Cross-sectional Echocardiography in the Diagnosis of Congenital Heart Disease

Identification of the Relation of the Ventricles and Great Arteries

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SUMMARY Using a mechanical sector-scanner, two-dimensional echocardiograms were obtained from 28 normal subjects, 15 patients with tetralogy of Fallot, 11 patients with complete transposition of the great arteries and six patients with double outlet right ventricle. The image obtained perpendicular to the long axis of the left ventricle at the base of the ventricular septum was superimposed on the image obtained perpendicular to the long axis at the origin of the great arteries. In normal subjects, these superimposed images demonstrated that the aorta originated posterior and to the left of the ventricular septum. In patients with tetralogy of Fallot, the aorta was displaced anterior and to the right resulting in the aorta overriding the ventricular septum. In patients with double outlet right ventricle both great arteries originated anterior to the ventricular septum (i.e., from the right ventricle). In patients with complete transposition, the aorta originated anterior and the pulmonary artery posterior to the ventricular septum. Thus, cross-sectional echocardiography permits noninvasive identification of the relation of the ventricles and great arteries and, therefore, provides important information for the diagnosis of patients with congenital heart disease.

RECENT DEVELOPMENTS IN CROSS-SECTIONAL (two-dimensional) imaging of the heart with ultrasound have allowed an accurate noninvasive visualization of cardiac anatomy that has been particularly useful in evaluating patients with congenital heart disease.** Previous studies have described the application of this technique to the identification of the great arteries and the determination of ventricular situs. We used cross-sectional imaging to determine the relation of the ventricles and great arteries to each other.

Methods

Patient Population

We studied the following groups of patients: 1) 28 normal subjects (13 males and 15 females; four months to 49 years of age), 2) 15 patients with tetralogy of Fallot (nine males and six females; four to 22 years of age), 3) six patients with double outlet right ventricle (three males and three females; two to 19 years of age), 4) 11 patients with complete transposition of the great arteries (seven males and four females; two to 18 years of age).

In each of the patients with congenital heart disease, the diagnosis had been made by angiography, operation, or both, and was not known by the individual performing the cross-sectional study. Patients considered to have tetralogy of Fallot had the aorta overriding a ventricular septal defect plus infundibular pulmonic stenosis. Patients with double outlet right ventricle had normal atrial and ventricular situs, and two great arteries originating from the right ventricle. Subaortic and subpulmonic conus tissue separated both semilunar valves from the mitral valve. Of these six patients, four had either valvular or subvalvular pulmonic stenosis. In five patients the ventricular septal defect appeared to be subaortic while in the sixth it was more closely related to the pulmonic valve. Patients with complete transposition of the great arteries were those with normal atrial and ventricular situs, and D-transposition of the great arteries. Patients with a diagnosis of common or single ventricle, as well as those with dextrocardia, were excluded from this study.

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Echocardiographic Technique

Cross-sectional echocardiographic images were obtained with a mechanical sector-scanner. Studies were performed with each subject lying supine with the head and back elevated 30 degrees. Most subjects were turned partially onto their left side. The scanner was in direct contact with the patient's chest in an intercostal space just lateral to the left sternal border. The scan plane of the sector-scanner was oriented perpendicular to the long axis of the heart (i.e., roughly parallel to a line connecting the patient's left shoulder and right hip). The scanner was tilted in a cephalad direction until the base of the ventricular septum was visualized at or slightly caudad to the level of the mitral valve tip (fig. 1 and fig. 2, left panel). In each patient, the interspace was chosen so that the tips of the mitral valve leaflets could be visualized with the scanner perpendicular to the chest wall. From this level, the scanner was tilted in a cephalad direction until the origins of the great arteries were seen (fig. 2, right panel). In patients with complete transposition of the great arteries and those with double outlet right ventricle, the scan plane usually had to be oriented nearly perpendicular to the sternum in order to visualize clearly the great arteries.

Image Analysis

All studies were recorded on one-half inch video tape via a video camera. During the subsequent analysis of each patient study, the video tape recorder was placed in a stop-frame mode. A simultaneously-recorded QRS marker was used to select two late diastolic frames; one when the scanner was imaging the base of the ventricular septum at or slightly caudad to the tips of the mitral valve leaflets, and the other when the origins of the great arteries were being imaged. With the ventricular septal image on the screen, a sheet of transparent plastic was placed over the video screen and the right and left surfaces of the ventricular septum were traced onto the plastic sheet (fig. 2, left panel). The tape recorder was advanced until the origins of the two great arteries were seen. The outline of the ventricular septum on the plastic sheet was then compared to the image of the great arteries to determine the spatial relation of the ventricular septum and the great arteries (fig. 2, right panel).

