ASPECTS OF CONGENITAL CARDIO-VASCULAR DISEASE

BY

CARROLL JAMES CLARK

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INTRODUCTION

In the not too distant past congenital heart disease was looked upon by the medical profession, in general, as a clinically impenetratable field, and left its mysteries to be solved by the autopsy table. Medical men were content to make just the diagnosis of congenital cardiac lesion without trying to determine or differentiate what type of defect existed in the individual patient, and condemned all of the afflicted to a grave prognosis and early death. There were a few intrepid early clinicians who ventured into the field and between them laid the fundamental basis for its understanding; namely, Jean Baptiste de Senac (1693-1770), Giovanni Baptista Morgagni (1682-1771), Carl Rokitansky (1804-1878), Thomas Peacock (1812-1882) and Sir Arthur Keith (1866-), but the far greater majority of doctors were not only ignorant about the subject, but also uninterested in it. Indeed, much of this same disinterest exists today among far too many of the general practitioners and specialists who do not realize the importance of detailed diagnosis in respect to prognosis, nor do they realize that a certain number of congenital cardiac defects are compatible with a long and useful life.

It remained for Maude E. Abbott to bring the subject before the medical profession in such a way as to create widespread interest and to enable the general practitioner to have a much better understanding of a field which he had hitherto avoided for any one of several possible reasons. Dr. Abbott's analysis of 1,000 autopsied cases of congenital heart disease is the most complete and thorough piece of work in the entire field. Her classification has made it possible to arrive at a quite accurate prognosis. She has stimulated much research in the subject and has particularly instig-

ated the cardiologist, internist, pediatrician and recently the surgeon to acquire a healthy interest in it. Gradually but definitely the attitude toward congenital heart lesions is changing.

Congenital heart disease is relatively rare, comprising one to two per cent of the total incidence of organic heart disease in the United States, although some authors quote figures as high as seven to ten per cent. Epstein places the incidence at four per cent in children with heart disease under ten years of age. In New England, where the incidence of rheumatic heart disease is high, White and Jones found in a series of 448 patients under twenty years of age that six per cent were congenital in origin. This indicates that the incidence is sufficiently high to warrant the current interest in the subject.

CLASSIFICATION

There are two main types of classification, namely clinical and anatomical. Both types have their respective merits and are worthy of consideration. Maude Abbott's clinical classification is perhaps the most widely known and used today and is presented below in detail. She divides the cases into three groups:

I. A Cyanotic Group. Cases in which no abnormal communication exists between the systemic and pulmonary circulations, but in which the anomaly is liable to become the seat of strain.

A. Less important group.

1. Simple dextrocardia
2. Anomalies of the pericardium. Defects and diverticula.
3. Anomalous chordae
4. Uncomplicated quadricuspid and bicuspid semilunar valves.
5. Double auriculoventricular orifices
6. Pure coarctation of the aorta; adult type.
7. Anomalies of aorta, of the aortic branches, coronary arteries, pulmonary arteries and of the great veins, unless these are extreme.

B. More serious group.

1. Ectopia cordis
2. Primary congenital hypertrophy of the heart
3. Pure subaortic or aortic stenosis, with considerable left ventricular strain.
4. Pure mitral stenosis
5. Pure coarctation of the aorta, infantile type.

II. Cyanosis Tardive group. Cases of arterial-venous shunt with possible transient or terminal reversal of flow.

1. Patent ductus arteriosus
2. Localized defects of aortic septum (communication between base of aorta and pulmonary artery or base of right ventricle).
3. Localized defects in the interauricular septum, patent foramen ovale, persistent ostium primium

persistent ostium secundum.
4. Localized defects of the interventricular septum.

III. Cyanotic group. Cases of permanent veno-arterial shunt (Morbus coeruleus).

A. Moderate cyanosis.

1. Defect of interventricular septum with dextroposition of the aorta.
2. Cor triloculare biatriotum
3. Pulmonary stenosis with patent foramen ovale
4. Tricuspid stenosis
5. Tricuspid atresia with septal defects.

B. Marked cyanosis.

1. Pulmonary stenosis with defect of ventricular septum and dextroposition of aorta.
2. Pulmonary atresia with defect of ventricular septum and dextroposition of aorta
3. Transposition of arterial trunks with defect of ventricular septum.

