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Ever since the day that man discovered the heart to be the most vital portion of the body, his innate curiosity has driven him to study it. It is only natural too, that we take a special interest in the rare mistakes in nature's plan that result in what we call congenital cardiac anomalies or congenital heart disease.

As physicians, an understanding of congenital heart disease is necessary to be able to give an intelligent prognosis. In the future, when our methods of treatment of these conditions improve, the understanding of them will be more necessary than ever.

This paper makes no attempt to be an anatomical dissertation, nor an embryological study, nor a theoretical discussion. Nor is it an extensive study of all reported cases or writtings on the subject. Congenital heart disease has been treated in the above manners by many authors. An attempt has been made, however, to select the more important facts about the subject and to assemble them into a short paper that may be of use to the clinician not well versed in the present day concepts of congenital cardiac disease.

Abbott (1932) gives a very well worded definition of congenital heart disease. "Congenital heart disease is that condition in which, owing to the disturbance of development or disease occuring in intra-uterine life, abnormalities in the anatomical structure of the heart or great vessels exists, which set up irregularities in circulation, interfering in certain cases with the adequate oxygenation of the blood in the arterial or capillary stream." The cyanosis complex which is so commonly associated in the minds of many with congenital heart disease is present only when there is a permanent venoarterial shunt or stasis in the venous system. More often, the defect gives only cardiac strain which gives no cyanosis, but only a group of atypical signs, and may lead to cardiac failure.

Congenital heart disease is usually considered a rarity. However, as our clinical knowledge of these conditions grows, more cases are noted. Altho the average physician may see but few such cardiac patients in his practice, most cardiac specialists have a group of congenital heart patients in their practice.

In the Boston Public schools (Robey, 1927) a survey of those with cardiac disease showed that 11% could be diagnosed as congenital, altho but few of them had any symptoms. Autopsies on those dying in childhood of cardiac disease reveal that from 2.8 % to 11.8 % of these deaths have been due to congenital anomalies of the heart. These figures vary greatly with different institutions and authors (Abbott, 1932). Wilson, Lingg and Croxford (1928) report from a heart clinic for children between the ages of 2 and 22 years, saying that 11.2 % of their cases, or 56 cases in all were diagnosed as congenital cardiac disease. White and Jones (1928) compiled statistics on 3000 cases; 1000 from hospital and dispensary practice, 1000 from those seen in consultation and 1000 seen in private practice. In these 3000 cases they found 37 which they diagnosed as congenital heart disease. This was 1.6 % of all the organic cases, and 1.2 % of the total

group of 3000.

It has been found that some of the congenital lesions of the heart are more common in one sex than the other, and apparently for no reason at all. In the 37 cases of the series of 3000 of White and Jones (1928), 56.7 % were in males and 43.3 % were found in females. Poynter (1919) found that the percentage was 55.69 % in males.

In the same 37 cases of White and Jones (1928), 27 % were in their first decade of life, 40.6 % were in their second, 18.9 % were in their third, 10.9 % were in their fourth, and 2.7 % were in their fifth.

Wilson, Lingg and Croxford (1928) found that they discovered 9 % of their congenital hearts in children during the first year of life, 21 % between the second and fifth years, 70 % between the sixth and thirteenth years.

To indicate the average number of congenital hearts that might be found at autopsy in a general hospital, we give below the report of Houck (1929). It reports the number of congenital hearts found in 5000 consecutive, routine post mortem examinations at

the Massachusetts General Hospital.

Patent foramen ovale	601 cases
Patent ductus arteriosus	96
Hypoplasia of the aorta	38
Interventricular septal defect	5
Bicuspid aortic valve	3
Pulmonary stenosis	3
Malformation pulmonary valve	2
Coarctation of the aorta	2
Congenital aortic stenosis	1
Malformation of aortic valve	1
Congenital idiopathic hypertrophy	1
TOTAL	753 cases

It was not known how many of the 601 cases of patent foramen ovale were mere anatomically patnet, nor what percent could be classed as tru functional anomalies.

The comparative occurance of the different lesions is better judged from Abbott's (1932) statistical study of 1000 cases of congenital heart. These figures are given in the section dealing with classification.

There are two possible causes of congenital cardiac disease. Intra-uterine fetal endocarditis and endarteritis causes some of the cases of aortic and some of the pulmonary stenosis and atresia. How the fetus is infected by the mother thru the placenta is not known for sure, but it is known to occur late in intra-uterine life, usually after the heart is well developed with all of its valves and septa (Abbott, 1932).

The other possible cause of congenital heart disease is distortion of the development of this organ. The distortion may result from fetal disease such as syphilis, rheumatic endocarditis, and the like. Syphilis has been incriminated in a few cases. However, this mode of altering the development has not been proven (Abbott, 1932).

It is quite possible that the distortion is due to some inherent property of the germ plasm. This thought has been prompted by several observations. Other congenital anomalies have often been found with congenital heart disease which are known to be inherited, such as polydactylism, hemophilia, clubbed foot, bilateral dislocation of the hips, fetal lobulation of the kidneys, and patent urachus (Abbott, 1932). Too, the same lesion

has been found in identical twins. Smith (1929) reports finding patent ductus arteriosus in each of identical twins, and dextrocardia in each of another set of identical twins.

Distortion of development is thought to be most often brought about by conditions surrounding the embryo. It has been found that congenital heart disease is more common among siblings than near ancestry. Wilson, Lingg, and Croxford (1928) report that five of their 56 cases had brothers or sisters with congenital hearts. Often there is evidence of some infection in one or both parents. Certain influences on the mother in the first few weeks of pregnancy are occasionally held responsible for the causing of the congenital cardiac anomalies. The fact that there are often associated anomalies (in 18.8 % of cases) is used to uphold this contention (Abbott, 1932).

It is noteworthy that 9.3 % of Mongolian idiots have congenital hearts (Von Hofe, 1922), and that 6 % of congenital cardiac patients are mongolian idiots (Abbott, 1932).

Poynter (1919) believes the conditions about the embryo cause the distortions in development as

pointed out above. He states that the distortion is due to changes in the rate of oxidation at the specific time a certain anlaga is prominent in development. He was able to produce various congenital cardiac anomalies in chicks by lowering the temperature of the eggs to near freezing for a time and at a certain time, before continuing the incubation. The--- "only action of the cold is to inhibit oxidation and so disturb the growth balance between the parts." He produced the same results by incubating the eggs in an atmosphere low in oxygen. So, he establishes the contention of Mall that congenital hearts defects are the result of an insult to the embryo.

There are a number of symptoms which may be caused by congenital cardiac anomalies. Most common are cyanosis, dyspnea, dyspneic attacks, weakness, headache, syncopic attacks, cold feet and hands, parasthesias, epistaxis, hemoptysis, cough, abnormal susceptibility to infection, palpitation, biliousness, etc.

Cyanosis is not found in the group I of anomalies (see classification). In group II, cyanosis is only transit or ternimal. In group III there is cyanosis. However, it may not develope at birth, and may be evident only after some pulmonary infection. Often it does not develope before the indificual is two years old (Abbott, 1924).

Cyanosis is a bluishness of the skin, best noted on the lips, and at the tips of the fingers and toes. It is modified by the thickness of the skin, variations in the skin pigments, the oxygen disassociation curve, etc.

The determining factors of cyanosis as worked out by Lundsgaard and van Slyke (1923) are as follows: Diminished oxygenation of the blood in the lungs due to alterations in the alveoli or lowered oxygen tension of the inspired air, known as the "1" factor: a permanent

venoarterial shunt, the alpha factor; increased peripheral de-oxygenation of the blood due to either stasis or increased oxygen usage, the "D" factor; and a high hemoglobin content of the blood, the "T" factor. Usually these factors work in pairs, the "1" and "T" together and the alpha and "D" together. The later pair are the factors usually active in producing cyanosis in congenital heart disease.

In experimental work methods have been worked out to calculate the volume of the venoarterial shunt. It has been found that it is necessary for one third or more of the venous blood to be shunted into the arterial circulation to produce a cyanosis of sufficient degree to be recognized clinically. This explains why so many congenital heart cases do not have cyanosis. For many years, and in the minds of many today, the cyanosis was thought to be always due to the venoarterial shunt, but as far back as 1844, Stille (1844) proved that venous stasis was also a factor, and in many cases, the only cause.

The color of cyanosis varys a great deal from a questionable tinge of the nail beds to a dark purple color of the entire body.

Dyspnea, especially on exertion, is the second most importnat symptom. At first it is a compensatory measure, and attempt to alleviate the embarrassed circulatory system. Later it is a true dyspnea due to the capillary dilatation and thickening of the alveolar walls which gives decreased respiratory area and decreased vital capacity (Abbott, 1932). Dyspnea is found in all degrees (White, 1931).

Dyspneic attacks in the form of paroxysmal exacerbations of the respiratory distress with increased cyanosis are found in some types of congenital heart cased. These attacks are probably due to a sudden increase in the volume of the venoarterial shunt. They may last for a few seconds or for hours.

Synocopic attacks, and also epileptiform attacks sometimes are found. They are due to cerebral anoxemia.

Other symptoms of congenital heart lesions are common, but are in no way characteristic.

Many cases of congenital heart disease are discovered by the finding of atypical physical signs. These may be of value in making the diagnosis of congenital heart disease and also in the naming of the particular type of lesion. Such findings include; cyanosis, cyanosis retinae, clubbed fingers and toes, decompensation, precordial bulging, paradoxical embolism, malnutrition, murmurs, thrills, alterations of the pulmonic second sound, alterations of cardiac shape and size on percussion and teleoroentgenograms, abnormal fluoroscopic shadows, abnormal cardiographic tracings, polycythemia, abnormal blood pressure, etc.

Cyanosis may go unnoticed by the patient and be found by the physician. Cyanosis retinae is found earlier than clubbing of the fingers. It is a condition in which the retina is a rosy, bluish hue. The capillaries are dilated, elongated and increased in number, and the arterioles and venioles are wavy and dialted. It is found in some degree in about one half of congenital heart patients (Abbott, 1932).

Clubbing of the fingers and toes is a general thickening of the distal parts of the body due to the low oxygen tension in the capillaries. There are changes in the capillaries and the soft tissues. It

takes much longer to develope than cyanosis, and so is not found in early infancy. It is found in all cases of long standing cyanosis.

Decompensation is found only in cases in which the cardiac strain has persisted long enough to weaken the myocardium to a stage at which it no longer maintains adequate circulation. It differs in no way from the decompensation found in any other form of cardiac disease.

Precordial bulging is found in cases in which the heart has been enlarged over a great period of time. It too, is in no way specific.

Rare paradoxical embolism which consists of embolism in the arterial tree from venous thrombi, is found in cases with defects of the interauricular or interventricular septum, especially the former. Occassionally such emboli set up brain abcesses. When paradoxical embolism can be definitely diagnosed, it proves the existance of a defect in the cardiac septa.

A tall slender habitus with a delicate frame, undevelopment, malnutrition, and delayed puberty and pallor are signs which point to aortic hypoplasia and

are often found in certain congenital heart patients. It is not specific, but often quite suggestive, (Abbott, 1934).

The murmurs found in congenital cardiac patients are almost always systolic, and are found at abnormal sites. Often they are course and rough in character, lound and high pitched and often holosystolic. Too, they can be found in young patients where there is no history to even suggest preceeding, causative disease. The transmission of these murmur is often abnormal and characteristic of the condition (Abbott, 1934).

