

Ballooning Deformity (Mucoid Degeneration) of Atrioventricular Valves

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In the past three years several reports have drawn attention to a type of mitral incompetence associated with a peculiar deformity of the mitral valve. This is characterized by an increase in area of the affected leaflets which become voluminous and prolapse into the atrium in systole. Various names or descriptions have been applied to the condition, based either on pathological findings (described as myxomatous transformation or floppy valve syndrome by Read, Thal, and Wendt, 1965, and Read and Thal, 1966; and as billowing sail deformity by Oka and Angrist, 1961) or on cine-angiographic appearances (described as ballooning of posterior leaflet by Linhart and Taylor, 1966, and Behar, Whalen, and McIntosh, 1967; as prolapse of posterior leaflet by Criley *et al.*, 1966, and Stannard *et al.*, 1967; as aneurysmal protrusion and billowing of posterior leaflet by Barlow *et al.*, 1968, and Barlow and Bosman, 1966). It appears identical to the two cases described in 1958 by Fernex and Fernex as mucoid degeneration. Most of these reports have been from centres in the U.S.A. and South Africa, but necropsy observations suggest that similar cases are not uncommon in this country.

SUBJECTS

The 35 cases seen in the past 6 years consisted of 23 men and 12 women whose ages ranged from 51 to 98 years. Of these, 30 occurred in the 3083 adult necropsies performed in this hospital, an incidence of 1 per cent. The available clinical and pathological data are summarized in the Table. Though the presence of heart murmurs had been known for up to 25 years, none of the patients was under 50 years at death, and the average age was 73.6 years. Their age distribution was similar to that for necropsies generally in this hospital.

Only 16 patients had presented with cardiovascular signs or symptoms. In 5 cardiac failure was clinically

considered secondary to pulmonary disease, in 1 to a combination of hepatic and respiratory failure, and in 2 to ischaemic heart disease; 3 patients were thought to have rheumatic valvular disease, and 3 (including one of the rheumatic cases) bacterial endocarditis, 1 congenital heart disease, and 3 "senile" heart failure. Nineteen patients were in cardiac failure on their terminal admission, with atrial fibrillation in 5 patients. None had been hypertensive. Electrocardiograms in 14 patients showed no constant abnormalities. Apart from one (Case 34), who had a large recent infarct, ischaemic changes were reported in 3 patients, only one of whom had complained of angina.

Systolic murmurs had been present in 24 patients; these too had no constant features; descriptions ranged from short apical to loud pansystolic with thrill. The presence of murmurs was not assessable in 7 patients whose heart sounds were either obscured by pulmonary adventitious sounds or were too faint. In 4 patients, heart sounds had been recorded as normal. The murmurs had been attributed to rheumatic heart disease in 3 patients; Cases 25 and 31 had histories of attacks of rheumatic fever, and the rheumatic aetiology in Case 23 was suggested by a radiological finding of calcification in the mitral area (later shown to be in the valve ring). Case 2, a woman aged 51, with longstanding kyphoscoliosis, was thought to have tricuspid incompetence due to right ventricular dilatation, and Case 26, a woman aged 98, was considered to have mitral ring calcification. Mitral incompetence of undetermined aetiology had been diagnosed in Cases 11, 19, and 35. One patient (Case 5), whose murmur had first been noted 20 years previously, had been labelled "functional", and another (Case 31), also known to have had a murmur since army medical examination 25 years before death, was considered to have congenital heart disease.

Only 2 patients had past histories of rheumatic or scarlet fever (and neither showed evidence of old carditis at necropsy). Cases 7 and 18 had pulmonary tuberculosis and pneumonia, Case 29 had been gassed in the 1914-1918 war, and 6 others were chronic bronchitics.

Family history was non-contributory in most instances. Parents or sibs of 3 patients had died of heart disease, but the nature of this was not ascertained.

