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
Ultrasound
Transposition of the great vessels

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 be-
tween 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 diagno-
sic 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
concaue pulmonary artery segment, and oli-
gemc 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 subpulmo-
nary 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. 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. 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

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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.

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transposition, the right parasternal position was also used because the mitral (systemic) atrioventricular 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.
mitral-semilunar valve discontinuity

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).
nary 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 anterior leaflet of the mitral valve is in fibrous continuity with the pulmonary valve.4 The relationship of the anterior mitral leaflet to the aortic root (or pulmonary root in transposition) may be observed in good quality angiocardiograms 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.1-2 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.4 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.
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}\n\nGramiak 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}\n
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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