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CONGENITAL ATRESIA OF CONUS ARTERIOSUS—INCOMPLETE SEPTUM VENTRICULORUM—AORTA, MAINLY ARISING FROM THE RIGHT VENTRICLE.

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NOTE ON A CASE OF CONGENITAL MALFORMATION OF THE HEART.

CONGENITAL ATRESIA OF CONUS ARTERIOSUS—INCOMPLETE SEPTUM VENTRICULORUM—AORTA, MAINLY ARISING FROM THE RIGHT VENTRICLE.

A STRIKING contrast exists between the right and left sides of the heart before and after birth in regard to structure, functions, and proneness to disease. In the adult heart the left ventricle is all powerful and dominant, and vigorously resists the effects of disease, which so often attacks its orifices and valves; but on the right side the valves are but rarely primarily diseased, it is the thin-walled cavities that first suffer, then the orifices are widened, and finally the valves become incompetent. In the fœtus, on the contrary, the wall of the right ventricle is as thick as that of the left, and it has the larger share of work; its valves are submitted to greater pressure, and it is on the right side that the results of fœtal endocarditis are most commonly seen, and in more than half the cases, at the origin of the pulmonary artery, which has to transmit blood through the ductus arteriosus into the descending aorta, and thence to the umbilical arteries and placenta, where transitory interruptions in the circulation will readily occur. The frequency of lesions of the pulmonary valves at this period may be contrasted with the frequency of mitral disease in childhood, and of both mitral and aortic disease in adult life. Disease of the tricuspid valve before birth is rare, but yet may be said to be commoner than after birth, even though bearing in mind Byrom Bramwell's recent researches which indicate that post fœtal right-sided endocarditis is oftener to be found, if carefully looked for, than is generally supposed. Fœtal endocarditis affecting the aortic valve is usually stated to be very rare, but Rauchfuss says that fewer cases are seen owing to the very short duration of life, and that, leaving out the influence of congenital defects (oftener met with on the right side), it is really not very uncommon. Haranger, too, collected 49 cases for his thesis on congenital endocarditis of the left heart.

The etiology of fœtal endocarditis is still obscure. We cannot, as in the child or adult, point to rheumatism as the most likely cause, for there is no evidence to show that the parents of cyanotic children are more rheumatic than those of children who have healthy hearts; or that rheumatic fever is common during the pregnancies of women who bear

babies with diseased hearts; nor is there proof that the fœtus is subject to rheumatism. Von Hoffman has suggested that from hæmorrhagic foci in the placenta, pathological products may be introduced through villous absorption into the fœtal circulation, and so occasionally induce inflammatory lesions. Whatever the explanation, it is curious and suggestive that in a fœtal heart one should see all the lesions—vegetations, thickenings, shrinkage, calcareous degeneration, &c., of the valves—encountered in adult hearts, and known there to be the result of rheumatic endocarditis. In the fœtal heart we invoke some arrest of development on which an inflammatory lesion has been grafted, but do not think it necessary to find a weak spot as a predisposing cause for the point of selection of endocarditis occurring after birth.

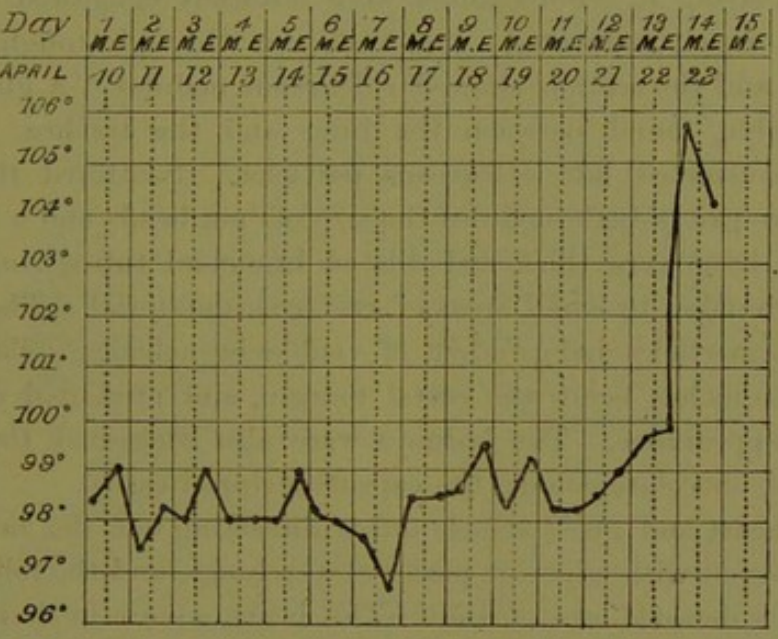
In the following case, the heart, belonging in its broad features to one of the commonest types of congenital malformation, presents certain peculiarities, of which one of the most interesting is the complete absence of any trace of recent or of old inflammation.

