CLINICAL AND PATHOLOGICAL BEARINGS OF MALFORMATION OF THE HEART AS SEEN IN (A) PATENT FORAMEN OVALE; (B) IMPERFECT VEN- TRICULAR SEPTUM.¹

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WITHOUT preface I shall pass at once to a clinical description of two or three cases of malformation of the heart that have lately come under my observation.

A boy, aged 4, sent to me by Dr. Hardy, of the Gateshead Dispensary. He is smaller than most children of his age. There is marked cyanosis, the finger-nails and those of his toes being quite black. Finger ends are bulbous. Cannot walk a few yards without feeling faint or gasping for breath. In bed he lies on his hands and knees, his face on the pillow. Has no cough; never expectorated blood. Was not dark at birth. As an infant he cried a great deal. The discoloration has only come on within the last two years. There is a good family history, and there is no history of maternal shock. Pulse 94, extremely feeble; tongue is black and moist. Chest: nipples extremely dark; no distended jugulars in neck. Heart: apex felt in natural position, but is not well marked. Cardiac dulness is carried one inch to right of sternum, measuring three inches and a half. A soft systolic murmur is heard over the mitral area. Over the base of heart and mid-sternum a loud blowing and somewhat rasping systolic murmur is heard; it is faint as you approach the aortic area, but is loud and prolonged as you approach the pulmonary artery along which it is conveyed for about one inch and a half. Here the second sound, which is healthy, is loud and accentuated compared to that over the aorta. The maximum point of intensity of the murmur is the junction of the third and fourth left costal cartilages with the sternum. Lungs healthy. Liver enlarged; no pulsation detected in it. Spleen normal.

In this case there is not the least doubt about the existence of some communication between the right and left chambers of the heart. The little fellow cannot walk across the room without bringing on a severe attack of cardiac dyspnoea, relief to which only comes by his sitting down on the floor, bending the chest forward, and resting his elbows on his knees. In this case cyanosis was not present at birth. It was two years afterwards before it became pronounced.

I shall briefly relate another case, one which ended fatally. Rose K., aged 25, married, was admitted into the Royal Infirmary, Newcastle-upon-Tyne, under my care on November 29th, 1887. Nine weeks previously she had been confined of her first child, whom she suckled for seven weeks. Three months after her marriage, fifteen months ago, she had a miscarriage. Until three weeks ago, when she began to cough and to suffer from shortness

of breath, her health had been good. She had never been told that her lips were bluer than they ought to have been, or that she was the subject of heart disease. Her father died from bronchitis, and her mother from old age; and whilst nine brothers and sisters, out of a family of twelve, had died, no cause of death could be given by patient of any of them.

When admitted she was extremely cyanosed, was suffering so much from orthopnoea, that only a cursory physical examination could be attempted. Her chest and spine were deformed, pulse was very rapid and indistinct. There was dulness on percussion over the base of the right chest; here the respiratory murmur was distant and indistinct; moist crepitation and coarse respiration were noticed higher up. Left chest presented similar signs. Heart: apex beat was fairly distinct, and the sounds, though feeble, were healthy. The area of cardiac dulness was increased. By means of the aspirator a few ounces of serum were removed from the right chest. The abdomen presented nothing remarkable. Urine 1020, no albumen, no sugar. Patient died slowly asphyxiated.

Post-mortem Examination.—Heart is enlarged, right ventricle dilated. Towards the anterior border of the fossa ovalis is a slit-like opening, which admits a full sized catheter. It passes forwards and to the left, forming a channel of communication between the two auricles. The opening into the left auricle is guarded by a valve which could only have permitted blood to pass from right into left auricle. The walls of the right ventricle are unusually firm and fibrous. The tricuspid orifice is constricted, admits the tips of three fingers. Left ventricle normal in size; walls are somewhat thick. Aortic valve competent; mitral valve normal, except at free margin a few soft and oedematous vegetations are noticed. Pleural cavities contain a few ounces of serum; lungs oedematous. Liver 2½ lbs. in weight, capsule thickened, nutmeggy. The spleen and kidneys are all very loosely attached to their normal positions. The right kidney has a range of motion extending down to brim of pelvis. In the posterior wall of the stomach was found a small round ulcer towards the pylorus, in the neighbourhood of which the mucous membrane was found to be thickened and congested. In spite of the existence of a patent foramen ovale, we regarded this case *post mortem* as one of those in which, owing to the prolonged fixation of the diaphragm which occurs

Fig. 1.—Left ventricle of heart and aorta laid open—showing at (a) large opening, immediately below aortic valves, communicating by the interior of right ventricle.

in the latter months of pregnancy, and occasionally, too, in large ovarian tumours, the heart had undergone softening and degeneration.

