Association of prolapse of posterior cusp of mitral valve and atrial septal defect

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Eleven patients with fossa ovalis atrial septal defects and prolapse of the posterior cusp of the mitral valve are described. Six patients had clinical evidence of mitral regurgitation, and in 2 others the electrocardiogram was unusual for uncomplicated fossa ovalis atrial septal defects. The varied appearance of the prolapsed cusp was shown by left ventricular angiography. The principal significance of this association is in its differentiation from atrioventricular defects.

The coincidence of prolapse of the posterior cusp of the mitral valve with fossa ovalis atrial septal defects has not previously been emphasized though the association occurred in 2 of 90 patients reported by Barlow *et al.* (1968) and 2 of 40 patients reported by Hancock and Cohn (1966). Prolapse of the posterior cusp of the mitral valve is now described in 11 patients who had, in addition, fossa ovalis atrial septal defects. A preliminary account of this work was given at a meeting of the British Cardiac Society in London in November 1969 (McDonald *et al.*, 1970).

Material and methods

The 11 patients were aged from 4 to 57 years; 3 were male and 8 female. Electrocardiograms, chest radiographs, and phonocardiograms were obtained in all patients and cardiac catheterization and left ventricular angiography were performed. The findings at operation were available in 5 patients.

Results

Symptoms Two patients were asymptomatic. Four had slight, and 4 moderate exertional dyspnoea; supraventricular tachycardia caused paroxysmal palpitation in 3, and ventricular ectopic beats occurred in the other patient. A past history of rheumatic fever was obtained in Case 6.

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Signs Apart from the usual clinical findings of an atrial septal defect (Cleland *et al.*, 1969), an abnormality of the mitral valve was suspected clinically in 6 patients (Table) who had a mitral pansystolic murmur which was associated with a mid-systolic click in 2. In one patient (Case 8) who had moderate pulmonary valvar stenosis and subvalvar stenosis with reversal of the atrial shunt there was moderate central cyanosis and finger clubbing. The skeletal manifestations of Marfan's syndrome were present in Case 2.

Electrocardiography Eight patients had partial right bundle-branch block and 3 complete right bundle-branch block (Table). The mean frontal QRS axis showed right axis deviation (greater than $+90^\circ$) in 7 patients and was normal in 3. The frontal axis was unusual for uncomplicated fossa ovalis atrial septal defects in 2 patients. Abnormal left axis deviation ($> -30^\circ$) was present in one case, and the axis was indeterminate in another, both having partial right bundle-branch block.

Radiology The findings in 10 patients were typical of atrial septal defect with cardiac enlargement, dilatation of the pulmonary artery, and pulmonary plethora. There was absence of pulmonary plethora in one patient (Case 8) who had moderate pulmonary stenosis. In 2 patients (Cases 4 and 6) there was enlargement of the left atrium suggesting a mitral valvar lesion.

TABLE Clinical findings and investigations in eleven patients

Case No., age, and sex	Dyspnoea	Palpitation	Apical pansystolic murmur	Mid-systolic click	Mean frontal QRS axis	Pulmonary systemic flow ratio	Pulmonary vascular resistance	Mitral regurgitation on angiography
1 4 F	—	—	—	—	Right axis deviation	2.5:1	Moderate	—
2 18 F	—	—	+	+	Right axis deviation	1.8:1	Normal	Moderate
3 28 F	+	—	+	+	Right axis deviation	2.4:1	Slight	Slight
4 28 F	++	—	+	—	Right axis deviation	2.6:1	Moderate	Slight
5 31 F	+	+	—	—	Indeterminate	2.3:1	Normal	—
6 34 F	++	+	+	—	Right axis deviation	1.8:1	Moderate	Slight
7 40 F	+	—	—	—	Left axis deviation	2.0:1	Slight	—
8 57 F	++	—	—	—	Right axis deviation	1.0:1.4	Normal	—
9 27 M	—	—	+	—	Normal	3.0:1	Normal	—
10 35 M	—	+	—	—	Right axis deviation	3.0:1	Normal	Slight
11 48 M	+	+	+	—	Right axis deviation	2.0:1	Slight	Moderate

+ present, — absent.

