CONGENITAL TRICUSPID ATRESIA

BY

JAMES W. BROWN, M.D., M.R.C.P.

(From the Grimsby and District Hospital.)

Congenital tricuspid atresia is an infrequent malformation of the heart. Abbott¹ in her analysis of 1,000 congenital cardiac cases gives sixteen examples. To these may be added the cases of Bellet and Steward², Murphy and Bleyer³, and Grayzel and Tennant⁴, making a total of nineteen cases in the available literature. It is probable, however, that many cases escape recognition from lack of a post-mortem examination, or from the assumption that the cyanosis invariably present is the result of some obstructive lesion of the pulmonary tract. In the case here recorded laboratory examination enabled the exclusion of a stenotic pulmonary lesion, but the diagnosis was not certain until autopsy. Two main types of tricuspid atresia are described. Isolated tricuspid atresia, with which are associated interauricular and interventricular septal defects and other secondary changes is the commoner type. The other group comprises those cases in which the tricuspid atresia is associated with transposition of the vessels or other abnormality. Duration of life is longest in this second group. This communication is concerned with isolated tricuspid atresia.

Case record.

A male child came under observation at the age of five months on account of cyanosis dating from birth. He was poorly developed and weighed 5 lb., and had failed to gain weight. He was dyspnoeic at rest. Marked cyanosis was present, and the least movement or such simple acts as feeding or crying led to increased cyanosis and respiratory distress. There was no clubbing of the fingers or toes.

On examination there was evident enlargement of the heart, the apex beat being forcible in the anterior axillary line. A mesocardial systolic thrill of variable intensity was present. A systolic murmur, rough in quality, was heard over the precordium and was loudest over the base of the heart. It was heard widely over the back but not in the neck. The liver was palpable but the spleen was not enlarged.

X-ray examination (fig. 1) showed a globular heart with exaggerated convexity of the right and left borders suggesting right auricular and left ventricular hypertrophy.

The electrocardiogram (fig. 2) showed marked left axis deviation. The P wave in lead II was large and the T wave in lead I diphasic.

Death from bronchopneumonia occurred at the age of eight months.

FIG. 1.—X-ray appearance of heart.

FIG. 2.

POST-MORTEM EXAMINATION. The heart was greatly enlarged and globular in shape (see fig. 3). The anterior surface of the heart consisted mainly of the left ventricle and the dilated right auricle. The aorta appeared to be much larger than normal and arose in its normal relationship to the pulmonary artery. The left ventricle was dilated and hypertrophied with walls 10 mm. thick. There was an oval shaped defect of the upper part of the interventricular septum (fig. 3 B and C), anterior to the membranous septum and beneath the non-coronary cusp, from which latter it was separated by a band of fibrous tissue. The defect measured 5 by 4 mm. The sinus of the right ventricle was small and rudimentary, and of about the capacity of a hazel nut. There was no evidence of tricuspid leaflets or papillary muscles. The conus of the right ventricle was narrow with thick walls. The defect of the interventricular septum opened beneath the crista supraventricularis. The pulmonary artery was of normal calibre and the pulmonary cusps were normal. The right auricle was dilated and hypertrophied. There was no tricuspid orifice and its normal site was marked by an elliptical depression (fig. 3 D). Relatively thick muscular tissue intervened between the right auricle and ventricle. A large patency of the foramen ovale admitted the little finger. The left auricle was moderately dilated but its walls were not hypertrophied. The mitral valve was normal. The aorta was enlarged relative to the pulmonary artery but its walls were healthy. The aortic cusps, three in number, presented no abnormality. The ductus arteriosus was closed.

An accessory spleen was present.

ANATOMICAL SUMMARY. Atresia of the tricuspid orifice; interauricular septal defect; interventricular septal defect; hypertrophy and dilatation of the right auricle and left ventricle; rudimentary cavity of the right ventricle.

Discussion.

Such a combination of defects as are present in this case are the constant findings in isolated tricuspid atresia. The primary lesion is atresia of the tricuspid orifice which may be so complete that not even a scar indicates the site of the opening. More frequently a slight depression or a few muscular bands, as in the present case, are found to mark its normal site. The right ventricle is always extremely small and aplastic, in size scarcely the volume of a nut (Laubry and Pezzi³), or existing as a small irregular flattened space in the wall of the right ventricle, which might well be overlooked (Robertson⁶). Some cases, as for example that of Robertson, have been described as a type of cor biatriatum triloculare. The right auricle is dilated and hypertrophied. There is always an interauricular septal defect, either a widely patent foramen ovale or other abnormality of the septum. There is nearly always a defect of the interventricular septum (87.5 per cent. Abbott's series) and this may open, as in the present case beneath the crista, or directly into the conus of the pulmonary artery. The ductus arteriosus was patent in only 37.5 per cent. of Abbott's cases. In the absence of a patent ductus, which would assure a limited supply of blood to the pulmonary circulation, it is evident that the defect of the interventricular septum is essential for the maintenance of the circulation. The increased work performed by the left ventricle leads to its hypertrophy.

