CASE REPORTS

Double Outlet Right Ventricle with Long Survival

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The entity of double outlet right ventricle is being increasingly recognized in recent years (Neufeld, DuShane, and Edwards, 1961a; Neufeld *et al.*, 1961b, 1962; Morgan *et al.*, 1962; Mehrizi, 1965; Venables and Campbell, 1966). The oldest patient in the series reported by Mehrizi (1965) was 22 years and in the 10 patients of Venables and Campbell (1966) the oldest was 9 years. We report a patient of 27 years, with necropsy details.

Case Report

A woman of 27 years was admitted in December 1966, and died 6 days later. She had cyanosis from the age of 6 months and had repeated syncopal attacks up to the age of 6 years. She had a history of squatting, progressively increasing effort intolerance, and recurrent bouts of haemoptysis during the last few years. There was no history of prolonged fever. Her physical development was poor.

Examination showed stunted growth, poor general condition, central cyanosis, clubbing of fingers, and prominent "a" waves in the neck veins. All peripheral arterial pulsations were normally felt and blood pressure was 135/95 mm. Hg. There was no evidence of congestive cardiac failure. The apex beat was in the left 5th intercostal space just outside the midclavicular line. There was a moderate left parasternal heave. The first heart sound was normal and the second showed close splitting with a normal pulmonary component. There was a grade 3/6 ejection systolic murmur and a short early diastolic murmur in the left 2nd and 3rd intercostal spaces. The spleen was not palpable, and the other systems were essentially normal.

X-ray film of the chest revealed pulmonary oligoemia, small main pulmonary artery segment, and right atrial enlargement (Fig. 1). Cardiothoracic ratio was slightly increased. The electrocardiogram (Fig. 2) showed isorhythmic AV dissociation, mean frontal axis of $+105^\circ$, right ventricular hypertrophy, and a prominent S wave persisting in V6.

A clinical diagnosis of transposition of the great vessels

with pulmonary stenosis was made. No haemodynamic studies could be carried out because of the poor condition of the patient. She died on the sixth day in hospital.

At necropsy, heart weighed 210 g. Both aorta and pulmonary trunk arose from the right ventricle (Fig 3). There was a large ventricular septal defect of 1.5 cm. in diameter in the membranous portion of the septum. The aorta was situated to the right of the pulmonary trunk. The anterior leaflet of the aortic valve had prolapsed through the septal defect. The pulmonary valve cusps were partially fused resulting in stenosis with an opening of 4 mm., which showed healed, firm vegetations. The right ventricular wall was 18 mm. in thickness. The

FIG. 1.—Skiagram of the chest.

FIG. 2.—12-lead electrocardiogram and rhythm strip lead III showing isorhythmic AV dissociation.

myocardium showed a moderate degree of diffuse fibrosis. The kidneys had areas of small infarcts and focal pyelonephritis. Other organs did not reveal any significant abnormality.

Discussion

The present case is perhaps the oldest patient on record of double outlet right ventricle. According to the classification of Mehrizi (1965) this patient belongs in group 1a. An additional feature was the prolapse of the anterior leaflet of the aortic valve through the ventricular septal defect, which explained the early diastolic murmur in the left parasternal region. This association in double outlet right ventricle has also been described (Mehrizi, 1965). Other features were the evidence of subacute bacterial endocarditis and isorhythmic AV dissociation in the electrocardiogram.

Summary

A patient with double outlet right ventricle with survival up to 27 years is reported. She also had valvular pulmonary stenosis, prolapse of the pulmonary leaflet of the aortic valve through a ventricular septal defect, evidence of subacute bacterial endocarditis, and isorhythmic AV dissociation.

FIG. 3.—Necropsy specimen of heart showing the right ventricle with both aorta and pulmonary artery arising from it. AO, aorta; PA, pulmonary artery; RA, right atrium; RV, right ventricle; LV, left ventricle; VSD, ventricular septal defect.

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