William M., aged 10, was brought to the Clinical Hospital at the beginning of April, 1887. He was fairly grown and of average stature for his age. His nose and cheeks were purplish in colour; his lips, tongue, and the inside of his mouth presented a dark purple congested appearance. The ends of the fingers and toes were livid and much clubbed. There was no œdema, and the urine was free from albumen. On examining the chest there was slight bulging of the præcordial region. The heart's impulse was normally placed; it was a little stronger than natural, but not distinctly heaving; no pulsation was detected at the lower end of the sternum or in the epigastrium; there was no thrill; the cardiac dulness was increased laterally, extending from a finger's breadth to the right of the sternum to the left nipple line. On auscultation, a harsh systolic murmur was heard at the base, of maximum intensity at the second left costal space near the sternum; it was conducted a little better towards the right than towards the left clavicle; was heard on moving the stethoscope down the sternum, or towards the left apex, but was not heard in the axilla nor behind—the heart sounds being quite clear and pure at the angle of the left scapula and over other parts of the back. The pulse was quite regular and of normal quality. As regards the lungs, the resonance was impaired at the right apex, and the breath sounds there were harsh and prolonged, but not distinctly bronchial. Occasionally some fine moist sounds were audible at the end of a deep inspiration; elsewhere the lungs seemed to be healthy.

On inquiring from the boy's mother, she stated that he was "blue when born," and that the midwife who attended her remarked that "his heart was wrong." He was born at the full time—the mother had a good confinement, and was quite well during her pregnancy; did not

fall, had no fright. The boy has always had fair health; was never laid up with severe illness—never had inflammation of the lungs, nor any chest complaint, though at the beginning of last winter he had pains over the right side of the chest in front and behind. Lately he has been spitting blood. She has never noticed any swelling of his feet, but says that he has complained much of pains in the legs and feet. He often has diarrhœa. His father is quite healthy; his mother's father died of consumption, and her brother died of some lung affection. She lost two babies from bronchitis, has five other children living, younger than William—they are all quite well.

The day after admission to hospital, patient vomited about three oz. of fluid blood, and became intensely cyanosed, but seemed quite well an hour later.



On April 22nd his chest was again carefully examined, but nothing fresh was discovered; the heart sounds were distinctly heard over the back, and were quite pure; in front the systolic murmur had the same character as before. His temperature, (as shown in the accompanying chart) which previously had been more or less normal, was 99·8° in the evening of the 22nd, and the next morning, at eight o'clock, was 105·8°; at 10 a.m. the house surgeon, Dr. Scott, noted that the heart's action was very rapid, the pulse 186, and respirations 48 per minute. The murmur was not distinctly audible. The breath sounds, except at the right apex, were normal, and no crepitation was heard. There was some general tenderness of the abdomen, and a little diarrhœa. At 7 p.m. his condition was much the same. The bowels had been moved three times since 4 p.m. The pulse was very rapid and small, and difficult to count. He was drowsy, but said he had no headache. He died somewhat suddenly a few hours later.

At the autopsy made about 34 hours after death rigor mortis was well marked. On opening the thorax, the thymus was seen to be much enlarged, its tip reached the sternum opposite the third cartilage, and overlaid the upper part of the right auricle. The pericardium was unduly exposed, measuring 3½ inches transversely, and 3 inches vertically in the mid-sternal line. On opening the pericardium there was no excess of fluid. The heart was placed more horizontally than usual, its apex being tilted up; its anterior surface was made up of a much-distended right auricle and of the right ventricle, from the base of which sprang an unusually large artery, subsequently seen to be the aorta.

The lungs were bound down by extensive adhesions, which, for the most part fine and easily broken through, were exceedingly firm about the right apex, and it was impossible to detach the right upper lobe without tearing it. Both lungs were intensely congested. The right apex was much indurated; the knife cut through tough and gritty material, and the section showed calcareous nodules. A gland, the size of a marble, found between the lung and the trachea, was almost entirely converted into calcareous particles. Scattered through both lungs were gritty little nodules the size of pin heads and a little larger. Many of these were thickened dilated bronchial tubes, some of which were surrounded by small areas of cretaceous material. There were also calcareous particles independent of visible bronchioles. The liver projected two inches below the costal margin, and presented on section a nutmeg appearance. The kidneys were also congested, the pyramidal portion of chocolate colour, the capsule stripped easily.