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Case No.	Age (yr.)	Sex	Clinical presentation	Cause of death	Past medical and family history	Blood pressure (mm. Hg)	Pulse	Cardiac failure
1	54	M	Cor pulmonale; kyphoscoliosis	Bronchopneumonia and cardiac failure	Congenital kyphoscoliosis	?	?	+
2	51	F	Cor pulmonale; kyphoscoliosis	Pulmonary thrombosis, cardiac failure	Potts disease at 3 yr.	130/90	Fibrillating	+
3	69	F	Coma and pyrexia	Staphylococcal endocarditis with multiple emboli	None obtained	120/90	Regular	+
4	80	F	Pleural effusion	Carcinoma of bronchus with metastases	None obtained	130/70	Regular	+
5	58	M	Retention of urine	Carcinoma of prostate with metastases	Angina for 5 yr.; rejected for military service 20 yr. ago because of "murmur"	170/90	Regular	-
6	83	F	Dyspnoea and backache	Pulmonary embolism	None obtained	120/90	Fibrillating	+
7	81	M	Acute-on-chronic bronchitis	Carcinoma of bronchus	Old pulmonary TB	130/70	Regular	-
8	63	M	Acute-on-chronic bronchitis	Cardiac failure	None relevant	160/80	Regular	+
9	84	F	Arteriosclerotic gangrene	Bronchopneumonia	None relevant	130/90	Regular	-
10	90	M	Senile dementia	Bronchopneumonia	None obtained	140/70	Regular	-
11	62	M	Cerebral tumour	Metastases from carcinoma of bronchus	None obtained	120/90	Regular	-
12	79	M	Cardiac failure	Cardiac failure, bronchitis, pulmonary embolism	None obtained	?	?	+
13	84	M	Laryngeal stridor	Acute pancreatitis, tracheobronchitis	Previous attacks of cardiac failure	130/80	Regular	+
14	89	F	Scleroderma	Carcinoma of bronchus	None relevant	100/70	Regular	-
15	76	M	Carcinoma of bronchus	Carcinoma of bronchus	None relevant	140/70	Regular	-
16	61	M	Dyspnoea	Carcinoma of bronchus with metastases	None relevant	100/70	Regular	-
17	84	F	Abdominal pain, distension	Carcinoma of stomach	Parent died at 33 from "heart trouble"	190/90	Regular	-
18	87	M	Coma and hypothermia	Bronchopneumonia	Pneumonia 20 yr. previously	130/70	Regular	+
19	79	M	Chest pain	Carcinoma of bronchus	None relevant	170/100	Regular	-
20	83	M	Urinary symptoms	Bronchopneumonia and suppurative pyelonephritis	Several yr. "heart trouble"	120/70	Regular	+
21	54	M	Chest and neck pain	Carcinoma of bronchus	None relevant	180/95	Regular	-
22	63	F	Asthma	Haemorrhage from gastric ulcer	None obtained	125/75	Regular	-
23	58	M	Cardiac and hepatic failure	Cardiac failure and cirrhosis of liver	Chronic bronchitis and heavy drinking	110/100	Fibrillating	+
24	85	F	Pneumonia, kyphoscoliosis	Bronchopneumonia	None obtained	?	?	?
25	75	F	Recurrent hemiplegia	Cerebral infarction and bronchopneumonia	Rheumatic fever at 15 and 40 yr.; cardiac symptoms 15 yr.; mother and brother died in 50's of "hearts"	110/70	Fibrillating	+
26	98	F	Pneumonia, pancytopenia	Lobar pneumonia	None obtained	140/65	Regular	-
27	60	M	Carcinomatosis	Carcinoma of pancreas	None obtained	110/60	Regular	-
28	73	M	Abdominal pain	Carcinoma of caecum	None relevant	160/80	Regular	-
29	84	M	Cardiac failure, gangrene	Aortic thrombosis and carcinoma prostate	Gassed in 1914-1918 war	130/80	Regular	+
30	63	M	Sudden death	Cardiac failure; ruptured chorda tendinea	Several yr. progressive cardiac failure; murmur in army medical 25 yr. ago	120/90	Regular	+
31	79	F	Cardiac failure (thought to be rheumatic)	Cardiac failure	Rheumatic fever	150/90	Regular	+
32	61	M	Cardiac failure and cerebral signs	Emboli from bacterial endocarditis	Dyspnoeic for 4 yr.; mother died of "heart trouble"	120/90	Regular	+
33	57	M	Jaundice, cardiac failure	Cardiac failure; acute-chronic bronchitis	Chronic bronchitic	90/65	Fibrillating	+
34	84	M	Acute myocardial infarct	Myocardial infarction	None obtained	110/70	Regular	+
35	78	M	Chest pain, cardiac failure, cyanosis, leg vein thrombosis	Cardiac failure; pulmonary embolism and thrombosis; ruptured chordae tendineae	A 10-yr. previous admission and 2 recent ones (for pulmonary embolism from deep leg veins)	150/100	Regular	+

