Echocardiographic Recognition of Mitral-Semilunar Valve Discontinuity
An Aid to the Diagnosis of Origin of Both Great Vessels from the Right Ventricle

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SUMMARY
The angiographic recognition of mitral-semilunar valve discontinuity is a strong indication of origin of both great vessels from the right ventricle. This study shows that the same finding may be elicited by echocardiography, whether the great vessels are normally related or transposed. When mitral-semilunar valve continuity is present, the mitral valve echo at the onset of systole is continuous with and at the same depth as echoes from the posterior margin of the aorta (the great vessels being normally related) or from the pulmonary artery (the great vessels being transposed). In five cases where both great vessels originated from the right ventricle, echocardiography demonstrated mitral-semilunar valve discontinuity, with the posterior border of the adjacent great vessel lying anterior to the mitral valve echo. The degree of separation measured on the angiogram and by ultrasound was virtually identical. This atraumatic technique is a valuable adjunct to the evaluation of patients with complex congenital malformations, particularly when mitral-semilunar valve relationships are difficult to assess angiographically.

Additional Indexing Words:
Double-outlet right ventricle
Taussig-Bing malformation
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Ultrasound

In the angiographic investigation of patients with cardiac malformations, the presence of a large ventricular septal defect subjacent to the aorta and the pulmonary artery may obscure the precise ventricular connections of these vessels. Difficulty may thus be encountered in distinguishing between conditions such as tetralogy of Fallot with marked override of the aorta, origin of both great vessels from the right ventricle\(^1,2\) (double-outlet right ventricle),\(^3\) and occasional instances of complete transposition of the great vessels. It has been pointed out that this difficulty may be resolved angiographically by evaluation of mitral-semilunar valve relationships. Characteristically, in cases of origin of both great vessels from the right ventricle there is mitral-semilunar valve discontinuity, whereas in complete transposition and tetralogy of Fallot with marked override of the aorta, mitral-semilunar valve continuity is present.\(^4\)

The role of ultrasound cardiography in cardiac diagnosis is being extended. Recently,
the aortic valve has received considerable attention, and several authors have reported their findings in large numbers of patients with both normal and diseased aortic valves.\(^5\)\(^{-8}\) After identification of the familiar mitral valve echo, the aortic root echo is relatively easy to record by slight rotation of the ultrasound transducer. The ability to accomplish this maneuver is, in fact, a manifestation of the normal continuity between the base of the anterior mitral leaflet and the aortic root and suggested to us the potential for the diagnosis of origin of both great vessels from the right ventricle.

This paper reports the echocardiographic findings in five patients with origin of both great vessels from the right ventricle in whom it was demonstrated that the mitral valve was not continuous with a semilunar valve.

**Materials and Methods**

Two groups of patients were studied. Group 1 consisted of 34 patients, aged 3 months to 9 years, who were placed in the following diagnostic categories:

(a) Clinically normal (13 cases).
(b) Tetralogy of Fallot (nine cases).
(c) Pseudotruncus arteriosus (three cases).
(d) Ventricular septal defect (two cases).
(e) Transposition of the great vessels without pulmonary stenosis (five cases).
(f) Dextrocardia with situs inversus (normal heart) (one case).
(g) Corrected transposition (one case).

Group 2 consisted of five patients whose ages ranged from 4 to 15 years. These patients were placed in two diagnostic categories, as follows:

(a) Origin of both great vessels from the right ventricle with pulmonary stenosis and side-by-side relationship of the aorta and the pulmonary artery (four cases).

These four patients presented with a clinical picture similar to that of tetralogy of Fallot, i.e., cyanosis, clubbing, and on auscultation, an ejection systolic murmur and single second heart sound. Roentgenograms of the chest showed dilatation of the ascending aorta, a concave pulmonary artery segment, and oligemic lung fields. Cardiac catheterization demonstrated severe pulmonary stenosis with equal ventricular pressures and large right-to-left shunts. Angiography, performed in both frontal and lateral views, showed both great vessels arising side by side from the right ventricle, dilatation of the ascending aorta, and a narrow pulmonary trunk. The only route of egress from the left ventricle was via the ventricular septal defect, and the anterior leaflet of the mitral valve was separated from the root of the aorta. The diagnosis was confirmed at surgery in all four patients.