I am deeply obliged to my colleague, Dr. Drummond, for having placed the *post-mortem* notes of one of his cases at my disposal, that of a man, aged 44, who died deeply cyanosed.

"The heart weighed 27 ounces. The mitral orifice was contracted, admitting the tips of two fingers. Foramen ovale patent; allowed the tip of little finger to pass from one auricle into the other. A thin cribriform diaphragm filled up the foramen, barring the opening referred to. Pulmonary artery was enormously dilated. Wall of right ventricle very much hypertrophied, as thick as left, measuring nearly one inch. Right auricle very much dilated; tricuspid orifice admitted four fingers easily; curtains thickened; aorta small. Branches of pulmonary artery of very large calibre, particularly the left. The left auricle is relatively

¹Read in the Section of Medicine, at the Annual Meeting of the British Medical Association, held in Glasgow August, 1888.

small; liver, kidneys, and spleen were well-marked instances of the induration met with in these organs in old-standing heart disease."

The next case is one in which the inter-ventricular septum was found to be deficient, and the pulmonary artery deformed. T. D., aged 25, a miner, was admitted into the Royal Infirmary on April 16th, 1888, complaining of pain in back and thighs. He is extremely sallow, and his lips are cyanosed; he looks at times so ill, and is so collapsed, that we cannot make a careful physical examination of him. Diffusible stimulants have had to be freely resorted to. His illness began four months ago, but from childhood he is known to have been the subject of heart disease. He is rather under the average size and weight. At the age of 14 he was cut for stone in the bladder. Right chest resonant in front; posteriorly there is dullness at the base, where the respiratory murmur is deficient. Heart: apex-beat is ill-defined. A loud blowing systolic murmur is heard internal to apex, midsternum, and just below aortic orifice, is not conducted in any direction. There is rather an absence of the second sound over the aortic area; its maximum of intensity is at midsternum. The systolic murmur is well heard over the base of the left lung. Nipples are extremely dark; abdomen presents nothing remarkable. Pulse extremely feeble, can scarcely be counted, 108; knee-jerk is absent. There is a good deal of muscular tenderness in the calf of left leg. He is the subject of recurrent muscular tremors. Nothing seemed to arouse him out of the extremely depressed and collapsed state into which he had fallen. Death supervened in a few days from the increasing asthenia.

Post-mortem examination made April 26th, and at which, unfortunately, I was not present. Heart weighs 10 ounces. There is considerable hypertrophy of the wall of the right ventricle; it is as thick as the left. There is a peculiar pendulousness and thickening, with perforation of the tricuspid valve, particularly at the tips, the edges of which are irregular. The orifice is normal. There is marked narrowing of the conus arteriosus, and constriction of the pulmonary artery, which only admits the tip of the little finger. The semilunar valve is composed of two segments, and allows of slight regurgitation. At the undefined space at the base of the septum is a large perforation, with smooth borders, readily admitting two fingers, and allowing thus of the freest intercourse between the two ventricles. This opening lies immediately below the aortic valves. Aortic and mitral valves are healthy. The ductus arteriosus is about the size of a large crow quill; its orifice at either end is patent, but its channel is occluded except for a few lines at each extremity. In the apices of the lungs are a few old tubercular nodules of small size, which have undergone cicatrization. The stomach unfortunately was not examined, nor the other viscera.

