medical literature. The complete recovery which occurred in Mr. M.'s case could not have occurred had there been either haemorrhage or softening in the motor regions affected, or in the part of the optic radiation which determined the hemianopia.

A WORKING HYPOTHESIS.

The explanation, moreover, provides a working hypothesis of great practical value, and this is the reason of my bringing the subject before you to-day. Take the phenomena in Mr. M.'s case. Extending over three months there were symptoms commonly regarded as premonitory, or as the threatenings of cerebral haemorrhage. But what is meant by *threatening* of cerebral haemorrhage? It is either haemorrhage or not; there is no half-way stage in haemorrhage, although the quantity varies greatly. But these threatenings also precede thrombosis and softening; they also precede the paralysis due merely to temporary arterial closing, as in Mr. M. The "paralytic stroke," due to any of three different pathological processes, has the same premonitory threatenings, and it is of no small practical importance that there should be a clear picture of the clinical pathology of this stage. These threatenings can only, I take it, be due to irregular vessel action, and so to irregular blood flow through the part. There must be vessel constriction, local in site, varying in degree and in extent, coming and going, intermittent.

EFFECTS OF INTERMITTENT CLOSING.

The effects of intermittent closing may be placed in four groups:

1. It leads to impairment or suspension of the function of the part affected, as manifested by sensory disturbances and motor faltering or paralysis.
2. It favours thrombosis in certain states of the vessel wall, or of the blood itself, or of both combined.
3. If the heart be weak the blood flow is more easily arrested, thrombosis is favoured, and the risk of softening is greater.
4. If the heart be strong and the vessels not sound, haemorrhage is liable to occur. The haemorrhage, however, does not take place in the closed portion of vessel; it must take place on the proximal side, where the pressure is raised because of the distal closing.

The significance of the premonitory warnings and threatenings is thus seen, and the aim of practical therapeutics is made clearer.

ETIOLOGY AND TREATMENT.

We have thus a picture of intermittent closing of arteries occurring in different parts of the brain, occurring in persons of very different types, and occurring, as in our case, at the early age of 50 years. If you ask about the etiology of this irregular vessel action we are at once brought face to face with fundamental problems. There are questions of constitution such as are embraced in the words "gouty" and "rheumatic"; there are questions in metabolism, such as the accumulation of the waste products of cell activity, the fatigue toxins, and the inefficient removal of such. There are questions of food and drink; there is the condition of the bowels, and the absorption of toxins from the intestinal tract. There are questions of skin and kidney action. There are questions of mental work and of mental repose; there is emotion and passion. In no department of our work is it so necessary to form a sound all-round estimate of the patient—habits, digestion, bowel action, kidney efficiency, the cardio-vascular system, have all to be reviewed and estimated aright, if we are to be guided aright. What is to be aimed at is the lowering or removal of the abnormal vessel irritability. In the robust plethoric subject the threatenings of which we have spoken have long been treated by active purgation, diminished food supply, physical and perhaps mental rest. All these measures are good, but the plethoric type does not supply the greater part of the cases we are now considering. In many cases active purgation, and all extreme measures, are to be avoided as positively hurtful. Rest, physical and mental, gentle bowel action, spare diet with no red flesh, are the general lines of treatment. The irritable tonicity of the vessel wall is in many cases wonderfully controlled by spiritus aetheris nitrosi. When symptoms appear to be more urgent I have much confidence in erythrol tetranitrate in $\frac{1}{4}$ to $\frac{1}{2}$ grain

doses, repeated every three, four, or six hours, as may be required. The action is temporary, but it allows time for the bowels to act fully and for the elimination of the excess of waste products in the tissues. Thereafter restricted diet and daily bowel action may prevent the recurrence of symptoms. In other cases the vessel irritability is markedly allayed by the administration of iodide of potassium. I would very strongly warn you against violent purgation with calomel and similar drugs in persons with a feeble circulation, for the lowering of such a circulation is the surest way to ensure softening, and even subsequent haemorrhage. The feeble circulation has to be recognized behind thickened and incompressible vessels; see to it that you are not deluded into thinking that there is a strong heart because there is a high haemomanometer reading. A tonic as nux vomica may be a valuable addition to spirit of nitrous ether or to iodide of potassium in cases of this kind.

By a true estimate of the patient as a whole, by the wise use of the simple methods indicated, the irritability of the arterial wall disappears and can be prevented returning to any alarming extent. In this way we can by foresight prevent such truly pathetic cases as must be known to most of you.