(cont'd)

Murmurs	ECG findings	Heart weight (g.)	Extent of ballooning	Other pathological changes
? (heart sounds obscured)	—	410	Both AV valves, all cusps	—
Loud pansystolic, noted 6 yr. previously (diagnosed as tricuspid incompetence)	Atrial fibrillation	300	Both AV valves, all cusps	Fibrinous endocarditis
Mitral pansystolic with prediastolic crescendo (diagnosed as mitral incompetence)	—	350	Posterior cusp mitral only	Bacterial endocarditis with ruptured chordae tendineae
? (heart sounds faint)	—	300	Posterior cusp mitral only	—
Harsh systolic, maximal at apex (diagnosed as functional)	Normal	360	Both cusps mitral, most marked in posterior	Fibrinous endocarditis
Loud apical systolic, conducted toward axilla	Suggestive of old infarction	400	Both cusps mitral	Marked left atrial dilatation
? (heart sounds obscured)	—	?	Posterior cusp mitral and anterior and septal cusps tricuspid	Post-inflammatory adhesions of mitral commissures (slight) and vascularization of mitral cusp
? (heart sounds obscured)	Normal	230	Posterior cusp and medial half anterior cusp mitral	Marked dilatation left atrium; posterior cusp chordae adherent to ventricular wall, with active inflammatory changes still visible
Short apical systolic	—	270	Both cusps mitral	—
Doubtful apical diastolic	—	300	Mainly posterior cusp mitral	Old myocardial infarct
Loud apical pansystolic, also heard 1 yr. previously (diagnosed as mitral incompetence)	Normal	320	Posterior cusp mitral only	Large area of endocardial roughening with adherent fibrin on posterior wall of left atrium
Apical systolic	—	510	Both cusps mitral	Minor congenital chordae tendineae abnormalities; small area myocardial infarction
? (heart sounds obscured) but apical systolic 4 yr. previously	—	490	Both cusps mitral	Roughening of left atrial posterior wall endocardium with adherent fibrin
Systolic, at junction of apex and aortic area; first heard 5 yr. previously	—	220	Mainly posterior cusp mitral	Endocardial "pocket" anterior cusp mitral, also small aneurysm membranous septum
None heard	—	250	Posterior cusp mitral only	Fibrous "jet" lesion anterior mitral cusp
None heard	—	310	Posterior cusp and mild anterior cusp mitral	—
None heard	—	270	Posterior cusp mitral only	—
Systolic (details obscured by adventitious sounds)	—	330	Both cusps mitral and mild anterior cusp tricuspid	—
Harsh systolic, maximal 4th space conducted to axilla (diagnosed as mitral incompetence)	Incomplete left bundle-branch block	320	Both cusps mitral and slight changes anterior tricuspid	Congenital abnormalities of chordae
Apical pansystolic	Suggestive of digitalis overdosage	240	Posterior cusp mitral only	—
None heard	—	320	Mainly posterior cusp mitral, slight anterior cusp only	Small vegetation on thickened area on anterior mitral cusp
? (heart sounds faint)	—	310	Posterior cusp mitral only	—
Loud apical systolic with thrill over mitral area (diagnosed as mitral incompetence)	LV hypertrophy and atrial fibrillation	670	Both cusps mitral and slight changes anterior tricuspid	Fibrinous endocarditis mainly over severe ring calcification ulcerating through cusp; congenital aortic cusp fusion
Mitral pansystolic, conducted to axilla	—	250	Both cusps mitral	—
Apical pansystolic conducted to axilla, and softer mid-diastolic (diagnosed as mitral stenosis and incompetence)	Atrial fibrillation	420	Both cusps mitral and anterior tricuspid	Marked left atrial dilatation; atrial septal defect; severe mitral ring calcification
Apical pansystolic (diagnosed as mitral ring calcification)	—	230	Both cusps mitral and slight changes anterior tricuspid	Small foci fibrinous endocarditis both mitral cusps
Soft apical systolic	—	250	Both cusps mitral	Brown atrophy of myocardium
Soft apical systolic	? Ischaemia	370	Mainly posterior cusp mitral	Old "jet" lesions posterior wall of left atrium
? (heart sounds obscured)	—	?	Both cusps mitral	Fibrous adhesions of postero-lateral chordae tendineae to posterior wall of left ventricle
Loud praecordial systolic with thrill (diagnosed as congenital, probably ventricular septal defect)	No significant abnormality	610	Both cusps mitral	Rupture of 2 postero-medial chordae tendineae; fibrinous endocarditis
Pansystolic in mitral area, conducted to axilla (diagnosed as rheumatic mitral incompetence)	—	435	Both cusps mitral mainly anterior, and anterior and septal tricuspid cusps	—
Apical pansystolic conducted to axilla and neck	Ischaemic changes	710	Both cusps mitral, mainly posterior	Bacterial endocarditis both cusps mitral
? (sounds inaudible)	—	460	Both cusps mitral	—
Apical systolic	Anterior infarct.	?	Both cusps mitral, mainly posterior	Myocardial infarction
Loud apical systolic conducted to axilla (diagnosed as dilatation of mitral ring)	Occasional ectopic beats; tall P waves, lead III	450	Both cusps mitral, mainly posterior	Rupture of 3 postero-medial chordae tendineae; mitral ring calcification



