Parachute deformity of the tricuspid valve

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ABSTRACT A parachute deformity of the tricuspid valve occurred in a heart with atrioventricular concordance, double outlet right ventricle, and straddling mitral valve. Although to the best of our knowledge parachute deformity of the tricuspid valve has not previously been reported, in this case its presence was insignificant in relation to the other lesions.

The malformation of an atrioventricular valve in which the tension apparatus springs from a single papillary muscle or muscle group was first described by Swan *et al* (1949). The name "parachute" to describe the anomaly was coined by Schiebler *et al* (1961). Thus far, to the best of our knowledge, parachute deformity has been observed only of the morphologically mitral valve, and its association with other left-sided lesions was so frequent that Shone *et al* (1963) introduced the concept of the "parachute mitral valve complex." We here describe a case of parachute deformity of the morphologically tricuspid valve in a heart further complicated by the presence of a straddling mitral valve and doubled outlet right ventricle.

Case history

A boy, born on 26 April 1968, was the product of a full-term, normal delivery and weighed 2800 g at birth. He failed to thrive, and at the age of 5 weeks was first admitted to hospital, suffering from upper respiratory tract infection and vomiting. At age 10 weeks he weighed only 3700 g. In August 1968 he was admitted to the Hospital for Sick Children at Great Ormond Street for investigation. He was underweight and undersized for his age. There was slight dyspnoea and central cyanosis. All peripheral pulses were palpable. There was evidence of right ventricular hypertrophy, and a systolic thrill and pansystolic murmur down the left sternal edge. There was also a soft apical mid-

diastolic murmur. The child was not in congestive heart failure. Radiological examination of the chest showed an enlarged heart with plethoric lung fields. ECG showed a P-pulmonale with bi-ventricular hypertrophy and left ventricular predominance. Catheterisation and angiography (table a) were interpreted as showing transposition of the great arteries with ventricular septal defect. A Rashkind atrial septostomy was performed. The child improved and was transferred back to his home town hospital. He was reviewed as an outpatient and admitted again in September 1970 when a second cardiac catheterisation was performed (table b). After this it was decided that a pulmonary-systemic-shunt was necessary, and on 22 April 1971 a right Waterston procedure was performed. The child did well until September 1973, when the shunt murmur was no longer audible and he again became cyanosed. He was restudied on 15 February 1974 (table c) and after this he was considered for a Rastelli operation, which was performed on 5 December 1974. On opening the ventricle, the finding was of a large anterior ventricular chamber with straddling of the papillary muscles and the chordae tendineae of the mitral valve of both sides of the rudimentary ventricular septum through an anterior defect. Repair therefore proceeded by constructing a new interventricular septum within the anterior ventricle which returned the straddling part of the mitral valve to the left ventricle and directed left ventricular blood to the aorta. The pulmonary valve was then closed and a conduit placed from the right ventricle to the pulmonary artery. A second, posterior ventricular septal defect unrelated to the straddling valve was identified and closed. After these procedures cardiopulmonary

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Haemodynamic data. (All pressures in mmHg)

Haemodynamic data	a Preoperative data		b Preoperative data		c Preoperative data		Operating room pressures		
	10.9.70		15.2.74		5.12.74		(age: 6 yr 8 mth)		
	(age: 2½ yr)		(age: 5 yr 10 mth)		(age: 5 yr 10 mth)		(age: 5 yr 10 mth)		
	Pre-Rashkind procedure	Post-Rashkind procedure	Dynamic Mean pressure	Dynamic Mean pressure	Dynamic Mean pressure	Dynamic Mean pressure	Dynamic Mean pressure	Dynamic pressure	Mean pressure
	Dynamic Mean pressure	Dynamic Mean pressure							
Right atrial pressure	a = 4 v = -4 x = -1 -2 y = -2	a = 4 v = -1 x = -4 -2 y = 1 2		9	a = 10 v = 8 x = 4 4 y = 2				
Right ventricular pressure	112/-4 - +4	96/0-2	110		110/6		105	35-40 syst	
Pulmonary artery pressure		46//23	14/10 12		8/2 4		18/10		
Central aortic pressure	100/46	96/38 65	100/70 75		100/60 75		110/80	80 syst	
Left ventricular pressure	100/4-8	118/-2 - +2	110		100/60		105	75-80 syst	
Left atrial pressure	a = 14 v = 9 x = 5 y = 4	a = 6 v = 4 x = -4 y = 2	8		a = 6 v = 8 4 x = 0 y = 0		4		13-15

by-pass was terminated satisfactorily, but his cardiac performance deteriorated after the operation. Because of suspicion of cardiac tamponade a re-exploration was carried out but the pericardial sac was dry. His condition continued to deteriorate and despite vasopressor support he died 24 hours later.

