STENOSIS AT THE LOWER BULBAR ORIFICE OF THE INFUNDIBULUM

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Stenosis at the lower bulbar orifice is not very rare, as Keith (1909) in the examination of two hundred and seventy abnormal hearts found it to be present in nineteen. We consider, however, that the associated abnormalities of this heart make the case worthy of publication.

The right ventricle is a composite chamber, formed partly by the transverse part of the primary ventricular loop and partly by the lower part of the bulbus cordis ; the bulbus lies between the common ventricle and the truncus arteriosus in the embryonic heart. The bulbus cordis persists in elasmobranchs and most of the reptiles as a chamber that remains separate from the ventricle, but in the mammalian heart part of the bulbus is incorporated in the right ventricle. Greil (1903) was the first to suggest that the part of the bulbus absorbed by the right ventricle is represented by the infundibulum (conus arteriosus) in the adult human heart. Keith (1924) considers that the function of the infundibulum in the mammalian heart is essentially akin to that of the bulbus cordis in the lower vertebrates, in which it persists as such ; that the bulbus acts as a guarding mechanism for the vascular network of the lungs against the raised blood pressure that occurs during increased cardiac activity ; and that the musculature of the infundibulum passes into action later than that of the body of the right ventricle.

The component parts of the right ventricle in the human heart, however, may retain their individuality and only communicate with each other by a constricted orifice. This condition has been referred to by Keith as *stenosis at the lower bulbar orifice of the infundibulum*.

In such a heart the infundibulum itself is expanded above the orifice and forms a dilated, but relatively thin-walled chamber, which communicates freely with the pulmonary artery. The wall of the right ventricle below the orifice, i.e. the ventricle proper, is hypertrophied and is usually thicker than the wall of the left ventricle. The size of the lower bulbar orifice varies greatly in different specimens, and the duration of life tends to bear a direct relationship to the size of this orifice.

In the great majority of examples so far described there has been a deficiency

in the interventricular septum and in most of these the communication has been such as to provide a shunt between the two chambers. In four cases, recorded by Lafitte (1892), Jackson (1893), and Eakin and Abbott (1933), there was no interventricular foramen. Associated with this deficiency there is usually some degree of dextro-position of the aorta. This combination of defects is not surprising, as the incorporation of the bulbus cordis in the right ventricle normally takes place early in development before the completion of the septa and before the normal torsion of the arterial trunks. For the same reason a patent foramen ovale and a patent ductus arteriosus are sometimes found in association with these other changes.

CASE RECORD

A. L., aged 38, was admitted to the Eastern General Hospital, Edinburgh.

History.—He had “heart trouble” of unknown nature when 17 years of age, from which he recovered well. He never had rheumatic fever, but suffered frequently from sore throats, the last being one week prior to the onset of his terminal illness. Throughout his life he had been unable to take violent exercise because it caused attacks of extreme breathlessness and cyanosis. No other illnesses were admitted.

One month before admission he began to notice weakness and marked tiredness on exertion and inability to walk more than a quarter of a mile without a rest. His feet, face, and tongue began to swell. He developed a cough with sputum, and for three weeks had suffered from diarrhoea. As the condition was progressing rapidly, he was sent to hospital.

Examination.—He was of medium size, with a sallow complexion, cyanosed lips and hands, and puffiness of the face. The fingers and toes exhibited clubbing. No purpuric spots were noted. There was œdema of the feet and legs and also a small lumbar pad.

The pulse was rapid (100 per minute), regular, and of good volume. Blood pressure, 150/100 mm. The vessel walls were not palpable. The apex beat was visible and diffuse; on palpation it was forcible and was localized in the sixth interspace in the anterior axillary line. There was an apical diastolic thrill. On auscultation there was a harsh loud diastolic murmur of maximum intensity in the fourth interspace at the left border of the sternum, propagated in all directions. There was also a systolic murmur in the pulmonary area, propagated upwards for one inch towards the left.

Both bases were dull to percussion posteriorly and the respiratory sounds were diminished. The respiratory rate was 30. The sputum was slight in amount and consisted of a frothy mucus.

The abdomen was distended. The liver was not enlarged. Otherwise no abnormality was detected on clinical examination.

The urine contained much albumen and a faint trace of blood. Microscopically there were granular and epithelial casts and red blood cells. The hæmoglobin was 45 per cent.

Course.—The temperature swung between 97° and 101° during the first six days in hospital, but thereafter remained normal or subnormal. A blood culture was carried out twice during this pyrexial stage, but no growth was obtained. The œdema became worse, despite therapy. The spleen, which had not been palpable, enlarged progressively and became tender. At no time were any purpuric spots noted. The patient remained lethargic in his reactions for the first week, after which he became more torpid and eventually died in a coma two weeks after his admission.

