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CASE OF

MALFORMATION OF THE HEART,

IN WHICH

DEATH RESULTED FROM OBSTRUCTION IN THE TRUNK OF THE PULMONARY ARTERY.

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The following case is conceived to be sufficiently interesting, from the unusual form of the malformation of the heart and the infrequency of the cause which occasioned death, to render it worthy the notice of the Society.

William Holland, a milk boy, aged 15, was admitted into the Royal Free Hospital, on the 20th of February 1847. He stated that on the 12th he had bruised the left knee by a fall, and had since suffered from constant pain in that joint, and also, for two or three days before admission, in the right knee. The day after the accident he began to experience pain in the left side of the chest and difficulty of breathing.

When admitted he was much collapsed and the extremities were cold and livid. The cheeks were of a deep purple colour and the lips blue. The fingers and toes were club-shaped, and the nails incurvated and very dark-coloured. The pulse was 124 in the minute and extremely feeble. The tongue dry and covered with a whitish fur. The respiration

was peculiarly rapid and panting, and he was compelled to lie on the back, partly inclined towards the right side, and with his head low. He complained of pain in the region of the heart, palpitation, and difficulty of breathing. Both knee-joints were swollen and tender, and there existed a red and swollen patch over the left trochanter.

The chest yielded a clear sound on percussion, except in the præcordial region, where the resonance was impaired over a larger space than natural. The liver could be felt extending a little below the edges of the ribs. The sternum was arched and prominent, more especially towards its base. The respiration was puerile in its character, and was attended with occasional mucous rales. A loud murmur was heard in the præcordia, accompanying the impulse of the heart. It was of a soft or blowing character, and was most intense at the cartilage of the third left rib near the sternum, or at a point half an inch above the nipple and between that body and the sternum; it was here prolonged throughout the whole period, so as to render the second sound inaudible. From this point it continued to be heard very distinctly, though decreasing in intensity, along the upper part of the sternum, in the subclavian and carotid arteries, and on the left of the spine in the interscapular and dorsal regions. It was also heard less distinctly in a line from between the nipple and sternum, towards the middle of the left clavicle. Below the level of the nipple the murmur became shorter and more feeble; and, at the point of pulsation of the apex, towards the epigastrium and on the right side of the lower half of the sternum, it was followed by a very clear second sound.

The boy was much exhausted, and his intelligence so impaired that it was found impossible to collect any satisfactory information of his state of health previous to the present attack. He stated, however, that he had lost flesh and strength; and had been very subject to affections of the chest, and of a livid complexion, since he was thrown from a cart twelve months before.

These notes were taken about 4 P.M. on the day of ad-

mission; and, notwithstanding the free use of stimulants externally and internally, he gradually sank, and died at eight o'clock on the following morning. On inquiry I ascertained that he had always been of a somewhat livid complexion; but was stout, healthy, and capable of a full amount of exertion, till the occurrence of the accident he referred to. He was then riding at the back of a cart, when it toppled up, and he was thrown out and fell upon his head. He was admitted into the hospital on the 6th of February 1846, under the care of my colleague, Mr. Gay, with symptoms of concussion of the brain. He continued there six days, and the only peculiarity then observed in his appearance, was some slight lividity of the lips. Since that time he had been gradually getting thinner and weaker; he was constantly chilly, and very subject to take cold. He complained occasionally of palpitation, difficulty of breathing, and pain in the region of the heart; and his hands and face were always very blue, but especially so in cold weather, or when he was suffering from affection of the chest. His appetite was generally defective, and he occasionally vomited his food. His father is of a livid complexion and has a "pigeon breast."

The *post-mortem* examination took place at 4 P.M., on the 22nd, thirty-two hours after death.

The brain was healthy, though much congested. It weighed 49 oz. 4 dr. avoirdupois.

The surfaces of the pleura on the right side were adherent by a small cellular band. The left lung was entirely free. Both lungs were engorged with blood, sparingly crepitant, and contained several masses in the state of pulmonary apoplexy. The bronchial mucous membrane was somewhat reddened. The pericardium was free from adhesions, and did not contain fluid. The heart was about 10 oz. in weight. It was broader from side to side than from above downwards. Its total circumference was $8\frac{1}{2}$ inches, of which the right ventricle constituted $4\frac{1}{2}$.