The pulmonic second sound is often altered, particularly with lesions of the pulmonary artery and its semilunar valves. It may be absent, weak or accentuated.

Thrills are found in many of the congenital lesions of the heart. Often they are very course and forceful. Usually they are localized over the lesion but may be transmitted in a characteristic manner. They are found in cases of stenosis and shunts between the venous and arterial circulations.

The size and shape of the heart is often altered to the roentgen ray and percussion. Holman (1925) says the heart chambers respond by dilatation and hypertrophy to increases in the volume flow of blood thru them In pulmonary stenosis and atresia the pulmonary are is small or missing. The shape of the upper part of the heart shadow is changed in many of the anomalies of the aorta. Enlargement of the right or left ventricle is found in many of the lesions. Ofcourse, the teleoroentgenogram is quite characteristic in dextrocardia.

Fluoroscopic examination of the heart at times reveals pulsating hilar shadows, a condition known as "danse hilaire." It is caused by a high pulse pressure in the pulmonary circulation. As Hubeny (1920) points out, the pulmonary arc can also be seen to pulsate, and when compared to the pulsation of the aortic arch, it is greater in its movement instead of less as in normal chests. These hearts usually show dilated lung hila on simple plates. This sign is found in defects of the interauricular septum, patent foramen ovale, and others.

Pendergrass and Allen (1934) have described a displacement of the trachea to the right and post-

eriorly during inspiration in transposition of the great trunks on fluoroscopic examination. Enlargement of the thymus which gives the same sign must be ruled out first. However, he found in one third of the cases with this sign there was no transposition, but instead, multiple other defects.

The electrocardiogram is often useful in making the diagnosis. Ofcourse, in congenital heart block the disassociation between the ventricular and the auricular beat is found, with a slow ventricular rate.

McCullock (1916) pointed out that the electrocardiographic tracings in congenital heart vary from the normal only as the anomalies secondarily affect the heart chambers.

In patent foramen ovale there is a high P wave, due to the hypertrophy of the auricles (Abbott, 1934). In pulmonary stenosis there is a right preponderance and usually a high P wave (Alexander, Knight, and White, 1925). In general, there is left preponderance in lesions of group I, the acyanotic group and right preponderance in lesions of the third group, the cyanotic group (Abbott, 1934). Inversion

of Lead I and reversal of leads II and III means mirror-picture dextrocardia (White, 1931).

In the later stages of some of the cyanotic congenital heart diseases there is a polycythemia. It may vary from five to twelve million. It is a compensatory mechanism, but usually is a bad prognostic sign (Abbott, 1932). Polycythemia may cause epitaxis (Leaman, 1933). Usually there is a parallel rise in the hemoglobin content of the blood. It may vary from 100 % to 200 % altho it only rarely reaches the later figure (White, 1931).

The blood pressure is usually not at all characteristic. It is, however, in coarctation of the aorta in which there is hypertension in the upper extremities and hypotension in the lower. White (1931) says that in general the blood pressure in congenital heart disease is apt to be low with a low pulse pressure. In widely patent ductus arteriosus, the pulse is full and the pulse pressure is high as a result of a low diastolic pressure.

Congenital cardiac anomalies can be classified on an embryonic, anatomical, pathological, or clinical basis. However, only the last is useful in the hands of the practicing physician. Abbott (1929, 1932, 1934, Abbott and Dawson, 1924) has developed such a classification and it is her latest form which is given below.(Abbott, 1934). It is followed, with minor alterations by most clinicians.

The clinical classification is based primarily on the symptom of cyanosis, and in this way, all lesions are divided into three groups; those which have no cyanosis, those with transit or terminal cyanosis, and those with true cyanosis.

In group one, the acyanotic group, there are no abnormal communications between the venous and arterial circulations, but the lesion is apt to be the seat of strain. These lesions are usually minor, and usually left-sided lesions. They are usually due to an arrest of development at a late stage.

Group two, known as the "cyanosis tardive" group, consists of lesions in which there are communications between the arterial and venous systems, which allows an arterio-venous shunting of the blood. The

transit and terminal cyanosis results when, due to alterations in the normal pressures within the heart, the abnormal flow thru the shunt is reversed and becomes a veno-arterial shunt. These lesions are due to arrests in the development before the septa are formed or the passages are closed. In patent ductus arteriosus and defects of the aortic septum the cyanotic attacks are more common as the pressures here are more subject to fluctuations. On the other hand, the small defects of the interventricular septum cause cyanotic attacks only very rarely and terminal cyanosis comes on very late.

Group three, the cyanotic group includes lesions in which there is a permanent veno-arterical shunt and increased deoxygenation in the capillaries. These lesions are due to arrests or alterations in the development very early in fetal life. Also included in this group are certain right-sided lesions without interventricular septal defects which have been caused by fetal inflammation after the septa are closed. Then the cyanosis appears late and is due to the "D" factor plus, perhaps a small alpha factor, a patent foramen ovale. In group three there are often associated lesions which have developed after the primary lesion as a compensatory measure. Such lesions

are patent ductus arteriosus and patent foramen ovale.

Below is the classification as used in this discussion, taken with slight modifications from Abbott's (1934) classification. With each condition is given the average age that the time of death of the patients with that lesion, and the number of such cases which Abbott has included in her studies.

In general, the lesions are listed in each group in accordance with their average degree of oxygen unsaturation in the arterial blood.

		Age	Cases
I	Acyanotic Group		
1	Coarctation of the aorta	36 yr	70
2	Double and right aortic arch	32	19
3	Anomalous semilunar cusps	31	44
4	Pericardial defects	45	60
5	Anomalous atrioventricular cusps	27	10
6	Pulmonary insufficiency and dilatation	28	8
7	Subaortic stenosis	19	12
8	Anomalous septa in auricles	23	14
9	Congenital arteriovenous aneurysm	21	6
10	Congenital aortic or mitral stenosis	3	17
11	Congenital heart block	7	3

12	Primary congenital hypertrophy	4 mo	15
13	Ectopia cardis	1	7
14	Dextrocardia	8 yr	29
II	Cyanose Tardive Group		
1	Localized defects of interauricular septum and patent foramen ovale	28 yr	73
2	Localized defects of interventricular septum	42	7
3	Patent ductus arteriosus	29	92
4	Localized defects of the aortic septum	15	22
III	Cyanotic Group		
A	Moderate Cyanosis		
1	Pulmonary stenosis with patent foramen ovale	18	16
2	Tricuspid atresia with ventricular septum defect and transposition of great trunks	56	1
3	Dextroposition of the aorta with defect of ventricular septum	13	10
4	Cor biatriatium triloculare	7¾	13
5	Persistent ostium atrioventriculare commune	1½mo	10
B	Marked Cyanosis		
1	Pulmonary stenosis with ventricular septum defect	12½yr	83
2	Pulmonary atresia with ventricular septum defect	6¼	30

3	Persistent truncus arteriosus	4	21
4	Complete transposition with ventricular septum defect	1½	16
5	Cor biloculare with transposition	15½	2
C	Extreme Cyanosis		
1	Pulmonary atresia, patent ductus and foramen ovale	2½mo	10
2	Transposition of great trunks, patent ductus and foramen ovale	1¾	31
3	Mitral atresia	10	5
4	Tricuspid atresia	6	15
5	Cor biloculare	1½	7
6	Aortic atresia	4 da	12

Coarctation of the aorta is a condition of stricture or narrowing or even atresia of the aorta between the left subclavian artery and the ductus arteriosus. There are two types, the so-called adult form and the infantile form. Coarctation of the aorta is a rather common type of acyanotic congenital cardiac anomally. Abbott (1928) and Hamilton and Abbott (1928) report on 200 cases they have collected. Strangely, they find it to be about three times as common in males as in females. Blackford (1928) states that it is found in one in about every 1,550 patients going to autopsy.

The fetal type is a persistance or exaggeration of the fetal anatomical condition where the pulmonary artery, thru the patent ductus arteriosus and descending aorta supplies the lower part of the body. It is a narrowing of the entire isthmus, that is, the aorta between the left subclavian artery and the point of the ductus arteriosus. There is little collateral circulation so that death usually occurs soon after birth because of this inadequacy. Hamilton and Abbott (1928) report the average age in nine cases as one and three-fourths months.

The adult type of coarctation on the other hand consists of a sudden narrowing of the aorta at the

point of the ductus arteriosus. It appears as tho there were a ligature about the aorta at that point. It is probably due to presence of abnormal tissue in the mural structure of the aorta at this point, similar to that of the ductus arteriosus, and which contracts after birth (Hamilton and Abbott, 1928). However, Blackford (1928) states that there is no evidence of ductus tissue in the aorta and that the coarctation is due to the absence, atrophy or imperfect development of the left fourth brachial arch which forms this portion of the aorta. Formerly, coarctation was thought to develope postnatally due to the pull of the ligamentum arteriosum as it matured and contracted, or, due to an obliterating endarteritis (White, 1931).

Postmortem examinations of these cases reveal a narrowing of the aorta between the left subclavian artery and the ligamentum arteriosum. Usually the narrowest point, or the point of the atresia is very near the ligamentum arteriosum. Usually the mural structure of the aorta is normal, tho it may contain considerable fiberous tissue. The aorta is usually dilated above the narrowing, and hypoplastic below it (White, 1931).

There is usually no symptoms until the heart,

begins to fail under its increased load. However, there may be many symptoms among which are violent headaches, sleeplessness, tinnitus, epistaxis, hemoptysis, thoracic, epigastric or abdominal pain, and long standing vomiting. Cyanosis is rare. In the later stages the symptoms of decompensation develope slowly and insidiously.

The physical findings are always quite numerous and sometimes startling. The collateral circulations is very marked, usually including the supra and infrascapular arteries, the internal mammeries, the intercostals, the highest intercostals and others. The superficial ones can be palpated as large, dilated, pulsating arteries, and serve as a very characteristic finding in coarctation of the aorta. In rare cases, the major portion of the collateral circulation is within the thoracic cage and in such instances, these pulsating collaterals are not found on palpation. There is an elevated blood pressure in the upper extremities and it may not be the same in the two arms. There is an unusually high pulse pressure (Blackford, 1928). When the pressure is taken on the lower extremities, it is low and with a low pulse presure (Abbott, 1932). On palpation, the pulse in the lower extremities is plateau shaped , and without the characteristic thrust. On most cases, the systolic pressure in the lower limbs is 100 mm. of mercury or below, while in the

upper extremities it is usually 200 mm. or over (Abbott, 1928). On auscultation, a long systolic murmur is found over the precordium which is transmitted to the back and to the vessels of the neck. There is also a loud systolic murmur in the left interscapular space (Blackford, 1928). A thrill may be palpated over the precordium. The roentgenogram shows a widening of the ascending aorta while the aortic knob is decreased or absent. If there are symptoms of cardiac failure, the left heart appears enlarged and in many asymptomatic cases, the entire organ is enlarged. Holt and McIntosh (1933) point out that an erosion on the lower border of the ribs caused by the hypertrophied intermammeries and intercostals, can be seen by the x-ray in adolescents and adults.

Rosler (1931) finds with the capillary microscope that in the skin above the constriction, as on the elbow, the capillaries show elongation and stretching, broad venous ends, sluggish flow and capillary aneurisms.

The ductus arteriosus is usually not patent (Abbott, 1932). Coarctation is usually uncomplicated by other lesions of congenital origin. Abbott (1928) found only 17 cases in 200 collected that were complicated. It is when the coarctation is extreme that the associated congenital anomalies are most common. The

most common associated lesion is bicuspid aortic valves which are found in about one-third of the cases. The aorta is often the site of thrombosis, rupture, endarteritis, dilatation and aneurysms. The terminal stages present the picture of ordinary cardiac failure.

