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Congenital Affections of the Heart.

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CONGENITAL AFFECTIONS OF THE HEART

CONGENITAL AFFECTIONS OF THE HEART

BY

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PREFACE.

At my out-patient clinique at the Evelina Hospital for Children, I have so frequently been asked by students for demonstrations on the congenital affections of the heart, and complaint has so often been raised that the ordinary text books are not sufficiently precise and explanatory of the subject, that I have for some time past been in the habit of giving them demonstrations on these affections. These demonstrations I now publish in book form without any intention or desire of rivalling the classic works of Rokitansky, Kussmaul, Peacock, and others, but inasmuch as they have proved of service to my classes, they may, I trust, be equally useful to other students and practitioners.

George Carpenter.

12, Welbeck Street,
Cavendish Square, W.
January, 1894.

CONGENITAL AFFECTIONS OF THE HEART.

The heart, in its earliest embryonic condition, appears as two tubes which lie on the ventral aspect of the embryo immediately under the head. Subsequently, with the development of the neck, it assumes a position further back. These tubes very soon coalesce into a single one, which posteriorly receives the vitelline veins, and anteriorly divides into two branches, the rudimentary aortas. The heart at this period is median in position, but soon afterwards becomes more or less obliquely placed. The anterior portion, consisting of the ventricle and bulbus arteriosus, deviates to the right, the posterior portion now forming the sinus venosus and auricle to the left. At the same time a bend occurs, and the auricular portion becomes, with regard to the ventricular, dorsal

in position. The cavities are next differentiated, so that ultimately two auricles and two ventricles are formed. It is to be noted that in the partition between the auricles a special aperture is constructed which, in the course of development, becomes closed by a distinct and separate septum of its own. This aperture is the foramen ovale.

The Eustachian valve appears as a fold of the lining membrane of the auricles. The septum between the ventricles is not for some time fully competent above, thus allowing a free communication between the two ventricles at its upper portion. On its completion this is termed the undefended space. This is an area of triangular shape, in which the ventricles are only separated by the endocardium and fibrous tissue on the left side, and by the lining membrane and a thin layer of muscular substance on the right. Its base is formed by the muscular substance of the septum, its sides by the attachments of the right and posterior aortic valves.

It has already been mentioned that the bulbus arteriosus divides into two branches or

primitive aortas. These branches are placed in the branchial arch, and encircle the body,

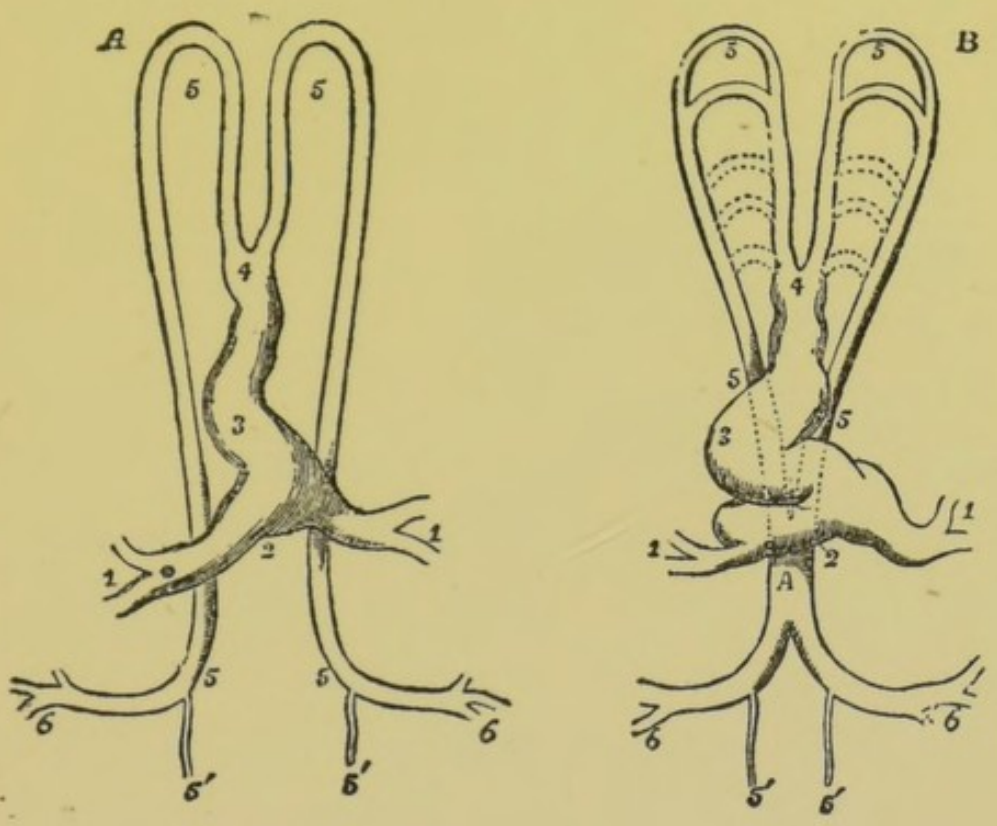


FIG. 1.—DIAGRAMMATIC OUTLINES OF THE HEART AND FIRST ARTERIAL VESSELS OF THE EMBRYO, AS SEEN FROM THE ABDOMINAL SURFACE. (After Quain.)

A, at a period corresponding to the 36th or 38th hour of incubation in the chick; B, at the 48th hour of incubation; 1, 1, primitive veins; 2, auricular part of the heart; 3, ventricular part; 4, aortic bulb; 5, 5, the primitive aortic arches, and their continuation as descending aorta; these vessels are still separate in their whole extent in A, but at a later period, have coalesced into one tube in a part of the dorsal region; in B, below the upper 5, the second aortic arch is formed, and farther down the dotted lines indicate the position of the succeeding arches to the number of five in all; 5,′ 5′, the continuation of the main vessels in the body of the embryo; 6, 6, the omphalo-mesenteric arteries passing out of the body of the embryo into the vascular area of the germinal membrane.

being continued on the ventral aspect above the alimentary canal. These are known as the anterior and posterior aortic roots, *vide* fig. 1. At a somewhat later period four other arches are formed, placed posteriorly to the first pair, and are continued in the same manner through the corresponding branchial arches to open into the primitive aortas. The bulbus arteriosus itself becomes divided by a septum into the aorta and pulmonary artery. This septum grows downwards from between the fourth and fifth branchial trunk of the left side, and first separating the truncus arteriosus into the aorta and pulmonary artery is further continued, so as to divide the bulbus arteriosus into the conus venosus and conus arteriosus. This septum begins at the back of the common efferent trunk, and having its broad surfaces at first directed laterally, twists as it grows downward, so that the left hand one comes almost to look directly forward, and the right hand one backward. Only the fourth and fifth branchial arches of the left side remain permanently as the aorta and pulmonary artery respectively, the rest in part disappearing.

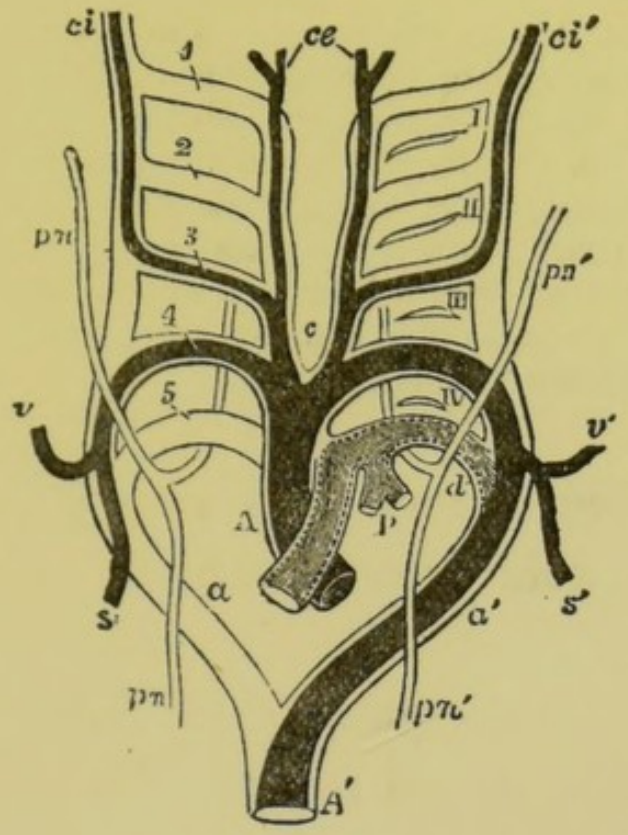


FIG. 2.—DIAGRAM OF THE AORTIC OR BRANCHIAL VASCULAR ARCHES OF THE MAMMAL, WITH THEIR TRANSFORMATIONS GIVING RISE TO THE PERMANENT ARTERIAL VESSELS (according to Rathke, slightly altered). (After Quain.)

A, P, primitive arterial stem or aortic bulb, now divided into A, the ascending part of the aortic arch, and P, the pulmonary; *a*, the right; *a'*, the left aortic root; A', the descending aorta. On the right side, 1, 2, 3, 4, 5, indicate the five branchial primitive arterial arches; on the left side, I., II., III., IV., the four branchial clefts, which, for the sake of clearness, have been omitted on the right side. It will be observed, that while the fourth and fifth pairs of arches rise from the part of the aortic bulb or stem, which is at first undivided, the first, second, and third pairs are branches above *c*, of a secondary stem on each side. The permanent systemic vessels are represented in deep shade, the pulmonary arteries lighter; the parts of the primitive arches, which have only a temporary existence, are drawn in outline only. *c*, placed between the permanent common carotid arteries; *ce*, the external carotid arteries; *ci*, *ci'*, the right and left internal carotid arteries; *s*, the right subclavian rising from the right aortic root beyond the fifth

arch ; [*v*, the right vertebral from the same opposite the fourth arch ; *v'*, *s'*, the left vertebral and subclavian arteries rising together from the left or permanent aortic root opposite the fourth arch ; P, the pulmonary arteries rising together from the left fifth arch ; *d*, the outer, or back part of the left fifth arch, forming the ductus arteriosus ; *pn*, *pn'*, the right and left pneumogastric nerves, descending in front of the aortic arches, with their recurrent branches represented diagrammatically as passing behind, with a view to illustrate the relations of these nerves respectively to the right subclavian artery (4) and the arch of the aorta and ductus arteriosus (*d*).

The primitive aortas terminate in the umbilical or allantoic arteries which supply the placenta.

The aortas subsequently coalesce into a single tube in the dorsal region.

The development of the venous system is shortly as follows. At an early period of development two short transverse veins, the ducts of Cuvier, enter the auricle.

Each duct of Cuvier is formed by the junction of a primitive jugular vein and a cardinal vein.

The primitive jugular veins return the blood from the head and upper extremities, the cardinal vein from the Wolffian body and trunk. At this stage the inferior vena cava has no existence. Several small branches open into the duct of Cuvier, and ultimately form the internal

jugular and subclavian veins. The direction of the ducts of Cuvier is gradually altered in conformity with the change of position of the heart as it passes backwards from beneath the bend, and, as development proceeds, a short transverse branch connects the primitive jugulars. This branch in due course becomes the left innominate vein. The right cardinal vein becomes the azygos major, the left the superior intercostal vein, and the right duct of Cuvier the superior vena cava. In the normal process of development the left duct of Cuvier is obliterated, with the exception of the lower part which persists as the sinus of the coronary vein, but occasionally persists as a left superior vena cava, the transverse vein then dwindling.

There is considerable variation in the arrangement of the venous system in the neighbourhood of the heart, and this depends entirely upon the extent and mode of obliteration of the various vessels to which reference has just been made.

The pulmonary veins are formed in the lungs —as are also the arteries—and subsequently establish a communication with the left auricle

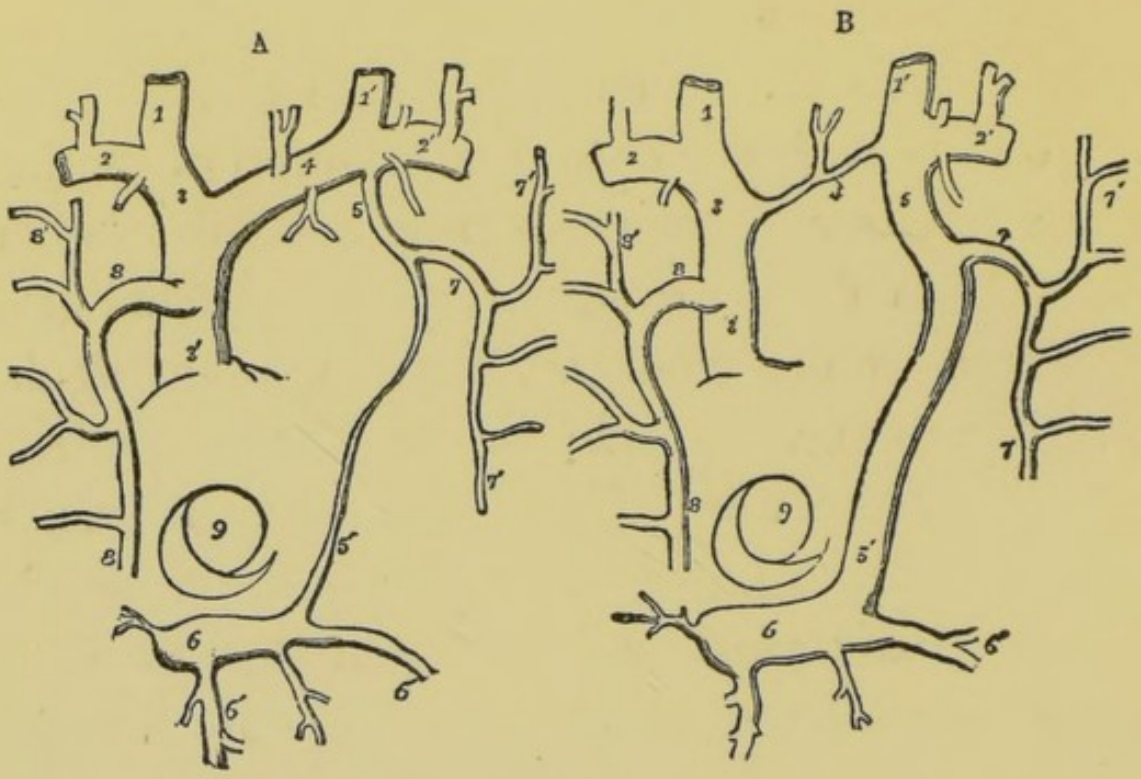


FIG. 3.—A and B.—DIAGRAMMATIC OUTLINES OF THE VESTIGE OF THE LEFT SUPERIOR CAVA AND OF A CASE OF ITS PERSISTENCE (sketched after Marshall). $\frac{1}{3}$ (after Quain.)

