Severe heart failure in child with ventricular septal defect and acute tricuspid regurgitation

J R PEPPER, M C JOSEPH, P B DEVERALL

From the Departments of Paediatric Cardiology and Cardiothoracic Surgery, Guy's Hospital, London

SUMMARY A case is reported in which acute tricuspid regurgitation developed in a child with a ventricular septal defect resulting in a left ventricular-right atrial shunt. This was successfully treated by closure of the defect and tricuspid valve replacement. The anterior leaflet of the tricuspid valve was almost completely destroyed by endocarditis, though in previous reports of tricuspid valve endocarditis in association with ventricular septal defect in children, it has been invariably the septal leaflet which is damaged.

Case report

A 2-year-old Libyan boy was referred for investigation because of shortness of breath and cyanosis. On examination he was small for his age and had mild central cyanosis and clubbing. He was in sinus rhythm with a raised jugular venous pulse and a marked V wave. The liver was enlarged 4 cm below the subcostal margin and was pulsatile. On auscultation there was a grade 4/6 systolic murmur accompanied by a thrill at the left sternal edge which increased in intensity on inspiration. There was a short grade 2/6 mid-diastolic murmur medial to the apex beat.

The chest radiograph showed moderate cardiac enlargement and plethoric lung fields. The electrocardiogram showed sinus rhythm, axis +120, right atrial hypertrophy, and right ventricular hypertrophy. Blood cultures were sterile. An echocardiogram showed abnormal tricuspid valve motion with a high amplitude vibration in the outflow tract of the right ventricle throughout the cardiac cycle (Fig. 1). The timing of tricuspid and mitral valve closure was normal. Cardiac catheterisation showed a bidirectional shunt at atrial level and a 5-mm gradient across the tricuspid valve. Angiography disclosed gross tricuspid regurgitation, the valve appearing completely flail. A ventricular septal defect was also seen. The tricuspid valve was attached normally at the atroventricular ring. There was a large end-systolic volume in the right ventricle.

At operation there was obvious tricuspid regurgitation caused by almost total destruction of the anterior leaflet of the tricuspid valve. There were old vegetations on the small area of valve remaining, suggesting healed endocarditis (Fig. 2). There was a perimembranous ventricular septal defect and a persistent foramen ovale. The ventricular septal defect was closed by direct suture and a 27-mm Carpentier-Edwards porcine xenograft inserted in the tricuspid position. The persistent foramen ovale was closed by direct suture. The heart came off bypass without difficulty.

The patient made a satisfactory recovery and eight months later appeared to be extremely well, with no evidence of failure or of tricuspid regurgitation.

Discussion

Endocarditis of the tricuspid valve in association with a left-to-right shunt through a ventricular septal defect is uncommon, but has been described. Inspection of the tricuspid valve in these cases reveals damage to the septal leaflet. The unusual feature of this case was the damaged anterior leaflet resulting in its almost complete destruction. It has been shown experimentally by Wait and Mustard that the septal leaflet of the tricuspid valve acts as a baffle against which the anterior and posterior leaflets synchronously close. Hence it is not surprising that in this case gross tricuspid regurgitation occurred.
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![Acute tricuspid regurgitation](image)

Fig. 1 Preoperative echocardiogram. Irregular valve motion is seen in the right ventricular outflow tract (RVOT) throughout the cardiac cycle. IVS, interventricular septum; LV, left ventricle.

![Tricuspid valve](image)

Fig. 2 Tricuspid valve seen through right atriotomy at operation. Note encrusted vegetation (V) arising from area of ventricular septal defect and remnants of anterior leaflet (AL) of tricuspid valve. SL (septal leaflet). Posterior leaflet not seen.

Left ventricular to right atrial communication via the tricuspid valve is well described. The ventricular septal defect opens into the right ventricle behind the septal leaflet of the tricuspid valve. Whereas there is usually a second deficiency in the septal leaflet, in this case a destroyed anterior leaflet gave rise to a clinical picture dominated by acute tricuspid regurgitation, rather than a left-to-right shunt. In fact, the cardiac catheterisation disclosed a left-to-right shunt through the ventricular septal defect and a right-to-left shunt through the persistent foramen ovale. This bidirectional shunting together with the gross tricuspid regurgitation probably accounted for the mild degree of central cyanosis.

Echocardiography is being used increasingly in the study of tricuspid valve disease. It has been suggested that systolic flutter of the tricuspid valve caused by the passage of left ventricular blood into the right atrium is of value in the recognition of congenital left ventricular–right atrial communication. In this case the near total destruction of the anterior leaflet and a consequently unsupported posterior leaflet resulted in a valve which was completely flail, and abnormal tricuspid valve motion was observed throughout the cardiac cycle.

We thank Dr M Tyanan who performed the cardiac catheterisation.

References


Requests for reprints to J R Pepper, Esq, FRCS, Department of Cardiothoracic Surgery, St Thomas' Hospital, Lambeth Palace Road, London SE11.