VOLUME 1.

CONGENITAL MORBUS CORDIS

WITH SPECIAL REFERENCE

TO

SEPTAL DEFECTS

AND

PULMONARY STENOSIS.

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C O N T E N T S.

INTRODUCTION.

In the choosing of a subject for the writing of a thesis, it would appear desirable firstly to select such a topic as is of interest both to the profession at large and to the writer in particular. Secondly a sufficiency of clinical or other material must be available to allow of sound conclusions being drawn, hoping therefrom to contribute some further truth, no matter how little, in the elucidation of some perplexing purpose of nature. The subject of Congenital Heart Disease has been chosen, the writer believing it to fulfil the above-mentioned conditions. It is indeed a subject of interest, certainly to the parents of the many children who are sufferers therefrom. Whether it is of equal interest to the medical profession at large is a matter for consideration. It is indeed true that Cardiology and its daughter sciences have been a matter of profound interest to the practitioners of medicine from the distant ages. The Alexandrian anatomists, Herophilus and Erasistratus, at that early date were carrying out dissections on the heart and blood vessels, and so throughout the succeeding ages, aided step by step by the work of such great men as Harvey, enquiries into the mysteries of the circulation have been answered by discoveries of greater and lesser importance; and yet, by many, the ugly sister of Congenital

Heart Disease has been relegated to the limbo of the incomprehensible and the untreatable. To Laennec, that frail and phthisical regimental surgeon of the early nineteenth century, albeit a man preeminent among clinicians of all ages, must be ascribed the title of discoverer of the clinical side of cardiology, opening up vast fields of research and understanding in the matter of theclinical interpretation of physical signs; and yet the very ease with which such sounds and rubs as he demonstrated were heard, and the fact that noise and death's approach were in so many cases coincidental, intensified the awe inspired by the audition of the bruit to the exclusion of the patient's response to the routine efforts of his daily life. Such fears were magnified a hundred-fold when inspired by the rasping bruits of the newly born, and so there arose the belief, fostered by the early demise of some few of such children that congenital morbus cordis was a lesion of the greatest rarity, and when found merited a grave prognosis, debarring the child from all the physical joys of work and play. It is because this belief is embedded fast, not only in the minds of the laity, but still more so in the minds of a number of general medical practitioners throughout the country, and because one has been fortunate enough to have access to some 42 cases of this defect, that this subject has been chosen for

this thesis, hoping to show that congenital heart defects are by no means rare, thereby meriting a fuller consideration at the hands of the examiner, and further trying to instil a more optimistic view as to the child patient's expectation, not only of life, but of physical usefulness.

P A R T 1.

NOTES ON THE DEVELOPMENT OF THE HEART.

1. Early development.
2. Segmentation of the cardiac tube.
3. Sinus venosus and auricle.
4. Bulbus cordis.
5. Histology.
6. Bulbar differentiation.
7. Foramen ovale.
8. Bundle of His.
9. The foetal circulation.

Before passing to the symptomatology, signs and differential diagnosis of Congenital Heart Disease, some little time must be spent in attempting briefly to explain the salient points in the development of the human heart, realising how impossible it would be to understand the clinical significance of, for instance, a "subaortic stenosis" without some understanding of the embryological processes whereby such a departure from the normal may be brought about.

EARLY DEVELOPMENT. Broadly speaking, the functional activity of the heart is the propulsion of blood into the vessels arising from it, but the mechanism whereby this is brought about undergoes profound alteration, the heart changing from a simple tube without valves, propelling its blood by means of purely rythmic peristaltic pulsations, into a complex, two-sided, four chambered organ, its two halves without

direct communication, and fulfilling its function by means of alternating succeeding contractions of the auricles and ventricles of the right and left halves simultaneously.

It is to attain a knowledge of the detailed occurrences taking place in the embryo heart, whereby such evolutionary changes as those outlined above, may be understood, without which an intelligent comprehension of the pathology of congenital morbus cordis would be impossible, that this chapter has been written.

The very earliest development of the human heart is unknown, but according to Mollier (95), from comparative studies in other vertebrates, the earliest signs are the appearance of a number of cells, angioblastic cells, or as Mollier describes them "Vascular" cells, on the ventral wall of the pharynx in the distal part of the head, and seen about the end of the third week of foetal life. These cells are arranged doubly, but central fusion unites them, thereby forming a single heart tube of delicate endothelial cells. These cells project laterally, bulging into the wide pleuropericardial space which at this stage is continuous with the peritoneal cavity. Only the endothelial lining of the heart arises from them, the myocardium and the pericardium arising from the visceral coelomic wall.

SEGMENTATION OF THE CARDIAC TUBE.

We have thus reached the termination of the first stage in the cardiac development, a single cylinder draining the afferent veins and continuous cranially with the truncus arteriosus and the aortic arches. There are two fixed points in this primitive tube, caudally where, according to Tandler (135), the developing septum transversum exercises a fixing action where the sinus venosus becomes embedded in the myocardium, and cranially where its terminal segment perforates the pericardium. Laubry and Pezzi (80) assign the cause of fixation to the necessarily slower growth of the afferent and efferent vessels than of the cardiac tube. Whatever may be the cause, these two points remain fixed while the remainder of the tube grows, necessitating a doubling of it upon itself. This it does, not in the one plane, but in the shape of the letter "S" or as Waterston (147) more aptly describes it, in the shape of the letter "V" with the caudal limb lying to the left and posteriorly, and the cephalic limb to the right and in front - so ∠ . The tube, while this change is occurring, loses its regular calibre, and segmentations appear, unequal and as yet incomplete, but distinct. These are from a caudal to a cephalic direction: First: the transversly widening sinus venosus lying immediately

above the septum transversum , into which the afferent veins drain. Second: - the primitive auricle, which is later in its appearance than the other "chambers" of the heart, not being marked until the fifth week, the other segments being determinate during the early part of the fourth week. The auricle lies in front of and cephalic to the sinus venosus. A narrow constriction, the anlagen of the auriculo-ventricular canal separates the primitive auricle from the third cavity, the common ventricle, folded back upon whose anterocephalic surface lies bulbus cordis or fourth chamber, which is continuous with the truncus arteriosus.

SINUS VENOSUS & AURICLE.

The development of these two chambers is closely associated, the one with the other, and they will be considered at the same time. The relationship of the two alters with the advancing development of the heart; the sinus which at one time lay caudal to the primitive auricle now ascends, coming to lie high up on its posterior wall, whilst the auricle itself rises higher, revolving round the vertical axis of the heart, and presenting over the superior border of the ventricle. By its rapid growth it dilates principally in a transverse direction, coming to occupy the whole width of heart. The sinus venosus which as described above,

drained all the systemic veins of the body, is divided into three parts, a main central chamber and two lateral extensions or horns. Into the left horn flows the oblique vein of Marshall or the left vein of Cuvier, into the right horn run the superior vena cava and the inferior vena cava, whilst into the body drain the vitelline veins. There is later an alteration in the venous arrangement, the one of importance to us being the atrophy, and final disappearance of the left vein of Cuvier, with the result that the sinus now empties into the right half of the common auricle. Around this opening, protective flaps appear which are invaginations into the auriclar cavity of the wall where the bodies of the sinus venosus and auricle meet. This was noticeable at an earlier stage as a narrow groove which deepened and thus formed these two flaps or valves, the right and left venous or sinus valves. They are of importance as not only do they act as preventors of regurgitation of blood back into the great veins, but also as anlagen of future cardiac structures. These valves meet above and below to form two fornices, a superior and an inferior, the inferior running down on the posterior wall of the auricle to fuse with thickenings which later appear on the posterior wall of the atrial canal - the posterior endocardial

cushion, while the superior fornix is continued upwards as the septum spurium. Contemporaneously with these changes a significant alteration is taking place in the relationship of the sinus and auricle. The sinus meanwhile becomes absorbed in the atrial substance, eventually showing no sign of its earlier existence except that its site may be determined by the points of entrance of the great vessels into the auricle, the superior and inferior venae cavae, and the coronary sinus.

We see now the early stage of the process whereby the auricle is eventually divided into the two non-intercommunicating compartments. As the common auricle enlarges, its ventricular exit becomes more and more buried under the growing auricle and ventricle, and at the same time passes towards the right and the bulbo-ventricular cleft. We can understand how the bulbus cordis represented by the superior limb of the V now crossing the antero-superior surface of the auricle becomes embraced by its winglike lateral expansions. The pressure of the bulbus on the auricle produces a cleft in the auricular wall and corresponding with this on its internal surface there appears a sickle-like fold of membrane, extending down the anterior and posterior walls until it reaches the atrial canal -

the septum primum of Born (12). The lower margin of the septum primum does not quite reach the level of the atrial canal and the ostium thus formed between the two auricles constitutes the foramen primum. The upper part of the septum primum now thins out and gradually disappears, leaving a gap between its free upper edge and the superior auricular wall, the foramen secundum; the septum primum thus forms a band which stretches forwards and upwards across the auricle. These changes are taking place when the foetus is no more than about 6 mm. maximum length.

In the left auricle, immediately to the left of the lower part of the interauricular septum can be seen the single opening of the pulmonary vein. This vein originally grew from the sinus venosus, but as the sinus shifted to the right its opening came to lie in the posterior left auricular wall. As the lungs enlarge, their venous system multiplies, four veins being formed which join into a common trunk, which as we have seen enters the left auricle. As the left auricle increases in extent it absorbs in its substance the stem of the pulmonary vein. Later, by the inclusion of the proximal parts of the two pulmonary trunks of each side, there appears the adult arrangement of four pulmonary openings into the left auricle. This process may fail to occur, the four

pulmonary veins thus uniting into a common stem and opening into the posterior wall of the left auricle. There now appears however, not in the line of origin of the original septum, but a little to its right, a structure about which there is controversy, viz: the septum secundum. According to Tandler (135), the septum secundum is a tissue growth de novo; in other words, a true septum. Retzer (115) first called this to account, stating that it was formed passively on account of the fixation of the conus arteriosus and the growing around it of the atria with a passive bulging inwards of the wall. Waterston (147) corroborates this, pointing out how even in the adult heart one can separate the two atria almost to the margin of the limbus fossa ovalis, merely by dividing the epicardium on the dorsal wall of the atria, along a vertical line extending between the right pulmonary veins on the one hand and the line joining the left borders of the inferior and superior venae cavae on the other, the two atria separating merely by dividing a little loose connective tissue. The septum secundum extends downwards towards, but not reaching at this stage, the superior margin of the septum primum. The two septa moreover do not grow in the same plane. Their manner of approximation will be considered more

fully later.

BULBUS CORDIS.

The relation of the bulbus cordis and the ventricle is a matter of great future interest. The folding of the bulbus on the ventricle produces a duplication of the cardiac wall where their surfaces oppose one another. This redundant tissue disappears, not through any actual degeneration of tissue, but, as is seen in all similar embryological eventualities, by a simple cessation or lagging behind of growth of the part in question, with the result that outwardly the bulbo-ventricular cleft shallows and there is thus formed a common ventricular cavity which encloses in itself the one time adjacent portion of the bulbus cordis.

Having indicated the final destiny of the bulbus cordis it seems pertinent here to digress from our description of the cardiac development. The fate of the bulbus is of the greatest importance, for according to Keith (68 - 75), it is due to an arrest in its development or in its dysgenese, that the majority of congenital defects are due, particularly those involving the pulmonary artery and valve, the position of the aorta, the conus of the right ventricle and indeed of any defect of the interventricular septum, and Greil (50), working independently of Keith came to the

same conclusions, that the upper part of the bulbus cordis becomes incorporated in the truncus arteriosus giving origin to the semilunar valves, whilst the lower part becomes invaginated in the right ventricle to form the infundibulum and contributing to the occlusion of the interventricular septum. The site of the bulbus can yet be seen in the adult human right ventricle, namely that part above the level of the tricuspid valve. The musculature of this area is quite distinct from that of the lower part of the right ventricle. In the upper portion, in the interior of the heart, the fibres are placed longitud - inally, whilst externally they have a distinctly horse-shoe shaped arrangement, extending down almost to the base of the tricuspid valve. The aorta terminates abruptly in the left ventricle, but the pulmonary artery ends in this series of horse-shoe shaped fibres, continuous with the right ventricle. This apparent continuity can be demonstrated in any heart on viewing the specimen from above. Keith (74) suggests that a portion of the bulbus may remain even in the left ventricle, persisting as a subaortic compartment though normally completely disappearing. The bulbus had and probably still has a very definite function. Keith (74) pointed out first how in fish and amphibians, where there is a single ventricle

and a distinct bulbus, the bulbus with its two layers of valves acts as a buffer, or as a gill valve in regulating the pressure at an even level in the gill circulation, no matter what the difference in pressure be between ventricular systole and diastole. The bulbus may and probably does fulfil some similar function in mammals. The pulmonary capillaries are of exceptional delicacy and would be quite unable, unaided, to withstand sudden variations in pressure such as occur in the pulmonary circulation. The necessity for and the efficacy of this mechanism is appreciated when it is remembered that the right ventricular output may rise in times of stress from three to thirty litres per hour, which must represent a very marked rise in pressure.

HISTOLOGY.

For a fuller understanding of later changes in the endocardium one must pause to examine certain changes that are taking place in the microscopical structure of the heart. It must be realised that of the chambers mentioned, e.g. ventricles and auricles, only a small portion is occupied by the actual blood carrying canals. The endothelial lined tube is surrounded by a wide space, reticulated and enclosed by the epicardium. These two layers do approximate each other however, but in different degrees in different areas of the heart; early in

the auricle, later in the ventricle, and at a much later date in the auricular canal. At certain situations the loose intervening reticulum becomes arranged into definite masses; in the auriculo-ventricular canal it becomes aggregated into an anterior and posterior thickening or cushion, whilst in the bulbus cordis the masses are termed the bulbar cushions which have been described by Born (12). The thickenings in the atrio-ventricular canal enlarge and their musculature becomes undermined by the growing in of the surrounding endothelium. This undermining alters their shape, they expanding laterally and becoming flattened, and the canal thereby assuming the shape of two letters T placed base to base. Their opposed surfaces fuse across the middle line thus forming two openings, one from either auricle into the ventricle of its own side. When the endocardial cushions fuse, the lower margin of the septum primum joins with their superior surface, thus obliterating the foramen primum. In the ventricle there is a differentiation of the epicardium as a continuous row of cells, whilst in the myocardium we find the cells arranging themselves into two layers. On the internal surface the cells, by their duplication, form elevations projecting into the cavity of the ventricle and becoming undermined by the overlying and enveloping

endothelium, as happened in the auriculo-ventricular canal, freely traverse the ventricle. This is the trabecular layer, while external to it lies the cortical layer, much thinner and not as yet showing the formation of well-formed muscle fibrils as noticed in the trabecular layer. This trabecular formation is entirely confined to the ventricle, the auricular musculature which is continuous with that of the ventricle, being similar to the ventricular cortical layer except along the line of attachment of the posterior mesocardium, where muscle cells are completely wanting - the Area Interposita of His.

BULBAR DIFFERENTIATION.

