Morphological Studies in Tricuspid Atresia

By Barbara Guller, M.D., and Jack L. Titus, M.D., Ph.D.

SUMMARY

Fourteen hearts with tricuspid atresia were reviewed at autopsy. The study included examples of normally related great arteries and of transposition of the great arteries; instances of pulmonary stenosis were present in both groups. In these examples of tricuspid atresia the size of the right ventricle depended on the size of the ventricular septal defect. The ventricular septal defect and the right ventricle usually were smaller in proportion to the size of the heart in specimens with normally related great arteries. In the absence of transposition of the great arteries, the ventricular septal defect was found to be the site of obstruction to pulmonary flow, and its size in relation to the size of the aorta determined the size of the pulmonary artery. Morphologic abnormalities exist in the region in which the proximal portions of the major atrioventricular conduction system (A-V node, bundle of His, origin of bundle branches) normally are located; thus, anatomic abnormalities in the course of the atrioventricular conduction tissue could be expected in tricuspid atresia. Based on histologic evidence, severe elevation of pulmonary vascular resistance, in general, does not occur in infancy in tricuspid atresia.

Additional Indexing Words:

Pulmonary vasculature
Ventricular septal defect in tricuspid atresia

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LTHOUGH the general morphologic features of tricuspid atresia are known, exact knowledge of the anatomy of associated conditions, such as pulmonary stenosis and transposition of the great vessels, often presents problems. Current clinical and hemodynamic studies of tricuspid atresia necessitate a semiquantitative morphologic knowledge of the varieties of tricuspid atresia. Therefore, we reexamined at autopsy the hearts and lungs from 14 patients who had had tricuspid atresia.

Results

The major elements for the diagnosis of tricuspid atresia at autopsy were (1) four recognizably distinct cardiac chambers, and (2) one atrioventricular valve which guarded the inflow into the morphologically distinct left ventricle and was of the general configuration of the normal mitral valve, whether in mirror image or in normal orientation.

The ventricles were identified as right and left on the basis of their internal topographic morphology1 and not according to their anatomic position in the body.

The classification used in the 14 cases was that proposed by Edwards and Burchell,2 in which two major types of tricuspid atresia are recognized according to the interrelationship of the great vessels at the base of the heart and in which each type is subdivided according to the state of the pulmonary artery. A further subdivision of type I (type Ic), proposed by Keith and associates,3 also was used. Thus, type I includes instances in which the great arteries are normally interrelated (no transposition), and type II includes those instances with transposition of the great arteries. Type I is subdivided into cases of pulmonary atresia (type Ia), pulmonary stenosis of some form (type Ib), and no pulmonary stenosis (type Ic). Two subgroups comprise type II: type IIa, pulmonary or subpulmonary stenosis, and type IIb, no pulmonary or subpulmonary stenosis. In all 14

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cases the heart was in the position of levocardia, that is with its major base-to-apex axis directed to the left, and the thoracic and abdominal viscera were in the position of situs solitus.

The morphological appearances, but not sizes, of the right atrium, the left atrium, and the left ventricle were similar in all 14 specimens. In all instances, the right atrium was moderately to greatly enlarged with prominent trabeculations and pectinate muscles and a thickened muscular wall. The floor of the right atrium was smooth without evidence of a right atrioventricular orifice or a remnant of valvular tissue. In nine of the 14 specimens, a tiny, endocardial-lined blind pocket was present in the floor of the right atrium in the region of the expected tricuspid orifice (fig. 1A). In two hearts with transposition of the great arteries (one of type IIa and one of type IIb), the right atrial appendage was situated on the left side of the great arteries, creating juxtaposition of the atrial appendages.

In 13 of the 14 hearts, interatrial communication occurred through a patent, valvular-competent foramen ovale. The size of the patency and the size of the valve of the foramen ovale in the left atrium varied from case to case. In only one of the 14 was there true deficiency of atrial septal tissue, which was a defect in the region of the fossa ovalis ("secundum type atrial septal defect"). The left atrium was enlarged in all cases, but the size of the left atrium varied greatly according to the size of the interatrial communication. In all instances, the pulmonary veins were normally connected to the left atrium. A right superior vena cava was present in all specimens; a persistent left superior vena cava was not found in any of the specimens. Enlargement and hypertrophy of the left ventricle existed in all 14 cases. The left atrioventricular valve (mitral valve) was large in all instances and appeared competent in every case.

