Echocardiographic Recognition of the Congenital Bicuspid Aortic Valve

By Navin C. Nanda, M.D., Raymond Gramiak, M.D., James Manning, M.D., Earle B. Mahoney, M.D., Elliot O. Lipchik, M.D., and James A. DeWeese, M.D.

SUMMARY
Twenty-one patients (age four to 25 years) with bicuspid aortic valves proved by surgery and angiography (12) or angiography alone (9) were studied by echocardiography. Seventeen patients had aortic valve disease (11 stenosis, 6 incompetence) and four had coarctation of the aorta with normally functioning aortic valves. A comparison group of 16 patients (9 with aortic valve disease and 7 without aortic valve disease) who had proven tricuspid aortic valve disease was also studied. Echocardiograms in tricuspid aortic valve patients showed the closed position of the cusps near the middle of the aortic lumen. In contrast marked eccentricity of the aortic valve cusp echoes in diastole could be demonstrated in all with bicuspid aortic valves. The Eccentricity Index (1/2 aortic lumen diameter/minimum distance of the diastolic cusp echo from the nearest aortic margin) was low (range 1.0-1.25) with tricuspid aortic valves and high (range 1.5-5.6) with bicuspid aortic valves (P < 0.001). This index was not significantly affected following successful valvotomy. Bicuspid aortic valve cusps showed normal systolic motion pattern resulting in marked asymmetry of leaflet images. About half of patients with bicuspid aortic valves also showed multilayered echoes in diastole in the absence of fluoroscopic evidence of valvular calcification. Echocardiography appears to be specific in the recognition of the bicuspid aortic valve.

Additional Indexing Words:
Ultrasound
Tricuspid aortic valve
Aortic valve stenosis
Coarctation of the aorta
Aortic regurgitation
Eccentricity index

The Bicuspid Aortic Valve is believed to be the most common congenital malformation of the heart or great vessels, its frequency approaching 2% of the general population1 and 85% of patients with coarctation of the aorta.2 Its susceptibility to infective endocarditis has been known for a long time.3, 4 It is also the most common structure of the aortic valve seen in patients with isolated aortic stenosis, with or without regurgitation.1, 5 It would thus be useful if a noninvasive, nontraumatic technique like echocardiography could contribute to the diagnosis of this condition. The purpose of this report is to describe an echocardiographic complex of findings characteristic of the bicuspid aortic valve.

Material and Methods
The clinical material consisted of 21 patients with proven bicuspid aortic valves in whom echocardiographic studies of the aortic root were performed. There were four females, the rest were male. Their ages ranged from four to 25 years, the average being 12 years. Nine patients had pure aortic valve stenosis, two had predominant aortic valve stenosis with minimal insufficiency, six had pure aortic regurgitation, while the remaining four had isolated coarctation of the aorta with no evidence of dysfunction of the aortic valve. In all patients the aortic valve anatomy was defined by aortography with subsequent surgical confirmation in 12 cases. A comparison group of 16 patients with proven tricuspid aortic valves was also studied. There were four females, the rest were male. Their ages varied from two to 25 years, the mean age being 11.7 years. Six patients had pure aortic valve stenosis, one had predominant aortic valve stenosis with minimal incompetence and two had pure aortic incompetence. The remainder were without structural disease of the aortic valve (three had subaortic membranous stenosis, three had isolated coarctation of the aorta and one had idiopathic hypertrophic subaortic stenosis). The tricuspid structure of the aortic valve was demonstrated in all cases by thoracic aortography with subsequent surgical or pathological proof available in thirteen cases. None of the patients had abnormal distortion or position of the aortic root as judged by aortography.

Seven patients with aortic valve stenosis (four with bicuspid aortic valves and three with tricuspid aortic valves) were also studied by echocardiography postoperatively following successful valvotomy with abolition of the gradient across the aortic valve.

Patients with calcific aortic valve disease were excluded.
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from the study. The clinical data in the present study are summarized in table 1.

