

# Parachute deformity of the mitral valve

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A case of parachute deformity of the mitral valve, a rare congenital form of mitral stenosis characterized by insertion of the chordae tendineae into a single posterior papillary muscle, is described in an 11-year-old girl. The eleven other cases in the English literature are reviewed.

Shone, Sellers, Anderson, Adams, Lillehei, and Edwards (1963) described a developmental deformity of the mitral valve characterized by insertion of the chordae tendineae into a single papillary muscle, producing a funnel-shaped valve which is to some degree stenotic. They employed the term 'parachute' first used by Edwards (1961), and noted an association with other left-sided cardiac deformities. We have discovered only 11 such cases published in English. We record the finding of such a parachute mitral valve in association with a bicuspid stenotic aortic valve.

To avoid possible confusion the term 'parachute mitral valve' is used throughout this paper in the way originally used by Edwards, and not to describe a ballooned mitral valve cusp (Hudson, 1965).

## CASE REPORT

Our patient was a white girl, who was first found to have a heart murmur during a routine school medical examination at the age of 5 years. She was slightly more short of breath on exertion than her fellows, and when aged 6 was first admitted to the Cardiac Unit, Papworth Hospital, for assessment. She was normally developed for her age. Her right brachial blood pressure was 90/60 mm. Hg, and her pulse was soft and regular. Her apex beat was described as forceful but not displaced. An ejection click preceded a loud systolic murmur and thrill of maximal intensity in the second left intercostal space adjacent to the sternum. Only one element of the second sound was heard. No murmurs indicative of mitral valve disease were heard at this time or subsequently.

The electrocardiogram showed sinus rhythm and was within normal limits. The chest radiograph was suggestive of left ventricular enlargement. No enlargement of the left atrium was apparent.

Later that year she developed measles, and thereafter suffered from recurrent respiratory infections. At the age of 7 the diagnosis of aortic stenosis was confirmed by direct left ventricular puncture. The peak systolic gradient was 40 mm. Hg.

At the age of 11 she was admitted to Huntingdon Hospital with a three-day history of fever, dyspnoea,

and cough productive of purulent, blood-stained sputum. She was extremely ill, with a rapid, feeble pulse and the signs of right basal consolidation. The electrocardiogram showed first degree heart block (P-R interval 0.24 sec.) and T-wave inversion in the left ventricular precordial leads. She was treated for pneumonia and heart failure without response, and on the eleventh hospital day was transferred to the Cardiac Unit, Papworth Hospital. By this time she was dyspnoeic and pallid, with cold cyanosed extremities. The apex beat was felt in the anterior axillary line, and a soft systolic murmur and gallop rhythm were audible. Signs of consolidation were evident at both lung bases and the liver edge was palpable 4 cm below the right costal margin. Despite continued intensive treatment she died of pulmonary oedema two days later.

Her younger sister has been noted to have an ejection murmur which has been attributed to congenital aortic stenosis. She is without symptoms and has not yet been investigated. Her elder brother has been seen by one of us because of an ejection murmur maximal along the right border of the sternum. He also has no symptoms related to his heart. His electrocardiogram shows an incomplete right bundle-branch block pattern and his chest film demonstrates that the cardiac shadow is not enlarged, but that there is some prominence of the main pulmonary artery.

**PATHOLOGICAL FINDINGS** At necropsy the significant lesions, apart from evidence of heart failure, were confined to the heart and lungs. There was a millimetric spot, 1.5 cm. in diameter, on the anterior surface of the left ventricular pericardium. The heart appeared slightly dilated and weighed 380 g. The atria were of nearly equal size, the larger right atrium containing 50 ml. of post-mortem thrombus. The foramen oval was closed. The mitral orifice was 2.2 cm. by 1.5 cm. and was placed rather posteriorly in the floor of the left atrium. There was no supravulvar ring. The left atrial muscle was no more than slightly hypertrophied. The antero-lateral papillary muscle was absent and appeared to be represented by a few parallel muscle bundles running in a basal-apical axis on the antero-lateral wall of the dilated left ventricle. The mitral valve cusps were thin and pliable, without commissures, and funnel-shaped. Thin chordae inserted into the paired postero-medial papillary muscle (Figure

