SUMMARY Echocardiograms were performed in thirty-six patients (aged 4 to 36 years) with proven coarctation of the aorta. Nineteen patients (53%) were found to have multilayered diastolic eccentricities of their aortic valves (Eccentricity Index >1.5), indicating the presence of bicuspid aortic valves. One of these patients also had multiflared aortic root echoes in diastole. Five patients had angiographic proof of their aortic valve morphologies which corroborated the echo findings. Five patients with bicuspid aortic valves showed mitral valve diastolic flutter indicative of aortic regurgitation. Idiopathic hypertrophic subaortic stenosis (IHSS) was suspected in four patients (11%) with abnormal systolic anterior motion of the mitral valve; three of these patients also had asymmetric septal hypertrophy. There was catheterization proof of IHSS in one patient. Two patients (5.6%) demonstrated mitral valve prolapse.

Materials and Methods

Thirty-six patients with coarctation of the aorta were studied by echocardiography. They represent the total number of patients seen in our laboratory with aortic coarctation and echocardiographic examinations. Fourteen patients had preoperative studies only and fifteen had only postoperative echocardiograms. The remaining seven patients had both pre and postoperative echo evaluations. The patients ranged in age from 4 to 36 years; there were 21 males and 15 females.

All echocardiograms were obtained using a commercially available ultrasonoscope (Picker Model 103) and a 2.0 MHz transducer. A Tektronics 565 dual beam oscilloscope was operated as a slave and displayed the ultrasonic data in a B mode on the upper beam. The lower beam was used in multi-trace operation to record the electrocardiogram. Continuous recordings were made on 35 mm film using an oscilloscope record camera. Film transport speed was selected to equal 125 mm/sec as obtained by ordinary strip chart recorders.

Mitrail valve echograms were obtained by placing the transducer in the third or fourth left intercostal space and directing the beam posteriorly and slightly medially. Aortic root echoes were obtained by medial and cephalic rotation of the transducer from the mitral valve position. Echoes from the left ventricular cavity, ventricular septum, and the posterior wall were all obtained by shifting the ultrasonic beam inferiorly and laterally so that it passed just below the mitral valve.

The diagnosis of a bicuspid aortic valve was made using the eccentricity index of the aortic valve cusps in diastole. Our laboratory has previously reported the computation of this index as follows: one-half the width of the aortic lumen at the beginning of diastole (L/2) is divided by the minimum distance between the diastolic cusp position and the closest aortic margin (d) (fig. 1). An eccentricity index of 1.5 or more was considered diagnostic of a bicuspid aortic valve. Multilayered echoes in the aortic root, a feature commonly present with bicuspid aortic valves, were also noted. Multilayering is probably due to valve thickening or redundant folds on the valve cusps in the absence of valvular calcification.

Aortic insufficiency was diagnosed echocardiographically by the presence of typical fine fluttering motion of the mitral valve leaflets in diastole. The presence of abnormal systolic anterior movements (SAMs) of the anterior mitral valve leaflet and asymmetric septal hypertrophy (ASH) of the type seen in idiopathic
hypertrophic subaortic stenosis (IHSS) was noted.\(^7\) Amyl nitrite was administered to patients to determine its effect on the SAMs. When a SAM, at its most anterior displacement, occupied more than 50% of the left ventricular outflow space, it was Defined as being large. Conversely, a small SAM occupied less than 50% of the left ventricular outflow space. Asymmetric septal hypertrophy was diagnosed if the width of the interventricular septum was at least 1.5 cm and at least 1.5 times as great as the posterior wall thickness measured at the same level.\(^8\) The thicknesses of the interventricular septum and posterior left ventricular wall were measured just below the tips of the mitral valve leaflets at the peak of the P wave on the electrocardiogram. The left ventricular outflow tract width was defined as the minimum distance between the left endocardial surface of the ventricular septum and the closure point (C) of the mitral valve. In children, the left ventricular outflow tract was considered small if there was prolonged apposition of the mitral valve and interventricular septum in diastole.\(^9\)

Mitrail valve prolapse was diagnosed echocardiographically if there was typical mid to late posterior displacement of the mitral valve in systole.\(^10\)

**Results**

Nineteen of thirty-six patients included in this study (53%) were found to have significant diastolic eccentricities of their aortic valves typical of bicuspid aortic valves. Only one of these 19 patients exhibited multilayered echoes in diastole. Examples of tricuspid and bicuspid aortic valves are shown in figure 2. A total of five patients had angiographic studies where the aortic valve morphology could be determined (4 bicuspid, 1 tricuspid). The echoes and angiograms were evaluated independently, but not in a double-blind fashion. In each instance, the angiographic determination agreed with the echocardiographic diagnosis. A bicuspid aortic valve was discovered at the only autopsy performed in this series, confirming both angiographic and echocardiographic findings.

The echocardiographic diagnosis of aortic insufficiency was made in five patients, an incidence of 14%. All of these patients demonstrated high frequency fluttering of their mitral valve leaflets in diastole (fig. 3); each had a bicuspid aortic valve demonstrated echocardiographically.