FIG. 1.—Photomicrograph of part of a mitral cusp showing loose myxomatoid tissue replacing the fibrosa. A narrow zone of normal collagen remains immediately below the atrialis. (Haematoxylin and eosin. $\times 290$.)

PATHOLOGY

The hearts were mainly of normal size, weights ranging from 220–710 g., average 366 g. In 23 cases, both cusps of the mitral valve were voluminous, opaque, and thickened, resembling a parachute. This ballooning deformity tended to be more conspicuous in the posterior cusp, which was often of larger area than the anterior. In 11 cases only the posterior cusp was macroscopically affected. Similar but less well marked changes were seen in one or more of the tricuspid valve cusps in 9 patients. Microscopically the affected cusps showed replacement of the fibrosa by a loose metachromatically staining myxomatous tissue with fibro-elastic thickening of the surrounding endocardium (Fig. 1). Both these changes were present in variable degrees in all the affected valves, but the basic abnormality appeared to be the fibrosal degeneration. Loss of this normally dense collagenous supporting structure would clearly allow stretching of the cusp by

normal variations in intraventricular pressures, and would result in the characteristic voluminous ballooned leaflets. The accompanying fibro-elastic proliferation was of the non-specific type associated with various haemodynamic stimuli (McMillan and Lev, 1959; Pomerance, 1967) and presumably provoked by the stretching process.

Superficial fibrinous “endocarditis” was seen in 10 patients (Fig. 2). The lesions consisted of foci of amorphous eosinophilic material, staining as fibrin, either in the atrialis, or adherent to atrial endocardium. Red blood cells were often present in the deeper lesions but no inflammatory reaction was seen. Bacterial endocarditis was present in 2 patients, both dying of systemic emboli. The infecting organism in Case 3 was *Staph. pyogenes*, and in Case 32 (Fig. 3) a non-haemolytic streptococcus. Three cases had ruptured posterior group chordae tendineae; in Case 3 this had occurred under a staphylococcal vegetation, but Cases 30 and 35 (Fig. 4) had no evidence of current or previous bacterial infection. Subvalvular areas of opaque white thickening were present in the posterior wall of the left ventricle in two hearts. These involved the adjacent chordae tendineae, which had become adherent to the ventricular endocardium, and in Case 8 chronic inflammatory cells were still visible in the abnormal fibrous tissue. Case 7 was the only example of previous valvulitis—slight adhesions were present at both mitral commissures, with macroscopical and microscopical vascularization of the anterior cusp.

Striking anatomical evidence of mitral incompetence was seen in 8 patients, 3 of whose hearts showed gross left atrial dilatation, and “jet” lesions were present on atrial walls or anterior mitral cusps in 6.

OTHER CARDIAC PATHOLOGY

Coronary embolism had occurred in Case 32, resulting in a giant cell myocarditis and pericarditis. Coronary atherosclerosis appeared less than expected in patients of this age-group. Case 34 was the only patient dying with an extensive recent myocardial infarct, though a small area of recent infarction was also present in Case 12, and an old fibrotic lesion was seen in Case 10. The coronary arteries were almost free from atheroma in 8 cases and showed mild narrowing only in a further 7. Marked atherosclerotic changes were present in only 8 of the 35 cases. The only other abnormality encountered with any frequency was calcification of the mitral valve ring, present in 9 cases, but severe in only 3 (Fig. 5); 8 of these were over 70 years, and the incidence was no higher than anticipated in this age-