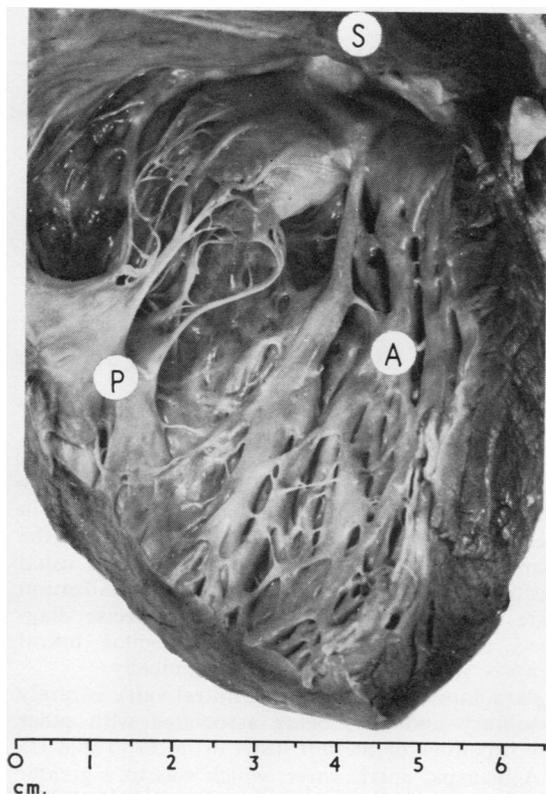


FIGURE Left ventricle showing mitral chordae tendineae inserting into bifid postero-medial papillary muscle (P) and the parallel muscle bundles in the position of the antero-lateral papillary muscle and chordae (A). Interventricular septum (S).

The left ventricular muscle was of normal colour and texture but hypertrophied, being 1.5 cm. thick. The chamber was dilated and contained 90 ml. of blood. The aortic valve was bicuspid, the anterior inter-coronary commissure being absent. The remaining two commissures were slightly fused, producing a competent but slightly stenotic valve orifice measuring 1.3 cm. by up to 0.6 cm. There was no sub-aortic stenosis and no endocardial fibrosis. The aortic arch and coronary arteries were normal, as were the right heart and pulmonary arteries.

The lungs showed several congenital abnormalities of lobation—an azygos lobe, a fissure separating the apical segment from the remainder of the right lower lobe, and a fissure separating the lingula from the left upper lobe. Both lungs were affected by haemorrhagic citrobacter bronchopneumonia which was more severe on the right. There were small (30 ml.) pleural effusions.

Microscopically the axial abnormal left ventricular bundles showed some patchy pallor and hypertrophy of muscle fibres. Their arrangement was orderly,

however, and not reminiscent of the disorderly arrangement of the septal region of the ventricle in sub-aortic stenosis. The muscle in the sub-aortic region was normal.

The lungs showed some degree of vascular change consistent with the development of mild pulmonary hypertension. The changes were essentially of intimal fibrosis rather than medial hypertrophy in a few of the pulmonary arterioles.

#### DISCUSSION

The normal papillary muscles maintain atrio-ventricular valve competence by contracting during ventricular systole to prevent prolapse of the cusps (Burch, DePasquale, and Phillips, 1968). It appears that this function may be adequately performed by a lone papillary muscle, although in three cases of 'parachute' mitral valve incompetence has been demonstrated by left ventricular angiography (Prado, Levy, and Varco, 1965; Terzaki, Leachman, Khalil Ali, Hallman, and Cooley, 1968). On the other hand, such an abnormal valve must be to some extent stenotic, as its free opening in diastole is prevented. The severity of obstruction is difficult to assess from measurement of the valve orifice at necropsy, as at least part of the impediment to flow is produced by the funnel-shaped mitral valve and chordae tendineae.

The true incidence of this form of congenital mitral stenosis is difficult to assess from the literature. Taussig (1960) describes the congenitally stenotic mitral valve as having thickened fused leaflets with short chordae tendineae, and noted the frequent association with other malformations of the left heart and aorta. More recent reports (Van der Horst and Hastreiter, 1967) confirm this association, but generally lack detailed description of the valve anatomy, so that some cases of this anomaly may have been included. However, unless the whole mitral valve region is grossly distorted and masked by endocardial fibrosis the absence of the antero-lateral papillary muscle should be a sufficiently striking necropsy finding not to be readily overlooked. The fact that only 12 cases are known to us indicates that it is a rare condition.

The clinical features of 11 cases are summarized in Table I.

The presence of congenital mitral stenosis has been frequently unsuspected in life, as the diagnosis may not be indicated by any specific features, and may be masked by the more readily discernible effects of associated lesions.

