ON TWO CASES
OF
MALFORMATION OF THE HEART.

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COMMUNICATED BY
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Received June 18th, 1856.—Read June 24th, 1856.

THE two cases of malformation of the heart described in this memoir are of extreme interest ; the first, as an instance of anticipation of the adult condition of the heart during foetal life ; and the second, as an instance of that rare and opposite condition, in which the three great elementary cavities of the heart have undergone hardly any fission at all.

CASE 1.—On January 5th, 1856, I delivered a woman of a female child, which gave hardly any indications of vitality for some minutes after birth. It ultimately respired pretty freely, but soon became very blue, and about an hour after birth relapsed into its original feebleness. The pulse became weaker, slower, and intermittent. On applying my ear to the infant's chest, I found that the natural cardiac sounds were replaced by two loud blowing murmurs ; the rhythm of these murmurs corresponded to the rhythm of the natural sounds—thus there was first *bruit*, second *bruit*, and pause—and the number of rhythms per minute was but little below the standard of a healthy new-born child. Blueness

of the surface increased, difficulty of breathing supervened about two hours after birth, and augmented rapidly, and four hours and a half after birth, having been convulsed two or three times, it died.

On examining the body of the infant three days after its decease, the following abnormalities of the thoracic viscera were discovered. As regards the heart—the right auricle was larger than natural, and its parietes were very thin. The foramen ovale was closed, or in other words the septum of the auricles was totally imperforate, there being nothing more than an oval depression in the usual site of the foramen. The tricuspid valve was incompetent; and formed rather a festooned imperfect diaphragm between the auricle and ventricle, than a valve. The right ventricle gave origin to two vessels; one of these immediately divided into a right and left pulmonary artery, it neither communicated with nor was attached to the aorta, and was of less calibre than usual; the second vessel was the aorta, which arose in part from this cavity, and in part from the left ventricle. The common origin of the aorta from both ventricles was the result of an imperfect septum ventriculorum, the deficiency occurring at the upper part of the septum. The left ventricle was small; the mitral valve was perfect; the left ventricle communicated with the right ventricle at its upper part, as before described, and gave partial origin to the aorta; the semilunar valves were perfect. The lungs were very imperfectly inflated, and were situated at the back of the chest, purple, and gorged with blood. The rest of the body was apparently well formed, but rather smaller than usual.

The course of the circulation during intra-uterine life would appear to have been as follows: all the blood returned by the *venæ cavæ* into the right auricle, except that which regurgitated, was propelled through the right ventricle into the aorta. A small proportion only of blood passed into the pulmonary artery; the left ventricle was only concerned in returning to the general circulation the small quantity of blood which had passed through the lungs. Thus before

birth there was regurgitation of blood through the tricuspid valve, and admixture of venous and arterial blood in the aorta at its very commencement. The physiological result was that the head and upper extremities, instead of receiving better oxygenated blood than the trunk and lower extremities, received blood of similar quality. As soon as birth took place, and the left instead of the right side of the heart became the arterial side, the faulty development, and the narrowing of the pulmonary artery became productive of fatal consequences. The errors of circulation after birth were three—

- I. Regurgitation from tricuspid incompetency.
- II. Admixture in the aorta of venous and arterial blood, from imperfection of the septum ventriculorum.
- III. Deficient supply of blood to the lungs, from narrowing of the pulmonary artery.

If it were not for one circumstance, I should not have thought this case worthy of particular notice. It is the type, but for this exceptional addition to its abnormalities, of a large class of malformations of the heart. Partial deficiency of the septum ventriculorum, with narrowing of the pulmonary artery, is, perhaps, the commonest cardiac defect, of a congenital origin, known; with, perhaps, the exception of patency of the foramen ovale. But the fact that the septum of the auricles was totally imperforate, and that there was for a considerable time during foetal life no communication between the right and left auricles, rescues the case from the category of familiar irregularities of development. Malformations of the heart are in general “per defectum;” the present one is manifestly “per excessum.” Imperforation of the cavities of the heart is said to be extremely rare. M. Geoffroy St. Hilaire states, in his work ‘On Anomalies of the Organization,’ that an instance of it had never come under his own observation. Whether this statement refers to an imperforate condition of the septum of the auricles at birth, I cannot say, but

