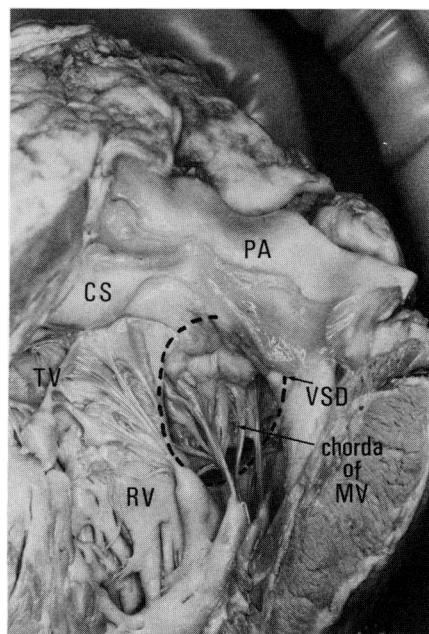


Figure 5

Right ventricular view of case 3. The entire anterior leaflet of the mitral valve passes through the VSD and falls into the right ventricle. PA = pulmonary artery; CS = crista supraventricularis; TV = tricuspid valve; RV = right ventricle; VSD = ventricular septal defect; MV = mitral valve.

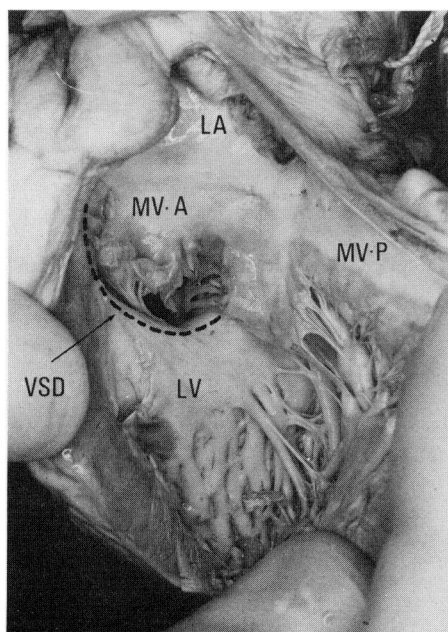


Figure 6

Left ventricular view of case 3. MV-A falls into the RV through a large VSD. LA=left atrium; MV-A=anterior mitral valve leaflet; MV-P=posterior mitral valve leaflet.

the pulmonary trunk. The ratio of the diameter of the aorta to the pulmonary trunk was 1 to 3, indicating that the latter was far more dominant.

The interior of the right ventricle revealed a fully developed cavity (fig. 7). The aorta originated entirely from the right ventricle. The aorta then passed to the right of the pulmonic trunk. Each great vessel had a conus under its semilunar valve. The two valvular rings were situated side by side, on the same level, and there was a fully developed parietal band between them. The pulmonic valve was located immediately above the VSD but originated completely from the right ventricle without overriding the VSD, which lay in front of and above the crista supraventricularis. The VSD viewed from the right ventricle was covered with two thick muscle bands which hung down from the subpulmonary conus (conal free wall), obstructing a full view of the VSD. The view from the left ventricle revealed that the VSD was large, 30×30 mm (fig. 8).

This case also had an underdeveloped left ventricle. All chordae of the anterior mitral valve and some chordae of the posterior leaflet passed through the VSD into the right ventricle and joined the two papillary muscles arising from the right side of the septum. No associated anomaly was observed.

Discussion

The complex heart malformation which is now commonly known as Taussig-Bing Heart was first described by Taussig and Bing in 1949¹ in a case report entitled "Complete transposition of the aorta and a levoposition of the pulmonary artery."

