3. A CASE OF CONGENITAL MITRAL ATRESIA WITHOUT AORTIC VALVE ANOMALY

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Mitral atresia is a rare form of congenital heart disease. It is invariably present with other cardiac anomalies, and many combinations have been recorded in the literature. This case is presented not only because of the rarity of mitral atresia, but also because of the unusual combination of associated cardiac defects. In this case of mitral atresia, the aortic valve was normal, the foramen ovale and ductus arteriosus were patent, there was a ventricular septal defect in the muscular portion of the septum, and the great vessels were normal.

Case Report

A three month old baby was admitted on 28/9/65 with cough, fever, and difficulty in breathing. An accurate history was not obtained, as the mother was mute. On examination, the child was moribund, with respiratory distress and crepitations over both lung fields. Tachycardia (pulse 160) was present and a systolic and a diastolic murmur were heard over the mitral area. Apart from a palpable liver, other systems were normal. No note was made of cyanosis. The child was treated with digitalis, bronchial dilators, antiobiotics, steroids and analeptics, but died three hours later. A clinical diagnosis of congenital heart disease and bilateral bronchopneumonia was made.

Autopsy findings

The body was that of a thin undernourished baby girl (weight 3,800G). The pertinent post mortem findings were confined to the lungs and heart. The spleen was present; no accessory spleens were found and no extracardiac malformations were detected. The lungs showed slight congestion only.

The heart was enlarged (80G, normal 23-27G—Coppoletta and Wolbach, 1933) and the right side bulged. The venous return of both systemic and pulmonary circulations was normal and no extracardiac arteriovenous shunts were demonstrable. The coronary veins drained normally into a normal coronary sinus. The great vessels were all present and retained their correct anatomical relationships. There was a patent foramen ovale, 5mm in its longest diameter (Fig 4). The right atrium was dilated and hypertrophied and communicated with its ventricle through a normal tricuspid valve. The ventricle on the right was hypertrophied (thickness at outer border 9 mm, thickness at outflow 8 mm), and it bulged into a small hypoplastic left ventricle; the papillary muscles on the right were also greatly hypertrophied (Fig 3). There was a moderately large ventricular septal defect in the muscular part of the septum (Figs 2 & 3). A moderate-sized pulmonary artery led from the right ventricle through a normal tricuspid pulmonary valve (circumference 2.8 cm). The pulmonary artery communicated with the aorta through a patent ductus arteriosus of small calibre (Fig 1); the coronary arteries arose from their normal positions, were patent and of normal distribution, and a normal sized aorta arose from the left ventricle through a normal tricuspid aortic valve (circumference 2.0 cm). The left atrium was small as compared to the right. No communication existed between this atrium and its ventricle, the mitral valve being represented by shallow grooves on the floor of the atrium (Fig 4). (The weight and cardiac measurements were obtained after 72 hours fixation).

Histology

The main histological changes were confined to the heart and lungs.

The site of the mitral atresia was represented by thickened endocardium without any actual fibrosis.

In the lungs there was capillary congestion throughout. Protein exudate, red cells, mononuclear cells, many of which contained haemosiderin granules, were present in the alveoli in various combinations. The lung changes were thus those of chronic venous congestion. No evidence of pulmonary hypertension was present.

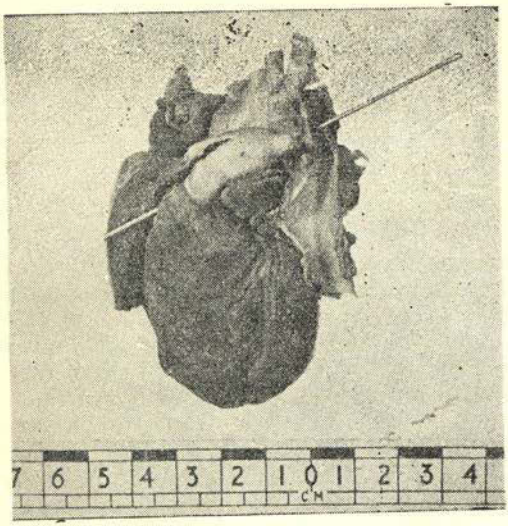

Fig. 1. *General view of the heart from the front. The probe in the right ventricle passes into the pulmonary artery, then through the patent ductus arteriosus and into the aorta. Part of the hypertrophied wall of the right ventricle is visible on the left and the normal anatomical relationship of the great vessels is also seen.*

