Radiologic Notes in Cardiology

Angiographic Differentiation Between Tetralogy of Fallot and Double-Outlet Right Ventricle
Relationship of the Mitral and Aortic Valves

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SUMMARY
The anatomic hallmark of a double-outlet right ventricle is the separation of the mitral and aortic valves by a band of myocardium. The two valves remain in contact with each other in tetralogy of Fallot no matter how far anteriorly the aorta is displaced. The mitral-aortic relationship can usually be well demonstrated in both anomalies by selective right ventricular angiocardiography.

The clinical and hemodynamic pictures associated with a double-outlet right ventricle with pulmonic stenosis are often identical to that seen in tetralogy of Fallot. In both of these complexes, the aorta arises at least in part from the right ventricle, a high ventricular septal defect is present, and the pulmonary outflow tract is narrowed. In general, the techniques for surgical correction of the lesions are somewhat different and, not uncommonly, the ultimate prognosis is more favorable in the tetrad. The two anomalies can be differentiated quite accurately by angiocardio- 

draphy if both the aortic and mitral valves are visualized.

Embryologic Considerations
In the early embryo, the heart is represented by a single tube which becomes folded upon itself. The ventricles at this stage are aligned in series. Blood flows from the common atrium into the primitive left ventricle, which must empty across the incomplete ventricular septum into the right ventricle. The only channel of egress from the heart is the bulbus (conus) cordis, connecting the right ventricle with the truncus arteriosus. Both the pulmonary artery and the aorta eventually form from the truncus. In other words, both great vessels, in essence, arise from the right ventricle at this stage in development.

Where the cardiac tube is bent most sharply, its wall becomes folded upon itself and results in a ridge of embryonic tissue that projects into the lumen. This ridge lies at the junction of the bulbus and the right ventricle and is termed the bulboventricular flange (fig. 1). As the heart matures, this flange is resorbed and is not represented in the adult heart.

The bulbus is divided into two by the growth of opposing ridges of mesenchymal tissue which join to form the conus septum. The cardiac end of the septum joins the ventricular septum, while its upper end becomes continuous with a similar septum dividing the truncus. The anterior channel of the conus connects the right ventricle with the pulmonary artery. Largely because of the alignment of the conus septum, the posterior conus channel, which leads to the aorta, becomes transferred to the left ventricle (fig. 2). This channel becomes foreshortened as the heart develops, allowing the aortic valve to descend to its final position adjacent to the mitral valve.

In order for the transfer of the posterior conus channel to be accomplished, the bulbo-ventricular flange must resorb. If the flange is
not resorbed, both conus channels remain with the right ventricle and both great vessels will arise from this chamber. The persistent bulboventricular flange will develop into myocardium which is interposed between the aortic and mitral valves in the mature heart.

Tetralogy of Fallot is due to a single embryonic fault, displacement of the conus septum, which develops too far anteriorly. As a result, the conus is divided unequally and the anterior channel is compromised, leading to stenosis of the outflow portion of the right ventricle. Because of its anterior position, the cardiac end of the conus septum is not aligned properly with the ventricular septum, leaving a sizable defect between the two ventricles. The position of the aortic valve is largely determined by the conus septum and, therefore, is also displaced anteriorly, arising astride the ventricular septal defect. The bulboventricular flange, however, does resorb normally, and the aortic and mitral valves are in continuity.

**Angiocardiography**

In the normal heart, the aortic and mitral valves lie adjacent to each other. Actually, the anterior mitral leaflet and a portion of the noncoronary cusp insert on a common tendinous ring. This can be demonstrated angiographically.

The mitral valve can be identified on a selective left ventricular angiocardiogram, especially during diastole. As the leaflets swing open, they trap contrast material between themselves and the wall of the ventricle. The orifice of the valve is filled with nonopaque blood entering from the left atrium, and the trapped contrast material is seen as a ring surrounding this lucency (fig. 3). This mitral ring indicates the line of attachment of the leaflets. The aortic valve is also best seen during diastole, as the cusps remain relatively quiet when in the closed position. During systole, there is considerable motion and vibration of the cusps so that they are blurred and cannot be clearly identified. Before ejection of the contrast material occurs, the valve cusps are outlined only on their cardiac surface by the opaque blood in the left ventricle. When there is contrast material in both the aorta and ventricle, the cusps are seen as curvilinear lucent lines against the background of opacified blood above and below.
Figure 3

Normal selective left ventricular angiogram, lateral view. During diastole, the line of attachment of the mitral leaflets is seen as an opaque ring (arrow heads) surrounding the radiolucent blood within the valve orifice. In this early film, the ventricle has not yet contracted, and there is no contrast material in the aorta. Only the undersurface of the aortic valve is outlined (arrows). The aortic and mitral valves are in direct continuity.