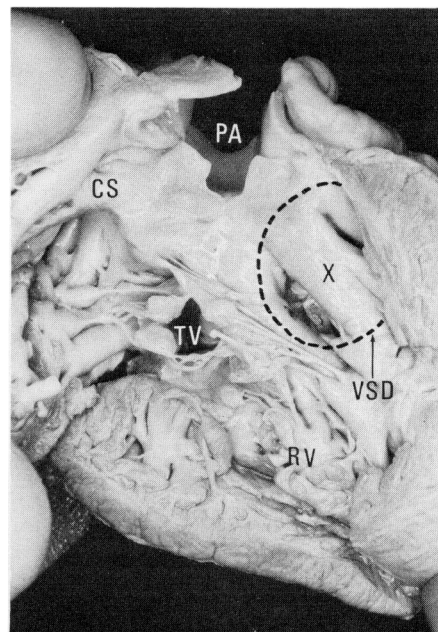


Figure 7

Right ventricular view of case 4. The pulmonary valve is located immediately above the VSD. PA=pulmonary artery; CS=crista supraventricularis (parietal band); VSD=ventricular septal defect; RV=right ventricle; TV=tricuspid valve; X=abnormal muscle ridge which covers the right ventricular side of the VSD.

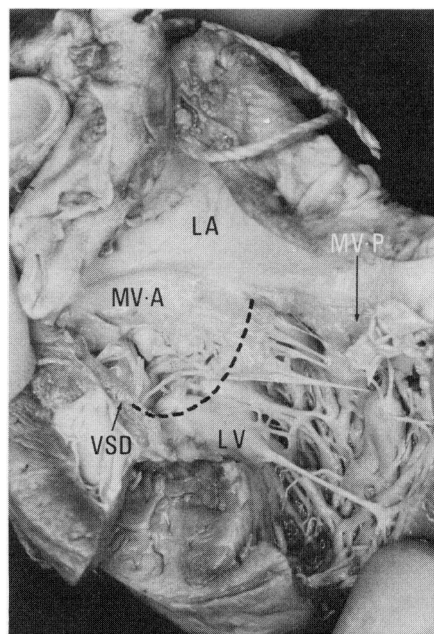


Figure 8

Left ventricular view of case 4. LA=left atrium; MV-A=anterior mitral valve leaflet; MV-P=posterior mitral valve leaflet; VSD=ventricular septal defect; LV=left ventricle.

Although the anomaly was presented schematically (fig. 9), the report gave only a generalized description of the location of the VSD, the spatial relationship of the valvular rings of both great arteries, and the extent to which the pulmonary valve overrides the VSD. Furthermore, it failed to mention the presence of a bilateral conus. Some confusion arose over the diagnosis and definition of this anomaly because of this broad description. Some groups¹⁰⁻¹³ classified "complete transposition with a posteriorly overriding pulmonary artery" as the Taussig-Bing malformation. Later this variation was referred to as false or spurious⁴ Taussig-Bing Heart, a change that only aggravated the confusion.

Keith et al.¹⁴ stressed the differences in life expectancy and prognosis between these types and favored classifying the two anomalies separately. Beuren¹⁵ published detailed descriptions of these two different anomalies. The definitions of transposition, by Van Mierop,^{3, 4} Grant,¹⁶ and Van Praagh and Van Praagh,¹⁷ and the descriptions by Neufeld et al.^{2, 18} and Lev et al.¹⁹ of "Origin of Both Great Vessels from the Right Ventricle" clarified the anatomy of the Taussig-Bing Heart. In an attempt to confirm these facts, Van Praagh²⁰ reexamined one of the original cases of the Taussig-Bing Heart. He found that only an edge of the pulmonary valve extended over the septum rather than the gross overriding of the pulmonary valve shown in figure 9.

There are a number of intermediate types between "Origin of Both Great Vessels from the Right Ventricle" and transposition of the great arteries, and these are called by some "partial transposition." While it is clear that the classification of the Taussig-Bing Heart has been given to cases of malformations which occupy a very specialized part of the spectrum of double outlet right ventricle, its definition varies slightly from one author to another. Lev et al.¹⁹ and Kawashima²¹ accept a range of pulmonary artery overriding too wide to completely coincide with the morphology of the original Taussig-Bing Heart. Lev et al. set

two extremes in this spectrum—the right-sided type in which the pulmonary trunk arises completely from the right ventricle, and the left-sided type in which the pulmonary trunk originates completely from the left ventricle.