A, brachio-cephalic veins with the superior intercostal, azygos, and principal cardiac veins.

B, the same in a case of persistence of the left superior cava, showing its communication with the sinus of the coronary vein. The views are supposed to be from before, the parts of the heart being removed or seen through.

1, 1′, the internal jugular veins; 2, 2′, subclavian veins; 3, right innominate; 3′, right or regular superior cava; 4, in A, the left innominate; in B, the transverse or communicating vein between the right and left superior venæ cavæ; 5 in A, the opening of the superior intercostal vein into the innominate; 5′, vestige of the left superior cava or duct of Cuvier; 5′, 5′, in B, the left vena cava superior abnormally persistent, along with a contracted condition of 4, the communicating vein; 6, the sinus of the coronary vein; 6′, branches of the coronary veins; 7, the superior intercostal trunk of the left side, or left cardinal vein; 8, the principal azygos or right cardinal vein; 7′, 8′, some of the upper intercostal veins; 9, the opening of the inferior vena cava, with the Eustachian valve.

and pulmonary arteries respectively. The valves are formed from endocardial projections, which in the semi-lunar are early noted to be tri-partite. In the auriculo-ventricular the projection is first annular and subsequently divides into the normal number of segments.

In a few words the pecularities of the circulation of the fœtus are as follows :

The venous blood is carried to the placenta for aëration by the umbilical arteries, and is returned by the omphalo-mesenteric or vitelline veins. By these veins the blood is carried partly to the liver and partly through the ductus venosus to the ascending cava, and thus directly to the heart. The blood from the head and upper extremities is returned to the right auricle by the descending vena cava. It passes into the right ventricle, and thence by the pulmonary artery and ductus arteriosus into the descending aorta, and thence to the placenta. The blood returning to the heart by the ascending vena cava is, of course, arterial, and, in all probability, the greater portion of this blood on entering the right auricle is directed by means of the Eustachian valve towards the foramen ovale,

through which it passes, into the left auricle, and thence into the left ventricle and aorta. The arterial blood is then transmitted through the aorta to the head and upper extremities, whose development is required to be in advance of that of the rest of the body, which thus receives their supply in advance of that to the trunk.

In early fœtal life the heart is considerably larger in proportion to the body than later on, but even at birth it is still proportionally large. The ventricular walls are equal in thickness, the patent ductus arteriosus equalising the work they they have to perform in utero.

The Pulmonary Artery and its Orifice.—Of all congenital affections of the heart those which attack the pulmonary valves, the artery, or its orifice, are the most common. Generally in cases of [this description the number of the pulmonary valves is defective. Associated with these conditions will be found either a perforate septum ventriculorum in varying degrees, a perforate foramen ovale or inter-auricular septum, the valve being perfect, or both auricular and ventricular septa may be more or less patent.

When the narrowing is not very marked the ventricular septum may be intact, and the foramen ovale closed.

Stenosis of the valvular orifice is occasioned by adhesion of the semi-lunar cusps which, when they are three in number, project funnel-like into the lumen of the vessel. The apex of the funnel is perforated by an orifice, which is either triangular in outline, or more or less rounded or puckered; the edge usually being thickened, and small vegetations may be found on its pulmonary aspect. Should there be but two valves the orifice will be slit-like.

The resulting channel varies from one of moderate stenosis to a lumen, not more than a No. 1 catheter in circumference, or there may be complete obliteration. The whole valve may be thickened, opaque, and the seat of calcareous deposits. When the diaphragm thus formed originally consisted of three valvular cusps, in the majority of instances, frænа more or less marked, corresponding to the lines of adhesion, will be noted on the pulmonary aspect with corresponding depressions on the ventricular.

Again, in others the bases, as well as the free

edges of the valves, are contracted, the resulting outlet then assuming a barrel shape. Such changes in the valves in no way differ from those found in after life, and the probabilities are largely in favour of them being due to intra-uterine endocarditis. Moreover, such damaged valves are rendered liable to future endocarditis, and consequent deposit of vegetations, entailing further narrowing, or complete obliteration of the already stenosed orifice.

In some cases disease of the fibrous zone at the outlet of the ventricle is accountable for the contraction in this situation, and in others a thickened endocardium, with or without vegetations, and surrounding muscular hypertrophy are the cause.

All varieties of narrowing are met with up to atresia with consequent obliteration of the conus arteriosus.

The trunk of the pulmonary artery presents many variations from the normal. Its coats may be thin and vein-like, or thickened and narrowed, or dilated, or partially obliterated, or there may be atresia, the artery being represented by a fibrous cord in whole or in part,

and finally no vestige of it may be found. In some instances the pulmonary artery is merely undersized in all its parts, but otherwise perfectly formed. Absence of the ductus arteriosus during fœtal life is possibly responsible for this condition, the artery being starved. When the septum ventriculorum is imperfect, it will in addition be found pushed more or less to the left, so that the aorta starts from the right or left ventricle in varying degrees, according to the septal inclination. When the ventricular septum is intact the trunk of the pulmonary artery is generally dilated.

In some cases only a small portion of the aorta takes origin from the right ventricle, or it may be equally divided between the two ventricular cavities, or arise from the right ventricle only. The more the aorta communicates with the right ventricle the larger will be its ascending and transverse portions, and when it takes origin from the right ventricle it is always much dilated.

The presence or absence of a perforated septum cordis, for the time being omitting the mention of other causes giving rise to it, depends upon the date of the intra-uterine endo-

carditis affecting the pulmonary structures. When it starts in early intra-uterine life, before the septum is completed, this is found patent, and pushed over in greater or less degree to the left with consequent displacement of the aorta to the right. When the disease occurs subsequently to the shutting off of the ventricles, that is, later on in intra-uterine life, the foramen ovale will be found unobliterated, and failing that, a perforate auricular septum. In other instances *both* septa remain patent.

It is surprising how minute a channel in the pulmonary artery suffices for life, and cases are on record when the pulmonary circulation, unassisted by a patent ductus arteriosus, has been carried on, even to adult life, with a lumen of only a few lines in diameter.

In other instances the supply of blood to the lungs has been supplemented by a perforate ductus arteriosus of varying capacity. Should obliteration of the pulmonary artery occur whilst the septum is in process of formation, the right ventricle becomes considerably hypertrophied, together with the right auricle, and corresponding diminution in size will take place in the left auricle and ventricle cavities. If, on

the contrary, closure takes place subsequently to the completion of the ventricular septum, the right ventricle will atrophy together with the tricuspid orifice and valve. The foramen ovale will then be pervious, and the left auricle and ventricle cavities hypertrophied. In other words, those portions of the heart which are thrown out of the circulation will atrophy, and those cavities which are in active work will hypertrophy.

In stenosis with perforate foramina the right heart will be hypertrophied, and the more the aorta communicates with the right ventricle the greater in proportion will be the hypertrophy of this chamber. When the septum ventriculorum is perfect, the right cavities will also be found large. In slight stenosis without patent septa, the ventricle hypertrophies sufficiently to overcome the increased resistance, and the balance of circulation is not disturbed, often for very many years, possibly for life. When the pulmonary artery is completely obliterated, blood to the lungs is carried, in the large majority of instances, by the ductus arteriosus (see fig. 2 on p. 11 and description). Failing this, the lungs

may be supplied by the bronchial arteries which enlarge for the occasion—it will be remembered that the bronchial arteries anastomose with the pulmonary radicles—or special branches may arise from the large vessels of the aortic arch such as the innominate artery. The lungs may be supplied by special vessels from the aortic arch, or there may be a direct communication between the aorta and pulmonary artery. Irregular vascular supplies to the lungs, such as those just enumerated, are due, with the exception of the direct communication, to faulty development or premature obliteration of the left fifth branchial arch.

Associated with the lesions just described, changes may also be found in the tricuspid and mitral valves. These may be thickened and opaque but competent, and with or without vegetations on their auricular aspect. The folds may be united and their orifices more or less stenosed, or the chordæ tendineæ may be too short, tying the cusps down and so rendering them incapable of performing their functions.

The reader must bear in mind that stenosis

of the mitral or tricuspid orifice, particularly the former, is exceedingly rare. Osler gives a picture of tricuspid stenosis occurring in a child of 4 months in Keating's "Cyclopædia of the Diseases of Children." Further, such diseased hearts are occasionally the subjects of other developmental irregularities. The following case illustrates the condition of atresia of the pulmonary orifice.

Arthur R. E——, aged 5½ years, in whose family there was no history of rheumatism, enjoyed excellent health until an attack of scarlet fever at the age of 2 years, from which date cyanosis occurred, accompanied by clubbing of the fingers and toes. There was a history of several severe falls happening to the mother during the last few weeks of pregnancy. In addition to extreme cyanosis, the face and body being dusky, the mucous membranes slate coloured, the conjunctivæ suffused, and the extremities blue, he suffered from time to time from convulsions, during which the cyanosis became increased greatly. His pulse on these occasions was barely perceptible, and in one of the attacks he died.

The heart's dulness commenced at the third left costal cartilage, and was carried well to the right of the sternum, the impulse being in the fifth interspace just internal to the nipple line and somewhat weak. There was a loud blowing systolic bruit, which varied in intensity from time to time, audible at the scapular angles and more or less all over the chest. It was loudest in

the third left interspace, close to the sternum, just appreciable under the right clavicle, and falling short of the left. The second sound was accentuated. Some days before his death the bruit completely disappeared, and there was considerable dyspnœa at this time. Whilst under observation he had an attack of chorea. His liver was slightly enlarged. The urine, which was free from albumen was scanty, the quantity passed averaging ten ounces per diem. He was never dropsical. His eyes, which were examined ophthalmoscopically shortly before his death, showed the retinal veins to be *very* large, the arteries of good size, and both sets of vessels most intensely cork-screwy. There was no edge to the optic disc, and the red reflex apparently started from the physiological pit. The pulse was, for the most part, about 84 to the minute and irregular, but shortly before his decease it increased to 106 beats to the minute, being feeble and compressible. The respirations as a rule were about 28 to the minute, but at the last were noisy, numbered 48, and the dyspnœa was very great. A slight cough was noticed for the first time the night preceding his decease. His temperature was generally subnormal, averaging about 97° F., but he had occasional rises of temperature.[1] Death ocurred when he was $7\frac{1}{2}$ years of age.

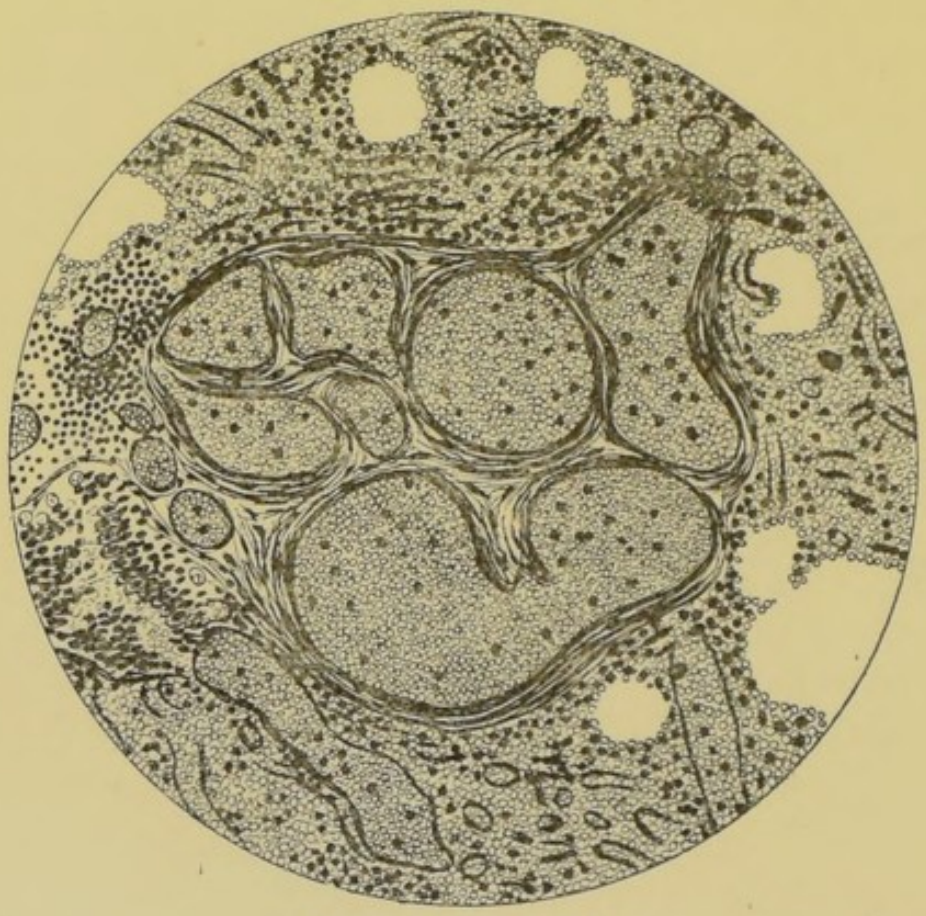
Both ventricles were hypertrophied and dilated, the right being in excess. The right auricle was twice the thickness of the left, but the left was dilated, and the

[1] "Subnormal" temperatures of a degree or so Fahrenheit are so very frequently found in children, that the standard 98.4° F. cannot be considered the absolute normal for them. Although such a temperature (97° F.) might be deemed subnormal for an adult, in the child it must be looked upon as fairly within normal limits.

right auricular appendix four times the size of its fellow, which was decidedly small. There was a patent septum ventriculorum, capable of admitting the index finger. The foramen ovale was closed. The conus arteriosus of the pulmonary artery was completely occluded, the endocardium at this part being united, and surrounding this there was considerable muscular hypertrophy. The pulmonary valves were two in number, small, but well-formed, and of equal size. The artery was capable of admitting a crowquill. The aorta arose equally from both ventricles, and the arch passed over the right bronchus. It gave off, first a left innominate, then a right carotid and sub-clavian respectively. A large vessel arose from the innominate for the supply of the left lung, and midway between its origin and this lung, gave off a vessel the size of a crowquill for the supply of the right lung. Microscopical examination of the organs showed extreme capillary dilatation and thickening with a tendency to fibrillation as also venous engorgement and thickening of the lungs, brain, kidneys, liver and heart. The lungs were much congested but crepitant, though firmer than natural. Some minute solid areas were visible, and in the right lung there was an infarct the size of a walnut. In addition to the extreme capillary dilatation small broncho-pneumonic areas, and blood extravasations with capillary remnants were visible, as also blood-filled alveoli. The smaller bronchioles contained blood and inflammatory products.[1] See figs., pp. 26, 27.