In order to understand these malformations, let us revise briefly the development of the heart. The heart originates in two layers of cells, arranged each in a linear manner; cells which pulsate rhythmically long before the mass which they compose has become tunnelled out to form a chamber. Pulsation is said to occur as early as the fifteenth day after impregnation. From being a straight tube, the heart gradually becomes incurved in the form of the letter S, and soon after this it is found to be divided into three cavities; the uppermost, which furnishes the aortic arches, becomes the bulb of the aorta; the lowest becomes the auricle, whilst the middle cavity becomes the ventricle. A further dividing off occurs. At the end of the first month the heart is

partitioned off at the level of the ventricles, and soon after this the two auricles are mapped out. The auriculo-ventricular orifices are noticed, and the aortic trunk is partitioned off into aorta and pulmonary artery. At the time of the division of the auricle, the vena cava is guarded at its entrance into the auricle by two valves—the left, the valve of the foramen ovale, and the other the valve of Eustachius. When the aortic bulb divides into two arteries, which become respectively the aorta and pulmonary artery, one part remains in its primitive condition, and allows of a communication being maintained between the aorta and pulmonary artery; this is the ductus arteriosus. Blood, therefore, passes in the foetus from right to left auricle, and from pulmonary artery into aorta.

"It is not a little curious," says Pettigrew,² "that in the foetal life of mammals, many of the peculiarities of the reptilian circulation reappear. Thus in the foetus the auricles communicate with each other by means of the foramen ovale; and the great vessels of the ventricles communicate by means of the ductus arteriosus."

At the early part of this and toward the close of last century, pathologists, influenced by biological doctrines then in vogue, were teaching that the human heart, before it reached its final development, passed through various stages, each of which found its counterpart permanently in the heart of beings lower down in the scale of existence. Each stage of the development of the human heart had its representative in that met with in the

various grades of animal existence. In other words, the heart of insects, crustaceans, and amphibians, was each in turn transiently represented in that of man. Whenever, therefore, a congenital cardiac lesion was observed there was the tendency to regard it as an arrest of development, and an attempt was made to establish the point at which the process of arrest had occurred. The pathological result was thus made to correspond with the physiological or normal of the lower animal. Opposed to this are the more purely pathological views of Cruveilhier, Rokitsky, and others, who maintain that many of these congenital cardiac lesions are the result of inflammation occurring in the foetus—for example, a myocarditis. In the foetus, as in the adult, all cardiac lesions

Fig. 2.—Interior of right ventricle—showing at (a) constricted orifice of pulmonary artery, (b) opening communicating with interior of left ventricle, (c) swollen tips of tricuspid valve, (d) hypertrophied wall of ventricle.

tend to obstruct the flow of blood from the heart; but whilst in the adult every obstacle is followed by cardiac dilatation and stasis, in the case of the foetus there is not only stasis, but, according to the date at which the lesion appears, so is the complete development of the heart interfered with, and the normal closure of certain passages prevented. The fact, for instance, that in any case of congenital heart disease the interventricular septum is perfect is to us evidence that the diseased condition, in all probability, arose after the ventricles were divided off from each other—say the end of the first month. Should disease arise, however, within the first month, then the closure of this septum will be interfered with. Whilst, therefore, I cannot but admit that many cases of heart malformation depend upon inflammatory affections arising during foetal life, most of them are really due to an arrest of development, and are caused by mechanical conditions arising during intra-uterine life, imperfect nutrition, owing to incomplete arterio-capillary development or thrombosis of blood-vessels, and possibly by maternal shock operating during the very earliest days of foetal life, disturbing thus either the normal multiplication of the primitive cardiac cells, or interfering with

² *Physiology of Circulation*, p. 99.

the proper relationship of individual cells to each other or groups of cells.

I am convinced that a foramen ovale may be more or less pervious, and the patient may be apparently not only none the worse for it, but there may be neither symptoms nor physical signs to indicate the fact. When pervious to any extent, however, it is generally associated with some obstruction in the pulmonary artery, as was shown by Morgagni. Anything, in short, which tends to increase right-sided intra-cardiac pressure will prevent the valve which guards the opening of the foramen ovale from becoming adherent to the margin, as it ought to be after the pulmonary circulation is established.

Fig. 3.—Pulmonary artery opened from above, and showing (a) orifice guarded by two instead of three segments in valve.

