Congenital Tricuspid Incompetence

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Rare congenital malformations of the tricuspid valve or its attachment producing tricuspid insufficiency have been reported by a number of authors (Hotz, 1923; Ariel, 1930; Abbott, 1936; Dubin and Hollinshead, 1944; Palladino and Kinney, 1948; Barritt and Urich, 1956; Kincaid *et al.*, 1962; Reisman *et al.*, 1965; Jordan and Taylor, 1966). The purpose of the present communication is to report a case in which congenital tricuspid incompetence was caused by absence of the chordae tendineae and papillary muscles of the posterior valve leaflet. To our knowledge, this is the first time this type of malformation has been described.

Case Report

A 3-year-old Nigerian boy was referred to the paediatric cardiac clinic, University College Hospital, Ibadan, on account of breathlessness, cough, tiredness, and facial

and abdominal swelling for one month. The previous medical history was non-contributory.

Physical examination revealed a well-nourished child weighing 11.5 kg. He had tachypnoea. There was neither cyanosis nor clubbing of the fingers. The eyelids were puffy. The lung fields were clear. The pulse was regular and of small volume. Pulse rate was 120 a minute. Blood pressure measured 90/60 mm. Hg. The jugular venous pressure was raised and there was systolic expansion of the vein. The praecordium was quiet; apex beat was in the 5th left intercostal space, 6 cm. from the midline. There were no thrills. A grade 3 rough pansystolic murmur was present at the left and right lower sternal borders. Diastole was clear. There was a gallop rhythm. Minimal oedema was present in the legs. There was an enlarged, non-tender, pulsatile liver 8 cm. below the right costal margin. The spleen was not palpable. There was no ascites.

A chest x-ray (Fig. 1) showed an enlarged heart and normal vascularity. The electrocardiogram revealed sinus rhythm, mean QRS frontal axis, $+165^\circ$, wide P waves in lead II, T inversion in chest lead V5, and flat in

FIG. 1.—Chest x-ray showing cardiac enlargement and normal vascularity.

lead V6. These features were interpreted to indicate right atrial dilatation, right ventricular hypertrophy, and non-specific T changes in the left chest leads.

Laboratory investigations included haematocrit, 36 per cent; white blood cells 5800/cu. mm; Hb genotype AA; normal liver function tests; total serum protein, 7.2 g./100 ml. (albumin, 3.2 g.; globulin 4.0 g.).

The clinical diagnosis was congestive cardiac failure due either to right-sided endomyocardial fibrosis or to idiopathic cardiomegaly.

Treatment consisted of digoxin, hydroflumethiazide, potassium chloride, and restricted salt intake.

Progress. Seven months after the initial visit, the child developed massive ascites which caused further respiratory embarrassment. A paracentesis abdominis yielded 1.2 litres of clear, straw-coloured fluid, with a total protein content of 5.9 g./100 ml. (albumin 3.8 g., globulin 2.1 g.). His general condition continued to deteriorate, and two months later, in spite of digoxin, diuretics, and restricted salt intake, there was rapid reaccumulation of ascitic fluid, the colour and total protein content of which were essentially the same as previously noted. Cardiac catheterization and angiocardiography were attempted, but during the procedure the child developed fatal cardiac arrhythmia.

Necropsy. The heart weighed 130 g. The epicardial surface was normal. The right atrium was very dilated. The tricuspid valve with normal basal attachment (Fig. 2) measured 5.5 cm. There were three valve leaflets; the anterior and septal leaflets with their attachments were normal. The posterior leaflet was semilunar in

shape; its free margin was folded backwards and attached directly to the endocardial lining of the right ventricle immediately below the valve ring. The chordae tendineae and papillary muscles were absent. The right ventricular cavity was grossly dilated, without evidence of fibrosis. The pulmonary valve, the left atrium, the mitral valve, and the left ventricle were normal.

The liver showed evidence of chronic venous congestion. Histological examination revealed dilatation of the centrilobular hepatic sinusoids and moderate increase in fibrous tissue in the portal tract, with some early incursion into the parenchyma.

Comments

Congenital tricuspid insufficiency due to an anomaly of the tricuspid valve or its attachments is a rare lesion. Thirteen such cases have been previously reported (Table). The tricuspid valve

TABLE
CONGENITAL TRICUSPID INCOMPETENCE

Author	Year of publication	No. of cases
Abbott	1936	1
Ariel	1930	1
Barritt and Urich	1956	1
Dubin and Hollinshead	1944	1
Hotz	1923	2
Jordan and Taylor	1966	1
Kincaid <i>et al.</i>	1962	2
Palladino and Kinney	1948	2
Reisman <i>et al.</i>	1965	2

FIG. 2.—Semilunar-shaped posterior leaflet of the tricuspid valve, attached directly to the endocardial lining of the right ventricle. Note absence of chordae tendineae and papillary muscles.

malformations in all these cases consisted of abnormally short chordae tendineae, rudimentary papillary muscles, nodularity, and thickening of the leaflets, and in some cases fusion of two of the leaflets. The present case, however, differs from those previously described in that both chordae tendineae and papillary muscles of the anomalous posterior valve leaflets were absent. The free end of the posterior valve leaflet was attached directly to the endocardial lining of the right ventricle. This case further differs from those previously reported in the absence of associated cardiac malformations.

The clinical features of tricuspid incompetence, enlarged pulsatile liver, massive ascites with high protein content, and little peripheral oedema in the present case, suggested the possibility of right ventricular endomyocardial fibrosis, an acquired heart disease of unknown aetiology, which is common in Nigeria. Without evidence of a cardiac lesion dating back to birth, an accurate clinical differentiation between the two conditions was virtually impossible. All the clinical diagnostic features of right ventricular endomyocardial fibrosis were present in this particular case.

Summary

A case of tricuspid incompetence due to absence of

chordae tendineae and papillary muscles is reported. We have found no previous reports of this type of tricuspid valve malformation.

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