C. Extreme cyanosis.

1. Cor biloculare with transposition of arterial trunks
2. Persistent truncus arteriosus with localized defect of interventricular septum.
3. Cor biloculare with persistent truncus arteriosus.
4. Complete transposition of arterial trunks without defect of ventricular septum, but with interauricular septal defect or patency of the ductus arteriosus.
5. Pulmonary atresia with closed ventricular septum, defective auricular septum and patent ductus arteriosus.
6. Mitral atresia with aortic aplasia, defect of auricular and ventricular septa and patent ductus arteriosus.
7. Aortic atresia, transposition of arterial trunks, closed ventricular septum, patent ductus arteriosus.

From this classification it may be said that, in general, the greater the degree of cyanosis, the more serious the lesion and the graver the prognosis.

The anatomical classification is of more value to the pathologist than to the clinician. There are several, and a combination of those presented by Dry and Mitchell is given below:

I. Anomalies of septal formation.

1. Cor biloculare
2. Cor triloculare biatriatum
3. Cor triloculare biventriculare
4. Auricular septal defects
5. Ventricular septal defects
6. Persistent truncus arteriosus.

II. Anomalies in association with torsion of the cardiac tube and with development of the bulbus cordis.

1. Subaortic stenosis
2. Pulmonary stenosis
3. Transposition of great vessels
4. Tetralogy of Fallot
5. Eisenmenger's complex
6. Anomalies of the aortic and pulmonic valve cusps.

III. Anomalies associated with development of the aortic arches.

1. Persistent right aortic arch with isthmus stenosis of the left arch.
2. Double aortic arches
3. Coarctation of the aorta
4. Anomalous origin of vessels arising from the aortic arch.
5. Patent ductus botalli.

IV. Anomalies in the size and position of the heart.

1. Transposition of heart chambers
2. Dextrocardia
3. Congenital hypertrophy
4. Ectocardia

V. Anomalies of the coronary vessels.

EMBRYOLOGY

In the field of embryology one can find the logical explanation of how the many intricate and bizarre defects of congenital heart disease occurs. From their studies of human embryos in the early weeks of development, embryologists claim that the cardio-vascular system recapitulates its ancestral history during the course of its own development, and they have traced the heart's growth from a structure similar to that of a fish down to the complicated human organ. If an arrest of development occurs during early fetal life, then the foetus will be born with a heart that will represent theadult form of one of the lower vertebrate animals, depending upon the stage at which the arrest took place. However, this does not account for certain congenital cardiac defects, which will be discussed later.

Thomas Dry has neatly summarized the cardio-vascular development in a simple form by dividing it into six stages: (1) Septal formations dividing the auricle, the ventricle, the bulbus cordis, and the common aorta or truncus arteriosus each into two sections; (2) torsion of the cardiac tube, (3) development of the bulbus cordis, (4) the incorporation of the sinusvenosus into the right auricle, (5) the evolution of the aortic arches, some being obliterated, others becoming the permanent aortic arch, pulmonary artery and their branches, and (6) closure of fetal channels after birth.

With the evolution of a lung-respiratory system the complete separation of the venous and arterial systems became imperative, and septal formations resulted. When an arrest in development occurs in this stage, the well-known septal defects and three chamber heart results, simulating the heart

of amphibious or of some of the higher reptiles.

The congenital defects, transposed great vessels and inverted ventricles, for a long time could not be explained upon an embryological basis, and they seemed to contradict the principles of evolution until Professor Spitzer formulated his theory. Normally as the primitive heart elongates, it kinks upon itself and there is a torsion or clockwise rotation of the bulboventricular end of the cardiac tube. This is necessary in order to bring the left ventricle into juxta-position with the aorta and the right ventricle with the pulmonary arch. In the process of torsion, the right aorta is obliterated, whereas the left remains open. Briefly, Spitzer's theory explains such congenital defects to the arrest or delay of the clockwise torsion that normally occurs in the growing primitive heart, thus leading to an apparent counter-clockwise shunting of the parts with resultant reopening of the reptilian right aorta and obliteration of the left aorta. Also the degree of transposition depends upon the degree of torsion that has occurred before the process is arrested. Sometimes the right and left aorta may fuse into a single large trunk. The dextroposed aorta may override the interventricular septum, which is often incomplete, and the right ventricle. Now, if there is an incomplete process of incorporation of the major portion of the bulbus cordis into the right ventricle to form the conus arteriosus and a smaller portion into the left ventricle, the corresponding ventricle will be stenosed in that region, e.g. subaortic stenosis or infundibular pulmonary stenosis. A combination of incomplete torsion of the cardiac tube and the development of pulmonary stenosis will give the genesis of the interesting anomaly, the tetralogy of Fallot, representing arrest at the reptilian stage.