All ultrasonic examinations were carried out using a commercially available echograph (Picker) and a 2 megaHertz collimated transducer (0.75 in diameter). Continuous records were made on 35 mm film by means of a Fairchild Oscilloscope Record camera and a dual-beam oscilloscope operating as a slave. Aortic root echoes were obtained by a method previously described. The mitral valve was first located from a left parasternal position, utilizing the 3rd or 4th intercostal space. Slight angulation medially and cephalically from the mitral valve position or occasionally placing the transducer one interspace above the mitral valve position and directing it slightly medially outlined the two walls of the aortic root and the valve cusps within. Time varied gain was adjusted in an attempt to equalize the amplitude of the echoes emanating from the anterior and posterior aortic walls. The transducer was angled slightly in various positions and any changes in the aortic valve image produced by this maneuver were recorded.

The positions of the aortic valve cusp echoes within the aortic lumen in systole and diastole were noted. Aortic root echoes were also studied for the presence of multi-layered images in diastole using different instrument sensitivity settings. The internal diameter of the aortic root was measured in millimeters at the onset of diastole. The minimum distance separating the diastolic cusp echo from the nearest aortic margin was also recorded. This measurement was obtained after studying a number of aortic root recordings. From these two measurements an eccentricity index was calculated as follows:

Eccentricity Index (in mm) =

\[
\frac{3}{4} \times \text{Width of aortic lumen at beginning diastole} - \text{Minimum distance of diastolic cusp echo from nearest aortic margin}
\]

Results

All patients with tricuspid aortic valves showed the diastolic position of cusp echoes near the middle of the aortic lumen with the cusps opening briskly to the periphery of the aortic root in systole (fig. 1). Little or no variation in the position of the diastolic cusp echo could be produced with changes in transducer location or beam angulation. The eccentricity index was low, ranging from 1.0 to 1.25. On the other hand, marked eccentricity of the aortic valve cusp echo in diastole with respect to the aortic lumen could be demonstrated in all patients with bicuspid aortic valves. The eccentricity index was high (range 1.5 to 5.6, average 2.43) in every patient in this group (P < 0.001).

In eleven patients with bicuspid aortic valves, the eccentric diastolic echoes within the lumen did not change significantly with transducer angulation. The diastolic position was located close to the posterior aortic wall in six and near the anterior margin in five cases. The cusps appeared markedly asymmetric with one leaflet image appearing much larger than the other (fig. 2). Some patients showed extreme asymmetry of the cusp images. One leaflet could be observed moving from the closed position near one aortic margin in diastole to the open position near the opposite aortic margin in systole practically traversing the whole width of the aortic lumen (fig. 1).

The remaining ten patients with bicuspid aortic valves presented variable positions of diastolic cusp echoes with respect to the aortic lumen. The cusp signal could be seen in the middle of the aortic lumen as well as in close proximity to the anterior and/or posterior aortic wall in the same patient. The same cusp appeared normal, diminutive or abnormally large in a given individual. These differences in the diastolic cusp position occurred in the same or successive cardiac cycles or could be elicited with slight changes in transducer angulation (fig. 3). In all patients the valve cusps could be seen regularly moving to the periphery of the aortic lumen in systole, exhibiting normal opening and closing velocities of motion.

Multiple short linear echoes were observed in diastole in eleven patients with bicuspid aortic valves (six with aortic stenosis, four with aortic regurgitation and one with coarctation of the aorta). Generally these echoes did not extend throughout the period of diastole (fig. 4). Multiple echoes were not noted in the remaining ten patients even with high instrument gain settings (fig. 2). Two patients with tricuspid aortic valve stenosis also showed multiple layered echoes in diastole.

Four patients with bicuspid aortic stenosis who underwent successful valvotomy retained the preoperative characteristic pattern of echoes, with no significant change in the eccentricity index. Echocardiograms obtained following valvotomy in three patients with tricuspid aortic valve stenosis also did not show any significant changes in the diastolic or systolic echo patterns when compared with preoperative studies.