The condition is usually discovered on finding the characteristic collateral circulation, the murmur, of the high blood pressure. The diagnosis ca be confirmed by the taking of the blood pressure of the lower extremities. Fray (1930) points out that the diagnosis can be made positive by a posterior-anterior-oblique roentgenogram which will actually show the break in the continuity of the aortic arch.

The prognosis varies a great deal depending upon the degree of constriction and the adequacy of the collateral circulation. All of the infantile type die in infancy, and many of the adult types also (Blackford, 1928). If the adult form survives infancy, the expectancy is good, the average in the adult group being 37 years (Abbott, 1932). Blackford (1928) found that of those living to adulthood, 40 % died between that ages of 16 and 30 years and more than 70 % died from cardiovascular causes. Death may be due to congestive heart failure, cerebral hemorrhage, thrombosis, bacterial

endarteritis at the site of the coarctation or on the associated bicuspid aortic valves, or rupture of the aorta above the constriction (Abbott, 1928). The most common among these are cerebral hemorrhage and rupture of the aorta (Abbott, 1932).

The only suggestion in the way of treatment is the prevention of cardiac strain and the removal of all foci of infection. Blackford (1928) insists that all activity causing cardiac strain should be eliminated from the patients life.

Double aortic arch is a condition in which both the right and the left aortae of embryonic life persist. This forms an arterial ring about the trachea and esophagus (Abbott, 1932).

The persistance of the right aorta instead of the left is another rare anomally. There have been other types of aortic anomalies described, but they are always isolated cases.

In all these conditions, which are extremely rare, the etiology is that of an idiopathic alteration in the usual development and regression of tissues in embryonic life. The pathology is usually only the altered gross anatomy. Usually there are no symptoms or findings, except perhaps a change in the aortic knob in the roentgenogram, from which it is conceivable a diagnosis might be made in some cases. The double aortic arch may cause dysphagia. The prognosis depends entirely upon the associated anomalies.

When the aorta and the pulmonary artery are normal, their valves seldom have anomalies. However, occasionally semilunar valves are found which are fenestrated. The fenestrations vary a great deal from insignificant points to apertures large enough to cause insufficiency. These might be the cause of cardiac strain and lead to symptoms of the same.

Bicuspid aortic and pulmonary valves are rarely found alone. Often, when they are present, they accompany a set of lesions which put it into the cyanotic group of congenital cardiac anomalies. However, Abbott, (1932) found that bicuspid valves in the aorta were most commonly found with the adult type of coarctation of the aorta. These bicuspid valves are often the seat of infective endocarditis. Abbott and Chase (1929) for this reason insists on the early removal of all possible foci of infection in those suspected of coarctation, aortic hypoplasia and congenital aortic valvular insufficiency.

Poynter (1919) reports cases of one, two, and four valve leaflets in both the aorta and the pulmonary artery.

White (1931) divides pericardial defects into three groups; total absence or small defects, herniations and diverticuli, and lastly, lack of normal attachments.

When the pericardium is absent, the heart lies free in the pleural cavity. It is very movable and can shift a great deal with respiratory movements and with changes of body position as can be seen by roentgen shadows and physical examinations. In ectopia cardia, the heart may or may not have a pericardial sac. The prognosis is much better in cases which do have. The common site for small defects is about the pulmonary artery. The size of these small defects varies a great deal. The total absence or defects in the pericardium allow easy passage of pulmonary and pleural infections to the heart so that pleurisy, effusion, pneumonia and other such conditions may easily lead to death. Too, with the increased movability of the heart, there is the possibility of kinking of the great vessels giving pain and even sudden death. Defects in the pericardium are diagnosible only when marked movability of the heart is found.

Herniation and diverticuli of the pericardium are not diagnosible or important. They may be the cause

of mediastinal tumors. Their orifices may become plugged, resulting in distension which may interfere with the heart action. The prognosis is fine as these seldom cause trouble and are not found until autopsy.

Lack of attachments of the pericardium is very rare and is diagnosed only at the autopsy. Usually the attachment at the diaphragm is the missing one. The prognosis is excellent (White, 1931).

There are several types of anomalies of the atrioventricular valves, as described below:

Congenital mitral insufficiency is very rare and is the result of hypoplasia of the valve leaflets. It might be diagnosed if the findings of mitral insufficiency are found in a young child with no preceeding disease to which it might be attributed.

Congenital tricuspid insufficiency is also very rare. In this condition, the tricuspid segment is only a fenestrated membrane. Too, it is also diaplaced downward so that part of the right ventricular musculature makes up part of the right auricular wall. This increases the incompetency. This condition might be diagnosed too, if insufficiency is found in those without preceeding causative disease.

A double atrioventricular orifice between the left chambers is very rare and is usually found in cases of double left auricle resulting from anomalous septa and anomalous entrance of the pulmonary veins into the left auricle.

Double tricuspid orifices is very rare. It may lead to valvular insufficiency (Abbott, 1932).

Congenital pulmonary insufficiency is die to the absence or defects of the pulmonary segment. Usually the roentgenogram shows a greatly enlarged pulmonary arc. This condition might be diagnosed if the signs of insufficiency are found in youth without an explaining history of causative disease.

Congenital dilatation of the pulmonary trunk and its branches is often associated with sclerosis of the pulmonary radicles. It may give signs of oxygen unsaturation. The right heart is greatly enlarged and the pulmonary veins may be very small. The foramen ovale may be patent. This congenital dilatation results from hypoplasia of the pulmonary artery mural structure, especially the media. There may be aneurysms of the pulmonary artery or small ones in the smaller arteries in the lungs. The roentgenogram shows a greatly enlarged pulmonary arc. (Abbott, 1932).

Subaortic stenosis results from a thin fibrous shelf which projects from the inner wall of the aortic conus about six millimeters below the semilunar valves. It is located at the junction of the heart and the primitive bulbus. Subaortic stenosis is rather rare and is the result of the arrest of development (Abbott, 1932).

The symptoms of this condition, as well as the physical signs are those of acquired stenosis. The symptoms are usually few but there is usually clubbing of the fingers and toes. There is a systolic murmur and a soft short diastolic murmur just after the second sound. Usually there is also a systolic thrill (Christian, 1928). As the patient grows older, this anomalous shelf is often the site of atheromatous changes.

Chiaris network is a network of fibers or a delicate membrane extending across the interior of the right auricle. It begins in the region of the eustachian or thebesian valve and crosses to attach to some point on the septum. Chiari believed it to be a mal-development of the valvula venosa sinistra or septum spurium. (Poynter, 1919). This rare anomaly may cause the formation of thrombi with resultant pulmonary embolism. In some cases a chord has been found in the right auricle estending from the anterior border of the fossa ovalis to the apex of the anterior mitral segment. In some cases a band has been found extending across the lumen of the aorta, probably a remnant of the primitive septum.

In some rare cases an anomalous septum divides the left auricle. This condition is usually found associated with a double mitral orifice and with the right pulmonary veins emptying into one left auricle, the left into the other.(Abbott, 1932).

Occasionally endocardial pockets are found. Most often they are in the upper part of the ventricles with their openings point toward the aortic valve. Endocarditis often begins in these pockets (Abbott, 1932).

Congenital arteriovenous aneurysms result when one of the coronaries arises from the pulmonary artery. Usually it is the right coronary. This condition leads to marked dilatation and aneurysm and extensive anastomosis because of difference in the pulmonary and systemic pressures thus connected and shunted.

There are other coronary anomalies such as altered number, distribution, size and origin. The seriousness of these lesions depends upon the effect they have on the cardiac musculature (Bland, White and Garland, 1933). There may also be a congenital absence of the left or the right coronary.

Aortic stenosis is very rare as a congenital anomaly. Most cases are of the subaortic type described above. They both give the same clinical picture and physical findings as acquired stenosis.

Mitral stenosis is also rare. It is associated with aplasia of the left ventricle and ascending aorta. The right auricle and ventricle are greatly enlarged and dilated. Usually the foramen ovale and the ductus arteriosus remain patent. Poynter (1919) believed it to be a result of inflammation. This condition is not consistant with long life, the average age reached by those in Abbott's (1934) series being 3 years.

This extremely rare anomaly has been diagnosed before birth (Aylward, 1928) (Dippel, 1934). The heart rate is usually about 50 best per minute. The electrocardiogram is, ofcourse, very characteristic (Leech, 1930).

Other anomalies are usually found associated with congenital heart block, especially defects of the interventricular septum which involve a portion of the bundle of His. Aylward, (1928) found congenital heart block in sisters born several years apart. The diagnosis in the youngest of the two was made before birth. Both had transit cyanosis during early infancy and evidence of cardiac enlargement.

In primary congenital hypertrophy of the heart, which is considered idiopathic, the shape of the heart remains normal, but the weight and size increase rapidly during the first few weeks of life so that it often may weight three times the normal. Carrington and Krumbhaar (1924) doubt if this condition is idiopathic. They consider some nine points which might be the cause of this hypertrophy.

To be placed in this class, there must be a total absence of degenerative processes on microscopic examinations. Clinically, it must be differentiated from mediastinal growth, and pericardial effusions (Emerson and Green, 1928). Infants with this condition rapidly develope a clinical picture of cardiac insufficiency, possibly with dyspnea and cyanosis. It fails to respond to digitalis. Death may be sudden, or as in slow cardiac failure. It is always fatal. Holt and McIntosh (1933) state that no child in which such a diagnosis has been made has been known to recover.

There are three types of ectopia cardis; cervical, pectoral and abdominal. The heart protrudes thru the medial line and ventral body wall and in most cases the parietal pericardium is absent beyond the chest wall. Usually there are other mal-developments and life lasts but a few hours. In some cases the diaphragm is defective and the heart lies in the peritoneal cavity. This in itself is no hinderance to long life (Poynter, 1919). The exact cause is not known, according to Poynter (1919).

Acardia is very rare and found only in non-viable monsters (Poynter, 1919).

Excluding the dextrocardia resulting from mere displacement of the heart to the right, there are two types, simple rotation and true dextrocardia. In the former, the heart lies in the right side of the chest, but the left chambers lie to the left and anterior while the right chambers lie to the right and posterior. It is usually accompanied by some other anomaly such as a common ventricle. The prognosis depends entirely upon the associated anomalies.

In true dextrocardia the heart is a mirror picture of the normal heart. It is usually associated with complete visceral transposition, altho there may be only partial visceral transposition. It can be diagnosed by physical examination, the electrocardiogram and the roentgenogram. The electrocardiogram shows and inverted picture in lead I and transposition of leads II and III (White, 1931). True destrocardia without general visceral transposition is usually accompanied by other cardiac anomalies, usually grave ones.

The two types of dextrocardia can be differentiated by the electrocardiogram and assuming the mirror type must be accompanied by a complete visceral transposition. Dextrocardia is clinically unimportant in itself and is no hinderance to longevity.

Under this heading are included all defects of the interauricular septum except small patent foramen ovale. A patent foramen ovale is not considered a true lesion until it is large and definitely of functional importance. Then it is considered as any other localized interauricular septum defect (White, 1931).