The systemic veins were free from disease. The right

auricle was large and its walls thick. The foramen ovale was closed, with the exception of a valvular aperture capable of admitting a goose-quill. The Eustachian valve was of moderate size. The right auriculo-ventricular aperture measured 39 lines in circumference. The valves were free from thickening and competent. The muscular column to which the cords of the anterior fold were attached was very large and firm. The aperture opened as usual into the sinus of the right ventricle; but this portion of the cavity was separated from the infundibular part by a thick muscular septum, defective only at its centre, over a space of sufficient size to admit the fore-finger, and perforated by one or two very small pores nearer the apex. The cavity thus formed communicated with the aorta by an orifice 30 lines in circumference, situated at its posterior extremity. Its walls averaged $5\frac{1}{2}$ lines in thickness, and were unusually firm and solid. In places they had undergone the fibro-cartilaginous degeneration throughout their whole extent, and the corresponding external serous envelope was opaque. The second or infundibular cavity of the ventricle was of smaller capacity than the former, and gave origin, as usual, to the pulmonary artery. Its walls averaged only 2 to 3 lines in thickness. The orifice of the pulmonary artery was very small, and was provided with only two valves which were extremely thick and opaque. The valves projected into the cavity of the vessel, so as to leave a deep sac behind each, and by their free borders occasioned a further contraction of the arterial orifice; which, while on the ventricular side it admitted a ball measuring 15 lines in circumference, could only give passage to one of 13 lines. The coats of the pulmonary artery were much indurated and thickened, and its canal was entirely obstructed by fibrinous coagula. At the sides of the vessel these coagula were of a dirty white colour, and were laminated and firmly adherent to the valves and diseased lining membrane; but towards its centre they were softer and less decolorized. The obstruction occupied the whole trunk of the vessel and extended a few lines into each division. The distal branches were free

from disease. The ductus arteriosus was impervious throughout the largest portion of its extent, but displayed a conical cavity, extending 2 or 3 lines from the bifurcation of the pulmonary artery. The pulmonary veins were natural. The left auricle was small and its lining membrane opaque. The auriculo-ventricular valves were healthy, and the aperture measured 36 lines in circumference. The left ventricle was of small capacity; its walls felt flaccid, and were $3\frac{1}{2}$ to $4\frac{1}{2}$ lines thick. The aorta communicated with this ventricle by an orifice of about the same size as that by which it arose from the right ventricle.

The aorta was very large from its origin to the sulcus marking the former point of communication with the ductus arteriosus. From that part its calibre greatly decreased. Its valves were three in number. They were free from disease, and completely closed the orifice. The coats of the vessel were natural. The bronchial and œsophageal branches were somewhat large.

The large veins and the several cardiac cavities were much distended with blood, chiefly fluid or feebly coagulated. The abdominal organs displayed no signs of disease, but were much engorged. The liver weighed 45 oz., the spleen $6\frac{1}{2}$oz., the left kidney $4\frac{1}{2}$ oz., and the right $3\frac{1}{2}$ oz. The latter was somewhat mottled.

Remarks.—The form of malformation of the heart in which the pulmonary orifice is contracted and the ventricular septum deficient, is not of uncommon occurrence; the combination, however, of a supernumerary septum in the cavity of the right ventricle with this malformation has, so far as I am aware, been very rarely observed.

When congenital perforations exist in the inter-ventricular septum, they are situated at that portion of its base which remains throughout life unprotected by muscular substance; and form communications between the origin of the aorta and the sinus of the right ventricle. When these communications are free and direct, the upper portion of the sinus of the right ventricle is usually found more or less distinctly sepa-

rated from the infundibular portion and origin of the pulmonary artery, by the column of muscle reflected from the pulmonary orifice along the upper and anterior border of the ventricle. A case well illustrating this condition was recently described by myself in the Monthly Journal of Medical Science.* In the present instance, however, the muscular mass referred to is of unusual size, and is combined with great enlargement of the fleshy column to which the anterior fold of the tricuspid valve is attached, and general hypertrophy of the walls of the ventricle, so as to produce a nearly complete septum in the centre of the cavity. Of cases of malformation similar to that now described I have only been able to collate four or five examples. Of these, one is reported by Mr. Holmstead ;† a second, of which the preparation exists in the Museum of St. Bartholomew's Hospital, is briefly described by Dr. Farre ;‡ and a third, bearing, however, a less close analogy to the present case, is related by Dr. Crampton, and subsequently by Dr. Todd.§ Dr. Theophilus Thompson has recorded a case in some respects similar ;‖ and a fifth has very recently been communicated to the

