Br Heart J 1985; 54: 179–83

Prevalence and clinical significance of aortic valve prolapse

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SUMMARY The prevalence and clinical significance of aortic valve prolapse were determined prospectively in 2000 consecutive patients undergoing routine clinical cross sectional echocardiography. Two hundred and twelve patients were excluded because the aortic cusps were not adequately visualised. Aortic valve prolapse was defined as downward displacement of cuspal material below a line joining the points of attachment of the aortic valve leaflets. Twenty four cases of aortic valve prolapse (1·2%) were identified. The patients were aged 12–64 years and nine were women. All had underlying valvar heart disease and the commonest lesion (in 11 cases) was prolapse of the larger cusp in bicuspid valves. Aortic valve prolapse was seen in four patients with mitral valve prolapse (two with severe regurgitation), one of whom had marfanoid aortic root dilatation. The remaining examples of aortic prolapse were seen in patients with various disorders including one with pulmonary atresia, two with aortic root disease (one with dissection and one with idiopathic dilatation), and one case of severe mitral regurgitation. Valves destroyed by infective endocarditis were seen in two cases. Aortic valve prolapse may be detected in various cardiac disorders and does not imply the presence of aortic regurgitation, but when bicuspid aortic valves are present it may well be important in producing such regurgitation. Although aortic valve prolapse may be associated with severe forms of mitral valve prolapse, these patients rarely have aortic regurgitation.

In patients with aortic regurgitation and a normal or dilated aortic root the M mode or cross sectional echocardiographic appearances of the aortic valve may be surprisingly normal. Often the only apparent abnormality is thickening of the valve or restricted cusp motion.[1] Aortic regurgitation in a dilated aorta is thought to be the result of malalignment and inadequacy of the cusps, but the valve may be competent and appear to be normal despite a grossly dilated aortic root.[2] The mechanism of aortic regurgitation in the absence of root dilatation is also often unclear, especially in bicuspid aortic valves, but anatomical studies suggest that such valves are usually regurgitant.[3] Aortic valve prolapse has been reported in surgical and necropsy studies[2 4] and may also be shown by echocardiography in patients with mitral valve prolapse.[5 6] The thin aortic valve cusps are less echo reflective than those of the more voluminous mitral valve, and therefore it may be more difficult to identify aortic valve prolapse. With the progressive improvement in image resolution by cross sectional echocardiography such features may be more readily seen.

The purpose of this study was to determine the prevalence of aortic valve prolapse in patients undergoing routine cross sectional echocardiography and to determine the relation of aortic valve prolapse to mitral valve prolapse and aortic regurgitation.

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Accepted for publication 7 May 1985

Patients and methods

Two thousand consecutive patients aged 9 to 83 years with all forms of heart disease who underwent routine M mode and cross sectional echocardiography were included. Patients with prosthetic aortic valves were excluded. The presence of an aortic early diastolic murmur was noted and graded out of four. The clinical details of patients were extracted from the case notes.

ECHOCARDIOGRAPHIC TECHNIQUE

Cross sectional echocardiograms were performed on

Hewlett-Packard phased array apparatus (model 77020A) with a 3·5 or 5 MHz transducer. Complete echocardiographic studies in the parasternal, subxiphoid, suprasternal, and apical views were routinely obtained and recorded on half inch (13 mm) video cassette tapes. To minimise difficulties with transducer angle and positioning we analysed only images obtained with the patient in the semi-left lateral position and taken in the long axis parasternal view. We measured left ventricular cavity size in systole and diastole (minimum and maximum cavity size respectively) and aortic root dimensions at end systole.

DEFINITIONS

Aortic valve prolapse was diagnosed when either or both of the right or non-coronary aortic valve cusps (seen in the cross sectional echocardiographic long axis view) showed backward bowing towards the left ventricle beyond a line joining the points of attachment of the aortic valve leaflets to the annulus (Fig. 1).