The Wightman Lecture

ON

CONGENITAL HEART AFFECTIONS,

ESPECIALLY IN RELATION TO THE DIAGNOSIS
OF THE VARIOUS MALFORMATIONS.

DELIVERED BEFORE THE ROYAL SOCIETY OF MEDICINE,
SECTION OF DISEASES OF CHILDREN.

BY GEORGE CARPENTER, M.D.,

PHYSICIAN TO THE QUEEN'S HOSPITAL FOR CHILDREN, LONDON.

GENTLEMEN,—In acknowledging the honour that you have conferred upon me by inviting me to deliver the Wightman Lecture, I must confess that the acceptance of this distinction proved far easier than the selection of a suitable subject for the occasion. Passing now to the subject of congenital affections of the heart, I will first draw your attention to the known causes which bring about the various departures from the normal, which are classed under the heading "Congenital Heart Affections."

The coincidence of congenital malformations of the heart with malformations in other regions of the body is a matter of everyday experience. Heart malformations are not always accompanied by other congenital bodily defects, but they are sometimes found in association with such diverse structural faults as hare-lip, cleft palate, ill-developed teeth, supernumerary auricles, and supernumerary nipples, polydactylism, syndactylism, webbed fingers, steeple-skull, defects in the abdominal wall, herniae, ill-developed and absent rib cartilages, undescended testis, atresia of the anus and rectum, post-anal dimple, congenital opacity of the cornea, coloboma iris, congenital defects of the orbits, congenital ptosis, not to mention many other congenital irregularities of development which are obvious to the naked eye—cretinism, for instance—which sometimes bear them company.

Since heart malformations have been shown to be a family complaint in some instances, it must be admitted that inherited as well as acquired defects in the sperm and germ cells of the parents are of some importance in the production of malformations of the heart, as in other regions, for erratic tendencies to produce deformities, when once acquired, are handed down from parents to children.

Inquiry has been directed to the state of the mother's health, both bodily and mental, during the pregnancy to serve as an explanation for the production of congenital cardiac defects. When it is realized that the heart is perfectly formed, though in miniature, in seven weeks' time from the date of conception, it is obvious that there must be some error in the various examples that have been recorded of congenital heart disease following upon frights and so on during the last two or three months

of pregnancy; these psychical disturbances cannot possibly have had any effect upon the fetal heart.

Several examples of fetal endocarditis have been shown to the Society for the Study of Disease in Children, or have been spoken of at its meetings. These include 6 cases, while a few others have been recorded by Planchu, Gardère, and André Moussous. Fetal heart murmurs have been recorded by various writers.

In the production, therefore, of congenital affections of the heart two distinct processes can be seen in action—namely, developmental disturbance and inflammation. The one can operate without the other, and both are frequently combined.

It is only in a very small proportion of the cases of congenital defect that endocarditis, when it is limited to the valves, can be viewed as the cause of the disease. The bulk of the cases of congenital malformation of the heart appear to be due to developmental errors when seen by the light of modern research on cardiac development.

Morbus coeruleus and congenital malformations of the heart have been looked upon as synonymous, but without reason, for children with severely congenital *morbus cordis* are frequently not blue, even when excited or crying. Many verified cases of congenital *morbus cordis* have been recorded where cyanosis was not a symptom during life.

Some children with congenital *morbus cordis* are fresh-coloured. Others are strikingly pale. In some the lips are dark crimson, perhaps inclined to blueness rather than redness, or to turn lilac on crying. Or the complexion may be quite good and only the fingers dusky and cold-looking. The majority, however, are cyanosed more or less, and all gradations of cyanosis may be noticed, from that of a dark rubicund face with similar coloured ears and reddened finger and toe tips, or a state of rather dusky-blue lips and cheeks and tongue, and a similar condition of the extremities, to one where the face is plum-coloured, the conjunctivæ suffused, the buccal mucous membrane and tongue the colour of slate, and the body generally dusky. In the majority of those who are blue the cyanosis dates from birth, but in others the children show no signs of blueness until after an attack of one of the exanthems, or following bronchitis or bronchopneumonia, with or without whooping-cough.

By far the most common malformations of the heart in association with cyanosis are those cases where the pulmonary artery or the conus are either absent, rudimentary, or constricted. Next in frequency in the list of malformations, though only to about one-seventh of their number, is transposition of the pulmonary artery and aorta. Apart from these maldevelopments, which form the bulk of congenital cardiac anomalies, the malformations which give rise to cyanosis are numerous and various.