* March 1847, p. 644, vol. vii., or N. S. vol. i.

† Medical and Physical Journal, vol. xvii. p. 455, 1822. In a girl who died at the age of 9 years. The description is accompanied by a woodcut, showing the aorta arising from both ventricles, and the aperture in the supernumerary septum. The pulmonary artery is stated to have been naturally formed in this case. The foramen ovale was closed.

‡ Malformations of the Heart, p. 26. The age of the person is supposed to have been about 14, but there is no history of the symptoms during life. The malformation is very similar to that in the present case, with the exception that the pulmonary artery, though of small calibre, has three well-formed semilunar valves at its orifice. The foramen ovale appears to be impervious.

§ Crampton, Trans. of Dublin College of Physicians, N. S. 1830, vol. i. ; and Todd, Cyclop. of Anatomy and Physiology, vol. i. p. 614. The pulmonary artery was in this case destitute of valves, but there existed a puckering at its orifice which occasioned contraction. The foramen ovale was open, and the septum of the ventricles imperfect. The heart was that of a boy aged 10 years.

‖ Med.-Chir. Trans., vol. xxv., 1842, p. 247. In this instance the su-

Society by Mr. Le Gros Clark.* In all these cases the supernumerary septa were situated in the right ventricle, at or near the same point, and co-existed with some form of defective development at the orifice of the pulmonary artery; and, with the single exception of the case of Dr. Thompson, with an imperfection of the septum ventriculorum, so that the aorta communicated in part with the right ventricle. In three of the cases the irregular development might, as suggested by Dr. Todd, be referred to the obstruction at the pulmonary orifice, occasioning dilatation of the infundibular portion of the ventricle and the general hypertrophy of the walls of the cavity, and more especially of the fleshy columns forming the septum in its centre. This explanation is, however, inapplicable to the cases of Mr. Holmstead, Mr. Le Gros Clark, and Dr. Thompson, in which no contraction of the pulmonary orifice existed.

It is evident that this form of malformation must greatly interfere with the passage of the venous blood into the pulmonary artery, and, to an equal degree, favour its entrance into the aorta. In the case now related, after the occlusion of the ductus arteriosus, certainly not half the column of blood returned to the right auricle can have been transmitted to the lungs; yet the degree of cyanosis, till within a year of the boy's death, bore no just relation to this free intermixture of the currents of blood.† During this period, it is probable that

pernumerary septum co-existed with an excess in the number of pulmonary semilunar valves, there being four of equal size and well formed. The pulmonary orifice was one inch wider than that of the aorta. In the septum there was an aperture one inch in length, and half an inch wide. The patient, a female, 38 years of age, was healthy till she had an attack of the oriental cholera; her symptoms were aggravated two years before death.

* See page 113. Supernumerary septa are mentioned as occurring in the heart's cavities both by Andral and Rokitansky, but without any case of the kind being described. The latter states that they occur in both ventricles.

† In Mr. Holmstead's case, though the proportion of venous blood entering the general circulation was scarcely less than in the present instance, the child had no signs of any cardiac affection, and was, indeed, of a re-

the pulmonary artery was free from disease, and its defective capacity being compensated by the direct passage of the blood from the right ventricle into the aorta, no material disturbance of the balance of the circulation was occasioned. At the time of the accident, however, the pulmonary artery seems to have sustained some injury, which gave rise to the disease of its coats, and a further contraction of its canal; the communication between the right ventricle and aorta became no longer equal to the transmission of the column of blood required to compensate the decreasing circulation through the pulmonary artery; the right ventricle was imperfectly emptied of its contents; the congestion gradually extended throughout the venous system, and the lividity of the face and extremities became proportionately more marked. This view of the case tends to confirm the explanation of the cause of cyanosis, first suggested by Morgagni, and since more fully developed by Louis.