It is extremely difficult to define the extent of pulmonic overriding. In a strict sense, the angle of inclination of the upper part of the ventricular septum should be taken into consideration but this complicates the definition without giving any information or clinical significance. Therefore, in our definition, we placed little emphasis on the extent of pulmonary valve overriding as long as the VSD is located in the pulmonic position.

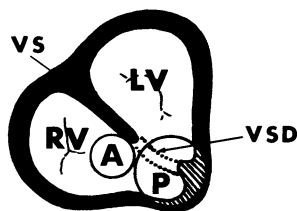
Other criteria of the Taussig-Bing Heart used at our Institute to define the malformation are as follows:

- 1) The ascending aorta originates entirely from the right ventricle, lying to the right of the pulmonary trunk
- 2) The aortic valve is higher than normal and is located at about the same cross-sectional level and on the same coronal body plane as the pulmonic valve, i.e., the aorta as well as the pulmonary trunk have a subvalvular conus
- 3) The anterior leaflet of the mitral valve lacks fibrous continuity with the aortic valve or the pulmonary valve.

The four hearts presented in this report are somewhat different from the original Taussig-Bing Heart in that the mitral valve straddles the ventricular septum. However, the other morphological features closely match the definition of the Taussig-Bing Heart. These cases were then classified as the "Taussig-Bing Heart with Mitral Valve Straddling." The mitral valve straddled the septum in all cases and the chordae of the anterior leaflet crossed over the septum through the large subpulmonary ventricular septal defect and were attached to the papillary muscles of the right ventricle. Three of the four patients had an abnormal muscle ridge, arising from the caudal part of the parietal band, which covered a large VSD as viewed from the right ventricle. The cavity of the left ventricle is seemingly underdeveloped. (This feature was especially marked in case 1.) The anterior half of the ventricular septum was abnormally shifted laterally to the left so that its upper margin was tilted over the left ventricular cavity. We postulate that this shift allowed the anterior leaflet of the mitral valve to straddle the septum (fig. 10) in these patients.

Figure 9

Modified schema from the original description of the Taussig-Bing Heart.¹ VSD = ventricular septal defect; VS = ventricular septum; RV = right ventricle; LV = left ventricle; A = aorta; P = pulmonary artery.



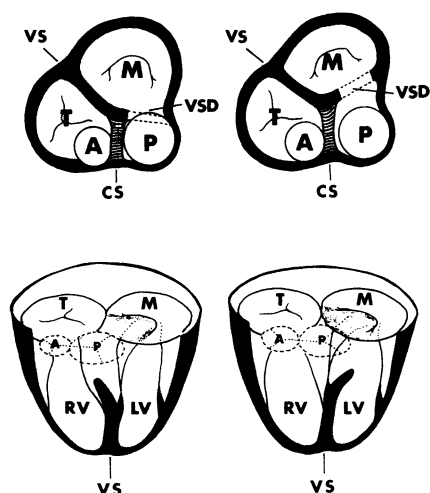


Figure 10

Original Taussig-Bing Heart (left panel) as compared with the Taussig-Bing Heart with straddling mitral valve. T = tricuspid valve; M = mitral valve; A = aorta; P = pulmonary valve; CS = crista supraventricularis; VS = ventricular septum; RV = right ventricle; LV = left ventricle.

Therefore the degree of straddling depends upon the degree of shift and the inclination of the septum. In case 2, a lesser degree of septal shift caused less straddling of the valve, and the mitral chordae were inserted on the upper margin or the right side of the septum.

A particular atrioventricular valvular malformation termed "Straddling of the mitral valve" has previously been described.^{22, 23} Characteristic features of this malformation generally include straddling of the mitral orifice over the ventricular septum and the presence of an atrioventricular communis type of VSD. In sharp contrast to these cases, the atrioventricular orifice in our patients is clearly different, with straddling of only a part of the mitral chordal structures over the anteriorly situated VSD being present. This type of straddling is associated with a unique shift of the anterior half of the ventricular septum, in which the upper margin shifts toward the left ventricular cavity as illustrated in figure 10. This shift of the septum has made it possible for the mitral chordae to be attached on the right ventricular side of the septum. Therefore, this type of straddling mitral valve differs significantly from the conventional straddling that is caused by a developmental anomaly in the sinus portion of the septum.

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