[1] "Microscopical changes in the organs found in a case of cyanosis with congenital malformation of the heart," by the author. *St. Thomas's Hospital Reports*, vol. xviii.

Sections of the skin to a considerable number were examined microscopically, but nothing ab-



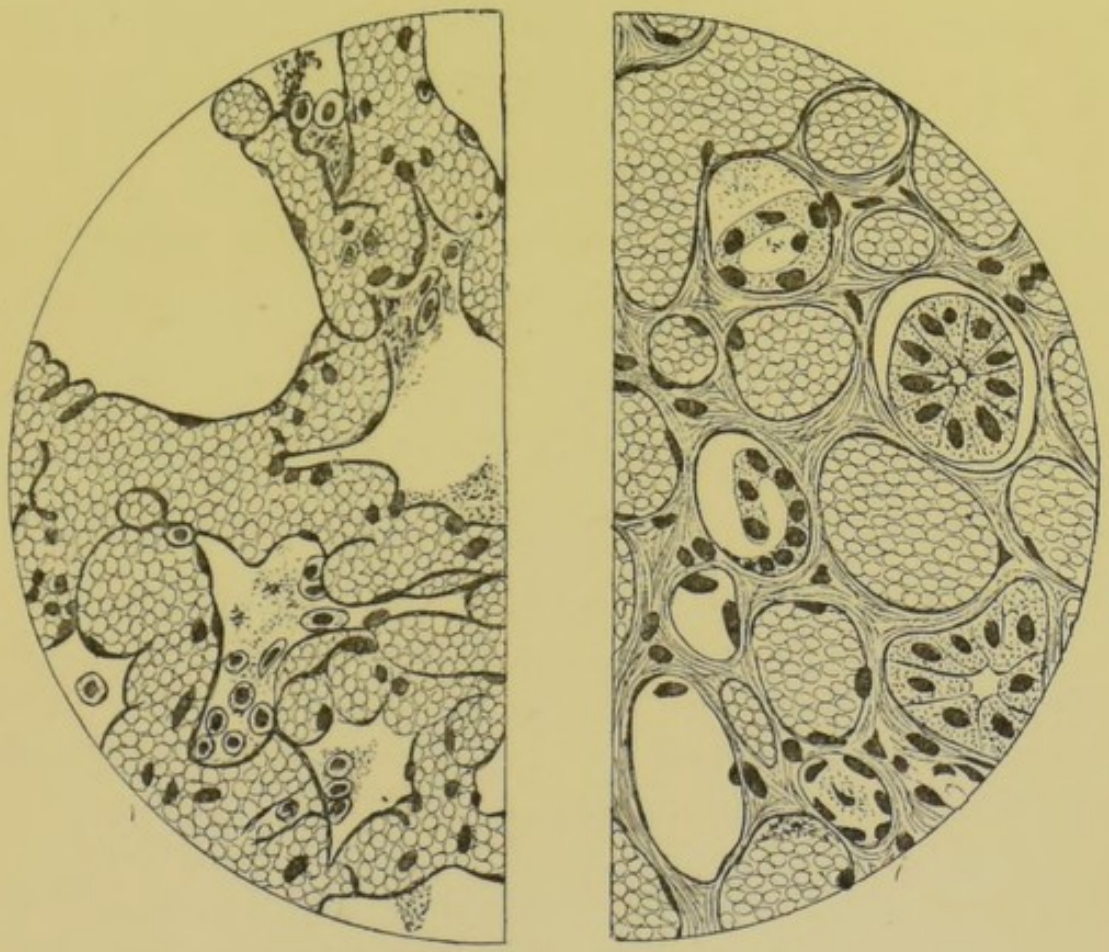
George Carpenter, del.

FIG. 4.— LUNC. Leitz, Obj. 4, Oc. 3.

Fig. 4 shows thickened capillary remnants; blood extravasations; an inflamed bronchiole containing red blood corpuscles, with a broncho-pneumonic patch above it; a peculiar multiloculated chamber of not very obvious origin constructed of fibro-nucleated material, and crammed with red blood discs and a sprinkling of white corpuscles. These chambers accompanied the bronchioles. The vacant spaces are artificial.

normal was detected. *Post-mortem* shrinking of the elastic cuticle might account for failure in the detection of capillary changes.

In this case the atresia occurred prior to the closure of the ventricular septum, and as a re-



George Carpenter, del.

FIG. 5. FIG. 6.

LUNG. Leitz, Obj. 7, Oc. 3. KIDNEY. Leitz, Obj. 7, Oc. 3.

Fig. 5 shows dilated, thickened and tortuous capillaries admitting from three to six or more red corpuscles abreast. In places, an indistinct fibrillation is seen. Under a low power the alveolar cavities are with difficulty picked out; the shapes shown are various.

Fig. 6 taken from the medulla shows a thickened fibro-nuclear stroma. The greater portion of the network formed by the stroma contains red blood corpuscles, the vasa recta being greatly increased in size and thickness.

sult is seen the altered aortic origin with right-sided ventricular hypertrophy. The changed position of the aortic arch was due to the obliteration of the left fourth branchial arch, the right fourth arch taking its place, and undergoing the necessary developmental changes.

A case illustrating a late onset of atresia of the pulmonary orifice, with consequent completion of the ventricular septum, was brought before the Pathological Society by my colleague, Mr. Clement Lucas.

A cyanosed child brought to the Evelina Hospital died on the sixth day. The heart was rounded in outline, the left ventricle very hypertrophied, and dilated, the aorta large, and the ductus arteriosus patent. Atresia of the pulmonary artery was present from union of its valves, but the artery was patent as far as this structure. The right ventricle was atrophied, and so small that the cavity would not admit a pea. The right auricle was hypertrophied and the foramen ovale patent.[1] There was a vertical septum in the right auricle formed by an unusually developed Eustachian valve.

Of the varieties of malformation just described, atresia of the pulmonary artery is a rarer form than stenosis.

[1] *Pathological Society's Transactions*, London, 1875.

One hundred and nineteen cases out of one hundred and eighty-one of various congenital cardiac affections collected by Peacock illustrate defects of the pulmonary artery, orifice, or conus arteriosus.

Septa in the Cardiac Cavities.—Passing from the consideration of the defects of the pulmonary artery, a much rarer form of malformation must be briefly touched upon, viz., a septum in the right ventricle. Septa, with extreme rarity, are found in the other cavities. The right ventricle consists of two portions, a sinus and an infundibulum (conus arteriosus), the latter communicating with the pulmonary artery from which it takes its origin. In the turtle's heart there are three ventricles, a right and left giving origin to the right and left aortic arch, and a central, in communication with the right ventricle only, to the trunk of the pulmonary artery. The septum arises, in man, at the junction of the sinus, and infundibular portions of the right ventricle, shutting off, more or less completely, the pulmonic portion of the ventricle from that of the sinus, as in the turtle. This septum consists of thickened endocardium, and surrounding

cardiac muscle. The orifice, connecting the two ventricles thus formed, is of varying sizes from a finger to a probe. It may be double. Vegetations may form on its margin.

Associated with this condition may exist a defective septum ventriculorum, or the septum may be perfect. In either case the sinus will be hypertrophied, and more so if the septum be patent. The pulmonary artery and infundibulum may be small, according to the amount of stenosis present. It may be of natural calibre, or it may present changes such as have been previously very fully described. Such a septum must be looked upon as a developmental error, which may occur early or late, and constitute the only pathological change, the various septa being completely closed.

A remarkable malformation allied to this condition was published by Dr. Stephen Mackenzie.[1]

A woman, aged 39, all her life had suffered from cyanosis and palpitation, and died of cardiac failure. The heart was very large. There were three ventricles. A *right*, not forming the apex, in front of the right seg-

[1] *Pathological Society's Transactions*, London, 1879-80.

ment of the tricuspid valve opening into the middle ventricle, and giving origin to the aorta. The *middle* ventricle communicated with the tricuspid orifice, the pulmonary artery, the left ventricle by an imperfect septum, and the right ventricle by a perforation in the septum. The *left* ventricle opened into the middle by the perforated septum, and was in communication with the mitral orifice. The pulmonary artery was large, the ductus arteriosus, the size of a goose quill, gave off the right and left pulmonary branches, and then continued to form the thoracic and abdominal aorta. The aorta was half the size of the pulmonary artery, and ended through the ductus arteriosus in the descending aorta of the pulmonary. The distribution of vessels from the arch was very irregular. The foramen ovale was large, and the pericardium adherent.

A case is recorded of a membranous septum in the left auricle by Dr. Church,[1] which is unique.

A woman, aged 38, was admitted into hospital, for orthopnœa, anasarca, and cardiac pain between the shoulders. She had suffered from hæmoptysis several times but there was no other history. The right and left ventricles were of normal proportions. The left auricle was divided by a membrane perforated by an elliptical opening. This membrane stretched completely across the auricular cavity, separating, on the one hand, that portion in connection with the auricular septum and into which the pulmonary veins entered,

[1] *Pathological Society's Transactions*, London, 1868.

from the side in connection with the auricular appendix and mitral orifice. The auricular wall was not hypertrophied. It was grooved on the outside corresponding to the internal septum.

An unusually developed Eustachian valve may form a septum in the right auricle as in Mr. Lucas's case.

Ductus Arteriosus supplying or giving origin to the descending Aorta.—Owing to developmental errors occurring in the left posterior aortic root, midway between the fourth and fifth branchial arches, that is, just beyond the origin of the left subclavian artery, the aorta at this point becomes extremely narrowed, so that its calibre is much less than it should be, and all stages of stenosis may be met with up to complete obliteration. This form of malformation is of rare occurrence, and when it exists the descending aorta is in part supplied by a patent ductus arteriosus.

In some instances the ascending and transverse aortas are entirely separate from the descending aorta.

Under these circumstances the right side of the heart will be found hypertrophied, and the pulmonary artery considerably larger than the

aorta, which will be small. Cases with this defect die early, but early decease is not the invariable rule, and existence may be prolonged to adult life; when cardiac symptoms due to muscular failure of the organ, from the increased demands made upon it, are likely to arise, and with the usual termination. On the contrary, prolonged and robust life may be enjoyed, and death may be due to some other complaint.

This malformation is a rare one under any circumstances, and it is, in the majority of instances, associated with other cardiac abnormalities incompatible with prolonged life, which have been, and will be, described under their respective headings further on in the text. As a very rare malformation it may be associated with transposition of the aorta and pulmonary artery.

Coarctation of the Aorta.—This malformation is closely allied to the preceding, in that the same portion of the posterior aortic arch is at fault, but it includes the extremity of the fifth left branchial arch which forms the ductus arteriosus. Here, as in the former deformity, the transverse and descending aortas may be united by a barely pervious cord, or the separation is

complete. In the absence of the ductus arteriosus the blood supply has to be carried on by other channels, and the usual methods of communication between the severed trunks is as follows :—The upper intercostal arteries from the second part of the subclavian are usually very tortuous, enlarged, and freely anastomose with the first aortic intercostals. The posterior scapular arteries arising from the third part of the subclavian with their branches, or as offshoots from the transverse cervicals of the thyroid axes (first part of subclavian), enlarge, become very tortuous, and freely anastomose close to the spine with the upper intercostal branches which are similarly affected. The same may be said of the subscapular branches. The internal mammary arteries from the firs part of the subclavians enlarge considerably, and anastomose freely with the aortic intercostal vessels by their intercostal branches. The musculo-phrenic branches of the internal mammary arteries, enlarged and tortuous, in like manner anastomose with enlarged phrenic arteries from the abdominal aorta. The superior epigastric of the internal mammary

anastomose with the deep epigastric vessels from the external iliacs, and both sets are enlarged and tortuous. Such cases are not incompatible with long life, but the cardiac hypertrophy such a condition occasions is considerable, and in the event of cardiac symptoms occurring in a patient with enlarged and tortuous arteries, in the scapular, or abdominal regions, a shrewd guess might be made as to the probable condition of affairs.