The observations of Baron* and Paget† have attracted attention to the formation of coagula in the pulmonary artery as a cause of death; so far, however, as I am aware, in the cases of this description observed, the coagula have been found in the branches of the pulmonary artery; and I have not met with any recorded instance of primary obstruction of the trunk of that vessel.‡ In this case, the diseased condition of the arterial tunics, the complete decolorization of the outer layers of coagulum, and their firm adhesion to the lining membrane of the vessel, render it most probable that the obstruction had been in progress for a considerable period, without having occasioned any urgent symptoms.

markably healthy appearance up to the age of three and a half years, when cyanosis supervened on an attack apparently of inflammation of the lungs.

* Arch. Gen. de Med. III^me^ et nouvelle série, t. ii. 1828, p. 6.

† Med.-Chir. Trans., vol. xxvii., 1844, p. 162; and vol. xxviii., 1845, p. 353.

‡ The case of M. Hélu, quoted by M. Baron, from the Bulletin de la Société Anatomique, Oct. 1837, may possibly be a case of primary obstruction of the trunk of the pulmonary artery. Arch. Gen. 1828, p. 22.

The final closure of the canal of the vessel appears, however, to have been a more acute process, and to have commenced coincidently with the rheumatic affection following the fall, eight days before death. In the drawing and preparation, the obstruction in the pulmonary artery appears less complete than it really was, owing to several masses of lymph, adherent to the valves and filling the spaces behind them, having been removed, to expose and render more evident the faulty conformation of that part of the vessel. Mr. Paget has remarked, in reference to the sixth case related in his first valuable memoir on obstructions of the branches of the pulmonary artery,* that when only two valves have been found at the pulmonic or aortic orifices, those valves have very generally been the seat of disease. The present affords a further illustration of this fact; and I have, in several instances of defect in the number of valves at the aortic orifice, seen their folds extensively diseased. The case related by Mr. Paget presents, in reference to the state of the pulmonary artery, considerable resemblance to that here described.

The physical signs which have been reported as present in this case, corresponded very closely with those observed in the other instance of somewhat similar malformation of the heart to which I have alluded.† In both, a loud murmur accompanied the systole of the ventricle, and was most intense at the seat of the aortic orifice. It was of a soft or blowing character, and was very distinctly audible, though gradually decreasing in intensity, throughout the course of the thoracic aorta. At the base of the heart, it was so prolonged as entirely to mask the second sound; but below that point, at the seat of pulsation of the apex, towards the epigastrium, and on the right side of the sternum, the murmur was feebler and shorter, and was succeeded by a second

* Med.-Chir. Trans., vol. xxvii. p. 182.

† They will also be observed to be similar to those assigned by Dr. Hope, partly on theoretic grounds, as characteristic of these forms of malformation.

sound of a peculiarly ringing or flapping character. It was also audible in a line from midway between the sternum and left nipple, towards the middle of the clavicle, though it was less intense here than along the upper part of the sternum. From these observations it may be inferred, that the murmur was chiefly occasioned by the meeting in the aorta of the two currents of blood proceeding from the right and left ventricle, and also, in part, by the constriction at the orifice of the pulmonary artery. The peculiarly ringing or flapping character of the diastolic sound is, doubtless, referrible to the powerful reaction of the large column of blood in the ascending aorta, on the semilunar valves at its origin. I have at present under my care a child about five years of age, in whom the physical signs are precisely similar to those here related; and these, together with the symptoms of obstruction at the right side of the heart which have been present from early life, have led me to infer the existence of similar malformation.

EXPLANATION OF PLATE IV.

This plate illustrates Dr. Peacock's case of Malformation of the Heart.

A, Infundibular portion of the right ventricle, with the orifice and trunk of the pulmonary artery laid open.