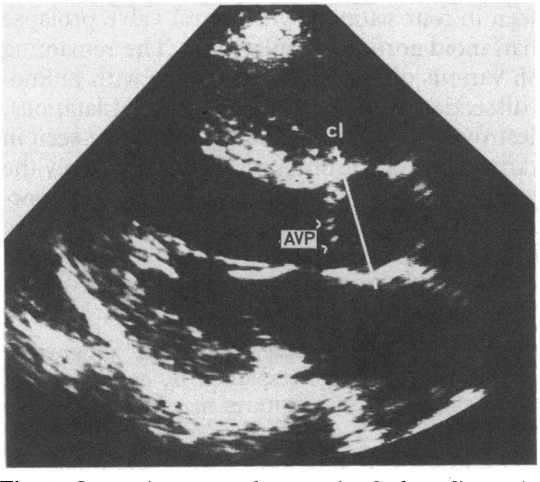

Fig. 1 *Long axis parasternal cross sectional echocardiogram in a patient with a bicuspid aortic valve and mild to moderate aortic stenosis. Aortic valve prolapse (AVP) is clearly seen with cuspal material below a line (cl) joining the points of attachment of the aortic leaflets to the annulus.*

Mitral valve prolapse was diagnosed when there was a systolic mitral click and a late systolic murmur or both. Systolic buckling or hammocking of the mitral valve detected by M mode echocardiography or mitral valve prolapse detected by cross sectional echocardiography also had to be present.

Aortic root dilatation was diagnosed when the end systolic aortic measurement exceeded 3·8 cm (2 SD from normal).

A bicuspid aortic valve was identified if either of the following was present (*a*) if in a diastolic short axis parasternal view the aortic valve did not form a normal Y pattern with commissures at 2, 6, and 10 o'clock and only two well defined cusps were identified with a single closure line; (*b*) if inspection at operation or necropsy showed a bicuspid aortic valve.

Results

Of the 2000 patients, 216 had no evidence of cardiac disease and in 212 the aortic valve cusps were not adequately imaged. The presence of valvar calcification made the aortic valve much more difficult to image and the aortic valve was seen sufficiently well in only 38 of 86 such patients. A total of 325 patients had rheumatic heart disease, 296 had valve replacement, 336 had coronary heart disease, and 320 had aortic valve disease (of whom 37 clearly displayed bicuspid aortic valve). Hypertrophic cardiomyopathy was seen in 63, congestive cardiomyopathy in 126, and mitral valve prolapse in 68 cases. The remaining 260 patients had various congenital heart diseases, pericardial disease, hypertension, and other less common conditions.

In patients without cardiac disease, the point of aortic valve closure was always above a line joining the valve attachments. We identified 24 patients with aortic valve prolapse, all of whom had evidence of cardiac disease; the Table shows their clinical details. They were aged 12 to 64 years and nine were women. In 10 the right cusp, in six the non-coronary cusps, and in eight both cusps had prolapsed. In seven cases the thickness of the aortic valve appeared to be normal, in one patient with infective endocarditis there appeared to be a vegetation on the aortic valve, and the remainder had some valve thickening. In only one case was there evidence of aortic valve flutter (valve destroyed and flail after endocarditis). Eleven of the 37 patients with obviously bicuspid valves had aortic valve prolapse, two had associated subvalvar aortic stenosis, and two had previously undergone aortic valvotomy. In seven, prolapse of the larger cusp was seen (Figs. 2a, 2b, and 3) and in four there was prolapse of both cusps (Figs. 1 and 4).

Only four of the 68 patients with mitral valve prolapse had aortic valve prolapse and seven had aortic root dilatation; one patient had both. Aortic valve prolapse was identified in only three patients without root dilatation (Fig. 4). Two other patients had aortic valve prolapse together with a dilated aortic root due to unknown causes or pulmonary atresia. Aortic valve prolapse in three patients was associated with rheumatic mitral and aortic valve disease. Although all these patients had aortic regurgitation, the predominant haemodynamic abnormality was related to the mitral blood flow. Seven patients with aortic valve prolapse did not have clinical evidence of aortic regurgitation. They included one patient with a bicuspid aortic