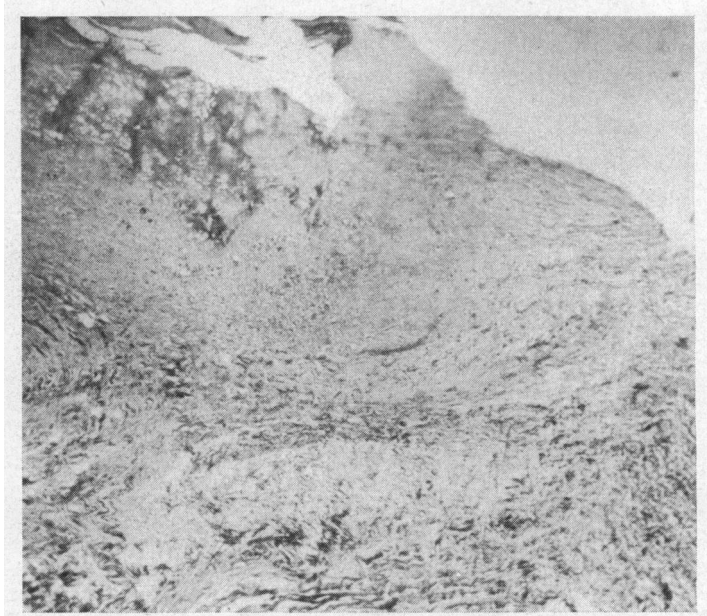


FIG. 2.—Photomicrograph showing tearing with fibrin and red blood cells in superficial zone of mitral valve atrialis. (Haematoxylin and eosin. $\times 470$.)

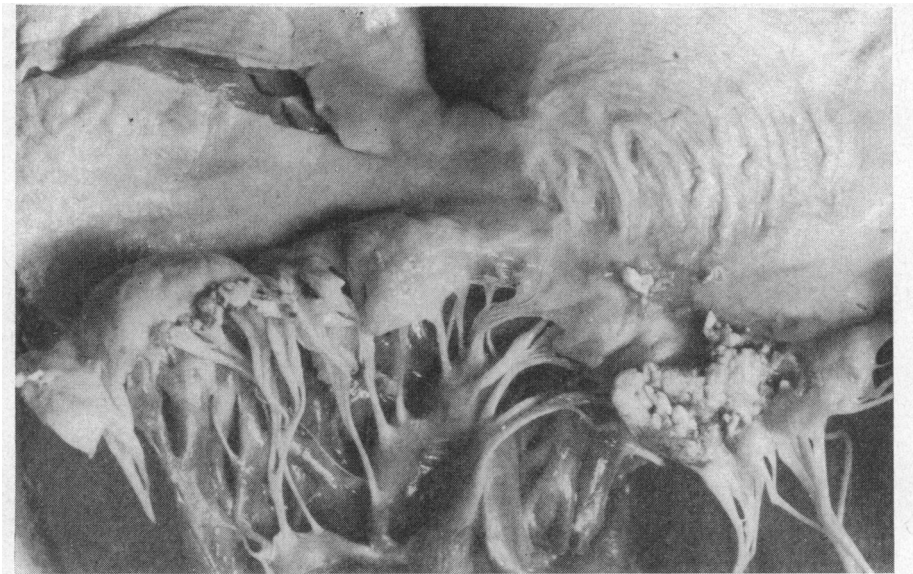


FIG. 3.—Opened left side of heart showing ballooning, more marked in the posterior cusp, with large vegetations on both cusps. An area of thick ridged endocardium above the anterior cusp indicates chronic mitral incompetence. Case 32, a man aged 61 admitted complaining of severe dyspnoea, palpitations, and loss of weight for 4 months. Dyspnoea on exertion had been present for 4 years. He was pyrexial, in congestive failure, and had a loud apical pansystolic murmur, conducted to axilla and carotids. Other signs and symptoms suggesting renal and cerebral embolism were present, and blood culture yielded a non-haemolytic streptococcus. He became afebrile on antibiotics, but emboli continued and he died with multiple infarcts, including cerebral.

