Multiple Crystal Echocardiographic Evaluation of Endocardial Cushion Defect

By David J. Sahn, M.D., Richard W. Terry, M.D., Robert O'Rourke, M.D., George Leopold, M.D., and William F. Friedman, M.D.

SUMMARY

A prototype multiple crystal echocardiographic system developed by Bom and associates was used to evaluate cross-sectional cardiac anatomy in real time in twenty infants and children with endocardial cushion defect (ECD). The findings were compared to fifty normal infants and children and nineteen patients with normal mitral valve anatomy but right ventricular enlargement (RVE). Three standard transducer positions for evaluation of sagittal and transverse cardiac cross-sections are outlined and the normal group and RVE subgroup described. Studies in patients with ECD demonstrated several distinctive abnormalities, consisting of multiple echoes in the mitral valve area, anterior mitral leaflet — septal apposition in diastole with reduced posterior motion in systole, and reduced excursion of the anterior mitral leaflet. In patients with complete atrioventricular (A-V) canal defects, the anterior leaflet was often observed passing across the plane of the ventricular septum into the right ventricle during diastole, and in some of these patients the anterior mitral leaflet was related to the tricuspid annulus. Multiple crystal cardiac ultrasonography allows a more precise determination of intra- and extracardiac spatial relationships and is easier to perform than single crystal echo. The new technique provides clinical information helpful in evaluating patients with possible ECD and gives some indication of the severity of the defect.

Additional Indexing Words:

- Ultrasound, noninvasive
- Cross-sectional echo
- Atrioventricular canal
- Echocardiography
- Multiscan
- Ostium primum atrial septal defect

A wide range of clinical presentations exists in patients with endocardial cushion defect (ECD). Anatomic details may vary from those associated with ostium primum atrial septal defect to the more complex findings in the complete form of common atrioventricular canal. The clinical diagnosis is facilitated by detecting the physical findings of atrial and/or ventricular septal defect with or without mitral regurgitation, and occasionally tricuspid regurgitation. A helpful diagnostic aid is the presence of a superiorly oriented, counterclockwise, frontal plane QRS vector similar to the pattern of left anterior hemiblock. It is clear, however, that not all patients with endocardial cushion anomalies demonstrate these ECG-VCG findings, that occasional patients with atrial septal defects of the secundum type have this abnormal conduction pattern, and that this abnormal vector loop may also commonly be seen in such diverse conditions as double outlet right ventricle, tricuspid atresia, and supravalvar pulmonic stenosis. While the precatheterization diagnosis of most patients with endocardial cushion anomaly is not especially difficult, a distinct group of patients exists in whom the diagnosis may be less clear without resorting to hemodynamic and angiographic investigations. Thus, the present study was undertaken to develop accurate criteria for the diagnosis of endocardial cushion anomalies employing a new, noninvasive, multiple crystal echocardiographic approach (multiscan).

Considerable interest exists in the noninvasive evaluation of cardiac anatomy using ultrasound cardiology. Studies utilizing single crystal echocardiographic techniques have outlined abnormal findings in patients with ostium primum atrial septal defects and common A-V canal defects. Among the most helpful findings have been an enlarged right ventricular cavity, abnormal mitral valve orientation, abnormal septal apposition of the anterior mitral...
leaflet in diastole, multiple mitral valve echoes, and a common mitral-tricuspid leaflet. However, it must be recognized that single crystal echocardiographic methods may be limited in the study of complex alterations in cardiac anatomy because they provide only a "flashlight beam" view that is subject to a variety of artifacts and places a great premium on the expertise of the examiner. Recently, a multiple crystal echocardiographic system has been developed that permits visualization of cardiac cross-sectional anatomy in real time.25-27 The method allows a substantially more precise definition of intracardiac spatial relationships. In patients with ECD, abnormalities in the location and motion of the atrioventricular valves and interventricular septum are recorded easily and accurately. Our experience with the multiscan approach in this group of patients forms the basis of the present report.