This is a disease which only comes under notice in adult life. I can find no record of such a case being detected in childhood and no pathological specimen. It appears to be a disease like stenosis of the mitral valve, if that be congenital, which takes time to produce its effects upon the circulation or the heart, and in this particular instance would seem to come under notice as early adult age is reached.

Pulmonary Artery arising from the left Ventricle. — Instances have been given of deviation of the septum ventriculorum to the left side, but this structure may deviate to the right, the pulmonary artery and aorta both taking origin from the left ventricle.

Failing this the pulmonary artery receives its blood supply indirectly from the left ventricle, communicating with this cavity by a perforate septum ventriculorum, and arising from a rudimentary right ventricle. This form of malformation, in comparison with deviated septum to the left, seldom occurs. Malformed hearts such as these usually present other abnormalities which are not at all constant.

Transposition of the Aorta and Pulmonary Artery.—In this condition, one of the most unusual, the aorta arises anteriorly from the right ventricle, the pulmonary artery posteriorly from the left. In the remarks on development, the natural division of the bulbus arteriosus into the aorta and pulmonary artery was described. Owing to the want of rotation of the septum on its long axis, it still maintains the lateral position of its broad surfaces as at the commencement, and with the above result. This transposition may be the only misplacement, as is usually the case, but the auricles and ventricles together with the veins may be in like manner transposed with these vessels. Such a condition may be found in transposition

of the viscera. The foramen ovale and ductus arteriosus are often patent together. In the large majority of cases the foramen ovale has been found open, but the ductus arteriosus is occasionally impervious. The septum of the ventricles may be more or less patent, but this is not a frequent concomitant. Associated with this malformation there may be other abnormalities, an enumeration of which need not be made. Of twenty-five cases collected by Rauchfuss twenty did not survive the first year. Transposition may be incomplete ; thus the aorta may arise from the infundibulum and the pulmonary artery, placed behind the aorta, from the sinus of the right ventricle. Such a condition is extremely uncommon. One case the subject of this malformation with a perforate ventricular septum, died at the age of eight years.

Failure of the Septum of the Bulbus Arteriosus to develop—Heart with four Cavities. —As an extremely rare malformation associated with four cavities the septum of the bulbus arteriosus fails to develop. The common trunk or aorta then arising from both

ventricles supplies the right and left pulmonary arteries in addition to those usually arising from the aortic arch. In the cases recorded other malformations were noted in addition.

Direct Communication between the Aorta and Pulmonary Artery.—As a further instance of septal irregularity may be noticed a direct communication between the aorta and pulmonary artery.

Perforate Septum Ventriculorum. — This condition presents all degrees and all varieties. It may be represented by not more than a pin puncture to a condition in which the merest rudiment of the septum is all that can be detected as a means of separation between the two ventricles. Communication between the left ventricle and right auricle is of very rare occurrence, but all four cavities may be in communication from slight failure of the auricular septum in immediate contiguity with the upper part of the septum. The aperture may be triangular, rounded, oval, or fenestrated, and it may be protected by a bulging of the corresponding segment of the tricuspid valve. Further, several perforations may be

found in the septum cordis, and rarely the left ventricle communicates with the infundibular portion of the right. Occasionally fibrinous deposits are found at the margins of these perforations, and in one, that of a child under my care, suffering from ulcerative endocarditis of the left side, vegetations were found in the right ventricle opposite the perforation. Many of the causes of this condition have already been enumerated, but it remains further to say that there are other states, such as disease affecting the mitral, the aortic, and tricuspid orifices, which will bring about such patency. Septal defects are extremely common with pulmonary bicuspid valves, but unusual with a similar aortic condition. It must be remembered as a rarity that the septum is occasionally perforated from disease in after life. Finally large apertures may be discovered in the *post-mortem* room in persons dying of some totally different disease in whom the presence of such was never suspected, and for which there is no discoverable reason. Rokitansky,[1] in 1875,

[1] *Die Defecte der Scheidewand des Herzens, Wein.*

pointed out that it is not the undefended space, as had been previously supposed, which is affected in these perforations. The portion in front of the undefended space lies between the arterial orifices, that behind between the auriculo-ventricular orifices. The usual situation for perforation is the anterior portion in immediate contiguity with the undefended space, next frequently the anterior portion in front of this, and least of all the part posterior to the undefended space. This latter is as a rule associated with defective auricular septum as also the membranous portion. Congenital deficiency of the undefended space occasionally occurs.

Patent Foramen Ovale.—In conducting autopsies on children, it is not at all a rare occurrence to meet with small valvular orifices which during life have led to no signs or symptoms, and which probably did not transmit any blood between the auricular cavities; anyhow, they are of no clinical moment. In many cases a wider aperture remains for a year or so of the first year of infancy. In addition to small valvular orifices from want of adhesion of the

valve to the cornua, there are other cases in which the valvular opening is large, and transmitted blood during life between the cavities. In other cases the valvular fold is completely absent, or not sufficiently large to cover the orifice. Many cases are on record of persons dying at all ages, and of various diseases, in whom, as the result of a congenital defect, there was no trace of the normal membrane covering the foramen ovale, this being patent. Such, when unassociated with other cardiac abnormalities, may never be attended by symptoms.

Again in other instances the valve may be quite adherent with a single opening in the septum, or the opening may be cribriform and situated either in the valve or the septum. Finally the auricular septum may be completely wanting, or give rise to the communication of the four cavities such as has been described under the heading *Perforate Septum Ventriculorum.* Many of the causes of this abnormality have already been enumerated, but there are other conditions capable of giving rise to the defect. Such as have been described under the heading *Perforate Septum*

Ventriculorum are they, and these need not be repeated. Not only may there be no symptoms during life, but after death no cause can be ascertained in some for the patency. Traumatism may lead to rupture of the valve in after life, and with it the symptoms of heart disease. The margin of the orifice may be thickened, and vegetations appear on it as on the margin of a patent septum ventriculorum.

Patent Ductus Arteriosus. — The ductus arteriosus is rarely patent beyond the tenth day, and by three weeks in almost all cases, it has completely closed. This obliteration primarily depending on contraction, is quickly succeeded by thickening of the coats. As in the case of the foramina just mentioned much has already been noted concerning the patency of this channel. It may be found in hearts that are otherwise well-formed, and its patent condition probably arose from the existence of some obstruction at the time of birth, such as atelectasis pulmonum, which passing away left this as a relic of the former mischief. Premature delivery has been likewise assigned as a cause. Patients with patent ductus arteriosus,

without other abnormalities, may reach adult life, and without symptoms — there may be trifling cyanosis. Closure of this channel usually commences at the aortic extremity. Its size, as has been already gathered, is very variable. The position of the vessel may be regular, or there may be two ducts. Again, it may show irregularity of origin. It may be absent. It may remain pervious, in addition to the causes already enumerated, from disease affecting the mitral, the tricuspid, or aortic orifices. Atheroma is common in this vessel in those cases which live some years.

Premature Closure of the Fœtal Channels.— This has been briefly touched upon in the account given of diseases of the pulmonary artery, but it must be further noted that any of the fœtal orifices may suffer in like manner.

Foramen Ovale. — In cases of premature closure of this structure, the right heart is very large, together with the pulmonary artery and ductus arteriosus. The left heart is small, and atrophied, together with the aorta. The septum ventriculorum may be patent, and the aorta then arises from both ventricles. The

condition is a rare one. The closure is never absolute, a valvular slit being left.

Ductus Arteriosus.—Premature closure of this vessel has likewise been discussed. Such closure may take place prior to, or after the completion of, the interventricular septum. When it occurs early it is a cause of atrophy of the pulmonary artery. This vessel may likewise be found atrophied when the septum is unclosed and the foramen ovale open.

The Aorta.—There may be stenosis or atresia of the valves or of the conus arteriosus. In twenty-four cases recorded by Rauchfuss[1] the ventricular septum was perfect—the majority were due to endocarditis. The ventricular septum may be defective. Atresia is more common than on the right side. Dilg[2] has collected fifteen cases of stenosis of the conus arteriosus—a rare malformation—seven were inflammatory and eight developmental. Of thirty-three cases (Osler), with or without septal defect, but one lived more than a month. Of Dilg's cases the majority were in adults.

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[1] *Gerhardt's Handbuch der Kinderkrankheiten*, Bd. iv., Abth. I.
[2] *Virchow's Archiv.*, Bd. xci.

Mitral Valve and Tricuspid.—The mitral valve may be wanting, the auriculo-ventricular orifice being quite obliterated. The same remarks apply to the tricuspid valve and its orifice. Under these circumstances, the altered condition of the circulation has to be allowed for by corresponding patency of the other fœtal channels.

Diseases of the Valves.—Some knowledge has already been gained of these diseases in the account given of malformation of the pulmonary artery. The remarks made concerning the valves guarding that vessel equally apply to those situated at the aorta, with the reservation that fœtal endocarditis and its consequences are more frequently seen on the right side. Rauchfuss has, however, seen both sides affected almost equally. According to his observations fœtal endocarditis is only more common on the right side when in association with developmental errors, and apart from such, the left side is not less frequently involved.

Regarding the semi-lunar valves, these may be but a single curtain stretched across the orifice, or two only of the cusps may be united.

The valves may be completely wanting, or merely represented by a fleshy ring. When the valves consist of but two segments they may be equal in size and appearance, but more often one is larger than the other, with an ill-developed frænum on its upper surface. Sometimes a rudimentary valve is detected, situated in one of the angles. As has been remarked before, so it must be again insisted upon, that such malformed valves are exceedingly liable to disease in after life. Sometimes valves show redundancy, doubtless from imperfect developmental changes. The conditions met with are as follows:—

(*a*) Four curtains, one distinct and three united.

(*b*) Three or four cusps of about equal size and one or two smaller cusps, more or less adherent to their fellows.

(*c*) Four valves, three of nearly equal size and a smaller one, blended more or less imperfectly with one or other neighbour.

(*d*) Four valves about equal in size; two segments imperfectly separated.

(*e*) Three cusps with a smaller fourth, with a perfect corpus arantii.

(*f*) Cribriform perforations may be found in these valves.

Diminution of the number of semi-lunar valves is usually found in the aorta. Regarding bicuspid sigmoids, of cases collected by Dilg[1] from literature there were 64 in the pulmonary artery and only 23 in the aorta. Osler has found this most frequent in the aorta—19 as against 2 ; and Viti[2] in the proportion of 7 to 3.

When, owing to endocarditis, one of the sigmoid valve partitions becomes broken down, such can be distinguished from developmental errors by the fact that the bicuspid valves bear no sort of proportion to one another in point of size, as in the latter case.

Redundancy is met with more frequently in the pulmonary artery ; thus in Dilg's collection of cases there were 24 of 4 pulmonary valves, and 2 of 4 aortic ; 3 cases of 5 valves showed 1 at the aorta, and 2 at the pulmonary artery.

Redundant valves are not so liable to future disease and they are as a rule quite competent to perform their duties.

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[1] *Loc cit.*

[2] *Lo Sperimentale*, 1886.

Auriculo-Ventricular Valves.—The chief malformations to which these structures are liable have already been touched upon. As in the semi-lunar valves, the segments may be united, and the orifice narrowed. The latter condition is an important one, and is entitled to more than passing observation, for it is a nice point, whether or not in some instances of mitral stenosis this condition is due to intra-uterine endocarditis. Although such malformations are identical with the changes observed in the valve in acquired disease, it must be pointed out that such a condition of the valve may be found when other malformations are present. Further, that complete obliteration of one or other orifice is not a very unusual malformation, and that the tricuspid and mitral valves have been found in the fœtus with all the signs of endocarditis, displaying vegetations on their auricular surfaces. There appears then to be ample *prima facie* evidence for, in some instances, congenital mitral stenosis, but it still remains true that such cases are not met with until after six or eight years of age, and even then very uncommonly; *vide* remarks under the

headings — "The Pulmonary Artery and its Orifice," and "Coarctation of the Aorta." The youngest case of which I have a clinical record is that of a child 5 years of age. The valves occasionally show more or less irregularity about their cusps, and in a case published by Dr. Greenfield the mitral valve consisted of four segments, due, apparently, to developmental adhesion of the cusps at one spot, the auriculo-ventricular opening being double.[1]

Hearts of Three Cavities.—Briefly, such hearts may be formed as follows :—

(*a*) Two auricles with a perforate foramen ovale, a ventricle giving rise to an aorta and pulmonary artery in their usual situations.

(*b*) As the above, but the pulmonary artery and aorta may be situated to the left of the single ventricle.

(*c*) As the above, with a transposed pulmonary artery and aorta.

The auriculo-ventricular openings may be single or double, and either valve may be obliterated. The pulmonary artery may be rudimentary, or impervious with a closed or

[1] *Pathological Society's Transactions*, London, 1876.

open ductus arteriosus—its valves may be imperfect. The foramen ovale may be closed. The aorta may be a single tube giving off the pulmonary arteries. It may pass over the right bronchus. In the single ventricle there may be a trace of a septum. The abnormalities associated with this condition are so numerous and varied that it would be useless and unprofitable to enumerate them all, and it need only be remembered that any arterial or venous abnormality may be present. The condition is a rare one, but it is more frequently present than a biloculate heart.

Hearts of Two Cavities.—This is a rare condition. Such malformed hearts present an auricle and a ventricle with a single vessel arising from the latter, supplying both the systemic and pulmonary circulations. Hearts midway in point of development between those just described and the present condition may be found. Such malformations are of no practical importance, and would hardly pay the enumeration.

Malformations of the Arteries.—On this subject much has been already said in the text.

As a very rare malformation, both fourth branchial arches may develop, and the aorta will be found as a double tube surrounding the trachea. Such a condition is rather of developmental interest than clinical moment. The coronary arteries may originate high up on the aortic arch, or be given off by some one of the large vessels. The origin of the great vessels from the arch admits of many irregularities. A knowledge of the development from the branchial arches will serve for the comprehension of any abnormalities met with beyond those enumerated.

