The Hunterian lectures on malformations of the heart : delivered before the Royal College of Surgeons of England on March 8th, 10th, and 12th, 1909 / by Arthur Keith.

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The Hunterian Lectures

ON

MALFORMATIONS OF THE HEART

Delivered before the Royal College of Surgeons of England on March 8th, 10th, and 12th, 1909

BY

ARTHUR KEITH, M.D. ABERD., F.R.C.S. ENG.

HUNTERIAN PROFESSOR, ROYAL COLLEGE OF SURGEONS OF ENGLAND, AND CONSERVATOR OF THE MUSEUM.

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MALFORMATIONS OF THE HEART.

LECTURE I.

Delivered on March 8th.

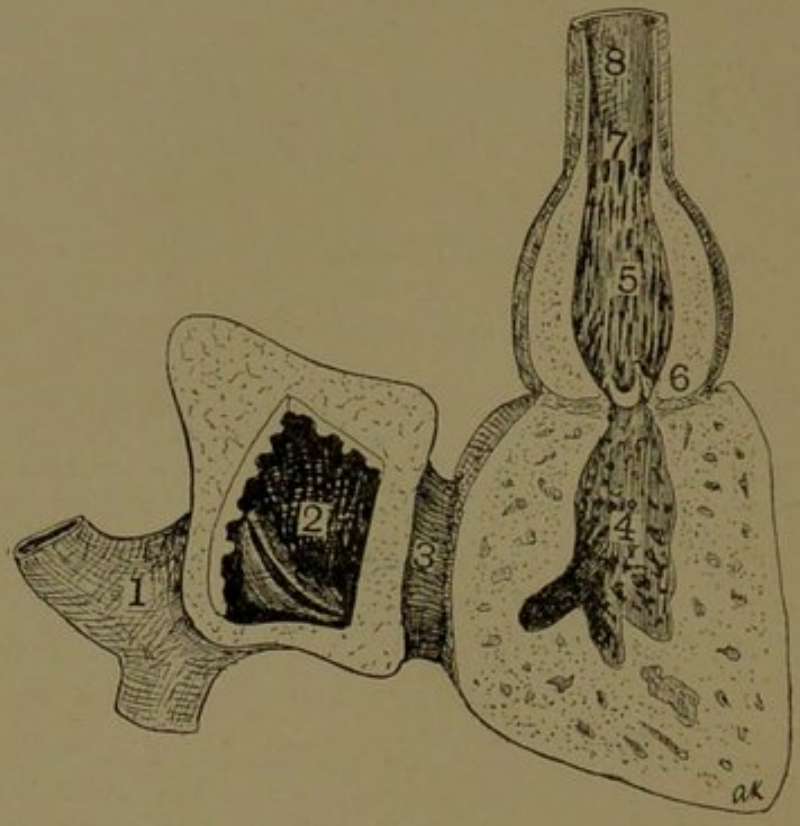
MR. PRESIDENT AND GENTLEMEN,—Although it is 43 years since the last edition appeared, Peacock's treatise on "Malformations of the Human Heart" still remains the standard English work on this subject. It is as fresh as the day on which it was written, for it is founded on accurate personal observation by the bedside and in the post-mortem room. After his death in 1882, the collection of malformed hearts which he had treasured and studied was presented to this Museum and forms a very important part of the series of specimens which I now bring before you as the subject matter of these three lectures.

Our knowledge of the heart has advanced since Peacock's time. One of the greatest discoveries of that period is now only dawning, but every year increases our assurances of its truth—viz., that there is a fourth part or chamber in the mammalian heart which hitherto we have taken no cognisance of. The three parts of the mammalian heart at present recognised are (1) the sinus venosus, (2) the auricles, and (3) the ventricles. The fourth part is the bulbus cordis, which is so well seen in the shark's heart. (Fig. 1.) It is usually supposed that the bulbus cordis has completely disappeared from the mammalian heart, but now we have good reason for believing that, in the same manner as the sinus venosus has become incorporated in the right auricle, the bulbus has become included in the right ventricle, forming that part loosely termed its infundibulum.[1] The

[1] The author has dealt more fully with the nature of the bulbus cordis and its malformation in Studies in Pathology, edited by William Bullock, 1906.

credit of this discovery belongs to Alfred Greil,[2] prosector in the University of Innsbruck. He traced the fate of the bulbus by a prolonged study of the hearts of developing vertebrates. Independently of him I had reached the same conclusion from an investigation of malformed human hearts and of the hearts of vertebrate animals. Professor Peter Thompson [3] has recently identified and described the develop-

FIG. 1.



The four primary chambers of the heart, from a specimen (heart of Xiphias gladius) in the Museum of the Royal College of Surgeons of England. 1, Sinus venosus; 2, auricle laid open showing the venous valves guarding orifice of sinus; 3, auricular canal; 4, ventricle; 5, bulbus cordis; 6, ventricular orifice of bulbus; 7, aortic orifice of bulbus; 8, common ventral aorta leading to gills.

ment of the bulbus in an early human embryo. A large number of the very commonest malformations of the human heart are due to an arrest of the process which ends in the incorporation of the bulbus cordis in the right ventricle. The

[2] Morph. Jahrb. 1903, Band xxxi., p. 123. See also Hertwig's Handb. der vergleich u. experim. Entwickelungslehre der Wirbeltiere, Parts 4, 5, 14, 15, 1903.

[3] Journal of Anatomy and Physiology, 1907, vol. xli., p. 159.

great majority of cases of congenital stenosis of the pulmonary artery are of this nature.

The 55 malformed hearts in the College Museum form the subject matter of my discourse, but my inferences are founded on a wider basis. Thanks to the liberality and courtesy of those who are responsible for the management of the museums of the metropolitan medical schools, I have had an opportunity of examining the specimens in their collections. Through Professor T. Wardrop Griffith—a leading authority on malformations of the heart—I have been able to examine the splendid collection in the museum of the University of Leeds. From these various sources and from my own dissections a series of over 270 specimens is included in the material with which I now deal.

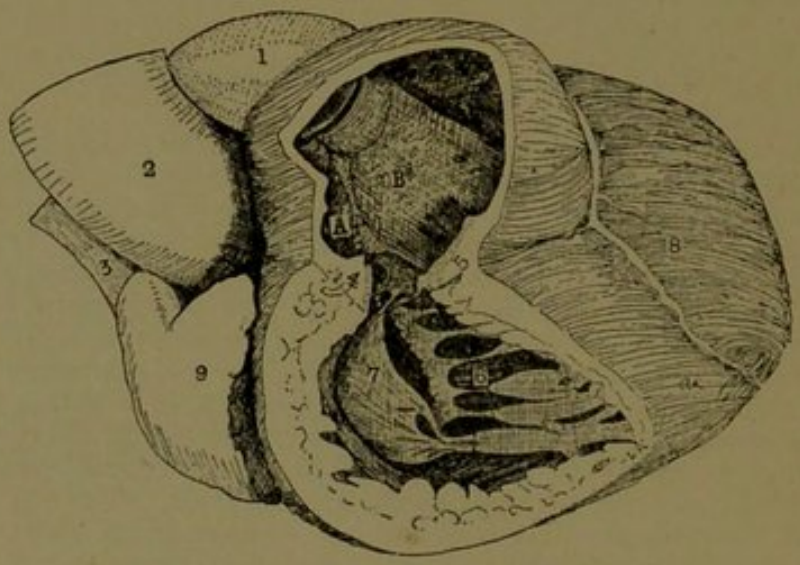
1. Sub-division of the Right Ventricle.

The first malformation I propose to consider is the comparatively rare one usually described as sub-division of the right ventricle. There are three specimens in the museum, and Dr. John Hay has recently presented a fourth. The heart, however, which I propose to describe as the type is one which Dr. Percy Kidd gave me the opportunity of examining. It is shown in Fig. 2. The body of the right ventricle is separated from its infundibulum by a muscular partition, the centre of which is perforated by a foramen large enough to take the little finger. The margin of the foramen is composed of fibrous tissue. The infundibulum is dilated and larger than in the normal heart. The semilunar pulmonary valves are normal. On the septal wall of the infundibulum is seen an important fibrous raphe extending from the semilunar valves above to the constriction or ostium infundibuli below. Below the ostium, in the position of the pars membranacea septi, there is a small foramen—the interventricular—by which blood in the body of the right ventricle could have passed into the left ventricle, or *vice versâ*, during life. There are 19 hearts with malformations of this nature in the series which I have examined. Two of these in the College collection are peculiar; the muscular partition between the two parts of the right ventricle is wholly muscular, and in one the infundibular chamber has reached the apex of the heart. The normal heart shows a muscular constriction at the junction of body and infundibulum of the right ventricle, a fact to which Peacock had drawn attention. The moderator band, which we now know to be a muscular bridge for the conduction and protection of the right septal division of the auriculo-ventricular bundle, forms part of the musculature of the constriction.

The majority of patients with this malformation of the heart live to adult age; indeed, in some the condition is

only discovered after death. The symptoms depend on the size of the ostium infundibuli. In some it measured only from 2 to 4 millimetres in diameter, leading to early death, but in the greater number the orifice measured from 6 to 15 millimetres in diameter. The maximum intensity of the systolic bruit heard in such cases lies to the right of the sternum below the insertion of the fourth costal cartilage. In nine cases there was an interventricular foramen present (see Fig. 2), so that the blood in the body of the right ventricle, obstructed by the narrow ostium infundibuli, might also escape into the left ventricle.

FIG. 2.



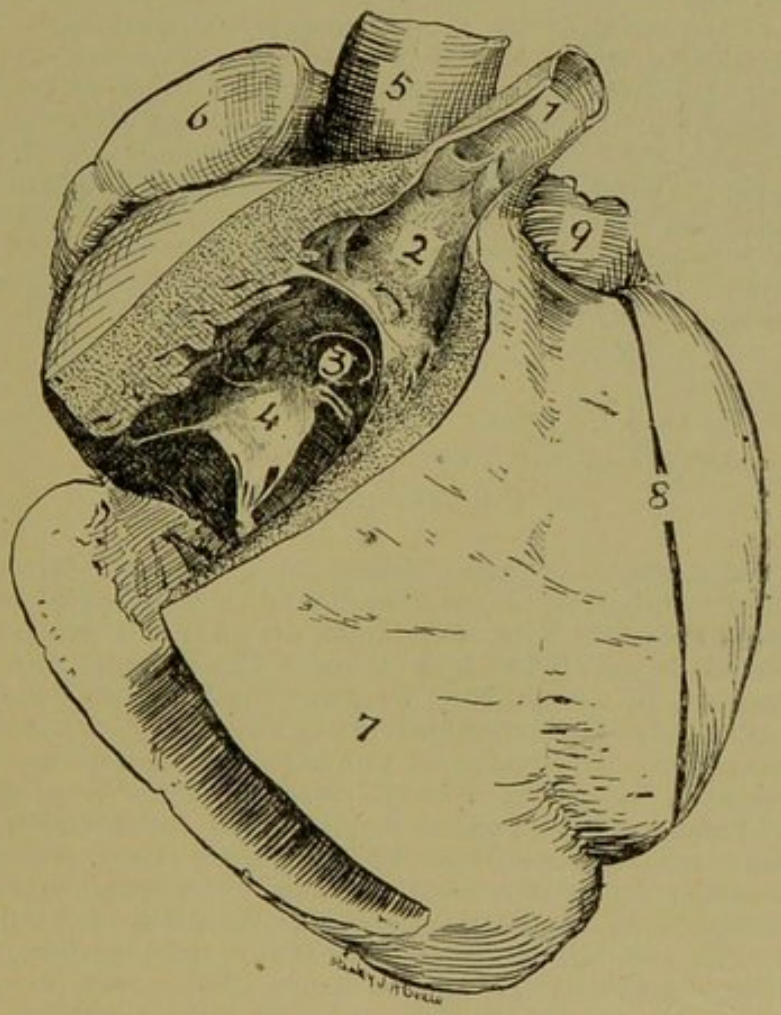
Heart of a man, aged 20 years, who died after two months' illness from acute infective endocarditis, showing subdivision of the right ventricle. 1, pulmonary artery; 2, aorta; 3, superior vena cava; 4, 5, on margins of lower ostium infundibuli—the interventricular orifice is seen between 4 and 5; 6, body of right ventricle; 7, tricuspid valve; 8, left ventricle; 9, right auricle; A, right infundibular band; B, left infundibular band.