Summary of Autopsy Findings.—The body was moderately developed. There was slight but generalized œdema. The pericardial and peritoneal sacs contained several

ounces of clear serous fluid. Both pleural cavities were almost obliterated by dense fibrous adhesions.

The heart exhibited gross abnormalities which are given in detail below. The outstanding feature was the tremendous hypertrophy of the right ventricle.

Both lungs were congested and slightly œdematous towards their bases ; no infarcts were detected.

The spleen weighed 750 g. No infarcts were present in the spleen or in the kidneys, both of which showed gross congestion.

No congenital abnormality was detected apart from that noted in the heart.

ANATOMICAL FEATURES OF THE HEART

The chief abnormalities present were : (a) stenosis of the infundibulum (conus arteriosus) at the lower bulbar orifice ; (b) hypertrophy of the right ventricle below the orifice ; (c) dilatation of the infundibulum ; (d) communication of the right ventricle with the anterior sinus of the aorta ; and (e) a small communication between the right and left ventricles.

The heart showed general enlargement (550 g. compared with an average of 330 g.), especially of the right auricle, which was markedly dilated, and of the right ventricle, which was more conspicuous than usual in the region of the infundibulum.

The walls of both right chambers were thickened. The wall of the right auricle had an average thickness of 4 mm., with prominent muscoli pectinati ; the wall of the left auricle was 2 mm. thick. Beneath the anterior limb of the limbus of the fossa ovalis there was an oblique passage 10 mm. long just admitting a fine probe. This is the standard type of patent foramen ovale which is found in a large percentage of hearts (25–30 per cent.).

The right ventricle was subdivided by an obliquely placed septum into (a) the right ventricle proper and (b) the infundibulum (conus arteriosus) ; the only communication between these two subdivisions was a foramen in the septum which measured 7×5 mm. ; the edges of the foramen were white and fibrous. The general disposition of these parts of the chamber are shown in Fig. 1.

The wall of the ventricle proper was especially noteworthy on account of its unusual thickness. Near the lower margin of the heart it was 13 mm. thick, gradually increasing towards the base of the chamber to 24 mm. ; the thickness of the ventricular septum was, on an average, 15 mm., with a corresponding increase in the trabeculæ carneæ, moderator band, and papillary muscles. The tricuspid orifice in the fresh specimen was large enough to admit five fingers ; the chordæ tendineæ were much thicker than usual and near the cusp margins flattened out into membranous bands.

About 10 mm. in front of the left end of the base of the anterior cusp there was an opening, with a maximum diameter of 7 mm., that led into the aorta. The margins of this orifice were thin and membranous and in the fixed specimen had come together so as to occlude the passage. This connection between the right ventricle and the aorta indicates that there was some dextro-position of that vessel. The site of the upper opening of the orifice was especially note-

FIG. 1.—Anterior view of the heart. The interior of the infundibulum and the communication of this chamber with the body of the right ventricle is shown.

worthy in that it was actually into the anterior sinus—which was much enlarged—and was about 10 mm. below the origin of the right coronary artery.

Immediately behind the aortic opening, i.e. between it and the tricuspid valve, there was a small, slit-like orifice, 3 mm. long and just large enough to admit a fine probe, leading into the aortic vestibule. The opening of this communication into the right ventricle was so guarded by a flap-like fold of endocardium that, quite apart from its size, there could be little doubt that it had not functioned during life.

The infundibular part of the right ventricle (Fig. 2) was much enlarged. In general it had a slightly saccular form, with a maximum length of 31 mm. and a width of about 20 mm. The posterior wall of the chamber was smooth, but the anterior wall was roughened by several well-defined trabeculæ carneæ.

FIG. 2.—Anterior aspect of the heart. The interior of both the right and the left ventricles is shown and three arrows have been inserted to demonstrate the communications between the chambers. The *black* arrow has been passed from the body of the right ventricle into the infundibulum ; the *stippled* arrow passes from the left ventricle into the aorta ; and the *white* arrow from the right ventricle into the aorta.

The opening into the right ventricle proper was on the right wall of the chamber about 13 mm. above its most dependent part ; above, the chamber communicated freely with the pulmonary artery through the pulmonary valve. The cusps of the pulmonary valve had the normal anatomical arrangement and showed no stenosis. The wall of the chamber was not of equal thickness throughout, as in its lowest part it varied from 5 to 9 mm. and as it was traced upwards it gradually thinned and so imperceptibly merged with the wall of the pulmonary artery.

The wall of the left ventricle was relatively thin as compared with the right, as near the base of the heart it was 14 mm. (cf. 24 mm. in the right) and at the apex it was 7 mm. (cf. 13 mm. in the right). The aortic orifice was displaced slightly to the right.