The arrest of development of the heart occurs in a great majority of cases between the fifth and eighth week of fetal life. Interference with growth during these weeks is apt to give rise to further anatomical changes of a compensatory nature, making the end-result confusing. However, further changes are merely adaptations of structure for the maintenance of the cardiac circulation under the various conditions imposed by the congenital defects, and they follow simple mechanical and hydraulic laws.

Several other congenital cardio-vascular defects have an embryological explanation, but are too detailed to be included here.

Another etiological factor in the production of cardiac anomalies is fetal disease, both endocarditis and myocarditis. Fetal endocarditis is relatively rare, and occurs in the later months of pregnancy after the cardiac septa have closed. Fetal myocarditis is of greater importance and has been shown by von Zolka to underlie many of the cases of aortic and pulmonary atresias.

The causation of the arrest of growth in the development of the heart is a subject of considerable speculation. Maude Abbott believes that the answer "should be sought chiefly in the environment of the developing embryo, in the form of disease in the fetal envelopes or maternal tissues, infective processes affecting the health of either parent at the time of conception, physical trauma or even psychic shock inflicted upon the mother in the first weeks or days of pregnancy, etc. All such pathological conditions set up or acting in utero are liable to interfere with perfect uniformity of growth." Other investigators believe that defects in

the germ cells cause a goodly proportion of the cardiac anomalies and point out that hereditary influence is evident in 97 of Abbott's 1,000 cases. Undoubtedly, both theories play their part in the explanation of the cause of the arrest, and neither one should be accepted to the exclusion of the other.

SYMPTOMATOLOGY

There may be no symptoms at all in congenital heart disease if there is no undue strain on the heart or vena arterial shunt. On the other hand, the symptoms may be extremely severe, as in cases of primary congenital hypertrophy, marked coarctation of the aorta, pure stenosis of any of the valves, or lesions with attendant marked cyanosis.

According to White, the most frequent symptom is dyspnea, particularly on exertion. Abbott noted it in thirty-two per cent of her 1,000 cases. If the dyspnea is severe enough, cough is common; hemoptysis may occur if there is pulmonary vascular engorgement, polycythemic congestion, or heart failure.

Cerebral symptoms are very important and are usually due to anoxemia, although sluggish circulation and cerebral thrombosis from marked polycythemia may also produce them. Weakness, faintness, syncope, dizziness, headache, convulsions, coma, delirium and transient or persistent paralysis may occur, especially if the degree of cyanosis is marked.

Gastro-intestinal symptoms are not important and are infrequently seen, except for dysphagia that may be caused by some anomalies of the aorta. Faulty abdominal circulation may cause anorexia, nausea, vomiting, hematemesis and constipation.

Coldness of the extremities with cyanosis or tingling has been noted. Some patients are abnormally susceptible to respiratory infections. Palpitation is frequently complained of, but pain seems to be quite rare.

Another unique feature of the cyanosis of congenital heart disease is the absence of edema when the cyanosis is quite pronounced.

INDIVIDUAL DEFECTS

THEIR PATHOLOGY AND DIAGNOSIS

Pericardial defects. There are three types of these defects: absence or defect of the parietal pericardium, diverticulum or hernia, and lack of attachment of the parietal pericardium - all being rare. The heart may lie entirely in the left pleural cavity, along with the left lung, and so be exposed to the diseases of the lung and pleura, invariably with a fatal result. With absence of the parietal pericardium, the heart is freely moveable, both on respiratory movements and with changes of body position; physical examination and x-ray study should reveal this extreme mobility. With this mobility there is the tendency for kinking of the great vessels and for sudden death. If the heart lies within the abdomen or outside the chest wall (ectopia cordis), the baby will either be born dead or survive but a few days.

