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Echocardiographic–Morphologic Correlations in Tricuspid Atresia

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Objectives. **Our aim was to clarify the anatomic substrate in hearts diagnosed as having tricuspid atresia by studying autopsy specimens and comparing the findings with those in two-dimensional echocardiograms.**

Background. **Traditionally, tricuspid atresia was thought, and is still believed by some, to be due to an imperforate valvular membrane interposed between the floor of the blind-ending right atrium and the hypoplastic right ventricle. Others argued that the dimple, when present, pointed to the outflow tract of the left ventricle rather than to the right ventricle, making the lesion more akin to double-inlet left ventricle.**

Methods. **We examined 39 autopsy specimens catalogued as having tricuspid atresia. We then studied 24 two-dimensional echocardiograms from patients with a primary diagnosis of tricuspid atresia.**

Results. **Of the 39 specimens, 37 had a completely muscular floor to the right atrium (absent right atrioventricular [AV] connection). The dimple, when identified, was (except in one case) directed to the left ventricular outflow tract. Only two hearts had an imperforate tricuspid valve. Two-dimensional echocardiograms in all cases showed an echo-dense band, produced by the fibrofatty tissue of the AV groove and representing absence of the right AV connection, between the muscular floor of the morphologically right atrium and the ventricular mass.**

Conclusions. **Tricuspid atresia is usually, but not always, due to morphologic absence of one AV connection. In most cases, the ventricular mass then comprises a dominant left ventricle together with a rudimentary and incomplete right ventricle.**

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Although the echocardiographic manifestations of tricuspid atresia are well recognized (1,2), disagreement remains as to the basic morphology of the lesion. Thus, Rao (3) perceives the lesion as an imperforate valvular membrane interposed between the floor of the blind-ending right atrium and the hypoplastic right ventricle. In contrast, most morphologic studies have demonstrated that the floor of the blind-ending atrium is usually completely muscular. A dimple, if present, has been shown to point to the outflow tract of the left ventricle or the ventricular septum, rather than to the incomplete and rudimentary right ventricle (4–6). Despite these anatomic studies, Rao (7) argues that echocardiographic studies typically show presence of an imperforate valvular membrane. In part, this disagreement is colored by differences of opinion concerning the "univentricular" nature of tricuspid atresia. We had thought that these differences had been ameliorated when it was acknowledged that only semantic sophistry had permitted the heart with tricuspid atresia to be considered a "univentricular heart" (8,9). To clarify further these controversies, therefore, and to establish the precise morphologic features that can accurately be determined with two-dimensional echocardiography, we reexamined hearts and echocardiograms catalogued as showing tricuspid atresia in the data base of the Children's Hospital of Pittsburgh.

Methods

Morphology. More than 2,500 hearts have been catalogued and preserved in the cardiopathologic museum at Children's Hospital of Pittsburgh since 1954. We extracted 39 specimens described as having tricuspid atresia. A protocol was designed and each specimen was observed in systematic fashion by at least two of three observers (R.H.A., C.A., J.D.O.). A dimple present in the floor of the atrium was identified as muscular, fibrous or imperforate and recorded as pointing toward the left ventricular outflow tract, the septal crest or the right ventricle. The right atrioventricular (AV) connection was carefully studied and, if an imperforate valve was present, its connection to the ventricular mass was detailed. The relative size and relation of the ventricles to each other were noted. The status and classification of any ventricular septal defects were studied. The interior of the right ventricle was scrutinized to assess which components were present and whether any false chords were visible. The right coronary artery delimiting the site of the ventricular septum was observed to note whether it descended

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from the AV junction at the site of the atrial septum (the crux) or at the acute margin.

Echocardiography. We reviewed the echocardiograms of all patients with the primary diagnosis of tricuspid atresia whose birth date was between January 1, 1980 and December 31, 1992. Although arbitrary, these dates were selected on the basis of the availability of two-dimensional echocardiography at our institution. A total of 33 patients were identified and suitable two-dimensional echocardiograms, obtained before a Fontan-type repair, were available for 24 of them. All studies had been performed on a Hewlett-Packard 500 or 1000 system and were recorded on VHS tape. These tapes were reviewed by two observers (J.D.O., J.A.E.). Special attention was paid to the anatomy of the AV connection, the interatrial communication and the ventricular septal defect. In all patients, multiple views had been recorded from the subcostal, parasternal and apical positions.

Results

Morphology. *Absence of one AV connection.* Thirty-seven (94.9%) of the 39 hearts examined showed complete absence of one AV connection. The floor of the blind-ending atrium was exclusively muscular in all 37. A dimple (Fig. 1) was seen in 32 (86%) of the 37 hearts and was muscular in all but 2 of these hearts, the 2 exceptional hearts showing fibrous dimples. As demonstrated by passing a pin (Fig. 1B), the dimple overlaid the outflow tract of the dominant left ventricle in 31 of the 32 hearts with a dimple; in the remaining heart, the dimple was related to the AV groove above the ventricular septum. The right-sided AV connection was absent in 35 hearts, and the left-sided connection in two; both the latter hearts had a mirror-image atrial arrangement ("situs inversus"). In all but 2 of the 35 hearts with an absent right-sided connection, and in both hearts with mirror-image atria, the morphologically left atrium was connected to a dominant left ventricle through a morphologically mitral valve. The left ventricle then fed the rudimentary and incomplete right ventricle through a ventricular septal defect in all cases except one. In this exceptional case, the ventricular septum was intact, the rudimentary right ventricle could not be identified by gross inspection and the pulmonary trunk was completely atretic. When viewed from the right ventricle, the defect in all those hearts with a dominant left ventricle had exclusively muscular borders (Fig. 2) except for two hearts in which it was doubly committed and subarterial because of the absence of the outlet septum.