Methods

Twenty infants and children with various forms of ECD, ranging in age from seven weeks to fourteen years (table 1), were examined with a prototype multiple crystal echocardiographic system designed by Nicholas Bom.25-27 Diagnoses and anatomic details were assessed and confirmed in each patient by standard hemodynamic and biplane cineangiographic techniques. The ultrasound findings in this group were compared to those observed in a group of fifty normal infants and children and a second group of nineteen patients with normal mitral valve anatomy but right ventricular enlargement due either to secundum atrial septal defect (8 patients) or pulmonic stenosis (11 patients) (age range 7 months to 10 years). A scheme of the multiscan instrument is shown in figure 1. Its use has been described in detail elsewhere.27-28 Sagittal and transverse cross-sectional images of cardiac anatomy viewed in real time on the oscilloscope were stored permanently on video tape and 8 mm motion picture film. Photographic enlargement of the small format motion picture film accounts for the increased grain and reduction of data seen on the illustrative examples accompanying this report. A 4.5 mHz, 20 element transducer was employed in all patients. The system produces images at a frame rate of 80/sec. When fired sequentially, each element generates an independent brightness modulated "B" mode echocardiogram which consists of two lines. A total of forty lines is displayed on the oscilloscope in an arrangement which duplicates their physical distribution. Therefore, the number 1, or most superior element on the transducer surface will generate the first two lines on the oscilloscope cross-section. An electrocardiogram (ECG) is displayed simultaneously on the oscilloscope.

Cross-sectional cardiac anatomy was evaluated in a systematic manner using three standard views:

Position 1, Sagittal: Transducer placed vertically along the left sternal border in line with the long axis of the heart (fig. 2). Position 2, Transverse: Transducer placed horizontally in the fourth intercostal space just to the left of the sternum with the superior crystal to the right (fig. 3). Position 3, Modified Sagittal: Transducer placed obliquely with the bottom element at the third intercostal space and left sternal border and the top element below the left clavicle (fig. 4).

Real time studies were analyzed independently by two observers for (1) right ventricular and left ventricular dimensions (Positions 1 and 2); (2) direction of systolic septal motion at the level of the chordae tendineae (Positions 1 and 2); (3) the excursion, position, and contour of the anterior mitral leaflet (Position 1 and 2); (4) the position and motion of the posterior mitral leaflet (Position 1 and 2); (5) left atrial dimensions (Position 1); and (6) the size of the pulmonary artery (Position 3). In a separate investigation validation of the relationship between echoes and anatomic structures viewed by the multiscan was obtained at hemodynamic study by selective injections of indocyanine green dye and catheter visualization.29 Age related normal values for echocardiographic dimensions were those obtained previously in our laboratory30 and by others.20

Results

Normal anatomy and findings in patients with a normal mitral valve but right ventricular enlargement:

Position 1 — Sagittal

As illustrated in figure 2, this long axis view shows the anterior right ventricle at the left of the image. The interventricular septum (IVS) is the next structure seen posteriorly, and is in continuity with the anterior wall of the aorta. In patients with right ventricular enlargement (RVE), the IVS appears to be more posterior the larger the right ventricular cavity. Tricuspid valve motion is visualized within the cavity of the right ventricle. Right ventricular cavity size is measured at its largest antero-posterior dimension. This dimension may be obtained reproducibly providing the sagittal plane depicts maximal excursion of both leaflets of the mitral valve simultaneously, thus assuring proper long axis orientation. When the same positioning criteria are satisfied, left ventricular dimensions may be measured at the level of the leading edge of the anterior mitral leaflet (AML) (fig.

Table 1

<table>
<thead>
<tr>
<th>Study Groups</th>
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<tbody>
<tr>
<td><strong>Endocardial cushion defect (ECD)</strong></td>
<td></td>
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<tr>
<td>(20 patients)</td>
<td></td>
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<tr>
<td>Ostium primum atrial septal defect</td>
<td>2 patients</td>
</tr>
<tr>
<td>Incomplete atrioventricular canal</td>
<td>8 patients</td>
</tr>
<tr>
<td>Complete atrioventricular canal</td>
<td>10 patients</td>
</tr>
<tr>
<td><strong>Right ventricular enlargement — normal mitral valve</strong></td>
<td></td>
</tr>
<tr>
<td>(19 patients)</td>
<td></td>
</tr>
<tr>
<td>Secundum atrial septal defect</td>
<td>8 patients</td>
</tr>
<tr>
<td>Pulmonic stenosis</td>
<td>11 patients</td>
</tr>
<tr>
<td><strong>Normal cardiac anatomy</strong></td>
<td></td>
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<td>(50 patients)</td>
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2). The left ventricular cavity is posterior to the IVS and mitral valve motion is detected within it. The AML extends inferiorly from the posterior aortic wall and chordae tendineae may often be seen inferior to the leaflets. The posterior mitral leaflet (PML) extends anteriorly from the atrioventricular junction. The left atrium lies posterior to the aorta. The posterior left ventricular wall sweeps inferiorly and anteriorly towards the apex.

