A HITHERTO UNDESCRIBED MALFORMATION OF THE HEART.

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THE specimen described below was sent to me in 1912 by my friend Dr J. A. Bell, M.C., Captain R.A.M.C. (T.), who was at that time Medical Officer at Camberwell Workhouse; I am greatly indebted to him for giving me the opportunity of examining and describing it.

The heart (which is now in the Museum of the Royal College of Surgeons of England) is that of a male infant, born in the workhouse; the child, which weighed 7 lbs., survived only thirty-eight hours. Other clinical details are lacking.

Unfortunately the specimen suffered considerably in the process of removal from the body—the more unfortunately, because it shows a condition which, so far as I am aware, has not up to the present been described, namely, that the aorta, though of normal size, has no communication with either ventricle, but was filled from near the apex of the left ventricle by way of the anterior interventricular branch of the left coronary artery.

In fig. 1 the heart has been drawn so as to show most of its abnormalities. There is a very obvious disparity in the size of the cavities of the right and left sides of the heart: this affects especially the auricles, the right auricle being large, while the left is remarkably smaller than normal.

The *right auricle* shows the normal division of its interior into two parts by a prominent crista terminalis, behind which is a large opening of the inferior vena cava; that of the superior vena cava appears to have been, in part at least, cut away; no Eustachian valve is present. At its left lower extremity the right auricle receives the wide opening (unguarded by any valve) of the left horn of the sinus venosus, which becomes continuous, round the left side of the left auricle, with a persistent left superior vena cava; a great cardiac vein is present in the left auriculo-ventricular furrow, and also opens into the sinus.

Enough of the interauricular septum remains to show that the foramen ovale was widely open.

The right auriculo-ventricular orifice and the tricuspid valve (fig. 2) show no gross abnormalities, though the cusps of the valve are unusually large. The opening measures 15 mm. in its longest diameter.

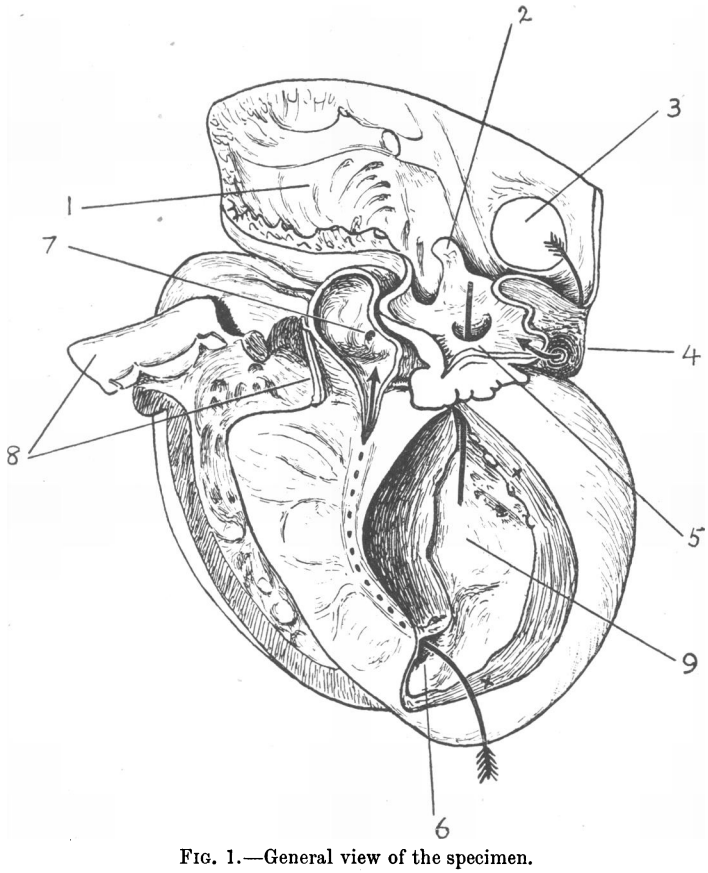

FIG. 1.—General view of the specimen.

1, right auricle; 2, interauricular septum, with patent foramen ovale; 3, opening of inferior vena cava; 4, left horn of sinus venosus and left superior vena cava, from which arrow emerges; 5, cavity of left auricle; a rod passes through the mitral orifice; 6, the abnormal communication between the left ventricle and the anterior interventricular coronary artery; a bent arrow passes along the latter into the aorta; 7, the point of origin of the right coronary artery from the aorta; 8, commencement of the pulmonary artery, in two pieces; 9, left side of interventricular septum; above is seen the oblique ridge which divides the cavity of the left ventricle into two parts.

The *right ventricle* shows considerable enlargement, which is confined to the body of the ventricle, the infundibulum, although it does not appear to be smaller than normal, being disproportionately small as compared with the rest. The cavity is seen to extend behind the left ventricle as far as

the left margin of the heart, the septum between the right and left cavities being very obliquely placed.