In four cases there was also a union or stenosis of the pulmonary semilunar valves; in three the ductus arteriosus was patent and in two the foramen ovale. In quite a large number the fibrous margin of the ostium infundibuli was covered by vegetations—a fact which led to the inference that the fibro-muscular partition between the infundibulum and body of the right ventricle is not a malformation but a cicatricial constriction. It is well known that the right side of the heart becomes more liable to endocarditis than the left if there are obstruction and hypertrophy on that side. The endocarditis is a result, not a cause, of the ostium infundibuli.

2. Cases in which the Developmental Expansion of the Infundibulum is Arrested.

We now pass on to a group of cases which occur much more frequently than those just considered and are usually classified under the term "congenital pulmonary stenosis." In reality they represent a condition in which the arrest in the development of the infundibulum is much more complete than in the last series. The heart shown in Fig. 3 may be

Fig. 3.



Heart of a girl, aged seven years, showing arrest in the expansion of the infundibulum (drawn by Mr. Stanley J. A. Beale). 1, Pulmonary artery laid open; 2, infundibulum separated from body of right ventricle by a fibro-muscular partition which is perforated by the ostium infundibuli; 3, interventricular orifice; 4, tricuspid; 5, aorta; 6, right auricle; 7, right ventricle; 8, left ventricle; and 9, left auricle.

taken as representative of this group—hearts in which there has been an arrest in the developmental expansion of the infundibular cavity. In the type specimen (Fig. 3) the infundibular cavity is fusiform in shape, measuring 12 millimetres from the orifice by which it communicates with the body of the right ventricle (the ostium infundibuli) to the pulmonary valves above. The lining of the infundibular cavity has the appearance of ground glass because the subendocardial tissue is fibro-cellular in texture, and embryonic rather than inflammatory in its microscopic structure. The ostium infundibuli may be as well marked as in the type specimen (Fig. 3), or it may be indicated merely by a circular thickening of the endocardium, or even that indication may be absent. The pulmonary valves in less than half of the hearts of this nature which I have examined—44 in number, of which 16 are in the College collection—were fused to a greater or less degree, the condition of arrest of the infundibulum being thus accompanied by a true pulmonary stenosis. In more than half of these cases—I do not give the exact numbers because in some of the specimens examined the heart was insufficiently opened—an interventricular foramen was present; the size of that foramen was variable. In only a small percentage was there an open condition of the foramen ovale or ductus arteriosus.

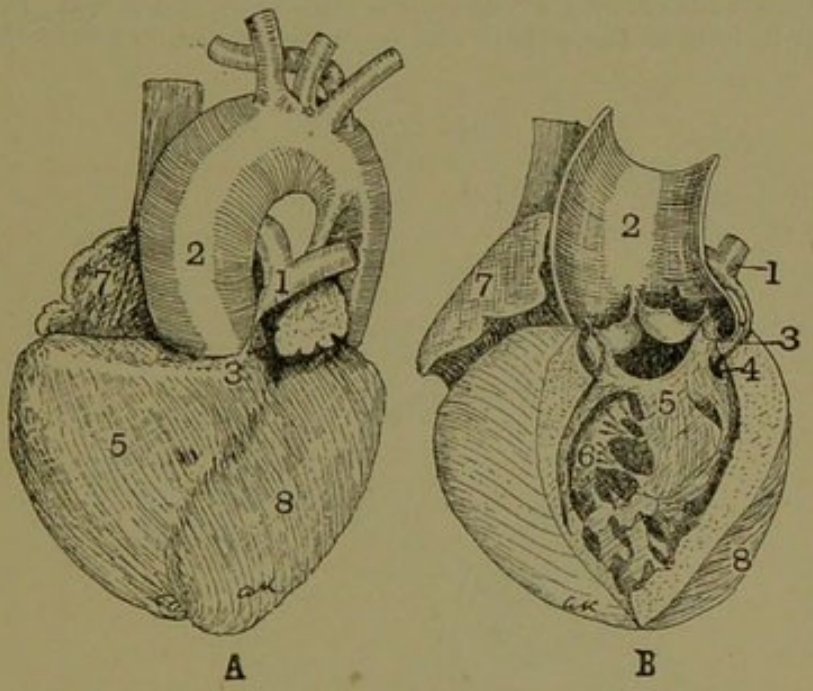
3. Hearts in which there is almost Complete Arrest in the Developmental Expansion of the Infundibulum.

A third group of cases represents an almost complete developmental arrest of the infundibulum. The typical condition is shown in Figs. 4A and 4B, drawn from a specimen presented to me by Dr. J. A. Milne. The infundibulum is represented by a mere slit or irregular chink situated at the orifice of the pulmonary artery and lined by thickened endocardium. The orifice of the pulmonary artery may be represented by merely a small cicatricial mass or a very small lumen may be present in which the fused semilunar valves can be distinguished. The pulmonary artery, perhaps represented by a fibrous thread at its origin, rapidly increases in calibre as it passes into its point of division into right and left branches. An intraventricular foramen is always present, but only in about 30 per cent. of the cases—which numbered 37 in all—was there a patency of the ductus arteriosus.

In such cases the branches of the pulmonary artery are very thin-walled and often greatly dilated. How are they filled with blood in such cases? The orifice of the pulmonary artery is closed or almost closed. Dr. Bertrand E. Dawson gave me the opportunity of injecting the arteries in a case

where there was marked congenital pulmonary stenosis. The bronchial arteries and other accessory branches derived from the intercostals were so enlarged that some of them were equal in size to the radial arteries, but one was especially struck by the number rather than by the size of the accessory arterial communications; they formed

FIG. 4.



Heart of a girl, aged two years, showing complete arrest of the development of the infundibulum. A, Unopened heart viewed from the ventral aspect. B, Right ventricle opened in the same specimen. 1, Pulmonary artery; 2, aorta; 3, position of infundibulum; 4, ostium infundibuli; 5, body of right ventricle and large interventricular orifice; 6, tricuspid; 7, right auricle; 8, left ventricle; and 9, left auricle.

a leash-like plexus. The arteries accompanying the phrenic nerves were greatly enlarged and sent branches to the lungs. In those cases where there is complete arrest of the infundibulum, the arterial system of the lungs is filled from the systemic circulation. To merely live—and that is the condition of patients with such hearts—a wonderfully small supply of oxygenated blood is apparently sufficient.

4. Cases in which the Stenosis is merely due to Fusion of the Semilunar Valves.

In my list of specimens I have noted 23 in which there was apparently no developmental arrest of the infundibulum yet the pulmonary semilunar valves were united, giving rise to a true condition of pulmonary stenosis. Five of the specimens on my list are in the College collection, but when they were taken out of their jars and examined it was seen that in four of them there was an arrested condition of the

Figs. 5 and 6.

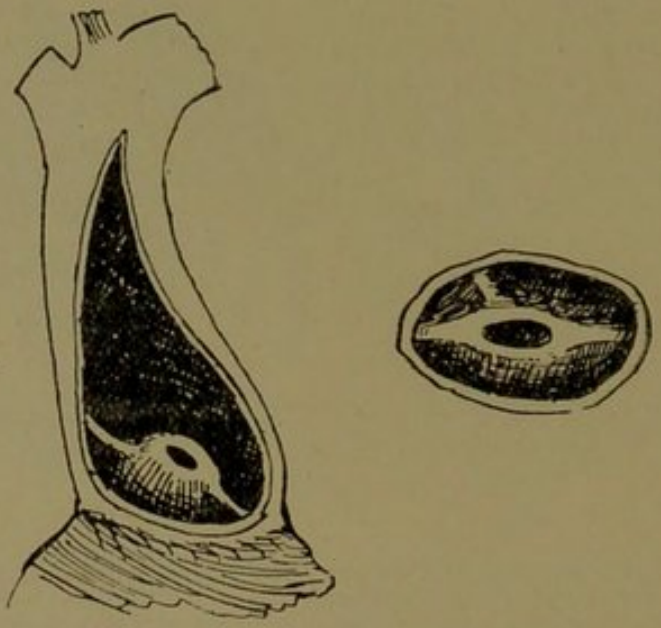


Fig. 5.—Pulmonary valves exposed by opening the pulmonary artery in a case of arrested development of the infundibulum to show the peculiar form of stenosis.

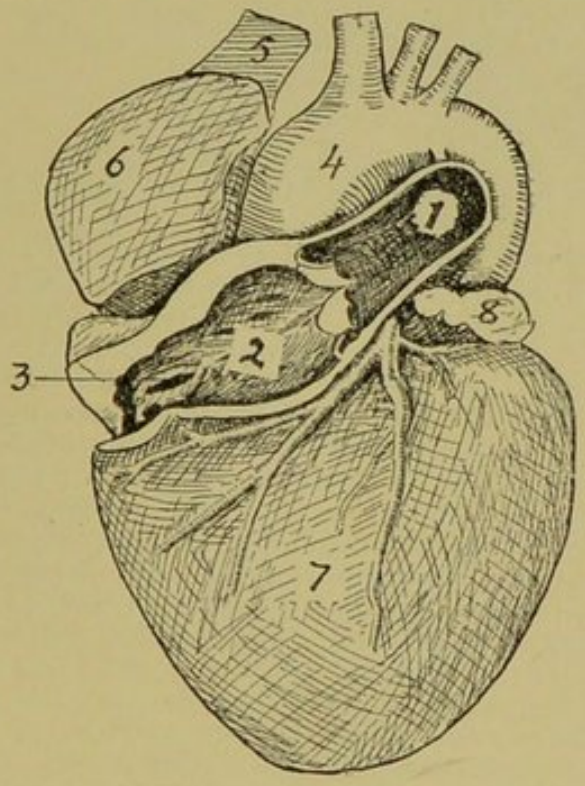
Fig. 6.—The pulmonary valves in a case of stenosis exposed by cutting across the pulmonary artery just above the valves. There is a trace of a third cusp.

infundibulum. It is very probable that if all specimens of this nature were so displayed as to show the infundibular part of the right ventricle there would remain only a few in which pulmonary stenosis is present by itself. Still, the condition does occur and is the only form which one could conceive to be the result of a fœtal endocarditis. In Figs. 5 and 6 is shown the usual condition of the valves. They are fused so as to form a bell-shaped perforated diaphragm. In most cases traces of only two valves can be made out, but occasionally a third can be distinguished.

Many patients who suffer from pulmonary stenosis die

from broncho-pneumonia or other forms of infection ; in such cases it is usual to find the margin of the pulmonary orifice carpeted with vegetations. It is difficult, however, to conceive that the peculiar form of the pulmonary valves in these cases could result from an inflammatory condition in the fœtus. The condition is totally unlike the stenosis which occurs after birth. Under the microscope the valves, in cases of congenital pulmonary stenosis, are seen to be composed of dense laminated fibrous tissue at their margins, but the body of the valves shows a reticulated tissue distinctly embryonic in character.

FIG. 7.



Heart showing infundibulum developed but the body of the right ventricle atrophied. 1, Pulmonary artery opened; 2, infundibulum opened; 3, ostium infundibuli, leading by an interventricular foramen into left ventricle; 4, aorta; 5, superior vena cava; 6, right auricle; 7, left ventricle; and 8, left auricle.

5. CASES IN WHICH THE INFUNDIBULUM IS PRESENT AND THE BODY OF THE RIGHT VENTRICLE IS SMALL OR ABSENT.

The group of cases I am now going to describe illustrates further the dual constitution of the right ventricle. The type specimen of this group is shown in Fig. 7, drawn from

a heart sent to me by Dr. F. S. Mackenzie. Here, while the infundibulum of the right ventricle is well developed the body of that chamber is atrophied or obliterated. The condition is usually accompanied by a congenital stenosis or complete obliteration of the right auriculo-ventricular orifice. In such cases the foramen ovale is patent, so that the venous blood can pass from the right to the left auricle and ventricle. From the left ventricle part of the blood passes into the infundibulum by a patent interventricular foramen. There is also another form in which the infundibular cavity is present and the body of the right ventricle is apparently absent—namely, those cases where the right and left ventricles are unseparated by an interventricular septum. Of the seven specimens belonging to this group five are cases in which the right ventricle is obliterated, two in which the two ventricles form a common chamber.