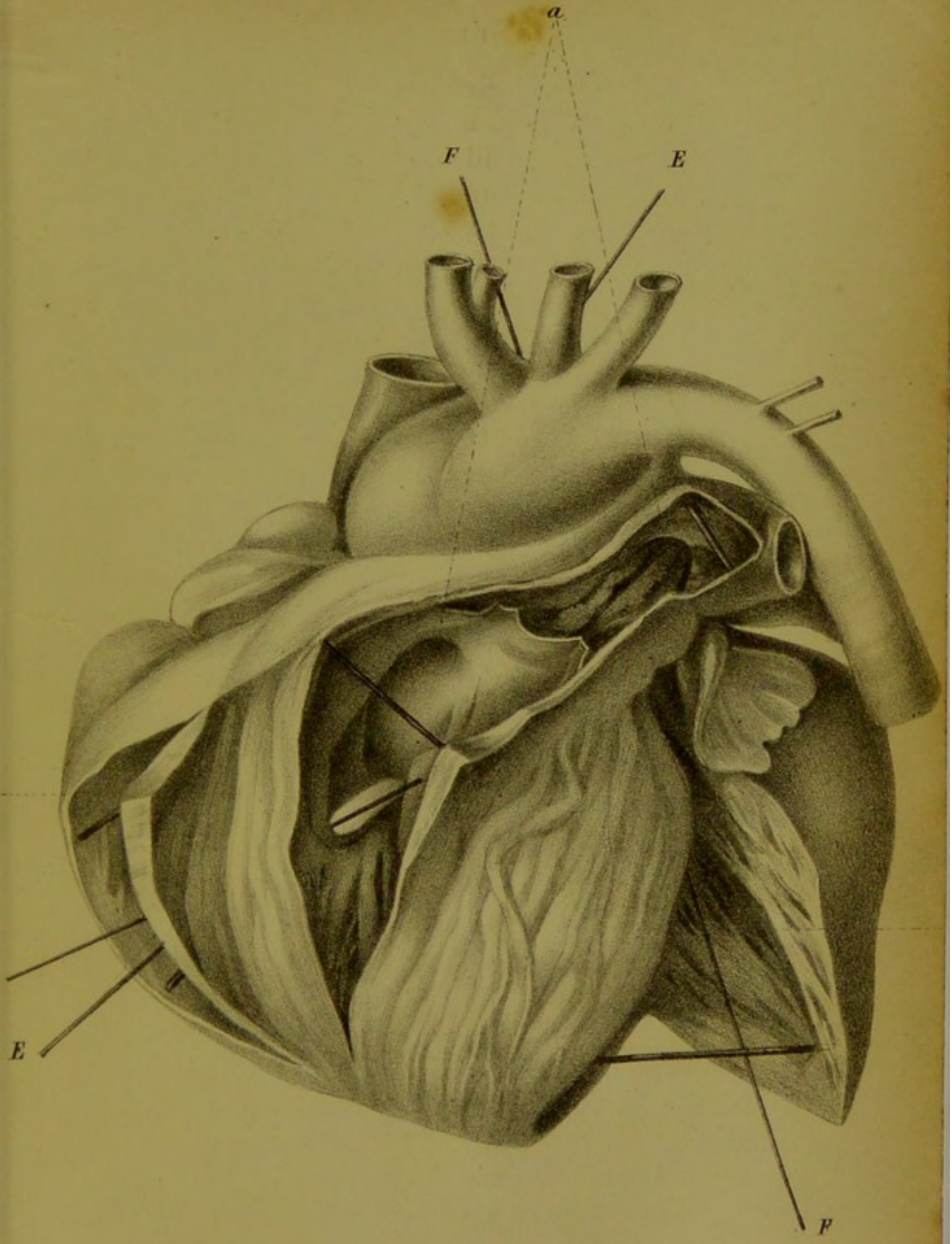
B, Portion of the anterior wall of the sinus of the right ventricle, turned back so as to expose the cavity.

C, Probe introduced into the opening in the septum, by which the two portions of the right ventricle are divided.

D, Cavity of the left ventricle laid open.

EE′ FF′, Probes passed from an opening in the posterior side of the aorta into the cavities of the right and left ventricles.

[*From Transactions of the Medico-Chirurgical Society*, Vol. xxx.]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