Table 1

Clinical Data

<table>
<thead>
<tr>
<th>Type of aortic valve</th>
<th>Age (years)*</th>
<th>Clinical presentation</th>
<th>Valve anatomy confirmed by</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bicuspid</td>
<td>Mean 12</td>
<td>11 AS, 6 AR 12 angio + surgery</td>
<td>4 CoA 9 angio only</td>
</tr>
<tr>
<td>(21 patients)</td>
<td>Range 4-25</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tricuspid</td>
<td>Mean 11.7</td>
<td>7 AS, 2 AR 13 angio + surgery</td>
<td>7 N 3 angio only</td>
</tr>
<tr>
<td>(16 patients)</td>
<td>Range 2-25</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Abbreviations: AS = aortic valve stenosis; AR = aortic regurgitation; CoA = coarctation of the aorta; N = no aortic valve abnormality; angio = angiography.

*The age of all except one patient in each group was 16 years or less.
Echocardiographic findings in patients with bicuspid aortic valves are summarized in table 2.

Discussion

The role of echocardiography in the diagnosis and evaluation of various forms of cardiovascular disease has been well established. It is a noninvasive, non-ionizing technique which does not pose any risk to the patient. Edler was the first to obtain ultrasonic signals from the aortic valve. Subsequently Gramiak described the technique of studying the aortic root routinely by echocardiography and also obtained validation of its anatomic components using indocyanine green dye during cardiac catheterization.

The echocardiographic technique has proved useful clinically in the evaluation of aortic valve disease as well as in the diagnosis of dissecting aneurysm involving the aortic root.

Normally parallel echoes arise from the aortic walls and between them are observed echoes from the valve cusps. An anterior and a posterior aortic valve leaflet are usually recorded making rapid excursions in systole to positions near the periphery of the aortic root forming a box-like configuration. In diastole, both leaflets make rapid closing movements occupying a mid-aortic position (fig. 1). Correlation with anatomic specimens indicates that the anterior cusp is the right coronary cusp and the posterior aortic leaflet is the non-crownary cusp. The left coronary cusp does not usually produce echoes as its movement is across the ultrasone beam in systole. A linear echo occasionally observed in the middle of the box may arise from the left coronary leaflet. The relationship of the diastolic echoes to the aortic valve margins does not change significantly with beam angulation, aortic root move-
Diastolic echoes in bicuspid aortic valve. On the left is a diagrammatic representation of the ultrasound beam traversing through redundant folds of a bicuspid aortic valve. On the right is an aortic root echogram from a patient with an incompetent bicuspid aortic valve showing multi-layered echoes in diastole, probably originating in the redundant tissue. AO = aortic root; PHONO = phonocardiogram; ECG = electrocardiogram.

Table 2

<table>
<thead>
<tr>
<th>Clinical presentation</th>
<th>Aortic valve in diastole</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Variable eccentricity</td>
</tr>
<tr>
<td>Aortic stenosis</td>
<td>5</td>
</tr>
<tr>
<td>(11 patients)</td>
<td></td>
</tr>
<tr>
<td>Aortic regurgitation</td>
<td>4</td>
</tr>
<tr>
<td>(6 patients)</td>
<td></td>
</tr>
<tr>
<td>Coarctation of aorta</td>
<td>1</td>
</tr>
<tr>
<td>(4 patients)</td>
<td></td>
</tr>
</tbody>
</table>

If the cusps of a bicuspid aortic valve are equal in size, with the line of closure in the middle of the aortic lumen, the echo patterns would be expected to show symmetrically placed leaflets. Ten patients with bicuspid aortic valves in the present study did show symmetric cusp images. However, small changes in transducer angulation were associated with wide variations in the diastolic cusp echo position so that the leaflet images exhibited symmetry, asymmetry as well as reversal of asymmetry in the same subject. Edwards has pointed out that the length of the free margin in a congenital bicuspid aortic valve may equal the straight-line distance between the two lateral attachments of the cusp to the aortic wall. If both the cusps are so configured and are therefore equal in length the valve would not open during ventricular systole. Therefore, one or both of the cusps would have to be longer than the straight-line distance between the points of attachment. This redundancy of leaflet tissue permits systolic opening of the valve but also results in folding and unfolding of the elongated cusp margins in diastole. These folds which result from redundancy could act as echo reflection surfaces and produce the various diastolic positions encountered in our study (fig. 5c). Beat to beat variation in the pattern of cusp folding or minor changes in their orientation to the ultrasonic beam appear to offer a reasonable explanation for the variation in symmetry which we have recorded. A normal tricuspid aortic valve, on the other hand, closes without cusp folding and therefore does not produce multiple diastolic echo sources along the lines of...
closure. Also, the presence of two relatively large cusps in the aortic root would make it more likely for the ultrasonic beam to traverse unequal parts of the cusps resulting in asymmetry of diastolic echoes.

