COR TRILOCULARE-BIVENTRICULARE WITH HYPOPLASIA OF THE TRICUSPID VALVE: SURVIVAL TO THE AGE OF 65 YEARS

BY

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Cor triloculare-biventriculare is a rare congenital malformation of the heart in which there is virtually a complete absence of the atrial septum.

Abbott (1936) in her analysis of a thousand cases of congenital heart disease found only 5 patients with this malformation, the average age at death being 6 years and the maximum age of survival 31 years. Brown (1950) gives several references to isolated case reports but few of these cases survived beyond middle age. Cunningham (1948) reported a man who lived to an age of 51 years with complete absence of the atrial septum together with a communication between the mitral and tricuspid valves, absence of the membranous part of the ventricular septum, and aneurysmal dilatation of the pulmonary arteries. Ellis *et al.* (1959) described 5 patients whose ages ranged from 3 years to 31 years, all of whom were submitted to operation. In a brief survey of the published material they found only 5 previous case reports and mention 3 other possible cases. These authors classified their cases into "partial" and "complete" defects. In the "partial" form the septal cusp of the mitral valve is usually cleft, the ventricular septum is intact, and the atrio-ventricular valves are separated by fused valvular tissue. In the rarer "complete" form both the mitral and tricuspid valves are cleft and communicate, and the lower edge of the defect is made up of ventricular septum which is itself in part deficient.

Dubost and Blondeau (1960) reported two patients aged 28 and 14 years, and Shaher and Johnson (1963) investigated 3 others with common atrium, one of whom was 30 years old. Giknis (1963) pointed out that 7 of 11 patients with the Ellis van-Creveland syndrome were found at necropsy to have a single atrium. In this rare combination of chondral and ectodermal dysplasia, polydactyly, and congenital heart disease, multiple cardiac malformations are common and most patients die in infancy.

The patient described here survived to the age of 65 years, and it seems likely that her longevity was due, at least in part, to hypoplasia of the tricuspid valve.

Case Report

The patient, a married woman without children, was aged 65 years at the time of her final admission to the Royal South Hants Hospital in 1963. As a child she had been told that she had valvular disease of the heart. She was first seen in 1954 as an out patient with pains across the shoulders thought to be due to mild arthritis. She was noted to be hypertensive (200/120 mm. Hg) and to have gallop rhythm. Her symptoms rapidly cleared. She next attended in 1959 when she reported short-lived attacks of palpitation and stated that in 1955 she had had a transient weakness of her left arm and leg which had recovered in about two months. Her blood pressure at this second attendance was 230/110 mm. Hg and the optic fundi showed Grade 2 hypertensive changes. The heart was in sinus rhythm and slightly enlarged. A murmur, which was thought to be presystolic, was heard just medial to the apex and the first sound was loud. The electrocardiogram showed left axis deviation and right bundle-branch block. A diagnosis of hypertension and

FIG. 1.—Chest radiograph taken six weeks before death showing enlargement of the heart and only slight pulmonary vascular engorgement.

FIG. 2.—Electrocardiogram showing left axis deviation and the pattern of right bundle-branch block.

mitral valve disease was made, and treatment was commenced with weight reduction and hypotensive therapy. In the next two years the patient had three attacks of undiagnosed pain in the left side of the chest during which her cardiogram and chest radiograph did not change. She also had a further attack of short-lived paresis, this time in her right leg which recovered quickly. She was moderately short of breath.

In 1962 she developed angina pectoris and was admitted to hospital with atrial fibrillation. Sinus rhythm was restored with quinidine but 10 months later atrial fibrillation recurred and persisted, ankle oedema developed, and she became increasingly short of breath. Treatment with digitoxin and diuretics was begun.

In August 1963 the patient was again admitted with a recurrence of angina pectoris for five days. She was in severe cardiac failure with orthopnoea and engorged neck veins but was not cyanosed. Atrial fibrillation was still present with an apex rate of 120 beats a minute. The apex was palpable in the anterior axillary line with a moderate left ventricular heave, and a mid-diastolic murmur could be heard at the apex.

FIG. 3.—View of heart from above with common atrium opened to show the hypoplastic tricuspid orifice, absence of the atrial septum, and the normal mitral valve. TV=tricuspid valve; CS=coronary sinus; MV=mitral valve.

FIG. 4.—Heart viewed from the front with the underdeveloped right ventricle opened to show the bicuspid tricuspid valve. A=atrium; TV=tricuspid valve; PV=pulmonary valve; RV=right ventricle.

No opening snap was heard and no abnormality of the second sound in the pulmonary area was noted. Chest radiograph showed a large heart with only slight pulmonary vascular engorgement (Fig. 1). The electrocardiogram showed left axis deviation (mean frontal QRS vector— 45°) with right bundle-branch block (Fig. 2).

Improvement was rapid with bed-rest, digitoxin, and diuretics, and she was discharged home. Three weeks later the patient was readmitted 20 hours after the sudden development of severe pain and numbness in both legs. There had been some subsequent subjective improvement. On examination there were signs of right-sided cardiac failure and atrial fibrillation. The legs were cold and the pulses were absent over the femoral arteries and below. A diagnosis of saddle embolus at the aortic bifurcation was made, and in view of the time interval, the subjective improvement, and the patient's poor general condition it was decided to treat her conservatively. Cardiac failure persisted, there was little improvement in her legs, and she died suddenly three weeks later.