Results

Satisfactory visualization of the heart both at the level of the base of the ventricular septum and at the origin of the great arteries was obtained in all 28 normal subjects and in 13 of the 15 patients with tetralogy of Fallot, five of the six patients with double outlet right ventricle, and 10 of the 11 patients with complete transposition of the great arteries. In three patients with complete transposition, the presence of a large ventricular septal defect resulted in unsatisfactory visualization of the base of the ventricular septum. These three patients were excluded from further analysis.

In all 28 normal subjects, the center of the aorta visualized in cross-section was located posterior and to the left of the ventricular septum. The right ventricular outflow tract was seen to course from right to left, anterior to the right side of the ventricular septum (figs. 1 and 2). In each of the 13 patients with tetralogy of Fallot, the center of the aorta was located either between the right and left septal surfaces or

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Figure 1. Drawing of the cross-sectioned heart of a normal subject. The upper drawing illustrates the plane of section relative to external landmarks. The bottom drawing is the cross-sectioned heart viewed from the region previously occupied by the cardiac apex (direction of view is also shown by arrow in upper panel). As a result, the chest wall will be shown at the top of the image, and the patient's right side will be shown on the left of the image in this and all subsequent illustrations. Also in all illustrations, the plane of section has been oriented so that the aorta is sectioned perpendicular to its long axis and hence will always appear as a circle in cross-section. In this normal subject, the aorta originates to the left and posterior to the ventricular septum (i.e., from the left ventricle).

Figure 2. Unretouched cross-sectional echocardiograms of a normal subject obtained in late diastole with the scan plane intersecting the heart at the base of the ventricular septum (left panel) and at the origins of the great arteries (right panel). The right and left surfaces of the ventricular septum are outlined by asterisks in the left panel. These two septal surfaces are shown in the right panel as smaller asterisks that are superimposed on the image of the origins of the great arteries. Note that in normal subjects the aorta originates posterior and to the left of the ventricular septum. RV = right ventricle, VS = ventricular septum, LV = left ventricle, MVA = mitral valve apparatus, RA = right atrium, RVO = right ventricular outflow tract, AWA = anterior wall of aorta, Ao = aortic root, PWA = posterior wall of aorta, LA = left atrium.
slightly to the right and anterior to the right side of the septum. Although a portion of the right ventricular outflow tract was narrowed in these patients, its overall course was similar to the normal subjects (figs. 3 and 4). Also, the cross-sectional images indicated that the aorta was not displaced entirely to the right of the ventricular septum in any of the patients with tetralogy of Fallot. In each of the five patients with double outlet right ventricle, the center of the images of both the aorta and the pulmonary artery (and the right ventricular outflow tract) were located anterior and to the right of the ventricular septum (figs. 5 and 6). In contrast to patients with tetralogy of Fallot, the entire aorta was displaced into the right ventricle and did not override the ventricular septum. In the seven patients with complete transposition of the great arteries, the center of the cross-sectioned aorta was located anterior to the right septal surface while the center of the cross-sectioned pulmonary artery (and the right ventricular outflow tract) was located posterior to the left septal surface (figs. 7 and 8).

**Discussion**

The results of the present study indicate that cross-sectional echocardiography may be used to determine the ventricle from which each great artery originates and thus is of considerable utility in the differential diagnosis of certain congenital malformations of the heart. For example, patients with tetralogy of Fallot were identified by demonstrating overriding of the ventricular septum by the aorta. This feature of tetralogy of Fallot has been previously documented by both M-mode echocardiographic sweeps and cross-sectional echocardiographic images obtained parallel to the long axis of the left ventricle. The method described in this study provides additional and complemen-

![Figure 3](image-url)  
**Figure 3.** Drawing of the cross-sectioned heart of a patient with tetralogy of Fallot. The upper drawing shows the plane of section relative to external landmarks. The bottom left drawing illustrates the cross-sectional anatomy at the level of the tip of the mitral valve. The bottom right drawing illustrates the anatomy at a slightly more cephalad level through the ventricular septal defect. In tetralogy of Fallot the aorta is displaced anterior to the right side and thus overrides the base of the ventricular septum.

