SUMMARY Aortic root echocardiograms were recorded from 89 patients whose aortic valves had also been adequately defined by selective angiography or viewed surgically or at autopsy. The eccentricity index (E.I.) of the aortic leaflets was measured at the onset of diastole and an E.I. of 1.3 or greater was taken as abnormal.

Of 31 patients with isolated nonobstructed or mildly obstructed bicuspid aortic valves (7 viewed previously at valvotomy and 24 diagnosed radiologically) 23 (74%) had an abnormal E.I. Varying eccentricity occurred in some of these patients. Central leaflet echoes (E.I. of 1.0 to 1.25) were present in the other eight patients. All 14 patients with nonobstructed tricuspid aortic valves had central echoes. Additional multilayered diastolic echoes were found in patients with bicuspid aortic valves as well as in two patients with abnormal tricuspid aortic valves.

The valves of 13 patients with aortic stenosis and incompetence were viewed surgically and the E.I. was abnormal in all patients with a bicuspid aortic valve in this group. Aortic leaflet echo findings were not diagnostically helpful in ten patients with tetralogy of Fallot, one of whom had a normal E.I. with a surgically confirmed bicuspid aortic valve. Of 21 patients with VSD only one had a bicuspid aortic valve but six had an abnormal E.I. This false positive sign was related to a high membranous VSD, sometimes with aortic valve prolapse.

It is concluded that an E.I. of ≥1.3 in the absence of an associated VSD is diagnostic of a bicuspid aortic valve and can be expected to be found in approximately three-quarters of subjects with this abnormality.

Materials and Methods

Definition

A bicuspid aortic valve is one which has only two functional cusps. Anatomically there may be two discrete cusps, two cusps with evidence of a small raphe on one, or evidence of fusion of two cusps of an apparently tricuspid valve such that there are only two functional segments.2, 6

Patients

A total of 89 patients whose ages ranged from 3 weeks to 27 years comprises this series (table 1).

GROUP 1

Patients with a substantiated diagnosis of a nonobstructed or minimally obstructed bicuspid aortic valve without associated intracardiac abnormalities were selected from the computerized records of the Cardiology Division of The Hospital for Sick Children, Toronto, and were recalled for echocardiographic assessment. All subjects had undergone cardiac catheterization including selective aortic root angiograms.

A). Those patients with a firm radiological diagnosis of a bicuspid aortic valve were accepted only if the measured gradient of pressure across the aortic valve was less than 25 mm Hg.

B). Patients with a surgically confirmed diagnosis of a bicuspid aortic valve had undergone previous aortic valvotomy. For inclusion, these patients had to have a precise description of the aortic valve made at the time of surgery and a residual gradient across the valve of less than 25 mm Hg on postoperative cardiac catheter study, or no clinical, radiographic or electrocardiographic evidence of re-stenosis.

Groups 2, 3 and 4 of this series consist of patients who un-
derwent cardiac catheterization at this hospital during the period July 1974 to January 1975. All were studied by echocardiography but only those who had adequate aortic root angiograms were reviewed.

GROUP 2

Patients with nonobstructed tricuspid aortic valves. These subjects had no associated intracardiac malformations, a gradient of less than 25 mm Hg and a radiologically tricuspid aortic valve.

GROUP 3

Patients with congenital aortic valve stenosis, or subvalve stenosis or aortic regurgitation. All in this group had subsequent confirmation of the aortic valve appearances at surgery, and thus enabled us to make further visual-echographic correlates.

GROUP 4

Patients with other cardiac anomalies but adequately assessed aortic valves (by radiology and/or surgery). This group consists of children with (A) ventricular septal defects and (B) tetralogy of Fallot.

Radiology

Only patients who had good quality selective aortic root angiograms, including a lateral or left anterior oblique view, were selected. It was essential for the catheter to be placed in the aortic root close enough to the valve to allow good delineation by injected contrast material, but not so close as to distort the valve appearances. The radiological diagnosis of a bicuspid aortic valve was made on the diastolic appearances of the aortic sinuses and the systolic appearances of the valve leaflets similar to the description by Simon and Reis\textsuperscript{5} for stenosed bicuspid valves. We documented diastolic valve appearances as follows: (a) three equal sinuses, (b) three unequal sinuses, (c) two unequal sinuses, and (d) two apparently equal sinuses. In addition, the systolic appearances of the leaflets, and evidence of complete cusp fusion, were recorded. Normal tricuspid aortic valves had sinuses classified as (a) or (b) and three leaflets. Bicuspid aortic valves had sinuses classified as (b), (c) or (d) and the systolic appearances of movement of only two cusps, one of which might be larger than the other. Since there is overlap of the diastolic appearances of the classification (b) to both groups, the systolic appearances are important (figs. 1, 2 and 3).