The diameter of the aorta was 21 mm. as compared with 17 mm. in the pulmonary artery; in the normal heart the pulmonary artery is slightly wider at its origin than the aorta.

Pathological Features.—In addition to these anatomical abnormalities there was endocarditis involving the opening between the right ventricle proper and the infundibular portion. Large fungating vegetations completely surrounded the orifice. No ulceration of the wall in this area could be detected, but the vegetations extended upwards from the orifice on to the wall of the conus arteriosus and involved the cusps of the pulmonary valve. They were growing through the orifice also and protruding into the right ventricle.

Microscopically.—The vegetations consisted of a fibrin thrombus with organization in its base; this and the adjacent muscle was densely infiltrated with cells in which polymorphs were prominent. On the surface of the vegetations were clumps of cocci. On the conus side of the pulmonary valve there was a similar formation, but with a more cellular fibrosis and less cellular infiltration and fibrin clot. A large patch of hyalinized fibrous tissue in the wall of the conus had apparently arisen from the overlying endocarditic changes. The myocardial fibres underlying the endocarditis were somewhat hyalinized, and in the right ventricle showed considerable hypertrophy. There was some periarterial fibrous thickening, and thin strands of connective tissue could be seen extending out between the muscle bundles from the vessels. This feature was much more marked in the right ventricle than in the left. No Aschoff nodules were present, but there was some intimal thickening with scarring in a few of the smaller myocardial vessels. This was obviously of long standing, but was not specific enough to warrant a diagnosis of an old rheumatic lesion. The wall of the right auricle contained more fibrous and less elastic tissue than did that of the left. As compared with a normal heart, however, this variation was not so prominent as was the relative difference in the ventricles with the definite fibrosis in the wall of the right chamber.

DISCUSSION

The case is a typical example of stenosis of the lower bulbar orifice of the infundibulum. Associated with this stenosis there was hypertrophy of the wall of the right ventricle, a minute opening in the ventricular septum, and a slight degree of dextro-position of the aorta.

The stenosis of the lower bulbar orifice in this case was essentially similar to that found in the cases of this anomaly reported previously. In these the measurements showed a wide variation, but the size in this specimen (7×5 mm.) was less than usual. The position of the orifice was not as usually described, however, as it was not in the most dependent part of the infundibulum, but about 13 mm. higher. The stenosed orifice was quite unguarded by any valvular mechanism and was probably responsible for the murmurs heard during life, though the vegetations around the orifice might have exaggerated them. The presence of bacterial endocarditis at the site of

the anomaly is in keeping with the view that such a lesion tends to occur in association with some pre-existing malformation.

The hypertrophy of the wall of the right ventricle proper was undoubtedly due to the small size of the pulmonary outlet. Associated with this hypertrophy there was a patchy fibrosis throughout the wall of this ventricle which was absent from the left ventricle. This fibrosis was, if anything, more pronounced in the subendocardial region of the myocardium, as had already been observed in a similar case reported by Eakin and Abbott (1933).

The opening in the ventricular septum was small and as it was guarded on the right side by a flap-like fold of endocardium it is felt there could not have been a veno-arterial shunt. Hence this case may be classed with these where the septum is complete. As already stated, stenosis of the lower bulbar orifice is generally found with a large defect in the ventricular septum, which allows free communication between the pulmonic and systemic circulations, so that these patients always have a slight cyanosis. The present case, however, only showed cyanosis during exertion, and it is justifiable to infer, therefore, that this must have been due to the stenosed bulbar orifice rather than to a veno-arterial shunt. This stenosis, while allowing a sufficient passage for normal activities, would interfere with the additional flow of blood that would usually pass to the lungs on exertion.

The aorta showed some degree of dextro-position and the manner of connection between the right ventricle and this vessel was of special interest. The opening was between the highest part of the right ventricle and the enlarged anterior aortic sinus, about 10 mm. below the origin of the right coronary artery. The orifice, which was 7 mm. in diameter, had a fibrous ring projecting into the sinus in the form of a cone. Probably, therefore, this had a valvular action and prevented the regurgitation of aortic blood in the right ventricle during diastole. This mode of communication, whereby the right ventricle opens into the aortic sinus, has never been recorded before so far as we are aware. The proximity of this ventriculo-aortic communication and the origin of the right coronary artery suggests that the former might have been responsible, by interfering with the blood supply, for the undue fibrosis present in the wall of the right ventricle.

SUMMARY

A heart is described in which there was stenosis of the lower bulbar orifice of the infundibulum. This feature is not in itself rare, but the absence of a functional interventricular foramen makes the case unusual. The communication of the right ventricle with the anterior aortic sinus is a further feature of interest.

Despite the gross abnormalities that were present in the heart, the subject lived to the age of thirty-eight, when the occurrence of bacterial endocarditis was responsible for death.

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