Necropsy. The heart weighed 420 g. A large common atrium was present, the right side together with the right atrial appendage being considerably larger than the left side. The atrial septum was completely absent apart from a low ridge between the atrio-ventricular valves (Fig. 3). The coronary sinus was large and opened on to the posterior part of the ridge but the valve of the coronary sinus was absent. The mitral orifice was of normal size (10 cm. circumference) and had two normal cusps. The tricuspid orifice was much smaller than normal with a circumference of 7 cm. (normal 12–13 cm.). The tricuspid valve had only two cusps, one anterior and one posterior in position, of approximately equal size and attached normally to the valve ring (Fig. 4). Anterior to the anterior cusp was an area of endocardial sclerosis into which two chordæ tendineæ were inserted and this was continuous with a small knob of tissue at the valve ring adjacent to the septal end of the anterior cusp into which a further chorda was inserted. The right ventricle was small in volume and the inflow tract was greatly encroached upon in its lower part by a large septomarginal trabecula. There was only one papillary muscle which arose from the wall of the ventricular septum and from which all the chordæ tendineæ arose. The outflow tract had a wall 5 mm. thick and the supraventricular crest was not well marked. The pulmonary valve was 7 cm. in circumference with normal cusps. The whole right ventricle appeared slightly underdeveloped.

The left ventricle was hypertrophied, the thickness of its free wall measuring 17 mm. Two normal

papillary muscles were present and the aortic valve was normal with a circumference of 8 cm. There was a tiny pin-hole defect in the ventricular septum at the junction of the membranous and muscular parts, which was just patent to a probe but which obviously had no functional significance. There was moderate atheroma of the aorta in its upper part and of the coronary arteries. The right main coronary artery was very narrowed but not completely occluded. The myocardium and pericardium appeared macroscopically normal. The aorta was blocked below the origin of the renal arteries by ante-mortem thrombus. No signs of mural thrombosis could be found in any of the chambers of the heart but the right femoral and iliac veins were obstructed by firm clot and it was thought likely that the aortic block was the result of a paradoxical embolus from the leg veins. Examination of the lungs showed no significant abnormality and the venous connexions of the heart appeared normal.

Discussion

Dexter (1956) showed that an atrial septal defect of more than 2 sq. cm. cross-sectional area resulted in diminution or abolition of the normal atrial pressure difference between the two atria thus producing a physiologically common atrium. Shaher and Johnson (1963) found that the haemodynamics of common atrium are similar to those of a large atrial septal defect. When a common atrium is uncomplicated by any other cardiac anomaly, the right side of the atrium and the right ventricle normally dilate and hypertrophy due to the increased blood flow through the right side of the heart. The pulmonary arteries are usually large and the lung fields are hyperaemic due to the increased pulmonary blood flow. According to Ellis *et al.* (1959) the cardiogram usually shows a variation of the rsR1 type of complex in lead V1, and P wave changes indicating atrial enlargement may occur. The mean QRS axes in their patients ranged from -60° to -150° . In the patient reported here there was only slight pulmonary vascular engorgement on the radiograph and the pulmonary arteries were not large. The electrocardiogram showed left axis deviation with right bundle-branch block. At necropsy hypoplasia of the tricuspid valve with a small right ventricle was found together with hypertrophy of the left ventricle, which was probably a result of the systemic hypertension.

Isolated congenital stenosis of the tricuspid valve has been reported infrequently. It is usually due to adherence of the valve cusps near their bases thus narrowing the orifice of the valve, though the valve ring is of normal size. Medd *et al.* (1961) reported two infants with congenital tricuspid hypoplasia and hypoplasia of the right ventricle where the valve ring was small but the cusps were normal and three in number. According to Lev (1953) this is the normal finding when there is tricuspid stenosis with hypoplasia of the right ventricle and is always associated with a defect of the atrial septum.

In the present patient the tricuspid valve had only two cusps which were not adherent and the valve ring was small. Although there was sclerosis of the ventricular endocardium adjacent to the septal end of the valve ring with some chordae tendineae inserted into it, there was no evidence that this represented an atretic septal cusp adherent to the ventricular wall. Lev (1953) stated that the tricuspid valve may have only two leaflets, a distinct medial leaflet being absent, when the angle between the entry and exit of the right ventricle is more acute than normal.

It is well known that patients with large atrial septal defects may survive to old age, and Rodstein, Zeman, and Gerber (1961) have reported a man with a defect 2 cm. in diameter who lived to an age of 84 years. In view of the fact that a large atrial septal defect gives a physiological situation similar to a common atrium it seems likely that the poor survival in the published cases of cor triloculare biventriculare is due largely to the associated cardiac anomalies that most of these cases have.

The patient reported here was never clinically cyanosed so that there could not have been a large right-to-left shunt at atrial level. On the other hand the pulmonary arteries were not enlarged and the right ventricle was small so that it is unlikely that there was ever a large shunt from left to right. A left-to-right shunt will occur in a patient with an absent or deficient atrial septum if the diastolic filling resistance of the right ventricle is lower than that of the left ventricle. This is a common situation and leads to increased work and output by the right ventricle with pulmonary artery enlargement and pulmonary vascular engorgement.

In the present case of common atrium it seems probable that the coexistence of tricuspid hypoplasia produced a compensatory effect, so that the diastolic filling resistance of the right ventricle remained similar to that of the left ventricle and gross unidirectional shunting was thus avoided. The patient's longevity may be attributed, at least in part, to the compensatory effect of her tricuspid hypoplasia. The eventual failure of the left ventricle, secondary to hypertension, probably upset this balanced situation and led to her death at the age of 65 years.

Summary

A woman aged 65 years who was found at necropsy to have a common atrium with hypoplasia of the tricuspid valve is described. Relevance of the latter to the unusual longevity is discussed.

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