from an attentive perusal of that section which treats of abnormalities of the vascular system, I have been unable to discover that that eminent teratologist was acquainted with the condition in question. Since the time of St. Hilaire, however, a case has been recorded by Mr. Ebenezer Smith, in the first volume of the 'Pathological Transactions.' The history of this case is very similar to mine, but the child lived twenty-one hours. The points of difference are two; in the first place, it appears that the closure depended simply upon adhesion of the valve of the foramen to its margins, whereas in the case I have related, the septum of the auricles was fleshy throughout; and secondly, in Mr. Smith's case, the pulmonary artery was wider than natural, while it was narrower in mine. Dr. Peacock informs me that another case of premature closure of the foramen ovale is recorded by Vieussens, but I am unable to refer to it more particularly, having failed to discover where it is related.

CASE 2.—On May 12th, 1856, I delivered a woman of an apparently healthy and robust male infant. On the 18th, I discontinued attendance, as both mother and child appeared to be perfectly well. Early in the morning of the 20th, a messenger was sent to say that the child had just died. The account given was that on the previous afternoon the infant began to pant, that the difficulty of breathing increased, and the skin became discoloured, that during the night the child was convulsed, and that it died at 7 o'clock a.m. The cause of death not being apparent, I demanded a post-mortem examination of the body.

The heart presented the following singular series of defects; The right auricle was large and more fleshy than usual. The foramen ovale was of greater area than in the normal condition. The valve of the foramen was unusually large and membranous, and was attached posteriorly to a fleshy column, which arose from the upper and posterior part of the auricle, and inserted itself into the ring at the base of the ventricular portion of the heart, rather to the left. This

fleshy column constituted, together with the valve of the foramen ovale, all that existed of an inter-auricular septum. The left auricle was very small, and, in fact, a mere diverticulum from the right auricle; it received *two* pulmonary veins, one from each lung, and communicated with no other cavity than the right auricle. The ventricular portion of the heart was single, and did not present the slightest vestige of a septum. The valve between the right auricle and the general ventricle, consisted of two parts. The largest of these was anterior, and to the left; it was attached, in the usual manner, by *cordæ tendineæ* to two well-defined papillary muscles of considerable thickness; the largest of these muscles arose from the anterior surface of the cavity of the ventricle, about one third of the length of the whole cavity from the apex of the ventricle. The smaller papillary muscle was attached by its base to the extreme right of the ventricle, and it was not so long as the other muscle; its *cordæ tendineæ* were not all inserted into the larger or anterior segment of the valve, some went to the posterior division. The posterior segment was situated more to the right than the former one; it was not so free, inasmuch as the tendinous cords attached to it were very short, and arose principally from *columnæ carneæ* on the posterior surface of the ventricle; it was neither so wide at its base nor so deep as the anterior division of the valve; it had not the distinct apex of the other portion, and its free border was wavy and irregular. A single vessel arose from the general ventricular cavity; it was much larger than either the aorta or the pulmonary artery taken alone, and was more decidedly bulbous at its origin than the normal aorta. The branches from the main trunk were as follows: first, two vessels arising from the posterior aspect of the ascending part of the aorta, these were a right and left pulmonary artery, and their aortic embouchures were close together. Then a third vessel arose from the summit of the aortic arch; this divided into the *innominata*, left common carotid, and left subclavian arteries. The aorta then continued its course downwards, as usual. No coronary arteries could be discovered,

until attention was directed to a vessel which coursed downwards, towards the base of the heart, from the innominate artery; at first this vessel appeared to terminate in a cul de sac, and I was at a loss to determine what structure it represented; subsequently, after more careful examination, it proved to be a common coronary artery.

The heart and great vessels were distended with black blood. The lungs were imperfectly expanded, so much so that their bulk was certainly not equal to more than one third of the usual dimensions; they were slightly crepitant throughout, more so at the apices than the bases; they were liver-coloured, and situated quite at the back of the chest, and did not overlap the heart, or displace the abdominal viscera sensibly from their foetal position. The most striking phenomenon in connection with the pulmonary organs was, that scattered over the anterior aspect of the superior lobes, there were small rosy coloured elevations, about the size of a millet-seed; each of these elevations appeared to consist of isolated portions of expanded pulmonary tissue. On inflating the lungs by blowing down the trachea, these patches disappeared, and the lungs then presented an homogeneous surface of the colour of the expanded lung in children newly born.