While considering the histological musculature of the heart one must pay special attention to the bulbar development where the myocardial changes which now take place are responsible for the later separation of the pulmonary artery from the aorta. These changes have been worked out by Born (12), whose classification of them is generally accepted. At six areas in bulbus, aggregations of the reticulated tissue between the endo- and the epicardium produce projections into the bulbar lumen. Of these there are six, two placed proximally A & B, and four distally, C & D and E & F. These six are arranged in pairs, each pair being diametrically opposed to the

other, but the succeeding pairs are arranged round the bulbar wall so that their union A with B, C with D and E with F forms a spiral septum aiding in the separation of the aorta from the pulmonary artery. Cranially the truncus arteriosus is divided into two by the septum aorto-pulmonale, which forms about the fifth week, as a sharp fold in the lumen of the truncus at the point of junction of the fourth and sixth aortic arches, which represent respectively the aortic and pulmonary trunks; this grows rapidly downwards. The distal swellings E & F unite with each other and also fuse with the lower margin of the septum aorto-pulmonale. More gradually the swellings A & B, situated respectively on the left of the posterior wall and on the right of the anterior truncus wall, unite across the middle line, thereby, at this stage nearly dividing the truncus arteriosus, with its incorporated distal demuscularised portion of the bulbus cordis, into two separate channels. This is later completed by the fusion of the distal swellings C & D. The origin of the aorta thus lies behind and to the right; that of the pulmonary artery in front and to the left. Resulting from the mechanism of separation of the two trunks a half of the swellings A & B lies in each channel and in the aorta swelling F, and in the pulmonary artery swelling E. From these three

swellings in each vessel are formed the three cusps of the semi-lunar valves.

There appeared at an earlier date on the interior wall of the ventricle a ridge corresponding with a sulcus seen on its exterior surface at the point of its flexion on the bulbus. This ridge increases in size forming a septum which passively extends further into the ventricular cavity, because of the evagination of the lateral halves of the common ventricle. As was seen above, the bulbar swellings A & B unite across the middle line of the bulbus, but only in their distal part, forming the bulbar septum. The interventricular and the bulbar septa approximate each other by their concave margins. The anterior extremity of the bulbar septum, uniting with the anterior prolongation of the interventricular septum, and similarly the posterior swelling uniting with the posterior endocardial cushion of the auriculo-ventricular canal, where it fuses with the posterior prolongation of the interventricular septum. There is thus left an elliptical area, filled in by membranous matter, constituting the pars membranacea septi, or as named by Peacock (110) the "undefended space"; it is near this region that the majority of interventricular septal defects occur.

The bulbar septum does not lie in the sagittal

plane. It will be remembered how swelling A lies to the left, while swelling B is to the right. The obliquity of the septum thus keeps the aorta to the right and behind, a point of importance in dealing with defects where there is transposition of the trunks or a dextroposition of the aorta. With the closure of this area, the right and left sides of the heart are finally separated, except where the foramen ovale still forms an intercommunication, but in their separation it is seen how the interauricular and interventricular septi are not in the same plane, the one deviating to the left and the other to the right. The interventricular septum comes to lie in the plane of the right tubercles of the endocardial cushions, much further to the right than the septum primum. Hence as Hochstetter (62) points out, that portion of the fused cushions between the attachments of the two septa, when it later comes to lie in the plane of the septa, does not separate ventricle from ventricle, but left ventricle from right auricle.

FORAMEN OVALE.

The foramen primum, as we have seen, is bounded above by the lower margin of the septum primum and below by the auricular-ventricular division. It closes by fusion of its inferior edge with the endocardial cushions of the atrial canal, while the

foramen secundum remains widely open, because of the poorer development of the septum secundum. The theory as regards the origin of this septum has been gone into more fully above. The septum secundum later increases considerably in extent, thereby narrowing the opening, but commensurately with this narrowing, alterations are taking place between the relative planes of direction of the two septa. The septum primum, which in the normal position of the heart was directed downwards and backwards, gradually alters its direction because of the lagging behind of the growth of its antero-superior prolongation as compared with the growth of its extremity abutting on the posterior auricular wall, which grows more rapidly upwards and forwards, thereby altering the direction of the septum from one of downwards and backwards to one of downwards and forwards. Coincidentally with this the septum secundum takes up a final position of downwards and backwards, thereby reinforcing the interauricular septum in that the posterior prolongation of the primary septum now overlaps the attachment of the secondary septum, the anterior end of which has similarly grown past the line of attachment, anteriorly of the septum primum. Eventually they close by the rising pressure in the left auricle equalling and exceeding that in the right

auricle.

Rouvière (122) points out how the septum primum and the septum secundum in their posterior and superior parts, are in the same plane sagittally but anteriorly they form a wide angle between their respective planes, and thus form a channel in an upwards and leftwards direction, which aided by the right venous valve helps to direct the blood entering from the inferior vena cava through to the left auricle.

BUNDLE OF HIS.

The question of the cardiac arrythmias does not affect to any extent the student of congenital morbus cordis in so far as it might be feared that any defect in the interventricular septum might cause an interruption of the conducting path. This we know not commonly to be the case, but the subject is of such undoubted cardiological importance that an account of the embryological development of the heart without mention of the origin and growth of the conducting bundles would be incomplete. Tandler (135) describes the earliest sign as seen in his specimens of 19.75mm. and 28.5mm. as a triangular area of peculiar colour situated on the upper margin of the interventricular septum on the lower surface of the as yet not completely differentiated endothelial cushion. He holds that it arises de novo and not as

a relic of previous embryonic tissue. Retzer (115), from investigations on pig's embryo believed it to have its origin in the left venous valve. Keith (67) is of the opinion that the system represents a persistent connection between the auricle and ventricle, in which a reduction and concentration has taken place in the course of phylogenesis. Mall (90), likewise from recent researches considers the A.V. bundle to represent the remains of the original auricular canal, and Mönckeberg (96) shows the correctness of Mall's work, namely that it is a relic of the auricular canal musculature.

THE FOETAL CIRCULATION.

We have now reached a stage where the heart is divided into two distinct non-intercommunicating halves, each with its own atrium and ventricle, and except for the foramen ovale, which remains patent till birth, in every way a model of the adult heart. It is a matter of importance and of wonder to note that these changes are complete by the eighth week of foetal life.

We can now review the path of the foetal circulation. The foetal blood arrives at the placenta through the umbilical artery, and there it is purified and returns to the foetus, principally via the inferior vena cava to the right auricle. Thence,

without passing into the right ventricle, and deflected in the right auricle by the larger right venous valve, which is bigger than the left, and by the septal arrangement as suggested by Rouviere (122) above, the major portion flows into the left auricle through the foramen of Botal, thence via the left ventricle into the aorta. Such blood as supplies the head and upper limbs returns to the right auricle by the superior vena cava, but deflected by the tubercle of Lower, it only feebly mixes with the stream returning from the inferior vena cava, and passes into the right ventricle, and so to the pulmonary artery. The greater part of the pulmonary content is directed through the ductus arteriosus into the aorta, of whose content a considerable proportion returns to the placenta, and so the cycle recommences.

LITERATURE.

It was not until 1749 that there was written the first treatise on this disease, by Senac (128). This was followed by publications by William Hunter (63) in 1784, Meckel (93 & 94) in 1802, Louis (84 & 85) in 1826, Farre (43) in 1814 - often quoted by Peacock - Paget (107) in 1831, Gintrac (48) in 1824, and Chevers (21) in a series of articles appearing in the London Medical Gazette from 1845 to 1851, and the father of all modern work on the subject, Rokitansky (118), a pupil of Virchow's, and in the eyes of many a greater clinician than his mentor, who in 1875 contributed his Defekte der Scheidewände des Herzens. During the latter half of the last century, the most comprehensive review on Congenital Heart Disease was made by Peacock (110) whose "Malformations of the Human Heart" with its exhaustive bibliography is still regarded as a classic. During the present century, four works stand out pre-eminent, that by Maude Abbott (1), with a detailed survey of 850 cases of Congenital Morbus Cordis; the brilliant clinico-pathological treatise by Laubry & Pezzi (80), numerous contributions by Keith (68 - 74) from an etiological point of view, founded on exhaustive researches into comparative embryology, and the monograph by Abbott & Weiss (2) in Blumer's "Bedside Diagnosis"; no vast

literature when one realises the frequency of incidence of the condition.

CLINICAL INTRODUCTION.

In a consideration of the clinical features of Congenital Morbus Cordis, it is essential to realise that even more so than in other pathological states, the expected may not happen. Again and again this will be stressed, and illustrated throughout the text, and in a review of the cases selected here, it will be seen how capricious are the physical signs. At one end of the scale one meets with cases with bruits, thrills &c. out of all proportion to the anatomical changes present, while at the other end one meets with the individual whose auscultatory phenomena are in no way consistent with the gross lesion involved. As a general principle in the study of Congenital Cardiac Disease, the case with few auscultatory signs is infinitely more serious than that in which they are rampant. Especially is this so when cyanosis is present. The following example will be referred to again, but illustration of this point may be made here.

In case No. 15 (Wright), were it not for her cyanosis and her radiological signs, she would reveal to the listening ear nothing other than a

soft bruit which by reason of its comparative silence, and its position might easily be disregarded. Further illustration on this point is given lower, namely the case cited by Muir (personal communication). This child, though deeply cyanosed showed no other clinical sign whatsoever of cardiac defect. At autopsy, however, an extreme degree of Fallot's Tetralogy was found; the specimen is now in the Middlesex Hospital Museum.

MATERIAL & METHODS OF INVESTIGATION USED.

The reasons for the choosing of this subject for the presentation of a thesis have been typified at the beginning of this paper, and the opportunity of studying Congenital Heart Disease has been greatly facilitated by the courtesy of Dr. D. C. Muir, in permitting the examination of cases presenting themselves at the Special Rheumatic Clinic, which is an integral part of the Hull School Medical Services, and to which he is examining physician. At that clinic 37 cases of Congenital Morbus Cordis have been diagnosed clinically, which together with 5 from the writer's own practice forms a series of 42. Of these, 29 showed evidence of primary or secondary septal defect. From that number, 15 have been selected as exemplifying typical examples of the lesion in question, or illustrating various departures from the usual conception of the condition. A classification of the selected cases will be found in PART 3.

Examinations have in the major number been confined to clinical investigation, corroborative mechanical evidence, apart from radiology, blood cytology and two electrocardiographs, being unavailable. Even as regards radiology, one has been unable to complete the series, because examination is not

compulsory, and parents through fear or suspicion of interference or experimentation in what they realise is an incurable ailment, have in many cases refused to allow or given permission for only a restricted radiological investigation.

The X-ray tracings etc. have been taken from films taken at 6 ft. and the advantage of orthodiagraphy over such a method is more apparent than real.

As regards final accuracy, the method here adopted whereby the tracing is taken over the viewing box, is satisfactory, though one must make allowance for an error of about 5%. On the other hand, screening must be done at a short distance from the stationary tube, which undoubtedly causes angulation of the shadows and from that point of investigation, - for instance, as regards the effect on the lower borders of the ventricles, - one loses a certain amount of accuracy. This drawback was later rectified by screening at 6 ft. with an increased milliamperage, when detail could be seen in its proper perspective, and so prevented alteration in position in the taking up of a new stance for the taking of the film. The exposures varied from two-tenths to eight-tenths of a second. In the early stages barium was not used as an adjunct to radiological examination. It is difficult to practice such a method of determining

an aortic displacement or an enlargement of the pulmonary artery in a young child, restless and nervous of a strange apparatus.

CLASSIFICATION.

Though one is not attempting to emulate the prodigious treatise by Maude Abbott (1), who described fully 64 different varieties of Congenital Morbus Cordis, one must give some views on the classification of such defects in general, and in particular on those under consideration here.

It has long been known how abnormalities in the foetal heart tend to associate themselves in many instances into certain groups of lesions. Others we know are found uncomplicated, e.g. we will note how patency of Botal's foramen is in the majority of instances unattended by associated defect in the heart; similarly with Maladie de Roger, and also some cases of Pulmonary Stenosis. On the other hand we know of certain groups of defect, e.g.

First: That anomaly commonly associated with the name of Fallot (41 & 42), in which there is found narrowing of the pulmonary artery, either atresic or stenotic, perforation of the interventricular septum, deviation to the right of the aorta, and hypertrophy of the

right ventricle.

Second: Atresias of the auriculo-ventricular valves are associated with an atrophy of the corresponding ventricle, and hypertrophy of the homolateral auricle and perforation of the interauricular septum, and patency of the ductus arteriosus.

Third: Coarctation of the aorta, and atresia of the pulmonary artery, are usually associated with persistence of the ductus arteriosus.

Fourth: Patency of the interauricular septum, if large, is associated with enlargement of the pulmonary artery and narrowness of the aortic shadow and "petitness" of build.

These are the four classical examples of the association of congenital cardiac anomalies, and with any one of them may be expected one or more of the same group. But there are numerous instances where the defects present do not associate themselves according to such a plan, and exact clinical diagnosis is not possible, allowing one only to determine such lesion as is exercising the major effect objectively.

Maude Abbott (1) divided congenital cardiac defects into those giving rise to cyanosis, and those not; Laubry and Pezzi (80) into those affecting the right and those affecting the left heart. Abbott's classification is from a clinical standpoint undoubtedly

superior, for the dividing line between health and morbidity in these children is the presence or absence of cyanosis. There are exceptions to this rule; for example with congenital mitral stenosis with patency of the foramen ovale, cyanosis is by no means always present, and yet the degree of morbidity is great. No classification on a clinical basis however, can hold good in all instances, but the division into the cyanotic (excluding those cases which later develop a cyanose tardive) and the non-cyanotic varieties, is without doubt the most useful.

INCIDENCE.

The comparative paucity of the literature on what is a subject of vast interest, indicates how rare the condition is supposed to be; that is by no means true. Botal (13) met with a defect of the inter-auricular septum so frequently at autopsy in persons who had been otherwise cardiologically healthy that he regarded it as a normal condition. Da Costa - Alvarenga (27), on the evidence of 213 autopsies on children of 2 years and under, placed the percentage of incidence of interauricular defects as high as 96.24%. Maude Abbott (1) quotes Zahn as having found the foramen of Botal open in 129 cases out of 711 examinations at autopsy, a percentage of 19, and Adami of Montreal placed its occurrence as high as 14.5%. One cannot insist on the Hull figures as being an accurate indication of the incidence of congenital cardiac disease for two reasons: Firstly, the diagnosis has not been corroborated by post mortem examination, which is essential where accuracy is desired, and secondly, because these cases were drawn from such children as were examined by the visiting School Medical Officers as showing some cardiac murmur, or other similar supposed departure from the normal, and were referred for cardiological examination during the first year of the Clinic, thereby representing a higher rate

than will appear from expert opinion during a normal year. Yet it is of undoubted interest to know that of 47,395 children in Hull, between the ages of 5 and 14, fit and able to attend the elementary schools, between the dates March 1930 and July 1931, 37 were found to be suffering from congenital heart disease.

ETIOLOGY.

The question of the etiology of congenital morbus cordis is one of the most vexed in the whole range of medicine. Participation in the controversy scarcely falls within the range of a clinical thesis on congenital heart disease and yet this paper would be incomplete without the exposition of the writer's views on the origin of such defects as are described above.

The conceptions commonly held may be broadly illustrated thus. That congenital defects of the heart have their origin in :-

1. An endocarditis acquired during gestation and carried to the foetal heart from the placenta.
2. An arrest or faulty progress of development.
3. An endocarditis implanted on an already developmentally deformed valve or orifice.
4. A combination of 1 & 2, a primary endocarditis so interfering mechanically with the free circulation of the foetal blood as to give rise to other anomalies, chiefly of septal nature.

Amongst the most inveterate adherents of the first line of thought may be included Morgagni (97), Peacock (110), Lancereaux (79) and Letulle (82 & 83), all of whom stressed the consistency of foetal endocarditis as the origin of such defects. The tendency of later writers has been to assign to some defects

an inflammatory etiology, whilst to other a purely developmental origin. Senac (129) held that their origin was accidental, "Des farces de la nature formatrice", or of embryological inception. To this view Laubry and Pezzi (80) hold. Rösler (121), while admitting that foetal endocarditis is not a rarity, yet maintains that there is always a pre-existing developmental error.

The several factors which might have some bearing on their etiology may usefully be reviewed, to enable one to arrive at a reasoned conclusion on this problem.