Table *Clinical details of patients with aortic valve prolapse*

Case No	*Age/(sex)*	*EDM/4*	*Blood pressure (mm Hg)*	*Prolapse of BCAV*	*MVP*	*Aortic root size*	*Comment*
1	67(M)	–	140/90			N	Rh, MVD, AVG 20 mm Hg, CAD
2	15(M)	2	110/70	+		N	AVG 35 mm Hg, SVAS
3	48(M)	2	150/70	*		N	Flail AV cusp, SBE
4	22(F)	2	110/50			E	Pulmonary atresia
5	68(M)	4	150/30		+	E	Marfan's syndrome
6	57(M)	3	140/45			E	ARD
7	59(F)	2	125/65			N	Rh MVD, AVG 25 mm Hg
8	34(F)	3	160/55			E†	
9	69(F)	–	130/80		+	N	Severe MR
10	13(M)	1	100/80	+		N	AVG 55 mm Hg, MVR, AVOT
11	16(F)	–	110/70	+		N	
12	28(F)	1	110/60	+		N	SVAS, AVOT
13	22(F)	2	120/65	+		N	AVG 40 mm Hg
14	24(M)	2	150/70	+		N	AVG 30 mm Hg
15	16(M)	1	120/85	+		N	AVG 25 mm Hg
16	58(F)	–	125/80			N	Rh MVD, trivial AVS
17	27(M)	3	150/60			E	AR due to SBE
18	54(F)	–	140/70		+	N	Severe MR
19	63(M)	–	130/80		‡	N	Severe MR
20	25(M)	1	130/70	+		N	
21	26(M)	–	110/70		+	N	Mild MR
22	60(M)	–	130/80	+		N	Mild AVS, CAD
23	19(M)	4	150/40	+		N	AVG 30 mm Hg
24	27(M)	3	130/70	+		N	

AV, aortic valve; AVG, aortic valve gradient (mm Hg); AVOT, valvotomy; AVS, aortic valve stenosis; AR, aortic regurgitation; ARD, idiopathic aortic root dilatation; Rh, rheumatic; BCAV, bicuspid aortic valve; MVD, mitral valve disease; MR, mitral regurgitation; MVP, mitral valve prolapse; MVR, mitral valve replacement; CAD, coronary artery disease; SVAS, subvalvar aortic stenosis; SBE, subacute bacterial endocarditis.
EDM/4, early diastolic murmur graded out of 4.
*AV destroyed by infection. †Aortic dissection present. ‡Redundant regurgitant mitral valve with no obvious prolapse.
+, present; –, absent; E, enlarged; N, normal.

valve, one with mild rheumatic aortic valve disease, and all four patients with mitral prolapse.

Discussion

We examined the prevalence of aortic valve prolapse in 2000 consecutive patients undergoing routine echocardiography. Although only 1·2% of patients had aortic valve prolapse, 30% of those with a bicuspid aortic valve showed prolapse. Both mitral and aortic valve prolapse were found in four patients, one of whom had marfanoid aortic root dilatation; aortic valve prolapse without root dilatation was seen in only three patients, two of whom had severe mitral regurgitation. Aortic valve prolapse was also seen in three cases with rheumatic heart disease and two with infective endocarditis. This latter finding is contrary to that reported by Becher *et al* who often found aortic valve prolapse in cases of endocarditis.[7] Aortic valve prolapse is known to be associated with a subaortic ventricular septal defect, but no such patient was identified in our series.[2]

There are several problems with the echocardiographic diagnosis of aortic valve prolapse. The aortic valve cusps may not be clearly seen and even with apparatus giving the highest resolution currently available, adequate images were obtained in only 89·3% of patients. There is no standard definition of aortic valve prolapse; that chosen for this study was of unknown sensitivity and specificity but in each case there was clear evidence of downward displacement and cuspal material below the aortic valve annulus. We excluded those echocardiograms that were not aligned along the left ventricular/aortic long axis since such echocardiograms often simulate aortic valve prolapse. This situation is analogous to that in mitral valve prolapse may be demonstrated in patients without evidence of cardiac disease.[8]