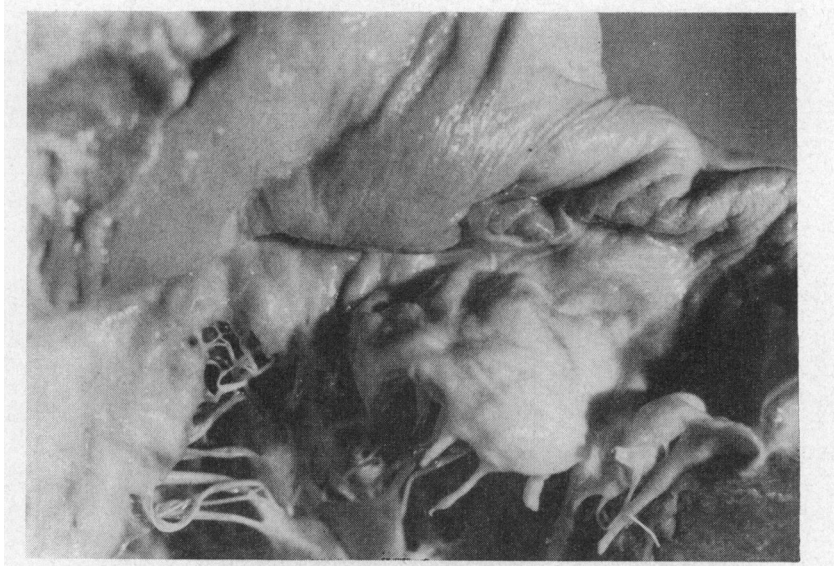


FIG. 4.—Part of opened left side of heart showing three ruptured chordae tendineae attached to the ballooned and thickened posterior cusp. Case 35, a man of 78, admitted for the third time in one year with pulmonary emboli from deep vein thrombosis. He was in failure, with a harsh apical systolic murmur conducted to axilla. The murmur had been heard on the previous admissions, but had not been conducted.

group. Case 23 (Fig. 6) was a comparatively young man, who also had hepatic cirrhosis. Brown atrophy was noted in 1 case, and 5 hearts had relatively minor congenital abnormalities consisting of

anomalous chordae tendineae in 2 and single examples of atrial septal defect, aneurysm of membranous interventricular septum, and bicuspid aortic valve in the other 3.

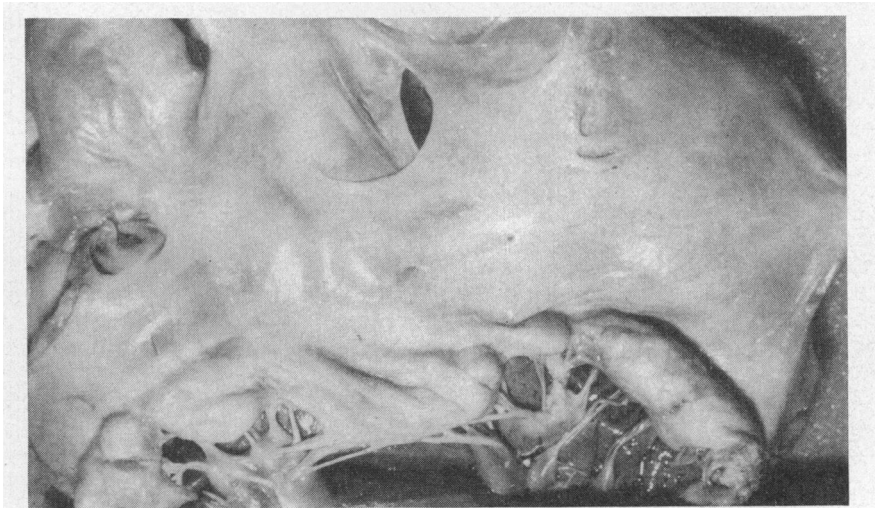


FIG. 5.—Part of opened left side of heart showing an atrial septal defect, and ballooning deformity of both mitral cusps. The posterior cusp is further distorted by a bar of calcification originating in the valve ring. No adhesions of commissures or chordae are present. Case 25, a woman aged 75, admitted with recurrent hemiplegia. She had a history of rheumatic fever at 15 and 40 years, but no further cardiac symptoms until 60 years. She was fibrillating but not in failure, and had an apical pansystolic murmur, conducted to axilla, and mid-diastolic murmur. These were attributed to rheumatic mitral stenosis and incompetence. Death was due to bronchopneumonia and cerebral infarction.