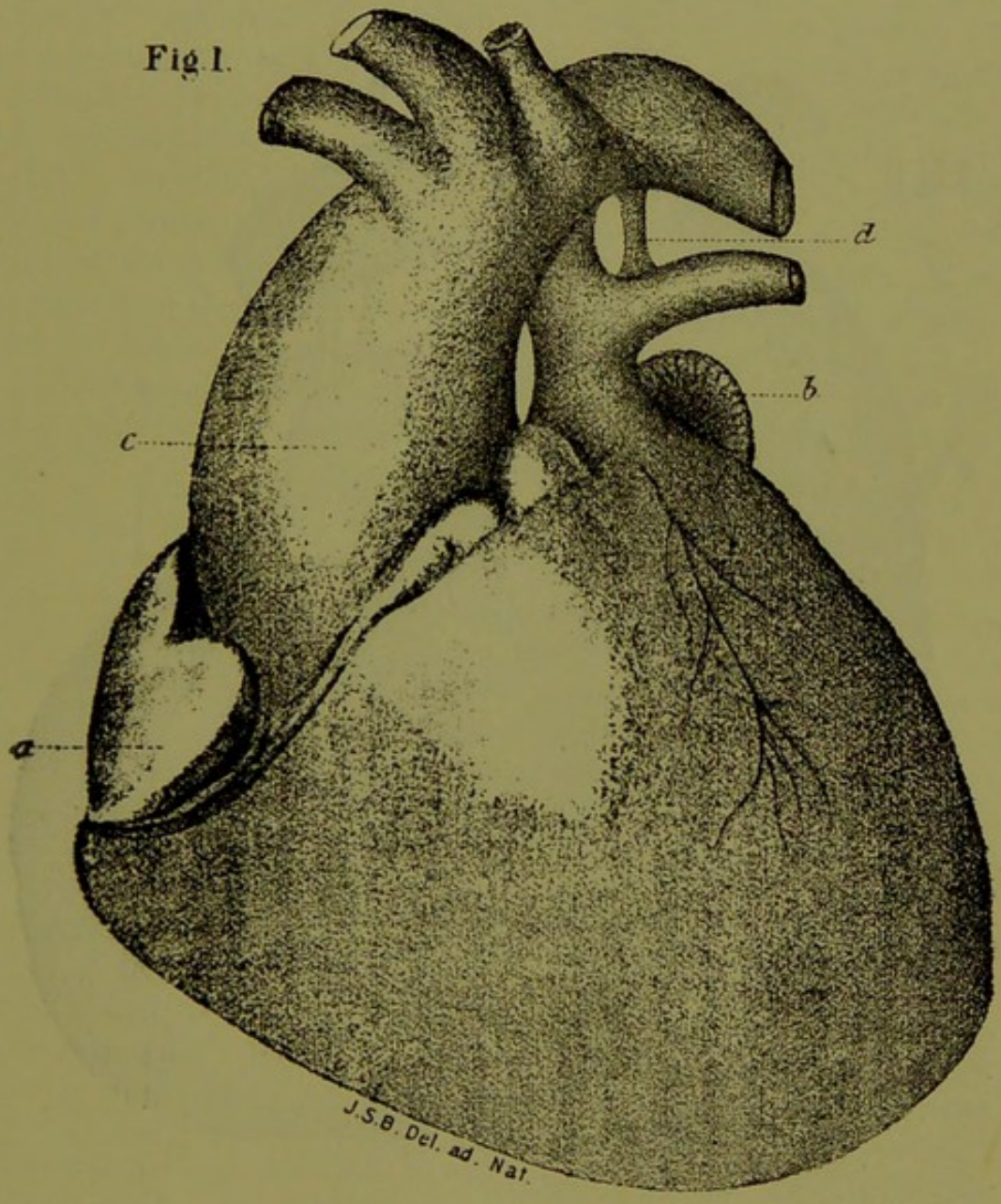
The heart and great vessels (after dissecting out the latter *in situ*) were removed with adjacent pieces of the lungs, and the following is the result of a detailed examination: The heart, square in shape, measured 3 by 3 inches, and weighed when empty 7oz.