It is at the best but a very mixed blood that circulates through the vessels of the fetus. The impossibility of pulmonary respiration occurring in the fetus is no doubt one of the reasons why there exists transitory communications between the two sides of the heart and between the pulmonary artery and aorta; but, after all, this mixed circulation may be the best for the fetus. As Pettigrew says, it not only meets the requirements of the fetus; it is a positive advantage, as it places the fetus in the condition of the reptilia, which are very tenacious of life, and endure much hardship without receiving positive injury, qualities of the utmost importance to the fetus *in utero*, the position of which is being constantly changed. Whilst the closure of the foramen ovale after birth is generally ascribed to the floating up of the valve above the margin of the opening, and to the fold being retained in apposition by the pressure of blood now entering the left auricle after the establishment of the pulmonary circulation, Peacock questions this purely passive method of closure. In his *Malformations of the Heart*, page 108, he shows that the obliteration is effected: (1) by shortening of the cornua and the drawing up of the fold of the valve considerably above the edge of the foramen; (2) by the approximation of the cornua; and (3) by the diminution of the opening itself through the contraction of various sets of muscular fibres, some of which surround the opening itself, whilst others can be traced from the walls of the left auricle through the cornua of the valve to be expanded on the curtain.

Inequality of tension between the blood in the two auricles is, I think, one reason why the valve is not rendered permanently and completely adherent. A slit is left which may allow blood to pass only in one direction, as in one of my own cases, or the valve may so overlap the edge of the opening that interpassage is prevented. It only requires the compensation to be disturbed—say by such a circumstance as an attack of bronchitis or injury to the chest—and what was up till then a well-ordered circulation will become one in which compensation can scarcely, if ever, be recovered. I do not think that minor degrees of inter-auricular communication are of any great importance, and in this I share the opinions of Farre, Peacock, and M. Paul. The orifice may be so guarded that blood will only pass from right to left auricle or *vice versa*, but there is no real impediment to the circulation, and no trouble of any kind seems to arise from the slight admixture of blood. It requires that some intercurrent pulmonary illness should be developed, or that the heart itself should undergo degeneration and, asystole thus arising, cyanosis, palpitation, and other symptoms of cardiac disease appear.

In my case of imperfect interventricular septum, not only is there a sacculated condition of either end of the ductus arteriosus, but there is a marked constriction of the mouth of the pulmonary artery, and its valve is composed of two instead of three segments. This is a very frequent association. There is a kind of correlative variation noticed in these cases, so that when diseased conditions arise in certain cellular areas, it is found to be fol-

lowed by disease in others. The existence of two instead of three segments in the valve of the pulmonary artery is explained by Peacock through adhesion of the contiguous sides of two of the three valves, and atrophy of the corresponding angle of attachment. This fusion must have occurred very early in foetal life, and is, in my opinion, rather a defect of structure than the result of disease. Not so with the opening between the ventricles. Whilst, no doubt, a deficiency in the number of cells originally laid down in the primitive cardiac area for the completion of the septum would explain the defect, I believe this to be partly of mechanical origin, and explained by increased right intra-ventricular pressure through a gradual constriction of the pulmonary artery.

Hunter adopted the view that if obstruction arose by which the right ventricle could not completely empty itself into the pulmonary artery, the blood naturally passed through the opening in the septum, and thus a final and complete separation of the ventricles became impossible.

Meckel, on the other hand, regarded the incomplete septum as a primary defect, and saw how, by the blood finding an easy outlet from the right ventricle through the aorta, the pulmonary artery might become abortive, and its orifice contracted during the progress of development. The tendency is for the septum in these cases to be carried to the left, and for the origin of the aorta to be drawn to the right, so that, as in my case, the aorta arises almost as much from the right as from the left ventricle; at any rate, blood had immediate access to the aorta from the right ventricle, and thus do we find an explanation of the hypertrophy of the right ventricle which is present in these cases.

Laennec, Bertin, and Bonillaud supposed that the entrance of aerated blood through the septum was the cause of the hypertrophy of the right ventricle, but I think we must admit that here, as elsewhere, it is entirely a question of the amount of work the ventricle has to do. Constriction of the pulmonary artery would alone be sufficient to induce hypertrophy of the right ventricle; but when, owing to the existence of an aperture in the septum, blood can be sent on directly into the aorta, the right ventricle, thus participating in the work of the systemic circulation naturally hypertrophies.