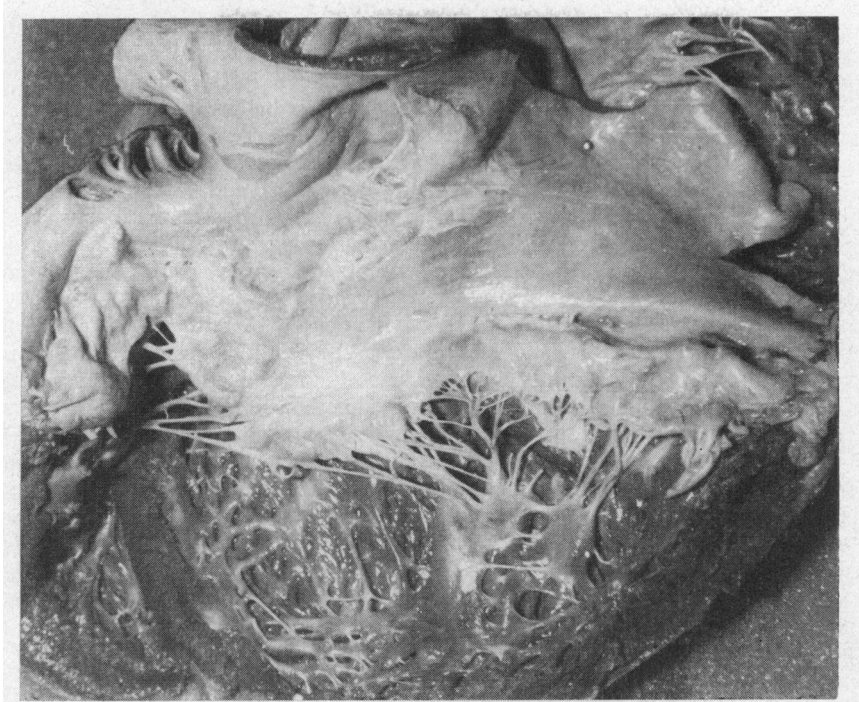


FIG. 6.—Opened left side of heart showing ballooned and floppy mitral valve. Spurs of calcium extend from the ring and distort the posterior cusp. Small patches of endocardial fibrosis are seen high on the posterior wall of the ventricle. Case 23, a man of 58 with two years of progressive cardiac failure and a loud systolic murmur over the whole praecordium, conducted to axilla. Calcification in the mitral area was seen on screening, and a diagnosis of rheumatic mitral disease was made. He was a longstanding epileptic and chronic bronchitic, and drank 15 pints of Guinness daily. Death was due to combined cardiac and hepatic failure.

DISCUSSION

There is no doubt about the identity of cases where affected cusps were examined pathologically, and it is difficult to envisage any process other than softening of cusp fibrosa which could produce a ballooning, billowing, or prolapsing appearance on cine-angiography. Stannard *et al.* (1967) also believed the anatomical deformity to be enlargement of cusps; this was confirmed in the only patient who subsequently came to necropsy (Barlow *et al.*, 1968).

Review of clinical findings in the present series showed no constant features which might assist detection of the ballooning deformity. Until recently the condition was not recognized as a clinical problem, but Behar *et al.* (1967) and Linhart and Taylor (1966) suggest that it may not be uncommon. Behar *et al.* (1967) showed ballooning in 6 per cent of cardiac clinic patients. In this hospital some degree was found in 4.5 per cent of necropsies on elderly patients with systolic murmurs (Pomerance, 1968). However, even knowing

its comparatively high incidence, clinical differentiation between ballooning, ring calcification, and other less common causes of mitral incompetence was not possible. As in clinical reports (Behar *et al.*, 1967), murmurs had ranged from short apical to diffuse pansystolic with thrills. Neither intensity nor localization could be related to severity of abnormality; both patients with thrills had had severe ballooning of both mitral cusps, but equally severe changes were present in cases without thrills.

Though all had a severe degree of ballooning, only 19 cases were in failure and those without cardiac symptoms included 4 of 8 with gross left atrial dilatation or "jet" lesions. This finding, together with a high average age compared with the clinical series, confirms views (Linhart and Taylor, 1966; Stannard *et al.*, 1967) that this type of mitral incompetence is relatively benign. The predisposition to bacterial endocarditis and spontaneous rupture of chordae tendineae was also confirmed, but review of clinical data showed nothing to distinguish patients in whom mucoid degeneration proved fatal

from those in whom it remained a benign condition.