Echocardiography

A commercially available Ekoline 20 ultrasonoscope interfaced to a Cambridge strip chart recorder was used with a variety of transducers varying from 2.25 MHz \(\frac{1}{2}\) inch diameter focused at 5 centimeters to 5 MHz \(\frac{1}{4}\) inch non-focused. Echocardiograms of the aortic valve were recorded by first identifying the anterior leaflet of the mitral valve and then rotating the transducer superiorly and medially until the aortic root was identified. Transducer position was then varied until the opening and particularly the closing movements of the aortic leaflets were recorded. The instrument sensitivity was adjusted to produce clear leaflet echoes. This maneuver was repeated several times using different

<table>
<thead>
<tr>
<th>Table 1. Patient Groups</th>
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<tbody>
<tr>
<td><strong>Number</strong></td>
</tr>
<tr>
<td><strong>Group</strong></td>
</tr>
<tr>
<td>1. Nonobstructed bicuspid aortic valves</td>
</tr>
<tr>
<td>A. Radiologically diagnosed</td>
</tr>
<tr>
<td>(i) with otherwise normal catheter study 10</td>
</tr>
<tr>
<td>(ii) with coarctation 6</td>
</tr>
<tr>
<td>(iii) with mild stenosis or incompetence 8</td>
</tr>
<tr>
<td>B. Surgically diagnosed at aortic valvotomy 7</td>
</tr>
<tr>
<td>2. Nonobstructed tricuspid aortic valves</td>
</tr>
<tr>
<td>Radiologically diagnosed 14</td>
</tr>
<tr>
<td>(i) with normal catheter study 6</td>
</tr>
<tr>
<td>(ii) with coarctation 5</td>
</tr>
<tr>
<td>(iii) with mild stenosis 2</td>
</tr>
<tr>
<td>(iv) with endocardial fibroelastosis 1</td>
</tr>
<tr>
<td>3. Aortic valve disease viewed at surgery 13</td>
</tr>
<tr>
<td>(i) bicuspid aortic valve 7</td>
</tr>
<tr>
<td>(ii) tricuspid aortic valve 6</td>
</tr>
<tr>
<td>4. Other intracardiac lesions 31</td>
</tr>
<tr>
<td>A. Ventricular septal defects 21</td>
</tr>
<tr>
<td>B. Tetralogy of Fallot 10</td>
</tr>
<tr>
<td>Total patients assessed 89</td>
</tr>
</tbody>
</table>

**FIGURE 1** Angiograms of a normal tricuspid aortic valve in the lateral projection (left) in diastole showing three sinuses of apparently slightly unequal size. (The noncoronary sinus is the lowest in this projection.) Right) In systole showing three cusps opening.

**FIGURE 2** Angiograms of a bicuspid aortic valve (left) in diastole showing three unequal sinuses and (right) in systole showing only two cusps. The right and left cusps are fused. The aortic root is dilated.
transducer angulations. Measurements of the "eccentricity index" (E.I.) of the closed aortic valve leaflets were made specifically at the onset of diastole (see discussion). The E.I. was calculated as follows:

\[
E.I. = \frac{1}{2} \text{ width of aortic lumen}{\text{minimum distance of the cusp echo to nearest aortic margin}}
\]

The internal diameter of the aortic root was used and both measurements were taken in millimeters at the onset of diastole (fig. 4). In each patient at least three measurements were taken. Where wide variation was noted the maximum E.I. was recorded and used in the assessment. Those with "central echoes" had an E.I. of 1.0 to 1.25 maximum. An E.I. of 1.3 or greater was taken as abnormal (fig. 5). This contrasts with a previously published series\(^{14}\) in which an undefined zone occurred between tricuspid aortic valves with an E.I. of 1.0 to 1.25 and bicuspid aortic valves with an E.I. of 1.5 to 5.6. The presence of additional multilayered echoes within the aortic root was also recorded. All the echocardiograms were reported routinely, but reviewed again by one person for consistency. Only those in which the diastolic closure of the cusps and anterior and posterior aortic walls were clearly recorded were used.

Surgery

We attempted to obtain as much correlation of echo findings with visualized anatomy as possible. Groups 3 and 4 were therefore included in the study for this purpose. The description of the aortic valve was obtained from the surgeon or from the surgical records, which usually included a drawing of the aortic valve if it was abnormal.

Results

Groups 1 and 2

The aortic root echo findings for the patients with nonobstructed aortic valves are shown in table 2. Eccentric aortic valve leaflets were recorded in 18 of 24 patients in group 1A considered to have a bicuspid aortic valve on radiological assessment (75%) and in five of seven patients in group 1B proven to have a bicuspid aortic valve at surgery (71%) thus giving an abnormal E.I. in 74% of patients with a nonobstructed bicuspid aortic valve.