Phonocardiography The records confirmed the auscultatory findings; an illustrative sound recording from Case 2 is shown in Fig. 1.

Cardiac catheterization The presence of an atrial septal defect was shown in all patients. In 10 patients the atrial shunt was from left to right, the pulmonary/systemic flow ratio varying from 3 to 1.7 (Table). In the patient with pulmonary stenosis there was a

right-to-left shunt at atrial level resulting in moderate arterial desaturation. The pulmonary vascular resistance was normal in 7 patients and there was a slight to moderate rise in resistance in 5.

Angiography Left ventricular biplane angiograms (Elema-Schonander), or cineangiograms in the right anterior oblique were performed. Mitral regurgitation was seen in 6 patients; it was slight in 4, and moderate in 2 (Table). Prolapse of the posterior cusp of the mitral valve was shown in all. The angiographic appearance of prolapse varied particularly with regard to the size and the direction of the prolapse. In the anteroposterior plane the prolapse appeared in some cases as a ballooning deformity at the right border of the left ventricle (Fig. 2), in others the prolapse occurred to either side of the mitral ring (Fig. 3). In a few the ballooning cusp was mainly directed posteriorly (Fig. 4) and was not well seen in the anteroposterior plane. The prolapse into the left atrium occurred in systole; in some cases a little medium was seen to be caught among folds of redundant cusp tissue, as the posterior cusp swung into the left ventricular cavity during diastole. The size of the prolapse was not related to the degree of mitral regurgitation. For example, in Case 5 (Fig. 4 and 5) a large prolapse was shown but the mitral valve remained fully competent.

FIG. 1 Sound recording, Case 2. H.F., high frequency; P.A., pulmonary area; M.A., mitral area; CAR., external carotid pulse; L2, lead 2 of electrocardiogram; INSP., inspiration; C, systolic click; A₂, aortic component of second sound; P₂, pulmonary component of second sound.

FIG. 2 Left ventricular angiogram, antero-posterior view, Case 11. The arrow indicates the ballooning posterior cusp of the mitral valve outlined by contrast in the left atrium and left ventricle.

FIG. 3 Left ventricular angiogram, antero-posterior view, Case 7. The arrows indicate prolapse to either side of the mitral valve ring.

Surgery Surgery was undertaken in 5 patients and a fossa ovalis defect closed by direct suture. The appearances of the mitral valves at operation varied. In one patient (Case 6) who had a past history of rheumatic fever slight mitral stenosis was found and mitral valvotomy was performed, and no significant abnormality of the posterior cusp or chordae

was noted. In 2 patients (Cases 3 and 11) redundant posterior cusp tissue was found and in the latter patient (Case 11), who had moderately severe mitral regurgitation, this was particularly noticeable and a posterior cusp valvuloplasty was performed. This procedure appeared to reduce the amount of regurgitation, yet after operation the mitral pansystolic murmur became a late systolic murmur. In the remaining 2 patients (Cases 4 and 10), in whom slight mitral regurgitation was shown, no significant abnormality was noted on viewing the mitral valve from the atrium. It may be impossible with a non-beating empty heart to show the dynamic abnormality of a prolapsing posterior cusp of the mitral valve, and this seems likely to apply to Cases 4, 6, and 10.