The contrasting sex incidence in necropsy (23 men: 12 women) and clinical (17:32) series is less easily explained. Possibly women develop symptoms and complications earlier than men, but the total number of cases is, as yet, too small for speculation. Furthermore, cases complicated by rupture of chordae tendineae occurred predominantly in men. The appearances in the cases of Marchand *et al.* (1966) were characteristic of mucoid degeneration; 5 of the 6 were men, as were both my cases.

The predisposition to bacterial endocarditis noted in clinical reports (Read *et al.*, 1965; Read and Thal, 1966; Linhart and Taylor, 1966) was confirmed in our necropsy series, and pathological examination offered an explanation, since fibrinous "endocarditis" was seen in 10 cases. The endocardial damage appeared secondary to the underlying fibrosal changes. With transformation of this normally rigid plate into loose myxomatous tissue the cusp would be stretched by normal intraventricular pressure changes. Overlying endocardium would inevitably also be subjected to abrupt changes in tension, and resulting loss of endothelial continuity and rupture of subendothelial connective tissue fibres would invite the deposition of fibrin seen in these cases.

The pathogenesis of mucoid degeneration of heart valves is unknown. It seems unrelated to rheumatic fever or any particular past illnesses, and no histological evidence of past inflammation has been found. A similar change occurs in Marfan's syndrome (Goyette and Palmer, 1953; Shankar *et al.*, 1967), and Read *et al.* (1965) suggested that it might therefore be a "forme fruste". Most of their 9 patients or their families had some of the musculoskeletal stigmata, but few patients in other series have shown similar findings. Furthermore, these valve changes have also been reported in other congenital heart diseases and in normal infants (Shankar *et al.*, 1967), and small areas of similar metachromatically staining material are often seen in random post-mortem sections at all ages.

In some reported cases ballooning does seem genetically determined (Read *et al.*, 1965; Stannard *et al.*, 1967; Linart and Taylor, 1966, Barlow *et al.*, 1968), but these seem a minority. Marchand *et al.* (1966) also thought that the abnormality was congenital, and they are supported by the early age at which some murmurs were first heard. However, most necropsy cases have been elderly; it seems improbable that Case 26, for example, with gross left atrial dilatation, could have survived 98 years if her mitral deformity had been congenital.

In contrast, Oka and Angrist (1961) suggested that the deformity was an ageing change. A similar

condition in dogs (Pomerance and Whitney, 1969) is strikingly related to advanced age. Though a slight increase in the comparatively common localized areas of ballooning occurs in men (Pomerance, 1967), no definite correlation with age was demonstrated in severe ballooning. The highest incidence was between 80 and 90 years, but in the 50-69 year groups it was greater than between 70-79 years. Furthermore, a third of cases diagnosed at operation or cine-angiography were under 30, and almost all were under 50 years.

It appears, therefore, that the small number of cases studied includes individuals providing evidence both supporting and contradicting any proposed pathogenesis, which suggests that there is no single cause of mucoid degeneration of valves. The most satisfactory explanation of currently available facts is that this is simply a tissue reaction, comparable to endocardial fibro-elastic thickening, and similarly the end result of many aetiological factors, including both congenital and ageing changes as well as a variety of diseases of intermediate ages.

SUMMARY

A severe ballooning deformity of the mitral valve was found in 1 per cent of necropsies over a six-year period. The 35 cases (23 men, 12 women) were aged between 51 and 98 years. Clinical presentation had been varied, and only 19 cases had been in failure. Systolic murmurs, heard in two-thirds of the patients, showed no diagnostic characteristics and were not related to severity of valvular deformity. Electrocardiograms were non-contributory. The pathological changes ranged from enlargement of the posterior cusp only, to involvement of the tricuspid valve as well as both mitral cusps. Microscopy showed replacement of cusp fibrosa by metachromatically staining loose myxomatous material, with fibro-elastic thickening of adjacent endocardium. Fibrinous "endocarditis" was common and appeared secondary to stretching of a fibrosa no longer able to resist intraventricular systolic pressures. Two cases were complicated by bacterial endocarditis and two by ruptured chordae tendineae without endocarditis, but though mitral incompetence (as evidenced by left atrial dilatation, "jet" lesions, or murmurs) was present in most cases, 16 patients had no symptoms related to their cardiac pathology.

The cases diagnosed clinically and at necropsy provided evidence both for and against all previously discussed pathogenesises, and it is suggested that mucoid degeneration of heart valves is a non-specific tissue change and may result from many possible aetiological processes.

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