Only the very commencement of the pulmonary artery is preserved, and that in pieces, but it is clear that the orifice was approximately of normal size; and it has a normal valve with three cusps.

The *left auricle* (fig. 1) is many times smaller than the right; no pulmonary veins are seen opening into it; and although they may have

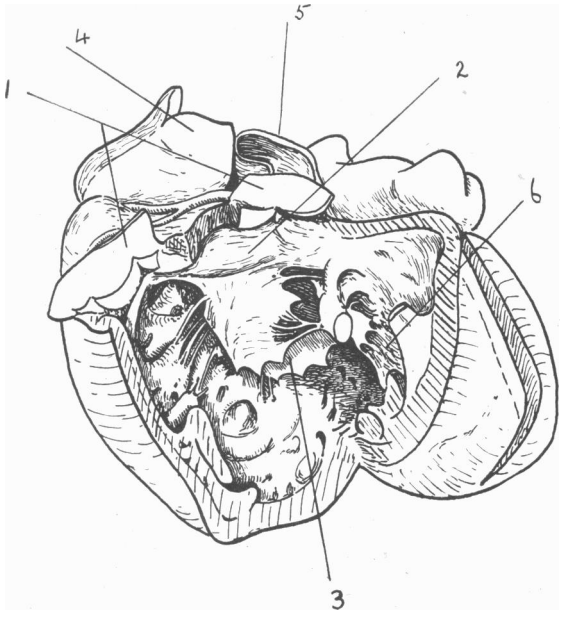

FIG. 2.—The right ventricle has been widely opened.

1, the two parts of the pulmonary artery; 2, infundibulum of ventricle; 3, tricuspid valve; 4, right auricle; 5, ascending aorta; 6, right side of interventricular septum; the septal cusp of the tricuspid valve is applied to its upper part. Note the cavity of the ventricle, in shadow, carried far to the left behind it.

been cut away, yet the incised wall of the auricle can be brought together in such a way as so nearly to reconstruct the chamber that it is difficult to see how the veins could have reached it. There is a small auricular appendix, with miniature musculi pectinati in its interior.

From the auricle a smooth, rounded mitral orifice, 4 mm. in diameter, leads into the *left ventricle*; this latter is of approximately the normal size, and has a well-developed muscular wall; but in almost all other respects it is a highly abnormal chamber. Its upper sixth part is partially separated from the rest by a prominent oblique muscular ridge (see figs. 1 and 3).

The two parts are very different in appearance and connexions; columnæ carneæ, though small, are prominent features of the ridge and of the upper subdivision of the cavity: certain of them are developed into musculi papillares, which have attached to them the chordæ tendineæ belonging to the two tiny cusps of the mitral valve. But in the lower subdivision muscular columns are absent: the endocardium is smooth, becoming thicker and opaque towards the apex, where also it is somewhat puckered.

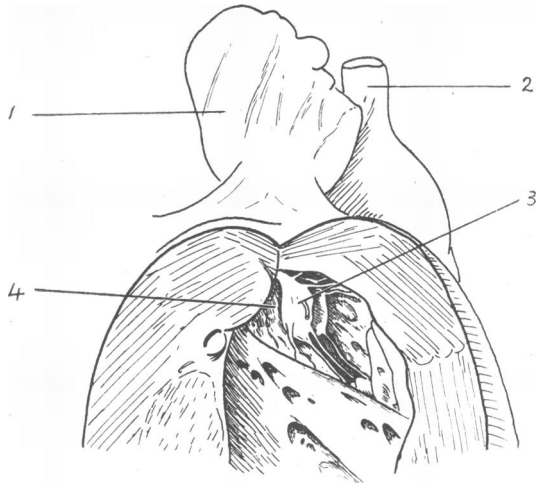

FIG. 3.—Enlarged view of part of the left ventricle, opened widely to show the upper subdivision of the cavity, bounded below by a thick muscular ridge.

1, left auricle, turned up; 2, left superior vena cava; 3, anterior cusp of mitral valve; 4, the blind angle between the anterior cusp of the mitral valve and the uppermost part of the interventricular septum.

The interventricular septum is entire, and of good thickness throughout its extent; the part of it that is related to the upper subdivision of the left ventricle has applied to its right side the septal cusp of the tricuspid valve; on the left side (see fig. 3), where it should lead up to the aortic orifice, no such opening is to be seen, the narrow space between the septum and the anterior cusp of the mitral valve ending in a blind angle above.

The question as to how the ventricle emptied itself is solved by an examination of its lower portion; here, near the apex, the cavity bends forward and to the right, and narrows to a funnel-shaped opening, 2 mm. in diameter, which leads directly into an anterior interventricular channel on the surface of the heart: at the junction of this vessel with the cavity

of the ventricle the channel is slightly narrowed by an irregularly circular thickening of the endocardium (fig. 4).