As regards symptomatology, one would naturally expect, seeing that we are dealing with a malformation of the heart arising during intra-uterine life, that symptoms would be present from birth. In many of these cases, no doubt, the children when born are found to be deeply cyanosed; but in others cyanosis, which may have been present, by degrees disappears.³ If the heart is greatly malformed, cyanosis will in all probability be present from the first. It is cyanosis, in fact, of a deep, livid, almost black colour, affecting the surface generally, but seen particularly in the face, the hands, and feet, which is the one symptom more or less pathognomonic of cardiac malformation; and particularly is this the case if there are paroxysms of difficult or gasping respiration. These children scream a good deal, partly from terror, and thus do they induce the attacks of difficult breathing they are so much in dread of. The finger-nails are generally very black; the finger-tips are bulbous, the so-called Hippocratic finger, and the external temperature is low. It is said that the heart's action is more rapid than in healthy children. Children thus afflicted assume certain attitudes in which they obtain relief—the very young lies placed on the nurse's lap face downwards, an attitude preferred in later days by such children when in bed; or, again, the knee-and-elbow position when sitting on a chair or on the floor.

Whilst cyanosis persisting from birth is the one symptom characteristic of malformation of the heart, M. Paul maintains that before the child is born there can be detected on auscultation a systolic *bruit* instead of a normal first sound. And whilst a few of these murmurs have subsequently disappeared, so much so as to raise the question of the existence of functional cardiac murmurs in foetal as in adult life, M. Paul insists that when cardiac murmurs have been detected in the fetus they are, as a rule, persistent, and if these cases are watched they are found to be afterwards cases of malformation of the heart.

Various theories have been advanced to explain the cyanæmia. It was the opinion of Morgagni, as also of Corvisart, Louis, and

³ In a very interesting case of univentricular, or tricocclian, heart, reported by Drs. Elliot and Embleton, cyanosis was not observed until three months after birth. The lad died in his twentieth year. In addition to there being only one ventricle, the positions of the aorta and pulmonary artery were reversed.

Laennec, that the deep discoloration was due to a general congestion of the venous system caused by obstruction of the pulmonary artery. That it cannot be due to this has been shown over and over again (there is no enlargement of the external jugular veins); its presence to a very marked degree in Dr. Drummond's case, where the pulmonary artery was enormously dilated, bids us look for another explanation. The freedom of communication noticed between the two sides of the heart in nearly all of these cases led Bouillaud, Hope, Walshe, and others to regard the intermixture of arterial and venous blood as the cause of the cyanosis; but the facts as pointed out by Stillé that cyanosis may exist without intermixture of the currents of blood, that there is no just proportion between the cyanosis and the intermixture, and that frequently there is intermixture without cyanosis, compel us to admit that this alone cannot be the explanation. Besides, as I pointed out in the early part of the paper, patent foramen ovale may exist for years, and there may be no cyanosis. So long, in fact, as the pressure on the two sides of the heart is pretty equal, even though there be a defective septum, there may be admixture of blood to any extent, and it only requires in such that this balance should be disturbed by some intercurrent pulmonary affection, by an attack of rheumatism or some other blood disease disturbing the nutrition of the heart, and cyanosis will appear. Peacock adopts the theory of congestion of the venous system, and lays stress upon the fact that the cyanosis of cardiac malformation is as a rule much more intense than that from any other cause. Of modern writers, Sansom adopts the theory of "intermixture of the blood." This I accept admitting, however, that one or two factors are introduced which aggravate it: central, as seen in cases where the heart undergoes degeneration or the intracardiac pressure becomes unequal; peripheral, as seen in the cyanosis of children immediately after birth, in whom, owing to the thin, transparent condition of the skin, and the distensible character of the capillaries, allowing a larger amount of blood than is usual to be retained in the veno-capillary system, we have a condition apt to be perpetuated by the difficulty which the arteries offer naturally to the circulation of the venous blood. Oxygenation is thus diminished through the constriction of pulmonary vessels, and possibly through the inefficient action of the lungs, owing to the deadened condition of the respiratory centre, from the long continued action upon it of venous blood. In support of the influences of the peripheral conditions already referred to is the great relief to the paroxysmal dyspnoea when the child is placed in a hot bath. Thus are the cutaneous vessels dilated, circulation through them promoted, as well as interchange of gases effected.