Malformation of the Veins.—The formation of the veins of the heart has been described in the portion devoted to development. The most frequent abnormality is the existence of two ascending or two descending venæ cavæ. In complete transposition of the heart the pulmonary veins enter the right auricle, the venæ cavæ the left. The pulmonary veins may be deficient. Both sets of veins may enter either auricular cavity, or the veins may enter the ventricles directly, or discharge into the left innominate vein. In a case reported by

Mr. Shattock[1] of a full time fœtus, the pulmonary veins were confluent at the back of the auricle in the space where this is uncovered by the pericardium, and they had no direct communication with the auricular cavity. Blood probably reached the heart by the bronchial posterior mediastinal veins through the normal communication existing. The right bronchial vein opens into the azygos vein near its termination, the left into the superior intercostal vein.

Malposition of the Heart.—The heart may be situated wholly or in part external to the thorax. It may occur with or without pectoral fissure, and is usually associated with protruded abdominal viscera. It may be extruded through the diaphragm, and in some instances forms an abdominal tumour. In one recorded case, that of a man who had been a soldier, the heart was found in the situation of the right kidney. Again, it may be in front of the neck, but this condition has been found in fœtuses only.

[1] *Pathological Society's Transactions*, London, 1883-4.

Transposition of Heart and Viscera.—The heart itself may be transposed as regards its position only. It may be transposed both as regards its position and cavities, together with their respective vascular supplies. This malformation may be associated with transposition of the viscera, and in such cases not only are the various organs transposed from left to right and *vice versâ*, but the several anatomical peculiarities meet with a like transposition; for instance, the cardiac extremity of the stomach points to the right, the pyloric to the left, and so on with regard to other organs. Such irregularities of development must be remembered, or tumours may be diagnosed when they do not exist. It should not be forgotten that transposed hearts not infrequently present other malformations incompatible with prolonged life.

Hypoplasia of the Heart and Aorta.—In 1856, Virchow called attention to the fact that the heart and aorta were small in connection with chlorosis. Subsequently Beneke[1] brought

[1] "Die Anatomischen Grundlagen der Constitutionsanomalien des Menschen," Marburg, 1878.

forward the following from careful measurements conducted by himself. Up to the age of puberty the collective lumina of the arteries are relatively narrow in comparison with the length of the body. At puberty the heart acquires more power, and the vessels enlarge. Beneke looks upon congenital smallness of the organ as the cause of retardation or disturbance of development at this period, and of general feebleness. A rapidly growing body and general arterial enlargement, with a small and badly developing heart, may give rise to palpitation from heart strain in young adults. Such a condition is usually associated with nervousness and anæmia.

Absence of the Pericardium.—This is frequently found in combination with ectocardia, but may be present in association with a normal heart. The condition may be general or partial.

Ætiology. — Congenital affections of the heart owe their origin to different causes. Certain valvular alterations are occasioned by intra-uterine endocarditis; other valvular changes, such as certain cases of diminution and redundancy, are occasioned by develop-

mental errors in the shape of arrested development. Virchow[1] espouses the view of fœtal endocarditis for a certain number of cases, but the anomaly of a bicuspid sigmoid, without a trace of endocarditis, clearly shows that in a certain proportion the error is developmental. The same remarks apply to the tricuspid and mitral valves. From Schipmann's cases it would appear that intra-uterine endocarditis is rare before the fourth month of fœtal life. Practically there is but one form of intra-uterine cardiac inflammation, and that is allied to the sclerotic variety met with in the adult. Vegetations, however, do occur. Stenosis of the conus arteriosus is due to developmental disturbance. Overlying endocardial thickening is probably occasioned by subsequent localised endocarditis. Patent foramina are brought about by obstructions at the arterial orifices, or at the auriculo-ventricular apertures ; thus an obstructed pulmonary artery induces a patent septum ventriculorum. Meckel, however, thought otherwise, and considered the primary defect to be in the ventricular septum,

[1] Virchow's *Archiv.*, Bd. ciii., S. 103.

the pulmonary artery failing to develop in consequence. Further, any portion of the heart thrown out of the circulation dwindles in consequence. Deficient or absent septa, and supernumerary septa are also occasioned by developmental irregularities.

Valvular affections of the right heart are common in intra-uterine life; those of the left cavities unfrequent. This liability to disease on the part of the right cavities has been supposed to be due to the increased calls made upon this side of the heart whilst in utero, but another solution is that valvular developmental errors are common on the right side in comparison with the left. In a certain proportion of cases a history of maternal injury or emotional disturbance whilst carrying can be obtained, and some of the infants are obviously syphilitic, or there is a syphilitic history. The influence of such on the fœtus yet remains to be proved.

Symptoms.—The symptoms of congenital heart disease are various, and foremost amongst them is cyanosis, but this is entirely wanting in some cases. Cyanosis varies from

slight duskiness of the features to the most intense blueness of the whole body, so that the mucous membranes are almost black in colour, the conjunctivæ of a dusky hue with enlarged and tortuous vessels coursing over them, and capillary stigmata are seen on the cheeks. Some cases present no alteration in colour, but on exertion or crying the lips may be noticed to become somewhat livid. Some, again, are never cyanosed, and others show unusual pallor. Cyanosis, if present, is always increased by any emotional disturbance, or bodily efforts on the part of the patient. Cyanosis, too, is of variable onset, sometimes noticed at birth, at others not for several months or years, and often then succeeding some illness, such as scarlet fever, measles, typhoid fever, bronchitis, and the like. More rarely it may be absent for years. When once present, it nearly always persists, but even then varies from time to time in its intensity. Cyanosis is met with in diseases of the heart, lungs, and upper air passages, the respiratory centre, and its muscular connections, or any condition which limits the quantity of blood

despatched to the lungs for aëration, or interferes with the necessary ingress of air to these organs, but it is rarely ever so intense as that met with in cardiac malformations. Occasionally, however, pulmonary disease, such as chronic emphysema, causes as extreme cyanosis as that met with in the worst forms of cardiac malformation.

The theory as to its causation has been debatable ground for some time, one side asserting that it was due to admixture of venous and arterial blood ; another, supported by Morgagni, that it was owing to venous congestion. That it is not due to admixture of venous and arterial blood through patent foramina there can be no shadow of a doubt, as there are now numerous instances on record where such admixture must have been of the freest possible character, and yet there had not been a trace of cyanosis. Dr. Lees'[1] view is that cyanosis depends on deficient aëration of blood.

Deficient blood aëration may be caused by unsuitable vascular connections with the lungs,

[1] *Pathological Society's Transactions*, Lond., 1880.

and as an illustration take a case of transposition of the aorta and pulmonary artery, with either a patent septum ventriculorum or a patent foramen ovale. Make a mental picture of this condition, and the fact will at once be revealed that only a small portion of the blood can become aërated. No congestion theory is necessary to account for the blueness. Or again, it may be owing to an insufficient and dammed-back blood stream to the lungs, and as an example take a case of stenosis of the pulmonary artery or a supernumerary septum in the right ventricle.

A large percentage of cases are cyanosed in association with developmental or other errors in connection with this vessel. After a very variable period, a sufficient quantity of blood with difficulty finds its way to the air. The lungs becoming gorged with blood further add to the obstruction, and limit the blood-aërating area.

In speaking of this condition it has been remarked that cyanosis does not always date from birth, and this has been noted in many and very various malformations of the heart.

Clearly, then, there was a time in the life-history of the child when its damaged heart was fully equal to keeping the blood sufficiently arterialised and its lungs capable of bringing about the necessary oxygenation of the blood carried to them. There comes a period, however, when more being required of it, the vascular connections are insufficient for the purpose. On the other hand, it is equally possible that the heart remains *in statu quo*, and that the additional mischief which produces the cyanosis is some added hampering of the pulmonary capillaries (bronchitic, &c.). With deficient arterial connections follow venous congestions of the lungs, the *vis a tergo* being utterly at fault. Then again, doubtless, as in the case of A. R. E., if life be sufficiently prolonged, various pathological changes arise in the viscera such as have been described.

On perusing the records of cases of congenital heart affections, where the state of the lungs is reported, it is no uncommon occurrence to find that the lungs were firmer than natural, or collapsed, or in a state of splen-

isation such as that met with in cardiac cases. In some, however, the lungs have appeared healthy, apparently. What the pathological changes met with in some such lungs may have been it is impossible to say, but whatever the condition, it must have further obstructed the blood flow through these organs and greatly diminished the blood-aërating area, and it is reasonable to suppose from the case of A. R. E., that had they been examined microscopically, the pulmonary capillaries would have been found similarly affected.

The lung capillaries in infants doubtless allow of much greater stretching and adaptability to the venous pressure than later on in years, and the increased adaptability gives rise to increased congestion and stagnation. Not only may the lung capillaries be affected, but venous congestions arise in other parts with dilatation of the capillaries. I think it may be safely inferred from A. R. E.'s case, that like capillary and venous changes would be found in the organs of others where the disease is of sufficiently long standing.

When the child succumbs a few days after birth, the cyanosis must be in the main due

to deficient aëration of blood, for any marked capillary change can scarcely have had time to arise.

In the lungs these various pathological changes diminish the blood-aërating area and further hamper the heart. In the systemic circulation a widely dilated venous system produces a sluggish stream which allows a complete robbery of the impoverished red corpuscles of the small supply of oxygen they can obtain in their passage through the lungs. The latter condition, however, when present, is secondary in importance to the former.

A stenosed pulmonary artery without septal or other imperfections of the heart may be readily overcome by suitable hypertrophy of the right ventricle for months or many years. Little by little, however, the natural balance is upset, either by further resistance in front, or by imperfect power behind, and the case which showed defective blood aëration under emotional excitement only, or perchance not at all, may pass on by slow stages to the development of well-marked and unmistakable cyanosis. With considerable tubercular mischief in the lungs, in association with a greatly narrowed

pulmonary artery, cyanosis may be absent, the ventricular hypertrophy being competent to overcome the resistance in front, sufficient lung tissue being left to carry on efficient oxygenation. It was so in the case of a small child under my care who was at no time cyanosed.

In all cases where there is no short circuiting of the pulmonary circulation, as in the first illustration, for instance, it is probable that cyanosis is an indication of defective lung-functions. Insufficient aëration of blood from defective lung-functions is the chief, and sometimes the only, factor, but congestion of the systemic veins, when present, doubtless plays a subordinate part in its production.

Associated with cyanosis is often found clubbing of the fingers and toes, a condition in which the terminal phalanges become enlarged and bulbous. In this, however, there is nothing peculiar to congenital affections of the heart, for the most extreme clubbing is often noticed in chronic phthisis and chronic empyema.[1] Any

[1] For a sketch of this condition *vide* "Pleurisy in Childhood," by the author, "International Clinics," vol. iv., second series, p. 167.

pressure on the veins can produce it, such as that of a subclavian aneurysm pressing on the axillary vein. Bulbous fingers and toes are merely congestive phenomena. In a case of this description, examined by Dr. Norman Moore,[1] the clubbed fingers could be reduced by cutting into them. Microscopical examination showed this condition to be due to engorgement, and some thickening of the walls of the blood vessels. There was not any general increase of connective tissue in the finger end.

Dr. Lees has pointed out that clubbing is due to congestion of the systemic veins in cases where the foramen ovale is closed, but that when that passage is open the congestion is so much relieved that clubbing does not result.

Although this is frequently the case, the foramen ovale is occasionally found open in association with marked clubbing of the extremities.

Other phenomena which may be present, and as a rule, are so, will be extreme irritability, dyspnœa, paroxysmal or constant cardiac pain,

[1] *Pathological Society's Transactions*, London, 1885-6.

palpitation, syncopal attacks, headache, convulsions, and coma.

A small boy was admitted under my care at the Evelina Hospital, quite comatose. He was somewhat cyanosed. No cardiac bruit was detected. An ophthalmoscopical examination revealed fundal changes similar to that noted in A. R. E.'s case, and suggesting to my mind the nature of his complaint. The next day he was sitting up in his cot playing with his toys.

Children circumstanced such as these are liable to gastro-intestinal troubles, such as diarrhœa and vomiting. Others, again, are subject to bronchitis or broncho-pneumonia from slight causes. Tuberculosis of the lungs is not an unusual complication, and some cases succumb to general tuberculosis. Some of these patients suffer from albuminuria; others have attacks of hæmoptysis, unconnected with phthisis. These attacks are often rather beneficial than otherwise, relieving the overloaded lungs. Jaundice is an occasional complication. Some patients suffer from sores about the extremities, genitals and anus, and gangrene may bring their lives to a close. Irregular ulcerated tracts may be found in the intestines. The catamenia are absent as a rule in females reaching adult life.

Children with congenital heart disease, in addition to their other ailments, are often wasted both in mind and body, but this is not always the case. Such children of necessity lead a more or less inactive life, but the amount of physical and mental restriction depends in great measure on the extent of the cardiac mischief and associated lesions. Some may develop rheumatism or chorea. The respirations are liable to many variations. In some cases the dyspnœa is extreme, in others there is but little, if any; and again, in others, attacks of dyspnœa may occur from time to time. The pulse too may be natural, or weak, or rapid, and in the paroxysms it may intermit, be irregular both as to force and frequency, or barely perceptible at the wrist. As regards the temperature, such children, although showing no alteration of the internal temperature, yet are liable to cold extremities and chilliness of the surface. Dropsy occurs very infrequently in children.