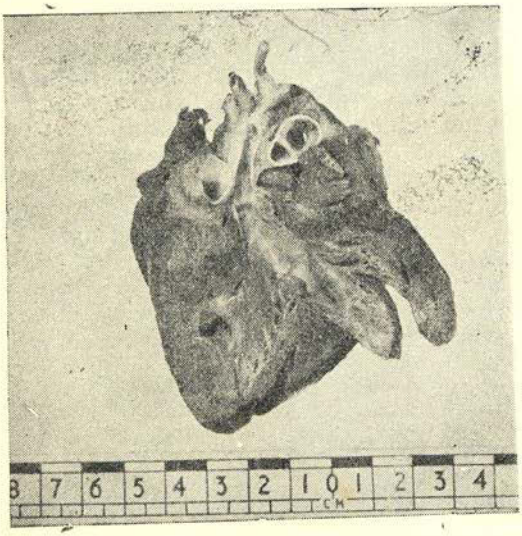

Fig. 2. *View of left side of heart with left ventricle opened. The thin musculature of the left ventricle is seen, as well as the ventricular septal defect. The aortic valve, coronary ostia, and the aorta curving upwards, can also be seen. Careful examination of the transected pulmonary artery reveals the left and right branches and the patent ductus arteriosus (top right of picture).*

Fig. 3. *View of right ventricle. The probe in the chamber of the right ventricle enters the defect in the muscular part of the septum. The hypertrophied myocardium (bottom right, between the thumb and finger) and papillary muscles are also seen.*

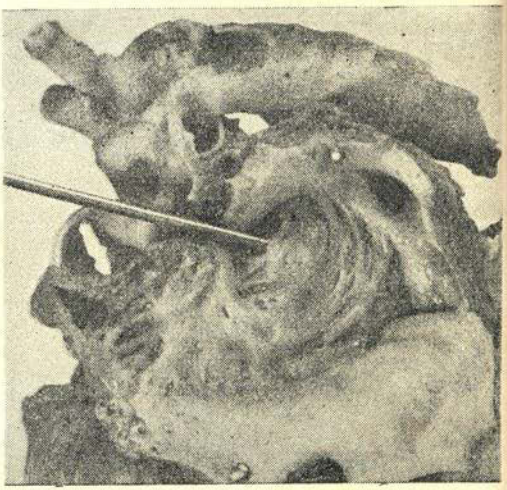

Fig. 4. *View of the left atrium. The floor of the left atrium shows no communication with the left ventricle (mitral atresia). The white, thickened, and grooved endocardium is seen. The probe enters the patent foramen ovale.*

Discussion

Many authors have described their own experiences and reviewed cases collected from the literature (Eliot *et al.*, 1965; Redo *et al.*, 1961; Watson *et al.*, 1960). They are all agreed on the rarity of mitral atresia. That

mitral atresia very rarely, if ever, occurs as an isolated lesion is well recognised (Edwards, 1953), and its combination with a variety of both cardiac and extracardiac anomalies has been described (Eliot *et al.*, 1965; Large, 1960; Shone and Edwards, 1964; Redo *et al.*, 1961). In all cases of mitral atresia there is right sided cardiac hyperplasia, and, except in rare instances, the so-called 'hypoplastic left heart syndrome' (Lumb & Dawkins, 1960). For survival in the immediate neonatal period, there must be a communication between left and right atria; this communication, in a majority of cases, takes the form of a patent foramen ovale, or sometimes an atrial septal defect, but rarely a common atrium may exist (Eliot *et al.*, 1965; Watson *et. al.*, 1960). One author described a case of mitral atresia, in which the interatrial septum was intact but the left atrium was connected with the left innominate vein by a laevoatriocardinal vein, thus establishing an indirect link (Edwards & Dushane, 1950). Ventricular septal defects occur frequently with mitral atresia, but usually the defect is in the membranous part of the septum and only occasionally in the muscular portion (Edwards, 1953). A case in which several defects were found in the muscular part has been described (Edwards and Dushane, 1950), but in our case only a single moderately large defect existed. Without a ventricular septal defect the left ventricle does not take part in the circulation, and its right counterpart acts as a single ventricle; indeed, sometimes the left ventricle may be entirely absent or only rudimentary. Probably the commonest lesion found in association with mitral atresia is aortic atresia (Eliot *et al.*, 1965). When both the mitral and aortic valves are atretic, there is also left sided hypoplasia, and usually no ventricular septal defect. An anomaly of the aorta, such as coarctation, dextroposition, or hypoplasia, or of the aortic valve, such as hypoplasia or stenosis is also a common finding (Brockman, 1950; Edwards and Dushane, 1950; Watson *et al.*, 1960). When the aortic valve is patent, there is invariably a ventricular septal defect, and the left ventricle participates in the circulation. In our case the aorta, in its whole length, and the aortic valve, were normal, and the left ventricle was hypoplastic. Because of the frequent occurrence of aortic hypoplasia and coarctation there is often a widely patent ductus arteriosus associated with mitral atresia (Lam *et al.*, 1953; Lumb & Dawkins, 1960; Redo *et al.*, 1961); in our case the ductus, although patent, was of very small calibre. Pulmonary valvular atresia or stenosis in conjunction with mitral atresia has been described (Lam *et al.*, 1953), but a hyperplastic pulmonary artery is the usual finding. In our case the pulmonary valve and artery were normal. Abnormalities in the anatomical relationships of the great vessels, anomalous venous returns to the heart, and extracardiac defects, none of which was present in our case, have also been described (Brockman, 1950; Shone and Edwards, 1964). One group (Watson *et al.*, 1960) mentioned the relatively frequent occurrence of asplenia or multiple spleens, especially when there was an anomaly in the cardiac venous return.