In all cases, the AV component of the membranous septum was seen just to the left of the posterior aspect of the ventricular septum as an integral part of the central fibrous body (Fig. 3). This was the area corresponding to the dimple in the blind-ending right atrium in all but one heart with juxtaposed atrial appendages (the heart in which a fibrous dimple overrode the posterior extent of the ventricular septum). On the ventricular side, when viewed from the right ventricle, the membranous septum was separated by a discrete muscular border from the rim of the septal defect. The right ventricle always lacked its inlet component, and the delimiting branch of the right coronary artery marking the site of the septum between dominant and rudimentary ventricles always descended from the AV junction at the acute margin of the ventricular mass (Fig. 4). It never descended at the point of insertion of the atrial septum (the crux). In the overall group of 37 hearts with an absent AV connection, the ventriculoarterial connections were concordant in 25, discordant in 9 and showed double-outlet right ventricle in 2. The other heart, with absence of the right ventricle, had pulmonary atresia with the aorta arising from the left ventricle.

In two hearts with absence of the right-sided AV connection, the left-sided AV valve was of tricuspid morphology. However, the atrial anatomy was indistinguishable from that in the other hearts. In one of these two hearts, the valve straddled (Fig. 5) between ventricles of similar size showing left-handed topology ("*l*-loop"). The ventriculoarterial connections in this heart were discordant. In the other heart, the morphologically tricuspid valve was exclusively connected to a dominant right ventricle that supported both arterial trunks, each arterial valve having a completely muscular infundibulum. The left ventricle was a slitlike cavity found posteriorly and to the right of the dominant right ventricle. In both of these hearts, the ventricular septal defect was perimembranous.

Imperforate AV valve. Imperforate valves were encountered in only two hearts, both with concordant AV connections. In one, the hypoplastic right ventricle was divided, with the distal part reminiscent morphologically of the rudimentary and incomplete right ventricle found in the most common variant of tricuspid atresia. However, the inlet part of the right ventricle was well formed, albeit small, and was connected to a hypoplastic annulus in the floor of the right atrium (Fig. 6A). It was difficult to be sure that the valve itself was atretic rather than severely stenotic. The ventricular septal defect in this heart was perimembranous (Fig. 6B). In the other heart (Fig. 7), the imperforate right valve overrode the ventricular septum, but its tension apparatus was located exclusively in the complete but hypoplastic right ventricle. The ventricular septal defect was again perimembranous. The ventriculoarterial connections were concordant, but the aorta was left-sided and anterior ("anatomically corrected malposition").

Echocardiography. Two-dimensional echocardiograms were reviewed in 24 patients. There was the usual atrial arrangement (situs solitus) in 23 patients and a mirror-image arrangement (situs inversus) in 1. All patients had an interatrial communication within the oval fossa, and two also had a communication through the coronary sinus. The floor of the right atrium in all patients was separated from the ventricular mass by an echo-dense band, typical of the fibrofatty tissue of the AV groove (Fig. 8, top). A dimple could not specifically be identified echocardiographically in any patient with absence of the AV connection but, in some, the AV membranous septum was seen between the front of the right atrium and the left ventricular outflow tract (Fig. 8, bottom). Ventriculoarterial connections were concordant in 20 patients and discordant in 4. The ventricular septal defect had complete muscular borders

