Hypoplasia of the Inflow Portion of the Right Ventricle: An Angiocardiographic Sign of Tricuspid Atresia

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SUMMARY
Atresia of the tricuspid valve is constantly associated with a large interatrial communication and an underdeveloped right ventricle. The ventricle adjacent to the right atrium is usually represented by a solid mass of myocardium without a lumen and, therefore, remain unopacified on an angiocardiogram. The resulting triangular lucency between the contrast-filled right atrium and left ventricle is the angiocardiographic hallmark of tricuspid atresia.

Additional Indexing Words:
Atrial shunt Congenital hypoplasia

THE DIAGNOSIS of atresia of the tricuspid valve is most accurately established by angiocardiography. The sequence of opacification of the cardiac chambers, reflecting the pattern of intracardiac blood flow, is quite characteristic. Contrast material injected into the blind right atrium cannot enter the right ventricle directly and must flow through an interatrial communication into the left side of the heart. The right ventricle is the last chamber to be visualized, opacified blood reaching it from the left ventricle by way of a defect in the ventricular septum. In some cases, especially infants and children with rapid circulation times, the diagnosis cannot be made in this manner because it is difficult to determine the exact order in which the cardiac chambers become filled on the angiocardiogram. However, absence of the tricuspid orifice is always associated with a maldevelopment of the right ventricle and this produces an abnormality on the angiocardiogram which is readily detected and is pathognomonic for tricuspid atresia.

The normal right ventricle can be divided into two portions, a heavily trabeculated inflow tract which includes the tricuspid valve, and a relatively smooth outflow tract leading to the pulmonic valve. The two regions of the ventricle have different embryologic origins. The inflow tract is derived from the primitive ventricle. As the heart develops, the lumen of the ventricle enlarges by progressive excavation and resorption of the surrounding embryonic cardiac tissue. This resorption is probably stimulated, at least in part, by the flow of blood through the area. If the tricuspid orifice is not patent, the primitive right ventricle is not hollowed out normally, the lumen of the inflow tract is underdeveloped, and this portion of the ventricle is represented in the mature heart by a solid mass of myocardium.
The outflow portion of the ventricle is the adult derivative of the conus arteriosus, a segment of the original cardiac tube upstream to the primitive ventricle. Most commonly, when the tricuspid valve is atretic, the upper portion of the ventricular septum is incomplete, allowing blood to flow from the left ventricle into the conus portion of the right ventricle. If the interventricular shunt is large, the outflow tract of the right ventricle can attain considerable size. The pulmonic valve is often normal. However, when the tricuspid valve is atretic and the ventricular septum is completely formed and intact, blood cannot enter the right ventricle, and neither the outflow nor the inflow portions will develop.

The right ventricle then is represented only by an endocardial-lined slit within the wall of the single left ventricle, and the pulmonic valve is atretic.

The inflow and outflow portions of the right ventricle can be separated on the frontal angiogram by a line extending from the upper aspect of the tricuspid valve downward and to the left, to a point a short distance above the ventricular apex (fig. 1). Thus, it is apparent that the entire diaphragmatic surface of the ventricle is a part of the inflow tract. The right atrium and the left ventricle lie on either side of the inflow portion of the right ventricle. When all three chambers are opacified, their shadows completely fill the inferior aspect of the cardiac silhouette on the frontal angiogram, forming a continuous density that is interrupted only by the

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**Figure 1**

Selective right ventricular angiogram, frontal projection. A venous catheter has been advanced through the tricuspid valve (T) into the right ventricle. A line drawn from the upper margin of the tricuspid valve, downward and to the left, indicates, approximately, the boundary between the inflow (In) and outflow (O) portions of the ventricle.