Cases of mitral atresia rarely survive the first year. In a review of 52 cases (Watson *et al.*, 1960) the average survival was found to be six months, death occurring anywhere from shortly after delivery to the maximal age of five years ten months. However, in anotherreview of 19 cases (Redo *et al.*, 1961) three cases, the eldest being seventeen, survived over ten years. That survival is related to the combination of cardiac lesions has been pointed out by Redo *et al.*, (1961), who noted that the longest survival was anticipated in those cases of mitral atresia in which there was mild pulmonary stenosis, an adequately patent foramen ovale or atrial septal defect, and an obliterated ductus arteriosus; for, with this combination, pulmonary vascular obstruction, the factor which precipitates congestive cardiac failure, was lessened. We believe that in our case, although there was no pulmonary stenosis, the blood flow through the lungs was not excessive, that the unusual combination of defects was largely compensatory, but we are unable to explain the mechanism responsible for the decompensation and subsequet failure.

Until recently, the diagnosis of mitral atresia was generally considered important only for the purpose of excluding it from other cyanotic and possibly correctable congenital heart diseases, and its diagnosis was regarded as of academic interest only, most cases being found at autopsy. By the use of angiocardiography and cardiac catheterisation, Redo *et al.*, (1961) recently

diagnosed four cases of mitral atresia during life. Three of the four cases underwent cardiac surgery with one success in an eight year old boy. In this case preoperative investigations revealed mitral atresia, a left to right shunt at the atrial level, a right to left shunt at the ventricular level, corrected transposition of the great vessels and pulmonary stenosis. The operation, consisting of widening of the atrial septal defect, resulted in symptomatic improvement, and the boy was alive and well at the time of publication eighteen months after the operation. Until then the treatment of mitral atresia was largely symptomatic, for respiratory infections and congestive cardiac failure. With the advances of diagnostic techniques and cardiac surgery, the prognosis of mitral atresia would appear to be less pessimistic.

Summary

A case of mitral atresia has been presented and the post mortem findings reported. The combination of associated lesions makes this case interesting. The chances of survival, the more common associated anomalies, the diagnosis, prognosis and the possible surgical treatment in selected cases have been briefly discussed.

Acknowledgement

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References

Abbott, Maude E. (1936). Atlas of Congenital Cardiac Diseases, New York, The American Heart Association p. 50.

Brockman, H. L. (1950). Congenital Mitral Atresia, Transposition of the Great Vessels and Congenital Aortic Coarotation. *Am. Heart J.* 40: 301.

Coppoletta, J. M. and Wolbach, S. B. (1933). Body length and normal weight of more important vital organs between Birth and Twelve years of age. *Am. J. Path.* 9: 55.

Edwards, J. E. and Dushane, J. W. (1950). Thoracic venous anomalies. *Arch. Path.* 49.: 517.

Edwards, J. E. (1953) in Gould S. E. ed. Pathology of Heart, Springfield, Thomas p. 386.

Eliot R. S. ,Shone J. D., Kanjuh V. I., Ruttenberg, H. D., Carey L. S., Edwards J. E. (1965). Mitral Atresia. *Am. Heart J.* 70. 6.

Lam C. R., Knights E. M., Ziegler R. F. (1953). Combined Mitral Atresia and Pulmonary Atresia. *Am. Heart J.* 46: 314.

Large, H. L. (1960). Congenital Mitral Atresia. *Am. J. M. Sc.* 219: 268.

Lumb G., and Dawkins, W. A. (1960). Congenital Atresia of Mitral and Aortic Valves with vestigail left ventricle (three cases) *Am. Heart J.* 60: 379.

Redo, S. F., Farber S., Gross R. E. (1961). Atresia of the Mitral valve, *Arch. of Surg.* (Chicago) 82: 696

Shone J. D. and Edwards, J. E. (1964). Mitral Atresia with pulmonary venous anomalies. *Brit. Heart J.* 26: 241.

Walls, E. W. (1941). Bilateral Trilocular heart with atresia of the mitral valve. *Lancet* 2: 668—669.

Watson, D. G., Rowe R. D., Conen P. E. Duckworth J. W. A. (1960). Mitral Atresia with normal aortic valve. *Pediatrics* 25: 450.