As regards the other points in the diagnosis of these cases, I shall say little. The paroxysms of dyspnoea, occurrence of epileptiform seizures, the existence of a systolic *bruit* at base of heart or mid-sternum, and with no definite line of conduction, enlargement of the area of cardiac dulness to the right, are all suggestive of the existence of malformation of the heart; but as to what the actual malformation is, I do not think we can regard the evidence obtained from auscultation or percussion as positive.

It is interesting to note that many of those who are the subjects of malformed heart live to a good age. M. Fallot's case of moderate constriction of pulmonary artery lived to 63; Peacock mentions a case of patent foramen ovale reaching 57; in Dr. Drummond's case it was 44; and in cases of constriction of pulmonary artery with imperfect septum, the age of 39 has been reached; in my case it was 25.

CLAY AND WAX MODELLING OF THE LIVING URINARY BLADDER UNDER ELECTRIC LIGHT.

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ALTHOUGH Mr. Pearson-Cooper and I have been at last able, after overcoming great difficulties, to obtain good photographs of artificial growths in male bladders *post mortem*, yet our vesical camera will have to be considerably improved before we can lay before the profession a practical apparatus for obtaining faithful negatives of the mucous membrane of the living viscus in health and disease. But, until we are able to thus graphically record the many new and interesting clinical facts which the electric cysto-

scope is constantly revealing, I wish to advocate a substitute which I have employed for some time, and with considerable advantage. I refer to modelling in some plastic material the interior of the living bladder as it appears illuminated by electric light (the Nitze method). The changes in the aspect of the mucous membrane of the bladder, produced by relaxation, congestion, or infiltration are so varied and often so remarkable, that it is only by systematically accumulating a record of these appearances that a sure basis for establishing a sound diagnosis, prognosis, and treatment of vesical disease upon visual grounds can be acquired.

Drawings in pencil, pen, or colour are most valuable if carefully taken, but they fall far short of clay or wax modelling in conveying to others an exact idea of the disease depicted. The reasons for this are obvious. The cystoscopic field from which the artist draws is small, and the area to be portrayed is often large; hence a number of drawings is generally necessary to represent the disease in its entirety. The cystoscopic field changes with the slightest movement of the patient or the operator. Even a cough, a deep inspiration, or a slight involuntary vesical contraction is sufficient to puzzle a non-professional artist, by suddenly displacing or distorting some salient feature which he may have taken as a "fixed point." Moreover, if bleeding should occur before the painting or drawing is completed, the transparent medium has to be renewed, and it is often difficult so to redistend the bladder as to obtain the same view of the object as before.

Fig. 1.

The plan I have lately adopted of making clay or wax models of the diseased living bladder is similar to that which I recommended at last year's Surgical Congress¹ in Berlin, for recording the changes of shape in the living prostate.² It is easily carried out, and only needs a small bowl or its shape-equivalent, a little oil, a penknife, and a handful of sculptor's clay.

As an example, suppose we wish to retain a record of an epithelioma of the bladder in an early stage. The clay is dropped into the bowl, and flattened out into a thick layer. A concave surface is thus formed, which will roughly represent the interior of the bladder. A little oil previously smeared over the inside of the bowl will permit the model to be subsequently turned out after drying. The bladder is now carefully examined in all parts by electric light, and the diseased section localised. Each part of the tumour is next observed; its size and the character of its surface appreciated. The light is switched off, and the handle of the cystoscope given to the patient or the assistant to hold, or it may be laid on a scrotal cushion. A fresh piece of clay, the size of the tumour, is

¹ Ueber Thonabdrücke der Prostata am Lebenden, Langenbeck's Archiv, Bd. xxxvi, Heft 2.

² JOURNAL, May, 1887.