Congenital idiopathic hypertrophy. These cases have been extremely rare in recent years, because of special studies which have separated from it cases of von Gierke's disease and cases of hypertrophy due to an abnormal coronary blood supply. In this anomaly, the heart is two or three times the normal size and weight, as is easily elicited by examination and x-ray. It is uniformly enlarged, roundish in shape, and the electrocardiogram is usually within normal limits. The symptoms and signs of circulatory embarrassment soon become evident, and the average duration of life is ten months (Abbott).

Dextrocardia. There are two types of congenital dextrocardia, occurring with about equal frequency. In one type the heart is merely shift-

ed to the right side of the chest and slightly rotated clockwise. There is no transposition of the heart chambers or abdominal viscera. Almost invariably there are associated grave cardiac anomalies, and the prognosis depends upon these anomalies, not the dextrocardia. In the other or "mirror type" there is a transposition of the chambers, the systemic chambers lying on the right side and the pulmonic chambers on the left side. Almost invariably the abdominal viscera are also transposed. There are usually no associated cardiac anomalies, unless the dextrocardia is isolated, and the condition is clinically unimportant. Routine physical examination and roentgen ray examination usually disclose the dextrocardia and a pathognomonic sign of the "mirror type" is electrocardiographic; complete inversion of Lead one and transposition of Leads two and three.

Transposition of the great vessels. This is a serious anomaly, in which the aorta arises from the right ventricle and the pulmonary artery from the left ventricle, occurring more frequently in males than females by a four to one ratio. If uncomplicated, survival of the infant is but a matter of hours or a few days. Usually, though there is an associated defect which permits survival for years, but rarely to an adult age. Abbott reports that in a series of 94 cases of transposition, the ductus Botalli was patent 63 times, a patent foramen ovale or auricular septal defect 79 times, and an interventricular septal defect 31 times. Cyanosis is almost invariably present, frequently very marked and accompanied by clubbing of the fingers. There is right ventricular enlargement and the electrocardiogram shows marked right axis deviation. Orthodiagram may show the anomaly in the oblique views. There are no murmurs unless the complicating defects mentioned are present.

In the condition known as corrected transposition of the great vessels, the aorta and pulmonary artery are in abnormal position regarding each other, but do arise from the correct ventricles. This condition is of no clinical importance unless associated with other anomalies.

Partial transposition of the great vessels is a third type in which both the pulmonary artery and aorta arise from the same ventricle. Frequently there is cyanosis and the average duration of life is $7\frac{1}{2}$ years in the fifteen cases collected by Abbott.

Interauricular septal defects. Patency of the foramen ovale is the most frequent of all congenital cardiac anomalies and, fortunately, of the least importance. In many cases it is anatomically patent, but closed functionally, having no effect upon cardiac efficiency. Clinically unimportant patency has been reported in nearly one quarter of all autopsied cases. Wide patency is usually associated with other more important congenital anomalies. In 112 cases of widely patent foramen ovale, only 32 were instances of pure or primary patency (Abbott). Uncomplicated well-marked patency may or may not be the cause of serious trouble, although usually there is right sided enlargement of the heart, with right axis deviation. In about one-third of the cases the pulmonary artery is dilated and the aorta is hypoplastic. Presystolic and systolic murmurs over the upper part of the sternum, accompanied in rare cases by thrills, have been reported in about one-fourth of the cases. With a history of cyanotic spells or undue cyanosis during pulmonary infections, patency of the foramen ovale should be given consideration. Subacute bacterial endocarditis is a rare complication, but there have been several instances of paradoxical embolism.

The defect may also occur in the upper or lower part of the interauricular septum. These are more serious than foramen ovale patency, and almost invariably are accompanied by other cardiac abnormalities. The clinical signs are the same as for patency of the foramen ovale. In any of these anomalies, cyanosis is infrequent but may occur as a terminal event.