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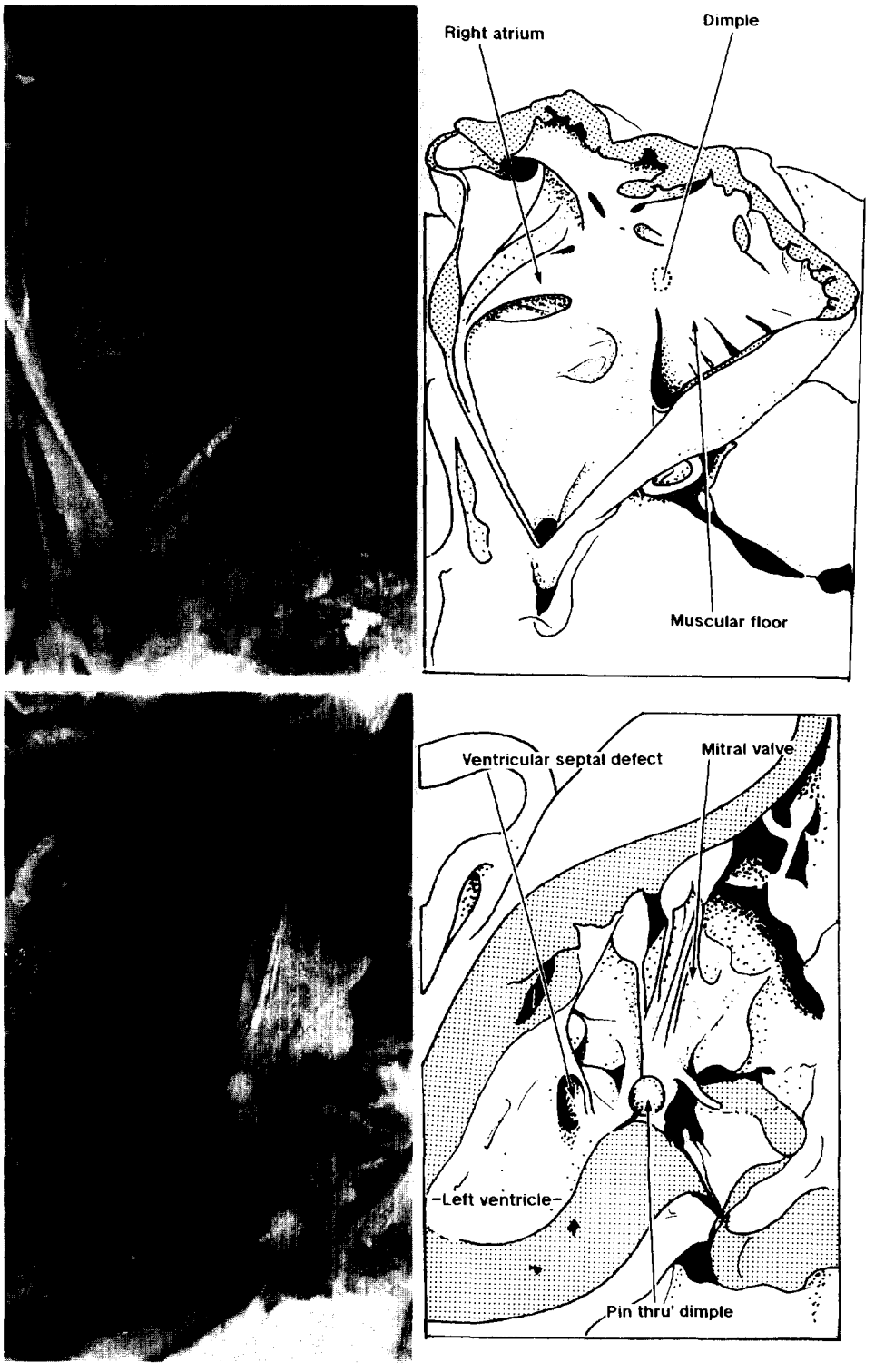

Figure 1. Heart with typical tricuspid atresia and complete absence of the right atrioventricular connection. The dimple in the muscular floor of the blind-ending right atrium **(A)** overlies the outflow tract of the dominant left ventricle, demonstrated from the left ventricular aspect in **B,** where a pin has been placed across the dimple.

in all patients. The origin and course of the coronary artery branches could not be discerned in any of the studies.

Discussion

This series, albeit relatively small, encapsulates the problems and realities of tricuspid atresia. It shows that, although in almost all cases the morphologically right AV connection is absent and the left atrium is connected to a dominant left ventricle in the presence of an incomplete and rudimentary right ventricle, cases can be found with an imperforate tricuspid valve. Why, then, has there been (10), and does there continue to be (3,7), so much controversy about a relatively straightforward lesion? In part, the controversy surrounds the