Statements concerning the physiological closing time of the foramen ovale vary a great deal among the different authors. Christie (1930) cleared this point up conclusively. He studied the time of closing of the foramen ovale and gives us the following figures deducted from 590 autopsied cases with normal hearts:

Age	% remaining patent
2 weeks	82.5 %
4	51.8
6	37.1
8	25.0
10	16.2
12	13.3
28	8.2
52	5.7

He concludes that the normal lenth of time for anatomical patency is about 12 weeks. He states that normally the

foramen ovale is physiologically impervious shortly after birth.

Auricular septum defects are the most common of all congenital heart anomalies, being found in about 36 % of all cases (Poynter, 1919). These defects are due to the failure in development of the primary septum or the secondary septum of the auricles and may be prompted by abnormal blood currents resulting from other anomalies in the heart and its great vessels.

In studying the pathology of these lesions we may find them to be only a defect in the septal wall. Some, however, show considerable thickening of the borders of the defect, and often the border shows signs of endocarditis.

The patient is usually slender, pallid, slow in development and with delayed puberty. They may show other signs characteristic of aortic hypoplasia. Often there are no symptoms whatsoever. There is transit and terminal cyanosis. There there is no cyanosis, there is an arteriovenous shunting; with cyanosis, a venoarterial shunting thru the defect. The later results when the right-sided pressure is increased to equal or exceed the left-sided pressure. Late in the condition, there

are signs of cardiac failure. In 43 cases Abbott (1928) found 4 cases with slight or transit cyanosis, 2 with moderate cyanosis, 1 with marked cyanosis, 13 with terminal cyanosis and 23 with absence of cyanosis.

The findings too, may be scarce or absent. Usually there is a presystolic and systolic precordial murmur of varying intensity, with its point of maximal intensity in the third or fourth left intercostal space, just to the left of the sternum. There may or may not be an accompanying mesocardial thrill. The above findings vary a great deal. Usually there is some cardiac enlargement, especially of the right auricle and ventricle. The right auricle may be greatly dilated. Occasionally the aorta is hypoplastic and the pulmonary artery is dilated. Since this lesion will allow passage of the contents of the right auricle into the left, it allows paradoxical embolism, that is, venous thrombi causing arterial, systemic embolism (Abbott, 1932).

When interauricular defects are complicated with acquired mitral stenosis, the defect is very large. The right heart is greatly enlarged, also the pulmonary artery and the great veins and the left auricle. Aortic hypoplasia also appears in this picture. The findings include a presystolic murmur and a systolic murmur in

the left third intercostal space, accompanied by a faint precordial thrill. The individual is underdeveloped and with other signs of aortic hypoplasia. There is a marked terminal cyanosis and decomposation (Abbott, 1932).

On roentgenographic study, the above mentioned enlargements of the right side of the heart may be seen, and alos a large pulmonary arc and a small aortic knob. Too, the lung hila will appear dilated and if watched under the fluoroscope, the hila appear to pulsate during systole (McGinn and White, 1933).

The diagnostic points in favor of interauricular defects are a tall, slender habitus, delayed puberty, and signs of aortic hypoplasia, a large pulmonary arc, right sided enlargement, dilated lung hila, a presystolic murmur and thrill in the third and fourth left intercostal spaces, and transit cyanosis (Abbott, 1932).

Individuals with this type of congenital anomaly usually live to middle life, and then die of cardiac decompensation. In Abbott's (1932) series, of 73 cases, the average age was 28 years.

This group includes all congenital anomalies in which there is an openning between the two ventricles, except gross defects which make the ventricles functionally a single chamber. It is a rather common type of anomaly, altho as a pure lesion it is not nearly as common as auricular septal defects. These defects are due to failure in the development of the ventricular septum.

Usually the defect is at the base of the interventricular septum just anterior to the pars membranacea, and opens into the right ventricle just below the tricuspid valve. Occasionally the defect is more anterior and opens into the right ventricular conus. Rarely, the defect is found in the lower part of the septum. The defect is usually small, circular or oval and about one or two centimeters in diameter and usually with thickened edges. The endocardium on the wall of the right ventricle opposite the defect is usually thickened and fibrous, indicating the point at which the stream from the left ventricle strikes (White, 1931). Usually there is little or no change in the ventricles as the volume and pressure changes resulting from the lesion are slight.

If the defect is larger than usual, the right ventricle is hypertrophied and dilated and the pulmonary

artery is slightly dilated. The left ventricle may be slightly enlarged (White, 1931).

Under normal circumstances, the higher pressure in the left ventricle forces a stream of blood thru the defect into the right ventricle with each systole and there is no cyanosis. At times of embarrassed circulation and cardiac failure, the pressure in the two ventricles may become equal, or the right-sided pressure may become the greater, and a transit or terminal cyanosis results. However, in this lesion, the cyanosis is apt to appear late and be of little or no consequence (Abbott, 1932).

The symptoms are few or none, unless there are associated lesions.

The findings are usually very characteristic. They were first described by Rogers and this congenital lesion and its clinical picture is often called by his name, "Maladie de Roger." Below is an extract of the summary of his original article as given by White (1931): "1. Interventricular openings do not cause cyanosis. 2. The murmur is long, starting in systole and covers both sounds. It is maximal in the upper third of the precordium and is not transmitted to the back or vessels

of the neck. It is accompanied by a harsh thrill. 3. The murmur is very constant, and never changes. 4. This condition is found in the nursing infant, before acquired disease is present in the child. 5. The prognosis is poor but they may live as long as normal individuals. 6. The exact diagnosis usually means persistant treatment. Here, refraining from treatment is a service to beth the patient and the doctor."

Abbott (1928) found in 34 cases, 6 with slight or transit cyanosis, 2 with moderate, 5 with terminal and 21 with no cyanosis.

The murmur is long, drawnout, harsh, loud, blowing sound, which may extend as far as to cover the second sound. The point of maximal intensity is usually in the third left intercostal space. It is not transmitted. There is an accompanying systolic thrill. The intensity of these findings varies inversely as the size of the defect. There may or may not be any cardiac enlargement on physical and roentgen examination. The electrocardiogram is normal, unless the defect affects the bundle of His, giving a congenital heart block.

Bacterial endocarditis on the border of the

defect and at the point of impaction on the opposite right ventricular wall may be complications. There may also be a slight deformity in the tricuspid valve leaflets. The most common associated congenital defects are pulmonary stenosis, dextroposition of the aorta, and right ventricular hypertrophy, which, if they are all present, make up the "Tretralogy of Fallot," a congenital heart lesion of the cyanotic group. This condition is discussed later.

The diagnosis can be made upon finding the characteristic murmur and thrill, without any cardiac symptoms.

The prognosis is good, the average age of Abbott's (1934) group of 7 cases being 42 years.

The ductus arteriosus should cease to function soon after birth and be converted to a fiberous cord in a few weeks. Just when it should anatomically close is not so well known. Christie (1930) found from studying 558 normal infant hearts at autopsy the following figures:

Age	% still patent
2 weeks	64.7 %
4	44.3
6	21.8
8	12.0
10	8.4
12	4.9
20	3.4
32	2.0
52	1.2

He concludes that 8 weeks is the normal lenth of anatomical patency of the ductus arteriosus.

Poynter(1919) found patent ductus arteriosus in slightly more than 25 % of all congenital hearts. It was present as an isolated lesion in about 3.7 % of all cases of congenital hearts.

When the ductus remains patent, it is usually

because of a need for compensation, as in transposition of the great trunks, pulmonary stenosis, and atresia, and coarctation of the aorta. At times, there is no apparent cause, and this patency must be called idiopathic. The degree of patency varies greatly as does the lenth and shape (White, 1931). If the patency is marked, it leads to pulmonary artery dilatation and right and left ventricular hypertrophy and dilatation, especially of the left ventricle. Holman (1925) states that hypertrophy of the ventricles is due to the increased volume of blood flowing thru the chambers concerned. So, when the patency acts as an arteriovenous shunt, there is left ventricular hypertrophy as the increased volume flows thru that chamber. When there is a venoarterial flow thru the ductus arteriosus, the right heart is the one with the increased volume of flow and so it hypertrophies. Since there is usually an arteriovenous shunting, this explains why the left ventricle is the one which usually hypertrophies.

Often there are attacks of transit cyanosis which are often accompanied with dyspnea. These attacks are the result of increased pulmonary system pressure which changes the arteriovenous shunt into a venoarterial shunt. This change can be brought about easily, thus explaining why these cyanotic attacks are common in this

condition. In infants, transit cyanosis is common because crying or holding the breath when nursing raises the pulmonary pressure. Abbott (1924) found in 64 cases there were 3 cases with slight or transit cyanosis, 4 with moderate cyanosis, and 10 with terminal cyanosis and in 37 cases it was absent.

In the infant there may be no murmur and when there is it is a short systolic one. In adults the murmur is very characteristic. It is systolic but prolonged thru systole or even thru the entire cycle. When it is continour there are systolic and diastolic accentuations. It is huming or machinery-like, and the point of maximal intensity is in the first or second left intercostal space. There is a prolonged systolic thrill in the first or second left intercostal space. Often the pulmonic second sound is accentuated. The murmur is transmitted slightly to the back, and only occasionally to the vessels of the neck (Abbott, 1932).

Often there is a small area of abnormal dullness in the first and second left intercostal spaces near the sternum on percussion. The x-ray studies show an enlargement of the pulmonary arc and occasionsome cardiac enlargement. Usually the left heart hypertrophies more than the right. Under the fluoroscope

the large pulmonary arc is seen to pulsate with each heart cycle. This can be compared to the aortic knob pulsation, and this condition, the pulmonary arc pulsation is much more noticable (Hubeny, 1920). The electrocardiogram usually show no preponderance. Occasionally there are calcerious deposites in the wall of the patent ductus which show up on x-ray examinations (Abbott, 1932).

When the ductus arteriosus is widely patent, the pulse is full, the pulse pressure is high and the diastolic pressure is low (White, 1931).

Patent ductus arteriosus is found in many of the complicated congenital hearts of the third, cyanotic group where it is a compensatory mechanism. The patent ductus is often the site of subacute bacterial endocarditis (White, 1931).

According to White, (1931) the diagnosis of patent ductus arteriosus is positive when the characteristic murmur is present and the teleoroentgenogram shows the enlargement of the pulmonary arc. Ofcours, the other findings help in many cases.

The prognosis in the pure cases of patent ductus arteriosus depends upon the size of the lumen.

When it is large and throws a heavy burden upon the heart, cardiac failure sets in early. If the patency is small, it may cause no cardiac burden what-so-ever (Wells, 1908). The average age of Abbott's (1934) series of 92 cases was 29 years.

A localized defect of the aortic septum is an opening between the aorta and the pulmonary artery, usually located just above the semilunar cusps. It is not a very common type of congenital anomaly. Poynter (1919) states he found it to constitue 1.7 % of all congenital heart anomalies. It is due to the failure of the primary aortic septum to completely divide the original ventral aorta into the aorta and pulmonary artery. This is but a minor degree of the same failure in development that is causative in persistent truncus arteriosus (Poynter, 1919). In many cases it is at first a mere thinning of the septum which becomes a congenital aneurysm and which later ruptures (Abbott, 1932).

The symptoms, when they are present, are the same as those found in patent ductus arteriosus. The findings also resemble those found in cases of patent ductus arteriosus, but the murmur is very near the ear and is slightly lower (White, 1931). Usually the point of maximal intensity is in the third or fourth left intercostal space, just to the left of the sternum. There is, ofcourse, a rough thrill accompanying the murmur (Abbott, 1932).