Various views have been and are still held as to the causation of the cyanosis. In many cases the explanation is invited, not that the lungs cannot oxygenate properly, but that they are prevented from dealing with a sufficient proportion of the total quantity of venous blood throughout the body by reason of various defects in the pulmonary region, perhaps combined with weakness of the heart muscle.

Cyanosis, therefore, appears to depend in some cases not upon defective aëration from defects in the lungs, but to be due to the conformation of the heart, which by reason of faulty construction and physical weakness is placed at a mechanical disadvantage, and is not adapted to deliver a sufficient quantity of venous blood to the lungs to be aërated.

But this will not explain all cases, for in some the lungs are not healthy and display considerable microscopical alterations. So wide and so tortuous were the thickened lung capillaries in the case of atresia of the pulmonary artery I examined that those in the alveoli admitted six or more red corpuscles abreast. Changes such as these, permeating as they do the whole of the lung structure, are not conducive to proper aëration. It would appear, then, that diffuse and not localized structural alterations of the lung tissues are favourable to the production of cyanosis.

Here, then, are two factors in operation: the difficulty of getting the blood to the lungs to be aërated, and the difficulty of aërating the blood when it arrives there should it so happen that the lungs are not in a position

to undertake the operation by reason of their structural defects.

Clubbing of the fingers and toes in congenital heart disease, the so-called Hippocratic fingers, depends almost entirely on congestive swelling and thickening of the soft parts of the terminal phalanges. The connective tissue is increased, and the capillaries are enlarged and increased in number.

The finger-tips are broadened and the nails cyanosed, a different appearance to that shown in chronic osteoarthropathy, where the nail is domed like a watch-glass, curls over the finger-tip, and bears a striking resemblance to a parrot's beak, and is combined with osteo-periostitis of the terminal phalanx. The finger looks like a drumstick, and the nail is pink coloured.

There appears to be two varieties of clubbed fingers: in one there is osteo-periostitis of the terminal phalanges, but in the other this has not yet been demonstrated. Whether there are really two varieties of clubbed fingers or whether the conditions are different stages of the same process has not yet been settled.

Common as is this clubbing of the fingers and toes in congenital heart disease, nevertheless it is a feature of this disease which very often is wanting. It is in pulmonary atresia and stenosis that clubbing most commonly arises, and it is in these conditions that congestion is most frequently observed.

Ophthalmoscopic examination in cases of marked congestion reveals tortuous retinal blood vessels of which the veins are very large. The blood in the arteries looks decidedly venous, though they are not quite so dark-coloured as the veins. The red reflex, which is of cyanosed appearance, often appears to start from the physiological pit, and the number of vessels on the face of the disc appears to be greatly increased. In children where the cyanosis amounts to but little more than a ruddy bucolic appearance a want of the usual colour contrast between the arteries and veins is quite noticeable. The retinal blood vessels early display evidences of congestion by enlargement and tortuosity of both arteries and veins. If there be considerable cyanosis retinæ, as sometimes is the case, without the associated appearance of curly retinal vessels, such an ophthalmoscopic finding, although not proof that there is no narrowing of the pulmonary region, nevertheless suggests that the pulmonary tract is either not involved, or that right-sided muscular compensation is sufficient to overcome the obstruction.

Blood changes such as polycythaemia, macrocythaemia, and increased viscosity along with a by no means invariable increase in the haemoglobin are not uncommon. In one blue child under my care, aged 2½ years, with clubbed fingers and toes, and who had been blue since birth, there was a blood count of 7,880,000 per c.mm., and the haemoglobin was 122 per cent. That is the largest number of red cells that I have met with in congenital heart disease.

Many of the children under my care with polycythaemia have shown a deficiency in the haemoglobin rather than an increase. In a blue infant, aged 1 year, with clubbed fingers and toes and lips, and finger-tips and toe-tips of purple hue, the blood count amounted to 5,400,000 per c.mm. red, with a haemoglobin percentage of 55 only.