Discussion

Prolapse of the posterior cusp of the mitral valve was shown by left ventricular angiography in all 11 patients, but in only 6 patients had a mitral valve abnormality been clinically diagnosed by the presence of a mitral pansystolic murmur. In the other 5 patients there were no auscultatory features suggesting a mitral valvar lesion. An unusual mean frontal QRS in 2 of these patients was considered a possible indication that an atrio-ventricular defect might be present. The characteristic deformity of the left ventricular outflow tract in atrioventricular defects (Baron *et al.*, 1964; Somerville and Jefferson, 1968) was not found on angiography. In the remaining 3 patients the prolapse was an incidental finding at angiography. A late systolic murmur has been shown to be due to mitral regurgitation (Barlow *et al.*, 1963; Segal and Likoff, 1964; Stannard *et al.*, 1967) and left ventricular angiography has shown that the regurgitation is related to prolapse of the posterior cusp of the mitral valve (Barlow and Bosman, 1966; Criley *et al.*, 1966). A mid or late systolic click is not uncommon and has been considered to be due to sudden chordal tension (Reid, 1961). The patients described here underline that prolapse of the posterior cusp need not occasion any abnormal physical signs, if mitral regurgitation is lacking, even when the size of the prolapse is considerable.

This type of deformity of the mitral valve has been noted in Marfan's syndrome (Read *et al.*, 1964; Criley *et al.*, 1966; Barlow *et al.*, 1968), and one patient in this series had the skeletal manifestations of Marfan's syndrome. In most instances, however, no definite aetiological factor is identified. Rheumatic endocarditis is not infrequently noted (Barlow *et al.*, 1968), as in one case in this series, but it

FIG. 4 Left ventricular angiogram, lateral view, Case 4. Arrow indicates large prolapse of posterior cusp of the mitral valve.

FIG. 5 Left ventricular angiogram, antero-posterior view, Case 4. Arrow indicates prolapse.

cannot be concluded that this abnormality was the consequence of rheumatic endocarditis. A familial incidence has been noted (Stannard *et al.*, 1967; Hunt and Sloman, 1969). In two series (Hancock and Cohn, 1966; Barlow *et al.*, 1968) two cases have been reported where prolapse was associated

with atrial septal defects of secundum type. In our own experience and that of Barlow *et al.* (1968), prolapse has rarely been found with other congenital lesions such as persistent ductus arteriosus, ventricular septal defect, atrioventricular septal defect, and pulmonary stenosis. The familial tendency, association with congenital cardiac lesions, and its occurrence in children suggest that this abnormality of the mitral valve may be congenital. The coincidence of fossa ovalis atrial septal defects with prolapse could be a random association of two fairly common abnormalities. However, the authors have performed many left ventricular angiograms in patients with ventricular septal defects and have found the association of a ballooned posterior cusp to be extremely rare, suggesting that the association with fossa ovalis atrial septal defects may be more specific. There is little pathological information. A voluminous posterior cusp with elongated chordae has been found at necropsy in one case (Barlow *et al.*, 1963), and reference to a localized expansion of the cusp has also been made (Hunt and Sloman, 1969). In the latter report the findings at operation in two patients with symptomatic mitral regurgitation were of ruptured chordae and large posterior mitral cusps, and others have reported similar findings (Marchand *et al.*, 1966). In Marfan's syndrome the chordae have been found to be unduly lax (McKusick, 1955; Raghib *et al.*, 1965). These findings support the contention that the primary abnormality is chordal in origin leading to a progressive ballooning of the cusp (Barlow *et al.*, 1968). The natural history of prolapse of the posterior cusp of the mitral valve is not known. It is possible that the severity of the mitral regurgitation may increase in time and cause significant symptoms. The risk of infective endocarditis in atrial septal defect of the fossa ovalis type is exceedingly small (Bedford, Papp, and Parkinson, 1941), but it may occur on the abnormal mitral valve in patients with atrioventricular defects. In patients with prolapse of the posterior cusp infective endocarditis has been found (Linhart and Taylor, 1966) and prophylactic antibiotic therapy at the time of septic hazard is indicated. When atrial septal defects of fossa ovalis type are associated with prolapse of the posterior cusp, especially where there are signs indicating mitral regurgitation, it is important to distinguish this combination from atrioventricular defects. The left ventricular angiographic features are distinct.

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