Interventricular septal defects. This is the second most frequent congenital cardiac anomaly, occurring in 27.4 per cent of Abbott's 1,000 cases. The defects are almost invariably at the base, just below the aortic valve; in Abbott's series 257 of the 274 defects were basal in position. Other congenital defects are generally present, Abbott noting only 50 cases of the pure defect in the total 257 cases of basal septal defects. The septal defect is usually small, 1 to 2 centimeters in diameter, the edges thickened and fibrotic. The right ventricle is usually enlarged and the pulmonary artery slightly dilated. Frequently the electrocardiogram shows right axis deviation, and in rare cases, congenital heart block, when an abnormality of the bundle of His is associated with the septal defect. There are usually no symptoms and the murmur is very characteristic, commonly called "Roger's murmur". This murmur is usually loud and harsh, best heard in the third or fourth interspace, generally in the median line with equal intensity to left and right. It is not heard over the vessels of the neck. The length of the murmur is characteristic. It begins early in systole and may be prolonged into diastole, so that it obscures the second sound. Almost without exception there is an accompanying systolic thrill where the murmur is best heard. At the base of the heart the second pulmonary sound is usually accentuated. Cyanosis is rare in the uncompli-

cated ventricular septal defect, and if present is only part of a terminal condition. A rather frequent and serious complication is that the defect may become the site of a bacterial endocarditis, which greatly shortens the life expectancy.

Pulmonary Stenosis. Although fetal endocarditis is rare, when it does occur the pulmonary valve is by far the most frequently involved, with the aortic valve a poor second. The degree of stenosis may vary from mild to complete atresia of the pulmonary orifice and artery. In 101 of 110 cases of pulmonary stenosis in Abbott's series, there were complicating septal defects, either ventricular (51 cases) or auricular (16 cases), or both (34 cases). Two distinctive features are cyanosis and clubbing of the fingers and toes, the degree of which depends upon the degree of the defect. Adult life may occasionally be attained before the cyanosis is noticeable, but once it occurs, it is likely to be progressive, with polycythemia also entering the picture. The cardiac impulse may be heaving and diffuse in character. There is a systolic thrill and harsh systolic murmur at the pulmonary area, with transmission of the murmur towards the left shoulder, rather than to the vessels in the neck. The second pulmonic sound is feeble or absent. The right ventricle is hypertrophied, the right auricle enlarged, and there is a prominent pulmonary conus to x-ray and orthodiagram. The electrocardiogram may show right axis deviation and there may be T-wave changes in Leads II and III, significant of right heart strain. Not a few patients succumb to subacute bacterial endocarditis.

Tetralogy of Fallott. This well known congenital condition consists of a basal interventricular septal defect, pulmonary stenosis, dextro-

position of the aorta and right ventricular hypertrophy. It is a serious condition, usually limiting activity and duration of life considerably. Abbott has noted that "the entire pulmonary tract is usually narrowed and hypoplastic, the pulmonary valve is bicuspid and its leaflets frequently fleshy in character, and the conus is narrowed and deformed or constricted at its lower bulbar orifice, while the wide thick walled aorta rides above the large defect in the interventricular septum receiving blood through this from both ventricles." The far greater percentage of cyanotic people attaining adult life are those with this combination of defects. In addition to the cyanosis, there is frequently present clubbing of the fingers and toes (often marked) and polycythemia varying from 6,000,000 to 12,000,000 red blood cells per cubic millimeter. Murmurs may be absent, but usually there is a rough systolic murmur and thrill, maximum at the second left interspace - the murmur being transmitted into the vessels of the neck. This murmur is often less intense than that which accompanies isolated pulmonary stenosis. The pulmonary second sound is weak and may be absent. The systolic murmur generated at the defect may also be heard in the back, below the left scapula. The heart becomes sabot-shaped because there is a prominent right ventricle without enlargement of the pulmonary artery. The great vessels may be prominent on the right side because of the dextroposition of the aorta, and there is electrocardiographic evidence of right ventricular strain.

Eisenmenger Complex. This is much more rare than the tetralogy of Fallott, but quite similar to it pathologically. Here the congenital defects consist of dextroposition of the aorta overriding a basal interventricular septal defect, a hypertrophy of the right ventricle, but a normal

or increased size. of the pulmonary infundibulum, valve and artery. Obviously this is a much less serious condition than the tetralogy. Cyanosis and clubbing in these cases are moderate, and usually occur later in life. The physical signs are the same as those of isolated interventricular defect, as is the electrocardiogram. Orthodiagram and x-ray may reveal the destroposition of the aorta.