Congenital heart murmurs are of various characters. They may be heard as harsh, roaring, churning, growling, sawing, twanging, rushing, or rasping noises. Musical bruits are not common in my experience. If murmurs of such character be heard in a child under 3 years of age, and if they be conducted over wide areas, the presumption is strong that the affection is of congenital origin. Soft, low, very faint bruits are sometimes present. Murmurs are usually systolic, occasionally presystolic or diastolic, and there is that peculiar rumbling murmur carried through the systole and diastole, which points to a patent ductus arteriosus or a communication between the pulmonary artery and the aorta. Sometimes the bruits become inaudible shortly before death, or during a paroxysm of cyanosis, and I have noticed a loud murmur to almost disappear under the influence of an anaesthetic. A murmur may be detected at one auscultation and cannot be heard at the next, and the causes for this temporary disappearance are not quite clear. Murmurs are also influenced by posture and by respiration. In regard to conduction, in some instances systolic bruits may be

traced along the arteries over the brachials as far as the bends of the elbows, and also be heard in the thighs along the course of the femorals. They can not only be followed up the arteries of the neck, but exceptionally can be heard by applying the ear to the vertex.

In only a small proportion of the cases are murmurs absent. In my series this occurred in 7 per cent.

Thrills are not infrequent; they are usually systolic, occasionally diastolic, or even presystolic. These purring tremors may be localized to the site at which they are produced or be of wide conduction. Further, the site of greatest intensity by no means necessarily corresponds to the seat of production. A systolic thrill of greatest intensity at the second left interspace and conducted towards the corresponding clavicle is, however, pathognomonic of patent ductus arteriosus. A systolic thrill may be felt at the episternal notch over the aorta, and may be conducted into the carotids and subclavians.

Defects in the interventricular septum are the most common of all cardiac malformations. They are associated mostly with stenosis and atresia of the pulmonary artery, transposition of the great vessels, and occlusion of the tricuspid orifice. The combination of pulmonary stenosis, deviation of the aorta to the right, and patent septum ventriculorum is one of the most usual forms of congenital morbus.

The commonest situation for perforate septum ventriculorum is directly beneath the aortic valves and just in front of the undefended space. In front of it lies the fleshy part of the septum and behind it the membranous. On the right side of the heart this opening will be found beneath the septal and anterior cusps of the tricuspid valve. It is rare for a perforation to be found in the anterior fleshy part. The septum may be absent, as in the cor triloculare, or the septum may be represented by a falciform ridge springing from the lower and anterior wall of the ventricle, or irregular defects may arise in it.

Changes in the heart muscle do not necessarily follow septal defects. In some cases, however, hypertrophy and dilatation of both ventricles are seen, and this is especially likely to occur on the right side.

Defects in the septum do not always produce murmurs, but when bruits are present the following characteristics have been noticed by me in the pure cases which have come under my observation. In a child, aged 2 years, there was a loud rough systolic bruit audible all over the chest, the point of maximum intensity being over the pulmonary area. It was not audible in the big vessels of the neck; the second pulmonary sound was accentuated. In an infant, aged 8 months, the systolic bruit was heard loudest at the junction of the xiphoid with the gladiolus on the left, and it was conducted a short way into the axilla. A faint systolic bruit was heard only occasionally at the left base. It was not transmitted to the neck vessels. The second pulmonary sound was accentuated. In an infant, aged 3 months, there was a loud systolic murmur at the left base, which was traceable as far as the left mid-axillary line. The second sounds were normal, and in this infant the heart was not enlarged. In the oldest child there was a marked systolic thrill all over the cardiac area, and in the child aged 8 months a systolic thrill was occasionally felt at the left base, but this might have been due to the patent ductus arteriosus, and in the infant aged 3 months it was absent.

It is not possible to lay down hard-and-fast rules as to the area of maximum intensity of the systolic bruit occasioned by patent septum ventriculorum. It is located on the left side of the sternum on the costal cartilages or interspaces, somewhere between the second left interspace and the junction of the seventh costal cartilage with the sternum, sometimes between the nipple and the left margin of the sternum, and rarely at the apex. The systolic thrill which occasionally accompanies it is not confined to any particular cardiac area; it is sometimes to be felt in the episternal notch, sometimes at the epigastrium. An important feature in the diagnosis of patent septum ventriculorum is the detection at the apex of a healthy first cardiac sound audible through the bruit and the tactile sensation accompanying it of the mitral valvular snap. The second pulmonary sound at the left base is accentuated if the pressure of the right ventricle be increased, and exceeding purity of this sound is important and suggestive.