The heart is subject to the same strain and

therefore has the same prognosis as cases of patent ductus. The diagnosis can be made from the murmur and thrill, altho it is difficult to distinguish it from patency of the ductus arteriosus.

Pulmonary stenosis with patency of the foramen ovale is usually a valvular stenosis, probably due to fetal endocarditis occuring after the cardiac sept are closed. Cyanosis begins late, developes slowly and is very marked. There is clubbing of the fingers which may become very extreme.

The findings include a harsh, prolonged holosystolic murmur over the precordium with its point of maximal intensity in the second left intercostal space. It is not transmitted to the great vessels of the neck. Accompanying the murmur is a rough sustolic thrill with the same localization. The pulmonic second sound is weak or absent. Frequently there is a polycythemia, especially in the later stages of the condition (Abbott, 1932). Roentgenographic shadows of the heart show an enlargement of the right ventricle and the right auricle. (Abbott, 1934). The electrocardiogram shows right preponderance and an abnormally high P wave in lead I or II or both (Alexander, Knight, and White, 1925). The late picture is often complicated with endocarditis on the pulmonary valve which greatly increases the cyanosis. The diagnosis can not always be made. It can be based on the murmur, clubbing, cyanosis and the pulmonic second sound, together with the electrocardiographic findings.

Tuberculosis is common in these (Abbott, 1932). The average life span was 18 years in Abbott's (1934) series of 16 cases.

Tricuspid atresia with a defect of the interventricular septum and transposition of the great trunks is very rare. Abbott (1934) reports only one case among 1000 cases of congenital heart. This strange rearrangement of the cardiac constituents however does allow a reasonable circulation. The heart is functionally a three chambered, biatrial heart. The right ventricle is hypoplastic, in fact, it is a mere cleft in the wall of the hypertrophied left ventricle. The foramen ovale is always widely patent. The cyanosis may be slight, with occasional attacks of marked cyanosis and dyspnea. In the terminal stages it is very marked. The auricles are dilated somewhat and the left ventricle is hypertrophic. There is a systolic murmur in about half the cases. Diagnosis is seldom to be expected.

DEXTROPOSITION OF THE AORTA WITH DEFECT IN THE INTERVENTRICULAR SEPTUM

This condition was first described by Eisenmenger and is often referred to by his name. The large aorta rides directly over the interventricular septal defect or is also in the right ventricle and very near the defect. This condition is most commonly associated with pulmonary stenosis and then falls into the class of the "tetralogy of Fallot."

There is no Lundsgaar and Van Slyke "D" factor of delayed capillary circulation present in this anomaly. The only factor leading to cyanosis is the alpha factor, the permanent venoarterial shunt. The cyanosis is constant but moderate, as is the clubbing of the fingers and toes. There is dyspnea on exertion. There is a precordial murmur, systolic in time and heard best at the apex and is transmitted to the back, but not to the vessels of the neck. There is an accompanying systolic, mesocardial thrill. The roentgenogram shows an enlarged pulmonary arc, right heart hypertrophy and the fluoroscopic studies reveal pulsating hilar shadows (Abbott, 1932). The average lenth of life in Abbott's (1934) series of 10 cases was 13 years.

This rather rare condition is the result of failure of the interventricular septum to develope. It is usually marked by a very small ridge in the wall of the common ventricle where the septum should have been. There is moderate, continuous cyanosis resulting from the large, permanent venoarterial shunt. Strangely, it is often late in onset. The clubbing of the fingers is also often late in onset, and usually slight in degree. Usually there are no murmurs or thrills, or any other signs. The most commonly associated lesions are pulmonary stenosis and transposition of the great trunks. Diagnosis is usually impossible. Abbott (1934) found the average age to be 7¾ years in her 13 cases.

This condition, sometimes known as incomplete double heart, is rather rare. Robson (1931) reviews the etiological discussions in the literature and concludes that it is due to faulty development of the endocardial cushions in the fetal heart.

Persistant ostium atrioventriculare commune consists of a defect in the lower part of the interauricular septum and the upper part of the interventricular septum and a single atrioventricular orifice between these septal defects which is guarded by five or less cusps. There is transit cyanosis with pulmonary infections and a moderate terminal cyanosis. There is a loud systolic murmur heard over the mesocardium. Robson (1931) states that one half of these hearts, when uncomplicated by other lesions, occur in Mongolian idiots.

Pulmonary stenosis and a defect of the interventricular septum are usually accompanied by dextroposition of the aorta and hypertrophy of the right ventricle and as such is known as the "tetralogy of Fallot" after Fallot who first described this complex. The pulmonary stenosis is not commonly of the valvular type, but consists of an obstructive shelf of tissue just below the valve, or a narrowed or deformed conus. It is probably due to a partial or complete arrest of developmental expansion of the infundibulum (Poynter, 1919). This type of congenital heart constitutes ninety percent of those serious lesions found in individuals who live to adulthood, with congenital heart disease.

The physio-pathological factors in the causetion of the cyanosis are the permanent venoarterial shunt caused by the interventricular septal defect and the raised right ventricular pressure and venous stasis resulting from the pulmonary stenosis. Too, the dextroposition of the aorta allows venous blood to enter the systemic circulation. Then, the passive congestion resulting from the raised pressure in the right ventricle tend toward cyanosis. It is also possible that the stenosis may so reduce the amount of blood passing to the lungs for oxygenation as to be a factor adding to

the cyanosis.

The cyanosis is constant and of a bluish color which increases on exertion to a violet hue. The increased cyanosis is accompanied with dyspnea. There are dyspneic and syncopic attacks at times. Clubbing of the fingers and toes begins early and is rather marked in most cases by the time they reach adulthood. As a result of the oxygen unsaturation there is often a geographic tongue and angina attacks (Abbott, 1932).

Among the findings are cyanosis retinae, polycythemia, and increased hemoglobin. The murmur is rough, systolic, prolonged thruout systole and with its point of maximal intensity in the left second intercostal space just to the left of the sternum. It is very constant and may be transmitted to the vessels of the neck, but not to the back. It is accompanied by a systolic thrill which may be weak or absent. The thrill is usually found in the second, but occasionally in the third or fourth left intercostal space. The pulmonic second sound is absent, weak, or, rarely, accentuated. The electrocardiogram shows a right preponderance and a high P wave in leads I and II and

extra ventricular systoles. The right ventricle is hypertrophied to a slight or marked degree. The teleoroentgenogram shows the "coeur-en-sabot," in which the right side of the heart is enlarged and the pulmonary arc is small or missing.

Often there is developed a collateral circulation to the lungs as in pulmonary atresia, as described later (Abbott, 1932).

In making the diagnosis of "tetralogy of Fallot" one can consider that this lesion is by far the most common one found in adults with cyanosis due to a congenital lesion. The murmur and thrill are rather constant and characteristic and the other findings usually are sufficiently characteristic to fit well into the picture. This diagnosis can usually be made clinically. Blackford (1930) states that it can be differentiated from the Eisenmenger complex which has no pulmonary stenosis by the teleoroentgenogram. In Eisenmenger's complex there is a prominence of the pulmonary arc as found in teleoroentgenograms of patients with patent ductus arteriosus.

The prognosis is only fair as most of the

patients with this congenital heart condition sooner or later fall victims to cardiac failure. In Abbott's (1934) 83 cases of this condition, the average age was 12¼ years. However, many of them live to middle or even old age.

This condition is the same as the tetralogy of Fallot, except that instead of stenosis of the pulmonary artery, there is atresia. It occures only about one third as often as the tetralogy. It is always accompanied by dextroposition of the aorta and a well developed collateral circulation to the lungs. This collateral circulation may include a patent ductus arteriosus, anomalous vessels from the subclavian, the thyroidea ima, the aortic arch, the thoracic aorta, and the esophageal vessels, and lastly but often the greatest in extent, the bronchial arteries (Abbott, 1932).

The symptoms and findings resemble the tetralogy, but the cyanosis and clubbing are more marked, and the dyspneic and cyanotic attacks are less common. The pulmonic second sound is absent. The expectancy, judging from Abbott's (1934) series of 30 cases is about 6¼ years.

Persistant truncus arteriosus is a congenital heart in which the aorta and pulmonary artery are a common trunk which rides over a defect in the base of the interventricular septum or comes from the right ventricle. It is not a common type of lesion, there being only 21 cases in Abbott's (1934) collections. Poynter (1919) reports it to be present in 4.8 % of all congenital heart disease.

Failure of the aortic septum to develop in the primitive ventral aorta is the cause of this condition. There may be a slight ridge or veil to point out where the absent septum should have been and it may lead up to the arteries that go to the lungs. The common trunk valve may be made up of three, four or five cusps and the coronaries arise from behind two of them. There is no trace of the pulmonary artery, the arteries to the lungs arising from just above the valve, from the under surface of the transverse or descending aorta, or from the innominate, subclavians, or the right carotid. Often the bronchial arteries are enlarged. The cyanosis is due to the raised oxygen unsaturation resulting from the permanent and extensive venoarterial shunt.

The cyanosis may not develope until after some disease, especially pulmonary disease, but usually it is

intense and constant. The clubbing is also marked. Dyspnea usually developes upon exertion.

There is a murmur all over the precordium but it is maximal in the third left intercostal space. It is diastolic in time. There is a diastolic thrill with the murmur, and in some cases, a systolic thrill also. There is a polycythemia. The roentgenographic shadows show an enlarged pulmonary arc and upon fluoroscopic study, a Corrigan pulse, or "danse hiliare" is noted in the lung hila as evidenced by the pulsation of the pulmonary hila (Abbott, 1932).

The diagnosis can be cased upon the diastolic murmur and thrill, and the pulmonary pulse and the large pulmonary arc. Many of these cases die in infancy, some live to middle life, but the expectancy is about 4 years, judging from Abbott's (1934) 21 cases.

In this condition, the great trunks arise from the opposite ventricles and the interventricular defect is the only way in which the blood of the systemic and pulmonary systems are mixed. Abbott(1934) reports 16 cases in her series. The individual may live to childhood or early adulthood, but the average life in Abbott's (1934) cases was only 1½ years long. Usually the interventricular defect is so large that it produces no murmur, or thrill.

The two chambered heart with transposition of the great trunks is rare, Abbott (1934) reporting only 2 cases among 1000 congenital hearts. Usually there is just a rudimentary ridge to mark the position of the septa. There is but a single atrioventricular valve which has from 2 to 5 cusps. Usually there are no findings but there may be a systolic murmur. The cyanosis and clubbing of the fingers and toes is marked, but dyspnea is moderate. There may be a patent ductus arteriosus, dextroposition, or right ventricular hypertrophy.

In pulmonary atresia, there is a very serious impediment to circulation, and if life is to continue long after birth, a marked collateral circulation must develope. It is not common, Abbott's (1934) series having only 10 cases. The right auricle and left ventricle are enlarged, while the right ventricle is hypoplastic.

There is extreme cyanosis from birth, which is constant. The clubbing is extreme if the individual lives long. Dyspnea accompanies the cyanosis. Usually there is a systolic murmur. If the patient lives past the first few days, he is apt to develope infectious endocarditis at any time which would lead to death. The average age in Abbott's (1934) 10 cases was only 10 weeks.

With transposition of the great trunks there is usually defects of both the interauricular and the interventricular septa which somewhat relieves the circulatory embarrassment. But when the transposition is aleviated only by a patent foramen ovale and a patent ductus arteriosus, the condition is very serious. There is a constant, extreme cyanosis and dyspnea. Usually there is some type of systolic murmur. The right ventricle is enlarged and the pulmonary artery may be either enlarged or hypoplastic. These patients die in a matter of a few days, the average lenth of life in Abbott's (1934) 31 cases was 1¾ months.