(A) External view of heart.

I.V. Septal defect

(B) Right ventricle opened.

I.V. Septal defect

(C) Left ventricle opened.

Foramen
Ovale

Atresic Tricuspid
orifice

(D) Right auricle.

FIG. 3.

Various theories have been advanced in explanation of the abnormality. Unequal division of the common auriculoventricular orifice by the abnormal growth of the anterior and posterior endocardial cushions, so that these adhere to the right wall of the common orifice, would appear to be the most satisfactory theory. Such an idea has been embodied by Abbott in her statement that the abnormality 'arises as the result of malposition and irregular union of those parts of the cardiac septa dividing the mitral from the tricuspid ostium.' Obliteration of the tricuspid orifice takes place at about the fourth week of foetal life and closure of the septa occurs at the eighth week. It is probable that raised pressure in the left ventricle is an important factor in maintaining a conduit between the two sides of the heart.

No case of tricuspid atresia has so far been diagnosed during life. As some of the cases survive until late childhood, or even to puberty or later, the clinical features are worthy of consideration. The presenting symptoms are cyanosis of marked degree, and clubbing in cases that survive more than a year. Cyanosis exists from birth and is enhanced by movement or emotion. The heart is enlarged to both left and right sides and the apex beat is forcible. Laubry and Pezzi insist on the value of percussion of the back as a means of detecting enlargement to the right of the vertebral column in children. A mesocardial systolic murmur is commonly present, and may be heard widely over the chest. A systolic thrill is present in 12.5 per cent. of cases (Abbott). X-ray examination shows an enlarged and globular heart. The convexity of the right border, corresponding to the right auricle, is exaggerated. The left border is well rounded and presents the appearance of left ventricular hypertrophy. The gross enlargement of the right auricle and the slight enlargement of the left can be confirmed in the oblique positions. The electrocardiogram shows normal or left axis deviation. The P wave may be enlarged or notched suggesting auricular hypertrophy.

The dictum of Fallot that 74 per cent. of cyanotic cases were examples of his tetralogy has perhaps led to the assumption by the physician that any congenital cyanotic case surviving more than a few days must almost always be the result of a stenotic pulmonary lesion with associated septal defect permitting a shunt. Apart from the interest that a cyanotic case evokes, little effort has been made until recently to identify anatomical lesions in this group. The advent of laboratory aids in the investigation of cardiac disease has at any rate enabled the segregation of those cases in which there is a stenotic lesion of the pulmonary tract and septal defect from the smaller group of cases where some other abnormality causing cyanosis exists. The skiagram is distinctive in some conditions, notably the tetralogy of Fallot, and the auricular septal defect. In tricuspid atresia the enormous right auricle and the hypertrophied left ventricle are suggestive. Of more importance is the electrocardiogram. The finding of normal,

and particularly of left axis deviation in a cyanotic case should at once exclude the tetralogy or any lesion of the pulmonary tract. Pulmonary stenosis with a closed septum, which may rarely be associated with normal axis deviation, is not accompanied by cyanosis except as a late or terminal event. The tetralogy of Fallot is always accompanied by right axis deviation unless dextrocardia or gross conduction defect are present.

Summary.

A case of isolated tricuspid atresia has been presented and its main features discussed. It is suggested that this abnormality might be more frequently diagnosed if the radiological and electrocardiographic pictures were carefully considered.

REFERENCES.

1. Abbott, M., *Loose-Leaf Medicine*, New York, 1931.
2. Bellett, S., & Steward, H. L., *Am. J. Dis. Child.*, Chicago, 1933, XLV, 1247.
3. Murphy, G. R., & Bleyer, L. F., *Ibid.*, 1933, XLVI, 350.
4. Grayzel, D. M., & Tennant, R., *Am. J. Path.*, Boston, 1934, X, 791.
5. Laubry, Ch., & Pezzi, C., *Maladies Cong. du Coeur*, Paris, 1921.
6. Robertson, J., *Lancet*, London, 1911, i, 872.
7. Abbott, M., *Modern Medicine*, Ed. Osler, W., & McCrae, T., London, 1927, IV, 755.