Defects in the Interauricular Septum.—A patent foramen ovale is so frequently found at autopsy that it can hardly be classed with other congenital abnormalities, and Fisher states that slight patency of this foramen can be found in one-quarter of adult *post-mortem* examinations. A widely open foramen, however, must be looked upon as an anomaly. It occurs either alone uncommonly, or frequently in association with other interauricular septal defects. Defects in the lower part of the interauricular septum are rare, and are explained by failure of the septum primum to descend and unite with the endocardial cushion between the ventricles.

Open foramen ovale alone and along with other interauricular septal defects occurs in association with stenosis and atresia of the pulmonary artery, transposition of the great vessels, and occlusion of the tricuspid orifice. Defects in the interauricular septum happen nearly as frequently as defects in the interventricular septum. Very occasionally they are the *sole* lesions, and very slightly exceed in number as solitary lesions the similar conditions found at the interventricular partition.

Patency of the foramen ovale and defects in the interauricular septum are often diagnosed and seldom discovered, and for the reason that they do not often give rise to characteristic bruits; indeed, murmurs are but seldom produced by these abnormalities. Gross interauricular septal defects may be carried through life without causing any inconvenience or giving rise to any suspicion as to their presence.

Septal defects become of consequence if the auricles act unequally, and if there arises an overflow of blood into the right auricle, which produces stasis in the systemic veins. A reversal of the flow into the left auricle may bring about the same conditions by throwing extra pressure on the pulmonary artery and so on the right auricle. A point of clinical interest about defects in the interauricular septum, as in the interventricular septum, is that septic and other particles may be carried from the venous system direct to the arteries of the brain and body.

Stenosis of the pulmonary region is one of the most common of all the cardiac malformations, and in relation to atresia it occurs nearly three times as frequently. In the large majority of cases of stenosis the interventricular septum is patent. When this septum is closed the foramen ovale is often widely open. Both of these fetal passages may remain open. In rare instances both the fetal passages are closed.

In atresia the interventricular septum is more frequently found closed than in stenosis.

Of 13 cases, 6 of stenosis and 7 of atresia, I found the ductus arteriosus closed in all the cases of stenosis and patent in all the cases of atresia except 1, and in that case the lungs were supplied by a large branch from a left innominate artery, and not by the ductus arteriosus as is usual. If the ductus be closed in atresia the septum will be found defective, or the septum and foramen ovale will be open, and vary rarely the foramen ovale only. When there is a defective interventricular septum the aorta, which will then be found large, is frequently transposed to the right, and this irregularity occurs more often in atresia than in stenosis. The aorta may arise from both ventricles over the defective septum, or be thrust more to the right and take origin from the right ventricle only.

The stenosis may involve the whole pulmonary region, there being a hypoplasia of the pulmonary artery and its branches. In some cases the stenosis is valvular in character. It may consist of thickened nodular valve cusps associated with a normal pulmonary artery and normal interventricular septum.

In some cases stenosis has evidently originated later in intrauterine life after the closure of the septum ventriculorum. In the other and the more numerous variety the forms that are met with suggest a developmental origin and a secondary endocarditis infection implanted on a rudimentary pulmonary tract.

In developmental cases the pulmonary artery is usually thin-walled and vein-like. In the other variety the pulmonary artery may be thin-walled also, but it is often either dilated or normal. In stenosis hypertrophy and dilatation of the right auricle and ventricle are common, and if the aorta be deviated to the right these hypertrophies always arise, and this vessel will be found large and thick-walled. The combination of pulmonary stenosis,

deviation of the aorta to the right, and patent septum ventriculorum, is the most usual of all congenital abnormalities.

In *atresia* the obliteration occurs at the conus, or the valve, or the pulmonary artery may be entirely absent.

Regarding the detection of right-sided hypertrophy of the heart in pulmonary stenosis and *atresia*, a small amount of right-sided enlargement is rather apt to be overlooked, if reliance be placed solely on the superficial area of cardiac dullness. Deep percussion should always be resorted to in addition, and it is well, if possible, to verify the result by *x-ray* examination. The lung on the right side of the heart covers a moderate enlargement of the right auricle and ventricle, and so masks the signs.

The *bruit* of uncomplicated pulmonary stenosis is systolic in time, and is best heard over the second left interspace close to the sternum or on the third costal cartilage. It is usually conducted over the left front of the chest and crosses the sternum, being heard slightly to the right. It is carried up towards the left clavicle. It usually ends before the axillary fold. There appears to be some doubt as to whether the *bruit* can be heard in the back in uncomplicated cases. It certainly can; I have *post-mortem* evidence as to that. It can be heard all over the front of the chest and all over the back, the left side more than the right, and also in the left axilla. It is not heard in the carotids.