A complete description of the appearance of IVS motion in the presence and absence of right ventricular volume overload provides the basis of a separate report. In brief, normal IVS motion is characterized by an anterior systolic motion of the superior portion of the septum which is anchored to the aortic root. Consistently, the IVS is found to be “hinged” so that it pivots at the junction of its upper one third and lower two thirds. During systole, the lower two thirds of the IVS moves posteriorly, i.e., towards the anteriorly moving wall of the left ventricle. With progressive degrees of enlargement of the right ventricle in those patients with right ventricular volume overload, a shift occurs of the pivot point to a lower level of the IVS such that the entire IVS is seen to move anteriorly in systole and no pivot point is observed in patients with the largest volume overload of the right ventricle.

With normal motion of the mitral valve, at end systole the AML is directly inferior to the posterior aortic wall. This leaflet moves anteriorly towards the septum in early diastole and then exhibits a slow posterior sweep during left ventricular filling. Atrial systole produces an abrupt anterior motion of small magnitude. Valve closure brings the AML to a position posterior to the root of the aorta. During ventricular systole the AML gradually moves anteriorly. The motion of the PML is opposite to that described for the AML, i.e., posteriorly in diastole and anteriorly at the onset of systole. The two leaflets exhibit a “hand clapping” motion, moving together in systole and apart in diastole.

Position 2 — Transverse

In this position (fig. 3) a transverse section through the body of the ventricles is obtained at the fourth intercostal space. The tricuspid valve lies at the rightward, anterior portion of the cross section. The right atrium is visualized posteriorly. The interatrial septum is not visualized since it lies in a plane almost parallel to the incident sound energy. An oblique section of the right ventricular cavity bounded by the anterior portion of the IVS lies leftward and anterior to the tricuspid valve. The IVS is visualized only partially in this view and septal motion is difficult to assess. The mitral annulus and the AML are posterior to the IVS and separated from it by the width of the left ventricular outflow tract. The left, posterior por-
Position 3 — Modified Sagittal

In this position the transducer is aligned along the right ventricular outflow tract (fig. 4). The septal leaflet of the tricuspid valve may be seen just below the infundibulum. The right ventricular outflow tract sweeps superiorly and posteriorly and pulmonary cusp tissue marks the position of the valve. The aorta is poorly visualized in this view since it is seen only in partial cross section. Likewise, the interventricular septum is poorly visualized.

Endocardial Cushion Defect (table 2)

Position 1 — Sagittal

In all of the patients with an endocardial cushion defect, the AML produced a distinctive pattern of multiple echoes (fig. 5). No single, discrete AML echo could be recorded irrespective of gain control setting. Apposition during systole of AML and IVS was observed in nineteen of the twenty patients. In these nineteen patients, the AML failed to move to the normal position in systole posterior to the root of the aorta. The effect over-all when viewing moving images was a loss of the normal “hand-clapping” appearance of mitral valve motion.

The excursion of the AML appeared to be reduced in sixteen of twenty patients. In eight of these patients, the reduced excursion was associated with the visualization of echoes from the AML crossing the plane of the IVS during diastole, appearing in the right ventricular cavity (fig. 6). Thus, only the left ventricular aspect of mitral valve motion was observed initially, and further angulation towards the patient’s right was necessary to detect the right ventricular aspect of leaflet excursion. In the remaining patients in whom reduced excursions were noted the finding most likely resulted from an alteration in the axis of valve motion. The PML in the ECD patients was un-

Table 2

<table>
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<tr>
<th>Major and Minor Diagnostic Criteria for the Multiscan Diagnosis of Endocardial Cushion Defect (20 Patients)</th>
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<tbody>
<tr>
<td><strong>Major</strong></td>
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<tr>
<td>1. Decreased mitral excursion (16/20)</td>
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<tr>
<td>2. Multiple echoes in the mitral area with poor leaflet definition (20/20)</td>
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<tr>
<td>3. Mitral-septal apposition or excursion predominantly anterior (19/20)</td>
</tr>
<tr>
<td>4. Passage of the anterior leaflet across the plane of the ventricular septum into the right ventricular cavity (8/20)</td>
</tr>
<tr>
<td>5. Anterior mitral leaflet related to tricuspid annulus on transverse section (4/20)</td>
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</table>
usually prominent and it appeared inferiorly displaced into the left ventricle.