![Figure 4](image-url)  
**Figure 4.** Unretouched cross-sectional echocardiograms of a patient with tetralogy of Fallot obtained in late diastole with the scan plane intersecting the heart at the base of the ventricular septum (left panel) and at the origins of the great arteries (right panel). In the right panel, the aorta is seen to override the outlined surfaces of the ventricular septum. The image in the left panel was chosen because it clearly illustrates the ventricular septum, even though the mitral valve apparatus is not demonstrated in detail.

![Figure 5](image-url)  
**Figure 5.** Drawing of the cross-sectioned heart of a patient with double outlet right ventricle. The bottom drawing illustrates that the aorta and pulmonary artery are side-by-side at their origin and that both originate anterior to the ventricular septum (i.e., from the right ventricle).
Diagnosis of double outlet right ventricle by cross-sectional echocardiography was based on two findings: 1) the aorta originated from the right ventricle and did not over-ride the ventricular septum and 2) the pulmonary artery also originated from the right ventricle. Although double outlet right ventricle can often be suspected clinically, diagnosis usually has been made by angiography or by direct examination at operation.\textsuperscript{31-38} Recently, an echocardiographic technique for diagnosing double outlet right ventricle has been described.\textsuperscript{10, 13, 14, 17, 18, 30-38} This method involves detecting apparent discontinuity between anterior mitral leaflet and posterior aortic wall by slowly angling the ultrasound beam from the mitral valve to the aortic root. However, technical difficulties with this diagnostic sign have been encountered.\textsuperscript{17, 18, 22-35} The cross-sectional echocardiographic technique described in this study does not involve the detection of aortic-mitral discontinuity per se but rather is based on relating the spatial position of the origin of the great arteries to the position of the base of the ventricular septum. We believe this method may obviate many of the problems associated with defining and identifying aortic-mitral discontinuity. Thus even though we have not studied patients with the form of double outlet right ventricle characterized by aortic-mitral continuity via fibrous tissue extending through a defect in the ventricular septum,\textsuperscript{22} we believe this method likely will be useful even in this variant of double outlet right ventricle.

In patients with complete transposition of the great arteries, the aorta originates from the right ventricle. However, the pulmonary artery is usually located posterior relative to the aorta and hence originates posterior to the ventricular septum (i.e., from the left ventricle).\textsuperscript{36, 37} This is in contrast to patients with double outlet right ventricle in whom the pulmonary artery is usually positioned lateral to the aorta and hence is anterior to the ventricular septum.\textsuperscript{38, 39}
The identification of great arteries that are positioned in an anterior-posterior relation has been used as the diagnostic marker for complete transposition of the great arteries in both M-mode and cross-sectional echocardiographic studies. While the identification of anterior-posterior related great arteries is a useful marker for this malformation, semantic confusion may occur. For example, one of the patients in this study had anterior-posterior related great arteries (as in complete transposition) that both originated from the right ventricle (fig. 9). In this patient, cross-sectional echocardiography allowed us to correctly identify that the great arteries were not only transposed but that they both originated to the right of the ventricular septum (i.e., from the right ventricle).

The cross-sectional images shown in this manuscript were obtained perpendicular to the long axis of the heart at two levels: 1) the base of the ventricular septum and 2) the origin of the great arteries. The images at these two levels allowed the ventricular origin of each great artery to be determined. However, images at other levels and with different orientations of the scanner were also very useful in diagnosing patients with congenital malformations of the heart. For example, the two great arteries were seen in cross-section as two side-by-side circles in patients with double outlet right ventricle (fig. 6, right panel and fig. 10, left panel). Two side-by-side circles are also seen in patients with corrected transposition of the great arteries (unpublished observations). By angling the scanner in a slightly more cephalad direction it was possible to visualize the course of the great arteries and identify the pulmonary artery by the fact that it coursed in a posterior direction toward the lungs. (Although the transverse portion of the aortic arch also courses in a posterior direction, it does so at a much more cephalad level and in fact is often difficult to visualize from the anterior chest wall.) Thus patients with double outlet right ventricle could be identified and distinguished from those with corrected transposition of the great arteries by demonstrating that the lateral (and not the medial) great artery coursed in a posterior direction toward the lungs (fig. 10, right panel).