Should pulmonary stenosis be associated with a patent septum ventriculorum, as so frequently happens, a systolic *bruit* will probably be heard in the carotids. If that be heard the suggestion is that pulmonary stenosis is combined with this septal defect, as I believe I was the first to point out some years ago. It is not necessary for the aorta to arise immediately over the septal defect to conduct a *bruit* into the neck vessels.

The prospect of life varies in these cases according to the defect. In *atresia* with a closed septum the children die in infancy. If the septum be patent the child may live a few years in place of a few months. In stenosis with a closed septum middle age may be reached, but with a patent septum adult life is the limit.

The average diameter of the ductus arteriosus *post-mortem* at birth is stated by Thérémis to be 4.8 mm., and, according to Klotz, this measurement is no indication of its functional capacity during life, for he states, on experimental evidence, that the size of this vessel when in action is equal to the capacity of the pulmonary artery itself.

Patency of the ductus arteriosus as an isolated lesion is rare, very few cases being recorded in the literature, but as an associate of other congenital abnormalities of the heart it is quite common.

The patent duct may be cylindrical or conical, with the base of the cone on the aortic side, or globular. The lumen of the vessel in the cases that have come to autopsy has varied from a bristle to that of a finger. Hypertrophy and dilatation of the right ventricle and dilatation of the pulmonary artery commonly take place in chronic cases.

Attacks of dyspnoea, epistaxis, tachycardia, and angina have been recorded by various observers in association with this condition.

The characteristic rumbling murmur accompanying patent ductus has been verified by *post-mortem* examination on adults, but not, as far as I am aware, on children. Gibson times the murmur as commencing soon after the systole and gradually shading off in the long pause. Combined systolic and diastolic bruits have been recorded by Drasche, and a diastolic *bruit* by Fagge. Systolic bruits have been described by Murray, White, Simmons, and Bittorf.

The systolic *bruit* is heard loudest in the second or third left interspace close to the sternum. It may be heard only to a limited extent, or it may be carried all over the chest, back and front, and into the arteries of the neck, as in my verified cases. A thrill is by no means constant. When it occurs it is best felt in the second left interspace, and if the thrill be carried up to the left clavicle in the direction of the pulmonary artery, that is pathognomonic. The pulmonary second sound may be unaltered, but if the blood pressure be raised in the pulmonary artery by hypertrophy of the right ventricle or by the contribution

from the aorta, then an accentuated pulmonary second sound will be heard. If the *x-ray* picture suggests an enlargement of the pulmonary artery, that point is in favour of patent duct.

If the second pulmonic sound be loudly accentuated, there will probably also be observed visible diastolic pulsation or rebound in the second left interspace at the time of the closure of the pulmonary valves.

Bruits and physical signs cannot be expected to occur in patent ducts of small calibre. In order that a *bruit* shall be produced it is necessary that there shall be a sufficient space within the walls of the duct and pulmonary artery, in which the conflicting blood currents shall operate best to produce vibration and sound.

Systolic bruits in my experience are the usual accompaniments of patent ductus arteriosus in children, and the rumbling *bruit* occupying the systole and diastole in adults which is considered to be characteristic of that lesion is not common in them.

Tardy closure of the ductus arteriosus may well explain the disappearance of cardiac bruits in children who have been diagnosed as suffering from a congenital heart affection. Reviewing the morbid anatomy of the congenitally deformed heart, closure of this fetal channel appears to be about the only thing possible in the way of a cure open to these cases.

Coarctation of the aorta is the name which has been given to stenosis or *atresia* of that vessel, either at or below the isthmus, that part which lies between the left subclavian artery and the ductus arteriosus. Soon after birth that narrow section of the aorta which is not much used during intrauterine life is apt to be smaller than neighbouring parts of this artery, but when the circulation is reconstructed this part soon acquires its proper proportional relationship. Bonnet describes two types. In one, the *infantile*, there is diffuse narrowing of the aorta at the isthmus. In the other, the *adult* form, there is an abrupt constriction as of a cord at or near the insertion of the ductus arteriosus. The *infantile* type is developmental and may consist of a moderate stenosis only, or, owing to the complete failure of development in regard to that part of the fourth branchial arch which lies between the ductus arteriosus and the subclavian artery, the part corresponding of the aorta—the isthmus—is represented by a cord only or completely disappears. The *adult* form he considers is not of developmental origin, because it does not occur in the fetus and is not met with until the closure of the ductus arteriosus, and is brought about by the act of closure of the duct itself, elements of which are incorporated in the aorta, and which by their contraction constrict that vessel as with a ligature. In the *infantile* type of case patency of the ductus arteriosus is common, and in the *adult* type that channel is usually closed. In the *adult* type extensive collateral circulation is common according to Bonnet. In the *infantile* type associated cardiac developmental anomalies are common and often severe; in the *adult* type the anomalies are usually unimportant and rarely severe. In the *adult* type the percentage of cases without associated cardiac developmental anomalies is far greater than in the *infantile*.