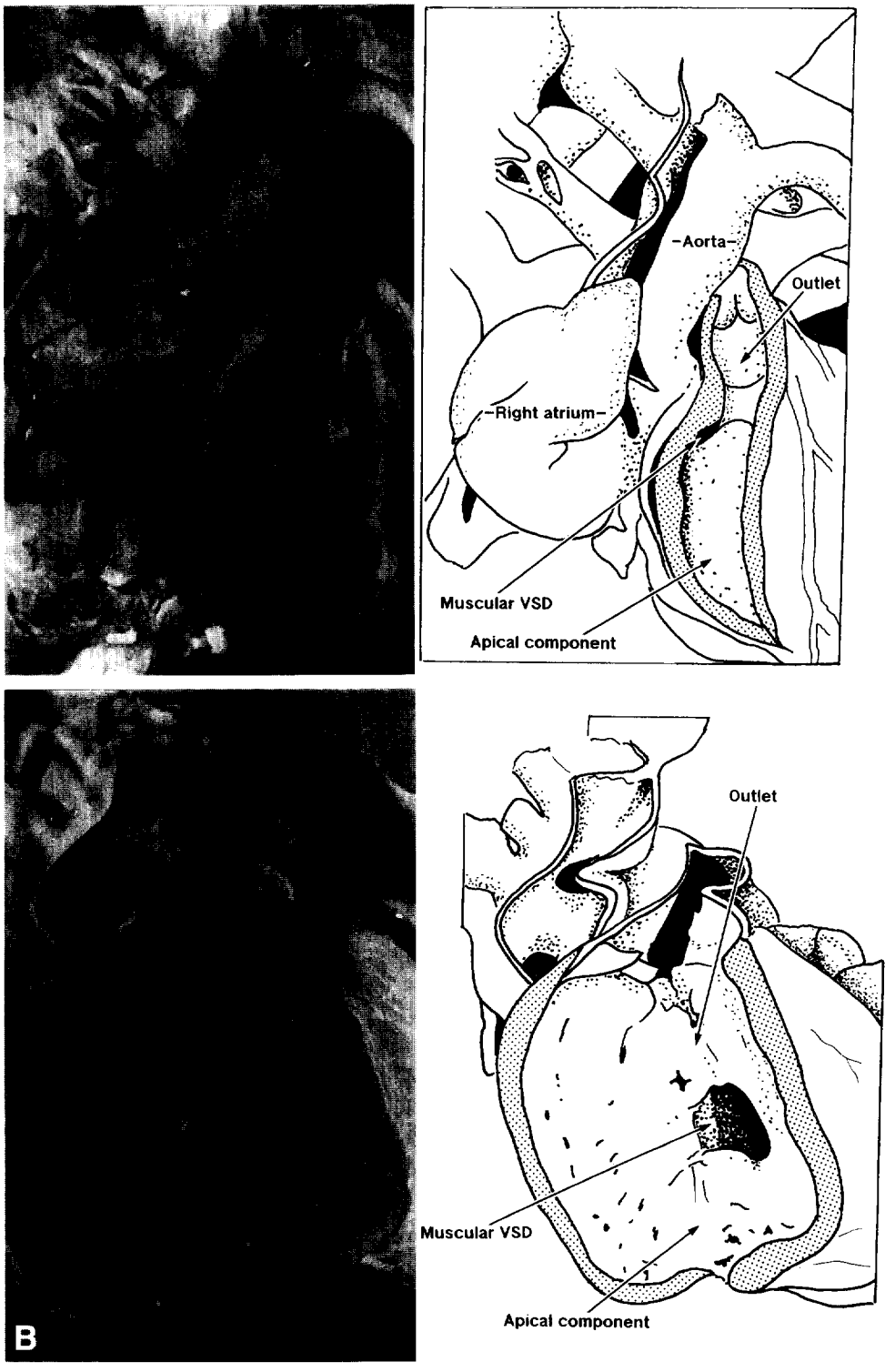

Figure 2. Hearts with typical tricuspid atresia. The rudimentary right ventricles have been opened to display muscular ventricular septal defects (VSD). The defect is tightly restrictive in **A,** with an additional constriction between the outlet and apical trabecular components, but it is larger and more centrally placed within the septum in **B.**

relation between tricuspid atresia and double-inlet left ventricle, and the propriety of describing either of these lesions as a form of "univentricular heart." But some of the disagreements remain related to the basic anatomy underlying the prototype of tricuspid atresia. We will address both these issues briefly before turning attention to the clinical significance of our morphologic and echocardiographic findings.

Tricuspid atresia versus double-inlet left ventricle. Although the ventricular mass in tricuspid atresia usually shows marked similarities with double-inlet left ventricle, the lesion itself is fundamentally different. In double-inlet left ventricle, both atrial chambers are connected to the dominant left ventricle, either through two AV valves or through a common valve. In the usual variant of tricuspid atresia, only the

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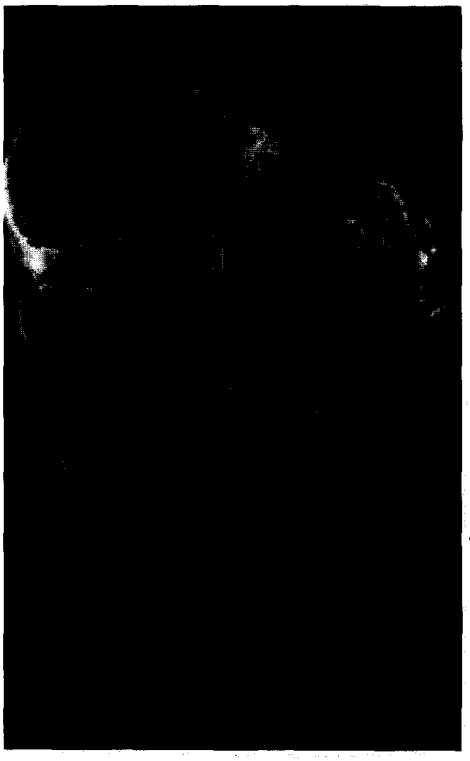


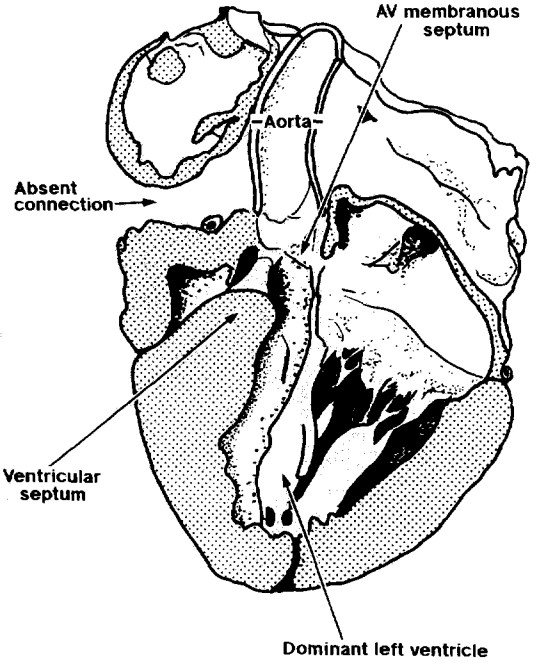
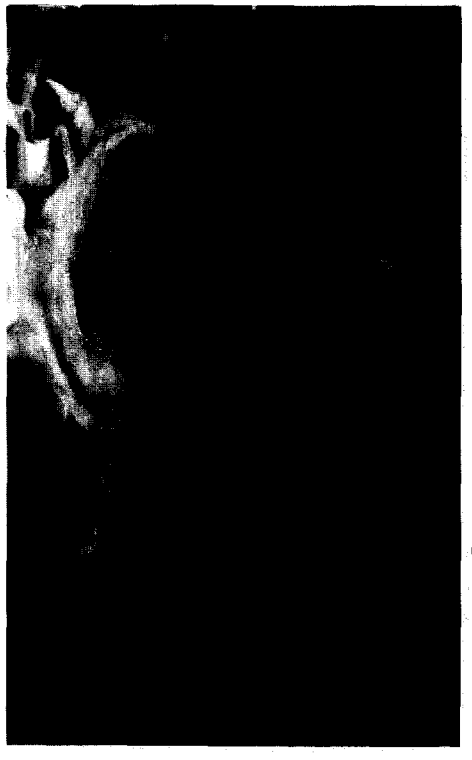