1. PARENTAL TRANSMISSION.

a. FAMILIAL TENDENCY TO CONGENITAL CARDIAC DISEASE

Where all other possible factors are excluded, the cases reported as falling within this group are extremely rare. One has, however, recently met with an instance of two brothers with radiologically proved dextrocardia and complete transposition of the viscera. Other writers instance similar cases; amongst others, De la Camp (29) of two sisters and four brothers in one family with congenital heart disease, Hess and Pearce (58) of three children of one family with cyanosis. Variot and Cailliau (140) cite a case of a child, born at eight and a half months, with a loud holosystolic murmur (heard at birth) audible over the precordium and transmitted out to the left

axilla and under the left clavicle, whose mother at the age of thirty showed all the classical signs of Maladie de Roger. She had, however, a positive Wasserman, which unfortunately brings into the question a second factor. In the case of the two boys, mentioned above, it does not appear credible that infection, whether spirochaetal or streptococceal, could give rise to such a defect.

b. FAMILIAL HISTORY OF HEART DISEASE. Histories, as elicited from the parents, especially with regard to other members of the family, are unreliable; cases of "weak heart" etc. are instanced as evidence of organic cardiac disease, which may or may not be so. In the present selected series a history of cardiac disease in the first or second family rank previous to that of the patient occurred twice, but from this small number no conclusions can be drawn. In the total series of 42 cases of congenital heart disease examined a positive history of "heart disease" was given 5 times, or a percentage of 10.9. When the uncertainty of the history is appreciated and the incidence of morbus cordis in the population is realised, -the death rate from heart disease in England & Wales in the age group 10 - 15 amounting to 10.8% of the total death rate (45)- it is seen that the incidence among the families of these children is in no way abnormal.

c. FAMILIAL HISTORY OF SYPHILIS. In no instance was it possible to establish a diagnosis of parental syphilis, questioning or examination on this point being impracticable. Statements in the literature on this point are not consistent. Rösler (121), in a series of 60 cases, found hereditary syphilis and congenital heart disease coexistent in only one instance. Irvine-Jones (66) in 4 out of 100, de Stefano (30) on the other hand cites 72% as the incidence of congenital syphilis among 32 cases of congenital heart disease investigated by him. Hochsigner (59), examining 500 cases found signs of syphilis in 7. Areas of degeneration, frequently myxomatous, may be seen in the path of the coronary circulation, and the spirochaeta pallida has been isolated from such areas, more especially in the region of atresic orifices. There are no records to show whether these syphilitic changes are commoner among children with congenital heart disease than among others of the hospital class from which such children were drawn.

d. FAMILIAL HISTORY OF ALCOHOLISM. It is difficult to determine what constitutes alcoholism, the normal consumption varying between individuals and also between different districts and different countries, but it is possible that a substance of

such known toxic powers may, if consumed in pathological quantities, so poison the "germ" as to abort or interfere materially with the normal course of development, especially of the heart which is the only organ which has a continuous labour to perform during its developing period.

e. CONSANGUINITY. In the present series the parents were questioned with regard to consanguinity of themselves or their parents. In no instance was a positive answer given. Rösler (121) found evidence of consanguinity in 6 cases in his series of 60. These were :-

1. Mother married maternal uncle.
2. Father is cousin of maternal grandmother.
3. Paternal great grandfathers were brothers.
4. Paternal grand parents were cousins.
5. Paternal grand parents were cousins.
   Paternal great grand parents were cousins.
6. Parents were cousins.
   Maternal grand parents were cousins.

Cassell (20) investigating 25 cases from this point of view found all with negative results. Egger (36), from 12 patients with congenital heart disease elicited positive answers - all distant relationships - in three.

To be of value, large series of cases must be investigated. Rösler's incidence is high, viz 10%, but the normal incidence of consanguinity, according

to Mayet (92) is :

France	10.45%
Italy	5.18%
Prussia	6.47%
Bavaria	6.53%

showing no pertinent variation from Rösler's figures.

2. CO-EXISTENCE OF VISCERAL AND CARDIAC DEFECTS.

These are commonly associated; there is no suggestion that hare lip, spina bifida, the hernias, undescent of the testicle, horse shoe kidney, etc. are inflammatory in their inception, and it appears inconsistent to suggest that similar cardiac anomalies have a different origin, particularly when they are so commonly associated with such defects. Rokitansky (118), among 24 complicated septal defects, all evidently of developmental origin, found associated visceral anomalies in 8 or one-third of his cases. Vierordt (143) found associated visceral defects in 11%.

One may mention here the high incidence of congenital cardiac defects in Mongols. Morse (98), in a report on 114 cases found Mongolism to be present in 6 individuals.

Certain cardiac anomalies, moreover, can in no way be considered other than developmental in origin, e.g. dextrocardia, persistence of the truncus arteriosus etc.

3. APPEARANCE OF THE LESION.

The appearance of the orifice in septal defects

is characteristic. It is round or oval, smooth and with a thin edge, without any tendency towards stenosis. Vegetations are commonly found around the ventricular perforations (rarely involving the foramen ovale) and can frequently be seen on the adjacent valve cusp or on the opposite right ventricular wall. Such vegetations are admittedly secondary implantations. With the septal patencies there is a complete unanimity of opinion as to their developmental origin. With the pulmonary lesions there is more diversity of views. Where the stenosis is valvular in character, as in the cases reported on page 104 , with a thickening, shortening and fusion of the cusps, there is every evidence to show that the lesion has originated in an inflammatory process subsequent to the date of completion of the ventricular septum. Conversely with certain other types of abnormality there can be no doubt as to their developmental inception. This latter class is typified in such cases as show thickened, fused and frequently bicuspid valves of rudimentary type. At times one finds a thin diaphragm with delicate raphe stretching across the pulmonary orifice and showing no sign of inflammatory change. Where the conus of the right ventricle persists as a separate chamber, often communicating with the sinus of the ventricle by a small pin-hole opening and surmounted by healthy valve cusps one is

dealing with a developmental defect. Not so simple in their differentiation are such cases as show similar anatomical changes but encrusted with a mass of vegetations. Such are, however, probably primarily of developmental origin with a secondary endocarditic implantation.

4. TIME FACTOR IN FOETAL ENDOCARDITIS.

The most unanswerable argument against the bacterial origin of congenital pulmonary obstruction lies in the apparent occurrence of the majority of such cases prior to the date of closure of the interventricular septum. Rheumatism will produce a more rapid valvular stenosis than any other infection, and it is the writer's experience that stenosis sufficient to produce obstruction to the free flow of blood through the valve does not develop under an average time of two years, or at a very earliest abnormal minimum, six months. The interventricular septum is entire by the eighth week of foetal life. In what manner is it possible to produce stenosis in these few weeks, of such a grade as to cause a persistence of communication between the two ventricles? In Abbott's series (c.f. p.110), out of 96 cases of pulmonary stenosis there were 7 with both septa closed, 16 with the auricular septum open and the ventricular septum closed, as opposed to 73 where the ventricular septum was open. Why with 40 weeks of gestation,

during which time the foetus is liable to infection, must over 76% occur during the first eight weeks, even supposing the infection and subsequent stenosis were of remarkable rapidity of progression? These figures scarcely appear consistent with the argument of some that the major number of such defects are inflammatory in their inception. Even were one to combine under the heading of pulmonary obstruction all cases of stenosis and atresia analysed by Abbott, it is found that only 30 or 30.92% of the total number are unaccompanied by patency of the ventricular septum, the date of whose closure corresponds to 20% of the gestation period.

6. VALVE COMMONLY AFFECTED.

Opponents of the embryological theory all stress the fact that the majority of the congenital cardiac anomalies affect the right side of the heart. This on superficial examination is quite consistent with placental infection. The major part of the blood however, entering from the inferior vena cava is propelled direct across the right into the left auricle, through the foramen ovale. Such blood as enters the right ventricle comes mostly from the head and neck via the superior vena cava. Why then do the aortic and mitral valves escape to such an extent as they do? In what manner is the foetal circulation fundamentally so different from that of the adult, that

whereas in the latter the mitral valve succumbs in 90% of cases of rheumatic valvulitis, in the foetus it escapes in an even higher percentage at the expense of the pulmonary valve?

That rheumatic or streptococcal valvular infection may occur in the foetus is not denied but it is felt that its incidence represents only a small proportion of congenital cardiac defects, the majority being of developmental origin; especially is this true of septal defects. With the atresias and with the valvular stenoses, syphilis undoubtedly may play a part; a syphilitic infection occurring in the earliest days of gestation may well result in complete atresia of one or other of the arterial trunks, if the valve is rendered imperforate, with resultant septal patency. That such an eventuality can take place we know; the incidence of its occurrence however, will only be appreciated after the microscopical investigation of a larger series than is at present available.

PHYSIQUE. HEIGHT & WEIGHT FACTORS.

On surveying the heights and weights, and notes on the general physique of these children, one cannot but be impressed by the fact that in many cases they are up to and even above the normal standard for their age. It was decided to compare their physical development with a consecutive series of children suffering from other forms of cardiac disease, e.g. mitral stenosis, and a consecutive number of normal children.

Of 38 cases of congenital morbus cordis investigated from this point of view

14 are over height & over weight
3 are over " " under "
5 are under " " over "
12 are under " " under "
1 is under " " average "
3 are average " " over "

The mean height and weight figures are extracted from the Annual Report of the Chief Medical Officer of the Board of Education, 1927. Of the 38 children, therefore,

22 are over weight
1 is average weight
15 are under weight

Of the 15 who are under weight, 6 are included in the cyanotic type.

For this comparison 22 consecutive cases of mitral stenosis and 25 normal children were selected. Their heights and weights were determined and the

average difference between their heights and the standard measurements was recorded. It was found that

Congenital heart disease	-	average difference height from normal	= — .41"
Normals	-	-do- -do-	= — .49"
Mitral stenosis	-	-do- -do-	= —1.03"

The respective tables were not recorded for weight as it was found that to come to a reliable conclusion very much larger numbers would have to be investigated. One unnaturally heavy child will upset the statistical record more so than in the case of theheight investigations where variations from the mean are not so marked.

As a diagnostic point, therefore, height and weight cannot be considered as having any direct bearing, except in so far as to disprove the prefixed idea of many that Congenital Morbus Cordis necessarily presupposes a child of under average physical development; nor is there any marked lowering of the intelligence standard. It is admitted that such individuals as suffer from the more severe forms of congenital heart disease, particularly when accompanied by cyanosis of a persistent or an intermittent nature, are peculiarly liable to recurrent attacks of bronchitis, especially in an area of low lying and humid atmosphere such as Hull. Again such patients number among themselves a normal proportion of sufferers from overgrowth

of adenoid tissue, and unlike their more normal brothers and sister s are refused operative relief therefrom, so that they are all the more subject to the ravages of bronchitis, with its absenteeism from school and attendant physical deterioration.

Reference will be made later to the effect of patency of the interauricular septum on physique and mental development. Though Weiss (148) and Morse (98) hold that such children are definitely below the normal standards of physique and intelligence, it is felt that their conclusions should be more restricted to those patients with a high degree of right left shunt. Such individuals as fall within the cyanotic class, do most certainly show a falling off below the normal weight and even height levels, but with the majority, and certainly those included in the non-cyanotic class, the standard of intelligence and physique is equal to and frequently above the normal levels.

COMPLAINT.

A careful review of the cases under consideration will reveal the fact that commonly, apart from the cyanotic group, together with those suffering from the grosser defects, the patient complains of nothing. Complaint when present on the part of the patient is commonly confined to older children whose cardiac abnormality has been discovered at an early date and who have been "coddled" at home and shielded from all physical hardships. It is frequently believed by parents, having been advised of the gravity of the condition, that the child may live with the greatest care to the age of seven, beyond which age its life is to be a burden and a precarious event. Seven years of anxiety, together with the attendant lavishing of maternal love on the child whose life is to be cut short, cannot but produce a life full of "Complaint". Such a circumstance is no exaggeration but a factor constantly being met with in the investigation of such children. When cyanosis is absent, and frequently when it is present in a mild degree, and when the parental outlook is healthy, the child's only complaint is because of restrictions in playing games and other exercises imposed by relations or the family doctor.

CYANOSIS.

This feature looms large amongst the most arresting signs of Congenital Heart Disease, because together with clubbing and dyspnoea, it is the only objective external sign of cardiac trouble. Maude Abbott (1) attaches so much importance to its presence that she, in her classification of Congenital Morbus Cordis differentiates her cases as cyanotic or not.

From the earliest days of the investigation of congenital cardiac disease, attempts have been made to account for its presence, and up till the last decade no one explanation has been able to account for it in each and every variety of disease in which it was present. In view of the importance of the question and the doubt which even now surrounds its etiology, it were well to survey the theories advanced for its causation.

1. VENOUS STASIS. This, though first advanced by Morgagni, might be termed the theory according to the English School, having as its sponsor, Peacock himself. Their view was that the obstruction to the circulation, especially at the pulmonary artery, produced a stasis of the peripheral circulation, which in the presence of increased oxygen consumption occurring there, gave rise to an alteration of the blood to a venous type, with resultant cyanosis.

Until in 1923 Lundsgraad and Von Slyke (86) published what is the most comprehensive survey of the subject, with reasonings which have shed light on the whole matter, ~~and~~ placing its etiology on a sure foundation, cases were reported with varying degrees of pulmonary obstruction, in whom cyanosis was absent, whilst with others cyanosis was present, without the necessary obstruction. Lafitte (78) in 1892 instanced a case of a woman, dying at the age of 21 of malignant endocarditis, never having shown any evidence of cyanosis, in whom a "closely constricting fibrous annular ring" was found one inch below the pulmonary valve. Bouillaud (14) in 1830 reported a case of Burnets,where pulmonary obstruction in the absence of other cardiac anomaly was unaccompanied by cyanosis. Perusal of their reports, however, gives one the impression that the degree of obstruction may have been over-estimated, cases of pulmonary and "sub-pulmonary" stenosis without septal leak or patency of the ductus arteriosis, compatible with life, being mostly of low grade. Ogle (104) and others have reported cases of pulmonary stenosis with slight or absent cyanosis. Peacock (111) himself admitted an observation on a patient with "extreme narrowing of the pulmonary artery" with all the accepted essentials of the Tetralogy of Fallot in whom cyanosis

was absent until three months after birth. In defence of his arguments, however, he explained this and such cases as have been quoted above, by stating that the resultant hypertrophy of the right ventricle was sufficient to overcome the obstruction.

2. ADMIXTURE OF TWO BLOODS. Waging war most fiercely with the exponents of this theory, that obstruction was the deciding factor, were those who held that the admixture of the bloods of the two circulations gave rise to the discolouration. Senac (129) first sponsored this belief, and was later supported by Gintrac (48), Bouillaud (14), Forget (44), Farre (43), and Walshe (146) and many others, among whom are included Laubry and Pezzi at the present time. Cyanosis and septal defects are certainly associated, Peacock (110) admitting that out of 124 cases which he investigated with varying degrees of cyanosis from all cases, septal defects were found in 112. Just as with other conceptions of the cause of the cyanosis, cases were reported in great numbers of undoubted and even extreme admixture of the two circulations which showed no cyanosis. Most famous is that reported by Breschet (16) and later shown at the Societié Anatomie by Martin (91), where the subclavian artery arose from the left pulmonary trunk without any discolouration of the left arm.