Necropsy findings

Only the heart and great arteries were examined. There was situs solitus of the atria, the apex was to the left, and the aortic arch left sided. The venous connections were normal, and atrioventricular concordance was present, the ventricles

being normally positioned. The atrial septum was intact. Two patches had been placed in the ventricles, one closing a posterior defect and the other diverting left ventricular blood to the right-sided anterior aorta. A conduit had been placed from the remainder of the right ventricle to the pulmonary artery. All suture lines were intact. Removal of the patches showed the ventricular morphology. The morphologically right ventricle was a large chamber receiving the tricuspid valve and part of a straddling mitral valve, and giving rise to both great arteries. Two ventricular septal defects were present, a posterior muscular inlet defect (fig 1a) and an anterior, larger, subpulmonary defect (fig 2). The tricuspid valve ex-

Fig 1 (a) illustrates right atrium (RA) and right ventricle (RV). Cusps and chordae of tricuspid valve (TV) converge into a single papillary muscle group (Single PMG). (b) shows a detail of tricuspid valve (TV) taking form of a parachute. An inlet ventricular septal defect (Inlet VSD) is pictured posterior to valve.

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Fig 2 (a) illustrates left atrium (LA) and left ventricle (LV). There is a large anterior ventricular septal defect (Ant VSD) over which mitral valve straddles (Str MV). (b) shows the right ventricle (RV) with the parachute tricuspid valve (TV) and straddling papillary muscles of mitral valve (Str MV) next to it. This ventricle gives rise to aorta (AO) and to pulmonary outflow tract (arrow).

hibited a parachute deformity (fig 1b). There was a single papillary muscle group giving rise to all the shortened and thickened chordae tendineae supporting the valve. The mitral valve opened 80% to the left ventricle and 20% to the right ventricle, straddling the anterior subpulmonary defect. Its anterolateral papillary muscle was displaced into the right ventricle (fig 2a), arising from the anterior aspect of the ventricular septum as a short muscle group (fig 2b). The posteromedial muscle arose from the parietal wall of the left ventricle. The aortic valve was anterior and to the right of the pulmonary valve and was supported by a complete muscular infundibulum (fig 2b). The pulmonary outflow tract originated from the right ventricle posterior to the aorta, and was separated from the aortic valve by the infundibular septum (fig 3). It was grossly narrowed by compression between the tricuspid valve and the straddling part of the mitral valve, the narrowing being exacerbated by a fibrous ring spanning the space between the atrioventricular valves. Via this ring the pulmonary valve was in fibrous continuity with the straddling mitral valve (fig 3).

Discussion

A parachute deformity of an atrioventricular valve, as first described by Swan *et al* (1949) and later correlated to specific associated malformations by Shone *et al* (1963) has always referred, to the best of our knowledge, only to the morphologically mitral valve. Indeed, Shone *et al* (1963) emphasised that the complex of supra-valvar ring of left atrium, subaortic stenosis, and coarctation of the aorta, which they termed the parachute mitral valve complex, was associated with the arterial side of the heart and vascular

Fig 3 Shows the relation of the parachute tricuspid valve (TV), and straddling mitral valve (Str MV) to pulmonary outflow tract (POT), which is "squeezed" between them and is partially bridged by a fibrotic ring (Fibr ring). (Ao-aorta).

system. Schiebler and associates observed a parachute anomaly affecting the right atrioventricular valve in corrected transposition as did El Sayed *et al* (1962), but of course the right atrioventricular valve in corrected transposition is the morphologically mitral valve.

Our case shows that a parachute deformity can equally affect the tricuspid valve, although its presence is likely to be less significant than a similar deformity of the systemic atrioventricular valve. Indeed, in our case the parachute deformity of the tricuspid valve was a bagatelle in terms of the problems facing the surgeon. These were the straddling mitral valve, the severe pulmonary outflow tract obstruction, and the second, posterior inlet ventricular septal defect. Although the anatomy was correctly identified, and although the ventricular patch redirected left ventricular blood to the aorta and restored the straddling portion of the mitral valve to the left ventricle, the patient did not survive. Possibly after the reconstruction the straddling valve blocked egress through the newly constructed left ventricular outflow tract. Since straddling of the mitral valve occurs with some frequency in association with double outlet right ventricle and subpulmonary defect (Kitamura *et al*, 1974), atrial redirection of blood, septation of the right ventricle as described above, and a conduit from the left ventricle to the pulmonary artery may be an alternative procedure to be considered for correcting these cases.

Addendum

Since submitting this paper we have read the report by Ariza *et al* (1979) describing a parachute deformity of the tricuspid valve in association with tetralogy of Fallot.

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