6. Cases of Subaortic Stenosis.

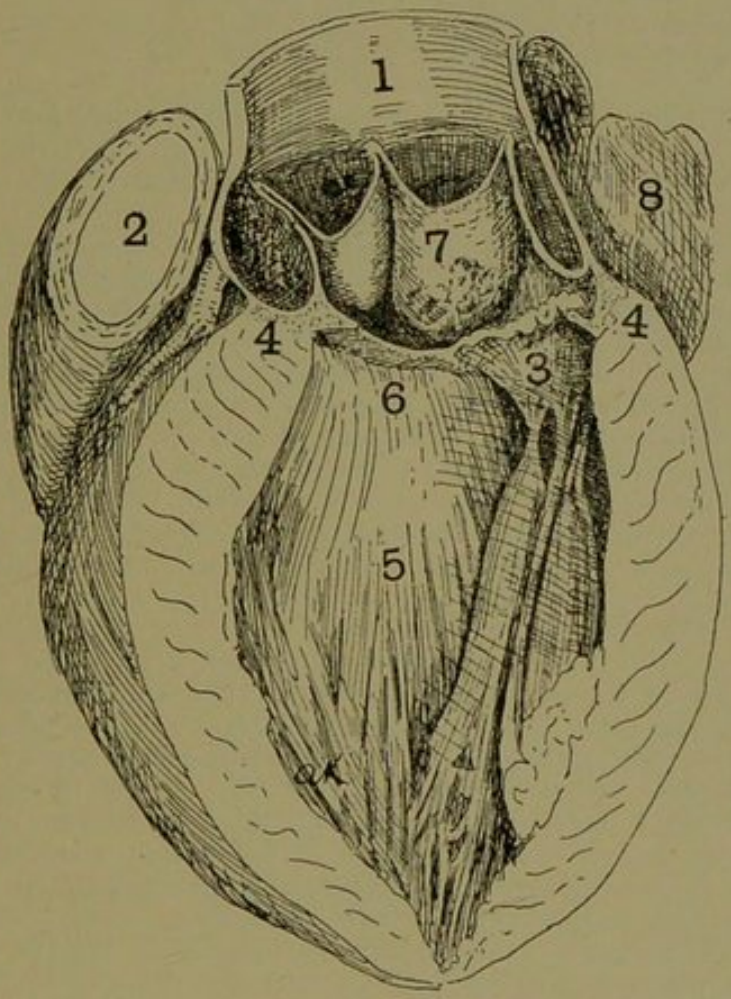
Turning now to the left side of the heart, a condition—very rare, it is true—is found similar in nature to the malformations just described on the right side with this difference. The part of the fœtal heart from which the infundibulum is derived—the bulbus cordis—normally disappears completely from the left side of the heart, but a trace of it may persist giving rise to the condition known as subaortic stenosis—a condition which has hitherto not received a satisfactory explanation. Thus, while the abnormality on the right side of the heart depends on an arrest of growth, that on the left side depends on an arrest of atrophy. The type specimen, which belonged to the museum of the London Hospital, and was lost in the disastrous fire at Toronto University, is shown in Fig. 8. Just below the orifice of the aorta a fibrous collar is seen to surround the infundibulum of the left ventricle. It is formed by an irregular thickening of the endocardium. The ostium infundibuli is in every way homologous to the corresponding abnormal constriction which occurs in the right ventricle. I have seen only four hearts showing subaortic stenosis—one in the museum of St. George's Hospital, one in St. Bartholomew's, one in Guy's, and the one figured here. We have, unfortunately, no specimen in the College collection.

7. Cases of Congenital Aortic Stenosis.

The condition of congenital aortic stenosis is rare in England. Altogether I noted only four cases in the museums of the metropolitan medical schools. The malformation is apparently much more frequently seen in St. Petersburg, for

in 106 malformed hearts Therémin found this abnormality 17 times. Among 14 stillborn children—all of them showing marked deformity of the body with malformation of the heart—I observed aortic stenosis in only three of them, two of these being in children of Polish Jews.

FIG. 8.



Heart showing subaortic stenosis (heart of man, aged 20 years). 1, Aorta; 2, pulmonary artery; 3, mitral valve; 4, 4, fibrous constriction surrounding aortic vestibule; 5, interventricular septum; 6, upper part of septum below ring; and 7, non-coronary cusp of aortic orifice, site of endocarditis.

SUMMARY OF CASES OF MALFORMATION OF THE BULBUS CORDIS.

So far I have dealt with malformation of the heart in the vicinity of the origins of the pulmonary artery and aorta—

the part into which the bulbus cordis is incorporated. The cases fall into seven groups :—

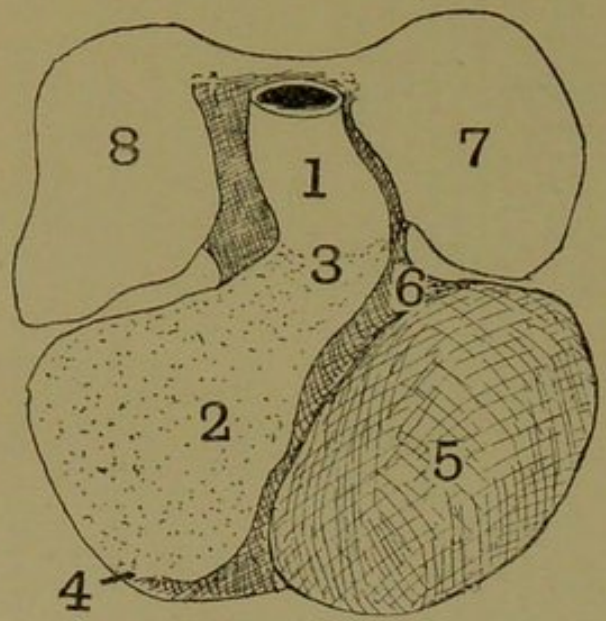
1.	Incomplete fusion of the infundibulum with the body of the right ventricle	19 cases.
2.	Partial arrest in the development of the infundibulum	44 ,,
3.	Complete arrest of the infundibulum...	37 ,,
4.	Fusion of pulmonary semilunar valves	23 ,,
5.	Partial or complete absence of the body of the right ventricle with development of the infundibulum ...	,,
6.	Subaortic stenosis	4 ,,
7.	Aortic stenosis	7 ,,

Altogether 141 cases out of a total of 272 malformed hearts are imperfect because of a lesion connected with the transformation of the bulbus cordis during the development of the embryo.

Applying now our present knowledge of the development and comparative anatomy of the heart to the explanation of these abnormalities, we find ourselves hampered at the very outset by a complete ignorance of the functional nature of the bulbus cordis. It is especially large in the shark tribe and is often lined with valves and thickened endocardium throughout (Fig. 1). Its musculature is striated but the fibres are more fusiform and less branched than in the other chambers. The nerve-supply is abundant. Systole occurs in it subsequent to contraction in the ventricles. Somehow its function is connected with the gill or respiratory system ; it is with the respiratory system that it is correlated. When the gills become replaced with lungs and the aortic stem divided into systemic and pulmonary trunks as in the amphibia, the part of the bulbus at the origin of the aorta atrophies more than the part connected with the pulmonary artery. In reptiles a part of the bulbus musculature persists and can be distinguished from the proper ventricular muscle ; I have also seen it well marked in an abnormal human heart. In the hearts of birds and mammals the bulbus musculature completely disappears, but its cavity persists, undergoes a great expansion, and forms the infundibulum of the right ventricle. How far the infundibulum is formed by a downward expansion of the bulbus cavity into the right ventricle and how far it is formed by an upgrowth over it of the ventricular muscle is difficult at present to decide, but the evidence of malformed hearts points to the former process being the true one. What the exact function of the infundibulum of the mammalian right ventricle may be is difficult at present to say, but its origin, its comparative anatomy, and the arrangement of its musculature make it certain that it has something to do with the regulation of the blood-supply to the lungs. There are many clinical phenomena connected with this part of the heart which need further observation and explanation.

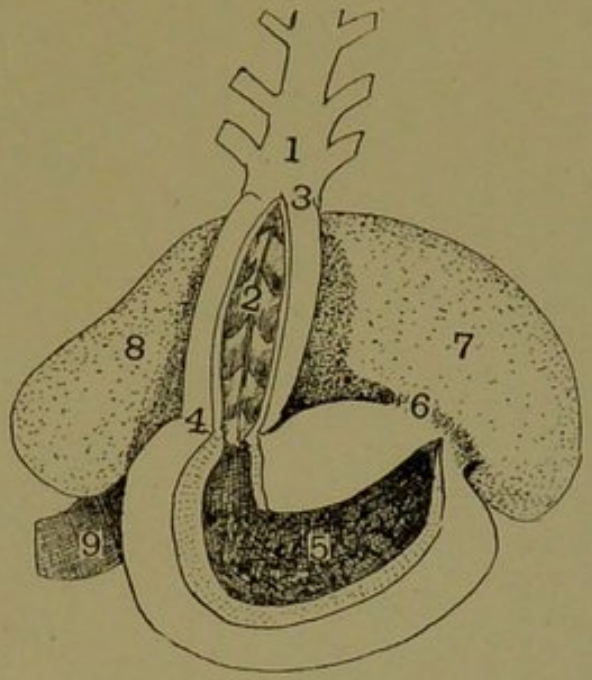
The embryological transformation which occurs at the aortic

FIG. 9 A.



Heart of a human embryo about three weeks old. (After His.) 1, undivided, ventral aortic stem; 2, bulbus cordis; 3, aortic orifice of the bulbus; 4, lower or ventricular orifice of bulbus; 5, left ventricle; 6, auricular canal; 7, 8, left and right auricle.

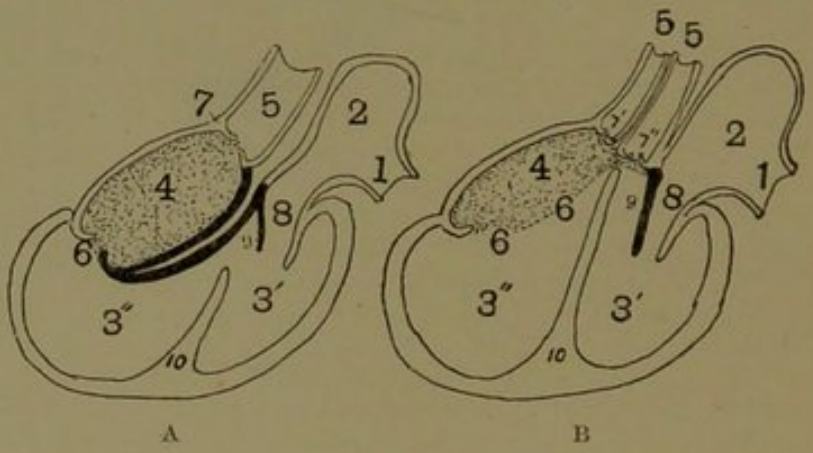
FIG 9 B.



Heart of shark (Echinorhinus spinosus) to compare with the heart of the human embryo (Fig. 9A). 1, common ventral aortic stem; 2, bulbus cordis; 3, upper orifice; 4, lower orifice; 5, common ventricle; 6, auricular canal; 7, 8, left and right auricle.

end of the cardiac tube during the latter part of the first, and earlier part of the second, month of development is very remarkable. In Fig. 9, A and B, are shown side by side the heart of a human embryo, probably about 21 days old, reconstructed and described by the late Professor His, and that of a shark. The bulbus cordis, from which the primitive aorta springs, is still well demarcated from the ventricular part of the tube—just as in the shark's heart. The ventricle at this time has, like the stomach, a greater and lesser curvature. As development proceeds a rapid atrophy of the lesser curvature sets in, so that the bulbus comes in contact with the auricular canal, just as an atrophy of the lesser

FIG. 10.



To show the manner in which the bulbus cordis becomes incorporated in the mammalian heart and forms the infundibulum of the right ventricle. A. The bulbus as a separate chamber. B. The bulbus as incorporated in the right ventricle. 1, orifice of sinus venosus; 2, auricles; 3′ 3″, left and right ventricles; 4, bulbus cordis; 5, common aortic stem; 6, ventricular orifice of bulbus (in B it becomes stretched out during the atrophy of the lesser curvature, indicated by thick black line); 7, 7′, 7″, aortic orifice of bulbus; 8, auriculo-ventricular orifice; 9, anterior cusp of mitral; 10, interventricular septum.

curvature of the stomach would bring the pylorus against the œsophagus. The atrophy also involves the corresponding or lesser curvature of the bulbus. When the primitive aorta becomes divided into aorta and pulmonary artery, the aorta is found to be connected with the lesser curvature of the bulbus and ventricle—where the bulbus atrophies, while the pulmonary artery, being situated towards the greater curvature, is connected with the part which persists.

The diagram given in Fig. 10 will help to make this transformation better understood. In the shark's heart one recognises the constriction or orifice between it and the common

ventricle—the ostium bulbi. The ostium bulbi is usually provided with valves; the pulmonary and aortic semilunar valves represent those at the aortic orifice of the shark's bulbus. In the process of development, as Fig. 10 shows, the ostium bulbi becomes stretched over the upper margin of the interventricular septum; the half remaining on the right side of the septum forms the ostium infundibuli and the half to the left may persist and form the subaortic constriction. The embryological and comparative evidence make it certain that the bulbus is incorporated in the ventricle of the heart; the ostium infundibuli in the right ventricle of abnormal hearts and the subaortic constriction on the left occupy the position at which the ostium bulbi should occur. In brief, the theory of the bulbus cordis explains a great number of abnormal conditions hitherto inexplicable.