In common with other forms of congenital mitral stenosis, congestive heart failure and recurrent respiratory infections, often from infancy,

TABLE I  
CLINICAL FEATURES OF CASES OF PARACHUTE MITRAL VALVE

Author	Age (yrs)	Apical Diastolic Murmur	Left Atrial Enlargement	Pulmonary Congestion	Left Atrial Enlargement on E.C.G.	Q.R.S. Axis	Right Ventricular Hypertrophy on E.C.G.	Pulmonary Artery or Right Ventricular Pressure (mm. Hg)	Wedge Pressure (mm. Hg)	Prolonged Opacification of Left Atrium
Swan <i>et al.</i> (1949) ..	18	-	+	+	.	R	.	.	.	.
Schiebler <i>et al.</i> (1961) ..	4	.	+	+	.	N	.	78/23	.	.
Shone <i>et al.</i> (1963) ..	2	+	+	+	-	R	+	56	23/17	+
Carey <i>et al.</i> (1964) ..	4	+	+	+	-	R	+	75	23	.
	13	+	+	+	-	N	+	55	16	.
	22	+	+	+	-	R	+	80	20	.
Anabtawi and Ellison (1965) ..	22	+	+	+	.	R	+	90/50	.	+
Prado <i>et al.</i> (1965) ..	3	+	+	+	+	N	-	30/15	25	.
Terzaki <i>et al.</i> (1968) ..	7	+	+	+	+	R	+	80/45	40/23	.
	5	+	+	+	+	R	+	110/62	23/16	.
Present case ..	11	-	-	-	-	N	-	.	.	.

+ Present. - Absent. . Not known. R, right axis deviation. N, normal axis.

were the prominent features. Our patient had recurrent respiratory infections and died of heart failure and pneumonia, but the necropsy findings indicate that her mitral stenosis was probably not haemodynamically severe, and that she did not have long-standing or gross left atrial or pulmonary hypertension. A rumbling apical diastolic murmur has been relatively inconstant. In two of the cases of Shone *et al.* (1963) the murmur was heard only after resection of an associated coarctation. In other forms of congenital mitral stenosis the murmur was similarly inconstant, being noted in only one of the nine personal cases of Ferencz, Johnson, and Wiglesworth (1954), and in 14 of the 23 cases of Van der Horst and Hastreiter (1967).

Left atrial enlargement was apparent radiologically in all cases except our own and that of Swan, Trapnell, and Denst (1949). Electrocardiographic evidence of left atrial enlargement was reported by Prado *et al.* (1965) and by Terzaki *et al.* (1968). Cardio-angiograms performed by injecting in the pulmonary artery showed delayed emptying of the left atrium in all cases where this procedure was done.

Electrocardiographic, radiological, and catheter studies, where performed, indicate the development of pulmonary congestion and pulmonary hypertension. The case of Prado *et al.* (1965) was an exception in that there was no elevation of pulmonary artery pressure.

The left ventriculogram in the case of Prado *et al.* (1965) was reported as demonstrating 'an unusual configuration, with a filling defect between the apex and the outflow tract'. It is not clear whether this was produced by the anomalous single papillary muscle or by a low sub-aortic stenosis, of the type reported by Gilbert, Aerichide, Bourassa, and David (1966). In both

the cases of Terzaki *et al.* a filling defect visualized along the medial wall of the left ventricle was attributed to the single papillary muscle, and in the second of their cases a filling defect in the shape of an inverted cone was seen in the usual position of the mitral valve. Unless visualization were possible by angiography, the precise diagnosis from other forms of congenital mitral stenosis would not be possible clinically.

Parachute deformity of the mitral valve is rarely a solitary anomaly, being associated with other abnormalities of the left heart or aorta (Table II).

A bicuspid aortic valve, which was to a greater or lesser degree stenotic, was found in half the necropsy cases, including our own. Coarctation of the aorta or sub-aortic stenosis was also present in about half the cases and they frequently occurred together. A supravalar ring was described by Shone *et al.* (1963) in all their cases, and by Anabtawi and Ellison (1965).

This spectrum of anomalies occurs also in association with other forms of congenital mitral stenosis (Van der Horst and Hastreiter, 1967), and with endocardial fibroelastosis (Moller, Lucas, Adams, Anderson, Jorgens, and Edwards, 1964), supravalar stenosis (Cassano, 1964), and the 'hypoplastic left heart syndrome' (Noonan and Nadas, 1958). It may be that reduction in the flow or pressure in the left heart during intra-uterine development secondarily affects the left ventricular outflow tract and aorta. This mechanism has been suggested as being responsible for the development of the 'hypoplastic left heart syndrome', with premature closure of the foramen ovale, and a similar concept was suggested by Folger (1968) to explain lesions associated with supravalar stenosis of the tricuspid valve. If this thesis is relevant it suggests that the mitral valve anomaly is the primary defect.