Another situation in which additional cross-sectional views clarified the diagnosis is illustrated in figure 4. In the right panel of this figure, signal drop-out resulted in suboptimal visualization of the anterior wall of the aorta. In addition, a portion of the right ventricular outflow tract was narrowed and hence difficult to visualize. By scanning medially and laterally in this patient, it was possible to visualize the right ventricular outflow tract on either side of the narrowing and thus determine that the outflow tracts in this patient crossed perpendicular to each other. This information combined with the information about the relation of the great arteries and ventricular septum allowed the diagnosis of tetralogy of Fallot to be made. Thus, as in most diagnostic decisions, the combination of several pieces of information facilitates diagnosis.

Although the images included in this manuscript were all obtained perpendicular to the long axis of the heart, it should be emphasized that images parallel to the long axis of the heart contain similar diagnostic information. For example the parallel view allows the detection of discontinuity between the ventricular septum and the anterior wall of the aorta (overriding) in patients with either tetralogy of Fallot or double outlet right ventricle. Also, the long axis view allows visualization of the course of the aorta and thus

Figure 9. Unretouched cross-sectional echocardiograms of a patient with transposition of the great arteries and double outlet right ventricle. The surfaces of the ventricular septum are outlined by asterisks in the left panel. In the right panel, the dilated aorta is located directly anterior to the small pulmonary artery as in complete transposition of the great arteries. However, both great arteries clearly originate well to the right of the ventricular septum.

Figure 10. Unretouched cross-sectional echocardiograms of a patient with double outlet right ventricle. The image on the left was obtained at the origins of the great arteries and demonstrates two side-by-side great arteries. The image on the right was obtained with the scan plane intersecting the heart at a slightly more cephalad level. The lateral vessel (pulmonary artery) is seen to course in a posterior direction before bifurcation into the right and left pulmonary arteries. MPA = main pulmonary artery, RPA = right pulmonary artery, LPA = left pulmonary artery.
facilitates identification of the great arteries. In our experience, it was difficult occasionally to distinguish patients with double outlet right ventricle from those with tetralogy of Fallot using only the parallel view of the heart at the base of the ventricular septum. As previously emphasized, however, this distinction could be made by imaging the heart with several different scanner positions.

The ventricular-great artery patterns most commonly seen in the four groups of patients in this study are summarized in figure 11 in terms of the amount of twisting or spiraling of the great arteries and their outflow tracts relative to each other. The drawings on the left side of this diagram were obtained by copying the cross-sectional images, while the drawings on the right were derived from diagrams made by cardiac pathologists. As emphasized in this diagram, the least amount of twisting occurs in patients with complete transposition of the great arteries and the maximum twisting occurs in normal subjects. Patients with double outlet right ventricle and tetralogy of Fallot appear intermediate between these two extremes. Also, the diagram emphasizes that it is now possible to obtain cross-sectional images of the heart at several levels and describe the abnormalities in a manner similar to that employed by pathologists at necropsy. This cross-sectional approach is particularly applicable to the segmental diagnosis of congenital heart disease since (as described in this and previous papers) it provides an improved noninvasive method for determining ventricular situs and ventricular-great artery connections.

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Echocardiographic Assessment of a Normal Adult Aging Population

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SUMMARY Echocardiograms were performed on 105 male participants in the National Institute on Aging's volunteer Longitudinal Study Program. All subjects (25-84 years of age) were physically active and had no evidence of hypertension or cardiovascular disease. Measurements were made of the initial diastolic (E-F) slope of the anterior mitral valve leaflet, the aortic and left ventricular cavity dimensions, and the thickness of the posterior left ventricular wall. Fractional shortening of the minor semi-axis of the left ventricle and the velocity of circumferential fiber shortening were also determined.

AS THE NUMBER AND PROPORTION of aged individuals in the population increases, knowledge of normal physiologic changes associated with aging becomes more important. Cardiovascular disease states can only be identified in an aging population, in fact, with reference to age-adjusted norms. However, characterization of the effect of age on cardiovascular structure and function is difficult because of the necessity to exclude those elderly subjects with cardiovascular disease and because invasive studies are associated with increased risk to these patients. Noninvasive phonocardiographic and carotid pulse tracing studies have indicated a prolongation of mechanical systolic and diastolic time periods with increasing age.1-4 The effect of age on the ballistocardiogram has also been thought to indicate prolonged ejection.4 Invasive studies using stroke volume and cardiac output as indicators of resting left ventricular function have been inconclusive. Most studies have shown a

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