Mitral atresia is a complete closure of the mitral segment due to the arrest in its early development. It is a rare condition. Usually the left auricle is small and the left ventricle is a mere cleft in the wall of the right ventricle, or, it disappears entirely. The foramen ovale is patent, and the right ventricle and auricle are greatly hypertrophied. The aorta is small or atresic, and the ductus arteriosus is widely patent.

There is extreme cyanosis with dyspnea from birth. There may or may not be any murmurs. This condition is often associated with other grave anomalies, especially aortic atresia (Abbott, 1932). The average lenth of life in Abbott's (1934) series of 5 cases was 10 months.

Tricuspid atresia is always accompanied by a patent foramen ovale and a defect of the interventricular septum just below the aortic valve. If the atresia has been the result of fetal endocarditis, there is scaring of the valve: if it is a true congenital anomaly, then the tricuspid segment is a mere muscular septum. The right auricle and left ventricle and auricle are dilated and hypertrophied. The right ventricle is hyplastic, just a cleft in the wall of the left ventricle. So, functionally, this type of heart is a "cor biatriatum triloculare."

There is extreme cyanosis, and dyspnea, with dyspneic attacks. There may be sustolic murmurs. This condition is often associated with transposition of the great trunks in which cases the circulation may then compensate somewhat. In 15 cases of Abbott's (1934) series, the average life lenth was 6 months.

The two chambered heart is rare, and its seriousness varies in each case. It is due to the total lack of development of the cardiac septa. These patients occasionally live to adolescence, and experience marked cyanosis, clubbing and dyspnea, and general underdevelopment. When this condition is associated with persistant truncus arteriosus, there is extreme cyanosis, and they die very early.

In some cases, the pulmonary veins are found to empty into the superior or inferior vena cava, the innominates or the hepatic veins. Some cases are accompanied with transposition of the great trunks, and still others are found with pulmonary stenosis. The average age in Abbott's (1934) series with 7 cases was 1½ months.

This rare cardiac anomaly is a very serious one. It is due to the primary arrest of its development, or, and more often, to fetal myocarditis, usually of luetic origin. Most often it is accompanied by mitral atresia, and then the left ventricle is a mere slot or thickened spot in the wall of the right ventricle, and the aorta only a cord to the transverse arch. Too, the pulmonary artery is large, and the patent ductus arteriosus is extremely large. The foramen ovale is widely patent. The coronaries arise from the aorta, but the blood flows "downward" thru the ascending aorta to them from the point of the patent ductus. The right auricle is very large, the left auricle is small. When the mitral valve is patent the left ventricle is large and concentrically hypertrophied. There is a very sever cyanosis and dyspnea and life is only four days long, according to the average of Abbott's (1934) series of 12 cases.

In a given case of heart disease it must first be determined whether it is a congenital or acquired disease. Altho in some cases this is very difficult, usually it is rather easy.

In some cases of congenital heart disease there may be no symptoms and the lesion is picked up in a routine physical examination. In other cases, the infant is born with cyanosis, and dies in a few hours or days. Heart symptoms in infants under two years of age point to congenital heart disease (Holt and McIntosh, 1933). In children with no history of previous causative disease, cardiac symptoms and findings are usually the result of congenital anomalies. In adults, such a history is merely suggestive. However, a history, and even symptoms and findings of organic heart disease does not rule out congenital heart disease in as much as many congenital lesions are complicated in childhood by organic disease, especially rheumatic cardiac disease and endocarditis.

In most cases, then, the diagnosis of congenital heart disease must be based on symptoms and findings which are abnormal for organic heart disease, but characteristic in congenital heart conditions. There are the atypical murmurs of early onset, cause unknown, and at abnormal sites and with abnormal transmission.

They are often rough, course, loud, and high in pitch. They are usually systolic and may be holosystolic. With the murmurs are often thrills. The x-ray shadows and the electrocardiographic tracings are often too incongruous to be called normal or characteristic of acquired heart disease. The cyanosis, clubbing of the fingers, and the habitus of the patient point to congenital lesions, when they are present.

After the diagnosis of congenital heart has been made, most clinicians are satisfied to cease the study of their diagnostic findings in the case. However, everyone will agree that an attempt should be made to go as far in the diagnosis as possible. Most cardiac men are now diagnosing the more simple congenital lesions clinically, and so the start has been made. Only by making the most of these rare cases can we advance our meager knowledge of them. Our only vital reason for urging the diagnosis of the exact type of congenital heart at present is for prognostic purposes and for the hope we have in the future improvements in our therapeusis.

The best method of diagnosing the exact condition is to know all the characteristics of the various types of the condition. In the first place, the rarity of congenital heart conditions makes this impossible except for the cardiac specialist with an extensive practice and in the second place, many of these conditions are so variable as to exclude the definite characterization of them. However, a very encouraging note is sounded when we say that the common congenital hearts which make up the majority of those which are of clinical interest have definite and well known symptoms and findings which make the

diagnosis easy and definite.

In attempting to summerixe the diagnostic points of each congenital lesion we can do so only fragmentarily. Griffith and Mitchell (1934) give the following outline of suggestions:

Cyanosis, marked, and no murmur, suggests transposition of the large vessels without other complicating lesions. Corroboratory evidence is accentuated pulmonic second sound and cardiac enlargement.

Cyanosis with systolic murmur in the second left intercostal space suggest pulmonary stenosis. The stenosis is isolated or with only a patent foramen ovale if the pulmonic second sound is feeble. If the pulmonic second sound is normal or accentuated there is also a septal defect.

Systolic murmur, loudest over the midsternum or in the third left intercostal space, without cyanosis suggests a septal defect. There is no conduction of the murmur.

Systolic murmur, very lound over the second left intercostal space or upper part of the sternum and carried to the neck vessels and with the pulmonic second sound accentuated suggests patent ductus arteriosus, with pulmonary stenosis. Also left side enlargement

and perhaps the right side also. Patent foramen ovale or interventricular septal defect may also be present. If the above is present without cyanosis, only patent ductus arteriosus is present.

Systolic murmur over the upper sternum and to its right with transmission to the neck vessels and left ventricular hypertrophy suggests coarctation of aorta.

Diastolic murmurs suggest pulmonary insufficiency.

Dunn(1914) arranged his congenital heart symptoms and findings in a chart. Reid (1931) followed the same plan but was more inclusive, both in the symptoms and findings included, and the number of lesions so studies. Following is a chart that is built upon the same plan. It may be of value in diagnosing the type of congenital lesion in a given case of congenital heart disease.

Lesion	Cyanosis	Dyspnea	Clubbing	Murmurs: time	site	quality	transmission	Thrills	Pul. second sound	Cardiac enlargements	E.K.G.
Aortic stenosis	none	none	slight	systolic	2, 3 rt	harsh	neck	systolic		L. V.	normal
Coarctation of the aorta	none	none		systolic	left of sternum		back neck				normal
Auricular septum defect	transit	transit	none	presystolic	2, 3 lt	roar	back		acc	rt side pul arc	normal
Ventricular septum defect	terminal	transit		systolic	3, 4 lt	harsh constant	none	systolic marked			normal
Patent ductus arteriosus	transit	transit	none	systolic diastolic	2 lt	harsh	back neck ?	systolic marked	acc	pul arc	normal
Aortic septum defect	transit	transit		systolic diastolic	2, 3 lt	harsh	none	systolic marked	acc	rt side	normal
Pulmonary stenosis with patent foramen ovale	gradual, marked in late stages		may be extreme	systolic	2 lt		back only	systolic rough	weak	rt side	high P in I, II rt prep
Dextroposition of aorta, ventricular septum defect	constant, moderate	moderate on exertion	moderate	systolic	apex		none	systolic		pul arc rt side	
Cor triloculare biatriatum	late, slight constant		late moderate	systolic	3, 4 lt			none			
Persistant ostium atrioventriculare commune	transit with pulmonary infections			systolic	3, 4 lt						
Pulmonary stenosis with ventricular septum defect	constant, worse on exertion	slight	extreme	systolic	2 lt	rough	neck	weak or absent 2,3,4 lt	weak	boot shaped heart	high P in I, II rt prep
Pulmonary atresia with ventircular septum defect	as above but more marked	slight	extreme	systolic	2 lt		neck	none	absent	boot shaped heart	high P in I, II rt prep
Persistant truncus arteriosus	inconstant, intense at times	marked	may be marked	diastolic	3 lt			diastolic		pul arc	
Transposition with ventricular septum defect	moderate constant	marked		none				none			

Abbreviations

lt	left	L.V.	left ventricle	2, 3 rt	2nd and 3rd right intercostal space
rt	right	pul	pulmonary	3, 4 lt	3rd and 4th left intercostal space
acc -	accentuated	prep	preponderance	high P in I, II	- high P wave in leads I and II

As in all departments of medicine, the general appearance of the patient, and a knowledge of the life he is leading is sufficient to make a fair tentative prognosis in congenital heart disease. However, since our knowledge is limited in congenital heart disease, each case needs study before a reasonable, far sighted prognosis can be given. Since we are limited in our therapeusis in this field at present, the prognosis is the main reason for wanting to make an accurate diagnosis of each case.

In general, cases which fall into group one have a good prognosis. They have an expectancy of living to about mid-life. Coarctation is always liable to rupture of the ascending aorta, so, they should avoid excessive strain. Subaortic stenosis is apt to have endocarditis. Bicuspid aortic valves are liable to subacute infection and also rheumatic endocarditis, leading to insufficiency.

Group two cases are the "potential cardiacs" of the congenital group. Defects of the auricular septum are liable to induce pulmonary insufficiency and resultant right-sided decompensation. Ventricular defects are more liable to be complicated by endocarditis. Patent ductus arteriosus and defects of

the aortic septum are often complicated by subacute bacterial endarteritis at the pulmonary orifice.

In group three, the prognosis is poorer. It is the exception rather than the rule for cases of this group to survive infancy and childhood. Here the prognosis in the individual case depends upon the degree of compensation of the heart as a functioning organ to its defect. Polycythemia is a compensatory mechanism but carries with it a bad prognosis.

The prognosis of individual lesions is mor fully explained in the discussions of the individual lesions.

The treatment of congenital cardiac disease is at present very limited, and at times, very discouraging. However, all cases of congenital heart should not be considered as hopeless. Many congenital heart patients can live useful lives, and there are instances where such patients have become famous because of their accomplishments in spite of serious lesions (White and Sprague, 1929). In many cases only palliative treatment can be offered.

The first place that treatment can be instituted is in the prenatal period. Heredity has been shown to play perhaps a minor part in the causing of some of the congenital heart defects, and prenatal environment has benn shown to play a major part (see section on etiology). Therefore, it is logical that the prenatal period should be as normal as possible, even from the first days following conception. Following are a few factors which have been incriminated or upon which suspicion has been thrown and which might well be avoided if possible during the prenatal period: Pyschic upsets, physical mistreatment and hardships, rheumatic fever, tuberculosis, small pox, alcoholism, and syphilis. In the later, the mother should be treated early, and as usual.

In the cyanotic group, blood letting has

been found helpful in cases where the right heart is burdened. Fifteen to twenty ounces are taken at a time. Stimulants are useful during dyspneic attacks. Brandy, ammonia and ether, cannabis indica, caffeine citrate, and epinephrine have been found suitable. When there is persistant irritability and cough, bromides are heopful. Sometimes morphine given with atropine in small doses is necessary. Barbiturates and choral hydrate serve to control insomnia.