The course of the circulation before and after birth would appear to have been the same. . In consequence of the non-communication of the left auricle with the ventricular cavity, the right auricle was the recipient of the aërated blood after as well as before birth; and all the venous blood in the body, whether belonging to the pulmonary or systemic circulation, found its way into that cavity. From the right auricle, the blood passed into the general ventricle, and was then propelled into the general aorta; from this vessel came off two pulmonary arteries, which were wholly inadequate to the purpose of passing a sufficient quantity of blood to the lungs. The nature of the error of circulation after birth would appear to have been this: that instead of the whole mass of the venous blood being passed through the lungs before being again sent into the systemic capillaries, only

about one third or one fourth was propelled into the aerating organ, and that even this fractional part of the blood, instead of finding its way into the systemic capillaries, was again partially diverted into the pulmonary circulation. Assuming, for purposes of illustration, that one fourth of the blood projected from the ventricular cavity found its way into the pulmonary circulation, and that that blood, on its return from the lungs, became intimately mixed with the blood in the right auricle, it would follow, that one fourth of the decarbonized blood, or one sixteenth of the whole mass, re-entered the pulmonary circulation, and that three fourths of the aerated blood, or three sixteenths of all the blood propelled into the aorta was aerated blood destined for the service of the economy at large.

It is perhaps hardly possible to conceive a heart more radically malformed, which should even for a short time subserve the necessities of the economy; the veins, the cavities, the valves, and the arteries, all deviate widely from the normal condition. There are certainly very few instances recorded of such extreme deformity, and I do not know of one exactly similar. Subjoined is a sketch of some of the most analogous cases hitherto published, and other cases are referred to merely.

A. In the first volume of the 'Pathological Transactions,' there is a case related by Mr. Forster, more like mine than any I have met with. There was one large auricle with the same number of veins opening into it, viz., five; the valve appeared to have been similar; the single ventricle gave off a single vessel, and the arrangement of the branches from this aorta was like that in the case described above, except that the coronary artery came off from the concavity of the arch of the aorta, instead of from the innominate, and it subdivided before reaching the heart. There was no rudimentary left auricle in Mr. Forster's case. The child lived seventy-eight hours, and died cold and livid during an attack of dyspnoea.

B. Dr. Ramsbotham reports a case on the same page of the 'Pathological Transactions' as Mr. Forster's, very similar

to both the preceding ones. There was only one auricle and one ventricle. A single vessel arose from the ventricle, and from this a branch was given off which subdivided into two pulmonary arteries. The child lived ten days, and was completely cyanotic, but neither respiration, temperature, nor muscular action appeared to be affected. The case is figured and described in the ninety-ninth vol. of the 'Philosophical Transactions.'

c. A case is described at page 49 of the first vol. of the 'Pathological Transactions,' by Mr. Crisp, in which there was a single auricle and a single ventricle. A single vessel arose from the ventricle, which divided near its origin into branches, the destination of which is only surmised. There was a rudimentary vessel also behind the aorta, and another artery, the origin and destination of which does not appear to have been determined.

d. A case is related by Martin Saint Ange, in the third number of the 'Bullet. de la Société Anatomique' for 1826, of a child which died six weeks after birth, having been subject to convulsions and vomitings. There was general transposition of the viscera. The heart consisted of one auricle and one ventricle; but of the state of the arteries I cannot find a description. The colour of the skin was livid, the child was habitually somnolent, and the temperature of the back and lower extremities was lower than that of the rest of the body.

e. Dr. Farre relates a case, in his 'Essay on the Heart,' which very closely resembles Mr. Forster's case and mine. The child survived its birth 79 hours. The respiration became hurried, the diaphragm laboured excessively, the skin became cold, and the face somewhat livid, but not blue. Torpor ultimately came on, and the respiration became more and more feeble until death occurred. There was a single auricle, a single ventricle, and a single artery from the ventricle. The single artery behaved as in the case I have related, except that the coronary vessels were differently arranged. There was not either any rudimentary left auricle.

F. Jackson relates the case of a child which died at the age of three years, in which the respiration was very irregular; but there was no blueness of the skin. The interauricular orifice was found patent, and there was a large opening in the inter-ventricular septum. The arteries were also abnormal. This case is a minor instance of the same developmental defect, which in the preceding cases resulted in hearts practically single.