(b) Origin of both great vessels from the right ventricle without pulmonary stenosis and transposed relationship of the aorta and the pulmonary artery (1 case).

This case was an example of the Taussig-Bing anomaly, but instead of the classical side-by-side relationship of the great vessels, these were transposed. This boy of 15 years presented with cyanosis and clinical signs of a large ventricular septal defect with severe pulmonary hypertension. Chest roentgenograms revealed pulmonary plethora and an egg-shaped heart with a narrow vascular pedicle. Cardiac catheterization demonstrated identical ventricular pressures, no pulmonary stenosis, a higher oxygen saturation in the pulmonary artery compared with that found in the aorta, and a pulmonary-to-systemic flow ration of 4/1. Angiography showed both great vessels arising from the right ventricle, with the aorta to the front and to the right of the pulmonary artery. The aortic valve was slightly higher than the pulmonary valve, and there was a subpulmonary ventricular septal defect. Mitral-semilunar valve discontinuity was present.

Ultrasound examinations were performed on these patients in the supine position and without sedation. A commercially available ultrasonic apparatus, transmitting bursts of 2.25 MHz vibrations 1,000 times/sec through a transducer 0.75 inches in diameter, was used according to techniques described previously.\(^9\) A water-soluble gel was used to obtain airless contact between the transducer and the skin. The reject control was set at maximum to obliterate low-intensity echoes. Sensitivity for display of near-field signals was adjusted by using depth compensation control to amplify echoes within the first 5 cm from the chest wall; adjustment of the "near-gain" control controlled the signals in this field.

The technique for locating the aortic root ultrasonically followed exactly the description of Gramiak.\(^5\)\(^{-7}\) The mitral valve echo was first obtained by placing the transducer in the left third or fourth interspace.\(^10\)\(^{-12}\) In the normal patient, slight medial and cephalic rotation of the transducer then passes the beam through the aortic root, which is recognizable as a pair of undulating signals moving anteriorly in systole and posteriorly in diastole. The posterior aortic root wall echo lies at the same depth as the closed position of the mitral valve at the onset of ventricular systole, and rotation of the transducer...
blends the mitral valve echo with the posterior component of the aortic root echo (fig. 1).

In the 39 patients studied in this series the mitral valve echo was located first, and the transducer was then angulated to locate the echo of the continuous or adjacent great vessel. Once the root of this vessel had been located, the points of most posterior excursion of the anterior mitral leaflet and the posterior margin of the great vessel at the onset of systole were carefully noted in the A-mode of the echocardiograph. With the apparatus then set on the B-mode, the beam was passed from the root of the great vessel to the anterior leaflet of the mitral valve or vice versa, and a polaroid photograph was taken of this sweep (fig. 1). Any difference in depth of the most posterior points of excursion of these two structures was then measured along the depth scale (graduated in centimeters). Other than the patients who were clinically normal and not catheterized, any degree of mitral-semilunar discontinuity was measured from the angiograms after appropriate allowance had been made for magnification. The measurement was compared with a similar measurement made ultrasonically. The depth calibration of the ultrasonic apparatus was checked by positioning of the transducer on the surface of a lucite test block of standard length. Signal reflection from the end of the block is shown as an echo correlating with the depth markers on the horizontal axis of the A-mode.

In the two patients with dextrocardia, the transducer was placed in the right parasternal position. In the one patient with corrected

Figure 1

Normal echocardiograms showing mitral-semilunar valve continuity. (A) Angulation of transducer from aortic root to mitral valve. (B and C) Composite recording of angulation of transducer from mitral valve to aorta. (D) Anatomic sketch illustrates transducer position for location of mitral valve in open and closed positions (1 and 2, respectively); tilting the transducer medially and cephalically locates the aortic root. The mitral valve echo is continuous with and at the same depth as the posterior margin of the aortic root at the onset of systole.