Although right ventricular dimensions were increased in patients with ECD, only four patients were observed to have true paradoxical motion of the entire ventricular septum. Two of these patients had only an ostium primum atrial septal defect.

**Position 2 — Transverse (Horizontal)**

The major findings in this view included multiple AML echoes, an over-all reduction in mitral valve excursion with the direction of AML motion primarily towards the IVS, and increased prominence of echoes from the PML (fig. 7). In this view, in the eight patients in whom the AML was seen in sagittal position to cross the ventricular septum, the phenomenon was also observed during the sweep of the left ventricular outflow tract produced by tilting the transducer superiorly.

In four patients, the AML appeared to be an extension of a large common leaflet that was related to both the tricuspid annulus and the mitral annulus (fig. 7).

**Position 3 — Modified Sagittal**

In this position the transducer was aligned along the right ventricular outflow tract. This view was employed to evaluate the size of the main pulmonary artery in the patients with ECD and the vessel appeared enlarged in thirteen of twenty patients (fig. 8).

**Discussion**

Multiple crystal echocardiography appears to be especially useful for establishing the diagnosis of endocardial cushion defect of whatever anatomical type. The multiscan finding most unique to these anomalies was the easily detectable presence of multiple mitral valve echoes associated with an abnormal excursion of the AML. Additional findings of importance included passage of the AML across the plane of the ventricular septum and the appearance in transverse view of one common atrioventricular valve. Of lesser significance diagnostically were observations of an increase in right ventricular dimension, paradoxical motion of the IVS in systole, and enlargement of the size of the main pulmonary artery.

A range of lesions existed in our study group from ostium primum atrial septal defect with cleft mitral valve, through transitional types of atrioventricular
Figure 6
Position 1: Sagittal section, complete atrioventricular canal. Still frame examples in two different patients of multiple echoes in the mitral valve area with the common leaflet (CL) in the left ventricle during systole (top panel — arrow) and the right ventricle during diastole (bottom panel — arrow). The right ventricle is enlarged in both patients. RV = right ventricle; Ao = aorta; LV = left ventricle; LA = left atrium; MV = mitral valve.

canal malformations, to the complete type of common atrioventricular canal. It should be recognized that the ultrasound findings are substantially more helpful in defining the presence or absence of one of the group of malformations arising from maldevelopment of the endocardial cushions than in describing the precise architectural details that allow separation of the subgroups of cushion anomalies. While the presence of a common A-V leaflet, or passage of the AML across the plane of the IVS, suggested the presence of more serious pathological anatomy, the remaining observations were less specific. In this regard, it must be emphasized that this new noninvasive tool is not offered as a substitute for more definitive hemodynamic and angiocardiographic analysis, but rather as an initial test to establish diagnosis, and as a precatheterization study to more appropriately plan the course of subsequent hemodynamic investigation. Unfortunately, as is well known, all of the complex anatomical details in these patients may not become clear until direct observation by either a surgeon or a pathologist.

Since the ultrasound diagnosis of an endocardial cushion defect centers upon detection of abnormalities in the mitral apparatus, it is appropriate to analyze our findings with respect to the types of derangement that may be observed in this valve’s anatomy in patients with ECD. Mitral valve abnormalities may be categorized in terms of (1) defective anchoring of the AML, (2) abnormalities of leaflet tissue per se, and (3) abnormalities of chordal suspension.

Figure 7
Position 2: Transverse (horizontal) section. Diagram and still frame show multiple echoes in the mitral valve area, right ventricular enlargement, and the relationship of the common leaflet to the tricuspid annulus in a patient with a complete atrioventricular canal. RV = right ventricle; LV = left ventricle; RA = right atrium; LA = left atrium; CL = common leaflet; TA = tricuspid annulus.

Figure 8
Position 3: Modified sagittal section. The enlarged main pulmonary artery (MPA) is well visualized in this view. RV = right ventricle.
Abnormal anchoring of the anterior-inferior portion of the AML is common to all forms of endocardial cushion defect. The absence of the atroventricular septum results in an abnormal line of attachment of the AML such that it is displaced inferiorly and anteriorly. This displacement results in a shift of the orifice of the valve towards the right side of the left ventricle, and a rotation towards the sagittal plane. This change in position of the mitral orifice and the associated elongation of the left ventricular outflow tract produces the gooseneck deformity seen angiographically. The echocardiographic manifestations of these abnormalities in orifice position and orientation are the result of a change in the direction of AML motion with respect to the IVS. Thus, the plane of motion of the AML is more parallel than perpendicular to the plane of the IVS. The altered antero-inferior anchoring of the AML results in the ultrasonic appearance of reduced excursions in the sagittal view, unusual proximity to the ventricular septum, and apposition between AML and IVS in diastole. A major finding in our patients was a reduction in the over-all excursion of the mitral or common A-V valve since motion appeared to be limited in all but an anterior direction.