In group 1A the range of values for abnormal E.I. in the 18 patients was from 1.3 to 2.7. The average value was 1.7. The six patients with "central echoes" had an average value of 1.1 and range 1.0 to 1.2. In group 1B, five patients had an
E.I. ranging from 1.4 to 2.3 with an average of 1.8. The maximum values for E.I. recorded in the other two with surgically proven bicuspid valves were 1.1 and 1.2, respectively. Figure 6 shows the echocardiogram of one of these latter patients with central leaflet echoes. No significant eccentricity was found on any view of the aortic root with altered transducer positions. Also, the echo recordings of the eight patients with normal E.I. (six from group IA and two from 1B) were reviewed to see if there was a change in leaflet position at any point in diastole. None showed changing of position or more marked eccentricity.

All 14 patients with a tricuspid aortic valve on radiological assessment (group 2) had a normal E.I. (1.0 to 1.25). One patient in this group had endocardial fibroelastosis and died. Postmortem confirmation of a tricuspid aortic valve was obtained.

Additional multilayered echoes were found in each group including two patients with tricuspid aortic valve. One of these patients had mild aortic stenosis with a measured gradient at catheterization of 21 mm Hg. The other patient died following rupture of an infected anastomotic site after coarctectomy. His aortic valve was shown at autopsy to be tricuspid but with thickened and ridged leaflets (fig. 7).

Group 3

The surgically confirmed diagnoses together with the echocardiographic and clinical findings of the patients with significant aortic valve disease are in table 3. It will be noted that all patients with a bicuspid aortic valve had an eccentricity index of greater than 1.3 and all with a tricuspid aortic valve had an index of less than 1.3.

Group 4A

Of 21 patients with ventricular septal defects (VSD) who had satisfactory angiography of the aortic root, six (29%) had eccentric leaflets (E.I. ≥ 1.3) by echocardiography. Of these patients, one who had a muscular VSD was considered to have a bicuspid aortic valve radio-

Table 2. Aortic Root Echo Findings in Patients with Nonobstructed or Minimally Obstructed Aortic Valves

<table>
<thead>
<tr>
<th>Echo features</th>
<th>Number of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Eccentricity index ≥ 1.3</td>
<td>Group 1A</td>
</tr>
<tr>
<td>(Additional multilayered echoes)</td>
<td>(3)</td>
</tr>
<tr>
<td>Central leaflets</td>
<td>(Index 1.0 - 1.25)</td>
</tr>
<tr>
<td>(Additional multilayered echoes)</td>
<td>(1)</td>
</tr>
<tr>
<td>Total patients in group</td>
<td>24</td>
</tr>
</tbody>
</table>

FIGURE 6 The aortic root echocardiogram of a patient whose aortic valve was viewed at surgery and observed to be bicuspid with right and left cusps. The echo of the aortic leaflets is central.

Table 3. Findings in Patients in Group 3 (With Aortic Valve Disease Viewed Surgically)

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Number of patients</th>
<th>Number with eccentricity index ≥ 1.3</th>
<th>Number with central index</th>
<th>Number with multilayered echoes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Valvular aortic stenosis with bicuspid aortic valve</td>
<td>6</td>
<td>6</td>
<td>0</td>
<td>3</td>
</tr>
<tr>
<td>Gross aortic incompetence with bicuspid valve</td>
<td>1</td>
<td>1</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Valvular aortic stenosis with tricuspid aortic valve</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Aortic incompetence with tricuspid aortic valve</td>
<td>1</td>
<td>0</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Subvalvular and valvular aortic stenosis with tricuspid aortic valve</td>
<td>4</td>
<td>0</td>
<td>4</td>
<td>0</td>
</tr>
</tbody>
</table>
right aortic cusp and mild aortic regurgitation. His E.I. was also 1.2. One patient with a central echo and a tricuspid aortic valve by angiography had autopsy confirmation of a normal tricuspid aortic valve.

Discussion

Normal aortic valves are tricuspid and have equal-sized cusps with commissures situated equally at one third of the circumference of the aortic ring, resulting in three sinuses of equal size. Variations occur in which there are three sinuses of unequal size and three unequal cusps without fusion. Bicuspid aortic valves may have three unequal sinuses and evidence of fusion of two cusps with a wide raphe visible, or there may be only two sinuses, two commissures and a small raphe. Other variations of the anatomy of aortic valves are rarer and include unicommisural unicusp valves, acommissural valves, and quadricuspid valves. It is important to be aware of these anatomical variations when endeavoring to study the aortic valve by angiography or echocardiography.