From the literature it appears that patients with *coarctation* of the aorta may live long and the condition not be suspected. In some death takes place suddenly and without obvious reason, sometimes from rupture of the aorta. Or death may occur from heart failure preceded by the signs and symptoms of mitral disease.

Congenital aortic stenosis and atresia are much rarer than the corresponding defects at the pulmonary artery. Conus stenosis in this situation takes the form of a thickened ring of the endocardium, just below the aortic valves, which involves the base of the anterior mitral cusp, and is ascribed by Keith to arrest of development. The danger of this rare anomaly lies in its tendency to become the seat of endocarditis. In *aortic atresia*, brought about by such conditions as obliteration at the root or by fusion of the cusps, the left ventricle atrophies and the circulation is conducted through the foramen ovale, patent septum ventriculorum, and patent ductus arteriosus. The life of such cases has been limited to a few weeks. In *aortic valvular stenosis* there are two forms—one in which the septum ventriculorum is perfect, the other in which it is deficient. In this latter variety other developmental anomalies are the rule.

Aortic disease of the acquired type is not as rare in children as is commonly supposed, and some of the cases are no doubt of congenital origin. Another point of clinical interest, a recognition of which will prevent falling into error, is that aortic disease in children is apt to be associated with a bruit which is heard loudest over the pulmonary area, and not over the aortic cartilage.

Hypoplasia of the aorta is characterized by diminution in size of the aorta and arteries throughout the body, the walls of which are thin and elastic.

A congenital origin has been claimed for this condition, because it is so commonly associated with anomalies in the generative and circulatory systems. A congenital tendency to the dwarfing of the arterial system has been ascribed to tuberculosis in the parents, and it may be due to other states of bad health.

Hypoplasia of the aorta is associated with a dwarfed heart in some instances, and in others with an enlarged heart confined to the left side or involving both ventricles. If the heart be small during childhood it hypertrophies at puberty. The period of danger in these cases appears to belong to adolescence, when heart failure is not unlikely to occur, and this is more likely to arise under physical strain.

Defects in the auriculo-ventricular valves are very uncommon. They are commonly combined with other cardiac abnormalities, uncomplicated lesions of the mitral and tricuspid valves being exceedingly rare. Of the two valves the tricuspid is more frequently attacked than the mitral, and of the lesions tricuspid atresia is the commonest.

Tricuspid stenosis in association with disease of the pulmonary valves is occasionally seen.

Mitral atresia is very rare. Cases have been recorded by Rokitsansky, Thérémín, and by Lawrence and Nabarro, and in each instance the condition has been associated with somatic abnormalities.

A specimen of *congenital mitral stenosis* was exhibited at the society for the Study of Disease in Children by Fisher in 1901.

Of *mitral insufficiency* Cautley has afforded an example in a girl aged 22 months. The physical signs were those of mitral regurgitation, and another case in a boy, aged 10½ months, has been quoted by Maude Abbott.

Hypertrophy of the heart undoubtedly arises as an idiopathic disease of congenital origin. Simmons was the first to record a case of its existence at birth. He found the heart of a newly born child, who died during the labour, to be enlarged to twice its natural size, all other organs of the body being quite healthy. Prior to that Hauser had recorded a case in a baby, aged 11 months, in which the heart was enormously dilated, with hypertrophied right and left ventricles. Virchow suggested that the hypertrophy might be congenital.

Cardiac displacements such as *dextrocardia* and *mesocardia* are the only congenital malpositions that are of interest to the physician and therefore that need be reviewed. *Dextrocardia*, accompanied by transpositions of the viscera, is commonly viewed rather as a rare developmental freak than recognized, as should be the case, as a not very out-of-the-way clinical experience.

Dextrocardia without associated transposition of the viscera is rare, and not only is it rare, but it must be viewed with suspicion, because the diagnosis of congenital *dextrocardia* has been made on some few occasions, and this opinion has not been confirmed by autopsy.