Lutembacher's Disease. This is an interesting condition of an acquired organic disease, mitral stenosis, added to a congenital anomaly, interauricular septal defect. The signs and course of marked mitral stenosis are somewhat changed by this combination. The left auricle tends to remain small and the right side of the heart hypertrophies. There is a prominent pulmonary conus and the pulmonary vessels are often increased in size. A mitral stenotic murmur is heard at the apex, often accompanied by a thrill, and frequently there is a high pitched murmur over the upper part of the sternum, which may have a thrill associated with it. The aortic knob may be smaller than normal and electrocardiogram may show marked right axis deviation. There have been several cases cited of persons with this disease living to the sixth and seventh decade of life, without much impairment of their activity (Lutembacher and Bonnabel). On the other hand, White does not feel that the septal defect aids much in relieving the heart or lungs in the presence of mitral stenosis. The end picture is usually one of right heart failure with terminal cyanosis.

Pulmonary regurgitation. This is one of the rarest of congenital cardiac defects and there is usually an abnormal number of cusps, two or four. Cyanosis may or may not be present and may occur only with exertion.

The right ventricle is usually hypertrophied, and the electrocardiogram is in accord with this. There is an early, blowing diastolic murmur along the left sternal border, maximum at the pulmonary area, and the pulmonary second sound is accentuated. A visible pulsation is usually evident in the second left interspace and a diastolic thrill may be palpable. The pulmonary artery may be dilated and pulmonary hilum shadows seen fluoroscopically. Right heart failure is frequently the end picture.

Aortic Stenosis. The stenotic part in this defect may be of the valve itself or of the infundibulum, a few millimeters below the valve. The latter is more commonly called subaortic stenosis. Stenosis of the valve itself is the result of fetal endocarditis, while the infundibular stenosis is structural in origin. The two types occur with about equal frequency. Naturally their effect upon the heart and body in general depends upon the degree of stenosis, consequently the prognosis is likewise variable. Of major importance is the tendency it presents to become the seat of an acute infective or chronic inflammatory process in early adult life. A harsh systolic murmur is heard at the base of the heart, especially in the aortic area, that is transmitted to the neck vessels, and associated with a systolic thrill. The forceful action of the left ventricle is out of proportion to the weak peripheral pulse. Symptoms are usually absent, especially in the subaortic stenosis type, until heart failure occurs, as it usually does eventually. There will also be evidence of left ventricular strain, both roentgenologically and electrocardiographically.

Coarctation of the Aorta. This is a localized narrowing of the aorta, occurring to a greater or lesser degree. There are two chief types, the

infantile and the adult. The infantile consists of a narrowing of that part of the ductus arteriosus, and sometimes also of the arch proximal to that region. Usually if an extreme degree, it is often associated with other important anomalies and is generally incompatible with long life. In a series of nine cases (Abbott) the maximum duration was nine months, with a mean of eight hours. Of course, if the narrowing were of mild or moderate degree the prognosis would be much more favorable. The adult type is a very localized constriction of the aorta at, or more frequently just below, the point of insertion of the ductus arteriosus, which sometimes remains patent. If the condition is extreme there is a greater liklihood of other cardiac anomalies, although frequently the adult type is uncomplicated. The most frequently associated cardiac anomaly is the bicuspid aortic valve. The result of pronounced adult coarctation presents an interesting picture. Above the constriction, the aorta is usually considerably dilated, sometimes with an aneurysm, and often narrowed below. A collateral circulation, often of high degree, is developed, with widely dilated, tortuous interval mammary, scapular and intercostal arteries carrying blood to the lower part of the body. The heart becomes hypertrophied and dilated in the majority of cases. There is a predominance in the male sex and the patients are usually of above average intelligence and of great bodily vigor. A pathognomonic sign of adult coarctation is the inequality of blood pressure and pulse fullness between the upper and lower extremities, the brachial systolic pressure usually being elevated (170 to over 200 millimeters of mercury), while the femoral systolic pressure is low (100 millimeters or less). The abdominal and femoral pulses are small, weak and difficult to palpate, while the radial pulse is full and strong. The collateral circulation may be