Figure 3. Heart sectioned in the long-axis plane, showing the position of the atrioventricular (AV) membranous septum just to the left of the posterior part of the ventricular septum. The membranous septum is an integral part of the central fibrous body and corresponds to the dimple in the floor of the right atrium (see Fig. 1).

morphologically left atrium is connected to the ventricular mass. Furthermore, tricuspid atresia is found most frequently with concordant, and double-inlet left ventricle with discordant, ventriculoarterial connections. It is the ventricular mass that shows the similarities, which are best appreciated when examples of each lesion are compared in the setting of the same ventriculoarterial connections (either concordant or discordant). Measurements of the ventricular masses from autopsy hearts (11) then confirm the similarities (12). Equally obvious differences, however, are found between the prototype



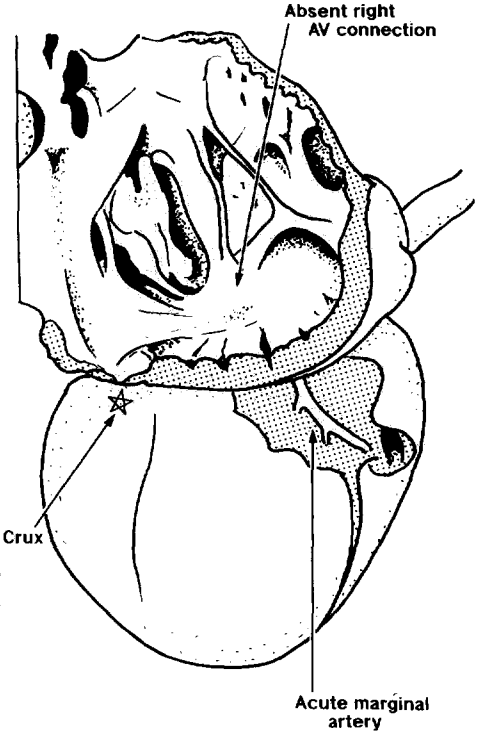


Figure 4. Heart with typical tricuspid atresia, dissected to show the delimiting branch of the right coronary artery descending from the atrioventricular (AV) junction at the acute margin of the ventricular mass rather than at the crux.

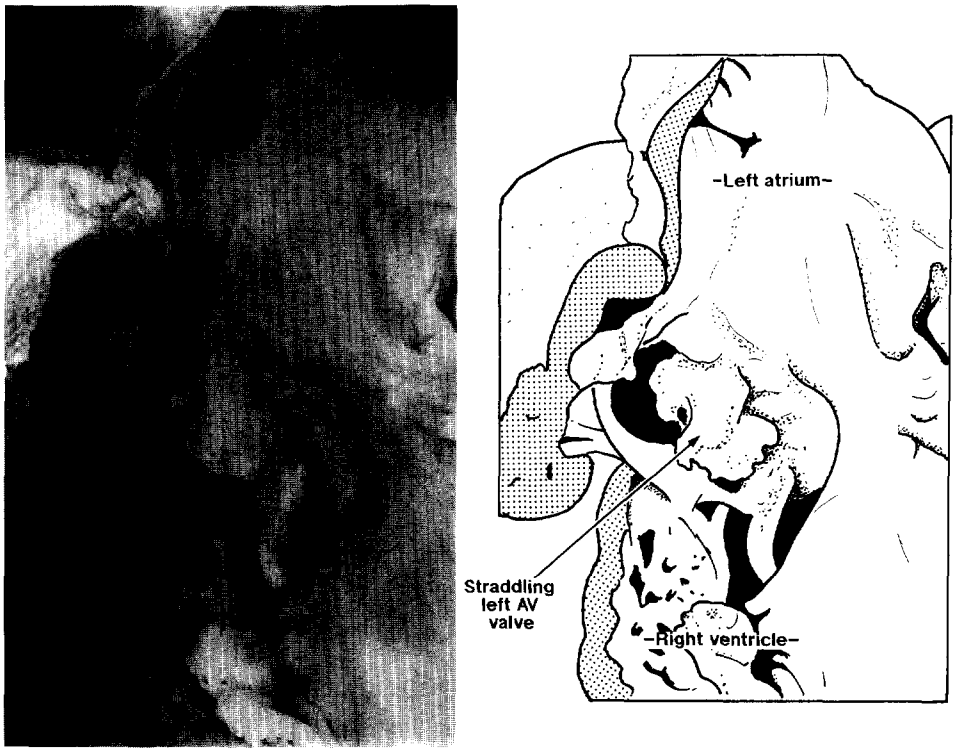


Figure 5. Heart with absent right atrioventricular (AV) connection. The left-sided AV valve is of tricuspid morphology and straddles and overrides between a dominant left-sided morphologically right ventricle and a right-sided morphologically left ventricle.