Since the cyanosis of congenital heart disease is not due to a pulmonary cause, oxygen by inhalation is useless. It helps only in cases where there is an added difficulty from atelectasis, alveolar thickening, or other factors which might cause poor aeration. Where the oxygen is needed is in the arterial system, and perhaps some day a method of intra-arterial administration of oxygen will be perfected.

Preventive treatment in the cyanotic broup of lesions is very helpful. There patients should have a good diet and good habits of health. All infections should be avoided if possible and even colds should be treated as major illnesses. These patients should not be allowed to become pampered

invalides just because crying will precipitate an attack of cyanosis and dyspnea. They should be taught self control and obedience just as other children are. Educating these children to live within their cardiac limits is difficulat at times, but very necessary. They should be taught to play active, but non-competitive games. With a little supervision, these little kiddies can be taught to live safe, yet fairly normal and happy lives. There is always the question of whether they should be placed in institutional schoold during childhood. The opinion of most authors is that they should be reared and accustomed to the environment and type of life they must lead all their days (Abbott, 1924).

In group two types of patients, preventive treatment is very helpful. Since these are the "potential cardiacs," all possible means should be employed to keep them in good health. All foci of infection should be cleared up early. Rheumatic fever, which they are very susceptable to, should be treated very early and thoroughly with rest, salicylates, tonics, etc. Too, they must learn to work, play, and in fact, live within their cardiac tolerance.

In all congenital heart patients, the cardiac

capacity and tolerance should be carefully studied, and frequently checked as it is the most dependable index to the patients progress.

When these patients are confined to bed or are in a hospital, a definite outlined course of physiotherapy would not only help the patient's physical condition, but also might be a wonderful aid in the building and maintainance of the patient's moral. Such work has been very effective in organic cardiac disease in children (Bronson, 1922).

The surgical treatment of cardiac disease is now only a dream, but is perhaps the field of brightest hopes in future therapeusis. Ofcourse, early tonsillectomy and carefull dental attention is of proven value. Intracardiac surgery is no in its infancy. Cutler and Beck (1929) summerize 12 operations which have been done on the heart for valvular stenosis and discuss the problems in general. In spite of a mortality of 83 % they believe this field of surgery is promising. Lockwood (1929) discusses all types of cardiac surgery and believes the future to be bright. In the future we may see not only the slitting of stenosed valves, but also the building of new ones and the repairing of septal defects. The imagination has no limits

in what the future may bring.

As long as 28 years ago, Munro (1907) suggested the operation of ligation of patent ductus arteriosus. To date it has not been attempted in humans. It still remains as a suggestion however.

It is conceivable that some day we shall be able to surgically repair coarctation of the aorta.

Intra-venous and intra-arterial administration of oxygen in cyanosis, at least during times of cardiac crisis, will be another type of treatment that time may bring us.

Summarizing, there is one point that should be stressed. Congenital cardiac disease is no longer the mere subject matter for the anatomist's discussion, or for the pathologist's museium of curiosities. Congenital cardiac disease is now in the clinician's realm, for he can now diagnose such disease almost as accurately as he does organic heart disease and the treatment is definitely more than palliative management.

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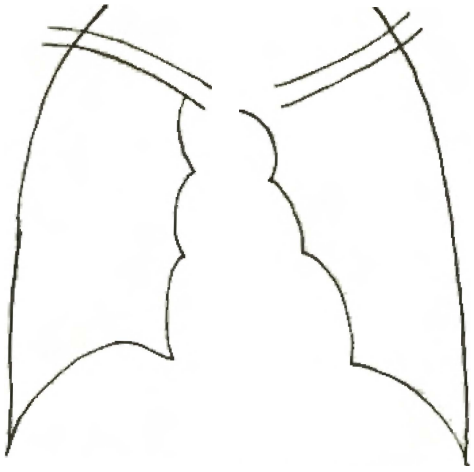

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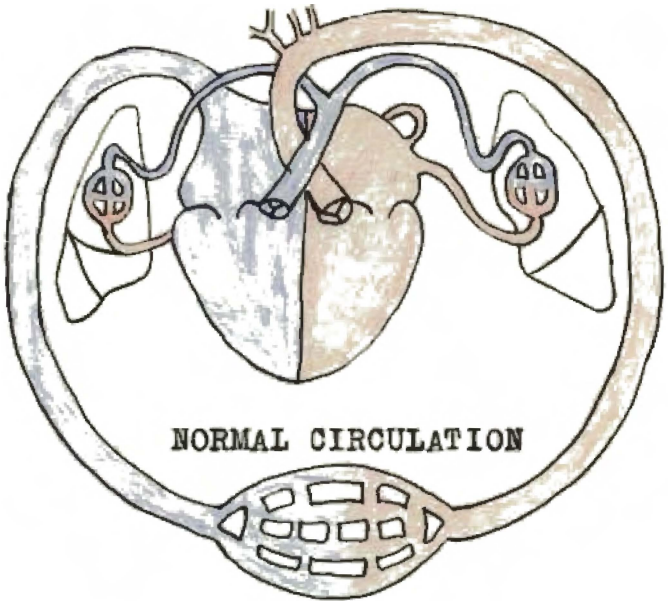

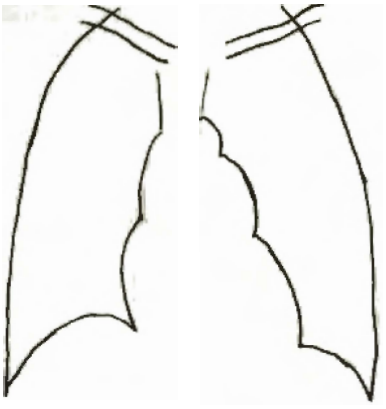

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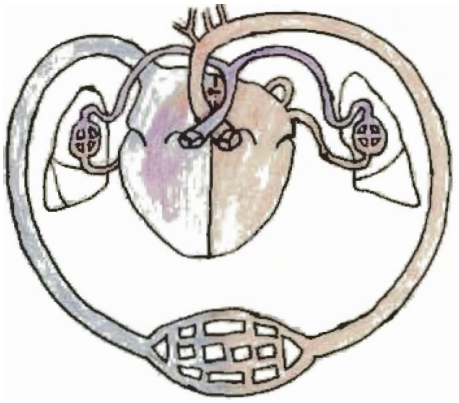

Circulation with
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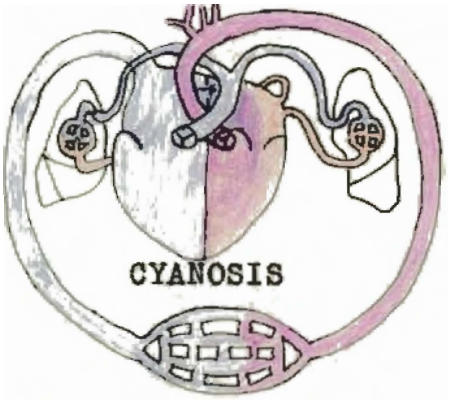
NO CYANOSIS



Circulation with
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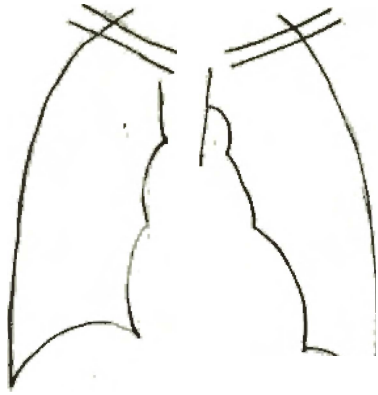

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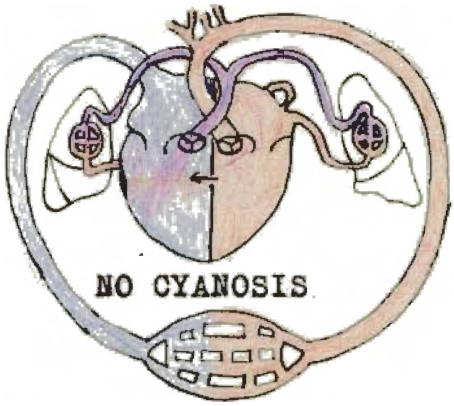
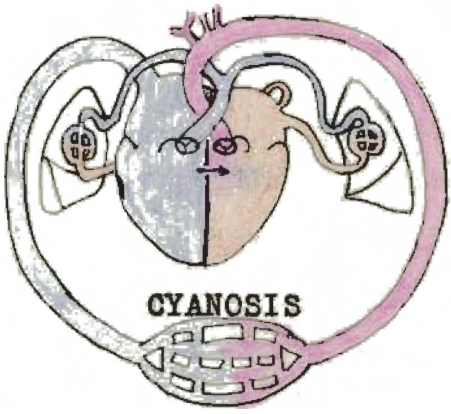
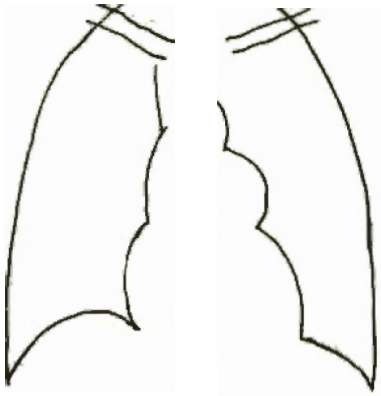


Circulation with
arteriovenous shunt



Rare transit and
terminal circulation



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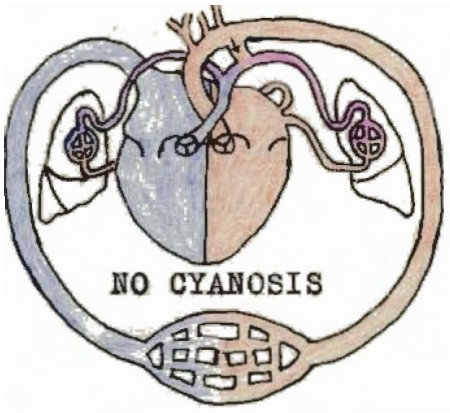
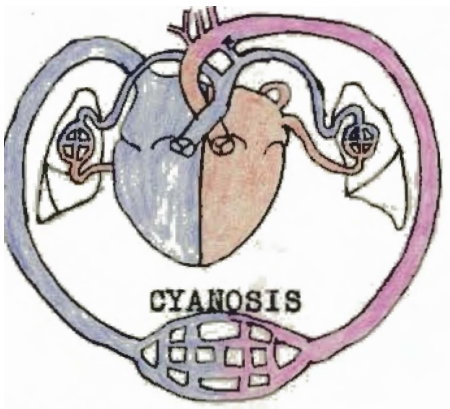
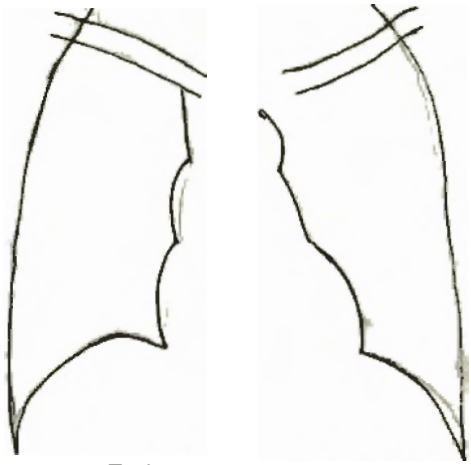