The angiographic diagnosis of a bicuspid aortic valve cannot be considered totally reliable, because there are variations in appearances. For this reason we have attempted to obtain visual confirmation of the echo and angiographic appearances whenever possible to minimize the possibility of error when one image technique is compared with another. For greater reliability a small group of seven patients whose aortic valves were seen at surgery and noted to be bicuspid, and who currently have no evidence of re-stenosis, have been included. In addition, the valves of all patients with significant aortic valve lesions have been visualized at surgery, and a small number of patients in each of the other groups have had follow-up of the radiological and echocardiographic assessments at surgery or autopsy.

Abnormal eccentricity of the aortic leaflets on echocardiography has been found in 74% of our patients who have nonobstructed bicuspid aortic valves, and in no patient with a tricuspid aortic valve, except where there is an associated high membranous VSD usually with aortic cusp prolapse. It would therefore appear that an eccentricity index of ≥1.3 is diagnostic of a bicuspid aortic valve if there is no associated VSD. However, the reverse is not true in that central aortic leaflet echoes have occurred in both bicuspid and tricuspid aortic valves. This fact has been confirmed by visualization of the valves at surgery.

Multilayered echoes in the aortic root, predominantly in diastole, have been observed in our study and described by others previously. We observed this sign with the instrument sensitivity set to give clear aortic cusp echoes and found that only additional nonspecific ultrasonic "noise" was produced by further increase in gains. This sign of diastolic multilayering has not been specific for bicuspid aortic valves in our series, but has indicated that the aortic valve is abnormal, as confirmed at autopsy in one case.

The main exception to the rule that an eccentricity index of ≥1.3 indicates a bicuspid aortic valve has been when there is an associated high membranous VSD. However, this false positive sign of bicuspid aortic valve is not present in every case of aortic valve prolapse through a VSD, so that it cannot in any sense be regarded as diagnostic of that clinical situation.

For assessment of E.I., we have obtained adequate recordings of the closing movements of the aortic leaflets and then made the measurements specifically at the onset of diastole at a precise and constant point. This varies from the method of Nanda et al., which we found to give false positive diagnoses in a few instances. When the closing movements of the leaflets are shown to merge into a line in diastole, it ensures that a true echo of the closed leaflets is recorded. Otherwise, eccentric linear echoes that really originate from the aortic ring, wall or sinuses may be falsely interpreted to be coming from the aortic valve leaflets. We made our recordings from several transducer positions and when variations occurred, documented the maximum E.I. In no case were the leaflets central at the onset of diastole and
eccentric in late or end-diastole. Since the undefined values of E.I. of 1.25 to 1.5 in a previous paper are an important zone, we made a cut-off value of 1.3 for abnormal eccentricity. From the observed values, a lower figure would not include all with a bicuspid valve, so some false negative cases appear inevitable. It has been possible to obtain suitable recordings in almost all patients.

We have encountered beat to beat variation in eccentricity at the onset of diastole, recorded from a constant transducer position. These variations occurred at a more rapid rate than respiration. Thus, they may be related to the forward systolic movement of the aortic root during the cardiac cycle resulting in the transducer beam transecting the root at different angles during this motion. Nanda et al. observed variations of leaflet position throughout one diastole and related this finding, and also the appearance of multiple diastolic echoes, to redundant folds of leaflet tissue.

The study of Glancy et al. indicated that abnormal aortic valves occur more frequently with tetralogy of Fallot than in the general population. We have not found echocardiography to be useful diagnostically in our small group of patients with tetralogy of Fallot whose aortic cusps were adequately recorded. E.I.s of 1.2 occurred in tetralogy patients with normal tricuspid aortic valves, and also in a proven bicuspid aortic valve and in a known case of aortic valve cusp prolapse through the VSD. The difficulties with echocardiographic diagnosis in this group may be related to the fact that the aortic root is large and anteriorly situated and there is an associated high VSD.

In conclusion, echocardiography of the aortic root is useful in making the diagnosis of a bicuspid aortic valve when the E.I. is ≥1.3, provided that there is no associated high membranous VSD. However, a normal E.I. does not exclude the presence of a bicuspid valve since central aortic leaflet echoes occur in approximately 25% of cases. Normal tricuspid aortic valves without associated VSD all have a central E.I. Multilayered echoes in diastole occur with both bicuspid and tricuspid valves, but this sign indicates that the valve is abnormal. Echocardiographic analysis of the aortic root is an important screening procedure in the detection of bicuspid aortic valves.

Acknowledgment

We thank Miss Connie Williams for technical assistance and Mrs. Jenny Visanji for secretarial work.

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Circulation. 1976;53:80-85
doi: 10.1161/01.CIR.53.1.80

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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