of tricuspid atresia with univentricular AV connection (only the left ventricular myocardium having a junctional connection with atrial musculature) and those much rarer examples with concordant AV connections and an imperforate AV valve. In these rarer lesions, the right ventricle is complete, albeit hypoplastic. In contrast, in the more common variant of tricuspid atresia, the right ventricle is incomplete and lacks its inlet component.

The concept of incompleteness of the ventricle was well described by Ando et al. (6). Although we agree with their interpretations on this point, our results do not support their contention that, in the usual form of tricuspid atresia, the posterior delimiting coronary artery descends at the crux of the heart. But this probably reflects the definitions used for the cardiac crux. If the crux is taken as the site of the posterior descending artery, as proposed by Bharati and Lev (10), then the septum is indeed at the crux. Nonetheless, this definition is self-fulfilling, because the septum, as long as it is marked by the artery, will be at the crux irrespective of its relation to the AV junction. When a separate criterion is used for definition of the crux, namely the location of the atrial septum as currently proposed, then there is an obvious difference in location of the ventricular septum between tricuspid atresia due to absence of one AV connection and the form characterized by an imperforate membrane blocking otherwise concordant AV connections.

Tricuspid atresia and the absent connection. Our echocardiographic and morphologic observations, in keeping with other more recent two-dimensional echocardiographic studies (1,2), show that the dense area of echoes beneath the muscular floor of the right atrium found in the most common variant is not produced by an imperforate tricuspid valve. These echoes, even when overlying the roof of the right ventricle, are generated by the fibrofatty tissue of the right AV groove. For those who doubt these findings, it is an easy matter to take any typical specimen of tricuspid atresia and completely strip away the muscular floor of the right atrium from the ventricular mass. Furthermore, recent embryologic studies (13) have shown how, during early development, the heart resembles closely the arrangement seen in the variant of tricuspid atresia with absent AV connection. Tricuspid atresia, therefore, can just as readily be explained on the basis of failure of formation of the right AV connection as by the concept of fusion of previously formed leaflets of the developing tricuspid valve. It is easy to envisage this latter mechanism producing tricuspid atresia, but as far as we know it has yet to be documented by serial fetal echocardiography.

Is tricuspid atresia a useful term? No one has doubted the clinical value of the term tricuspid atresia, least of all ourselves. Potential confusion can occur, however, if hearts with a blind-ending left atrium are described as having tricuspid atresia. Such hearts do exist, and were found in our autopsy collection. Had the left valve developed properly in these hearts, it would almost certainly have been of tricuspid morphology. Embryologically, therefore, these hearts show tricuspid atresia. But, hemodynamically, they present with total obstruction of pulmonary, rather than systemic, venous return. For this reason we excluded these hearts from this presenta-