Abnormalities of leaflet tissue are common in all varieties of A-V canal. Often, deficient anterior cusp tissue separates the leaflet into a superior and inferior segment. Either, or both of these segments may have attachments not only to papillary muscles, but to the superior rim of the IVS as well, which may result in restricting the motion of the affected segment. From an echocardiographic viewpoint, the two leaflet segments, when viewed in a sagittal position, will appear less discrete than a normal AML and their separation and abnormal redundancy produces multiple echoes. In the presence of a common atrioventricular orifice with cleavage of both the AML and tricuspid septal leaflet, anterior and posterior cusp tissue is common to both ventricles. The common valve leaflets may or may not have septal attachments. The plane of motion of these leaflets is parallel to the IVS. Since the common leaflets are related to both right and left ventricles, they may appear to cross the plane of the IVS during their excursion, depending on the angle from which they are scanned by the ultrasound transducer. In the present studies the transverse view was especially valuable for detecting the common A-V valve and defining its excursion.

The echocardiographic presence of systolic anterior septal motion has been emphasized as a specific finding in patients with right ventricular volume overload. However, this finding is difficult to assess in the presence of both atrial and ventricular left to right shunting as existed in most of the patients in the current study. Motion of the IVS is especially well visualized by the multiple crystal system since the entire septum may be visualized in the sagittal plane. Two patients with incomplete A-V canal and the two patients in our study group in whom an ostium primum atrial defect was an isolated lesion exhibited true paradoxical septal motion. We consider an abnormality of septal motion as a non-specific indicator of the presence of ECD. Enlargement of both the right ventricle and pulmonary artery were similarly regarded as non-specific indicators of the presence of ECD.

It is our view that the multiscan technique when applied to complex cardiac malformations offers many advantages when compared to a single crystal ultrasound method. The major limitation with single crystal techniques relates to recognizing the spatial orientation of the single beam and, therefore, the exact position of the echo-producing structure. Thus, standardization of views for the single crystal analysis of a deformed heart is difficult, and especially great reliance must be placed on individual examiner judgment for accurate diagnosis. The multiscan technique provides real time, two-dimensional, cross-sectional cardiac visualization. Satisfactory data may be obtained even if the small patient being examined cannot remain immobile during the study. Because the visual presentation of moving cardiac anatomy is more complete, the time required to fully describe anatomical alterations is substantially less than with single crystal methods.

We wish to stress that the illustrations that accompany this report are of single frames derived from real time motion studies visualized best on the oscilloscope. Furthermore, they have been subjected to photographic enlargement from a small format (super 8 mm) motion picture film except for figure 2 which is an ECG-gated polaroid print. The use of ECG-gated polaroid exposures postdated the accumulation of data from the group of patients discussed in this report. Even with a polaroid system it should be recognized that still pictures are quite difficult to obtain in children with rapid heart rates, and with slow camera shutter speeds that may avoid phase interference. Clearly, the full advantages of the multiscan system are best appreciated by viewing moving images. The limitations of the system with regards to depth and lateral resolution have been described by Bom et al. All or portions of certain intracardiac structures, e.g., the interatrial septum, may not be visualized because of their orientation parallel to incident sound energy in specific transducer positions. Nevertheless, in our experience the examiner learns to use the dimension of motion along with his a priori knowledge of cardiac anatomy and function to form a
mental image superior to that which can be conveyed in a static picture, i.e., he is able to make an accurate diagnosis of structures that are less well defined in the static image.

Since the major malformation in patients with endocardial cushion defects is a change in the spatial orientation of the orifice of the mitral valve, the cross-sectional view of cardiac anatomy provided in both sagittal and transverse views by the multiple crystal method was found to be a reliable and sensitive indicator of the presence of the congenital cardiac lesion. We would anticipate that this new technique will achieve increasing prominence to assist in the diagnosis and management of patients with endocardial cushion defect.

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Circulation, Volume 50, July 1974
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Circulation. 1974;50:25-32
doi: 10.1161/01.CIR.50.1.25

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

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