Physical Signs.—In many chronic cases considerable præcordial bulging will be noticed, but in this there is nothing to distinguish them

from the acquired variety. The position, extent and force of the cardiac impulse will depend on the amount of hypertrophy affecting the ventricles and also the side affected.[1] Affections of the pulmonary artery being the most common, the condition met with in abnormalities of this structure will be first given. They are as follows. The cardiac impulse may be in its normal situation, or more or less displaced downwards and to the left. More or less epigastric impulse will be noticed, depending on the extent of the enlargement of the right ventricle. The impulse is, as a rule, forcible. The heart's dulness may commence at the second or third left costal cartilage, and extend to the right margin of the sternum, or beyond it for a finger's breadth or so. A regurgitant venous pulse will in many instances be noticed in the jugulars depending on tricuspid incompetency, and there may be also undue pulsation of the arteries of the neck. The liver is often enlarged, may extend two or three fingers' breadth below the

[1] Right-sided hypertrophy displaces the apex to the *left*. In insisting upon this point I am not alone, for Dr. Goodhart tells me he has himself taught this for many years.

right costal margin in the nipple line, and pulsation may be detected. The urine is frequently scanty and may be albuminous. Scanty urine is probably owing to want of arterial tension in the glomeruli. Thus in A. R. E.'s case the malpighian corpuscles contained greatly enlarged capillaries. The afferent arteries admitted but two or three red corpuscles abreast, whilst the enormously dilated efferent veins contained seven or more.

Stethoscopic Signs.—The stethoscopic signs are of very varied character. In many cases they are absent.

J. S. K., aged $3\frac{1}{2}$ months, a case sent to me by my colleague, Mr. F. S. Eve, was very cyanosed about the lips and tongue. The face was leaden, and the hands and feet blue. The cardiac impulse was in the fourth[1] interspace external to the nipple line. No bruit. Chronic snuffles.

In others they appear after some considerable interval, after some illness, as an attack of bronchitis, if not present at first. In others the

[1] The cardiac impulse in children is naturally either in the fourth or fifth interspace internal to the nipple line. In my experience it is very unfrequently found *outside* the nipple line. The position of the nipple is not constant.

heart's sounds may be normal during rest, but under excitement a well-marked bruit appears, and again in others the converse holds true. Bruits, too, may be modified by the respirations, or disappear during certain of the respiratory phenomena.

As regards their intensity they are equally variable. They may be more or less localised, heard all over the præcordia, or all over the chest, and more or less over the back. They may vary in intensity from time to time, and finally disappear towards the close of the case. A thrill may or may not be present, and if so usually, though not invariably, situated over the left base and systolic in point of time. Affections of the pulmonary artery being by far the most common, bruits dependent upon such conditions will be first described.

In stenosis the bruit may be most intense anywhere between the second left costal interspace and the level of the xiphoid cartilage, and an inch or more on either side of this line, but the most common situation is perhaps either the second left interspace or third left costal cartilage. In point of time it is systolic,

and may rarely be attended by a diastolic whiff heard now and again. *Apropos* of this it may be noted that bicuspid valves and those formed by a single curtain are liable when diseased to regurgitation. If the bruit is carried to the left clavicle the pulmonary artery is probably stenosed, but it may be conducted also to the right, and heard in the carotids and other great vessels. Under these circumstances there is possibly a patent septum ventriculorum, the bruit formed at the septum being conducted into the great vessels of the neck with the current of blood. If the bruit is heard better towards the right clavicle than towards the left, and also in the vessels, the condition is possibly one of atresia with a patent ventricular septum, always of course providing that the symptoms point to some serious cardiac defect. In spite, however, of the above remarks, it does not follow because the bruit does not correspond to these lines that the condition is not one of atresia, or stenosis of the pulmonary artery after all, and the converse may be said, though this is much less likely. A perforate septum ventriculorum

may give rise to much the same murmur as that heard in atresia of the pulmonary orifice, and in atresia of this vessel the murmur heard may be manufactured at the septal perforation.

Where, however, there is no deviation of the septum, the pulmonary artery and aorta arising naturally, patency will not give rise to a murmur heard in the great vessels, but there may be a basic bruit audible over the pulmonary area, and often most intense about the third left costal cartilage. A perforation the size of a pin's head in the undefended space has been known to give rise to such a bruit. Stenosis of the aorta may originate a bruit heard all over the chest, the maximum intensity being at the apex, and not to the signs usually recognised as belonging to aortic disease.

Basic systolic bruits suggestive of stenosis of the pulmonary artery may be due to simple dilatation of the artery, and equally intense murmurs may be detected in simple anæmia. Moreover, a loud systolic bruit best heard over the base may represent a communication between the aorta and pulmonary artery. A basic systolic bruit may be heard in patent

ductus arteriosus. Tricuspid regurgitation may give rise to a systolic murmur best heard over the tricuspid area, viz., over the base of the ensiform cartilage; but in young infants an apical murmur may mean either tricuspid regurgitation, or mitral regurgitation, or both of them. A presystolic or diastolic murmur over the tricuspid area would settle the difficulty in favour of tricuspid disease. An associated sign in tricuspid incompetency is venous regurgitation in the veins of the neck, which would prove of assistance in diagnosis. Further, the extension of cardiac dulness to the right, and epigastric pulsation would be of some service in forming an opinion. The murmur, however, if present, may be drowned by or merged into a coexistent loud systolic bruit manufactured elsewhere, and so rendered difficult of detection. Tricuspid regurgitation has been detected *in utero* by abdominal auscultation by Professor Peter, who heard a systolic bruit replacing the first sounds of the fœtal heart. The child was stillborn, and at the autopsy the tricuspid valve covered with vegetations was

found incompetent, being tied down by shortened chordæ tendineæ. From what has been said it will be seen that the conclusions to be drawn from the point of maximum intensity, and the directions taken by cardiac bruits in congenital affections of that organ, are very unreliable in themselves and that great stress must not be laid upon them. The nature of the bruit and its attributes taken together with the cardiac symptoms, the condition of the ventricles as gathered by inspection, percussion, and auscultation, the history and age of the patient, will often, however, in combination give a tolerably correct knowledge of the malformation. Further, an important factor in the diagnosis is the fact that the percentage of affections of the pulmonary orifice in cyanosed patients who have survived the twelfth year is very high indeed—according to Peacock 86 per cent.

Prognosis.—The prognosis depends on the nature of the malformation, but many cases, whatever this may be, die during infancy. Should the children survive the first year or so the chances of a more prolonged duration of life are considerably increased. When the heart

consists of but two cavities, life can only be prolonged for a few days at most. With hearts possessing three cavities life is very precarious, but even with this condition adult life has been reached.

In simple stenosis of the pulmonary artery old age may be attained, and the same may be said when this condition is complicated by a patent foramen ovale, but the prognosis is then not quite so good. When the ventricular septum is deficient the probable duration of life is a little less. In atresia of the pulmonary artery the age arrived at depends on the presence or absence of a patent septum ventriculorum, the prognosis being much better in the former case. Even with this advantage adult life cannot be reached. With a patent foramen ovale only the children die very early. Transposition of the pulmonary artery and aorta is a defect incompatible with prolonged life — its duration may be counted in months. These remarks, however, pre-suppose that the cardiac condition can be accurately gauged. Bi- and tri-loculate hearts cannot be diagnosed, nor can, amongst other conditions, transposition of the

aorta and pulmonary artery. In the latter case a septal murmur from associated perforate septum ventriculorum may be there, but given a case over three years of age it is not in the least likely to be in association with that malformation.

When cyanosis is present, patients gradually become more or less accustomed to their semi-asphyxiated state. The future of the case may depend on a variety of circumstances. It may be connected with the integrity of the heart muscle and the duration of its compensatory capacity by hypertrophy for overcoming abnormal conditions. This compensatory capacity is seen at its best in children, compensation in the growing organism being far more durable and efficacious than later on in life. Then, again, the duration of compensation depends upon the mode of life. Muscular exertion and a life of privation are likely to bring this power of compensation more quickly to an end than the converse. An attack of rheumatic fever, from the likelihood of cardiac complication, is one of great gravity. When adult life has been reached, dropsy may terminate the case.

Further advanced cyanotic cases, although perhaps not likely to die from that cause alone, nevertheless are on the brink of a precipice. An attack of syncope, coma, or convulsions at any moment may terminate their career. These cases have but trifling resisting power, and slight causes may induce wide-spread inflammation in abnormally congested tissues. Thus are seen bronchitis and broncho-pneumonia, with a rapidly fatal termination.

Hæmoptysis, apart from and in association with phthisis, may bring life to a close. Many succumb to phthisis or general tuberculosis. The advent of puberty, with its cardiac and vascular changes, is one of added danger. Even under the best possible environment, life beyond thirty or so is quite a rarity. The prognosis, then, is somewhat intricate and uncertain, owing to the complex variety of the malformations and the difficulties attending the diagnosis of the exact nature of these conditions.

Treatment.—Patients should be completely clothed in flannel, and in the case of infants they should be swaddled in cotton wool fixed

by flannel roller bandages. Every care should be taken to prevent them catching cold, and the temperature, if possible, should be an equable one. The chances of a rheumatic attack for such as these is a very serious matter, as this would to a certainty affect the already compromised valves. Any exposure to cold and wet and sudden chills must be carefully provided against. Such patients should, if possible, be removed from cold, damp, and low-lying districts. The food should be light, nutritious, and easily digestible. Gastric indiscretions, may lead to a speedily fatal termination, or failing that, to attacks of dyspnœa or convulsions. In the relief of the paroxysms of dyspnœa the cause should be first enquired into. If due to indiscretions of diet an emetic will rectify matters, or if caused by mental excitement, or by physical exertion, absolute quiet must be enjoined on the part of older children, and in the case of infants every attempt should be made by the nurse to soothe their agitation. As regards drugs, alcohol or chloroform may be tried. Diarrhœa and vomiting must be treated on general principles.

The heart symptoms must be treated on the lines adopted in acquired disease. When digitalis is contra-indicated, hypodermic injections of ether will prove beneficial. Sir W. B. Foster recommends peroxide of hydrogen in 8-minim doses three times daily.

Attention should be carefully paid to regular action of the bowels, and daily warm baths followed by friction with the flesh brush in older children, and the hand in infants, will be found of the utmost service. Saline purgatives administered from time to time will be found beneficial. Patients should be encouraged to lead a quiet, regular life, both mentally and physically, and every care should be taken to prevent cerebral excitement or depression. Children the subject of such malformations are particularly prone to outbursts of passion, and such ebullitions of temper are most injurious. Complications must be treated as they arise and on ordinary principles. In the event of coma supervening, venæsection may be tried; it is sometimes successful. Dr. Peacock advocates a few leeches behind the ears or on the temples, but calls attention to the danger of hæmorrhage arising from the leech bites.

In conclusion, the following cases which I have selected from my note books, some of which contain *post-mortem* notes, may be read with interest. Many of them show the difficulties of coming to a conclusion as to the nature of the malformation present. Illustrative of many of the points which have been dilated upon in the text, they also convey an idea of the class of cases which will be encountered in practice from time to time.

Case I.—William H., aged 14 years, attended for hæmoptysis, cough, night sweats, and fainting fits. No history of rheumatic fever, and never convulsed.

On examination.—A pale, emaciated boy with livid lips, congested palpebral conjunctivæ, and marked clubbing and cyanosis, of the fingers and toes.

Heart.—Impulse in the fifth left interspace in the nipple line. A blowing systolic bruit of greatest intensity at the third left interspace, three-quarters of an inch external to the left sternal margin, and conducted from this point at radii of three inches all ways. Murmur not audible in the arteries.

Lungs.—Resonant. Râles and rhonchi.

Liver.—Dulness two fingers below the costal margin in the nipple line, and there seems some tenderness below and over this.

Spleen not enlarged.

Urine. — Small in quantity, and passed frequently. Albumen.

Eyes.—The conjunctival vessels ocular and palpebral, also the scleral veins very tortuous and injected—those on the palpebral conjunctivæ are even granular-looking from this cause.

Pulse 118 to the minute, weak and regular. Temperature 97° F. On making enquiries of the mother, I learned that the child died some months after ceasing attendance at the Evelina Hospital.

Case II.—Emily J., aged 7 months, was brought by her mother for "blueness," and on her arrival was in convulsions. The mother had borne four children, and had had two miscarriages. Two or three of the children had "thrush,"[1] which ran through them down to their feet.

On examination.—Child unconscious, breathing deep, and expiration noisy, the facial movements as those of the last effort of the respiratory centre. Child cyanosed, lips and tongue blue, eyes suffused, fingers and toes blue, but not clubbed. Pulse not felt at the wrist.

Heart.—No enlargement detected. No bruit. My clinical assistant heard a systolic murmur over the cardiac area prior to the fit.

Liver.—One inch below the costal margin. *Spleen* just below the ribs. *Lungs*—no adventitious sounds. Under treatment the child rallied, but died a month later.

A *post-mortem* examination could not be obtained.

Case III.—Nellie K., aged 2 months. An emaciated, syphilitic child, with considerable enlargement of the

[1] Thrush, which "passes through the body," and extends below the knees, is certainly a syphilitic eruption. Mothers who deny the presence of an *eruption*, will readily admit the former.

liver and spleen, mucous tubercles of the mouth, a macular syphilide, and arms and hands desquamating.

Heart.—Area of cardiac dulness not increased.

Sounds.—A systolic bruit, heard best over the left base, conducted to both clavicles, but better to the left, heard over both chests, back and front, louder behind on the left side, and heard in the great vessels of the neck. When she cries she becomes a little blue on the forehead. Lips of good colour. She remained under treatment for some months, with great benefit to her general health. She died subsequently, and the parents would not permit an autopsy.

Case IV.—Maud F., aged 8 years, did not speak until six years of age. When two years old a doctor told the mother that the child suffered from congenital heart disease. At that time the face was very frequently purple in colour. Always subject to cough and headache.

On examination.—The cheeks are flushed, but not cyanosed. *Pulse* 88, rather small, regular, and compressible. *Respirations* 24. *Temperature* 99.6° F.