Aortic valve prolapse was most commonly found in bicuspid aortic valves and this confirms the findings of a recent study.[9] We found that in all cases the echocardiographically larger cusp prolapsed, but in four cases both cusps appeared to prolapse. Bicuspid aortic valves dome upwards in systole and prolapse downward in diastole, reflecting their cuspal distortion and fusion. Ejection clicks are typical of a bicuspid aortic valve; however, some patients may have a prolonged and loud closure sound,[10] possibly due to the sound of cuspal prolapse. Bicuspid aortic valves may be predominantly regurgitant,[3 11] and in the absence of infection and extensive calcification aortic valve prolapse is presumably one of the mechanisms responsible for such regurgitation. There were 212 patients in whom the aortic valve cusps could not be

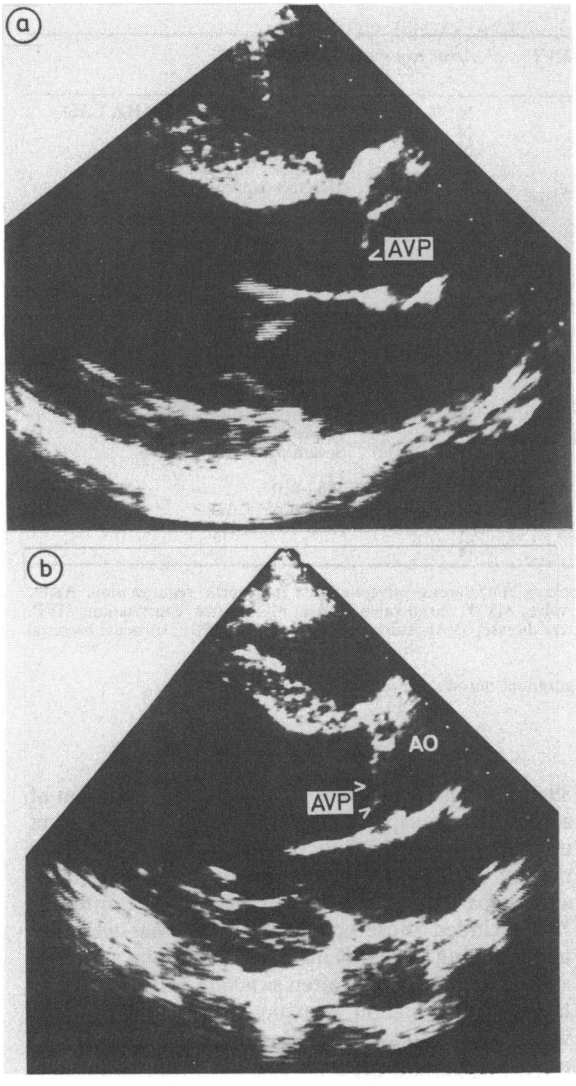

Fig. 2 *Two echocardiograms of patients with prolapse of the larger cusp of a bicuspid aortic valve. (a) Prolapse in a patient with no aortic regurgitation. (b) Similar appearance of the valve in a patient with severe valvular regurgitation. AO, aorta; AVP, aortic valve prolapse.*

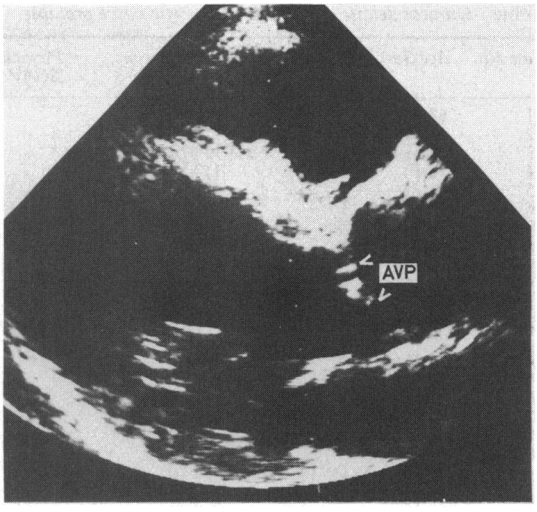

Fig. 3 *Echocardiogram showing prolapse of both aortic valve cusps in a patient with mild aortic stenosis only and no regurgitation. AVP, aortic valve prolapse.*