LECTURE II.

Delivered on March 10th.

Transposition of the arterial stems.—If the theory of a bulbus element in the human heart helps us to understand the numerous abnormalities which were discussed in my last lecture, it is of even greater assistance in clearing up the various abnormal and very puzzling conditions included under the term of transposition of the aorta and pulmonary artery. The pulmonary artery in these cases of transposition arises behind the aorta, over the left ventricle, while the aorta springs from the infundibulum of the right ventricle. Thus the venous blood is thrown by the right side of the heart into the aorta and systemic circulation while the arterial blood is carried back to the lungs by the pulmonary artery. Altogether I have examined 25 hearts in which the arterial trunks were transposed, six of them being in the College collection. Fig. 11 shows the arrangement of parts in a specimen which may be regarded as typical of the group. The aorta, recognised not only by its distribution but also by the fact that the coronary arteries are given off from it, arises from the infundibulum of the right ventricle—that part of the ventricle which is derived from the bulbus cordis. The pulmonary artery occupies the position of the aorta and springs from the left ventricle. The exact condition of parts is best seen by examining a section made at the base of the heart (Fig. 12). In the normal heart the aortic orifice is situated behind the pulmonary orifice and immediately anterior to the left auricle and left auriculo-ventricular orifice. The aortic cusps corresponding to the origin of the right and left coronary arteries—right and left coronary cusps—are situated towards the sternal margin of the aorta, the non-coronary towards the dorsal margin. In the abnormal heart the position of parts is reversed; now the aorta is in front and the pulmonary artery behind; the position of the aortic cusps is reversed, the non-coronary cusps being now not on the dorsal but on the sternal margin of the aorta. Further, it is to be noted that the aorta arises from the infundibulum or persistent bulbus cordis, while the pulmonary artery arises directly from the left ventricle, the part of the bulbus corresponding to it having atropied, but the atrophy is sometimes only partial.

How is transposition brought about? The explanation offered by Rokitansky—the only explanation yet given—is that the aortic end of the heart may undergo an abnormal

degree of rotation, either to the right hand or to the left, and the form of transposition depends on the extent of the rotation. Such a theory does not really help us. The clue to the problem is to be found in a study of the remarkable transformation of the mammalian embryonic heart. We

FIGS. 11 AND 12.

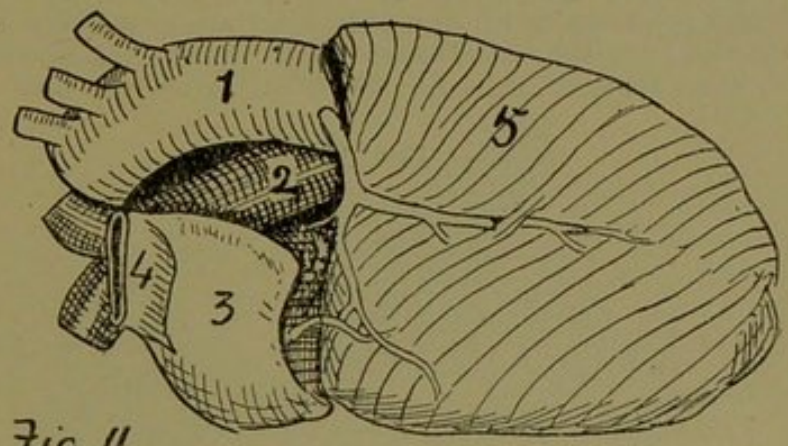


Fig. 11

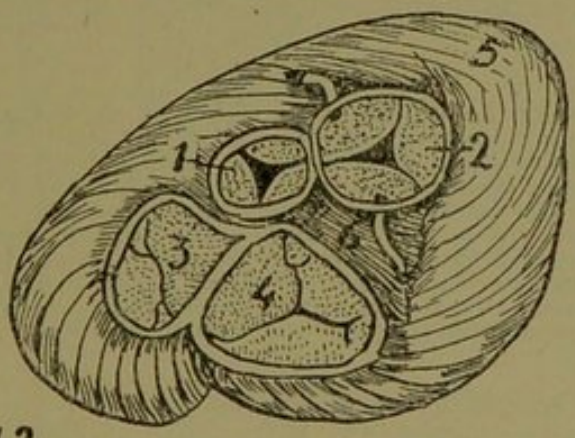


Fig. 12.

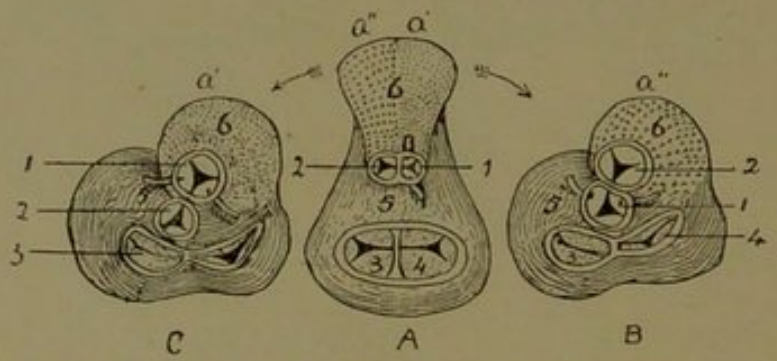
FIG. 11.—Heart showing transposition of the great stems (viewed on its right side). 1, Aorta; 2, pulmonary artery arising behind aorta; 3, right auricle; 4, superior vena cava; and 5, infundibulum of right ventricle giving origin to aorta.

FIG. 12.—The base of the heart shown in Fig. 11. 1, Pulmonary artery; 2, aorta (non-coronary cusp); 3, anterior cusp of mitral valve; 4, septal cusp of tricuspid; 5, infundibulum of right ventricle; and 6, triangle of persistent basal musculature.

have already seen how the lesser curvature of the bulbo-ventricular segment of the cardiac tube shrivels up during the early period of development. By the third week the ventricular segment of the primitive cardiac tube is bent transversely towards the right (Fig. 9, Lecture I.), so that it

occupies a position comparable to the adult stomach, the auricular canal representing the œsophagus and the bulbar part the pyloric end. In Fig. 13 A I have represented what I conceive to be the primitive form of the bulbo-ventricular part of the heart; in Fig. 13 B is shown the atrophy that leads to the aorta being brought into juxtaposition with the mitral orifice; and in 13 C the changes which lead to the pulmonary artery being brought into contact with that orifice. In the first case the bulbus connected with the aorta atrophies and the part connected with the pulmonary artery undergoes an expansion; in the second case the processes are reversed. The aorta instead of being the dorsal vessel is now the

FIG. 13.

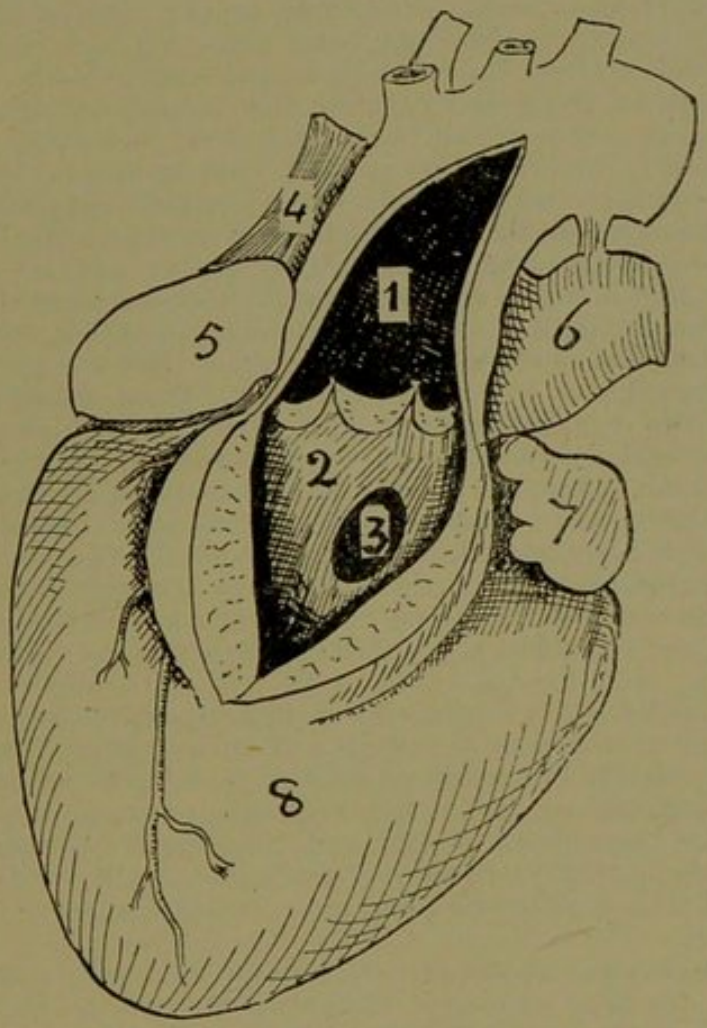


Showing the atrophy of the primitive cardiac tube which leads to the normal position of the arterial orifices (B) and to the abnormal position (C). A, Primitive ventricle and bulbus cordis viewed on the aspect of their lesser curvature. B, The normal position of the arterial orifices. C, The abnormal position. 1, Aortic orifice; 2, pulmonary orifice; 3, mitral orifice; 4, tricuspid orifice; 5, body of common ventricle; and 6, bulbus cordis. a', Right half of bulbus which atrophies in B, the normal form; a'', left half of bulbus which atrophies in C, the abnormal form.

ventral and situated towards the greater instead of towards the lesser curvature (Fig. 13 C). With the atrophy of the lesser curvature of the ventricle and pulmonary part of the bulbus, it is the pulmonary artery in place of the aorta that comes into contact with the auricles. In effect, the origin of the great vessels is reversed as regards the auricular canal (compare 13 B with 13 C). No one has seen an embryo in which the pulmonary part of the bulbus was undergoing a process of atrophy in place of one of expansion, but the assumption is justified by the fact that a complete explanation is thereby obtained of the condition seen in hearts with transposed arterial trunks. The theory explains why the aorta is in front of the pulmonary artery and also the reversion in the position of its valves. We see further why the bulbus cordis persists in connexion with the aorta and forms the infundibulum, while the part of the bulbus connected with the pulmonary artery atrophies.

Of the 25 cases of transposition 19 were of the type just described ; there are at least two modifications of this type. In one of these modifications the interventricular septum is so ill-developed that there may be said to be a common ventricle. Four such cases are included in my list. Professor

FIG. 14.



Transposition of the arterial stems. (After Rokitansky.) 1, Aorta ; 2, separate small cavity from which the aorta springs, probably derived from the bulbus cordis ; 3, orifice leading into common ventricle (the ostium bulbi); 4, superior vena cava ; 5, right auricle ; 6, pulmonary artery ; 7, left auricle ; and 8, common ventricle.

A. H. Young and Professor A. Robinson have lately described a fine example of this modification. The pulmonary artery arises directly from the common ventricle, while the aorta arises from the infundibulum or bulbus cordis, which communicates with the common ventricle by an orifice varying much in size and distinctness (Fig. 14). Another and still

rarer form of transposition—I have seen only two cases—is that in which the aorta arises from the left ventricle and the pulmonary artery from the right, but the origin of the pulmonary artery is situated behind the aorta and has no infundibulum or bulbus ; the aorta, although situated in front of the pulmonary artery, arises from an infundibulum attached to the left ventricle.

In transposing the great arterial stems Nature tries an impossible experiment ; two independent circulations are thereby arranged, the right heart supplying and receiving the systemic blood, the left side the pulmonary blood. Were the septa of the heart complete the child would die from suffocation as soon as born. But the septa are never complete. There are four possible communications in such cases between the pulmonary and systemic circulations : (1) the foramen ovale may remain open or some other defect may be present in the interauricular septum ; (2) an interventricular foramen may persist ; (3) the ductus arteriosus may remain open ; and (4) the communication between the bronchial and pulmonary arteries and veins may be very free. By one or more of these communications the venous and arterial bloods are allowed to mingle sufficiently to carry on the respiratory needs of the system. The prospect of life depends on the freedom with which such an interchange takes place. Indeed, the cases which live longest are those in which the two ventricles communicate so freely that they may be regarded as a common chamber ; in some cases of this nature the patient has passed into adult life without his defect being discovered and may reach the age of 45 or 50 years. In most cases of transposition life is supported with difficulty and the slightest affection of the lungs is sufficient to cause death. The apex of the heart is in its normal position, and I know of no case in which the condition has been diagnosed during life. Yet the superficial position of the aortic valves, the great hypertrophy of the infundibulum ought to give a second sound which is peculiar as regards character and position.