Both the aortic and mitral valves can be identified in any projection of a left ventricular angiogram. However, in the frontal view the upper portion of the mitral ring, the area that is normally in contact with the aortic valve, is obscured by the opacified outflow tract of the left ventricle. In addition, if there is separation of two valves, it will occur in the anteroposterior direction and cannot be seen in this view. Although the mitral valve is best studied in the left posterior oblique view, in which it is projected tangentially, any separation of this valve from the aortic valve will be foreshortened and may not be seen.

The right posterior oblique is the optimal projection not only for evaluation of the mitral-aortic relationship, but also for demonstration of the elongated appearance of the left ventricular outflow tract as it extends through the ventricular septal defect when both great vessels arise from the right ventricle. The lateral projection, however, serves almost as well. We prefer to perform biplane studies in the frontal and lateral projections with the patient in the supine position because it is easiest to accurately reproduce these projections on subsequent studies and to compare studies from one patient to the next.

The position of the aortic valve in relation to the ventricular septum, as seen in the lateral view, cannot be used to distinguish a double-outlet right ventricle from a tetralogy of Fallot. In a severe tetrad, the valve may appear to arise almost entirely from the right ventricle. However, no matter how anteriorly the aortic valve is situated, it remains in fibrous continuity with the mitral valve (fig.

Figure 4

Tetralogy of Fallot. Selective right ventricular angiocardiogram, lateral view. There is a sizable right-to-left shunt through a ventricular septal defect. The contrast material in the two ventricles outlines the undersurface of the aortic valve (arrows) which is situated astride the septal defect and overrides the ventricular septum (VS). The mitral valve (M) is in normal continuity with the aortic valve (X). There is marked stenosis of the infundibulum (S) leading to the pulmonary artery (P).
4). The anterior overriding of the aorta is due, in large part, to its increased diameter, the posterior aspect of the valve remaining approximately in its normal position.

In a double-outlet right ventricle, although the aorta arises entirely from the right ventricle, it may appear, on the angiocardiogram, to override the ventricular septum. The mitral valve is in its normal position, on the posterior aspect of the left ventricle, and is separated from the aortic valve by a segment of the ventricular wall. This segment is the adult derivative of the persistent bulboventricular flange, and usually forms the superior margin of the ventricular septal defect (figs. 5 and 6).

An abnormal mitral-aortic relationship is the *sine qua non* of a double-outlet right ventricle, and both valves should appear on the angiocardiogram if the diagnosis is to be established with certainty. This can almost always be accomplished if the left ventricle is separated from the aortic valve

 Contrast material flowing across the ventricular septal defect outlines the mitral valve (M), which is separated from the posterior aspect of the aortic valve by a segment of myocardium (arrows).

**Figure 5**

*Double-outlet right ventricle with infundibular stenosis. Right ventricular angiocardiogram, lateral view. Contrast material flowing across the ventricular septal defect outlines the mitral valve (M), which is separated from the posterior aspect of the aortic valve by a segment of myocardium (arrows).*
Double-outlet right ventricle with infundibular stenosis. Right ventricular angiogram, lateral view. (a) An early film shows the mitral ring (M) outlined by contrast material that has flowed through the ventricular septal defect (V). The mitral valve is separated from the aortic valve (arrows). The root of the aorta is faintly opacified, but largely obscured by the pulmonary artery (P). (b) A later film shows the aorta arising from the right ventricle (R), anterior to the ventricular septum (VS). The separation of the mitral and aortic valves can still be seen (arrows), although the mitral ring is not as well delineated as on the earlier film.

References
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_Circulation_. 1971;43:451-455
doi: 10.1161/01.CIR.43.3.451

_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

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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