TABLE II  
PATHOLOGICAL FEATURES OF CASES OF PARACHUTE MITRAL VALVE

Author	Age (yrs)	Sex	Parachute Mitral Valve	Supra-valvar Ring	Ventricular Septal Defect	Sub-aortic Stenosis	Bicuspid Aortic Valve	Coarctation or Hypoplasia of Aorta	Patent Ductus Arteriosus
Swan <i>et al.</i> (1949)	18	F	+	-	-	.	.	+	+
Gould (1960)	4 days	.	+	.	+	.	+	+	+
Schiebler <i>et al.</i> (1961)	4	F	+	-	+	-	-	+	-
Shone <i>et al.</i> (1963)	2	M	+	+	-	+	+	+	-
Carey <i>et al.</i> (1964)	4	M	+	+	-	+	+	+	-
	13	F	+	+	-	+	+	+	-
	22	M	+	+	+	+	+	+	-
Anabtawi and Ellison (1965)	22	M	+	+	-	.	.	+	-
Prado <i>et al.</i> (1965)	3	M	+	.	-	.	.	+	-
Terzaki <i>et al.</i> (1968)	7	M	+	.	-	.	.	+	-
	5	M	+	-	-	-	+	-	-
Present case	11	F	+	-	-	-	+	-	-

+ Present. - Absent. . Not known.

Van Mierop, Alley, Kausel, and Stranahan (1962) have described in detail the development of the atrio-ventricular valves. While the postero-medial half of the mitral valve leaflet develops from the superior and inferior endocardial cushions, the lateral halves of these valve leaflets arise from two left lateral cushions. It is the failure of these two lateral cushions to form valve tissue which produces a parachute deformity. In all the cases described the postero-medial papillary muscle or pair of muscles are present, and give origin to the chordae. The antero-lateral papillary muscle and chordae are missing and replaced by bands of muscle. It is possible that the occasionally observed presence of muscle bundles inserting directly into the mitral valve leaflets are due to partial failure of the endocardial cushions to form valve tissue. The case reported by Aldridge and Wigle (1965) is of interest in that there appear to be two papillary muscle masses shown in their Figure 4, as in a normal mitral valve, but the valve was abnormal in having muscle bands inserting directly into the valve, which was without commissures. Case 1 of Cascos, Rábago, Sokolowski, and Varela de Seijas (1966), although described as a parachute mitral valve, appears from their illustration to have a bifid antero-lateral papillary muscle and a broad mass of muscle in the position of the posterior papillary muscle which inserts directly into the mitral valve. Case 3 of these authors is also similar in having muscles inserting directly into the mitral valve. These cases of Aldridge and Wigle (1965) and of Cascos *et al.* (1966) have not been included in our analysis but may well be transitional stages between the parachute deformity and the normal mitral valve.

Right-sided parachute deformity has been reported in two instances, associated with cor-

rected transposition and with sub-pulmonary stenosis (Schiebler, Edwards, Burchell, DuShane, Ongley, and Wood, 1961; Todd, Anderson, and Edwards, 1965). In such cases the right-sided ventricle is morphologically the left ventricle.

If it is accepted that parachute deformity of the mitral valve is due to failure of development of the left lateral endocardial cushions, it is not surprising that the lesion occurs with other endocardial cushion defects. Gross centrally-sited endocardial cushion defects are, of course, incompatible with the formation of a parachute mitral valve.

Mitral stenosis due to parachute deformity of the valve can hardly be relieved by valvotomy, and successful treatment in the cases surviving mitral valve surgery was by valve replacement (Anabtawi and Ellison, 1965; Prado *et al.*, 1965; and Terzaki *et al.*, 1968). The diagnosis of parachute deformity from other forms of congenital mitral stenosis has been suggested clinically by Prado *et al.* and by Terzaki *et al.* Other forms of congenital mitral stenosis, including a supra-valvar ring, are almost equally unsuitable for closed operation, and surgery for congenital mitral stenosis should therefore, if possible, be undertaken with cardiopulmonary bypass. In young children definitive treatment may therefore be limited to the extracardiac malformations, as in two of the cases of Shone *et al.* (1963). The dangers of partial correction of anomalies were stressed by Swan *et al.* (1949). In the two children of Terzaki *et al.* (1968) the presence of left ventricular enlargement allowed insertion of a prosthetic valve thought to be large enough for adult life.

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