Circulation with
arteriovenous shunt



Frequent transit
attacks due to a
veno-arterial shunt



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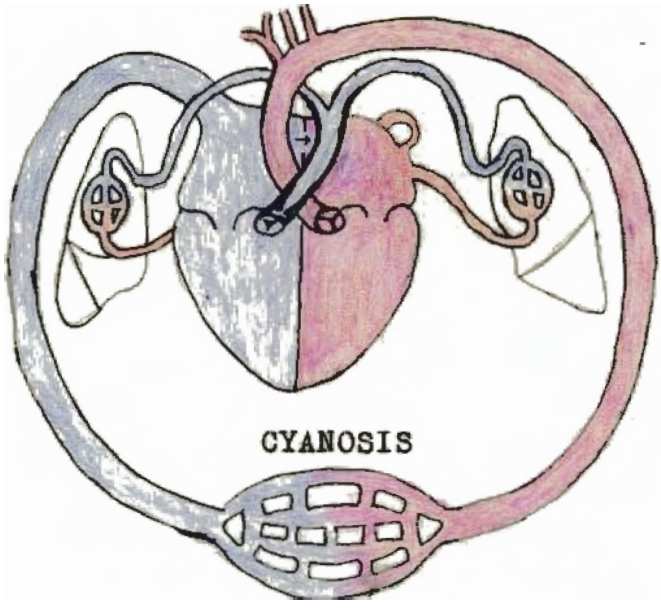
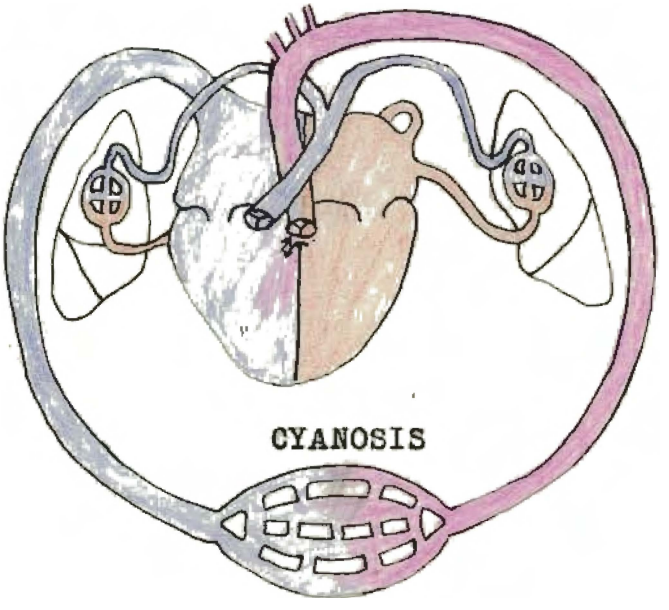
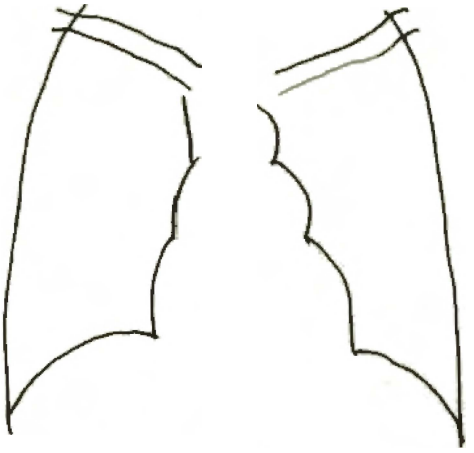


Gradual cyanosis, marked in late stages.

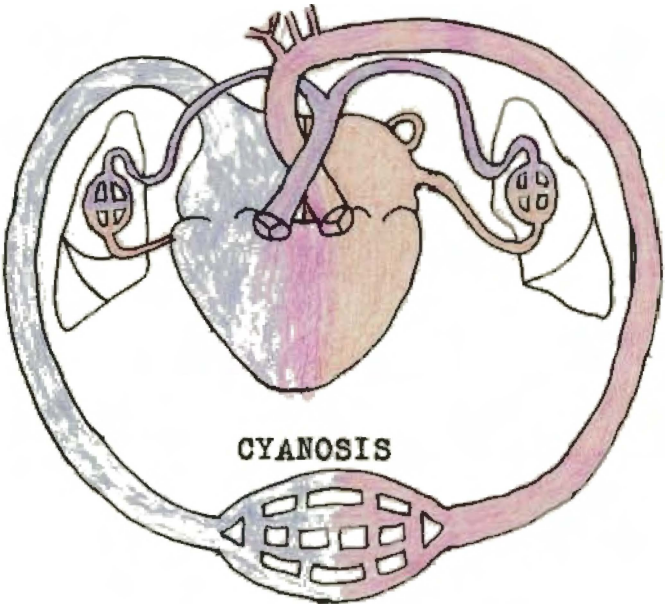
Teleoroentgenogram



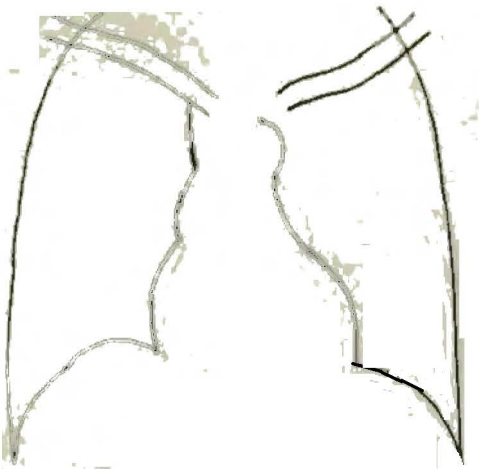
Moderate but constant cyanosis



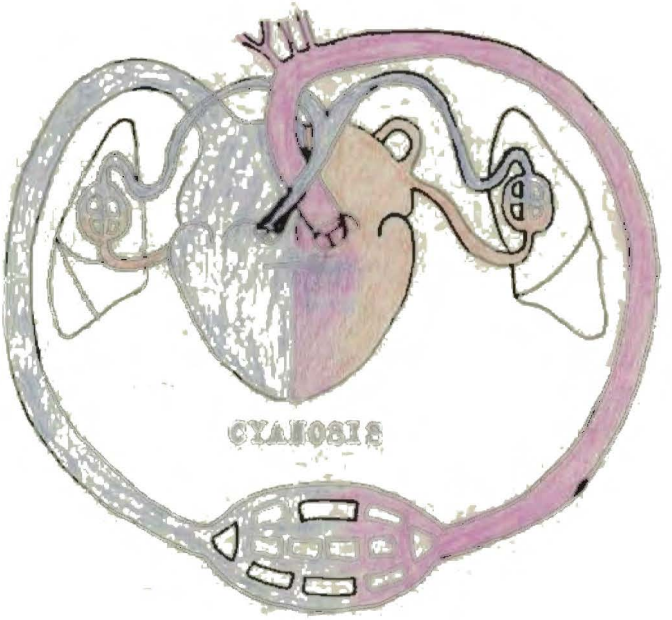
Teleoroentgenogram



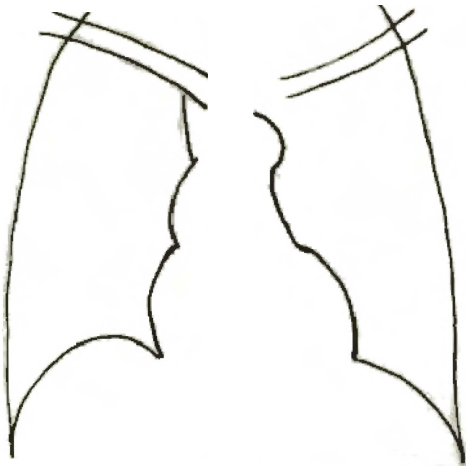
Slight but constant cyanosis, late in onset.



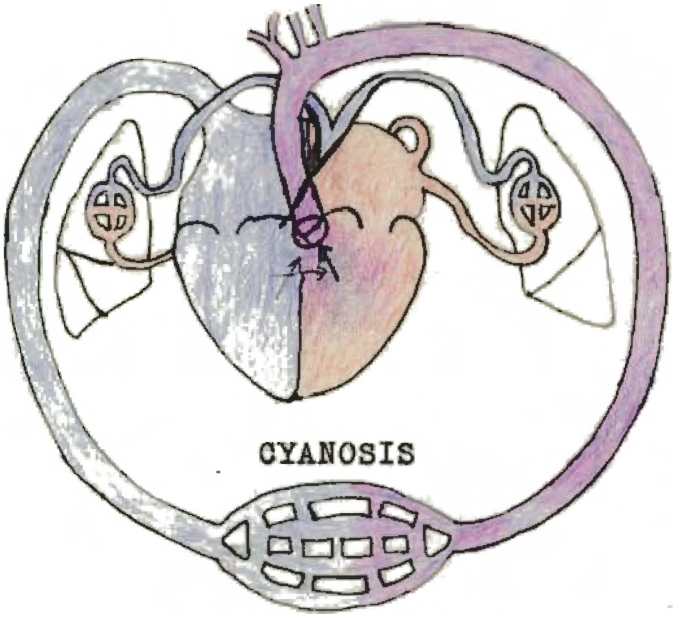
Teleoroentgenogram



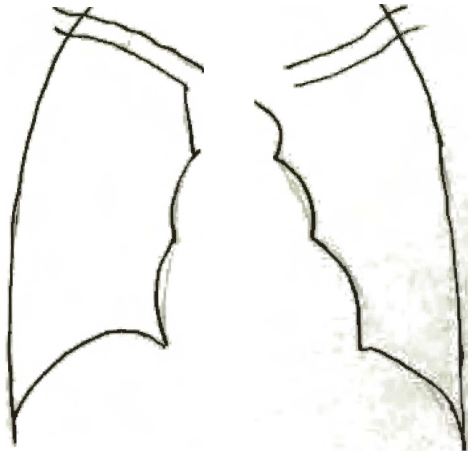
Constant cyanosis,
worse on exertion.



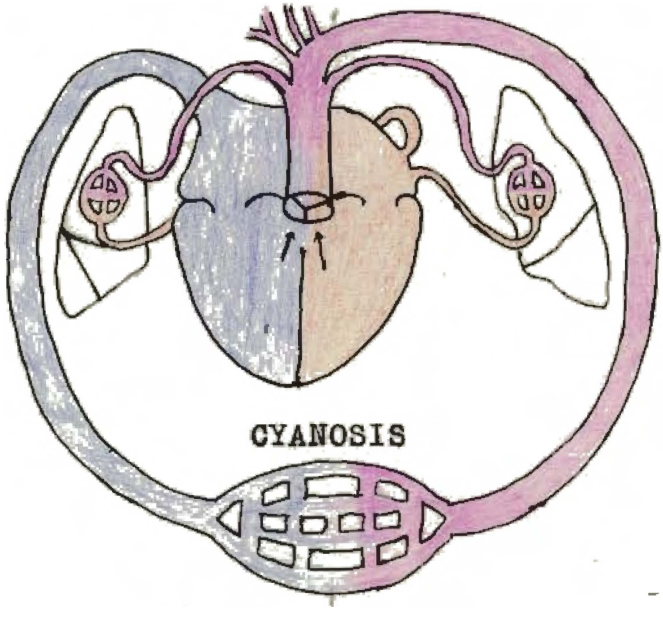
Teleoroentgenogram



Marked cyanosis, constant, worse on exertion.



Teleoroentgenogram



Inconstant cyanosis,
Intense at times.