Forster (45) in the first volume of the Transactions of the Pathological Society reported a case of a child, with one auricle and one ventricle, from the anterior surface of which rose the aorta, from whose posterior surface there arose two pulmonary arteries, who lived for 78 hours without cyanosis, though "cold and livid". Young (151) exhibited a specimen at Manchester of Cor Biatriatrum Trioculare with transposition of the pulmonary and aortic trunks, both of whom arose from the common ventricle, from a man aged 36, who had shown no sign of cyanosis until within three years of his death. As Stille (132) explained, there is no just proportion between the intensity of the cyanosis and the amount of venous blood which enters the arterial system; that complete admixture can take place without cyanosis necessarily being present. Roger (117) himself in his classical paper recognised this fact, and insisted that pulmonary obstruction must be superadded to entail cyanosis, uncomplicated defect in the septum being in itself insufficient to produce discolouration. This was elaborated further at a later date by Bard and Curtillet (10) who held that it might occur as a terminal event, that an intercurrent or terminal pulmonary lesion might so raise the pressure of the lesser circulation, as to establish a right to left shunt, the same reasoning applying not only in minor pure septal defects but also in those other graver

cardiac anomalies - pulmonary stenosis with associated septal leak, bi- and tri-locular hearts, transposition of the great trunks &c. in which cyanosis is a persistent and permanent feature. This fact had been realised by Peacock (112) as early as 1859, and it served to harmonise the divergent views of the English and French schools.

Such, then, were the only two theories meriting serious consideration. We know now how in part each was right. The essential feature, however, in cyanosis is not the degree of oxygen saturation, but the amount of reduced haemglobin present in the blood. There is a level above which any increase in the reduced haemoglobin present will cause visible cyanosis, and that level represents 6 grams reduced haemoglobin per cent, or 6.7 c.cms. oxygen unsaturation, and has been aptly named by Lundsgraad and Von Slyke (86) the threshold level.

It is clear therefore that the essential agencies are those which bring about an increase in the reduced haemoglobin in the blood, and as such have been called the determining factors, and will be discussed below. Individual peculiarities will however alter this arbitarily chosen threshold level, e.g. variations in the thickness or colouration of the skin, under whose influence cyanosis will be evident at a lower or higher percentage of reduced haemoglobin in one

patient than in another. These have been termed "modifying factors" each having at one time or another been thought to be an essential cause of cyanosis. There are otherfactors which can modify the threshold level, such as a variation in the number, width or length of the capillaries, which occurs in such pathological states as anoxaemia, toxaemia and venous stasis, when more capillaries remain open and dilatation of their lumen results; the threshold level can be further modified by variations in the colour of the plasma, as in lipaemia etc.

DETERMINING FACTORS.

1. In which all the blood passes through the lungs but where there is lessened or retarded oxygenation in the pulmonary alveoli :-

This is met with at rarified atmospheres where the oxygen tension is low, and in the presence of insufficient pulmonary ventilation, such as pneumoniosis, shallow breathing etc. This factor is of less importance in congenital heart disease, where administration of oxygen, which would remove cyanosis when exclusively due to this factor, is of less help. It must be realised, however, that this factor may be additionally present in the cyanosis of congenital heart disease. Cases of pulmonary stenosis are especially liable to bronchitis and the transient cyanosis seen

then is due entirely to this cause. If cyanosis were already present, such exaggeration of it as occurred would be due to this factor. Such individuals, under these circumstances, are much relieved by the administration of oxygen. The importance of this point is illustrated by a case reported by Campbell, Hunt and Poulton (19) of a man suffering from congenital heart disease with a red cell count of 9,300,000, whose oxygen saturation was raised by the administration of oxygen from 68.7% to 91.1% - a very considerable aid.

2. The presence of a right-left shunt :-

We are here reverting to the earliest theories of Senac, and the French, of the admixture of the two bloods; undoubtedly a primary cause of cyanosis in congenital heart disease. In the presence of such a shunt cyanosis may be absent, provided the shunt is sufficiently small as to allow of the reduced haemoglobin present remaining below the threshold level. In the majority of patencies of the ventricular septum, the aperture is small and the quantity of blood forced through into the other ventricle must represent an infinitesimal proportion of the whole volume of the blood, which has been estimated to be about 5-5½ litres, or $\frac{1}{15}$ to $\frac{1}{30}$ total body weight. The administration of oxygen has little or no influence on such cyanosis as is caused by this factor.

3. Increased reduction of oxygen in the tissue capillaries :-

This can be brought about by increase in the venous pressure retarding the rate of blood flow through the capillaries; a condition analogous to pulmonary stenosis. It has been shown by Barcroft (8 - 10) and others, how in cardiac insufficiency or shock there is a diminished capillary circulation rate, accompanied by high oxygen utilisation in the tissues. Physical exertion, cold etc. may so raise the level of the reduced haemoglobin by increasing the oxygen consumption that cyanosis will develop.

DIFFERENTIAL DIAGNOSIS.

Numerous other instances of cyanosis in children are quoted in general medical literature; it is seldom necessary to make a differential diagnosis in children. Cyanosis is seen in a minor degree in emphysema, bronchiectesis, massive collapse of the lung in influenza, and rarely in asthma, but is never of that degree met with in congenital morbus cordis, and clinical examination, especially when aided by radiography, will in most instances answer the question.

RADIOLOGY.

In the diagnosis of Congenital Heart Disease, a s in other cardiac investigations, radiology plays a part equal in importance to auscultation. Cases are frequently met with where in the absence of auscultatory signs, gross cardiac deformity is demonstrated orthodiagraphically. It appears disproportionate to devote, as is frequently the practice, a sufficiency of time and space to the description of the clinical phenomena, without a corresponding consideration of the radiological characteristics of the condition present. It is therefore proposed to review briefly the appearance of the normal cardiac shadow, enabling one to visualise such abnormalities as appear when considering the individual lesion present.

The dome of the diaphragm lies on the right side at the level of the 5th rib, whilst on the left side the summit is at a lower and more variable level. This variation in height depends on the amount of gas in the stomach, which if present in sufficient quantity can balloon the diaphragmatic dome to a height level with that of the right side. The amount of cardiac displacement is, however, little altered by this variation in height, the lower surface of the heart being frequently seen even 1" below the diaphragm, delving its upper surface. The right border of the

heart forms an acute angle with the upper surface of the diaphragm at or about a point 1" from the mid line, and passes upwards with little of convexity towards the vertebral margin at the level of approximately the lower edge of the fifth dorsal vertebra. This intervening border represents in its lower part the right ventricle and in its upper part the right auricle. Immediately above the upper limits of the right auricle lies the superior vena cava, normally invisible. When the right heart is enlarged with distention of the superior vena cava, a ribband of shadow can be seen passing upwards, quickly to be overshadowed by the lateral margin of the ascending aorta, which probably includes in its right margin the higher limits of this venous trunk. This ribband caused by the superior vena cava is well seen when the aorta is narrow, not over-riding the right vertebral margin, c.f. plate 25 . The upper level of the aortic arch lies opposite to about the level of the lower margin of the first rib, with the aortic knuckle appearing to the left side of the manubrium sterni for a varying distance before it bends downwards and medially to disappear behind the upper margin of the right ventricle. These levels for the aortic arch represent its average position. Vaquez and Bordet (141) give the normal distance between the upper margin of the aortic arch and the sternal ends of the clavicle as 20-30 mm, Abreu (4)

as little as 2-4 mm. This measurement must depend largely on the type of chest, height of the diaphragm, and the blood pressure. Surveying the clavicular-aortic gap in the present series, it is seen that there is no relationship between it and the size of the heart, or the heart-lung quotient. The quotient in cases with a gap of 10 mm. or over varying from 1 — 1.75 to 1 — 2.1, and in cases with a gap of under 10 mm. from 1 — 1.79 to 1 — 2.4 c.f. Table 1.

Table 1.

Name	Height.	Weight.	H./L.Q.	Cl.Ao.
Wallace	+ +	+	2.40	3
Leighton	- + -	—	1.79	9
Briggs	o	o	2.10	10
Brown	o	—	1.80	16
Markham	+	+	1.75	10
Tomlinson	— —	— —	1.93	5
Gladding	+	—	1.92	20
Taylor	— —	— —	1.96	5
Copeland	— —	— —	2.10	12
Latus	—	+	1.95	12
Powell	+ +	+ +	2.04	5

Plus and minus represent degree over and under average.

o represents parity with normal.

H./L.Q. represents Heart Lung Quotient.

Cl. Ao. represents Clavicular Aortic gap.

The pulmonary artery arises from the upper left margin of the heart in the second left space, and is normally invisible, but any enlargement either of

it or of the conus of the right ventricle will show as an elevation on what is the normally straight concave or even convex (108) line, passing from the lower limits of the aortic knuckle to the upper surface of the left ventricle. It must be realised that these deliminations must vary according to whether the heart is photographed in systole or diastole. With an exposure of 1 sec. or over, the two shadows may be seen. It would appear to be preferable to consider the cardiac shadow in diastole with the heart in a condition of rest. Automatic photography as practiced in America is the optimum method, with an instantaneous exposure in early diastole prior to auricular contraction.

The investigation of septal defects from a radiological point of view demands the interpretation of three features, (1) The general size of the heart. (2) The size of the right auricle and ventricle. (3) The pulmonary artery and conus of the right ventricle, as seen, a) on antero posterior inspection, and b) the oblique positions.

1. GENERAL SIZE OF THE HEART.

This, though a question of primary importance, is at once a stumbling block upon which no clear answer can be given. Percussion of the cardiac margins is not reliable; too much depends on the shape of the chest wall, as related to the surface of the heart,

and also to the position of the heart in the thorax, to enable one to determine with exactitude the cardiac borders by this method. Palpation likewise leaves too great a margin for error. An over enlarged heart will give a diffuse apical heave, whose point of maximal thrust it is impossible to determine. The impulse of mitral stenosis and nervous tacchycardia is localised and distinct, and some inference can be drawn therefrom in the estimating of the left side of the heart. As a general principle it may be stressed that unless a clear right angled thrust can be felt outside the nipple line, in which case the heart is obviously enlarged, no certain opinion can be given as to the size of the organ by palpation. Reference may be made to the present series, in which the greatest care was taken in estimating the extreme cardiac margin by palpation of the apex beat in the upright and prone positions, and comparison made between the estimated and the actual size as determined by teleo-radiography, c.f. Table 2.

It is thus seen how in only three cases did the two methods of mensuration coincide, how the majority showed a smaller size by palpation than by teleo-radiography, and how in only three cases was the heart estimated to be larger than it really was; in short how the tendency is to underestimate the size

of the heart when relying on either percussion or palpation. It is admitted that in the early series exposures of 0.2 — 0.4 secs. were made with the result that the photographs may have been taken either in cardiac systole or diastole.

Table 2.

Name.	Cms. from mid line Palpat.	Teleo.	Error palp. over teleo.
Markham	7.0	6.0	+ 1.0
Gladding	6.0	7.0	— 1.0
Taylor	6.0	7.0	— 1.0
Briggs	5.5	5.5	— — —
Wallace	5.5	5.7	— 0.2
Allen	5.25	6.2	— 0.95
Powell	6.0	5.8	+ 0.2
Copeland	6.5	5.5	+ 1.0
Tomlinson	5.5	6.5	— 1.0
Allott	5.0	6.5	— 1.5
Frisby	6.0	6.0	— — —
Smith	5.5	6.8	— 1.3
Leighton	7.0	7.0	— — —
Brown	5.0	6.2	— 1.2
Latus	5.0	6.1	— 1.1
Wright	6.0	7.2	— 1.2

Palpat. - - - Palpation.

Teleo. - - - Teleoradiogram.

Having determined the actual size of the heart, difficulty is immediately met with in the determination of whether the heart is enlarged or not; there is no constant normal heart size, there being several factors which influence the size in each individual.

a. POSITION IN CHEST. A large proportion of adolescents suffer from a minor degree of scoliosis,

any accentuation of which will alter the centering of the heart in the chest cavity. To whichever side the convexity of the curvature is directed, to that side will the heart be projected, and thus give a false impression of the relative sizes of the two sides. Kirsch (75) was the first to point this out.

b. SHAPE OF THE HEART. It will be seen that not only do individuals vary according to the breadth, but also according to the height of their hearts. A tall heart is always associated physiologically with a narrow heart, and a short heart with a broad heart. Excellent illustration of this point is furnished by comparing Wallace with Taylor in Table 3, in neither of whom is there any left sided enlargement.

Table 3.

Name	Age	Height	Weight	Chest	Heart Height	Heart Breadth	H./ L.Q.
				cms.	cms.	cms.	
Wallace	16	65"(+ 3.25)	116(+ 3)	23.7.	18.0	9.7	2.4
Taylor	8	42"(- 5.8)	39(-12)	19.8	11.5	10.1	1.96

Figures in brackets represent lbs. or inches over or under normal.

It is not meant that these two cases, being of different ages and suffering from different defects should be studied too literally, but they illustrate how a girl with an extremely tall heart may have such

a degree of narrowness as to give a H./L.Q. larger than in a boy half her age - neither of whom suffer from left-sided enlargement.

c. RELATIONSHIP TO CHEST SIZE. This has been the study particularly of Hammer (56), Dietlen (32), Grödedel (52 - 53), Deutsch (31) and Vaquez and Bordet (141). Theo. Grödedel (52) investigating a motley group of 100 recruits, 20 sergeant majors, and 38 children estimated a heart lung quotient of respectively 1 — 1.9, 1 — 1.92 and 1 — 1.95. The chest measurement is the internal horizontal chest measurement at the lowest point of lung translucency above the dome of the diaphragm. Hammer (56) after examination of 420 cases, suggested as a normal quotient - minimum 1.58, maximum 2.34, and mean 1.98. Danzer (28) basing his figures on 500 normal cases gives a rate of from 1.9 to 2.6. Parkinson & Cookson (108), on an investigation of 43 goitrous hearts found that their figures were (where comparison was possible) 23% higher than Hammer's, 62% higher than those of Vauqez & Bordet, and 40% higher than those of Schinz, Baensch & Friedl (126). There is doubtless a definite relationship between the size of the chest and heart width, but it is not possible where such a large margin of error exists to rely on this method alone.

d. SIMPLE INSPECTION OF TELEORADIOGRAMS. To anyone accustomed to the inspection of numbers of such films

a definite impression is readily given as to whether one or other side or the whole of the heart is enlarged, apart from mensuration.

2. RIGHT AURICLE AND VENTRICLE AND SUPERIOR VENA CAVA.

Enlargement of the right auricle when present is well seen on antero-posterior view. This chamber occupies the upper limits of the right cardiac border, and any enlargement thereof, as in patency of the inter-auricular septum, will show as a swelling whose outer limits occupy the furthermost part of the heart to the right - as in case 15. Dilatation of the right auricle, however, will not cause pressure on the oesophagus, as seen on oblique view. Enlargement of the right ventricle evidences itself more on the left side of the heart, the apex rising upwards and the lower border of the heart assuming a marked convexity downwards; this appearance has been aptly named by the French "Coeur en Sabot", and is well illustrated in Plates 15 & 22. The superior vena cava, when dilated, may not be seen, the shadow being incorporated in that of the ascending aorta. It may, however, be picked out as a ribband passing vertically upwards from the upper limits of the right auricle close to the ascending aorta.