The large size of the aorta, and its position more to the right than usual were very noticeable. It occupied 1½ inches of—that is, half—the width of the base. To its left was the small pulmonary artery The right cavities, previously found to be distended with dark blood and black clots, were first examined. The right auricle and its appendix were of large size; the two cavæ opened into it normally. The tricuspid orifice just admitted the tips of two fingers; the opening of the coronary sinus was natural. The foramen ovale appeared to be closed, but a small slit-like aperture measuring one-third inch in its longest diameter was found between the valve on the floor of the fossa ovalis and the annulus above. On opening the right ventricle the tricuspid valve made up of three segments appeared quite normal. The anterior part of the ventricle presented a remarkable abnormality. Instead of an infundibulum leading to a pulmonary artery, there was a

large sinus with the origin of the aorta at the top of it, and no trace of an infundibulum was present (see Fig. 2).

The aortic orifice was guarded by three semilunar valves; above the two posterior cusps were the openings into the coronary arteries, the right artery starting behind the right posterior cusp instead of behind the anterior cusp as normally, the left artery behind the left posterior

Fig. 1.

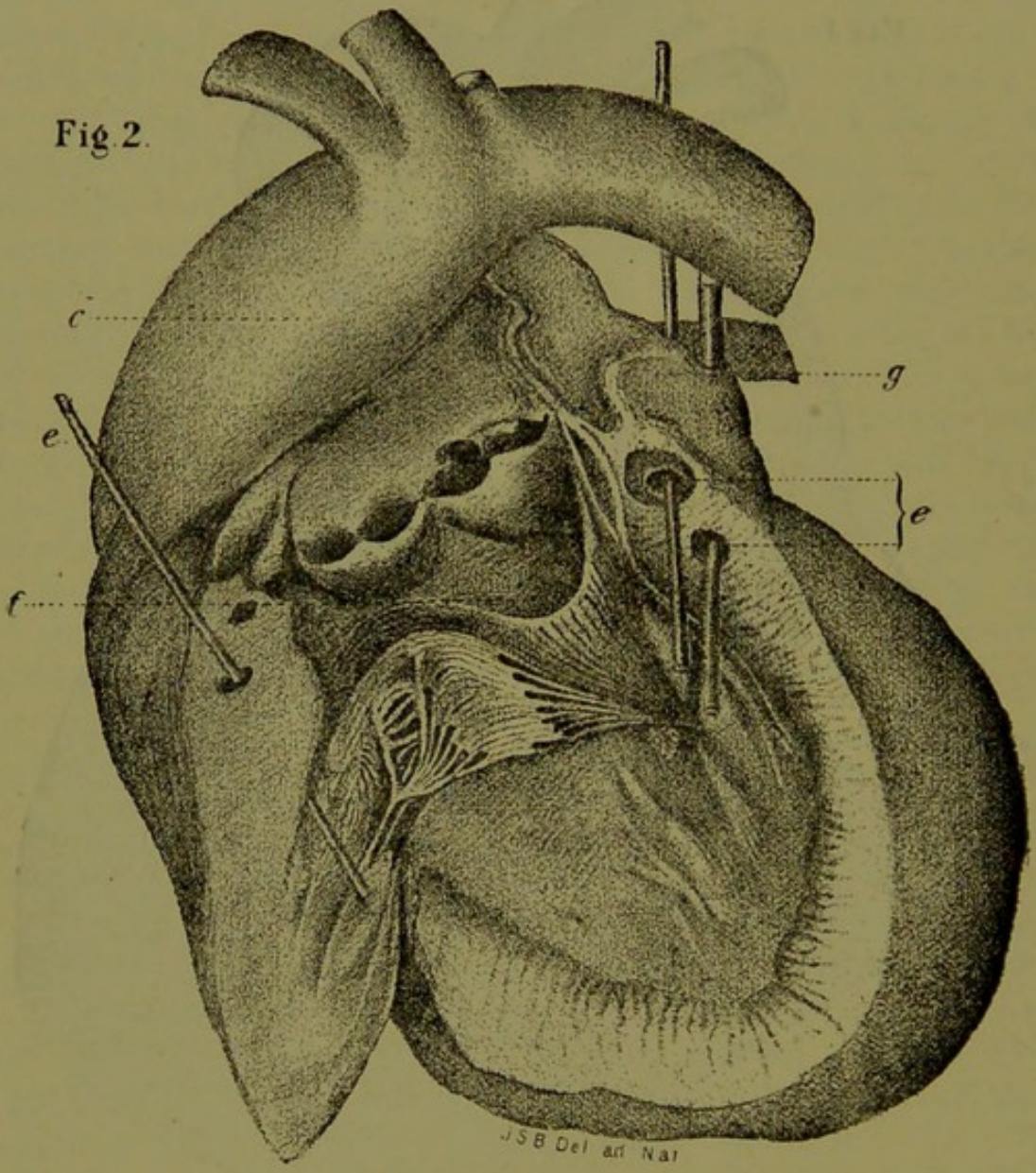


a—Right auricular appendix.
b—Left " "
c—Aorta
d—Impervious ductus arteriosus.

cusp, but higher up than usual, and this cusp had a remarkable downward oblique direction. Immediately below the right posterior cusp, and between it and the left posterior cusp, was a round hole with smooth

edges, leading into the left ventricle. It was bounded below by a prolongation of one of the columnæ carneæ, which spread up between the two posterior aortic cusps, and was joined by a slender bundle from below the anterior cusp; afterwards it was found that immediately behind the hole, and forming its posterior wall, was the anterior flap of the mitral valve. The muscular wall of the ventricle was very thick and its cavity large.

Fig. 2.



e—Probes passed through narrow channels of communication between pulmonary artery and ventricle.

f—Hole leading into left ventricle, the dark shaded part being the anterior cusp of the mitral valve.

g—Left branch of pulmonary artery.

No opening leading into the pulmonary artery could at first be discovered; on cutting into the artery above, it was found to be fairly capacious, though less than the normal size. There were two thin