**Type I: Tricuspid Atresia Without Transposition of Great Arteries (10 Cases)**

In all 10 examples of tricuspid atresia without transposition of the great arteries, the right ventricle was composed of a small chamber that represented mainly the infundibulum of the normal right ventricle. The size of the right ventricle was related to the size of the ventricular septal defect. This relationship, however, was less characteristic than the relationship of the size of the pulmonary

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*P = pulmonary; VSD = ventricular septal defect.
†Persistence of fetal state.

**Circulation, Volume XXXVIII, November 1968**
artery to the size of the ventricular septal defect.

The position of the ventricular septal defect, when viewed from the left ventricle, was similar in all 10 cases. In each instance it lay from 1 to 3 mm below the posterior and right aortic cusps and was entirely surrounded by muscle. In 12 cases the membranous septum was absent; in two cases it was represented by a remnant of fibrous tissue. In order to estimate the size of the septal defect it was compared to the size of the aorta. In all cases classified as type Ib, the ratio of the diameter of the ventricular septal defect to that of the aorta was less than 0.5; that is, the diameter of the ventricular septal defect was less than 50% of the diameter of the aorta (table 1, cases 2 through 9). In the single example of type Ic this ratio was greater than 0.5 (table 1, case 10). In type Ib, the pulmonary artery usually was smaller than the aorta so that the size of the pulmonary artery appeared to be related to the size of the ventricular septal defect. A similar relationship between the size of the ventricular septal defect and the pulmonary artery pertained for the specimen of type Ic. The major site of obstruction to pulmonary blood flow in type Ib was at the level of the ventricular septal defect and not at the level of the pulmonary valve or the infundibulum of the right ventricular outflow tract.

Type Ia (Case 1; Fig. 1)

The pulmonary atresia resulted from atresia of the pulmonary valve but a tiny pulmonary trunk was present. The right ventricle was represented by a tiny, endocardial-lined muscular chamber (fig. 1B), which was the smallest right ventricular chamber present in any of the 14 cases. The right ventricular chamber was isolated completely from the other cardiac chambers since no ventricular septal defect was present. Pulmonary blood supply occurred through a patent ductus arteriosus and prominent bronchial arteries originating from the aorta.

Type Ib (Cases 2 through 9; Fig. 2)

In each instance the site of obstruction was the ventricular septal defect. Thus, there was no pulmonary valvar or subvalvar stenosis in the usual sense. In all eight cases the

Figure 1

Case 1. Tricuspid atresia type Ia (no transposition of great arteries, pulmonary atresia). (A) Opened right atrium. "Dimple" (point of arrow) is anterior and to left of coronary sinus (CS). The dimple represents a tiny, endocardial-lined pocket in region of expected tricuspid orifice. Large patent foramen ovale (FO). SVC = superior vena cava; A = aorta. (B) Anterior view of base of heart with opened minute right ventricle (RV) at upper right cardiac border. Atresia of pulmonary valve (point of arrow). Tiny pulmonary trunk (P) opened. Large ascending aorta (A) behind and to right of pulmonary artery. LA = left atrial appendage; LV = left ventricle.
Figure 2

Tricuspid atresia type Ib (no transposition, subpulmonary stenosis). (A) Case 4. Anterior view of heart and great vessels. A small right ventricle (RV) leads to a small pulmonary artery (P). The right atrium (RA) is large. LV = left ventricle; A = aorta. (B) Case 4. Opened right ventricle of specimen shown in A. Small ventricular septal defect (point of arrow). (C) Case 3. Opened right ventricle. The small elongated right ventricle communicated with the pulmonary artery, which was of the same diameter. No infundibular or valvular obstruction. Small ventricular septal defect at apex of right ventricle (arrow). (D) Case 3. Opened left ventricle of specimen C. The small ventricular septal defect (white probe) is below the aortic valve and is surrounded by muscle.

Ventricular septal defect was a narrow muscular channel whose right ventricular opening was situated near the apex of the right ventricle. The right ventricle was an elongated chamber which lay obliquely along the base of the heart with the same diameter as that of the pulmonary artery with which it communicated (fig. 2A-C). Although the pulmonary valve was bicuspid in four cases, it was not stenotic in any case. The main pulmonary artery and its primary branches were of small diameter. In two hearts, prominent bronchial arteries arose from the aorta as collateral sources of pulmonary blood flow. The only examples of anomalies of the aortic arch encountered in any of the 14 cases of
tricuspid atresia were associated with this type. These were two examples of right aortic arch with mirror-image branching.4

Type Ic (Case 10; Fig. 3)

This specimen had the largest right ventricle of any in the type I category. A crista supraventricularis was present above the ventricular septal defect. The pulmonary artery was of normal size and the pulmonary valve was a normally formed, tricuspid, semilunar valve.