Four patients (11%), two with bicuspid and two with tricuspid aortic valves, exhibited abnormal systolic anterior movements of their anterior mitral valve leaflets (SAMs) of the type seen in IHSS. Idiopathic hypertrophic subaortic stenosis was not clinically suspected in any instance. Two patients showed large SAMs at rest while the remaining two had small SAMs which increased in amplitude following amyl nitrite inhalation (fig. 4). Subsequent to surgical repairs of their coarctations, all showed large SAMs at rest. None of the patients in the study group demonstrated apposition of the interventricular septum and SAM. Three patients with SAMs also had asymmetric ventricular septal hypertrophy (ASH) (fig. 5). Catheterization of the left ventricle and the outflow area was performed in one patient with large SAMs and ASH. No gradient was observed at rest. However, following amyl nitrite inhalation a gradient of 50 mm Hg developed across the left ventricular outflow tract, confirming the diagnosis of IHSS. Similar hemodynamic studies were not performed in the remaining three patients. Clinically, two of the three patients who had SAMs and systemic hypertension preoperatively showed persistent elevations of blood pressure postoperatively. The third patient, while becoming normotensive following surgery, clinically retained some obstruction at the coarctation site as
FIGURE 3. Incompetent bicuspid aortic valve. Diastolic fluttering of the mitral valve (MV) leaflets, characteristic of aortic regurgitation is shown. ECG = electrocardiogram.

evidenced by a continued significant disparity between the upper and lower extremity systolic blood pressures.

Two patients of the total series (5.6%) had mitral valve prolapse. One of them also had aortic insufficiency and a bicuspid aortic valve.

Discussion

Previous reports describing the incidence of various anomalies associated with coarctation of the aorta have necessarily reflected skewed populations because they have dealt primarily with autopsy findings.1-4 Where applicable, echocardiography as a noninvasive tool may accurately reflect the true frequency of certain anomalies associated with coarctation of the aorta.

Anatomic studies have found the incidence of bicuspid aortic valves in patients with aortic coarctation to be between 27 and 85%.5-4 Our echocardiographic study demonstrated an incidence of 53%. In our laboratory the echo diagnosis of bicuspid aortic valves has been shown to be very reliable when compared to the morphologic diagnosis obtained at angiography, surgery, or autopsy.4 Unfortunately, in the present study, the echo diagnosis could be confirmed in only a few instances by angiography or at autopsy. In most instances, this reflects the fact that aortic valve abnormalities were not suspected clinically. In those patients who did have angiographic studies, the absence of aortic valve obstruction and eccentric doming made the radiologic diagnosis of a bicuspid aortic valve difficult. Finally, many views of the aortic root are often required before a firm radiographic interpretation can be made concerning the morphology of a nonstenotic aortic valve; and this also adds to the risk of the procedure.12 In our series many of the X-ray views were taken in only one or two planes. However, it is significant that where a positive radiologic diagnosis of a bicuspid aortic valve was made, it agreed with the echocardiographic findings. A bicuspid aortic valve was found also in the only autopsy performed in this series, confirming the echocardiographic and angiographic diagnoses. Why multilayering did not occur more often in our patients with bicuspid aortic valves is not clear. It may reflect the non-

FIGURE 4. Systolic abnormalities of the mitral valve (MV). A small, resting systolic anterior movement of the mitral valve (MV) (upper panel) becomes large and very prominent following the administration of amyl nitrite (lower panel). PHO = phonocardiogram; ECG = electrocardiogram; VS = interventricular septum.

FIGURE 5. Asymmetric septal hypertrophy in coarctation. PHO = phonocardiogram; ECG = electrocardiogram; VS = interventricular septum; PW = posterior wall.
obstructive nature of the valvular lesions, since multilayering is observed with aortic valve thickening.

The finding of mitral valve flutter in diastole pointed to the diagnosis of aortic insufficiency in 14% of the patients studied. Not surprisingly, all of these patients had bicuspid aortic valves.

Idiopathic hypertrophic subaortic stenosis has been reported in patients with coarctation of the aorta. The presence of large SAMs of the mitral valve in four patients together with definite evidence of ASH in three of them strongly suggested this diagnosis. The one patient who underwent a left ventricular catheterization study had a significant gradient across the left ventricular outflow tract; this was elicited following provocative testing with amyl nitrite. This patient also demonstrated ASH on his echocardiogram. The remaining three patients did not have catheterization studies for IHSS as it was not clinically suspected. However, their echocardiograms were very similar to that of the patient with catheterization proof of IHSS.

None of the SAMs appeared to touch the interventricular septum, implying that the left ventricular outflow tract was not significantly narrowed. This may be due in part to some dilatation of the left ventricular cavity resulting from the effects of longstanding coarctation and/or systemic hypertension. Even postoperatively, three of four patients with SAMs retained residual obstruction at the coarctation level and/or systemic hypertension. All the patients with SAMs were in the pediatric age group, and the findings presented in this study may simply be a harbinger of the more typical IHSS syndrome that they might develop later in life. Nevertheless, it is important to consider the possibility of coexisting IHSS when contemplating repair of aortic coarctation. Significant left ventricular outflow tract obstruction could conceivably occur following the correction of the coarctation itself as the distal fixed obstruction tends to mask the proximal functional obstruction. Administration of certain inotropic drugs in the operative or postoperative periods could also create a significant gradient across the left ventricular outflow tract and may pose a serious clinical problem.

Finally, only 2 or 5.6% of our patients demonstrated mitral valve prolapse; one of them had a bicuspid aortic valve which was incompetent. Mitral valve prolapse is a common finding; some estimates claim it occurs in 8% of the general population. Thus, it appears from our study, that mitral valve prolapse does not occur any more frequently in patients with coarctation of the aorta than it does in the general population.

In summary, the bicuspid aortic valve appears to be the most common echocardiographic abnormality present in coarctation of the aorta. In addition, some patients with coarctation may have associated IHSS.

References

Echocardiographic studies of abnormalities associated with coarctation of the aorta.
J A Scovil, N C Nanda, C M Gross, A C Lombardi, R Gramiak, E O Lipchick and J A Manning

_Circulation._ 1976;53:953-956
doi: 10.1161/01.CIR.53.6.953

_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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