valvular curtains, one to the right with a pocket behind it; the other, a narrow thin fold, occupied the rest of the lumen and had a very shallow pouch behind it. On passing a probe downwards between the valves it came out through a small hole in the wall of the right ventricle; then another hole was found in the ventricular wall, leading by a narrow passage also into the pulmonary artery. On looking at the opposing cut surface of the wall corresponding holes were discovered, and a probe passed into them was pushed behind muscular bundles, and came out near the apex of the ventricle. These two narrow passages leading from the cavity of the right ventricle through its muscular wall into the pulmonary artery I have endeavoured to represent in the sketch. Each was lined by a thin membrane, and soft brownish fibrinous material was pulled out of the passages. The higher and larger of the two measured nearly one quarter inch in diameter. Their appearance on looking down at them from the pulmonary artery is shown in Fig. 3.

The pulmonary artery had thin walls; its widest part was near the valves, where it was dilated into a little pouch about $1\frac{1}{4}$ inches in cir-

Fig. 3.

cumference; it divided into two branches, the right, a little larger than the left, admitted an ordinary cedar pencil with difficulty; each divided into two branches, the upper breaking up at once into small vessels for the upper lobes of the lungs, the lower branches of larger size than, and not breaking up so soon as, the upper ones, supplied the lower lobes. The ductus arteriosus, quite impervious, was traced from the left branch of the pulmonary artery to the aorta. The four pulmonary veins were traced to the left auricle, which was of fair size. The left auricular ventricular opening admitted the end of the thumb. The left ventricle was very small, and the septum bulged into its cavity instead of being crescentic towards the right ventricle. The wall of the left ventricle was very thick, and about the same thickness as the wall of the right ventricle. The mitral valve appeared healthy; it consisted of a well-marked anterior flap, $\frac{1}{2}$-inch deep and $\frac{3}{4}$-inch wide, and a posterior one divided into two parts, each $\frac{1}{4}$-inch deep, the anterior part $\frac{1}{4}$-inch wide, and the posterior part $\frac{1}{2}$-inch wide. On passing the finger upwards

through the gap (which it just filled) above the septum it entered the aorta; but if the finger, instead of upwards, was directed forwards, it entered the right ventricle. The aorta, very wide at its first part, was much reduced in size after giving off the great vessels to the head and neck, the descending portion of the arch only just admitting the tip of the little finger. An irregularity in the arrangement of the great vessels for the head and neck was also noticeable.

DIMENSIONS OF THE HEART IN INCHES.