This remarkable vessel is the only communication between the left ventricle and the aorta; it passes upwards, accompanied by a microscopic cardiac vein, in the anterior interventricular groove, displaying in its interior the origins of several branches which pass obliquely upwards and laterally to the walls of the ventricles: it then opens in a funnel-shaped fashion into the commencement of a well-developed ascending aorta.

This interventricular vessel is, beyond doubt, the anterior inter-

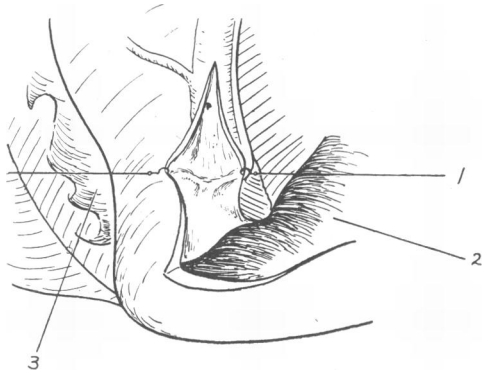

FIG. 4.—Enlarged view of the communication between the apex of the left ventricle and the anterior interventricular coronary artery.

The point of communication is hooked open (at 1) to show the slight transverse constriction; 2, cavity of left ventricle; 3, cavity of right ventricle.

ventricular branch of the left coronary artery; this, not only from its position and connexion with the aorta, its supply of vessels to the heart wall, or its microscopic structure (see fig. 6), but still more from the fact that the main trunk of the left coronary is seen arising from within the funnel-shaped upper end of the vessel, and then running to the left in the auriculo-ventricular furrow (fig. 5).

Examination of the beginning of the ascending aorta places the identity of the coronary vessel further beyond question. The aorta, as has been said, does not communicate with the base of the left ventricle; but its interior, in the situation where the communication should be, shows a shallow depression, divided into two lateral parts by a very low ridge: from the right-hand portion arises a normal right coronary artery. It is impossible to doubt that these shallow depressions represent two of the sinuses of Valsalva; and a less distinct indication of the third is seen where

the left coronary artery communicates with the aorta. The relation of these shallow depressions to one another and to the pulmonary artery will be recognised as being normal.

When this specimen was demonstrated to the Anatomical Society information as to certain points which have since been made clear by further dissection was lacking, and considerable doubt was expressed as to the correctness of the interpretation of the specimen given above. It

FIG. 5.—Enlarged view of the interior of the commencement of the aorta.

1, origin of right coronary artery from one of the rudimentary sinuses of Valsalva; 2, anterior ventricular branch of left coronary artery; within it is seen the opening of one of its branches to the right ventricle; 3, cavity of left ventricle; 4, left auricular appendix, turned aside; 5, left superior vena cava; 6, main trunk of left coronary artery; 7, part of pulmonary artery.

was suggested that the vessel seen in the anterior interventricular groove might not be a coronary artery, but that it might rather represent a part of the bulbus cordis, the aperture near the apex of the left ventricle being the ostium bulbi. Such a suggestion was at the time a tempting one; it gave prospect of a reasonable explanation, based on embryological knowledge, for an otherwise inexplicable condition. Indeed, the striking difference between the two portions of the left ventricle suggested that possibly only the small upper subdivision was really the ventricle proper,

the ostium bulbi being at the level of the muscular ridge marking the communication between the two portions of the cavity (figs. 1 and 3).

Microscopic examination of the ventricular wall from each portion of the chamber does not lend support to this view. Sections were made at the two points marked with crosses on the cut surface of the ventricle in fig. 1; they show no essential difference in structure: near the apex the muscle is much thinner, and, as the naked-eye examination suggests, the endocardium is thick (fig. 7), and beneath it bundles of cellular fibrous

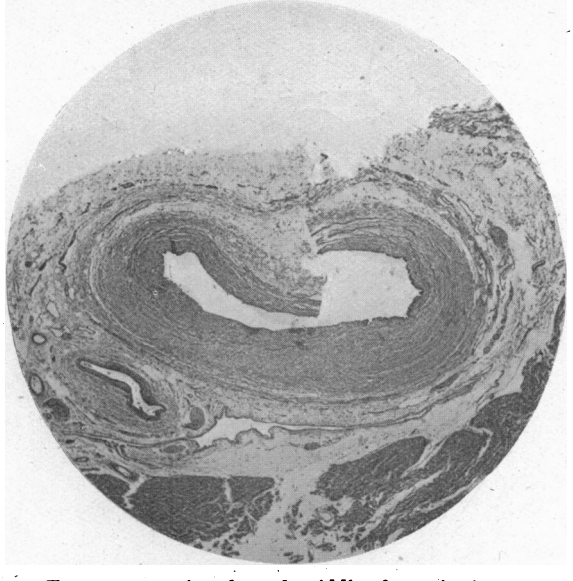

FIG. 6.—Transverse section through middle of anterior interventricular coronary artery and accompanying cardiac vein. × 19.

tissue, with a structure similar to that of the surface endocardium, pass in places between the muscular fasciculi—an appearance which is not seen in the upper section of the ventricle.