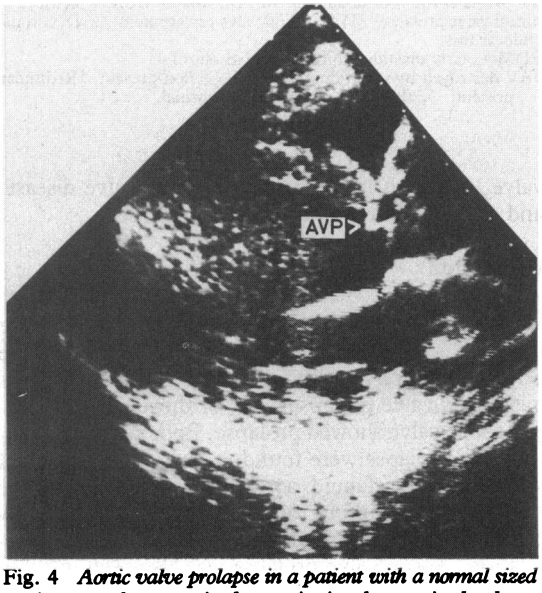

Fig. 4 *Aortic valve prolapse in a patient with a normal sized aortic root and severe mitral regurgitation due to mitral valve prolapse. There was no clinical or angiographic evidence of aortic regurgitation. AVP, aortic valve prolapse.*

adequately visualised and the cusps were obscured in 38 of the 86 with aortic valve calcification. A substantial proportion of the latter are likely to have had bicuspid aortic valves and therefore aortic valve prolapse may well be more common than indicated by this study.

An association between aortic valve prolapse and mitral valve prolapse has been reported by others. Sahn *et al* identified aortic root dilatation in 85% of patients with mitral valve prolapse[12] and aortic valve prolapse was seen in 20% of such patients.[5] In patients with associated aortic and mitral valve prolapse, aortic regurgitation was seen in four of 17[5] and eight of 12 cases.[13] Our findings contrast with these reports, since only seven of our 68 cases of mitral valve prolapse had aortic root dilatation and four of the seven had skeletal or other manifestations of Mar-

fan's syndrome. Only three patients without aortic root dilatation showed aortic valve and mitral valve prolapse, and two of these had severe mitral regurgitation requiring valve replacement. No patient had clinical evidence of aortic regurgitation. Aortic root dilatation or severe mitral regurgitation are manifestations of a more extreme form of the developmental connective tissue abnormality (unlike cases with minor echocardiographic mitral valve prolapse[8]) and they may represent a group with multivalvar myxomatous degeneration or a formes fruste of Marfan's syndrome.[5 14]

In conclusion, aortic valve prolapse was an infrequent finding in patients undergoing routine clinical echocardiography. Although these findings may represent an echocardiographic phenomenon rather than a diagnosis, in each case there was other evidence of valve disease, unlike the situation in individuals without evidence of valve disease in whom mild mitral valve prolapse may be demonstrated by echocardiography. In aortic root dilatation, the prolapse may occur with the formes fruste of Marfan's syndrome, presumably because of a defect in collagen metabolism, as occurred in the three patients with mitral valve prolapse without aortic root dilatation. In our series aortic valve prolapse was most commonly seen in the larger cusp of a bicuspid aortic valve. Three cases of rheumatic heart disease with predominant mitral valve involvement and two cases of infected valves were also seen. Presumably aortic valve prolapse may be the cause of aortic regurgitation, particularly in bicuspid aortic valves, valves destroyed by infection, and those malaligned because of root dilatation. Aortic valve prolapse did not imply the presence of aortic regurgitation, especially when aortic valve prolapse was associated with mitral valve prolapse.

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