Abbreviations: Ao = aorta; MV = mitral valve; LA = left atrium; LV and RV = left and right ventricles, respectively; pma = posterior margin aortic root.
transposition, the right parasternal position was also used because the mitral (systemic) atroventricular node is right-sided and the heart is partially displaced to the right because of dextroversion.

Results

Group 1

The mitral valve and aortic root (or pulmonary root in transposition) were located in all 34 patients. In all cases the mitral valve echo blended with the posterior component of the aortic or pulmonary root echo, and at the onset of ventricular systole these two echoes were located at the same depth.

Group 2

In the five patients with double-outlet right ventricle there was echocardiographic evidence that the mitral valve was not attached to a great vessel; the closed (systolic) position of the mitral valve lay deeper to the posterior margin of the adjacent great vessel (figs. 2 and 3). In figures 2 and 3, the vertical distance between the arrowheads pointing to the closed position of the mitral valve and the posterior margin of the aortic root, is the measurement of interest and represents the degree of mitral-semilunar valve discontinuity. In contrast to group 1, in group 2(a), after identification of the mitral valve echo, more rotation of the

Figure 2

Echocardiograms in double-outlet right ventricle with mitral-semilunar valve discontinuity. (A and B) Angulation of transducer from mitral valve to aorta. (C) Anatomic sketch illustrating double-outlet right ventricle and side-by-side relationship of great vessels. After identification of mitral valve echo, medial and cephalic rotation of transducer locates the aorta situated anterior to closed position of mitral valve. Black arrows = closed position of mitral valve; white arrows = posterior margin of aortic root; PA = pulmonary artery.
transducer was required to locate the adjacent great vessel. In two of these three patients the root of the ascending aorta overlapped the ventricular septum. In the one case in group 2(b) there was transposed relationship of the great vessels. Because the pulmonary valve was slightly lower than the aortic, identification of the mitral valve echo followed by slight cephalic rotation of the transducer recorded both the mitral valve and the pulmo-

Figure 3

Echocardiograms in double-outlet right ventricle with mitral-semilunar discontinuity. Posterior margin of aorta is situated anterior to closed position of mitral valve. Arrows mark closed position of mitral valve and posterior margin of aortic root.

Figure 4

(A) Anatomic sketch and (B) echocardiogram of double-outlet right ventricle, transposed great vessels, pulmonary valve slightly lower than aortic valve. After identification of mitral valve (MV), slight cephalic rotation of transducer records mitral and pulmonary artery echoes in the same plane but at different depths. Enclosed between parallel undulating signals of root of PA is echo from posterior cusp of pulmonary valve moving anteriorly to closure point just before forward movement of opening anterior mitral valve leaflet (MV).
ary artery echoes simultaneously in the same plane of the beam but at different depths. Enclosed between the parallel undulating signals emanating from the root of the pulmonary artery, the echo from the posterior cusp of the pulmonary valve was recorded moving anteriorly to closure point just before the forward movement of the opening movement of the anterior mitral leaflet (fig. 4).

The degree of mitral-semilunar valve discontinuity measured from the angiograms, with the corresponding echocardiographic measurement given in parentheses, was 1.2 (1.0), 1.5 (1.2), 1.0 (1.0), 1.0 (0.8), and 1.8 (2.0) cm, respectively.

Discussion

Normally, the base of the anterior mitral leaflet is in fibrous continuity with the bases of the posterior and left cusps of the aortic valve. In complete and corrected transposition of the great vessels, the pulmonary artery arises from the anatomical left ventricle and the anteriorleaflet of the mitral valve is in fibrous continuity with the pulmonary valve. The relationship of the anterior mitral leaflet to the aortic root (or pulmonary root in transposition) may be observed in good quality angiograms exposed in the lateral view.

