Echocardiography of the “Floppy” Aortic Valve

Report of a Case

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SUMMARY
The echocardiographic features of a patient with severe aortic regurgitation due to a “floppy” aortic valve are presented. The salient abnormality observed was marked fluttering of the aortic valve cusps in diastole. Fluttering of the anterior leaflet of the mitral valve and left ventricular enlargement were noted. The diagnosis of a “floppy” aortic valve was substantiated at surgery.

THE ROLE OF ECHOCARDIOGRAPHY in the assessment of various forms of heart disease has been firmly established. The echocardiographic features of the normal and diseased aortic valve were described by Gamiak and Shah. Abnormalities of aortic valve motion have also been noted in patients with idiopathic hypertrophic subaortic stenosis, discrete membranous subaortic stenosis and right coronary sinus of Valsalva fistula.

Diagnostic ultrasound has also proved to be a reliable tool in the evaluation of “floppy” mitral valve. More recently, the echocardiographic detection of tricuspid valve prolapse was described. Although the macroscopic and microscopic appearances of the “floppy” aortic valve have been described, its echocardiographic features have not been previously reported. The ultrasound findings in a patient with severe aortic regurgitation due to a “floppy” aortic valve are outlined in this report.

Case Report
A 56-year-old white male was admitted to another hospital with severe aortic regurgitation and congestive cardiac failure. He was noted to have a cardiac murmur for the first time in September 1974. He was treated with digoxin and diuretics and transferred to Mount Sinai Medical Center for further cardiac evaluation. On examination, his pulse was 80/minute and regular and his blood pressure was 140/50. He did not have any external stigmata of Marfan’s syndrome. A diastolic thrill was palpable at the left sternal border. A grade II/VI ejection systolic murmur and a grade V/VI musical low pitched holodiastolic murmur were audible at the left sternal edge. The ECG showed left ventricular hypertrophy. Cardiac catheterization revealed a left ventricular end-diastolic pressure of 18 mm Hg, and angiography revealed grade 4/4 aortic regurgitation and left ventricular enlargement. There was no calcification of the aortic valve. The ultrasound examination was performed with the patient supine. A 2.25 MHz, 0.5 inch, 7.5 cm focused transducer, an Ekoline 20 ultrasonoscope and an Electronics for Medicine DR8 recorder were used. The phonocardiogram was recorded from the second left interspace at the left sternal border, using a crystal microphone. Recordings were made using the logarithmic and low frequency settings.

Aortic valve replacement was performed with a #27 porcine heterograft. At operation, the aortic valve leaflets were grossly redundant, wrinkled and “floppy.” The leaflets prolapsed into the left ventricular outflow tract in diastole. The noncoronary cusp was the worst affected. The histology of the valve (fig. 1) showed many fibroblasts proliferating on both the ventricular and aortic margins of the valve. In these areas, the valve material was loose and fibrillar. There was an increase in dense, relatively acellular collagen throughout the remainder of the leaflets. Great increase in acid mucopolysaccharides as demonstrated by Alcin blue staining was noted in all areas of the leaflets.

Phonocardiographic and Echocardiographic Findings
Figure 2 shows the preoperative phonocardiogram. A low pitched holodiastolic murmur is seen. An echocardiogram of the aortic root is illustrated in figure 3. There is marked fluttering of the aortic valve leaflets in diastole. There are two discrete echoes in
Figure 1

This shows the histologic appearance of the noncoronary cusp of the aortic valve. There is a striking increase in acid mucopolysaccharides (dark areas) and an increase in dense, relatively acellular collagen. (Alcian blue stain × 100.)

Figure 2

This phonocardiogram demonstrates a loud, low frequency holodiastolic murmur. $S_1$ = first heart sound, $S_2$ = second heart sound, Log = logarithmic setting, 2nd LICS = second left interspace.
Diastole. Slight fluttering of the valve is also present in systole. Early systolic partial closure of the right coronary cusp of the aortic valve with reopening shortly thereafter is also seen. The anterior leaflet of the mitral valve exhibited marked fluttering in diastole (fig. 4). This was suggestive of aortic regurgitation. The left ventricular end-diastolic dimension (measured at the peak of R wave) was increased (6.4 cm). The end-systolic dimension (smallest dimension between septal and posterior left ventricular endocardium) was 3.8 cm (fig. 4).

Discussion

The echocardiographic features of the normal and diseased aortic valve have been studied extensively. Abnormalities of aortic valve motion may also occur in conditions in which there is no primary pathology of the valve. Characteristic echocardiographic abnormalities have been described in patients with idiopathic hypertrophic subaortic stenosis (IHSS), discrete membranous subaortic stenosis and right coronary sinus of Valsalva fistula. 7, 8, 15

The clinical and echocardiographic features of mitral valve prolapse have been the subject of many reports. 9-12, 18 Patients with the "floppy" mitral valve syndrome may have associated "floppy" aortic valves. 14 Subjects with myxomatous degeneration of the mitral, aortic or pulmonic valve may have Erdheim's necrosis of the aorta and femoral artery. 14 Read and Thal's description of the floppy aortic valve was very similar to that observed by us. 14 They stressed the frequency of dehiscence of the valve ring after valve replacement. The echocardiographic features of the "floppy" aortic valve have not been previously described.

The salient echocardiographic abnormalities noted in our patient were marked fluttering of the valve cusps in diastole and diastolic separation of the cusp echoes. A calcified aortic valve may give rise to multiple echoes in diastole and produce a spurious appearance of cusp separation. However, no aortic valve calcification was noted in our patient. Thus, the two discrete echoes seen in diastole probably represent separation of the right and the noncoronary cusp of the aortic valve. This echocardiographic finding was confirmed at surgery. However, it is possible that a large redundant cusp may give rise to multiple diastolic echoes. Prominent diastolic aortic cusp echoes may also be seen in endocarditis of the aortic valve. 17, 18 However, diastolic fluttering of the aortic valve has not been described in aortic valve endocarditis. Fluttering of the aortic valve cusps in systole may be observed in patients with myocardial dysfunction from coronary artery disease, IHSS, mitral regurgitation, ventricular septal defect or discrete membranous subaortic stenosis. 8, 15 Diastolic fluttering of the aortic valve cusps, on the other hand, suggests
lack of support of the cusps. Thus this finding may be suggestive of the floppy aortic valve although it is conceivable that such an abnormality may also occur in a patient with a torn aortic cusp. In the latter case, one would expect the fluttering to be limited to the cusp involved.

References

Circulation, Volume 52, November 1975

Figure 4
The left panel shows the left ventricle at the level of the chordae tendineae (c). The left ventricular end-diastolic dimension is increased with good motion of the interventricular septum (IVS) and the posterior wall of the left ventricle (LVPW). Fluttering of the anterior mitral leaflet (F) is seen in the last beat. The right panel shows anterior mitral leaflet (AMV) flutter (F) more clearly. PMV = posterior mitral leaflet.
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Circulation. 1975;52:959-962
doi: 10.1161/01.CIR.52.5.959

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

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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