3. PULMONARY ARTERY.

a.) ANTERO-POSTERIOR VIEW. As previously described, the upper left border of the heart may be straight,

concave or rarely convex. It follows, therefore, that in a heart with a convex or even straight upper left margin, an absolute enlargement of the pulmonary artery may be present though unsuspected. Further, that enlargement of the pulmonary artery though evident, may at a later date be hidden by subsequent increase in size of the right or left ventricle. It is therefore helpful to take serial photographs, and where that is impossible to combine antero-posterior with right anterior oblique view. Moreover, much help can be derived from the study of the relationship of the pulmonary artery to the oesophagus. Enlargement of the pulmonary stem itself may cause pressure on the oesophagus but such pressure is more commonly the result of dilatation of the right branch of the pulmonary artery immediately below the left bronchus, to the left of and immediately distal to the fifth thoracic vertebra. Barium swallowed during screen inspection will demonstrate this pressure on the gullet and has been used in the present series. Difficulty was met with through the use of barium sulphate as used for gastric photography, whereas citobaryum, especially when further emulsified by liquid paraffin, being thicker and more tenacious will better demarcate the gullet and remain in situ for a longer period to enable teleoradiographic plates to

be taken with the barium still distending the oesophagus. On anterior view, at this level and opposite to the convexity of the pulmonary arc on the left cardiac margin, can be seen an impression on the gullet, with the aortic bed immediately cephalic, c.f. plate 8 , due to pressure by the pulmonary artery. Various methods have been used to measure this enlargement of the pulmonary arc. Vaquez and Bordet (141) measure from the summit of the arc perpendicularly to the mid vertebral line. Abreu (4) measures the drop between the summit of the arc and the base line joining its upper and lower limits, giving 3 mm. as a normal; as previously suggested this is not reliable. The conus of the right ventricle may rarely be seen as a separate bulge at a lower level than the pulmonary arc, or may be incorporated in it.

b.) FIRST OBLIQUE POSITION.(With patient turned half left.) With the aid of barium in the oesophagus enlargement of the pulmonary artery may be seen. When present two areas of pressure can be made out - an upper or aortic bed and a lower, at the level of the fifth dorsal vertebra, due to the right branch of the pulmonary artery. The pulmonary stem itself may press backwards on the left bronchus and the oesophagus. This method is helpful in determining whether convexity of the upper right margin of the heart is due to a true enlargement of the pulmonary arc or of

the conus of the right ventricle or the appendage of the left auricle. In the presence of a single indentation of the gullet it must be determined whether it is caused by the aorta alone or the pulmonary artery in addition. Where both vessels are taking part in the production of this appearance the "bed" will be longer than would be the case were it due to the aorta alone. Plates 17 & 19 demonstrate this point well. In the former the large aortic bed is seen but with its deepest part at a higher level than the pulmonary artery. In plate 19 taken in the second oblique position there is seen to be no pulmonic pressure exercised on the gullet.

c) SECOND OBLIQUE POSITION (with patient turned half right) Here the pulmonary artery is not so well seen and this view is seldom used except to corroborate what has already been noted. In plate 19 can be seen a case of Fallot's Tetralogy with a large aortic shadow and a ~~much~~ dilated right auricle.

It must be remembered that carcinoma of the oesophagus or of the glands lying in the angle of the bifurcation of the trachea may simulate enlargement of the pulmonary artery by obliteration of the oesophageal lumen.

P A R T 2.

THE DIFFERENT DEFECTS.

AURICULAR SEPTAL DEFECTS.

Of all defects found in the newly born this is in all probability the commonest, and at the same time the one which may show the fewest, if any, signs. Only by reference to consecutive series of autopsies performed as a routine examination can one arrive at any conclusion as to its true incidence. Evidence on this point has been given above. The frequent impossibility of diagnosing this defect during life is stressed but the writer has included in this series two cases where the auscultatory and particularly the radiological appearances, strongly suggest its presence.

The foramen ovale is that portion of the interauricular septum lying between the lower edge of the septum secundum and the upper border of the septum primum. Unlike the foramen primum it does not close until a disputed date after birth. The actual date of closure has been a subject of controversy. Gray (49) gives the time of closure as ten days after birth, Holt and Houland (60) suggest that it remains open until the middle of the first year; Griffiths and Mitchell (51) deny its closure until the end of the first year or even during the second year; Patten and Taggart (109) believe closure to occur during the latter half of the first year;

Maude Abbott (1) refuses to specify any exact date of closure. This is a matter of clinical importance. The circulatory and respiratory requirements of a child of one year are proportionately little less than those of a child of four or five, and if normal closure is delayed until the second or third year without giving rise to pathological signs and symptoms it would appear to be, except when of large diameter, of little importance with regard to the child's health. To settle this point at variance Christie of America (22) performed 500 autopsies in non malformed hearts, i.e. gross malformation. He discovered that in 82.5% of cases the foramen ovale was open at the end of two weeks. By open he included those in whom only a fine probe could pass at the edge of the valve. At the end of ten weeks 16.2% were still patent, and at the end of the first year 5.7% were still open. These figures go far to explain the frequency with which the condition was found in other series of autopsies performed on young children in whom the "defect" was thought to be pathological, and to sustain the belief that even wide patency of the septum may be unaccompanied by gross pathological signs, at least until the shunt is changed to a right-left direction. The major number of such "defects" are minor slits or fenestrations.

Patency of the foramen ovale is without doubt the commonest type of auricular septal defect, comprising over 55% of auricular septal defects in Abbott's series of 58 cases. They are the type most commonly found at autopsy in young children, and except in extreme degree or as a member of an association of defects, symptomless.

The opening may be anything from a fine slit to a wide patency, even to almost complete absence of the septum, or the septum ovale may be perforated by fine pin holed fenestrations. Defects of the upper and lower parts are rare, and when present are usually accompanied by associated abnormalities of the adjacent structures. When involving the upper part of the septum, a very rare condition, there is usually interference with the entry of the great veins, the pulmonary veins frequently uniting into a common trunk, while deviation of the septum to one or other side may cause the pulmonary veins or the superior vena cava to "look into" both auricles.

Defects of the lower part of the septum are due to a persistence of the foramen primum, and according to Mönckeberg (96) always associated with a lack of development of the septum primum. Allied to this is a peculiar splitting vertically of the anterior flap of the mitral valve, the inferior extremities of

which are widely separated, with resultant incompetence. In the case of the foramen ovale, we have two membranes, whose adjacent edges approximate, overlap and fuse at an indefinite date following birth. It is understandable how an increase in pressure in the pulmonary circulation, arising physiologically with the first respiration, if it occurs before the overlapping of the membranes has reached a stage sufficiently to strengthen the septum, or arising postnatally from any sudden acute lung infection or congestion, may separate an already weakly united septum, and overcome the valve-like action of the membranes, which is only at its optimum in the presence of a marked superiority of the systemic over the pulmonary pressure. As elsewhere, such a free margin may in rare cases become the seat of an endocarditis, but it appears to be a confusion of cause and effect to suggest endocarditis as a primary cause of the anomaly. More frequently is the patency of the septum a sequel to an endocarditis of the pulmonary valve, where the ventricular septum is closed. It is indeed a peculiar thing that an implantation endocarditis is seldom present in this condition, unlike other septal lesions.

PHYSICAL SIGNS.

CYANOSIS. Cyanosis does not commonly occur in an uncomplicated defect of the interauricular septum

when of moderate size. Even when the heart assumes the form almost of a trilocular organ, cyanosis may be absent. Dufour and Huber (33) describe such a case in whom even in the presence of severe terminal lung infection, and complex rheumatic cardiac affection, cyanosis was not present. Tirard (137) cites a case of a child 5 months old, with patency of the foramen ovale, granulations onthe pulmonary and tricuspid valves, patency of the ventricular septum, and the ductus arteriosus, in whom no cyanosis was noticed. Of all signs occurring in the course of heart disease or defect, cyanosis is the most capricious and one must not lay too great weight on its presence or absence alone, as being proof for or against inter-auricular septal defect. The colouring of such children is pale, with a delicate transparency of the skin which is very typical. Cyanosis of a deep hue may, however, develop in the presence of marked venous arterial shunt, even 75% of the blood at times being shunted through the defect (Bedford, D. Evan - personal communication), but is never of so deep a hue as in Fallot's Tetralogy. Where cyanosis is present in a child suffering from gross congenital heart disease, patency of this septum may be and probably is there, but the cyanosis is evident rather because of its association with the other graver anomalies.

PHYSIQUE AND MENTAL DEVELOPMENT. Weiss (148) suggests that with patency of the interauricular septum the children are commonly of an infantile build. Provided it is accompanied by gross abnormality elsewhere in the heart, especially hypoplasia of the aorta, which we know to be a common accompaniment, or pulmonary circulatory obstruction, one may expect the height-weight factor to be below average, but per se, there is no reason why simple patency should entail underdevelopment. Further, Weiss states in the same paper that it is frequently associated with poor mental development. The same argument as against the former belief holds good here, and it is felt that his statement should be more restricted to those cases which show a large defect, or are complicated by other defects. It is to be remembered that such children may be deprived, frequently unnecessarily, of regular school attendance, sufficiently to warrant a labelling of them as "backward".

INTELLIGENCE. In writings on Congenital Heart Disease it is frequently asserted that it and poor mental development go hand in hand. It is appreciated that the incidence of Mongolism in such children is high. Morse (98) in his series of 100 cases occurring in his private practice, found six children so affected. In the writer's experience no such

association has been met with. Morse further, found five other feeble-minded children in his series, a percentage in all of mentally defectives of 11%. One reason for a degree of backwardness on the part of the child is given elsewhere, namely the frequency with which they are unnecessarily kept from school. In coming to a decision on the mental condition of the patient, one must bear two facts in mind. In the first place he may be mentally defective, whilst in the second he may be uneducated though cerebrally alert and potentially intelligent. One might, therefore, more usefully classify them in accordance to their response to education - such as they have had. The writer's experience is shown in Table 4.

Table 4.

STANDARD OF INTELLIGENCE.

Name	Age	Response to Education. Noncyanotic.	Cyanotic.	Age Defect Discovered.
Wallace	16	− + −		2 m.
Leighton	8½	− + −		2½ y.
Briggs	11	+ −		S.M.E.
Allen	10	+		6 y.
Frisby	11	+ +		S.M.E.
Smith	6	+ −		4½ y.
Brown	6	+ −		3 y.
Markham	5	+ −		6 m.
Latus	6	+ −		11 m.
Allott	9	+		S.M.E.
Powell	7	+ −		4 m.
Tomlinson	9		− + −	Birth.
Copeland	11		+ +	Birth.
Gladding	14		+ +	Unknown.
Taylor	8½		− − M.D.	Birth.
Wright	32.		+ −	Unknown.

Table 4 (cont'd.)

Abbreviations.

m - Months.	- -	very poor.
y - Years.	- + -	poor.
S.M.E. - School Medical Examination.	+ -	average.
M.D. - Mentally defective.	+	good.
	+ +	very good.

It is thus seen that out of 16 cases chosen at random, from this point of view, the response to such education as was given them was as follows :-

Very poor	-	1 - M.D.
Poor	-	3
Average	-	7
Good	-	2
Very good	-	3

Of these showing a poor response, the age of discovery was early. Wallace, Leighton, and Tomlinson were all "coddled" at home, and kept from school unnecessarily; they are all only children. Whilst the absolute degree of education in Copeland and Gladding is low they are mentally very acute and such education as their health has permitted them to enjoy has been taken full advantage of. Taylor is an illegitimate child with a mentally defective mother. It is thus seen that the standard of cerebral acuity compares favourably with children normal from a cardiological viewpoint, and that it is not unduly influenced by the degree of cyanosis present.

Apart from the risk of paradoxical embolism, which is an occurrence of no great rarity, twenty

cases having been extracted from the literature, eleven by Ohm (105), six by Ballet (7), two by Versé (142) and one by Beattie W.W. (11) where an embolism from the haemorrhoidal vessels 15 cms. long was found lying in the right pulmonary artery, with multiple recent infarcts in the spleen and kidneys, the condition, namely auricular septal defect, far from adding to the burdens of life of the patient, in many cases appears to aid the difficulties of the individual. Abbott (1) quotes Firket in 1880 as having published the records of a woman who lived to 74 having survived eleven pregnancies and three abortions, who had a large interauricular septal patency. Lutembacher (87) refers to a woman who died, also at the age of 74, with cyanose tardive, having a patency size 3.5 x 4.0 cms. with the mitral orifice thickened to a chink, having survived seven pregnancies.

AUSCULTATION. Depending on the position and size of the patency, and whether the auriculo-ventricular valves are involved, any murmur, systolic, systolic and diastolic or presystolic, may be heard. It is typified as being soft, harsh or musical, and with its maximum intensity parasternally either to the right or left of the sternum, or heard diffusely over the precordium, but usually in the third and fourth left space, and commonly accompanied by weak aortic sounds,

and accentuated pulmonary sounds, especially the second. - In other words the murmur may be present or absent, and if present may be of any character and heard over no specified area. It is seen then, how little help may be had from the auscultatory signs and yet gross defect in the presence of the typical radiological picture will cause little difficulty in diagnosis. If it is borne in mind that the pulmonary artery is not commonly enlarged in pulmonary "stenosis" and is always enlarged in patency of the interauricular septum, except where the perforation is small in size, the diagnosis will be greatly aided.

X-RAY. The right auricle is commonly enlarged, and the bulge caused thereby can be seen on the screen. Its vigorous pulsations are usually easily noticeable, and together with these there is often great enlargement of the pulmonary arc. Where hypoplasia of the aorta is present, narrowing of the aortic shadow with a less prominent knuckle may be seen. In minor degrees , the appearances are not characteristic.

In this condition where dilatation of the pulmonary artery is present, pulsation of the hilar vessels is commonly seen. One must be careful not to confuse transmission of the auricular impulse with true hilar pulsation in which latter case alternate variation in the hilar shadow occurs with each systole and not merely a pulsating heave.

DEFECTS OF THE INTERVENTRICULAR SEPTUM.

MALADIE DE ROGER.

Maladie de Roger is that anomaly wherein the only defect is an aperture of varying size to be found in the interventricular septum. Abbott (1) in her series of 850 cases found a defect in the interventricular septum on 255 occasions, of which 46 were pure or "primary". On page 18 we described the approximation of the ventricular septum to the bulbar septum, leaving there an elliptical space later to be filled in - the pars membranacea septi or the undefended space of Peacock. It is in this region that the defect is most commonly seen, seldom occupying the pars membranacea itself, but commonly the anterior part of the anterior septum - (Rokitansky's space) immediately beneath the aortic cusps, and establishing a communication with the right ventricle, beneath the septal cusp of the tricuspid valve, at times perforating it or bulging it in an aneurysmal like fashion. The defect may lie far forward in the septum anterior to the pars membranacea, immediately beneath the front wall of the heart, and opening into the conus of the right ventricle, as reported by Coupland (24) and Rolleston (119). Evidence of the left-right flow of blood through the defect may be seen in patches of fibrosis or

vegetations commonly to be found on the opposite ventricular wall, or on the tricuspid valve cusp. The tricuspid valve may be rendered incompetent by adhesions or stretching, opposite to the point of entrance of the defect, as has been reported by Hart (57) and Thompson (136) (where the tricuspid flap was pierced) and McIntosh (101). Defects may be found lower in the septum near to the apex, or in extreme cases, involving practically the entire septum, this constituting a condition of Cor Biatriatrum Triloculare.

It has already been described how in congenital cardiac disease certain defects tend to group themselves into recognised associations of lesions, and this patency is one member of such a group of anomalies, i.e. that associated with the name of Fallott (41 & 42), but first described by Sandifort (125) in 1777, as showing a narrowing of the pulmonary valve, deviation to the right of the aorta, ventricular septal defect, and hypertrophy of the right ventricle. The association may, however, occur without the aortic displacement, or in the absence of pulmonary stenosis the septal defect may be combined with rechtlage of the aorta. Patency of the ventricular septum is undoubtedly met with as an uncomplicated lesion. Reverting to the figures as given by Abbott which, as we have seen, have all been corroborated by

autopsy; of 255 cases of interventricular septal defect or 37% of her whole series, 54 occurred with the ventricular septal leak as the only lesion, except for eight cases of rechtlage of the aorta, which were included in this series of 54. This goes to discount the belief of Laubry and Pezzi (80), unsupported by figures, that uncomplicated, it is a lesion of rarity, being in practically all cases associated with pulmonary stenosis. Weiss (148) also, holds that it is commonly seen in conjunction with a degree of pulmonary stenosis. We will see later how Laubry and Pezzi's view may be substantially true, the incidence of true interventricular defect without conus or pulmonary stenosis being smaller than Abbott suggests.

There are thus two classes of ventricular septal defect :- firstly as a pure or primary lesion, and secondly as a member of an association of defects.