With transposition of the stems there is usually a greater or less degree of stenosis of the pulmonary orifice. In 17 of the 25 cases there was pulmonary stenosis ; the stenosis is not due merely to a union of the semilunar valves ; there is commonly also a remnant of the bulbus below the orifice corresponding to sub-aortic stenosis. That the pulmonary artery should be so liable to stenosis when there is transposition is a most interesting fact, for it occupies the position normally taken by the aortic orifice which is so rarely occluded. The inference I draw from the fact is that stenosis is more likely to occur in a side stream than in a main stream ; the pulmonary circuit is a secondary one in the fœtus ; the aortic is the primary one. In other words, congenital stenosis is to some extent the result of a mechanical condition.

Reversion of the heart.—Transposition of the great trunks must not be confused with a true reversion or transposition of the part of the heart seen in cases where the viscera are transposed. I have seen four examples of this condition, one of which is in the College collection. McCrae has recently described a most instructive case in which the reversion of parts was accompanied by complete stenosis of the infundibulum. Reversion of the heart is frequently accompanied by irregularities in the arrangement of its parts.

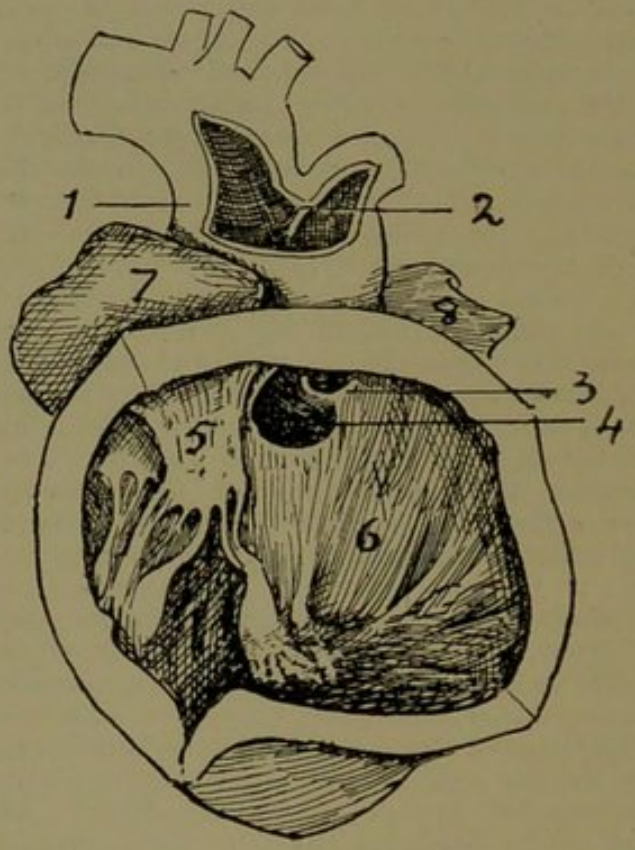
Imperfect separation of aorta and pulmonary artery.—All the defects so far described, and they make up a great majority of cases which need medical assistance, are related to a maldevelopment or irregular incorporation of the bulbus cordis in the heart. We now pass on to another structure which cannot rightly be said to be part of the heart—the truncus arteriosus, which becomes divided to form the root of the aorta and of the pulmonary artery. Irregularities in the division of the truncus arteriosus are rare; I have only seen three cases, two of them in Guy's Hospital Museum. There is no specimen in the College collection. The illustration I have used (Fig. 15) is borrowed from Rokitansky's work, and shows the commonest form where the truncus has not been divided, the pulmonary arteries springing directly from the ascending aorta. In one of the specimens at Guy's Hospital there is a perforation of the septum between pulmonary artery and aorta, which would have the same functional effect as a patent ductus arteriosus.

Patent ductus arteriosus.—A patent ductus arteriosus is usually correlated with some malformation of the heart, but cases do occur in which—as far as one can see—there is no other lesion. I have seen only two such cases. We have no specimen of the condition in the Museum collection, nor have we any specimen of an aneurysm of the ductus, which apparently is not a rare condition. Gibson of Edinburgh has pointed out that the thrill and bruit caused by a patent ductus persists into the diastolic pause—unlike a murmur of intracardiac origin.

Malformation of semilunar valves.—Although abnormalities of semilunar valves are very common—altogether I came across over 50 museum specimens—their study has been peculiarly barren of any useful knowledge. This is largely owing to the fact that many specimens are put up in such a way that it is impossible to know the relationship that existed between the pulmonary and aortic semilunar cusps when the parts were *in situ*. The normal relationship of the six cusps is shown in Fig. 13 B. It will be remembered that, on the division of the truncus arteriosus, two of its four valves are included in the division, giving rise to the two

non-coronary cusps in the aorta and the septal or posterior cusps of the pulmonary artery. In 25 cases where it was possible to distinguish the septal cusps from the others it was seen that the right septal cusp of the pulmonary artery was subdivided in six cases, the left also in six, so that in these 12 cases there were four cusps. In one case the non-septal or anterior cusp was absent. In the aorta the non-coronary cusp was absent in one, small in one, and sub-

FIG. 15.



Incomplete separation of pulmonary artery from ascending aorta. (After Rokitansky.) 1, Aorta; 2, pulmonary artery; 3, lower end of imperfect septum beween aorta and pulmonary artery; 4, interventricular foramen; 5, tricuspid valve; 6, interventricular septum; 7, right auricle; 8, left auricle.

divided in four. The right coronary cusp was divided in two cases and united with the left coronary in three.

Imperfect formation of septa.—Having passed in rapid survey the numerous malformations which occur at the aortic or distal end of the heart, I propose now to examine the various defects found in connexion with the development of the septa of the heart. The form of abnormality I shall

take first is the comparatively rare one where the septum between the right and left auriculo-ventricular orifice is incomplete. There are two specimens illustrating this condition in the College collection; I have seen 14 altogether, the lesion being remarkably alike in all. Professor T. Wardrop Griffith has described two cases. When the auricles of such a heart are opened (see Figs. 16 and 17) the anterior cusp of the mitral is seen to be continuous with the anterior or infundibular segment of the tricuspid, while the posterior cusp of the mitral is continuous with the mesial or septal segment of the tricuspid. When the left auricle and ventricle are

FIG. 16.

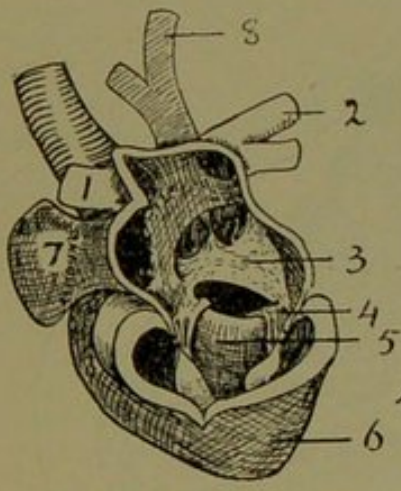


FIG. 17.

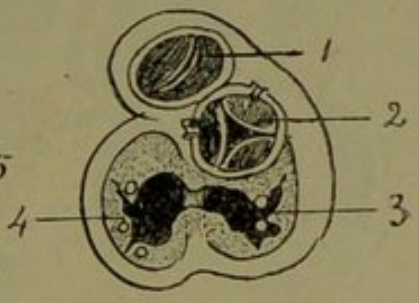


FIG. 16.—Heart of child showing a communication between the auriculo-ventricular orifices due to non-union of the endocardial cushions. 1, 2, Left and right pulmonary veins; 3, interauricular septum; 4, continuity between posterior cusps of mitral and tricuspid valves; 5, interventricular septum, forming lower margin of the abnormal foramen; 6, left ventricle; 7, left auricle; and 8, superior vena cava.

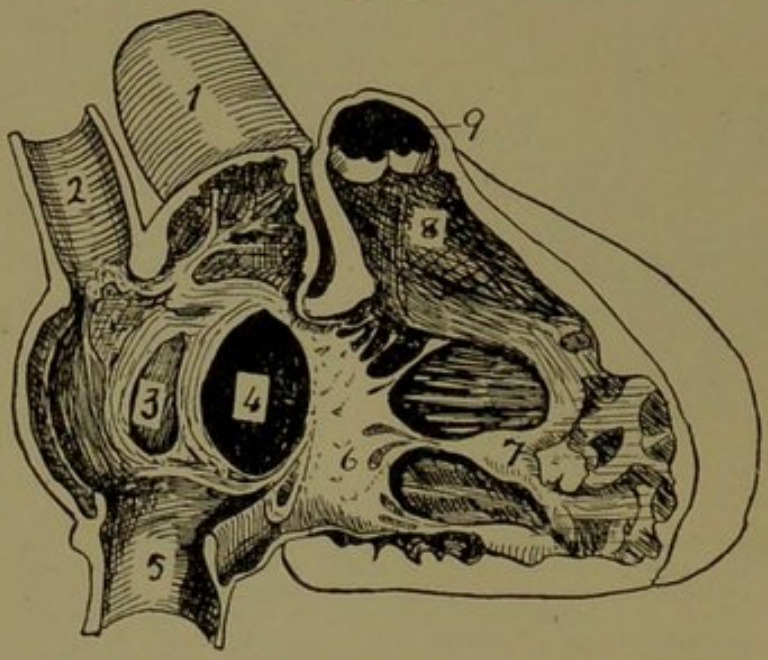
FIG. 17.—Section of base of same heart to show continuity of mitral and tricuspid valves. 1, Pulmonary artery; 2, aorta; 3 and 4, tricuspid and mitral valves.

opened the appearance seen is that represented in Fig. 16. The common auriculo-ventricular anterior and posterior curtains are draped over the interventricular septum, the parts of the valves situated on the septum being puckered and warty in appearance. These parts which in the normal course of development should have fused and joined in the formation of the central fibrous body of the heart, have remained apart, recalling the condition of hare-lip, where also the normal union of adjacent parts has failed.

The condition of parts seen in such specimens is directly comparable to that which occurs in the hearts of amphibia

and dipnoi—the air-breathing fishes. The common auriculo-ventricular orifice in these vertebrates is furnished with four valves—two small lateral and two larger valves, one on the dorsal, the other on the ventral side of the orifice (Fig. 17). In the human embryo the dorsal and ventral valves are named endocardial cushions; by the end of the fifth week, when the embryo is about 5 millimetres long, the central parts of these cushions or valves, lying directly over the interventricular septum, have fused, offering a substratum where the musculature of the interauricular and inter-ventricular septa may obtain a common point of purchase.

FIG. 18.



Right chambers of a child's heart exposed to show the foramen primum. 1, Aorta; 2, superior vena cava; 3, fossa ovalis; 4, foramen primum; 5, inferior vena cava and Eustachian valve; 6, septal cusp of tricuspid; 7, musculus papillaris of right ventricle; 8, infundibulum; and 9, pulmonary artery.

The condition of the interauricular septum in such abnormal hearts is shown in Fig. 16; the lower margin of the septum forms a crescent, which arches from the dorsal to the ventral valve—a condition exactly similar to that seen in the frog. Non-fusion of the endocardial cushions produces an amphibian condition; the free communication thus established between the two sides of the heart is interventricular, not inter-auricular. Non-fusion of the endocardial cushions is never present alone; it is associated with some grave lesion, either transposition of the great trunks or stenosis of the pulmonary artery. Death usually occurs in childhood. I have not seen the lesion in the heart of an adult.

The foramen primum.—The defect in the septum just considered leads naturally on to the next form, one in which the endocardial cushions have fused but are still separated from the crescentic lower margin of the interauricular septum by a space at which the blood in the two auricles may become mixed. This deficiency in the interauricular septum is known as the foramen primum. There is no specimen in the College Museum ; I have seen five cases ; the one from which the illustration is taken (Fig. 18) occurred at the London Hospital. Professor Wardrop Griffith and Professor Peter Thompson have given good descriptions and figures of this defect. It is not necessarily accompanied by any grave disturbance in the action of the heart.

Patent foramen ovale.—Patency of the foramen ovale is so common that I have excluded it from my list of heart malformations. In a combined investigation by Professor E. Fawcett and Dr. J. V. Blachford the foramen was found more or less open in 96 out of 306 individuals, all of them above the age of ten years. They found that it was open more frequently in the old than in the young. I have observed that patency of the foramen ovale is very frequent in those cases where the right auricle has become dilated and the septum expanded by any condition which leads to a distension of the right side of the heart. To what extent such cases are relieved by the blood in the right auricle being permitted to escape to the left or *vice versâ* there is no clinical evidence to show, but one would infer that it is an advantageous condition.