The systolic position of the anterior mitral leaflet of the closed mitral valve forms virtually a straight line with the posterior wall of the ascending aorta or the pulmonary artery (fig. 5).

When both great vessels arise from the right ventricle, both semilunar valves are usually located at the same level in the horizontal plane. The position of the great vessels in this condition is variable; they may be side by side or the aorta may be in front and to the right of or in front and to the left of the pulmonary artery. The tricuspid valve is separated from the pulmonary valve by the crista supraventricularis as in the normal heart, but the mitral valve is also separated from the semilunar valves by muscular tissue.

The diagnostic angiocardiographic feature in this malformation is the discontinuity between the anterior mitral leaflet and the semilunar valves, which is best demonstrated in the lateral view.

The identification of mitral-semilunar valve relationship is of more than academic interest since those malformations in which both great vessels arise from the right ventricle are now amenable to surgical therapy, and this relationship may determine the nature of the

Figure 5

(A) Selective left ventricular (LV) angiogram in systole (left lateral position) from a patient with obstructive cardiomyopathy. The aorta (Ao) and attached anterior mitral leaflet (AML) in closed position form a straight line. (B) Selective right ventricular angiogram (right anterior oblique view) in patient with mirror image dextrocardia, situs inversus, and double-outlet right ventricle. The mitral (m) and tricuspid (t) valves are separated (arrows) from the semilunar valves by muscle.

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surgical repair. In transposition of the great vessels with overriding of the pulmonary artery where there is mitral-pulmonary continuity, closure of the ventricular septal defect with the Mustard operation is indicated. When the Taussig-Bing malformation is present, there is mitral-pulmonary discontinuity, and it is necessary to construct a tunnel from the left ventricle to the right side of the pulmonary artery and then to carry out the Mustard procedure.15

Gramiak et al. have clearly demonstrated the feasibility of ultrasonic recognition of the aortic root echo as well as the aortic valve cusps. With electrocardiographic, phonocardiographic, and contrast study techniques, they have shown that medial angulation of the ultrasonic transducer beam from the mitral valve recording position blends the latter echo into one continuous record with the posterior component of the aortic root echo. In its most posterior (closed) position the mitral valve echo is continuous with and at the same depth as the posterior margin of the aortic root.5-7 This finding correlates with the normal angiographic findings and is a manifestation of the fibrous union between the base of the anterior mitral leaflet and the aortic root.

The technique described by Gramiak was used in this investigation. Our purpose in studying the 34 cases in group 1 was primarily to determine the frequency with which we could detect mitral-semilunar valve continuity after identification of the mitral valve echo. The fact that we were able to do so in all 34 cases, including those with cardiac malposition or transposed great vessels, attests to the accuracy of the technique, which is easy to perform in children where problems such as emphysema and thick chest walls do not impose technical difficulties.8 Mitral-semilunar valve continuity in this group was confirmed in all cases investigated angiographically.

In contrast, the patients in group 2 manifested different findings. After identification of the mitral valve echo, medial and cephalic rotation of the transducer located an adjacent vessel whose posterior margin was discontinuous with and lay anterior to the posterior (systolic) position of the mitral valve. Correlation with the angiographic measurement of mitral-semilunar valve separation was excellent and conforms with measurements made pathologically.16

Identification of mitral-semilunar valve relationships by selective angiography may be difficult even with high quality technique, particularly when malposition or rotation of the heart is present. In the investigation of cases of congenital heart disease, it has previously been shown that ultrasound may be of help in identifying the ventricular septum and the number of atrioventricular valves.9 Although ultrasound cannot establish which great vessel is continuous with the mitral valve, the ease and atraumatic manner with which this phenomenon may be detected in children should provide additional help in the investigation of complex cardiac malformations since, with few exceptions, the presence of mitral-semilunar valve discontinuity indicates the presence of double-outlet right ventricle.

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