The former is commonly coupled with the name of Roger (117), who in 1879 described as a separate clinical and pathological group those cases in which a "pure" uncomplicated patency of the interventricular septum without cyanosis is the only defect present. He considered it to be the commonest of all congenital cardiac defects consistent with life, and that it occurred in the upper part of the septum. This we know not to be strictly in accordance with fact,

cases falling within his descriptions where the lesion is situated at the lower part of the septum. He recognised how cyanosis was met with in rare instances of interventricular septal patency, but insisted on some degree of pulmonary stenosis being present in such cases.

PHYSICAL SIGNS. This disease has been aptly described by Laubry and Pezzi as one of "Functional Silence." The literature is replete with cases of Maladie de Roger in whom the defect was discovered accidentally often not until late in adult life. In the present series in not one pure uncomplicated case of ventricular septal patency was there any complaint mentioned or admitted on questioning referable to the heart and circulatory system.

BRUIT. The murmur is said to be characteristic in its consistency of localisation; this the writer has not found to be so. It may be heard down the left border of the sternum, most commonly in the third or even second space, but frequently int he fourth space or even at the fifth rib. The third intercostal space is the seat of election. Where heard in the second, which it may be at times, it is usually associated with some degree of narrowing of the pulmonary valve; when heard as low as behind the fifth rib, a lesion in the lower half of the septum is to be suspected. According to some observers - Laubry

and Pezzi (80), - the murmur may be heard with its point of maximal intensity to the right of the sternum; such the Writer has not found to be so except as the result of propagation. In the present series it was maximal :-

Once at the second left space.
Twice " " third space.
Twice " " third-fourth space.
Once " " fourth rib.
Twice " " fourth space.
Once " " fifth rib.

i.e. in practically 80% of cases the point of maximal intensity lay between the third and the fourth spaces.

It has been doubted that there is a flow of blood from the left to the right ventricle of sufficient force to produce a murmur, but there is abundant anatomical proof that such is the case; defects of the septum at times assuming a funnel shape, with the base of the funnel towards the left ventricle, which could only be so were there a strong flow in that direction. Horder (61) reports such a case where in addition, a patch of vegetation was found on the wall of the right ventricle opposite to the defect, carried thereto through the patency from the aortic valve.

The murmur may be, and usually is, rough, prolonged right through systole, accompanying and not replacing the first sound which however, it usually drowns in its loudness. Commonly it is not so unduly prolonged, and tends in a number of cases to tail off three-quarter way through systole. The

Writer has heard it of a much softer, whistling character, and even musical. Musical murmurs are rarely endocardial, but where Roger's disease gives rise to a musical murmur one is probably dealing with a lesion high up in the base, bulging or perforating the septal cusp of the tricuspid valve, or maybe with an anomalous cord passing through the aperture. In the cases here presented the murmur was noted to be musical in only one individual. The localisation of the murmur is constant, notwithstanding alterations in posture on the part of the patient; neither does it vary with the different phases of respiration, except that deep inspiration naturally tends to muffle it, nor with the passage of time. This latter point Roger stressed, insisting that it remained as it always was, over a period of years.

According to Laubry and Pezzi (80), only apertures of medium calibre will give rise to a murmur, variations in the bore of such average sized patencies determining the pitch of the bruit, whilst large and very small perforations give no auscultatory evidence of their presence. Abbott (1), on the other hand states that the murmur varies inversely with the size of the aperture, being heard only in those of medium or small size. Unfortunately, without corroborative post mortem evidence, one is unable to substantiate or refute either of these statements.

The other cardiac sounds are unaltered, although the pulmonary second sound may be accentuated, as occurs wherever continuous and forcible contraction of the right ventricle distends the pulmonary artery.

PROPAGATION. Of all systolic murmurs this is the most fixed. Examination discloses a central point of maximal intensity, with the murmur fading in all directions therefrom like ripples on a pond. Roger stressed that it was not conducted in any one direction, but experience shows, modifying what has been said above, that there are certain areas where it is better heard than at others. Firstly, transversely along the corresponding intercostal space, for a distance proportionate to the intensity of the bruit. Depending on its loudness it may be heard in the back; never, except when very loud, is it heard under the left clavicle; congenital murmurs heard best over the second or third left space, and carried upwards and outwards from there, being in practically all instances due to pulmonary stenosis, which it must be remembered may and frequently does co-exist with ventricular septal defect. Further, the murmur of a "pure" defect is never conducted into the vessels of the neck.

One must be careful, however, in diagnosing a dextroposition of the aorta solely on account of the

hearing of a murmur on the right side of the neck, for it is the Writer's opinion that the vertebral column may act as a sounding board, carrying the murmur up to the stethoscope placed firmly on the neck, especially in sparse subjects.

THRILL. What has been said of the bruit applies equally to the thrill in its localisation, and propagation, but even more so than with the bruit is the thrill seldom felt other than at the edge of the sternum. It is not constant in its presence; Roger (117) insisted on its being present, but Weiss (148) estimated that it was demonstrable in only about 50%, and Abbott in about 35% of cases. In the present series, in two cases of undoubted Maladie de Roger no thrill was felt.

POLYGRAPH. The graphic evidence will be discussed more fully when dealing later with the signs of right-sided heart enlargement, but it may be mentioned here how Laubry and Pezzi suggest as a point of diagnostic importance, the exaggeration of the "a" wave in the jugular tracings. This is easily understood, corresponding as it does to auricular systole, and can be demonstrated without difficulty in older patients, but tracings taken in children of the age included in this series are unsatisfactory because of the small dimensions of the neck, and the predominance of the carotid pulse.

X-RAY. The radiological appearances of Maladie de Roger are said to be characteristic, namely the assumption of a globular appearance of the heart due to right ventricular enlargement. It is difficult to understand why, if there is no other cardiological departure from the normal, there should be this appearance, and one does meet cases (in the present series, there are two such examples - Brown and Frisby) where, in the face of otherwise incontrovertible Maladie de Roger, the radiological picture shows no right-sided enlargement. When one says no such enlargement, it is realised how one may be overstating one's ability to guage minor degrees of hypertrophy; and why should one expect it to be present? There are four possible explanations. 1. With patency of the interventricular septum, the systemic diastolic pressure carried by the left ventricle is transmitted through the defect to the right ventricle, which has to carry this load, in a lessened degree because of the usual small dimensions of the aperture, in addition to its own. Nevertheless the head of blood to be carried by it probably represents at a minimum 40 mm Mercury, and to meet this it may enlarge. But can the pulmonary circulation withstand the strain of a constant head of 40 mm of Mercury? In moments of stress, the right ventricular output may

rise from 3 to 30 litres of blood per minute, but this is a temporary change, and entails no sudden strain on the alveolar capillaries by reason probably of the gill valve action of the ventricular infundibulum. To allow of the pulmonary circulation to carry a constant load of 40 mm Hg would undoubtedly cause rupture of the fine alveolar capillaries, with haemoptysis and atheromatous changes in the pulmonary artery.

2. Is the right sided enlargement the result of an attempt on the part of the right ventricle to raise its pressure to a state of equilibrium with that of the left, and so neutralise the shunt resultant upon the septal leak? If that were so there would be no reason for the production of either systolic murmur or thrill.

3. We know the common site for perforation of the septum to be high up in the anterior part of Peacock's undefended space; how immediately to its right side lies the septal cusp of the tricuspid valve, and how the force of blood projected upon this flap is sufficiently great at times to produce aneurysmal pouching of the valve cusp. It can be easily understood how such a force over a period of years may entail a considerable strain on the guy ropes of the valve, viz, the papillary muscles, and it requires no great imagination to picture a stretching of them, with a

resultant looseness of the cusp, and arising from that a defective closure thereof, with regurgitation into the auricle. This would certainly cause a varying degree of hypertrophy both ventricular and auricular, with characteristic polygraphic jugular tracings, namely a heightening of the "a" wave. The murmur of a tricuspid incompetence when present, is holosystolic, and is heard at a lower part of the sternum than is usual in Roger's disease, and is propagated to the right axilla, but is in the majority of cases completely silent.

4. In the presence of an arterio-venous shunt through the patency, an added quantity of blood must be accommodated in the right ventricle. This additional blood will require, for its expulsion, an increase in the propulsive power of the right ventricle, with consequent hypertrophy. It is in this manner that numerous cases of Roger's disease show radiologically a "pulmonary arc" caused by a distention of the conus of the right ventricle or the pulmonary stem and an accentuated pulmonary second sound.

CYANOSIS. Roger, in describing this syndrome, admitted that cyanosis might or might not be present, but that it was commonly absent, and this undoubtedly is so. In Abbott's series of 48 "pure" defects, cyanosis was absent in 24, terminal in 8, slight

in 3, and marked in only 1. Unlike so many French writers of his time, Roger did not adhere to the theory of Senac, that cyanosis was in all cases due to admixture of the blood of the two systems, and conversely that admixture of the two bloods must invariably give rise to cyanosis. His explanation which came near to the truth was that cyanosis was only met with in these cases, in the presence of pulmonary obstruction.

It was not until 1889 that Bard & Curtillet (10), reviving the admixture theories of Senac (129), Gintrac (48) and Corvisart (23) showed how any rise in the pulmonary circulation would tend to reverse the flow of blood from a left-to-right to a right-to-left direction, and this occurring most commonly as a terminal event, was named by them Cyanose Tardive. This reversal of flow will only produce clinical cyanosis provided oxygen unsaturation reaches and rises above the level of 6.7 c.cms. oxygen unsaturation %. This latter theory was evolved with relation to auricular septal defects, but applies equally to patencies of the interventricular septum.

DIFFERENTIAL DIAGNOSIS.

1. EXOCARDIAL MURMURS. With the exception of mitral stenosis, murmurs of exocardial origin present more similarity to congenital patency of the inter-ventricular septum than any other lesion. Exocardial

murmurs are not common in children, if one excludes pericarditis, but when present they have commonly a musical tone which the writer has heard only once in the congenital lesion. Exocardial murmurs are seldom present unaltered throughout all phases of respiration, which is a sine qua non of the congenital defect, and are never accompanied by thrill. Moreover, the note in the functional murmur is softer, the bruit is shorter, and the first sound clearly heard through it. The bruit of pericarditis is in many cases double, not truly synchronous with systole or diastole, is basal or diffusely precordial in position, and with it the child is ill.

2. SYSTOLIC MURMURS. Of much greater difficulty is the common systolic murmur, so frequently heard in adults and children. As will be pointed out when dealing with mitral stenosis, organic lesions of the mitral valve in children are all rheumatic, and are ipso facto stenotic. There may be a systolic murmur present, but there is always some alteration of the second sound at the apex, be it only a weakening thereof. There remains however, a residuum of cases where the patient is fit and well, and for whom there is no restriction even in violent exercise, provided attention has not been directed to the adventitious souffle, who present a soft blowing murmur systolic or late systolic in time, diversely heard in the mitral

or basal areas, not radically altered by respiration, never accompanied by thrill, and with a clear sharp second sound. What are they? One cannot say. They bear a superficial resemblance to Maladie de Roger, they are even suggestive of a mitral stenosis, and yet they are neither; of organic valvular origin they cannot be for many disappear, and none appear to inconvenience. According to Skoda (130 & 131) the diagnostic point between these and other non-organic systolic murmurs, and the true organic mitral murmurs, is the intensity of the pulmonary second sound, which in the latter instance is always accentuated or doubled. This is not so; in diphtheria the commonest auscultatory cardiac alteration is a splitting of the pulmonic second sound, frequently accompanied by a basal or precordial systolic murmur, both of which eventually disappear.

3. MITRAL VALVULITIS. Difficulty is admitted by even such an observer as Laubry in the diagnosis of Maladie de Roger from mitral disease. That this might be so in adults is admitted, but even without the help of the radiogram there is only a superficial resemblance in the case of children, which careful examination will always dispel. In approaching this matter, which is one undoubtedly of confusion to many, it were well to insist that in children, in whom the vast majority of congenital lesions are examined,

valvular disease is only of one type, namely, rheumatic, with the additional possibility of a superadded subacute or acute bacterial endocarditis. Rheumatic disease is invariably stenotic, and infection, sufficient to produce permanent auscultatory signs, is sufficient to produce a concurrent stenotic process, and if regurgitation does occur it only does so by virtue of the stenosis, and the stenotic clinical signs will be evident. It is therefore a misnomer to speak of mitral or aortic regurgitation per se in children.

Mitral stenosis has certain very definite signs, totally distinct from those of patency of the inter-ventricular septum.

a) IMPULSE. There is something quite characteristic about the impulse of mitral stenosis, which in many cases, can be diagnosed from this alone. It has a slapping, not the heaving character of other organic or functional cardiac lesions, and is extremely localised.

b) THRILL. Thrill is by no means always present, but when so it is in practically 100% of cases diastolic. The thrill of Maladie de Roger is never diastolic. Its localisation moreover is further out than in congenital heart disease, being most frequently felt immediately internal to the apex, c.f. cases Wright and Taylor.

c) MURMUR.

(1) Systolic. This is of a character very typical but modifications of which can be imitated by patent interventricular defect. It is explosive, rising up to a crescendo, and then suddenly fading out or ending with a bang, but usually ending quietly.

(2) Second Sound. The second sound is always faint or absent entirely. Mitral stenosis can never be present together with a clear sharp second sound at the apex.

(3) Diastolic Murmur. A diastolic murmur is reported as being not uncommon in the congenital lesion. Bard (9) states that a quiet diastolic murmur is frequently heard, lasting throughout diastole. Breccia (15), Galliard & Cawadias (47) and Abbott all corroborate this statement, and the writer himself has heard it in one case when examined in the ventral and extreme left lateral decubitus.

In mitral stenosis, the diastolic murmur never occupies the whole of diastole. It may start early i.e. immediately after the second sound, as a short "Woff" or it may be more prolonged as a rumble, and may take up half diastole, but never the whole. Its timing will alter more and more with the heart rate, exercise bringing an early diastolic murmur up to the first sound, or presystolic.

d) RADIOGRAPHY. The radiographic appearances of the

two conditions may not in themselves be dissimilar. In Maladie de Roger, the right ventricular enlargement if present, gives an appearance which is typical, namely the globular heart. This is by no means always the appearance presented, as is illustrated by cases Brown and Frisby; clinical examination with them offers no alternative diagnosis, and yet the classical radiological picture is absent. Mitral stenosis, further, is productive of right ventricular enlargement and dilatation of the pulmonary artery, c.f. plates Nos. 24 & 25. In these the teleoradiogram gives a picture suggestive of interauricular septal defect. Clinical examination alone was able to establish a diagnosis.

e) HISTORY. Too great reliance must not be placed on a previous history of rheumatism. The child with congenital heart disease is as liable as his more fortunate brothers and sisters to fall a victim to rheumatic fever, and is all the more prone to a subacute bacterial endocarditis. It is clear therefore, how little help may be expected here. In point of fact the placing of weight on previous history may be more conducive to error than its disregard.

4. AORTIC STENOSIS. Laubry and Pezzi (80) wish to differentiate here again between congenital morbus cordis and a lesion which is in children always rheumatic. The two conditions are in no way similar. It is true that the murmur of aortic stenosis may be

heard in the second left intercostal space or at any point along the left border of the sternum, and even out to a line half way between the sternum and the left nipple, frequently having to be hunted for, and being only demonstrable in the position of ventral or extreme left lateral decubitus, and perchance coinciding with the common site of localisation of the murmur of a patency of the interventricular septum. The type of murmur, however, is different in the rheumatic condition in children, where the altered sound is the second one, the first being frequently normal. Where present the murmur is propagated into the carotids, at times with a thrill in the episternal notch, and with other signs of reflux of blood from the systemic vessels into the left ventricle. The murmur in interventricular patency is systolic in time, though there may rarely be a diastolic murmur. It is loud, and it is rough, and much higher pitched than in disease of the aortic valve. 90% of aortic valvular disease occurs in male children, and in a large percentage of these pericarditis has been present with well marked clinical signs. The proportion of male children over female in cases of Maladie de Roger is not marked. Further in the congenital lesion the hypertrophy is right sided, whereas with the acquired condition the preponderance is to the left.