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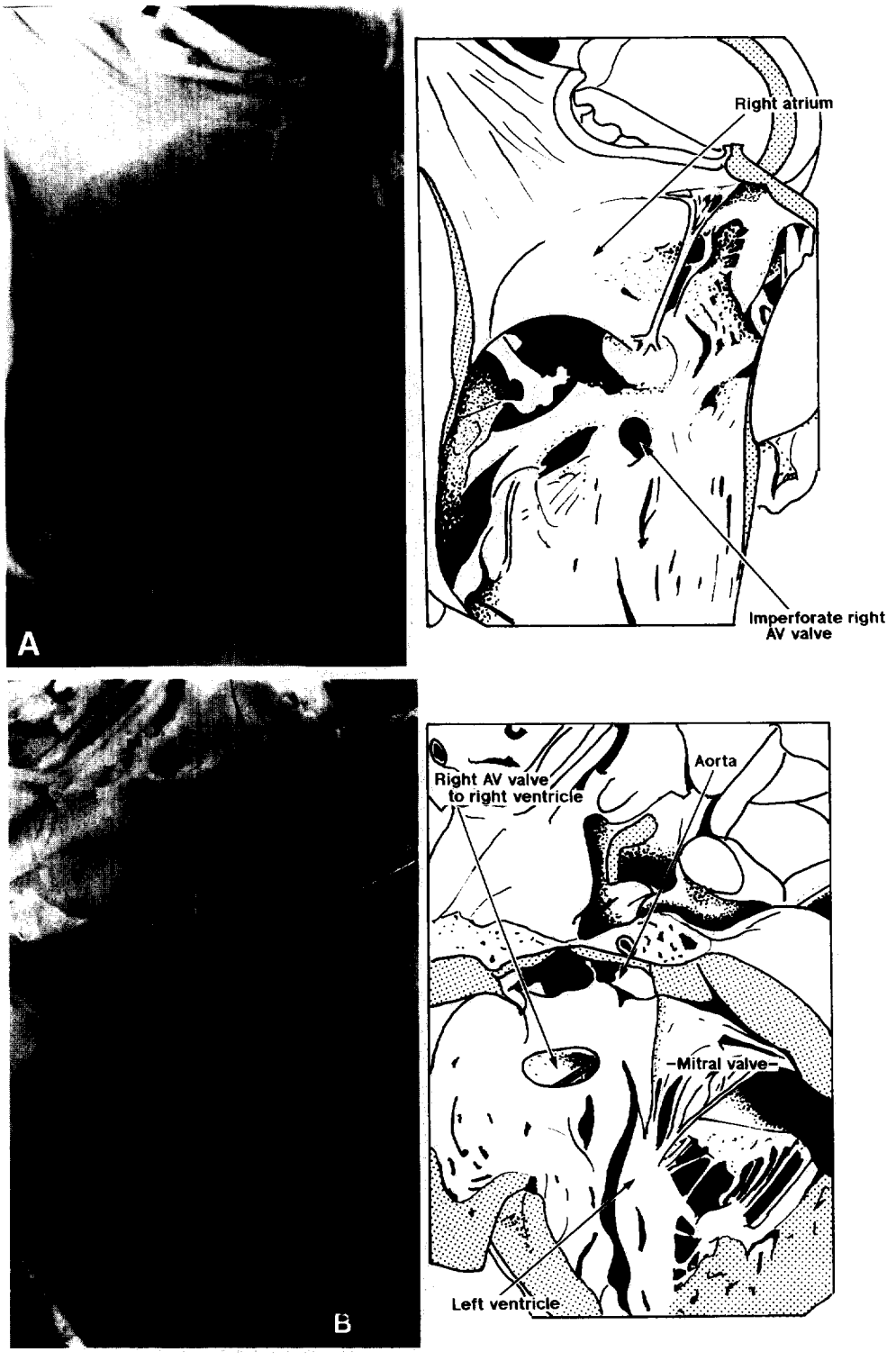

Figure 6. Heart with an imperforate tricuspid valve interposed between the right atrium and a hypoplastic right ventricle (**A**). The tension apparatus supporting the imperforate valve is seen through a perimembranous ventricular septal defect (**B**). AV = atrioventricular.

tion. In contrast, we included in our series two hearts with a blind-ending morphologically right atrium with absent right AV connection, an atrial morphology that is indistinguishable from the usual variant of tricuspid atresia. In these hearts, the left atrium was connected predominantly to a dominant right ventricle in a ventricular mass showing left-handed topology ("*l*-loop"). Morphologically and developmentally, therefore, these hearts would properly be described as having mitral atresia. To us, this term is again potentially confusing because, in such hearts, it is the morphologically right atrium that is blind-ending, and this is the clinical criterion for tricuspid atresia. Such variants are sufficiently rare to warrant a full

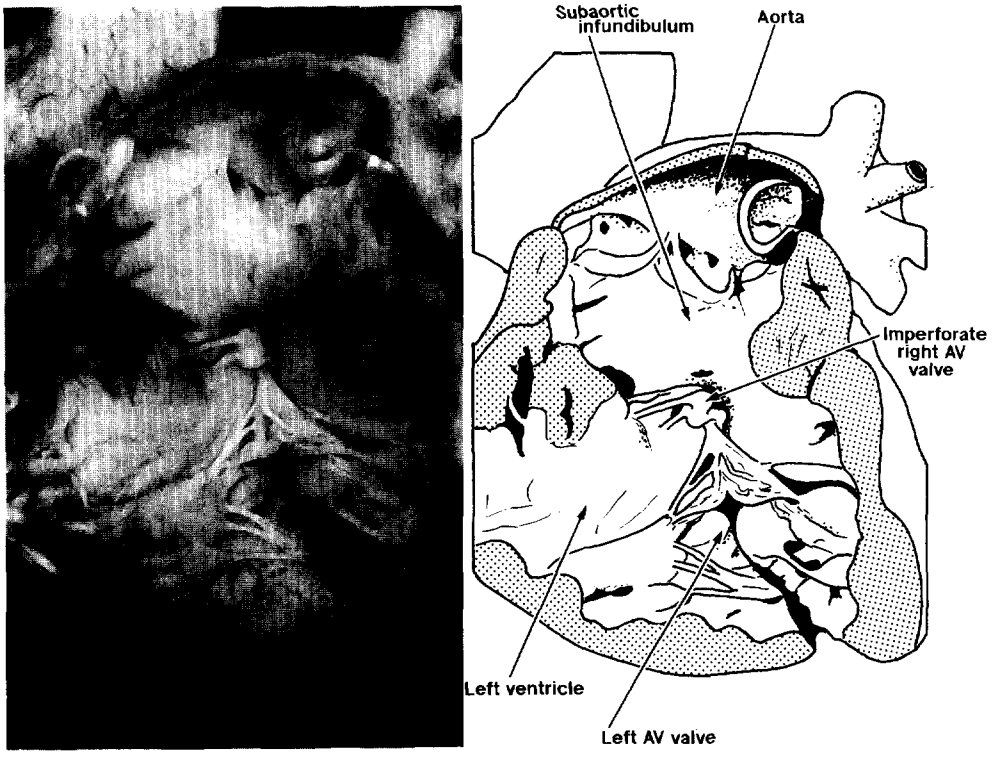

Figure 7. Heart with an imperforate right atrioventricular (AV) valve. The valve is connected to the dominant left ventricle along with the patent left AV valve–double-inlet AV connection. Note the origin of the aortic valve from a complete muscular infundibulum ("anatomically corrected malposition").

segmental description, as proposed by Ando et al. (6), but will only be recognized when searched for specifically. Whether such cases should be included with the commoner variant of tricuspid atresia depends on whether developmental or clinical criteria are used to make the groupings.