Observation of multiple echoes in diastole have previously been associated with calcific deposits in the aortic valve. Multi-layered diastolic echoes were noted in eleven of our patients with bicuspid aortic valves though none had evidence of valvar calcification either radiologically or subsequently at surgery. Small nodular excrescences present in the edges of thickened or stenosed valves may be the source of multiple echoes in addition to small wrinkles from redundant tissue in the cusps (fig. 5d). Multiple echoes were also seen in two of seven patients with thickened and stenosed tricuspid aortic valves.

The eccentricity index used in the present study denotes the deviation of the diastolic cusp position from the middle of the aortic lumen and takes into account the internal diameter of the aortic root. The index would equal unity if the valve cusps closed exactly in the center of the aortic root, while very high values (3.5 or above) would indicate extreme asymmetry of the cusp images. Although the diagnosis of the bicuspid aortic valve may be made in many instances on the basis of obvious marked eccentricity of the cusps in diastole, use of this index is helpful in cases where the eccentricity is less marked. Use of this index also served to establish the statistical validity of the differences in eccentricity between normals and patients with bicuspid aortic valves.

No differences in echocardiographic findings were observed when patients with apparently normally functioning bicuspid aortic valves were compared with those associated with definite valve malfunction. We have observed that echocardiographic findings in young patients with aortic stenosis are indistinguishable from those with normal valves. Apparent brisk opening and closing movements with normal amplitude of motion have been recorded even in the presence of severe aortic valve stenosis. This probably results from the passage of the ultrasonic beam through the base of a domed valve where the valvar elements are in close proximity to the aortic walls. Our experience with the malfunctioning bicuspid aortic valve has been similar and successful valvotomy with abolition of the gradient across the valve did not change the already normal systolic motion pattern observed preoperatively. In addition the diastolic eccentricity of the echoes was not altered and the multi-layered diastolic images persisted. It would appear, therefore, that fusion of the cusps does not contribute to the echocardiographic findings characteristic of the bicuspid aortic valve.

Deposits of calcium in the aortic valve produce confusing echo patterns in the form of thick multiple complexes within the aortic root, making it difficult to identify the underlying valve anatomy. Therefore patients with aortic valve calcification were excluded from the study.

In the present study, echocardiographic diagnosis of the congenital bicuspid aortic valve was made independently in 12 patients without prior knowledge of the valve anatomy as defined by angiography or later at surgery. In the remaining patients the diagnosis was arrived at retrospectively.

Recognition of the congenitally bicuspid aortic valve is of clinical importance. As many as 25% of patients with bicuspid aortic valves have been known to develop bacterial endocarditis. The bicuspid aortic valve has a special predilection to develop stenosis and/or incompetence and constitutes over 50% of all cases of aortic valve disease. From the surgical viewpoint, it is important in patients undergoing valvotomy not to incise the third rudimentary commissure or raphe present in one of the two leaflets, as severe valvular incompetence may result from improper suspension of the unsupported parts of the leaflet. A bicuspid aortic valve may leak spontaneously as a result of acquired systemic hypertension and an associated weakness of the sinuses of Valsalva may lead to aneurysmal dilatation or rupture. Recognition of the bicuspid aortic valve by a noninvasive method like echocardiography may further delineate its natural history which remains largely unknown. This technique may also be used to confirm or rule out the presence of this abnormality in young subjects with so-called "functional" systolic murmurs as well as in patients with loud aortic ejection clicks who have no other manifestations of aortic valve disease.

Demonstration of marked eccentricity of the aortic valve diastolic echoes with respect to the aortic lumen appears to be the hallmark of a congenital bicuspid aortic valve. An additional frequent finding is the presence of multi-layered echoes in diastole. We have not observed this combination of findings in any other condition and have made no false positive or false negative diagnoses using these criteria. It would appear that echocardiography is of diagnostic value in the recognition of the congenital bicuspid aortic valve.

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