Sometimes the position of the fetal heart in the centre of the thorax is maintained, giving rise to the condition known as *mesocardia*. The apex beat will then be felt in the epigastrium, and the position of the organ will readily be demonstrated by the *x* rays.

At the eighty-first annual meeting of the West Riding Medical Charitable Society, of which Dr. Gibson, of Harrogate, is now President, it was reported that forty-eight grants, amounting in all to £1,680, had been made during the year, and that since the foundation of the charity the total sum disbursed in grants had been £46,295 10s. Of those on the society's list of grantees four had died during the year, and five new applications had been received. Mr. Herbert Rowe, who for twenty years was co-secretary with the late Mr. Claude Wheelhouse, was unanimously elected Honorary Treasurer, and Mr. Michael Teale appointed Honorary Secretary.

THE PERONEAL TYPE OF MUSCULAR ATROPHY

WITH AN ACCOUNT OF A FAMILY GROUP OF CASES.

BY

J. RUTHERFORD HALLIDAY, M.D.,

AND

ARTHUR J. WHITING, M.D., M.R.C.P.,

PHYSICIAN TO THE PRINCE OF WALES'S GENERAL HOSPITAL, N.;
ASSISTANT PHYSICIAN TO THE MOUNT VERNON HOSPITAL
FOR CONSUMPTION AND DISEASES OF THE CHEST.

THE following cases of hereditary muscular atrophy belong to a family in which it appeared in at least four generations; the cases are examples of the still unclassified peroneal type first closely studied by Dr. Howard Tooth in this country, and by MM. Charcot and Marie in France.

The patients are a man, aged 45, and his two daughters, aged respectively 17 and 21; the remaining members of the group include the grandfather, mother, and younger brother of the present head of the household, all deceased.

CASE I (Father).

G. H. S., aged 45, married for eighteen years, has six children, three girls and three boys. One girl died in infancy of whooping-cough. The other children, the youngest of whom is 2 years of age, are living, but the two daughters are the subjects of the same condition as their father; the others are apparently sound. The patient is a cardboard box cutter, and for fourteen years held the position of foreman in the works. He is a man of more than ordinary intelligence, has been steady all his life, and, it may be taken as certain, has not suffered from any venereal disease.

Family History.—His father is still living and is 70 years of age; he is somewhat alcoholic and has been so for many years. The mother died when 50 years of age of heart disease; she suffered from clubbed feet. Such particulars as could be gathered of her case are set forth under Case V. An only and younger brother—there were no sisters—died, when 32 years of age, of pneumonia; he was similarly affected in the lower extremities, and particulars, so far as they could be obtained, are recorded under Case IV.

The following is the patient's family tree, details of the affected members of which follow:

History of Present Illness.—Up to 8 years of age the patient was quite healthy; he then had measles. Soon after recovery weakness of the feet was noticed, and he gradually came to walk on the outer edge of the feet. At 15 years of age he attended an orthopaedic hospital, and was given special boots, which were worn for two years. At 17 he went to another hospital and underwent an operation for the deformity, after which he wore iron supports for three years. He was able to walk fairly well on the level ground, but experienced a little difficulty in going upstairs. From adolescence up to the age of 32 the condition seems to have remained stationary, and he was able to attend regularly at work. Thirteen years ago, when 32, he began to "feel himself going all wrong," he became conscious of increasing weakness in the legs, and then noticed they were getting thinner, especially the left. At this time he also noticed commencing weakness in the hands, which showed itself in clumsiness of movement on attempting to pick up small articles. Gradually the backs of the hands became thin and hollows appeared between the metacarpal bones. He found difficulty in feeling small articles in the dark, so that he was often unable to take matches out of a matchbox, to pick up a coin, or to fasten his collar. About eight years ago an ulcer formed on the outer side of the right sole; this several times healed under treatment, only to break down again on using his foot. At varying intervals during the last thirteen years he has had attacks of pain down the limbs and also round the waist, and these have been more frequent of late. He was able to work up to February, 1904, when, in consequence of the extensive ulceration of the right foot, he was compelled to lay up. On recovery he found himself much weaker and unable to walk far without feeling exhausted.

State on Examination.—In 1904 the following notes were made: The patient's general condition in regard to his respiratory, circulatory, and digestive systems is good. Bladder and rectum are normal in their functions. Sight and hearing are good. The trunk muscles are well developed.