LECTURE III.

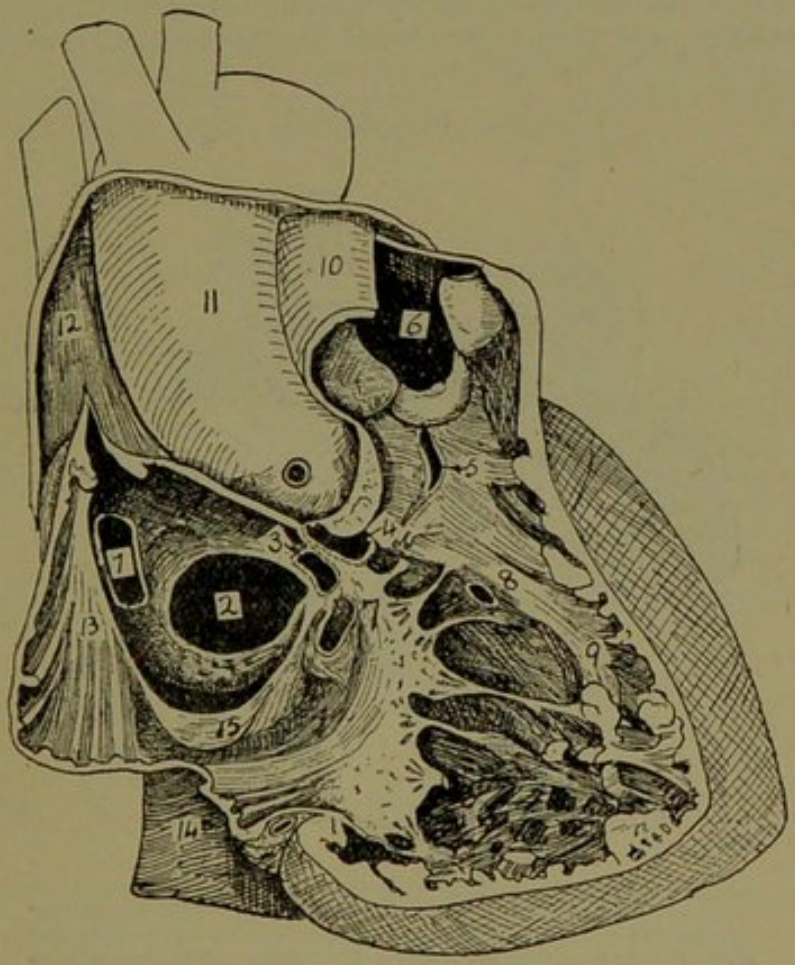
Delivered on March 12th.

Absence of the septum ovale.—The condition I now proceed to describe to you, although frequently regarded as patency of the foramen ovale, is really a more serious defect. The malformation is shown in Fig. 19; we have no specimen in the College collection and I have seen only five examples. Not only is the foramen ovale open, but the septum ovale—that part of the interauricular septum which forms the floor of the fossa ovalis—is absent, so that there is a large circular window between the two auricles. The lower margin of the orifice is formed by a septal strand which can be traced to the valves bounding the orifice of the coronary sinus, which it will be remembered are derivatives of the first chamber of the heart—the sinus venosus. The condition is interesting because it bears out the statement made by His, and afterwards denied by Born, that in the lowest part of the interauricular septum there is an element derived from the sinus venosus (Fig. 20). It is also important because the node or small mass of peculiar muscle in which the auriculo-ventricular bundle commences is found in the region of the interauricular septum formed by the sinus element. Indeed, Retzer regards the node as really composed of sinus musculature and the bundle and its branches as outgrowth from the node—a view which is not only in opposition to all that is known of the comparative anatomy of the bundle, but also unsupported by embryological evidence. In cases of absence of the septum ovale the element derived from the sinus and described by His under the name of spini vestibuli is present, while the real interauricular septum, at least the part which forms the septum ovale, is absent. In a model prepared by His the spini vestibuli grows out from the lower fornix of the venous valves which guard the orifice of the sinus venosus, and reaches the endocardial cushions to fuse with the lower margin of the real interauricular septum, known to embryologists as the septum primum (Fig. 20). In reptilian hearts the lower fornix of the venous valves ends on the auriculo-ventricular valves (see Fig. 2 in THE LANCET, Feb. 27th, 1904, p. 556).

Abnormal foramen in posterior part of the interauricular septum.—There is another defect of the auricular septum of which I have seen but two examples: one of them occurred

in the case of a woman, aged 45 years, under the care of Dr. F. J. Smith in the London Hospital. The defect, a large aperture, 15 by 20 millimetres, situated in the auricular septum above the fossa ovalis (Fig. 19), gave rise

FIG. 19.



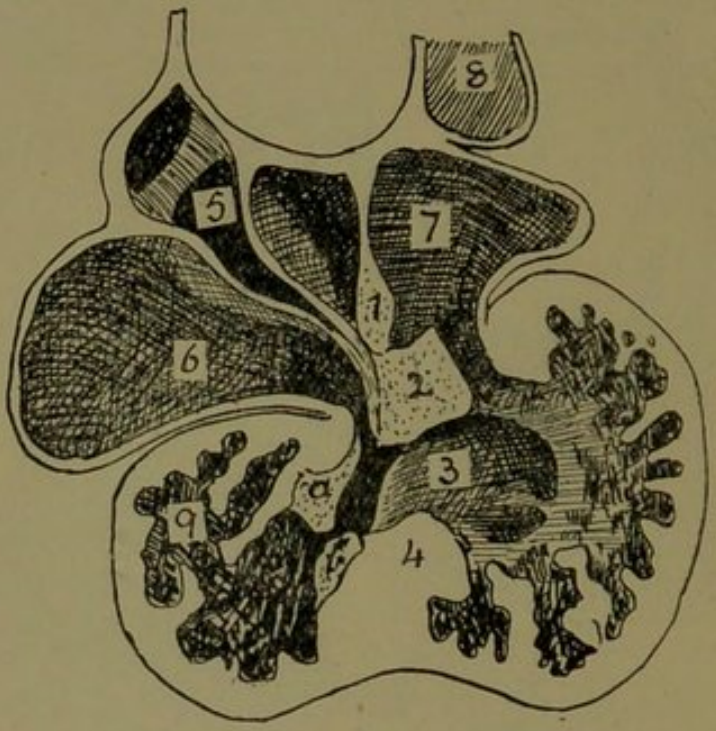
Right chambers of a human heart exposed to show the position at which various communications in the septum may occur. 1, Communication at posterior part of the interauricular septum (probably inter-sinus); 2, absence of the septum ovale; 3, the foramen primum; 4, the interventricular foramen; 5, infundibular or interbulbar foramen; 6, pulmonary artery laid open; 7, on septal cusp of tricuspid near the opening of coronary sinus; 8, unusual site of anomalous interventricular foramen; 9, moderator band; 10, pulmonary artery; 11, aorta; 12, superior vena cava; 13, taenia terminalis; 14, inferior vena cava; 15, Eustachian valve at opening of inferior vena cava.

to no symptoms during life. In this case the defect was correlated with an abnormal termination of the right pulmonary veins; they terminated in the right auricle between the openings of the upper and lower caval veins.

The mixed blood of the right auricle had by means of the interauricular defect free access to the left auricle. The communication in this case is probably one between the parts of the right and left auricle formed out of the sinus venosus.

Premature closure of foramen ovale.—How far imperfect formation and premature closure of the foramen ovale is a

FIG. 20.



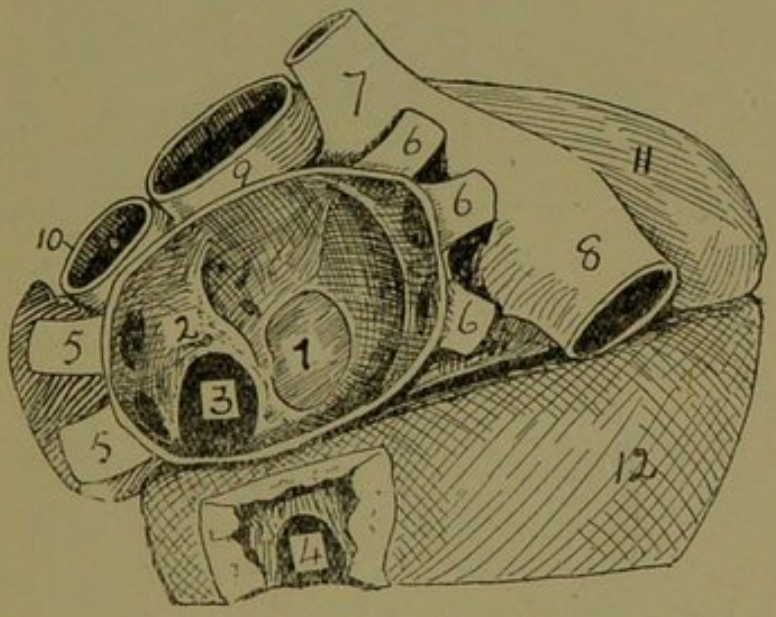
Coronal section of the heart of a human embryo about five weeks old. (After His). 1, Lower end of interauricular septum; 2, separation between auriculo-ventricular orifices formed out of the endocardial cushions—it is joined by the venous valves on the right side; 3, interventricular septum within ventricle; 4, the same on section; 5, opening of sinus venosus between right and left venous valves; 6, right auricle; 7, left auricle; 8, left superior vena cava; 9, infundibulum; *a*, *b*, lower ends of infundibular septum.

factor in the causation of other congenital malformations of the heart I am not yet in a position to say, but I have observed in several cases that the opening and fossa were abnormally small.

Abnormal septum in left auricle.—Time does not permit me to deal with the various abnormalities of the Thebesian, Eustachian, and other remnants of the two venous valves in the right auricle. In THE LANCET of Feb. 27th, 1904, p. 556,

I have figured a case where both valves persisted. I now pass on to another condition which is not yet represented in the College collection—viz., the presence of an abnormal septum in the left auricle. The three cases which I have seen are those already published by Professor T. Wardrop Griffith (two) and by Dr. H. D. Rolleston (one). The illustration is taken from a case well reported by Potter and Ranson in the *Journal of Anatomy and Physiology* (October, 1904, p. 69). The left auricle is imperfectly divided by the septum into two compartments, one of them receiving the pulmonary veins—the vestibular part, the other or body

FIG. 21.



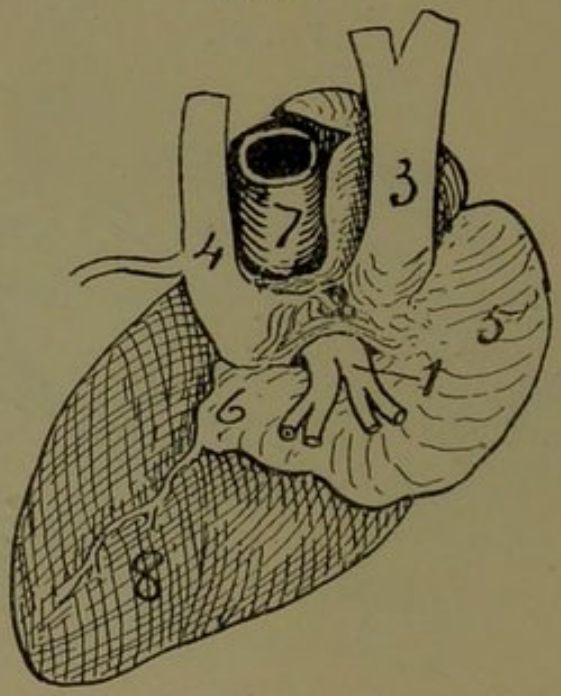
Left auricle opened to show subdivision by an abnormal septum. 1, Septum ovale in left auricle; 2, abnormal septum; 3, communication between vestibule and body of left auricle; 4, cavity of left ventricle opened; 5, left pulmonary veins; 6, right pulmonary veins; 7, superior vena cava; 8, inferior vena cava; 9, aorta; 10, pulmonary artery; 11, right auricle; 12, right ventricle.

of the auricle communicating with the appendix as well as the vestibule (Fig. 21). The condition is interesting because it shows the dual composition of the left auricle, one part, the vestibule, being derived from the opening up of the common termination of the pulmonary veins, the other from the primitive auricle. The abnormal septum occurs at the line of fusion of these two elements. In Fig. 22 is represented a heart in which there has been an arrest of the expansion of the pulmonary sinus to form the vestibule of the left auricle. I have seen three hearts showing this arrested condition of the vestibule

—all of them associated with other grave malformations. As in the case shown in Fig. 22, the left superior vena cava and left duct of Cuvier have persisted.