Type II: Tricuspid Atresia With Transposition of Great Arteries (Four Cases)

In all four cases, the aorta lay anterior to and to the right of the pulmonary artery at the base of the heart. This variety of transposition of the great arteries has been termed "dextrotransposition" to distinguish it from transposition of the great arteries in which the aorta lies anterior to and to the left of the pulmonary artery at the base of the heart, which has been termed "levotransposition."5

In order to compare the potential blood flow to the systemic circulation with that to the pulmonary artery, the sizes of the aortic valve, the pulmonary valve, and the ventricular septal defect were measured. The diameter of the ventricular septal defect was as large as or slightly larger than the diameter of the aorta in all four hearts (table 1, cases 11 through 14). In type IIa (transposition with pulmonary obstruction), the diameter of the ventricular septal defect was larger than that of the pulmonary valve. In type IIb (transposition without pulmonary obstruction), the area of the pulmonary valve was slightly larger than that of the ventricular septal defect.

The size of the right ventricle was related to the size of the ventricular septal defect in each instance and was larger in all cases in which the arteries were transposed than in those without transposition. In hearts in which the great arteries were transposed, the right ventricle had an infundibular chamber with a well-defined crista supraventricularis, and the muscular trabeculae were more

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**Figure 3**

Case 10. Tricuspid atresia type Ic (no transposition, no pulmonary stenosis). Anterior view of opened right ventricle and large pulmonary artery (P). The right ventricle was larger than in type Ia (fig. 1) or in type Ib (fig. 2). A large ventricular septal defect (arrow) was present.

**Figure 4**

Case 11. Tricuspid atresia type IIa (transposition of great arteries, subpulmonary stenosis). Opened left ventricle has narrow subpulmonary muscular outflow tract (opened) leading to a bicuspid pulmonary valve (PV). Large ventricular septal defect (VSD). AM = anterior mitral leaflet.
prominent than in any case without transposition of the great vessels.

Type IIa (Case 11; Fig. 4)

This type of tricuspid atresia is characterized by transposition of the great arteries and obstruction to pulmonary flow at or below the level of the pulmonary valve. The specimen showed obstruction below the pulmonary valve because of a narrow muscular subpulmonary tract which represented the left ventricular outflow tract. The bicuspid pulmonary valve was not stenosed. The pulmonary artery was smaller than the aorta.

Tricuspid atresia type IIb (transposition of great arteries, no pulmonary stenosis). (A) Case 14. Heart viewed in left anterior oblique position. Juxtaposition of atrial appendages and transposition of great arteries. LA = left atrial appendage; RA = right atrial appendage; LV = left ventricle; P = pulmonary artery; A = aorta. (B) Case 14. Opened right ventricle and aorta of A. The right ventricle is larger than in type I (no transposition of great vessels, figs. 1, 2, and 3). Arrow points to large ventricular septal defect. (C) Case 14. Opened left ventricle (LV) and pulmonary artery (P) of specimen shown in A and B. Arrow points to large ventricular septal defect. (D) Case 13. Opened right ventricle and aorta. The right ventricle and ventricular septal defect (probe) are large. (E) Case 13. Opened left ventricle and pulmonary artery of specimen shown in D. Probe in large ventricular septal defect.
Figure 6

(A) Case 5. Age 7 weeks. Tricuspid atresia type Ib (no transposition, subpulmonary stenosis). Small thin-walled pulmonary artery with recent thrombus; elastic van Gieson; × 45. (B) Case 9, Age 26 months. Tricuspid atresia type Ib (no transposition of great arteries, subpulmonary stenosis). Organizing thrombus in small, thin-walled pulmonary artery; elastic van Gieson, × 100. (C) Case 11. Age 12 years. Tricuspid atresia type IIa (transposition of great arteries, subpulmonary stenosis). Recanalized thrombus in small muscular pulmonary artery; elastic van Gieson, × 225.
The ventricular septal defect lay below the subpulmonary tract and was not directly related to the pulmonary valve. No membranous septum was identified. The right ventricular chamber was larger than the right ventricle in all cases without transposition of the great vessels.

Type IIb (Cases 12 through 14; Fig. 5)

In all three cases the pulmonary artery was as large as or larger than the aorta. The ventricular septal defect was entirely surrounded by muscle in two cases in which the membranous septum was absent. In the third specimen, a remnant of membranous septum was present at the posterior-superior aspect of the ventricular septal defect. In all three cases, the right ventricle was larger than in specimens without transposition of the great arteries. In one specimen juxtaposition of the atrial appendages to the left of the great arteries was present.

