

MALFORMED HEART WITH REDUNDANT AND DIS- PLACED TRICUSPID SEGMENTS AND ABNORMAL LOCAL ATTENUATION OF THE RIGHT VENTRICULAR WALL

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IN 1920 there were published by Dr Shaw and myself in the *Journal of Anatomy* (Vol. LIV, p. 163) particulars of a case under my care in the Great Northern (now Royal Northern) Hospital which showed combined cardiac and genito-urinary anomalies. The cardiac condition consisted in a redundancy of the anterior, low insertion of the posterior, and rudimentary state of the septal cusp of the tricuspid valve. The right ventricle was dilated and hypertrophied, the auriculo-ventricular orifice wider than usual and the clinical signs indicative of the condition were a tricuspid regurgitant bruit and strikingly marked accentuation of the first sound of the heart. The patient died of tuberculous meningitis.

On the 27th of February, 1922, there was brought into the Royal Northern Hospital the dead body of a man, 33 years of age, on whom an inquest was held and in the course of post-mortem examination, Dr Shaw, the Pathologist to the hospital, found a cardiac state which reminded him of the case to which I have already referred, and which we reported together to the Anatomical Society. He saved the heart for my inspection and examination in detail, and has given me permission to exhibit it to the Society on this occasion.

No particulars of previous illness transpired at the inquest and discovering the address of the widow later, I learned from her, to whom I addressed a series of questions, the following particulars which may be given in her own words: "He made no complaint of any kind during the years I knew him. His occupation was that of an electrical condenser maker. No doctor has attended him for the past twelve years. He appeared to be a fully healthy man and only appeared to be extremely tired at times. He appears to have had no knowledge of any defect until rejected when a candidate for the police at the age of twenty-one years." I was also informed from another source that he had been rejected when examined for military service during the war, on account of a cardiac defect which was detected. He therefore evidently gave some physical signs of his condition although not subjectively suffering.

The pathologist's notes of the necropsy are as follows: "*Summary.* Congenital malformation of tricuspid valve. Body of a well-nourished man. No evidence of external injury or violence. Hypostasis present posteriorly. *Head.* No wounds on scalp. Membranes natural, brain 51 ozs. natural. Skull vault and base natural. *Chest.* Pleura; old adhesions between parietal and visceral

layers. Lungs: right, 21 ounces, left, 17 ounces. Both (with) passive hyperaemia.

Heart and Pericardium. Weight 15 ounces. Right side much dilated and wall of ventricle very thin. Tricuspid orifice admits (the) whole hand easily. Segments attached very irregularly to wall of ventricle. Septal cusp small and vertical; posterior cusp attached low down; right cusp reaches about to apex. Large area of wall of ventricle situated above the attachments of the segments. Other valves natural. Pericardium natural.

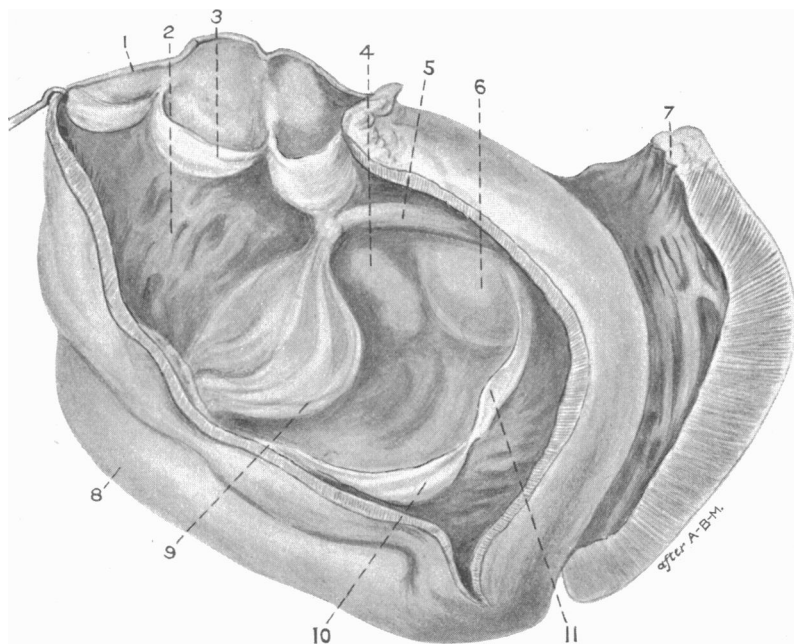


Fig. 1. Malformed heart with attenuated area in right ventricle and low insertion of tricuspid segments, the anterior cusp being redundant. 1, pulmonary artery; 2, conus arteriosus; 3, pulmonary semi-lunar valves; 4, large pars membranacea septi illuminated from the left ventricle; 5, papillary band; 6, internal cusp of tricuspid valve; 7, left ventricle; 8, right ventricle; 9, anterior cusp of tricuspid valve; 10, 11, posterior cusp of tricuspid valve.