Heart.—Impulse in the sixth interspace, one and a quarter inches external to the nipple. Dulness commences at the left second costal cartilage, and extends well to the right margin of the sternum.

Sounds.—A loud blowing systolic bruit over the pulmonary area, loudest in the third space, and here "churning." This can be traced up in the neck vessels on both sides, but more audible on the left. Audible all over the chest, but more intense on the left side. At the apex it changes its character to a blowing sound, is heard plainly in the left axilla, less so in the right. The bruit is audible behind, but very faintly. The bruit is

followed by a noisy laboured second sound, and then a pause. A marked systolic *thrill* at the third left space, and also to a less extent at the apex. No albumen.

Case V.—W. J. A., aged $1\frac{5}{12}$ years, was brought to the Evelina Hospital for whooping cough, and on examination was found to have slight broncho-pneumonia. His mother stated that his lips at times became very blue. Respirations 60; pneumonic grunting; temperature 99° F.

Heart.—Impulse in the fifth interspace just internal to the nipple line.

Sounds.—A loud blowing systolic bruit heard best at the left side of the xiphoid cartilage, and next best at the left base, conducted better to the left clavicle than the right, heard in the great vessels of the neck and over both chests, back and front, better perhaps over the left. The lips become a trifle blue on coughing, but this is of course frequently seen in children with whooping cough without congenital heart malformation.

Case VI.—Herbert A., aged $1\frac{7}{12}$ years, was one of twins. From birth fretful, peevish, and puny; he suffered from blueness of the fingers and toes.

On examination, March 26.—Child screaming, but does not become more cyanosed.

Heart.—Heaving impulse. Epigastric pulsation, and dulness well to the right margin of the sternum.

Sounds.—A loud systolic murmur heard all over the præcordium and loudest at the left base. Conducted to the left clavicle, not so well to the right, and heard in the great vessels of the neck. May 14.—During the week he was taken to Willesden and appeared better. Whilst away he became very blue and died during the attack.

Post-mortem.—On opening the pericardium the right side of the heart appeared dilated, and the dilated aorta seemed to rise chiefly from the right ventricle. The pulmonary artery was represented by a vessel the size of a crowquill. The ductus arteriosus was pervious and dilated to the size of the left common carotid. The branches conveying blood to the lungs arose from the pulmonary artery close to its junction with the ductus arteriosus. The lungs were normal.[1]

Section.—Right ventricle hypertrophied and three or four times the thickness of the left, and the cavity more capacious than the left. Auricular walls of equal thickness. The aorta arose from both ventricles, rather more from the right. Septum ventriculorum perforate. *Foramen ovale* admitted a crowquill, but the two surfaces of its valve were in close apposition.

Tricuspid valve edges somewhat crumpled and thickened. The commencement of the pulmonary artery was completely occluded, no trace of the valves being apparent. The artery itself was patent from its point of exit from the heart, its lumen being about the size of a piece of whip cord. The abdominal organs were not markedly congested.

Case VII.—Arthur B., aged 2 years—a seven months' child. Very blue at birth. Child thin and cyanosed, becoming worse when crying. Has never walked or talked.

Heart.—Impulse diffuse in the fifth interspace half-an-inch external to the nipple line, no bruit.

Hands, feet and lips blue.

Case VIII.—Arthur F., aged 2 years, becomes

[1] A microscopical examination was not made.

"black" in the face, hands and toes ever since birth; never walked. Subject to "insensible fits," but none for the last six months.

On examination.—Face cyanosed, eyes suffused, lips, fingers, and toes blue, with slight clubbing of the fingers. Child very thin.

Heart.—Impulse localised in the fifth interspace in the nipple line. Resonance impaired one finger's breadth beyond the right sternal margin.

Sounds.—A loud blowing systolic bruit all over the cardiac area, of greatest intensity at the second left costal cartilage, and conducted best to the left clavicle. Over the cardiac area there are heard normal first and second sounds. The bruit is heard over both chests back and front. A few rhonchi in the lungs. Front of chest prominent. Liver two fingers below the costal margin. Three weeks after his first attendance he developed jaundice.

Case IX.—Walter S., aged $2\frac{1}{2}$ years, was brought by the mother for convulsions of a week's duration. A bad fit when a year old, and another one four months ago. Rheumatism and heart disease in the family.

On examination.—Lips of good colour and no clubbing of the fingers and toes.

Heart.—Impulse fifth interspace just outside the nipple line. No epigastric impulse, but a systolic thrill there.

Area of dulness.—Commences fourth left rib and does not pass beyond the left sternal margin. A very marked systolic *thrill* over the second left interspace and neighbourhood. Here there is a loud, very harsh systolic bruit, and of greatest intensity at this point. Con-

ducted better towards the left clavicle than the right, and heard all over the cardiac area and front of the chest, to the great vessels of the neck, but not particularly loud there, and faintly over the back, louder on the left side.

Organs normal.

Case X.—Ernest S., aged 7 months, wasting since 3 months old. Dyspnœa, worse at night, and on exertion the fingers and toes become blue, but the latter for the last few weeks has not been noticed.

On examination.—Child thin, breathing rapid and difficult. No cyanosis. Respirations 68. Pulse 138. Temperature 98°.8 F.

Heart.—Impulse in the sixth interspace two inches outside the nipple line.

Dulness.—Well to the right side of the right sternal margin.

Sounds.—A loud blowing systolic bruit with a well-marked *thrill* of greatest intensity at the left second interspace, conducted more to the left side than the right, but heard over both, well to the left clavicle, and here loud, faintly to the right. Bruit just detected behind.

Lungs. — No abnormal sounds. *Liver* two fingers below the costal margin.

Case XI.—Walter S., aged 3 months. Brought for advice because from the first day of birth his breathing had been very "heavy," worse at times. He has attacks of crying, and goes "different colours in the face." Blue about the lips when he has these attacks. Rheumatism and heart disease in the family.

On examination.—Lips of good colour. Respirations very rapid and irregular.

Heart.—Impulse in the sixth interspace in the nipple line; no epigastric pulsation.

Sounds.—Loud blowing systolic, heard best at the apex and conducted well into the axilla. Heard over the superficial area of cardiac dulness, and not conducted to the clavicles.

Lungs normal. *Liver* one finger below the costal margin. *Spleen* normal.

Case XII.—Tom M., aged 6 months. Blueness of lips and extremities since birth. Snuffles since birth, and a history of a rash on the buttocks. Other children have not snuffled, but they had "slight rashes on the buttocks." No rheumatism.

On examination.—A fairly nourished child with cyanosed lips and extremities.

Heart.—Impulse forcible and diffused in the fifth interspace, just internal to the nipple line. Strongly felt in the costal angle.

Sounds.—A loud blowing systolic bruit over the cardiac area, loudest left sternal margin, conducted to right clavicle, also left, and here heard much better. Audible in the vessels of the neck.

Lungs, a few rhonchi. Pulse 174. Respirations 48.

Case XIII.—Ernest P., aged 4 years, cyanosed ever since birth. Becomes "black" if excited or fatigued. Not able to walk, subject to bronchitis. His breath is very short. He has convulsions occasionally, and is always more or less cold. Very fretful.

On examination.—The child is fairly well nourished. Fingers and toes very clubbed and blue. Tongue and lips blue. Conjunctivæ injected and blue.

Heart.—Impulse fifth interspace internal to the nipple line. Epigastric pulsation.

Dulness.—Third left costal cartilage, well to the right of right sternal margin.

Sounds.—The second sound over the pulmonary area is very sharp and metallic. No bruit.

Lungs.—No adventitious sounds. Pulse 120, of good volume and regular.

Case XIV.—The notes of this case were given me by my old friend and former colleague, Dr. Albert Martin, of Wellington, New Zealand.

Fred. R., aged 12, was first seen during an attack of influenza, and it was whilst undergoing physical examination that the following cardiac phenomena were detected.

Heart.—Impulse fifth interspace half-an-inch internal to the nipple.

Dulness not increased to the right.

Sounds.—At the apex a slight bruit accompanied the first sound, increasing in intensity across the sternum to the aortic cartilages on the right side. At a corresponding point at the left the sound is louder. Posteriorly at the upper internal angle of the left scapula during the systole there is a loud whistling bruit heard more or less over the whole of the chest and back, but whose points of greatest intensity extend in a line just to the left of the spine, from the top of the scapula to the base of the spine in the sacral region, and in fact corresponding to the line of the thoracic and abdominal aorta. The bruit is audible in the subclavian and carotids, with a preponderance in favour of the left vessels, while on applying the ear to the vertex of the skull a distant musical short bruit is heard. The bruit is audible in the femorals but not in the popliteals. *Pulse* 80, full, regular and equal both des. *Eyes.*—The

veins of the optic disc pulsate and appear beaded. No cyanosis or coldness of the extremities. He plays football when well. Examined three months later his heart was found in quite the same condition.

Case XV.—Nellie P., aged 10 months. A wasted, feeble child was brought to the Evelina Hospital with measles and a cough suspiciously like whooping cough. But recently convalescent from chicken-pox. *Mother.*—Three children, two miscarriages. All snuffles soon after birth. *Thrush.*—One child, "passed to bottom and right down to knees." *Pulse* 136. Respirations 36. Temperature 99.4° F.

On examination. — *Heart.* — Impulse fifth interspace just internal to the nipple line.

Sounds.—Systolic bruit loudest over left base, audible over front of chest, conducted best to left clavicle, and just audible in the vessels of the neck.

No cyanosis. *Lungs.* — Slight broncho-pneumonia. *Spleen* two fingers below the costal margin. Commencing rickets.

Case XVI.—William E., aged 16 years, of late has complained of pains in his head and back, but is bright and cheerful. He used to suffer from hæmoptysis.

On examination.—Face dusky, lips blue, and marked clubbing of the fingers and toes. *Pulse*, small but regular.

Heart.—Impulse fifth interspace just external to the nipple. No epigastric pulsation. Dulness third left costal cartilage, and well to the right margin of the sternum.

Sounds.—A systolic bruit, not of great intensity, heard best on the fourth left interspace, but close to the sternum. It is detected best along this margin. Not heard

under either clavicle or over the right base. The apex is free from murmur. Not heard in the neck. No thrill. *Lungs.*—A few râles and rhonchi. *Chest* bulged in front and broadly guttered on either side.

CASE XVII.—William C., aged $3\frac{5}{12}$ years, seen Oct. 4. A healthy child until four months back, when drowsiness, dyspnœa, and duskiness of the cheeks appeared. For the last fourteen days orthopnœa, œdema of the legs, puffiness of the face, and passage of small quantities of urine. Mother and one sister suffered from rheumatic fever.

On examination.—A well nourished but much cyanosed child. *Lungs.*—Nothing abnormal detected. *Heart.*—Impulse displaced outwards; dulness to right of sternum. *Sounds.*—Loud systolic bruit at apex, conducted to axilla and angle of the scapula. Heard all over the cardiac area and chest, but loudest at the apex and better towards the left clavicle than the right. Slight epigastric pulsation. *Liver.*—Two fingers' breadth below the costal margin; slight pulsation. Albumen a trace. *Pulse* 120, *respirations* 28, and *temperature* normal. The following day his temperature rose to 101.8° F. Passed rather more urine (5 oz.). Albumen $\frac{1}{5}$. Not quite so blue. On the 6th blood in the urine; albumen $\frac{1}{20}$th. On 7th both bases dull and deficient entry of air; dyspnœa very great, pulselessness and collapse. I aspirated him and withdrew $13\frac{1}{2}$ oz. from the right chest and 11 oz. from the left. Both fluids coagulated. No distinct improvement after the operation, and he died the following day. The temperature went up to 102°F.

Post-mortem.—Much fluid both bases; a little lymph on the pleuræ. *Abdomen.*—Some ascites; no lymph. *Pericardium.*—A little fluid. *Heart.*—Both ventricles hy-

pertrophied ; the wall of the right as thick as that of the left. *Left auricle* enormously dilated. Pulmonary artery stenosed greatly. Just above the pulmonary valves the lumen was as big as an average crowquill. Valves thickened and united into a cone. Left ventricle much dilated. *Mitral valve* healthy, but the pink excrescences so commonly met with in children were of unusual size and distinctness, giving a very characteristic, elevated dark red ridge at the margin of the valve. Everywhere the surface was perfectly smooth, and there was not a trace of fibrinous deposit.[1]

Aorta.—Somewhat reduced, valves thick, presenting at their edges a similar condition to that of the mitral. Here also the surface was perfectly smooth, and free from fibrin. *Kidneys* congested but not hard. *Spleen* not large but very red.

One cannot forbear remarking about this case that it well illustrates the difficulties or rather the impossibilities of making a correct diagnosis in many instances. The characteristics of the bruit undoubtedly pointed to mitral regurgitation, whereas the inspection proved conclusively that it was due to pulmonary stenosis.

Case XVIII.—Walter G., aged two months. The child has paroxysms of dyspnœa at intervals of half an hour, during which he is excessively cyanosed, the face, lips, tongue, gums and fingers becoming very livid. During the paroxysms he opens his mouth, gasps for

[1] To those not acquainted with these nodules on the auriculo-ventricular valves, such are frequently mistaken for early endocarditis ; they are, however, quite normal. With regard to the nodules on the aortic valves, they must, I think, be considered due to endocarditis. I am not acquainted with such structures in the semi-lunar valves normally, at least nothing of the nature described.

breath, and the alæ nasi dilate. Conjunctivæ injected, and the eyes are opened and shut drowsily. Finger ends not clubbed.

Heart.—Impulse fifth interspace just below the nipple. Percusses to left margin of the sternum. A systolic murmur is heard over both chests, back and front, louder behind on the left. The point of greatest intensity of the murmur is at the second right costal cartilage.

Liver.—Just below the costal margin.

Spleen not enlarged.

Mother has six children and one suffers from heart disease. Two miscarriages.