Histologic Pulmonary Vascular Findings

In the 14 autopsy cases, the pulmonary vessels were evaluated histologically\textsuperscript{6,7} and graded according to the criteria of Heath and Edwards.\textsuperscript{7}

With Obstruction to Pulmonary Flow

The walls of the small muscular pulmonary arteries were thinner than normal in eight of the 10 cases of tricuspid atresia with obstruction to pulmonary flow (type Ia, 1 case; type Ib, 8 cases; type IIa, 1 case). In four of these eight cases, small muscular arteries and arterioles were thrombosed (fig. 6; table 1; cases 5, 6, 9, and 11).

Without Obstruction to Pulmonary Flow

Three of the four cases without obstruction to pulmonary flow were type IIb tricuspid
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atria (with transposition) and one was type Ic (without transposition). All patients were less than 2 years of age and showed persistence of the fetal state of the small pulmonary arteries and arterioles which was manifested by a thick muscular media (case 10; fig. 7). No intimal changes or other vascular lesions were found.

Comment

In tricuspid atresia in the absence of pulmonary vascular disease, the blood flow into the right ventricle is governed, in part, by the size of the ventricular septal defect. In this study the size of the ventricular septal defect was expressed as a fraction of the diameter of the aortic valve, which was different for each of the five anatomic subgroups. In type Ia (one case; no transposition of the great arteries, pulmonary atresia) no defect was present. In type Ib (no transposition of the great arteries, subpulmonary stenosis) the ventricular septal defect was less than 50% of the diameter of the aortic valve. In type Ic (one case; no transposition of the great arteries, no pulmonary stenosis) the defect was more than 50% of the diameter of the aortic valve. In all cases of type II (transposition of the great arteries) the ventricular septal defect was larger than in any of the cases of type I and of the same size or even greater than the aortic orifice.

In type Ib the ventricular septal defect represented the only intracardiac obstruction to pulmonary flow. Thus, the stenotic region was the narrow, channel-like ventricular septal defect. The term “subpulmonary stenosis,” when used with this type of tricuspid atresia, refers to the obstruction caused by the ventricular septal defect.

The size of the right ventricle appeared to be dependent on the size of the ventricular septal defect where the size of the ventricular septal defect was expressed as a fraction of the diameter of the aortic valve. The right ventricle was largest in type II (transposition of the great arteries), and this group also had the largest ventricular septal defects. Theoretically, the aforedescribed relationship between the size of the ventricular septal defect, the type of tricuspid atresia, and the size of the right ventricle would not be maintained if the ventricular septal defect were to undergo partial or complete spontaneous closure. Spontaneous closure9 and narrowing8 of the ventricular septal defect in tricuspid atresia have been reported.

In many instances of tricuspid atresia (9 of our 14 cases), a minute, endocardial-lined pocket (dimple) generally has been considered to mark the theoretic site of the absent tricuspid valve. The dimple is situated just anterior to the ostium of the coronary sinus. Thus, it is in the area in which the atrioventricular node is located in the normal heart. The region of the membranous septum, which normally is related to the right atrium, septal parts of the tricuspid valve, and the left ventricle, is abnormal. No true membranous septum could be identified in 12 of the 14 hearts that we studied and in the remaining two it was diminutive. In view of these abnormalities in the region in which the proximal portions of the major atrioventricular conduction system normally are located, anatomic abnormalities in the atrioventricular conduction tissue could be expected in tricuspid atresia.

Other cardiovascular malformations observed in this series included a right aortic arch in two cases (cases 2 and 5) and a left patent ductus arteriosus in three instances (cases 1, 2, and 5). These associated anomalies have been reported previously.8 No examples of the following associated malformations, which also have been reported previously, were present in this study: coarctation of the aorta,10 origin of both great arteries from the left ventricle,11 and dextrocardia and common atrium.12

The histologic findings in the pulmonary vessels in cases of tricuspid atresia with obstruction to pulmonary flow were similar to those reported by Heath13 and Wagenvoort14 and their associates in cases of tetralogy of Fallot and pulmonary stenosis. In the presence of obstruction to pulmonary flow, the
small muscular arteries tended to be thin-walled and were thrombosed in half the cases. In general, morphologic evidence of severe elevation of pulmonary vascular resistance was not found in infants who had tricuspid atresia with increased pulmonary flow. In instances without obstruction to pulmonary flow, pulmonary vascular disease might develop at later ages than we encountered in this study.

References
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Circulation. 1968;38:977-986
doi: 10.1161/01.CIR.38.5.977

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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