Interbulbar or infundibular foramina.—Before describing the defects of the interventricular septum it is convenient to return again to the aortic end of the heart and to describe certain defects in the septum of the heart which are at present very imperfectly understood. The position in which such a defect occurs is shown in Fig. 19. The opening is

FIG. 22.

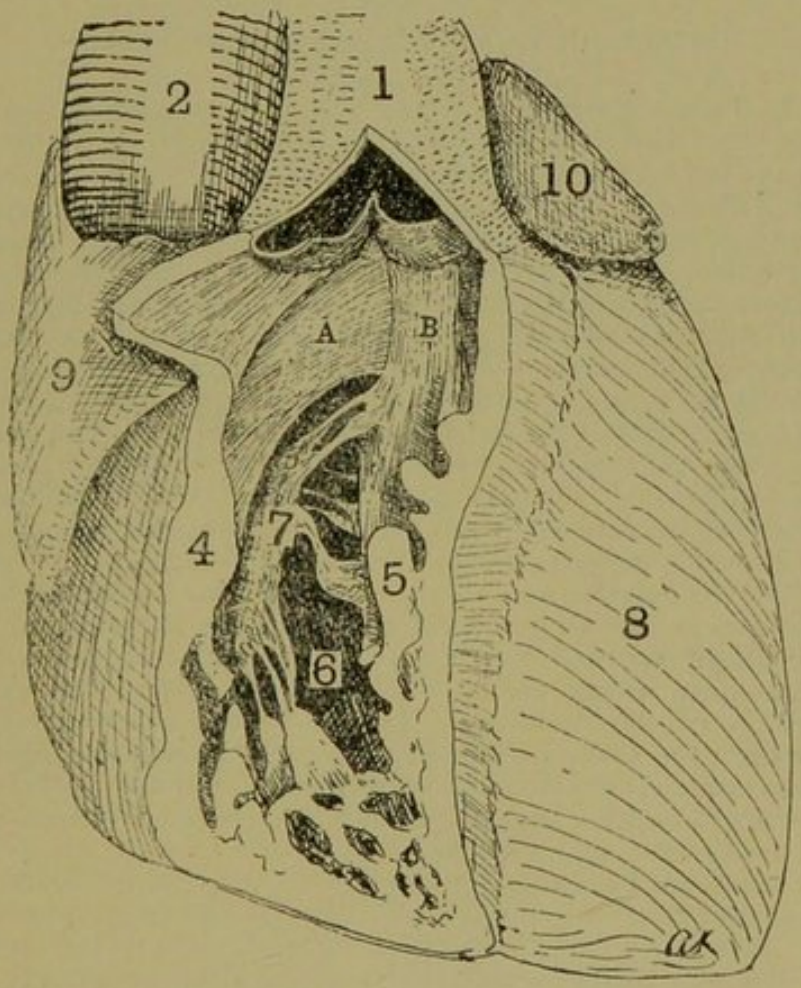


Arrest in the development of the vestibule of the left auricle. 1, Pulmonary veins; 2, unexpanded vestibule of left auricle; 3, right superior vena cava; 4, left superior vena cava; 5, right auricle; 6, coronary sinus; 7, aorta; 8, ventricles.

seen to be situated in the septal wall of the infundibulum of the right ventricle, and to communicate with the left ventricle under the right coronary cusp, which forms the upper boundary of the opening. It is situated in that part of the heart which we have seen is derived from the bulbus cordis, and we are dealing here with a defect, not really of the interventricular septum, but of the interbulbar. Rokitansky, in his classical monograph, represents exactly the condition seen in a specimen lately shown to me by Professor Wardrop Griffith, and in two others, one in the Museum of St. George's Hospital, and already described by Dr. Rolleston and another in the Museum of Guy's Hospital.

In the heart figured in my first lecture (Fig. 2, THE LANCET, August 7th, p. 360) a fibrous raphe (the interbulbar raphe) is seen to descend on the inner wall of the infundibulum, stretching from the semilunar valves above to the interventricular orifice below. It is in this raphe that interbulbar foramina occur.

FIG. 23.



The infundibulum of the normal heart. A, Right infundibular band; B, left infundibular band; between their origins is seen the infundibular raphe. 1, Pulmonary artery; 2, aorta; 3, pulmonary valves; 4, 5, constriction marking the site of the ostium infundibuli (5 is on section of moderator band); 6, body of right ventricle; 7, tricuspid valve; 8, left ventricle; 9, right auricle; 10, left auricle.

Even in the normal heart a trace of the interbulbar raphe may be seen (Fig. 23). On the septal wall of the infundibulum two great bands of musculature are seen to arise, and descend on the inner wall of the right ventricle—one towards the base of the ventricle, and therefore fitly termed the right or

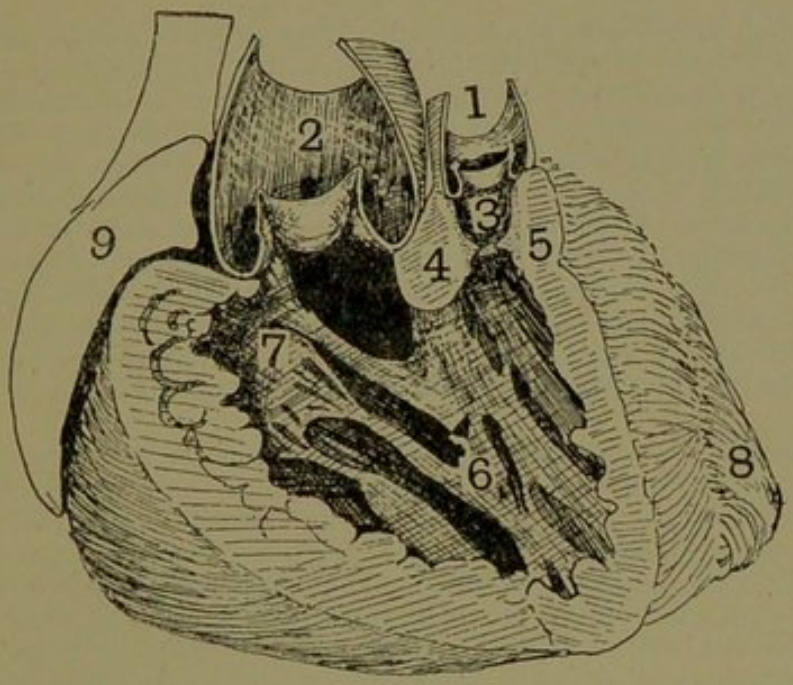
basal band, and another towards the apex of the right ventricle, and which may therefore be named the left or apical band. These bands take their fixed origin from the interbulbar raphe and orifices of the aorta and pulmonary artery and play an important part in emptying the ventricle. The interbulbar septum is laid down at first as an endocardial fold into which the two muscular bands just described push their origins.

Defects in the Interventricular Septum.

1. *Interventricular foramina.*—The only malformation of the heart discussed by Hunter in his writings is that found in the heart of a boy in his fourteenth year whom he saw with Dr. Poultney. The heart showed a typical interventricular foramen associated with that conditon of pulmonary stenosis already described in these lectures as due to an arrest in the development of the infundibulum of the right ventricle. In the majority of such cases (85–90 per cent.) an interventricular foramen is present. Hunter regarded the obstruction at the entrance to the pulmonary artery as the cause of the interventricular foramen ; Meckel, on the other hand, regarded the foramen as primary and the pulmonary stenosis as a sequence. There is not yet sufficient evidence to decide which of these two theories is true, but what there is points to Hunter's suggestion being the more correct one.

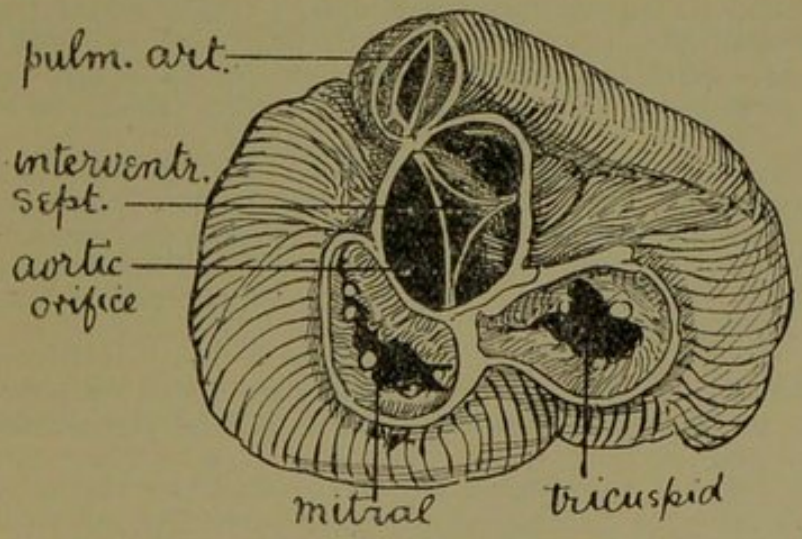
The boundaries and relationships of the interventricular foramen are shown in a heart represented in Figs. 24 and 25. The right ventricle and atrophied infundibulum below the pulmonary artery are laid open in Fig. 24. The lower margin of the foramen is formed by the interventricular septum ; the posterior boundary is formed by a band of fibrous tissue which is situated just below the non-coronary cusp of the aorta and derived from the mass of fibrous tissue formed out of the endocardial cushions. The upper boundary, cut across in the figure to display the interior of the aorta, is formed by the lower margin of the aortic orifice, while the anterior margin is formed by the interbulbar septum. Thus the interventricular foramen extends along the upper margin of the interventricular septum from the interbulbar septum in front to the intervalvular or auriculo-ventricular septum, formed out of the endocardial cushions, behind. Its size is extremely variable ; usually it is about 8 × 12 millimetres, but it may be little more than a pinhole or large enough to take three or four fingers at once. The exact sequence of events which lead to the normal closure of the interventricular foramen is not known, but from the fact that the foramen disappears as soon as the infundibulum has become opened up and incorporated with the right ventricle very early in the second month one may reasonably infer that its closure depends on the full development of the infundibulum.

FIG. 24.



Heart of a boy, aged six years, with right ventricle and aorta opened out to show relationships of the interventricular foramen. 1, Pulmonary artery; 2, aorta; 3, unexpanded infundibulum; 4, right infundibular band; 5, left infundibular band; 6, on moderator band which contains right septal division of the *a.v.* bundle; 7, tricuspid valve; 8, left ventricle; 9, right auricle.

FIG. 25.



Base of the heart of a girl, aged 13 years, in which there is arrest of the infundibulum with large interventricular foramen. The position of the aortic valves is shown and the subdivision of the aortic orifice by the upper margin of the interventricular septum.

Indeed, as one may see from an examination of the heart in Fig. 24, the expansion of the bulbus cavity to form the infundibulum must play a most important part in its closure. In that diagram the figure "4" marks the right infundibular band of muscle which springs from the interbulbar septum. It will be apparent that as the infundibulum expands this great band of musculature is thrown across and helps to occlude the interventricular foramen. The occlusion is completed by the fusion of the endocardial cushion which lines the margin of the foramen. The tissue of this endocardial cushion forms the substance of the pars membranacea septi —the curtain which closes the interventricular foramen.

The effect of an interventricular foramen on the action of the heart.—The functional significance of an interventricular foramen is best realised by such a view of the heart as is represented in Fig. 25. It is from the heart of a girl, aged 13 years, in which the infundibulum was only partially opened up. The upper margin of the interventricular septum is seen to divide the aortic orifice into two parts, one-third of it being in communication with the right ventricle, two-thirds with the left. The body of the right ventricle discharged its blood by two orifices: (1) a small part into the constricted infundibulum; and (2) a much larger part into the aorta. The walls of the body of the ventricle are hypertrophied so as to be able to compete on equal terms with the left ventricle in supplying the body with blood.