G. There is a case described by Professor Owen, in vol. 2, p. 664, of the 'Lancet' for 1848, which is very similar to mine.

H. Dr. Hale's case, in the 4th vol. of the 'Pathological Transactions' is another intermediate case of malformation. The auricular portion of the heart was double, the ventricular single, and there were two main trunks arising from the ventricular cavity. The child lived nineteen weeks, subject to dyspnœa and vomitings; it was found dead in bed one morning. Dr. Hale refers to analogous cases by Kreysig, Tiedemann, and Hufeland. His own case does not appear to have been cyanotic. Numerous other cases of malformation of the heart are described by Fleischman, Chemineau, Otto, Dr. Cheevers, and others; but none, I believe, present a more complete arrest of development than the cases reported by Mr. Forster and myself.

In speaking of the case I have related as an instance of arrest of development, I refer, of course, to the well-known fact that, before about the ninth week of embryonic life, the rudimentary heart consists merely of three dilatations of the main vascular trunk: the auricular sinus, the ventricular sinus, and the bulbus arteriosus. At the period mentioned, ridges appear in three sinuses which meet in the centre, and ultimately divide each cavity into two.

Reference to the cases I have quoted and referred to will justify, I think, the conclusion that cyanosis is the constant attendant of no single species of malformation of the heart. Dr. Reeder advanced an opinion that, in cases where there was patency of the foramen ovale or an hiatus in the inter-

ventricular septum, cyanosis or other inconvenience does not necessarily occur so long as no obstruction is offered to the course of the blood in the lungs, or elsewhere, and for the following reason, viz., that the right and left sides of the heart being nearly equal in size, acting simultaneously and with nearly equal force, it does not at all follow that any great amount of admixture of blood shall take place simply because the cavities communicate; but as soon as any obstacle is offered to the exit of the blood either from the right or left sides of the heart, the balance is destroyed, and admixture then takes place. This explanation seems applicable enough to a limited number of cases; but it will not account for all. The following general conclusions with regard to cyanosis appear to be safe:

1. That it may be connected with abnormalities of the great vessels only.

2. That it is not so necessarily connected with abnormalities of the orifices of the heart as always to be present when they are.

3. That there may not only be patency of the foramen ovale, but defect in the interventricular septum also, and yet no blue disease.

4. That it may be the result of veritable pathological changes, such as adhesions of the lungs to the pleuræ and pericardium (*vide* Marc, Tartra, and Gilbert, in the 'Bullet. des Sc. Med. '); and

5. That if any one condition be more inseparably connected with cyanosis than another, it is narrowing of the pulmonary artery or such abnormality as diminishes the supply of blood to the lungs, especially if such abnormality be of congenital origin.

EXPLANATIONS OF THE FIGURES.

FIG. 1.

- A, A, A, or a, a, a.* The general aorta.
- b.* The aortic embouchures of the right and left pulmonary arteries.
- c, c.* The origin and trunk of the brachio-cephalic vessel.
- d.* The left subclavian artery.
- e.* The left common carotid artery.
- f.* The innominate artery, which afterwards divided into the right common carotid and right subclavian arteries.
- g.* The common coronary artery, coursing downwards from the innominate artery to the base of the heart.
- h.* A probe passing through the left auricle and the imperfect septum auricularum into the right auricle, and thence into the cavity of the general ventricle, where its other extremity may be seen.

The similunar valves may be seen to be perfect, and the anterior segment of the auriculo-ventricular valve is conspicuous in the general ventricular cavity. The aorta is divided longitudinally.

FIG. 2.

- a.* The left or anterior segment of the auriculo-ventricular valve.
- a'.* The papillary muscle of segment *a*.
- b.* The right or posterior portion of the same valve.
- b'.* The papillary muscle of *b*.
- c.* The fleshy column, which, with the valve of the foramen ovale and another small column (*e*), constitutes the rudimentary septum auricularum.
- d.* The valve of the foramen ovale.
- e.* A small fleshy column, which defines the position of the foramen ovale.
- f.* A probe passed behind the valve, &c., into the left auricle, and thence into the right auricle again.

Fig 1.

Fig 2.