Is the heart with tricuspid atresia a univentricular heart? Our echocardiographic and morphologic studies show that, apart from the exceptional heart with total absence of the right ventricle and pulmonary atresia, none of the hearts described herein had a solitary ventricle. But, then, neither do the characteristic hearts with double-inlet left ventricle. Our original assertion concerning the potential univentricular nature of typical tricuspid atresia (11) was made in an attempt to show the similarity of the ventricular mass in hearts with tricuspid atresia and double-inlet left ventricle. The similarity unequivocally exists. This does not negate the fact that any argumentation specifying a univentricular arrangement for the ventricular mass in either of these entities has to be spurious. It is our belief that the controversy generated by such argumentation has confused the issue. This is clarified when hearts with tricuspid atresia in its typical form and hearts with double-inlet left ventricle are both described as having big left ventricles and small and incomplete right ventricles. Description of the ventricular mass in terms of big and small ventricles then permits the hole between them to be described simply and accurately as a ventricular septal defect.

Clinical implications. The morphologic differences among the various forms of tricuspid atresia are rarely sufficient to change the options for surgical treatment. Thus, those rare cases with imperforate valvular membranes are hardly ever found when the valve is of sufficient dimensions to be excised and replaced by a prosthesis, thus restoring biventricular AV connections. Only those variants with an atretic Ebstein malformation (14), not encountered in our series, are likely to permit this option. For the remainder, the surgical option will be one of the modifications of the Fontan procedure. It will then be advantageous to distinguish those rare hearts with left-handed ventricular topology in which the left atrium is connected to a dominant right ventricle. As discussed, although developmentally and morphologically these lesions are justifiably described as mitral atresia, clinically they are directly comparable to the usual variant of tricuspid atresia. The presence of the morphologically right ventricle as the dominant ventricle in these patients may have significance for long-term follow-up.

In the remaining greater majority of hearts encountered in our study, the ventricular septal defect was either muscular or, very rarely, doubly committed and juxta-arterial. Perimembranous defects were found only in the hearts with concordant AV connections and an imperforate tricuspid valve. We never encountered defects that included the AV component of the membranous septum as part of the rim when the AV connection was absent. Neither, apart from those hearts with an imperforate valve, did we find examples with remnants of the tricuspid valve within the right ventricle. We did occasionally find examples with false chords that could have appeared as

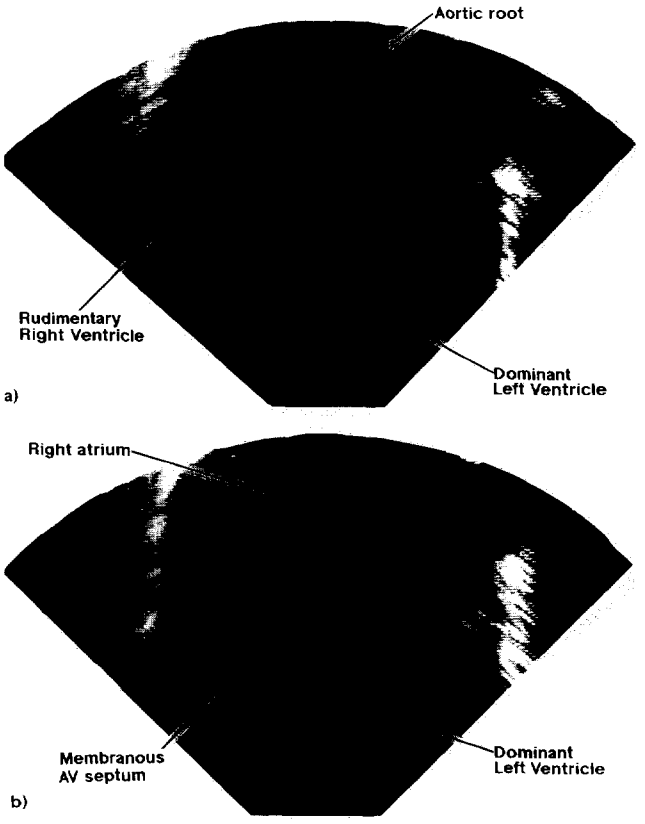

Figure 8. Two-dimensional echocardiograms showing the typical features of classic tricuspid atresia. The **upper panel** shows the aortic root interposed between the muscular floor of the right atrium and the rudimentary right ventricle. A more posterior cut, in the **lower panel**, shows the echo-dense wedge representing the fibrofatty tissue of the atrioventricular (AV) groove at the anticipated site of the right AV connection, which is absent. Note the location of the membranous AV septum.

tension apparatus, or examples with folds of tissue beneath the aortic valve. These remnants should not be interpreted as representing the tricuspid valve when the right AV connection is absent.

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