5. INTERVENTRICULAR DEFECT WITH DEXTROPOSITION OF THE AORTA. In this condition, first described by Eisenmenger (37 & 38), where the ventricular leak is associated with a misplaced aorta over-riding the septum, and with a normal or dilated pulmonary artery, the maximal site of the murmur and thrill is in the mesocordium, and cyanosis and clubbing are present, being absent in pure Maladie de Roger. It may be further differentiated from pulmonary stenosis because of the absence of any murmur over the pulmonic area and carried up to the left clavicle; the bruit is not heard in the carotids. On the other hand its conduction is peculiar in that it is carried down towards the apex, to the right side, and also through to the back.

"PULMONARY STENOSIS".

Before dealing with the symptomatology of Pulmonary Stenosis one must have a clear understanding of the anatomical changes which havetaken place. Pulmonarey stenosis is a term generally used to describe a number of lesions of a constrictive nature, affecting the region of the pulmonary valve. It must be appreciated that the defect may not be confined to the valve nor need stenosis be the pathological change present.

PATHOLOGICAL ANATOMY.

1. STENOSIS. By this is meant a narrowing commonly of inflammatory origin, as with mitral stenosis one means the constrictive aftermath of rheumatic affection of the mitral valve. In this type of case the valve cusps are not developmentally altered but their condition is pathological because of the presence of vegetations indicative of an endocarditis having occurred during foetal life. The degree of stenosis will depend on the stage of foetal development at the time of infection and will also be an indication of the severity of the endocarditis.

2. ATRESIA. - This may originate in one of three ways :-

(a) Developmentally. - Reference has already been made to the absorption of the bulbus cordis in the common ventricle and how even in the adult heart its limits may be differentiated in the incorporating

right ventricle. As Keith (68 - 74) pointed out, this incorporation may be arrested at any point, resulting in the conus remaining as a separate chamber, to be found even in the left ventricle, and he demonstrated how the lower opening, which may be narrowed to a pin hole, represents the embryological lower bulbar opening. This stage may be carried a degree further, causing atresia of the conus. This type is not common, the faulty bulbar differentiation more frequently causing a constriction, sometimes with complete imperforation, of the pulmonary valve orifice. Here the cusps may be deformed, bicuspid, or replaced by a membrane stretching across the small arterial orifice.

(b) Inflammatory. Syphilitic, streptococcal or other infection may so attack and obliterate the pulmonary valve as to produce a secondarily atresic condition of the pulmonary artery.

(c) It must not be forgotten that an infective process may be implanted on an already atresic and deformed valve.

In none of these three latter types is the pulmonary artery enlarged except in the presence of a patency of the ductus arteriosus.

EFFECT ON THE VENTRICULAR SEPTUM.

Briefly, the effect on the ventricular septum will depend on whether the constrictive process, whether stenotic, atresic or both combined, so narrows the

pulmonary valve as to constitute a severe obstruction to the outlet of the ventricular content, prior to the time of closure of the septum. If this limit of obstruction is reached before there is complete fusion and incorporation of the bulbar and ventricular septa, a permanent patency will remain. If by the time of closure of the septum, the narrowing process has not reached such a degree, then one of three things will happen. Either, the septum being entire, the right ventricle will so hypertrophy as to overcome the obstruction; or if the condition is progressive, patency of the interauricular septum and the ductus arteriosus will result, thereby easing the strain put on the right ventricle; or thirdly, if it progresses to complete imperforation, death either in utero or in the first few hours or days of life.

CLASSIFICATION. The common classification of these defects is in accordance with whether the defect is stenotic (inflammatory) or atresic (developmental). It is felt that the clinical picture produced is proportionate to the degree of pulmonary obstruction, independently of whether it is inflammatory or developmental in its inception. It is therefore, proposed to describe the lesion according to whether partial obstruction or complete imperforation is the condition met with.

A. Pulmonary obstruction. (1) True inflammatory stenosis of low grade without associated ventricular septal defect.
(2) Of higher grade, either stenosis, or primary or secondary atresia, with associated septal defect.

B. Pulmonary imperforation (Stenotic or atresic) (1) With associated septal defect.
(2) Without associated septal defect.

A.1. PULMONARY OBSTRUCTION WITHOUT ASSOCIATED SEPTAL DEFECT.

PATHOLOGICAL ANATOMY. In this condition one is dealing with a true inflammatory type with an endocartitic implantation on a normal valve. The infection is of low grade or has occurred at a later date in gestation with the result that the stenotic process has become arrested at a point where the hypertrophied right ventricle can still overcome the obstruction at the pulmonic outlet and allow of a sufficiency of blood to reach the lungs.

EFFECT ON THE ARTERY. With a stenosis of this low grade, where a normal or almost normal volume of blood is still pumped through the pulmonary artery no change in its calibre is to be expected. Any alteration met with will be either of a hypoplastic nature, indicative of a diminution in the right ventricular output or, as occurs in this type only of pulmonary obstruction, a dilatation of the artery.

EFFECT ON THE RIGHT VENTRICLE. In this type the degree of pulmonary obstruction at the eighth week of

foetal life is not sufficiently great to cause a persistence of the interventricular orifice, with the result that closure of this septum proceeds in the normal manner. The volume of the right ventricular content is not great during foetal life, and with the aid of the interauricular shunt and the ductus arteriosus no undue strain is thrown upon its musculature, and hypertrophy may not be marked. Postnatally however, with the closure of the foramen ovale and the ductus arteriosus its content is increased, and to overcome the effect of the narrowing at the pulmonary orifice, it hypertrophies. With minor degrees of obstruction this hypertrophy is sufficient to maintain an adequate circulation. The musculature of the conus of the right ventricle however, is of poorer development than that of the fundus and dilatation of this chamber frequently results and can be demonstrated on radiological examination.

EFFECT ON THE RIGHT AURICLE. The patency or otherwise of the foramen ovale depends on the degree of pulmonary obstruction and the capability of the right ventricle to overcome this obstruction post natally. A relatively heightened right auricular pressure resultant on a failure of the right ventricle completely to overcome the pulmonic valve stenosis, will tend to cause a persistence of this foramen with a consequent interauricular interchange of blood. Moreover, stenotic processes, whether rheumatic or

not, tend to be progressive, and it can be understood how just so long as the right ventricle can cope with such blood as it receives so will any extreme degree of right-left auricular shunt be avoided. When, however, with the advancing pulmonary sclerosis in later life, or a progressive failure of the right ventricle, its load becomes increased, the strain on the right auricle will similarly increase, with marked venous arterial shunt. This, we will see, is borne out by our clinical examination of such patients.

PHYSICAL SIGNS.

CYANOSIS. Cyanosis is a common manifestation of obstruction at the pulmonary valve. It is the custom to describe its presence as being dependent on whether the ventricular septum is entire or patent; this is scarcely correct. Where the obstruction is extreme, whether the ventricular septum is perforated or not, cyanosis will be present. Where, as in these cases, the obstruction is partial with consequent imperforation of the septum, cyanosis is by no means always found and illustrations of such cases, where it has been reported to be absent, have been given above; in no case, however, has cyanosis been absent throughout life, cyanose tardive invariably making its appearance at a later date, as failure of the right heart supervenes. When stenosis is present without associated ventricular septal defect the right ventricle is

frequently capable, as we have seen, by hypertrophy, of overcoming the pulmonary obstruction, and if the foramen ovale is open such backward pressure as is exerted on the right auricle is reduced by the safety valve-like action of this shunt. Later, with right ventricular failure producing a venous-arterial auricular shunt together with a retarded peripheral circulation, there arise the two most consistent causes of peripheral cyanosis which develops with concurrently advancing clubbing, often reaching an extreme degree. Persistent cyanosis developing late in life in the presence of pulmonary stenosis is in almost all cases indicative of a complete ventricular septum, and rapidly approaching cardiac failure. The concomitant sign of dyspnoea, constant or in paroxysmal attacks, may even precede the onset of visible cyanosis, but even at this early stage, excitement, or exposure to cold or exertion will demonstrate an alteration in the tint of the mucous membranes, passing off immediately there is a return to normal routine.

THRILL. Thrill is commonly present, more so here than when associated with a ventricular septal defect, but is by no means constant. McPhedran (102) reports a case of pulmonary stenosis with an entire ventricular septum, the pulmonary valve projecting into the arterial orifice in a funnel shaped manner for 2 cms., with the orifice of the funnel only 2 - 3 mm. in

diameter, in whom there was no thrill felt. In Abbott's (1) series, in 7 cases with both septa closed, thrill was present in 3, and of 16 cases with a complete ventricular septum and patency of the foramen ovale, it was present in 8 or 50%. When present it is situated over the pulmonary area - in the second or third left space, close to the sternum, but may have to be hunted for with the patient in the ventral decubitus.

MURMUR. The murmur of pure pulmonary obstruction is in the majority of cases characteristic in its site and its propagation. It is heard maximally over the second or third left costal space, close to the sternum, but may most unaccountably be heard low down, even approaching the apex. It may be harsh and high pitched, and varying in intensity, but is usually loud, systolic in time and of long duration, lasting frequently throughout systole, and right up to and even encroaching on the second sound. Reference was made above to the case published by Abbott, Lewis and Beattie (3) where in the presence of a pulmonary stenosis, without ventricular septal defect, the murmur was heard at the apex, and transmitted to the axilla and the left scapular region, and was at times accompanied by a pre-systolic murmur - such cases are however rare, but the type must be remembered. The murmur is propagated up and out under the left clavicle, following the direction of the pulmonary artery, and may be

heard if loud, high up in the left axilla. It is practically never heard in the neck, but Scremini & Montes-Parega (127) however, report a case of this type where a holosystolic murmur covering the first sound was heard in the arteries of the neck, no other abnormality being present (c.f. p.84, para. 3 .) The pulmonary second sound is reported to be at times slapping or normal in character; such cases are rare, it being in almost all instances weak. Loudness of the second sound in the presence of signs indicative of pulmonary obstruction, is suggestive of patency of the ductus arteriosus, ~~or=conus=stenosis=~~

SIGNS OF RIGHT SIDED ENLARGEMENT. These signs are not restricted to this individual lesion, but are found wherever there is hypertrophy of the right ventricle and right auricle. Consideration of them is of the greatest importance, because right-sided enlargement is met with always in all grades of "pulmonary stenosis", and in many cases of Maladie de Roger.

1. Precordial Bulging.

This depends largely on the development of the patient. In the thin sparse type, especially when of a rachitic diathesis its presence is obvious, but in the more robustly built it may not be seen.

2. Epigastric pulsation, and at a later stage liver pulsation.

3. Pulsations in the second and third spaces, left

and/or right.

4. Enlargement of the perithoracic veins.
5. Percussive evidence, which is notoriously difficult, of the enlargement of the right ventricle and auricle beyond the right sternal line.
6. Auscultation. Enlargement of the right ventricle is commonly accompanied by accentuation or doubling of the pulmonary second sound, a clapping which, in the presence of an abnormally situated or prolonged systolic murmur is very suggestive of mitral stenosis. It can even be accompanied by a precordial presystolic murmur, which is due to the forcible contraction of the right auricle.
7. Phlebographic. Most characteristic is the enlargement of the "a" wave in the jugular tracing, due to the forcible contraction of the auricle driving back a larger "back-wash" wave into the neck, and to the right ventricular stasis forming an obstacle to the flow of blood from the auricle. This "a" wave may be so large as completely to overwhelm the "v" and "c" waves. Laubry and Pezzi (80) state that a tracing of the femoral vein will show two waves; a first, synchronous with the "a" wave in the jugular vein, and a second larger wave, synchronous with ventricular systole. Three such tracings were taken in cases of pulmonary stenosis

with septal defect.

8. Radiological. On antero-posterior view the enlargement of the right heart is usually obvious to the trained eye, even without the help of the measuring tape, and in the upper part of the bulge made by the right margin, the auricle and the superior vena cava may be seen pulsating vigorously. These changes are, however, more obvious on right oblique or lateral view, when the backward enlargement of the right auricle is seen bulging into the mediastinal space, and tending to obliterate the clear area between the heart and the spine. Administration of barium at the time of screening will demonstrate how the right auricle, no matter how enlarged, cannot exert pressure on the oesophagus. This enlargement can be seen distinct from the pulmonary arc which is at a higher level, the middle of the three swellings - the aortic, pulmonary and auricular.

9. Electro-cardiographic. The "P" wave is always of greater than normal height, indicative of the hypertrophy of the right auricle, and the QRS complex of extreme amplitude in leads 1 and 3. There is also evidence of a right sided preponderance. Lead 2 shows exaggeration of waves "P" and "T" more than leads 1 and 3, whilst in this former lead, namely lead 2, the QRS complex is of smaller potential and commonly diphasic.

A.2. PULMONARY OBSTRUCTION WITH OPEN VENTRICULAR SEPTUM

The pathological changes in the former defect are completely altered, as is the clinical picture, when dealing with such cases as present a defective ventricular septum. As already described, the effect on the interventricular septum depends on two factors, either singly or in conjunction one with the other, namely the degree of obstruction and the stage in the development of the foetus that the limit is reached in the capability of the right ventricle to maintain an adequate pulmonary circulation in the face of pulmonary obstruction. In this type one is dealing with either a higher grade of endocarditis or a developmental error in the early weeks of intrauterine life before the ventricular septum is closed. One remembers how the bulbar swellings "A" and "B", situated respectively on the left of the posterior and the right of the anterior walls, joined so as to divide the truncus arteriosus into two equal channels, with the aorta behind and to the right and the pulmonary artery in front and to the left. Any misplacement of these two swellings or faulty torsion of one trunk on the other will result in deformity of either opening, commonly the pulmonary outlet, nonuniformity in the size of the two vessels, and deviation to the right of the aorta, which may arise at times more from the right than from the left ventricle, or in extreme cases entirely from the right ventricle.

The relative frequency of the complete association of these defects is seen by making reference to Abbott's series of 96 cases of pulmonary stenosis.

		Cases		D.A.Open
F.O.closed	V.S.closed	7)		0
F.O. open	V.S. closed	16)	23	1
F.O.closed	V.S. open	44)		5
F.O. open	V.S. open	29)	73	6

It is thus seen that ~~out~~ of 96 cases, in only 23 was the ventricular septum closed, whilst in 73 or 79% of the series the ventricular septum was open. In later writings (2) she quotes 10% and 90% as the respective proportions. Rauchfuss, in his series of 192 cases, found the interventricular septum open on 171 occasions, and Vierordt in 105 out of 130 cases, (quoted by Abbott).

PHYSICAL SIGNS.

CYANOSIS. The changes in the adult heart in Fallot's Tetralogy are in no way different from those in the newly born child; one would therefore expect cyanosis to be present from the earliest days, and such we find to be the case; yet cyanosis is not constant in the first weeks of life. Pulmonary obstruction with ventricular septal leak and rechtlage of the aorta typifies the text book type of morbus coeruleus, the only condition producing a deeper discolouration being pulmonary atresia without septal

-defect. The conditions necessary for its fulfilment are present in all their full development; pulmonary obstruction, a right-left shunt and a pouring of the right ventricular blood direct into the wide mouthed aorta. With true pulmonary stenosis without inter-ventricular septal patency,especially where the ductus arteriosus is open, a greater proportion, if not all, of the venous blood finds its way eventually into the pulmonary circulation, even in the face of an open foramen ovale, because until right ventricular failure supervenes, the auricular content is independent of the ventricular muscle. In the major defect, however, no such happy condition is present, and the cyanosis, the clubbing and the dyspnoea are extreme from early life. Patency of the ductus arteriosus, when it accompanies pulmonary stenosis, must have a beneficial effect for the relief of cyanosis; cyanosis is never extreme in the presence of patency of the ductus arteriosus. This is because of the negative pressure produced in the chest by respiration, whereby some aerated blood is sucked into the pulmonary from the systemic circulation with each inspiration, and to the difference in pressure of the two circulations.