But the section of the wall of the left ventricle near its apex is very different from that of the interventricular channel conveying blood to the aorta; the microscope confirms the conviction formed by dissection that this vessel is an artery, the interventricular branch of the left coronary (see fig. 6).

While the connexion of this artery with the left ventricle and the total obliteration of the normal aortic orifice seem at present impossible of satisfactory explanation, certain conjectures may be hazarded.

The two conditions are no doubt interdependent. The aorta did originally arise widely and directly from the ventricle; all that we know of its development, the saccular depression at its commencement, and the rudimentary sinuses of Valsalva, all make this certain. The obliteration, then, was a late process, occurring after the truncus arteriosus had been separated into its pulmonary and systemic parts. Was it the result of the abnormal communication between the coronary artery and the ventricle, or was the aortic orifice first obliterated, and the coronary communication opened up

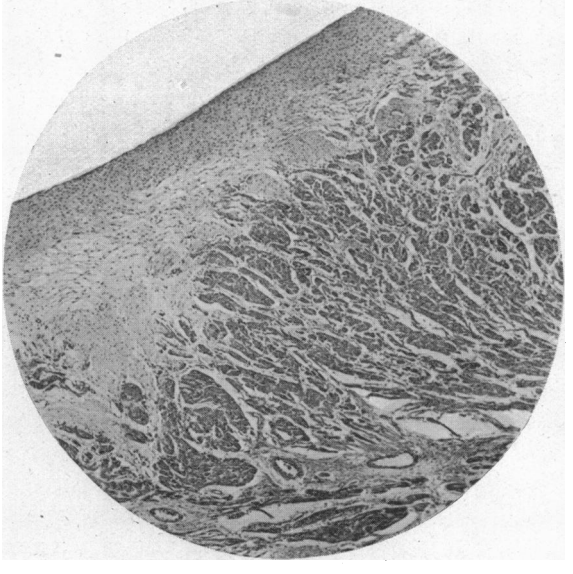

FIG. 7.—Section through wall of left ventricle in the position of the lower of the two crosses seen on the cut surface of the ventricle in fig. 1. × 35.

as a result? If the obliteration of the aortic orifice was the primary condition, we may suppose a pathological inflammatory process to have been probably at work; but perhaps not necessarily so: in the young embryo, the facility with which parts fuse together which before were distinct is only matched by the ease with which solid tissues may become pervious. But our wonder is constantly excited, not only by the rarity with which complex developmental processes go awry, but nearly as much by the fact that when they do so the very malformations are generally subject to laws, and are readily recognised, though not explained, as arrests of development and their results.

No arrest of development will explain the obliteration of the aortic

orifice seen in this heart, for from the earliest stage of its development the aorta and the ventricle are in free communication. The stenosis is an *irregular* abnormality; but since the causes, whatever they may be, which lead to abnormal development do not act entirely capriciously, it would be unreasonable to suppose that this irregular aortic stenosis was in fact capricious. The possibility of the process being syphilitic has to be considered. The mother cannot be traced; but the sections of the coronary artery and of the heart wall do not suggest a syphilitic lesion, and such an explanation would fail to cover the other anomalies seen in the specimen.

Less unlikely, then, than an explanation depending on a merely capricious (because causeless) closure, or upon an inflammatory stenosis of the aorta, is the view that the aperture into the coronary artery from near the apex of the ventricle may have been the primary anomaly, the aortic closure resulting from this.

While no details as to the development of the coronary arteries are yet known, it seems not impossible that during the absorption of the muscular sponge-work in the formation of the ventricular cavities such an abnormal communication between ventricle and coronary arteries might arise: the sharp difference between the upper and lower subdivisions of the ventricular chamber appears susceptible of some similar explanation, *i.e.* that the smaller upper subdivision alone is the ventricle proper, matching the auricle in size, while the lower part is an abnormal cavity formed by the opening up of the muscular sponge-work belonging rather to the right ventricle than the left. The meaning of the obliquity of what has been called above the interventricular septum, and of the extent of the right ventricle behind it to the left border of the heart, would thus become apparent. If then, as in this heart, extreme under-development of the left auricle and ventricle already existed, and the blood-flow through the left side of the heart were consequently much diminished, even such an apparently small cause as the opening up of the ventricle into the coronary artery might produce so great a result as the closure of the orifice of the aorta.