Stomach. Evidence of recent meal. Passive congestion of mucous membrane. Intestines natural.

Kidneys. Weight together $11\frac{1}{2}$ ounces. Firm. Capsule strips easily. Size natural. Passive hyperaemia. **Liver.** Weight 61 ounces. Passive hyperaemia. **Spleen.** Weight 5 ounces. Size natural. Firm. Slight congestion."

The following are the more exact measurements which I made and the details of the heart in question.

Pulmonary arterial circumference, 8 cm.; valves normal. Tricuspid valvular orifice (for it is not the atrio-ventricular orifice), 7.5 cm. by 7.5 cm.; anterior cusp 4.5 cm. deep, 6 cm. long; posterior cusp 2 cm. in depth at deepest, 5 cm. long; internal cusp 2 cm. deep, 3 cm. long.

Pars membranacea septi 2 cm. by 2.5 cm. Right ventricle 10.5 cm. by 8.5 cm.; ventricular wall 4 to 5 mm. at thickest.

Base of the anterior segment of the tricuspid valve attached at the auriculo-ventricular junction and the segment itself to either side of a large *flattened* papillary muscle from the lower end of which a second papillary muscle gives attachment to the outer end of the posterior segment; the *base* of which is

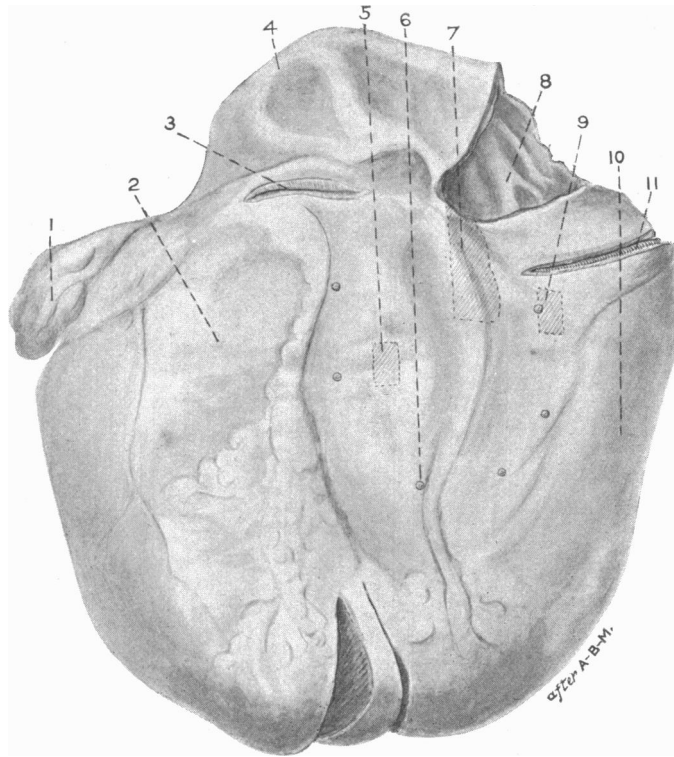


Fig. 2. Malformed heart with attenuated area in right ventricle and low insertion of tricuspid segments, the anterior cusp being redundant. 1, left auricular appendix; 2, left ventricle; 3, coronary sinus laid open; 4, right auricle; 5, 7, 9, portions removed for microscopic examination; 5, in attenuated area of right ventricle; 7, in auriculo-ventricular boundary of attenuated area; 9, in right border of attenuated area; 6, one of a series of pin-heads showing the extent of the attenuated area; 8, muculi pectinati at the entrance to the right-auricular appendix; 10, the left ventricle; 11, the right coronary artery laid bare. B.O. male æt. 33 years.

