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Incomplete commissure of the aortic valve

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Valvular aortic insufficiency presenting at birth is rare. Causes include congenital malformation at the subaortic, valvular, and supravalvular level. Subaortic deformities resulting in aortic insufficiency include subaortic annular aneurysm[1] and prolapse of aortic cusp or cusps into a ventricular septal defect.[2] Aortic insufficiency due to valvular deformity occurs in bicuspid aortic valves[3] and in fenestration of the aortic cusp or cusps.[4] Supravalvular deformities resulting in aortic regurgitation include congenital aortico-left-ventricular tunnel[5] and congenital aneurysm of the sinus of Valsalva.[6] A further cause of aortic insufficiency is a connective tissue disorder such as Marfan's syndrome[7] or its forme fruste,[8] in which aortic insufficiency follows aortic root dilatation. This report describes another condition causing aortic insufficiency at valvular level—namely, a partial cleft in a bicuspid valve with an incomplete aortic commissure.

Case report

A 14-year-old boy, who had been diagnosed as suffering from congenital aortic stenosis and insufficiency at the age of two weeks, was admitted to the Sheba Medical Centre for evaluation and treatment since clinical examination had indicated the presence of severe aortic valve disease. On examination the peripheral pulses were weak and left ventricular hypertrophy was palpable. Auscultation revealed a systolic click in the aortic area, a grade 4/6 aortic ejection systolic murmur, and a paradoxical splitting of the second heart sound. The electrocardiogram showed left ventricular hypertrophy and strain, and a chest radiograph showed prominence of the left ventricle and ascending aorta. Cardiac catheterisation was performed: the left ventricular pressure was 230/0–20 mm Hg, aortic pressure 85/70 mm Hg, and the transvalvar gradient 145 mm Hg. Supravalvar aortic injection (fig 1) showed aortic regurgitation (grade 2)[9] and poststenotic dilatation of the aortic arch. The coronary arteries appeared normal. Surgery was performed with routine cardiopulmonary bypass techniques, and with hypothermia and cardioplegia. A bicuspid aortic valve was found (fig 2). The right and left coronary cusps were fused. The raphé was 6 mm long, extending from the middle of

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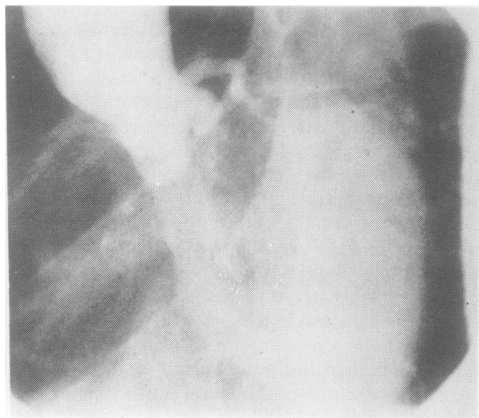

Fig 1 *Left anterior oblique view of diastolic phase in preoperative cineangiogram. Contrast material regurgitates mildly and outlines the outflow tract and the left ventricular cavity (grade 2). There is marked poststenotic dilatation of the ascending aorta.*

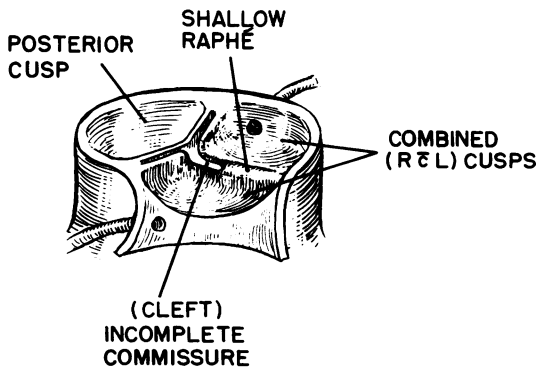

Fig 2 *Schematic illustration of the morphology. The commissures between the posterior cusp and the left and right coronary cusps were stenotic. A 6-mm raphé extended from the aortic wall towards the centre of the valve. The remaining distance was a cleft.*

the cusp towards the aortic wall. A cleft of 4 mm extended from the raphé to the middle of the fused cusp's free edge. The other two commissures were each fused for 2–3 mm at the periphery. Repair consisted of sharp division of the latter two commissures and the cleft was closed, two square sutures of 5–0 suture material being used. This left the patient with a bicuspid but competent valve with greater mobility than before operation.

The postoperative course was uneventful and the patient was discharged on the eighth postoperative day. Recatheterisation performed two weeks after discharge showed a decrease of the aortic pressure gradient to 50 mm Hg. There was no aortic incompetence.

Discussion

The morphological findings in this patient have not to our knowledge been described previously. One commissure was fused partially to a raphé in its outer third and a cleft was present in the middle of the leaflet opposite to it; there was mild fusion of the other two commissures. The morphology explained the clinical finding of severe aortic stenosis and insufficiency. Reuniting the edges of the cleft with 5–0 sutures (thus shortening the free edge of the malformed cusp) resulted in a competent valve owing to equalisation of the free edges of the two cusps. Commissurotomies could now be performed aggressively into the aortic wall, allowing the effective valve orifice to enlarge.

Commissurotomy alone, leaving the cleft untouched, could have been performed; but we believe that this would have resulted in diastolic prolapse of the malformed cusp into the left ventricle, causing a greater volume of blood to regurgitate than before. Closure of the cleft produced a bicuspid valve but was the key to the prevention of severe postoperative aortic incompetence. The presence of a cleft in an aortic valve at the central end of a well-developed raphé is interesting from a developmental point of view. It proves, we believe, the theory that the raphé actually represents a commissure that failed to develop.[3]

Lastly, we believe that, despite the fact that a prosthetic valve could have been inserted, it was preferable to leave the patient with his own tissues and a valvular gradient of 50 mm Hg. The exposure of a 14-year-old boy to long-term anticoagulation and the risk of prosthetic valvular complications carries a greater danger than leaving him with a malformed valve and a transvalvar gradient of 50 mm Hg. We are, however, aware that the natural history of such a valve makes gradual deterioration and eventual valve replacement likely. But we hope that this will be at an advanced stage in the patient's life.

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