As a compensatory effort, the red blood cells are increased in number to unusual proportions. As quoted above, Murray Leslie (81) reported a count of 12,750,000 in a case of this

association of defects. In the present series, case 13 (Gladding) showed a blood count of 9,470,000. On a very cold day Taylor's blood (case 14), was counted giving the extraordinary figure of 13,470.000. A sample was taken the same afternoon to a pathologist who corroborated this figure. A fortnight later on a warm summer's day it was counted again when the figure had dropped to 9,280,000. No opportunity for further corroboration of this red cell variation in accordance with the atmospheric temperature has been possible, but will be carried out as soon as the opportunity presents itself.

THRILL. Thrill is less commonly present than where the stenosis is unaccompanied by interventricular leak. Reverting to Abbott's series - with the ventricular septum closed, thrill was present in practically 48% of cases. With both septa open in only 16.5%, whilst in the cases with a closed foramen ovale and an open ventricular septum, in only 4 out of 44 or 9.1%. This has been ascribed by Rolleston (120) to the ease with which the right ventricle can pass its blood stream through what is usually a very large leak, even admitting the thumb or index finger, and also to the large aorta over-riding the leak. When present it is felt in the same area as in pure uncomplicated stenosis, or lower, even near to the apex, as in case 14 (Taylor); it is however, neither so pronounced nor

so prolonged as in pulmonary stenosis, uncomplicated by a defect in the interventricular septum.

RADIOLOGY.

The radiological picture of the Tetralogy of Fallot is characteristic. Where the pulmonary obstruction is of high grade and accompanied by ventricular septal defect, the pulmonary artery is small and hypoplastic, and no enlargement of the pulmonary arc is seen. In many cases this border of the heart is markedly concave, which combined with a broad aortic shadow gives an appearance not simulated by any other condition except pulmonary atresia with septal defect. On the right side the enlargement of the right auricle is seen and above that the greatly broadened aortic shadow, lying further to the right than in the normal heart. In the oblique views the aorta and the right auricle are seen to be enlarged. The aortic bed on the oesophagus is noted to be larger than normal but the right branch of the pulmonary artery takes no part in its formation as it does in some few cases of the valvular type of pulmonary stenosis which are unaccompanied by septal defect, or in mitral stenosis, or in patency of the ductus arteriosus where dilatation of the pulmonary artery is a characteristic feature. Plate 19 shows how even an unusual degree of dilatation of the right auricle may be present without exercising any pressure on the

oesophagus.

MURMUR. Laubry and Pezzi (80) suggest that the component lesions of this association of anomalies, as described by Fallot, give rise to their respective murmurs and that the auscultatory signs characteristic of each defect can be demonstrated in the one individual; that is by no means always possible. It is true that a low localisation of a precordial bruit, when accompanied by other signs indicative of apulmonary stenosis, would suggest a combination of a ventricular septal patency and an obstruction at the pulmonary valve. Similarly a bruit heard maximally at the 2nd left space, conducted upwards and outwards to the left clavicle and accompanied by a weak pulmonic second sound, if associated with an aorta shown to be enlarged and to lie well over to the right, would suggest a ventricular septal leak even though no auscultatory evidence of this patency were audible. As a general rule however, it is only by taking into consideration all the facts collectively, as revealed by examination clinical, radiological and electrocardiographical etc. that one can determine such lesions as are present. The murmur may be heard near to the apex, and at such times may be difficult to distinguish from mitral disease. The septal murmur, is however, situated at a higher level than that of the mitral lesion, and is more prolonged and

is rougher, and further a mitral murmur is never heard in the neck; moreover with the congenital lesion some evidence of the pulmonary valve defect is usually present. It is admitted that Maladie de Roger may give rise to a bruit heard over the pulmonary area, though its origin be in the interventricular septum. A further differentiation of lesions, although one upon which too great weight must not be placed, is the hearing of a systolic bruit in the neck, suggestive of an over-riding of the septum by the aorta.

It cannot be too strongly insisted upon that too great attention is commonly paid to a bruit heard over the right side of the neck as indicative of either aortic valvular disease or a dextroposition of the aorta. A murmur may be heard there in the absence of all cardiac disease, and in fact in many cases of the present series murmurs of septal or pulmonary valve origin were heard on either side of the neck in the absence of any deviation to the right of the aorta. If there is a bruit in the neck sufficient to lead one to suspect disease of the aortic valve or misplacement of the aorta, then there will be other signs of more certain import, e.g. increased pulse pressure, hypertrophy of the left heart, an aortic diastolic murmur, thrill in the episternal notch of the neck, marked carotid and axillary pulsations &c. indicative of the

lesion present. The auditive evidence of the defect may be extremely slight, or rarely completely silent as in a case of Muir's (personal communication) of a child deeply cyanosed but in whom there was no auscultatory evidence whatever of disease, yet who showed at autopsy a complete association of defects as in Fallot's Tetralogy. Reuben and Steffen (116), together with their own case, culled 24 further similar cases from the literature; such are frequently examples of pulmonary atresia. Examination of cases of Fallot's Tetralogy in their latter days with commencing heart failure, may disclose only the faintest murmurs.

Laubry and Pezzi (80) ingeniously suggest that where there is a right-left shunt, with the flow of blood towards the deeply buried left ventricle, no murmur is audible, the left ventricle being no longer that part of the heart abutting on the chest wall, and so able to carry the murmur up to the stethoscope. The pulmonary second sound, is, like in the sister condition, weakened or absent.

B.1. PULMONARY IMPERFORATION, WITH SEPTAL DEFECT.

When this association is present, one is dealing only with a more severe grade, but of the same type as in pulmonary stenosis with a defective ventricular septum, except that with complete atresia the narrowing may affect the whole or any part of the pulmonary

arterial system from the conus to the artery. The pulmonary systolic murmur is absent, and the aorta rises almost entirely from the right ventricle, and the only path of entry into the lungs is via the ductus arteriosus, which is invariably widely patent.

B2. PULMONARY IMPERFORATION WITHOUT SEPTAL DEFECT.

It does not appear probable that a developmental condition occurring after the eighth week could so alter the anatomy of the pulmonary valve as completely to occlude its lumen. The origin of these defects must lie in

1) An endocarditis occurring after the ventricular septum is closed of such a fungating type as altogether to occlude the pulmonary orifice, or
2) An endocarditis implanted on an already rudimentary and deformed valve, obliterating the lumen of the valve and artery.

To allow of even a minimal quantity of blood to enter the lungs the foramen ovale and the ductus arteriosus are always widely patent. The heart is greatly enlarged, and the lumen of the right ventricle small, which together with thrill and a systolic murmur over the precordium is the only pathognomonic sign in those infants, who die in most instances either in utero or with extreme cyanosis during the first few hours or days of life.

P A R T 3.

PROGNOSIS.

The determination of the expectation of health and life of a child suffering from congenital heart disease, is to the parent a matter of paramount importance, and to the physician a question of the greatest difficulty. There are certain general principles which apply here as in all cardiological eventualities, such as the response of the patient to the routine efforts of his daily life, his fundamental constitution and his social surroundings. Further than this it is necessary to assess the mechanical effect produced on the heart and circulation by reason of the defect, such pathological conditions as the patient is more liable to by reason of his defect, and the strain imposed by such physiological occurrences as menstruation and pregnancy.

LOCAL MECHANICAL EFFECTS OF THE LESION.

These have been fully considered in the previous text but may well be summarised here.

1. Pure ventricular defect. The presence of a pure ventricular septal defect of small calibre will impose little if any, additional burden on the heart. That in the majority of such anomalies the right ventricle is hypertrophied is admitted; why this is so has been discussed above in the description of the individual lesion. That, in the absence of any

obstruction at the pulmonary outlet this hypertrophy need not be marked, is seen by the frequency with which an electrocardiograph fails to show any right sided preponderence. If the hypothesis is correct, namely, that right ventricular enlargement in Maladie de Roger is in many cases indicative of a minor degree of tricuspid incompetence, the additional burden to be borne by the right heart will be so much the greater.

2. Pure pulmonary stenosis. The local effect on the right heart will be commensurate with the degree of obstruction present. Where slight, then no greater strain will be thrown on the right ventricle than by a ventricular septal defect, but as the degree of obstruction increases so will this strain be correspondingly greater, until where complete or almost complete imperforation is present, the right ventricle is unable to cope with the demands made on its resources, and death occurs either in utero or in the early days or hours of life.

3. Pulmonary obstruction with septal defect. The effect here on the right ventricle is not in all cases the sum total of the two previous conditions. The superimposition of the wide mouthed aorta provides an easier path of exit for the right ventricular content than the narrowed pulmonary artery. Further, in many cases the interauricular septum is defective which tends to ease the strain thrown on the right ventricle.

It will thus be seen how in practically all grades of the two defects under review here, the heart is subjected to some additional strain. This may not be great and may be easily borne by healthy children and young adults. Nevertheless there is a constant withdrawal on the balance of the reserve force of the heart. It is not by reason of this strain, however, that the expectation of life on the part of these children is shortened but as we shall see in the following paragraph from the greatly increased susceptibility of these individuals to bacterial endocarditis.

To what pathological conditions is the child more liable by reason of the cardiac defect.

1. Bacterial Endocarditis. The frequency with which such defects become the seat of bacterial endocarditis is best seen by referring to Abbott's statistical table of 850 cases of congenital cardiac disease, all of which have been autopsied.

Of 59 cases of primary interventricular septal patency, endocarditis was present in 22 individuals or 38.8%; from among 60 cases of primary defect of the interauricular septum on 18 occasions or 30%; of 89 cases of pulmonary obstruction complicated by septal defect it was met with 28 times, or 31.4%. It is thus seen with what readiness such lesions are susceptible to a secondary endocarditis. It is a matter of inter-

est that death was stated as being primarily due to bacterial endocarditis or endocarditis only once with interauricular septal defect, 9 times with interventricular septal defect, and on no occasion where pulmonary obstruction complicated by septal defect was present. No case of pulmonary stenosis with entire septa has been included here as in all of them endocarditis was present, it being assumed that foetal endocarditis is the causal factor of this defect.

2. Pulmonary tuberculosis. It is commonly supposed that there is an increased liability on the part of these patients to pulmonary tuberculosis. Personal experience has so far failed to corroborate this view and Laubry and Pezzi (80) state that in no case of congenital heart disease known to them has the tubercle bacillus been found in the sputum. Abbott (1) does not record the incidence of death from tuberculosis in her statistical record but reports a history of tuberculosis as occurring twice in her series of auricular septal defects, five times in her cases of ventricular septal patency, in three cases out of seven where pulmonary stenosis was unaccompanied by septal defect, and in twenty individuals out of a total number of 116 with pulmonary obstruction complicated by septal patency. These figures are certainly above the average incidence of tuberculosis and tend to show that there is without doubt, an increased susceptibility to

pulmonary tuberculosis in such children.

3. Bronchopneumonia & bronchitis. The frequency with which death results from these causes is comparable with that from endocarditis. In the present series pneumonia had been survived twice, - cases 9 & 15. In the cyanotic group bronchitis is of frequent occurrence and is commonly the principle complaint of the patient.

4. Paradoxical embolism. With a large septal leak, especially of the auricular septum, through which patency the blood from the abdomen and lower extremities chiefly flows, the danger of paradoxical embolism is ever present. Its occurrence has been reported upon on page 75.

RESPONSE TO CERTAIN PHYSIOLOGICAL EVENTS.

Puberty & Menstruation. The period of puberty is one where endocrinal evolution is typified in physical and temperamental change. It is however, a normal process during which there is no added strain thrown upon the circulation, and experience has not indicated that, apart from the increased necessity for rest and sleep during these years, there is any undue danger to the heart. Eger (35) suggests that the menstrual period, by reason of its diminution of the blood volume and reduction of the blood cell count, exercises a beneficent action on the patient. It is not thought that the effect is more pronounced than in

the normal person.

Pregnancy. Pregnancy must entail increased work on the part of the heart and as such is to be avoided and even terminated. With some it is borne without hurt, in many cases unaccountably so. Laubry and Pezzi (80) report personal experience of a girl of 25, a sufferer from pulmonary stenosis who survived five pregnancies and three abortions. In the presence of congenital or acquired mitral stenosis auricular septal defect appears to improve the prognosis,- c.f. cases reported on page 76. There is however, an ever present danger of a paroxysmal attack of dyspnoea and cyanosis with sudden death.

Borrowing terms descriptive of the causal factors of cyanosis one might describe as "determining factors" in the prognosis of congenital morbus cordis, the degree of right sided hypertrophy, the increased susceptibility to pulmonary tuberculosis and pneumonia, and the greater hazards of pregnancy. As modifying factors raising or lowering the morbidity or mortality threshold limit, might be described the hygienic surroundings of the patient, his hereditary physical birthright and the stresses and strains to which he subjects himself or is subject to.

TREATMENT.

In the present state of our knowledge of the origin of congenital heart disease we have no known

active means at our disposal for its prevention. In our etiological survey of the condition the possibility of alcoholism, syphilis, consanguinity and familial tendency has been considered. Until we arrive at a more certain opinion on the part these factors play we must strive to prevent marriage of or procreation from such individuals. This implies also the taking of all known precautionary measures against rheumatic and streptococcal infection with special attention to the care of the teeth and throat, and vigorous anti-luetic treatment where serological examination has shown syphilis to be present.

Treatment of the discovered lesion should be at once preventive and alleviative. Preventive because the occurrence of a raised pulmonary pressure or of a superadded rheumatic or subacute bacterial endocarditis may alter the immediate and the more distant outlook. The heart must be exercised as must any other muscle of the body but the patient must keep within the limits of his reserve force both as regard the avoidance of harmful exertion, physical and mental, and also, in so far as is possible, as regards the avoidance of bodily illness. By this it is not meant that the normal enjoyment of a healthy boy or girl should be unduly interfered with. Too frequently are these children deprived of all games and mental stimulus. So soon, however, as it is seen that any given degree

of exertion or exposure to cold induces an abnormal shortness of breath, listlessness or alteration in the colouring of the skin, so soon must it be realised that the limit of strain which may safely be imposed on the heart has been reached.

Palliative treatment is called upon for the relief of the paroxysmal attacks of dyspnoea and cyanosis so commonly met with. Warmth is of the first importance, the significance of which is to be seen in the alteration in the intensity of cyanosis as seen on a cold or hot day. Venesection is helpful but is secondary in its usefullness to the administration of oxygen, either by means of the Haldane apparatus or in an emergency through a Clover inhaler held tightly to the face. The giving of oxygen intravenously or intra-arterially is yet in the experimental stage. As explained earlier in the text, the administration of oxygen in congenital heart disease will not raise oxygen saturation to the normal level, especially where the cyanosis is due to venous arterial shunt but should always be given in an emergency because of the frequent presence of a lung factor. The experimental work of Cutler, Levine and Souttar has opened up vast fields for the relief of mitral stenosis. With congenital heart disease the incidence of a valvular type of pulmonary stenosis is of less relative frequency, and little hope can be held out in

the developmental types of pulmonary obstruction.

Such methods of treatment may have to be had recourse to, but the necessity for their employment would be much diminished were the child reared under optimum hygienic conditions and wisely shielded from such complications as tend to raise the pressure of the lesser circulation or lower that of the systemic system, thus favouring the occurrence of a venous-arterial shunt.