Case. XIX.—A girl, aged $4\frac{1}{2}$ years, suffering from spastic paralysis, attended as out-patient at the Evelina Hospital. She was a well-nourished, healthy-looking child, without any external evidence of cardiac disease. No rheumatic history.

Heart.—Area of dulness and impulse perfectly natural. A loud blowing systolic bruit was heard over the second left interspace, the third costal cartilage, the fourth, and a trifle below this. The point of maximum intensity was in the third left interspace. The bruit was not conducted into the great vessels, nor heard at or near the apex. The first and second sounds at the apex were clear and loud. At the right base clear valvular sounds were heard, and at the left the second sound was accentuated, and there was a marked valvular snap. She was under observation for six months, and no alteration was observed in the above.

Case XX.—Eliza E. K., aged 6 years, had been ill for a week with cough and pains in the chest, cheeks flushed, not a trace of cyanosis, finger ends normal. No history

of phthisis. Father died of bronchitis. Mother alive and strong; no miscarriages. Healthy child when born: no snuffles. No rheumatism. Pulse 104, small, somewhat irregular. Respirations 28, short little cough like pneumonia. Temperature 98.4° F.

Heart.—Impulse fifth interspace one inch external to the nipple. Doubtful exaggeration of the epigastric impulse. *Dulness.*—No extension to the right. *Sounds.*—A systolic bruit all over the cardiac area and front of the chest. When the patient lies down the bruit is loudest at the left base and in the erect position at the apex. Following the bruit at either base there is a second valvular sound, though not very decided; if anything it is more distinct on the left side. The bruit is conducted to the left clavicle, also to a less extent to the right clavicle, and is detected in the neck vessels, but here it has lost much of its intensity and is not loud. It is heard plainly over the left chest behind and not very distinctly over the right. Pulsating carotids. No venous regurgitation. Chest wall bulged over heart. A well marked systolic *thrill* felt best over the cardiac impulse and just detected over the left base.

Lungs.—Resonant all over except the right base, where there is dulness, deficient entry of air, bronchophony and tubular breathing. Elsewhere a few bronchitic signs.

Organs.—Other organs normal.

This child was under Dr. Frederick Taylor in the Evelina Hospital, when $3\frac{1}{2}$ years of age (Oct. 9, 1891), and the following notes are from his pen.

"*Present Condition.*—A pale, delicate child. No cyanosis. Glands under jaw enlarged. Wrists slightly thickened. Pulse 136, weak.

"*Lungs.*—Loss of resonance front and back of right apex. Complete dulness over rest of right lung, with

feeble healthy breath sounds of better quality in vertebral groove and apex. No crepitations or tubular breathing. *Heart's* apex diffused; most marked in the sixth space nipple line. Very marked *thrill* to be felt varying with pressure and less marked on firm pressure. Loud systolic bruit at apex, heard all over the chest especially running up to the left base. The maximum intensity was as follows:—Loudest over the ensiform cartilage, heard next best over the apex, then over the left base and next over the right. Behind louder over the upper half of the left chest than the lower. No presystolic definitely.

"*Abdomen*, lax. *Liver*, three fingers' breadth below the ribs. *Spleen* not felt. *Bowels* confined. *Urine*, no albumen.

"*October* 10. — Explored with syringe at angle of scapula; nothing drawn off.

"*October* 16.—Loss of resonance over front of right lung with feeble breath sounds. No tubular breathing or crepitations. Dulness at extreme left base behind; fair resonance above with fair breath sounds all down lung.

"*October* 23.—Up and dressed. Heart the same.

"*October* 28.—Left hospital.

"I seem to have thought that a perforated septum was a likely thing, and with the position of the murmur and thrill and the absence of cyanosis, I think I should think so still."

Such are Dr. Taylor's views on this case, and breaking my rule in not expressing an opinion at the conclusion of the cases enumerated I would add the following.

My own impression is that there is a perforated septum ventriculorum because the bruit is conducted into the great vessels of the neck. The basic bruit may

mean, amongst other things, this, or pulmonary stenosis or atresia. The better conduction of the murmur to the left clavicle would half suggest pulmonary stenosis, but it not improbably denotes (*vide* Case VI.) a perforated septum or pulmonary atresia (then of course perforated septum). I think there is some right sided ventricular hypertrophy, and I am doubtful as to the presence or absence of mitral regurgitation, but in association with this read Case XVII. On the whole I too am slightly in favour of a perforated septum ventriculorum only, but I would add that this diagnosis is not by any means an indisputable one for cyanosis, and other symptoms of congenital heart malformation may develop later on. Stenosis of the pulmonary artery in association with a perforate septum ventriculorum is not at all unlikely in this case in spite of the absence of symptoms.

Case XXI.—Josephine H., aged 6 years, had attended the Evelina Hospital some two years for a heart affection. She had suffered from influenza and measles. Family history good *On examination.*—Hands cold, feet blue over toes and heel. A purple flush on the cheeks, and when she cries becomes slightly blue. A thin child.

Heart.—Prominent chest. *Impulse*, fifth interspace half an inch outside the nipple. *Dulness*, one finger to the right of the sternum, impulse felt here. Epigastric impulse does not seem more than normal.

Sounds.—Systolic bruit, very loud, and heard best over the pulmonary orifice. No thrill. Bruit conducted up to left clavicle, heard in the carotid arteries. Bruit loud all over the cardiac area, detected in the left axilla, not in the right, but behind is audible over both chests from apex to base, more intense on the left side.

Lungs.—A few moist sounds. *Liver*, percusses two inches below the costal margin.

During her attendance in the year 1893 she suffered from attacks of cyanosis and palpitation, but she was able to skip. In December of that year her lips were appreciably darker, the eyes slightly suffused, she became cyanosed on crying, and the fingers and toes showed signs of clubbing. *Heart.*—Thrill over the pulmonary area. Exaggerated epigastric impulse. Over the right base second valvular sound, over the left base this is absent. The bruit was much as before. The points of greatest intensity were as follows. (1) Over the pulmonary area ; (2) over the fifth left costal cartilage ; (3) over the apex. *Pulse*, small and regular. *Lungs*, resonant, breath sounds vesicular.

Case XXII.—Emma R., aged 6 years, was brought for ulcerative stomatitis. It was said she had suffered from heart disease since birth, and always had a cough. Occasional hæmoptysis. *Pulse* 126, respirations 42, temperature 97° F.

Heart.—Impulse, sixth space outside nipple, epigastric pulsation. *Dulness*, to right sternal margin.

Sounds.—A rough blowing systolic murmur loudest at the junction of the third costal cartilage and the sternum. Heard all over the chest, back and front, and in the neck vessels. The apical murmur is very similar to the basic. Over a small area round the apex there is a double murmur, and over a small space between the apex and left base, there is a position when the systolic murmur is not so loud as at the apex and base.

No abnormalities detected in the organs. *Chest*, bulged over the heart, and a rickety Rosary.

The fact that I have made no attempt to diagnose the probable pathological changes in the cases narrated may excite comment. Had I done so, my remarks would have merely conveyed my impressions of their nature founded on the data I have given, impressions which can with advantage be formed unassisted by those reading this work. It really amounts to this, that given a case with the signs and symptoms I have detailed in the text, the particular cardiac malformation is very possibly of such a nature as has been described in association with these signs and symptoms. After such an admission it is hardly to be wondered at that I shrink from committing my impressions to paper, not from any personal motives, but because by so doing I may mislead rather than guide the reader by my apparent readiness in coming to a conclusion. When there is no bruit nothing more can be said, but this is an illustration of a congenitally deformed heart, and when there is one the preceding remarks apply. As clinical records they well illustrate the class of cases under discussion, and I offer them as a clinical exercise in much the same way that

text-books in chemistry or what not present at their termination mental exercises in the shape of problems for solution, but with the distinction in this book that a correct solution is impossible, short of an autopsy; questionable they are, doubtful they must remain.

A loud systolic bruit of greatest intensity over the left base is in childhood *almost* certainly due to some congenital affection of the heart.

Very rarely indeed are the healthy pulmonary valves organically attacked in extra-uterine life.

Anæmic bruits have already been mentioned, and any intense bruits in childhood from this cause and at this orifice are practically unknown.

Soft and low systolic murmurs over the left base, rarely the right, occurring in infants and children are occasionally heard. These, however, are in no wise so accentuated, so definite, so obtrusive in them as the pulmonary bruit due to organic disease. The soft murmur is usually followed by a pulmonary reduplicated second sound.

Apart from anæmia, ulcerative endocarditis in adults, as a most unusual occurrence, may attack the pulmonary valves.

The associated pulmonary symptoms and fever would point to the nature of the disease.

In children under 12 years of age that disease on this side of the heart may be almost ignored.

According to Cheadle[1] only a single case of ulcerative endocarditis appears in the records of the Great Ormond Street Children's Hospital during twenty years, where patients are admitted under the age of 12. The child was a girl of 8, and the mitral valve was affected. This, however, in my experience, is far too low an estimate. I have myself seen five cases in children of 8, 7, 7, 6 and 11 years respectively. The four former were females. The last occurred in my out-patients during 1893, and presented himself with fever, hæmaturia and heart disease. Vegetations and ulcerations were found on the aortic and mitral valves.

[1] Keating's "Cyclopædia of the Diseases of Children," p. 787, vol. i., part ii.

From 1886 to 1892 inclusive, there were 768 deaths at the Evelina Hospital, and of these four were occasioned by ulcerative endocarditis. Ulcerative endocarditis, in my experience, then, would not be so uncommon as Cheadle's statistics show, but I have never seen a case in childhood involving the pulmonary artery.

The greater number of the cases detailed illustrate the left base as the seat of election for congenital bruits, and the wide area over which they are usually conducted is another feature of these murmurs.

It is usual to look upon all cardiac bruits occurring under 4 years of age as of congenital origin, because acquired heart disease in children is very rare under that age. The most prevalent cause of heart disease, viz., rheumatism, apparently so seldom attacks children under that age, and chorea is even more exceptional still. The youngest example of chorea that I can find in my notes is that of a child of 3 years, and of articular rheumatism a child of 2 years. The younger the child, however, the more atypical usually is the rheumatism. If, when we speak of rheumatism in children

under 4, we look upon joint affections as the manifestation of that disorder, then it is so, very.

Cases, however, are on record of joint implications even in infants. Thus Henoch[1] gives a case of a child aged 10 months, who suffered from articular rheumatism, and others are narrated in literature younger than that.

With children of these tender years, parents are apt to ignore so-called "growing pains" (pains in the joints, tenderness of the tendons and muscles), either because the child is incapable of giving expression to his feelings other than by crying, the cause for which may be readily overlooked, or even if they are recognised their gravity is not understood, and they are passed by as due *only* to the above and, therefore, of no moment.

Rheumatism is protean. Slight pyrexia may be the only manifestation, and the child thought "only a little feverish." Examination of the heart in some such cases may give a clue to the nature of the fever, which otherwise would have been passed over as some-

[1] "Diseases of Children," p. 311.

thing trivial and occasioned by being "out of sorts." Pericarditis is another rheumatic manifestation, and here again infants, though quite exceptionally, may suffer. West[1] records such a condition in a child of 7 months. The *post-mortem* showed that there had been a previous attack at the age of 4 months. Rheumatic pericarditis in infants and young children is, in my experience, quite exceptional, and when present is usually associated with endocarditis. Other rheumatic manifestations are pleurisy[2] and pleuro-pneumonia.

Of 190 cases of pleurisy there were sixty-eight under 4 years of age, seventeen of these being simple serous effusions. The youngest was 14 months. Eliminating tubercular cases, broncho-pneumonia, nephritic and those following the specific fevers, a moderate percentage of these had a family history of rheumatic fever or heart disease. It is probable, then, that some of them owed their disease to rheumatism. Of other rheumatic manifestations there are tonsillitis, erythema multiforme and possibly

[1] "Diseases of Infancy and Childhood," seventh edition, pp. 556-7.

[2] "Pleurisy in Childhood," by the author, "International Clinics," vol. iv., second series.

erythema nodosum. Tonsillitis is seldom seen, in my experience, in such young children, and the erythemata only occasionally. It appears, then, that rheumatism does occur in infants and children under 4 years of age and clinically is met with occasionally, but it is often difficult to diagnose and may easily be passed over, making it appear rarer than is really the case. Acquired heart disease can and does occur in infants and children under that age, though here again this is rare. Taking the records of the Evelina Hospital of 768 deaths, there was one case of mitral disease aged 3 years, and another aged 3 years of mitral disease and pericarditis, and another aged 5 months with mitral disease. Here again statistics, perhaps, do not show all they are intended, because at the Evelina Hospital cases are not admitted, as a rule, under 2 years of age. Although this is so, a large number of infants are admitted into the wards during the course of the year, but, perhaps, not quite so many as would otherwise be the case if this rule were not in operation.

Cases XI. and XVIII. in my list are possibly of that nature, but the history of the

former is rather in favour of a congenital affection. The former seems to point to mitral implication, and the latter may possibly be owing to aortic trouble. Apart from these I have met with but one case of mitral disease under 4 years, viz., in a child 3 years old. Goodhart[1] gives the history and autopsy of a child aged 2 months admitted to the Evelina Hospital, in whom the disease was apparently extra-uterine.

The mitral valve edge was thick, and on its surface were abundant inflammatory granulations uniformly distributed round the orifice, and quite sufficient to interfere with its efficient closure.

Dr. Goodhart also says that he could give notes of several other cases of infants but a few months old in whom the physical signs were in favour of mitral regurgitation.

It must not be assumed, therefore, because the patient is under the age limit just mentioned, that acquired heart disease need not be thought of, and the remarks I have made are to emphasize the fact of the occasional occurrence of that disorder in infancy.

[1] "Diseases of Children," fourth edition, pp. 601-2.