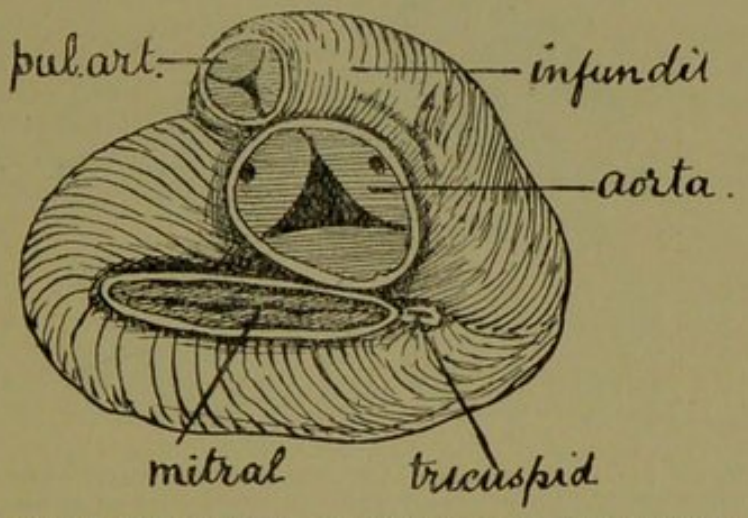
It is difficult to account for those comparatively rare cases where an interventricular foramen is present, yet the hearts seem otherwise perfectly formed. I have seen nine hearts of this kind. In all the foramen was of comparatively small size and had a membranous margin. The theory I have advanced, that the expansion of the infundibulum leads to the closure of the interventricular foramen, will not account for these cases. This class of hearts requires further study.

Irregular interventricular foramina.—Amongst the specimens examined for the purpose of these lectures there were four in which the interventricular septum was perforated by irregular foramina—apparently interstices in the original muscular sponge work of the septum which had remained unobliterated (Fig. 19).

Atrophy of left auricle.—The older writers on malformation in the heart endeavoured to utilise the classification employed in comparative anatomy and divide abnormal hearts into two-chambered and three-chambered hearts. A two-chambered human heart I have never seen; in three hearts of stillborn children, where there was complete stenosis of the mitral orifice, the left auricle was so small that it might easily have escaped detection; I have never seen the interauricular septum completely absent.

Hearts with one ventricle.—Amongst the malformed hearts which are said to have a common ventricle there are really three kinds : (1) those in which the interventricular septum is so little developed, or, to put it differently, the interventricular foramen is so large that the two ventricles are rightly said to form one chamber ; (2) those cases where the right ventricle is suppressed by the septum being applied to the right side of the heart ; and (3) those in which the left ventricle is suppressed by the septum being applied to the left wall of the ventricle. The first form is the commonest (nine cases) and is often associated with transposition of the arterial trunks ; of the second form I have examined seven cases and

FIG. 26.



Base of a heart in which there are (1) stenosis of the right auriculo-ventricular opening ; (2) suppression of the body of the right ventricle ; and (3) development of the infundibulum of the right ventricle. The common ventricle is really the left ; the auriculo-ventricular bundle in seen on the right lateral wall of the ventricle.

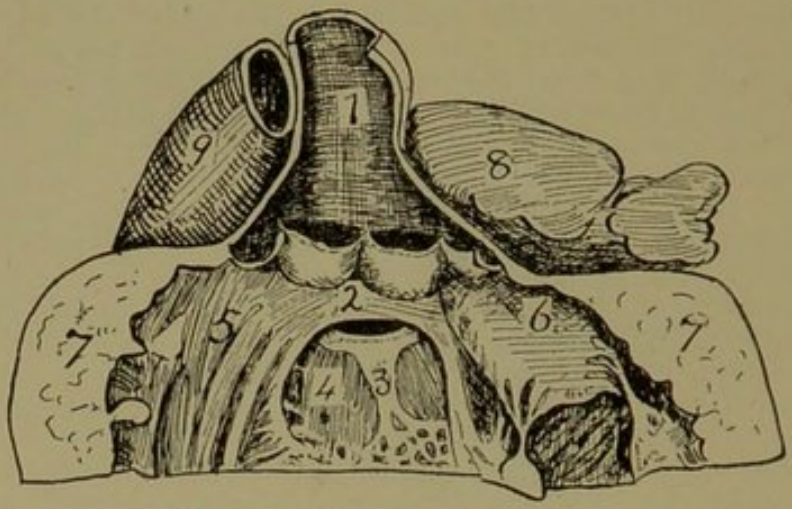
of the third five cases. In the second and third groups the absence of a ventricle is associated with a complete obliteration, or almost complete, of the corresponding auriculo-ventricular orifice (Fig. 26). The obliteration is probably due to the fusion of the endocardial tissue out of which the auriculo-ventricular valves and cushions are formed.

Development of the interventricular septum.—A bifid condition of the apex of the heart is not uncommon as an abnormality in man and is the normal form in some mammals such as the dugong. I refer to the condition here because it was difficult to give a satisfactory explanation when the interventricular septum was regarded as an upgrowth from

the floor of the primitive ventricle. We now know that the opposite is the case; the ventricular cavities are downgrowths or evaginations of the primitive cardiac tube, the septum being left between them during development. The upper margin of the septum thus represents a part of the lumen of the primitive heart; in the upper margin is developed the auriculo-ventricular bundle (Figs. 20 and 27).

The auriculo-ventricular junctional system in abnormal hearts.—I have investigated the condition of the auriculo-ventricular bundle, or, to use a term more expressive of its

FIG. 27.



The aorta and left ventricle of an abnormal heart opened to show the interventricular foramen and auriculo-ventricular bundle. 1. Aorta. 2. Musculature rising from the aorta above the interventricular foramen; this musculature frequently persists as the subaortic musculature and covers the auriculo-ventricular bundle. 3. Left septal division of the auriculo-ventricular bundle. 4. Interventricular septum, with main bundle on its upper margin. 5. Subaortic bundle. 6. Mitral. 7. Wall of left ventricle. 8. Left auricle. 9. Pulmonary artery.

real nature, the auriculo-ventricular junctional system, in malformed hearts. The main bundle of this system has a definite relationship to the interventricular foramen which is well shown in the heart drawn in Fig. 27. The heart is that of a boy in which the development of the infundibulum was arrested and there was an interventricular foramen. The bundle is seen passing along the upper border of the interventricular septum, just below the foramen; the left septal division is seen streaming down the septum and ending in branches on the musculi papillares. This specimen also shows another abnormal condition which has a special interest because of its relationship to the bundle. It will be

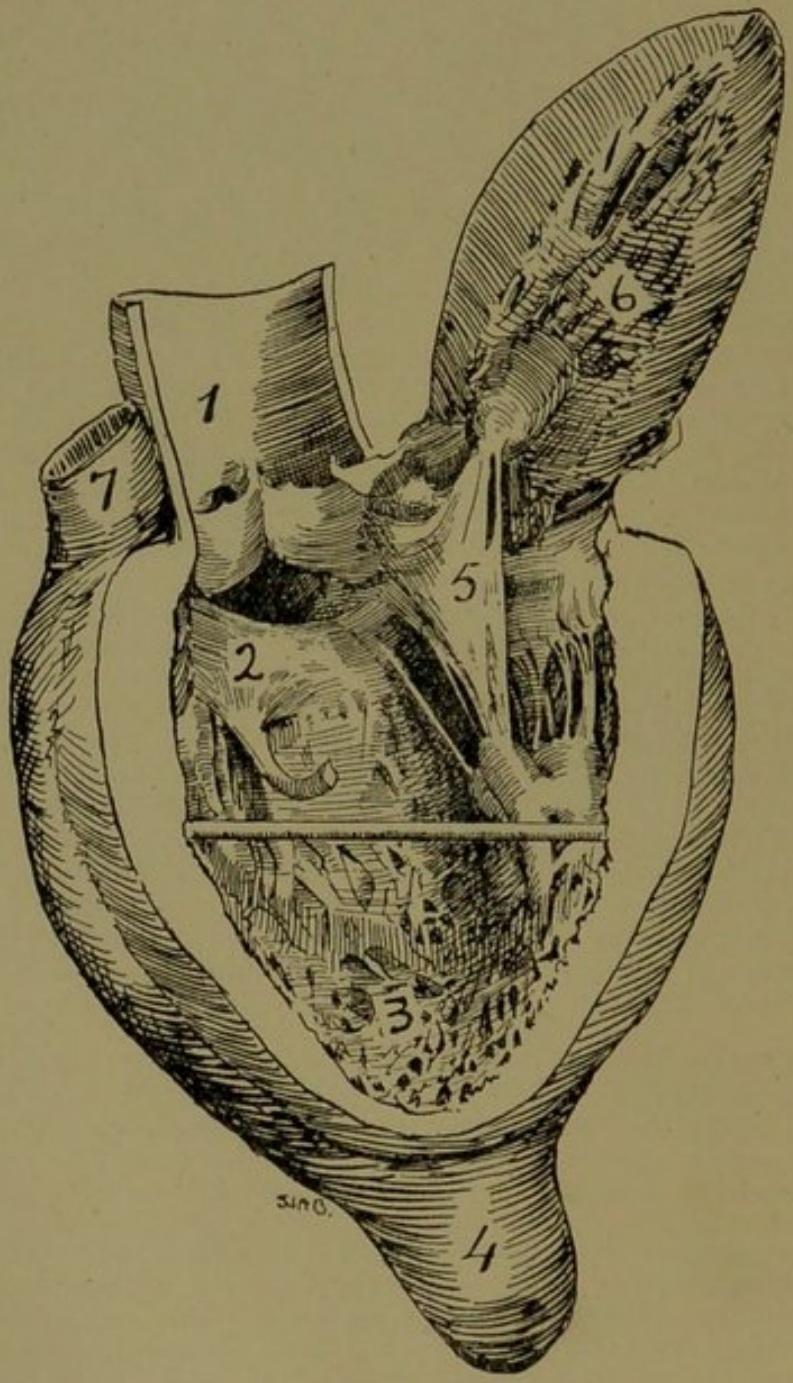
observed that there is a muscular curtain spanning over the interventricular foramen ; the part of the curtain above the foramen would, if that opening had become closed, have formed the pars membranacea septi. Now the pars membranacea septi does sometimes contain muscle in quite normal hearts ; there is a good specimen now in the Museum collection. Such an abnormal musculature—the subaortic musculature—overlies and hides the main bundle. This musculature is always present in the hearts of ungulates.

In Fig. 24 the position of the bundle on the septal wall of the right ventricle can be realised. It commences in the node situated behind the tendon of the septum (above point marked "7") and passing forwards on the lower margin of the foramen sends its right division into the moderator band of the right ventricle. In the heart shown in Fig. 24 the infundibulum is arrested, but had it gone on to normal development its lower margin would have extended as far as the moderator band, which thus marks the junction of the body of the right ventricle with the infundibulum—the part derived from the bulbus cordis.

The last abnormality I propose to deal with is that shown in a specimen just added to the Museum—the moderator band of the left auricle. The doyen of British anatomists, Sir William Turner, was the first to call attention to it, and Professor Wardrop Griffith has described several specimens. It is especially frequent in abnormal hearts and has a definite relation to the left septal division of the bundle (Fig. 28). The fibres of the auriculo-ventricular bundle pass by it across the left ventricle to the anterior musculi papillares in the same manner as the right septal division is conducted by the moderator band to the anterior musculus in the right ventricler. The left bundle is not a constant structure in any mammalian heart.

I have devoted these three lectures to bringing before you the results of an intermittent study of malformed hearts extending over some seven years. My reason for applying myself to this particular subject was not merely to gather cases as a collector may curios but with a definite, or rather a twofold, object—(1) I hoped to obtain some light on the development and morphology of the heart; and (2) the subject required systematisation, for the numerous cases annually recorded in the medical press, the mere references to which take up over 12 columns in the invaluable index catalogue issued from the office of the Surgeon-General of the United States army, were often rendered of little value because the recorder was unaware of the points which were most worth investigating and recording. These were my two objects. At least to some extent my study has been successful. It brought to light the fact that there is buried in the heart an unrecognised element which is more frequently malformed than any other part of the heart ; it

FIG. 28.



Left ventricle of an abnormal heart opened to show (1) the interventricular foramen; and (2) the moderator band (drawn by Dr. Stanley Beale). 1, Aorta, opened. 2, Upper margin of interventricular septum, at origin of left moderator band. 3, Apex of left ventricle. 4, Apex of right ventricle. 5, Anterior cusp of mitral. 6, Wall of left ventricle thrown up. 7, Pulmonary artery.

has helped to demonstrate that the left auricle is dual in its composition.

I am also convinced that these lesions are not the result of fœtal endocarditis for the following reasons. 1. It is impossible to conceive endocarditis giving rise to the great majority of the malformations which are here described. 2. The tissue in the neighbourhood of the lesions is not cicatricial, except in those cases which are at the time of death the seat of vegetations, the result of an infection after birth. 3. In 23 children, all of which showed some malformation of the body—hare lips, cleft palate, atresia ani, anencephaly, &c.—there were 14 with an associated malformation of the heart, especially pulmonary and aortic stenosis. Malformations of the heart evidently result from those unknown causes which give rise to malformations of the rest of the body. We are not dealing here with the results of an infection but with a disturbance of the conditions in which the embryo develops. The exact nature of that disturbance is being elucidated at present by the investigations of experimental pathologists.