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**Figure 2**

Tricuspid atresia. Venous angiogram, frontal projection. Contrast material reaching the right atrium (RA) from the inferior vena cava (IC) has flowed across the atrial septum and opacified the left atrium (LA) and the left ventricle (LV). The triangular lucency (arrow) between the right atrium and the left ventricle represents the hypoplastic inflow portion of the right ventricle.
Tricuspid atresia with transposition of the great vessels. (Top left) Right atrial angiocardiogram, frontal view. Contrast material flows from the right atrium (RA) into the left atrium (LA) and left ventricle (LV). The triangular lucency (arrow) adjacent to the right atrium is diagnostic of tricuspid atresia. The right ventricular cavity cannot be identified. There is considerable reflux of contrast material from the atrium into the hepatic veins (H). A is the aorta. (Top right) Selective right ventriculogram, lateral projection. The catheter was introduced into the brachial artery and advanced into the aorta. Because the vessels are transposed, when the catheter passed through the aortic valve (arrows), it entered the right ventricle (R). This chamber is small and is formed only by the outflow portion of the ventricle. There is a defect in the ventricular septum (D) and the pressure of the injection has forced some contrast material through it into the left ventricle (LV). The pulmonary artery (P) arises from the left ventricle, posterior to the aorta. The subpulmonic region (S) is stenotic. The aortic valve has not yet opened, and only its undersurface is outlined by the contrast material. (Bottom right) Right ventriculogram, frontal view. The diminutive right ventricular chamber is outlined by arrows and is projected directly over the left ventricle (LV). The aorta and the main pulmonary artery are superimposed in this view.

Lucencies of the tricuspid leaflets between the atrium and right ventricle, and the apical
Tricuspid atresia. Selective left ventriculogram, frontal projection. A venous catheter has been manipulated across an atrial septal defect and through the mitral valve. The left ventricular chamber (LV) is capacious, and there is a large left-to-right shunt through a ventricular septal defect. The opacified right ventricle (RV) reaches the diaphragmatic surface of the heart. S is the ventricular septum; P, pulmonary artery; A, aorta.

portion of the septum between the two ventricles. If the inflow portion of the right ventricle does not develop, a radiolucent gap will exist between the opacified right atrium and left ventricle.

A triangular lucency adjacent to the right atrium, appropriately named the right ventricular window, is indicative of atresia or, in rare instances, severe congenital stenosis of the tricuspid valve (figs. 2 and 3, top left). When the right ventricular cavity is small, the rudimentary chamber is located near the base of the heart and does not extend to the diaphragm (fig. 3). In this instance, the right ventricular window will be seen on all phases of the angiocardiogram.

In some cases of tricuspid atresia, the volume of the interventricular shunt is large and the cavity of the right ventricle, although incompletely formed, may be capacious and can reach the diaphragmatic surface of the heart (fig. 4). Once this chamber becomes opacified, the triangular lucency between the right atrium and left ventricle will be filled in. However, because the right ventricle is the last chamber to be opacified, early films, made when only the atria and the left ventricle are filled with contrast material, will show the triangular lucency indicative of tricuspid atresia.

The right ventricular window can also be partially obscured by opacification of the coronary sinus. The sinus lies in the atroventricular sulcus on the diaphragmatic aspect of the heart, immediately to the left of the right atrium. When the tricuspid valve is atretic, it is not uncommon for blood to be forced retrograde from the right atrium into the coronary sinus. The sinus becomes dilated and is projected, in the frontal view, over the area normally occupied by the inflow portion of the right ventricle. However, the appearance of the sinus is quite different from that of a trabeculated ventricular chamber, the sinus casting a sharply defined, smooth shadow (fig.
5). The sinus is usually not large enough to completely conceal the space left by absence of the inflow portion of the right ventricle.

Whenever there is an atrial septal defect and the right atrial pressure is sufficiently elevated, contrast material injected into the right atrium will flow directly into the left atrium, regardless of the underlying cause. However, so long as the tricuspid orifice is patent, some of the opacified blood will also flow, at the same time, into the right ventricle. Therefore, the presence of a lucent triangle between the right atrium and the left ventricle will not be seen, despite the right-to-left atrial shunt, except in tricuspid atresia.

References
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