3.5 cm. from the ventricular apex and 5 cm. below the auriculo-ventricular line. A very much attenuated portion of the posterior ventricular wall in the shape of an inverted triangle, measures 4.5 cm. by 5 cm.

The right auricle measures 7.5 cm. by 5 cm. Its wall is thickest, 1 cm., at the anterior part of the circumference of the superior vena cava and over the rest of the cavity is from 2 to 3 mm. thick. The foramen ovale is closed and large, measuring 2.5 cm. by 2.5 cm.

The coronary sinus is normal in position and has a short Thebesian valve. The inferior vena cava is normal and shows a Eustachian valve. The auricular appendix is likewise normal and the *musculi pectinati* well-marked.

The left ventricle measures 8 cm. by 7.5 cm., the average thickness of its walls being 1.5 cm. The aortic and mitral valves are normal. The right coronary arterial orifice is large, the left small. There is some atheroma in the sinuses of Valsalva. The aortic circumference is 6 cm.

The left auricle measures 10 cm. by 4.5 cm. Its walls average 2 to 3 mm. in thickness. Microscopic sections of the attenuated area, at the level of the coronary vessels (sulcus), and far above the insertion of the posterior segment of the tricuspid valve show the auricular to be quite distinct from the ventricular musculature and demonstrate the area in question to be purely ventricular. Sections from the most attenuated part of this area show a sheet of muscle, while its lateral junction with the thicker ventricular wall is in normal muscular continuity. Any muscular action of this area would follow auricular systole, but tend to open, not to close, the tricuspid valve.

The subvalvular portion of the ventricular cavity is thus limited abnormally, and, notwithstanding the capaciousness of the anterior segment of the tricuspid valve there must have been free regurgitation and all round blood pressure in the chamber which accounts for the abnormal size of the valvular orifice and the general thinness and flattening of structures in it. The weight of the heart being fifteen ounces, that is, rather over the average weight of the normal male heart, shows that some attempt at compensatory hypertrophy had been made to deal with the circulatory difficulty. A noteworthy fact is the relative position of the internal cusp of the tricuspid as regards the *pars membranacea septi* and as regards the papillary band, which is the left attachment of the anterior segment; the *pars membranacea* being posterior to and not covered by the internal or septal segment, a fact which may have developmental significance. Like the membrane closing the foramen ovale, the *pars membranacea* is abnormally large, but both the auricular and ventricular septa are complete, while the chief papillary muscle is quite flattened. The coronary sinus is normal in position, and the auriculo-ventricular bundle of His may be, but the latter point has not been determined positively. Sections of a large branch of the right coronary artery in the right auricle show considerable thickening of the intima and some flattening of the lumen, but there is no general increase of intima in the arteries, taking the membrane of Henle as a guide. There is neither naked-eye nor microscopic evidence of muscular deformation and sections of a papillary muscle from the left ventricle show no interstitial increase, but sections from the attenuated area in the right ventricle reveal a loose spreading intercellular connective tissue. The attenuated area is not degenerative and pathological, but developmentally deficient.

The break in the continuity of the fibrous ring at the auriculo-ventricular boundary of the area of attenuated ventricular wall which allowed distension, together with the low position and ineffective closure of the valve segments

resulting in the dilated dextral chambers, is a striking testimony to the importance of the fibrous support of the valvular apparatus and of the valves themselves, in the cardiac mechanism of the circulation. It was the fashion not long ago to decry the importance of the integrity of this factor. One rarely meets with such eloquent evidence to the contrary as is afforded by this case and the importance of this factor is not less but greater in the left or systemic arterial heart.

In the unfortunate absence of a record of the clinical signs in the case of B.O. some of the clinical physiology must be left obscure, but much may be deduced from the anatomical details given. His sudden death without a history of previous ill-health is interesting, as in many other cases of heart disease so dying. I shall not, however, comment further on this fact, beyond stating my belief that, if in the case of B.O., the foramen ovale had remained patent, it is probable that he would have lived longer. In support of this opinion, I may recall to the memory of members of this Society the case of an old man of whom I gave an account in 1917 (Vol. LII, p. 251), and who, with an almost obliterated pulmonary artery and very hypoplastic or rudimentary right ventricle, lived to a venerable age, because he had, like B.O., a sound left heart, but also a widely patent foramen ovale, which gave easy access of the right heart blood into the left chambers.