Circumference of heart .....7¼.
Girth of right ventricle .....4¼.
Girth of left ventricle.....3.
Thickness of walls of right ventricle .....¼—½, three times greater than normal.
Thickness of walls of left ventricle .....¼—½.
Circumference of right auriculo-ventricular aperture .....3½, half inch greater than normal.
Circumference of left auriculo-ventricular aperture .....2½.
Circumference of pulmonary artery .....1¼, half inch less than normal.
Circumference of aortic aperture.....2, nearly half inch greater than normal.
Internal circumference of aorta, higher up.....2⅜.
Internal circumference of descending portion of arch.....1½.
Width of cavity of left ventricle .....1—1½.
Length from top of septum to apex of left ventricle .....1½.
Circumference of orifice of superior vena cava...1⅜.
Circumference of orifice of inferior vena cava...1¼.

WEIGHTS.

Trachea, lungs, and heart.....28 oz.
Heart (empty).....7 oz.
Liver .....30½ oz.
Kidneys .....6 oz.
Spleen.....2 oz.

Remarks.—The sequence of pathological events in such a case is best understood by a reference to the normal development of the heart. Before the seventh week of fœtal life the heart consists of three cavities—ventricle, auricle, and arterial bulb. Of the partitions dividing each into two cavities, the ventricular septum is the first to appear, and at about the sixth or seventh week, starting from the lower border of the cavity, it rises as a crescentic partition till it reaches the base, where it is in relation with both the root of the arterial bulb and the auriculo-ventricular orifice. It is complete, and the ventricles entirely separated, before the end of the second month. About the same time the arterial bulb is divided into two channels, one anterior, the other posterior; the partition begins at the distal end and advances towards the ventricle. "There is a time, therefore, during which the ventricular septum and the

septum of the bulb, advancing towards each other, are incomplete and disunited, and from the difference in their general direction it is obvious that the septum of the bulb must be twisted upon itself in order that it may finally unite and become continuous with that of the ventricles."* The inter-auricular septum only begins to form after the separation into two ventricles, and its completion is much slower.

In the case described the presumption is that before the seventh week some obstruction existed at the origin of the pulmonary artery; then the resulting increased pressure would set up hypertrophy of the right ventricle, maintain the communication between the ventricles, deflect the septum towards the left, and so allow the aorta to grow into communication with the right ventricle, thus further increasing the work and, therefore, the hypertrophy of the latter. At this time the pulmonary obstruction was probably slight, certainly not excessive, otherwise a permanent inter-auricular communication would have been established in addition to the inter-ventricular, to relieve the tension in the right ventricle. What was the nature of the primary obstruction? Now, there is not the slightest trace of the infundibulum, unless the two narrow passages through the wall of the right ventricle leading into the pulmonary artery may be regarded as such. We may suppose that in early fœtal life the part which afterwards becomes the infundibulum was defective, perhaps smaller than natural near the origin of the artery; that this constituted the original obstruction, which was subsequently increased by muscular hypertrophy; the contraction resulting from this muscular stenosis, perhaps further increased by an acute or chronic myocarditis, or possibly there was no original malformation, the almost complete atresia being the consequence of gradual shrinkage from old myocarditis. However brought about, the communication between the right ventricle and the pulmonary artery during the latter part of the boy's life must have been extremely small, for along the two narrow channels but little blood could have been driven into the artery. How remarkable then is the relatively large size of the artery and its branches. How were they kept filled with blood and so prevented from dwindling into cords? The ductus arteriosus is impervious, and no other vessel supplies them with blood. In another case, where I found the pulmonary valves blended into a dome-shaped projection with a slit-like aperture at the top measuring only one-eighth of an inch, and the ductus arteriosus closed, the pulmonary artery was $2\frac{1}{2}$ inches in circumference. In this case I thought the size might be attributed to late closure of the ductus arteriosus, and that the blending of the semilunar cusps "attained its maximum sometime after this closure; and even with a small communication between the ventricle and the artery the latter would be kept full

* Quain's "Anatomy," 9th edition, p. 866.

of blood, and so maintain a fair size or dwindle very slowly."* Of this dwindling process we have evidence in the two very thin pulmonary valves, and in the thin arterial coat, and if the boy could have lived longer, no doubt the pulmonary artery would have been found still smaller. The aorta derived its blood from both ventricles, and the meeting of the two columns of blood in the ascending portion of the arch, together with the dilatation of the latter, sufficiently explain the presence of a systolic basic murmur and its better conduction towards the right than the left clavicle.

* "Congenital contraction of orifice of pulmonary artery, foramen ovale open."—*Lancet*, August 2nd, 1884.

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