visibly evident, or at least pulses in the intercostal and scapular arteries are frequently palpable and long systolic murmurs may be heard over them, sometimes accompanied by palpable thrills. Another pathognomonic sign, when present, is x-ray evidence of erosion and notching of the undersurface of the ribs due to the dilated intercostal arteries. The x-ray may also show a decrease or absence of the aortic knob shadow. In most cases there is a loud systolic murmur over the precordium, maximum at the base of the heart and well heard in the interscapular area. The orthodiagram and electrocardiogram reveal evidence of left ventricular enlargement and strain. Coarctation of the aorta is probably more common than is generally supposed and should at least be considered in young and middle-aged persons with hypertension. One fact that may be misleading is that the brachial systolic pressure is not always high in coarctation, but is always higher than the femoral systolic pressure. Subacute bacterial endocarditis may occur, but more often the terminal picture is that of congestive heart failure, sudden heart or aortic rupture or of cerebral complications.

Patent ductus arteriosus. This vessel, also called the "ductus Botalli", is normally patent in fetal life, shunting most of the blood from the pulmonary artery to the aorta. Usually it becomes obliterated shortly after birth, being converted into a fibrous cord, the ligamentum arteriosum, but may remain patent for several weeks. If it is patent three months after birth it is regarded as a congenital anomaly. (White). Patency of the ductus may be due to an unexplained arrest of development, but frequently it is a compensatory condition caused by other congenital cardiac anomalies such as infantile coarctation of the aorta or transposition of the great

vessels. The degree of patency may vary from that of a fine canal the size of a bristle to a canal the size of a finger. It may be very short (5 mm.) if the aorta and pulmonary artery are almost contiguous or it may be several centimeters long. The ductus usually communicates with the descending portion of the aortic arch, but may connect with the ascending or transverse portions instead. Usually it is cylindrical or funnel-shaped, but occasionally it may be dilated to form a kind of aneurysm. If the patency of the ductus is of considerable degree, the pulmonary artery is dilated and both ventricles are enlarged, the left usually more than the right. The right ventricle has to overcome pressure that is directed against its own stream by the ductus blood flow, while the left ventricle has to compensate by increased output for the diversion of blood from the systemic circulation. While uncomplicated cases of patency of a small degree may not impose a serious burden upon the heart, there is always the menace of subacute bacterial endocarditis being superimposed upon the defect. The pulmonary end of the ductus with its immediate adjacent tissue is always the initial seat of this infection (Abbott). A rare complication is rupture of the dilated pulmonary artery.

In a typical case of patent ductus arteriosus the diagnosis is relatively easy. Gross maintains that the physical signs of a patent ductus are almost always typical after the fourth year of life, but an analysis of Abbott's cases does not concur with this contention. The most pathognomonic sign of a patent ductus is a continuous, loud, harsh murmur with late systolic accentuation, maximum at the pulmonary area and usually transmitted towards the left clavicle - never to the neck. The quality of the murmur has been variously described as "machinery", "train in a tunnel" or "rolling

thunder". It is also well heard posteriorly in the left interscapular region on expiration. In infants it is only systolic in time, almost without exception. In Abbott's nineteen cases of infants with a patent ductus only one case had the murmur extending into diastole. Abbott reported that thirty per cent of her series of 92 cases of uncomplicated patent ductus had only a systolic murmur. With about twenty per cent of these occurring in infants, there is a remaining ten per cent that occurs in adults or in childhood. The presence of only a systolic murmur at the cardiac base may make the diagnosis of patent ductus more difficult but not remarkably so. The pulmonary second sound is accentuated. The murmur is generally accompanied by a thrill in the pulmonary area, either continuous or systolic in time. Bullock, Jones and Dolley strongly believe that after early childhood a patent ductus, without exception, produces a double or continuous murmur. To substantiate their belief, they present eleven cases with a typical murmur, and in reviewing the literature noted that it was described in 25 out of 27 cases. They also reviewed Abbott's cases and felt that the records therein were often incomplete and many were not carefully studied before death.

The second important sign is the unusual prominence of the cardiac shadow in the region of and just above the pulmonary artery, without the presence of mitral stenosis, evident by percussion and to the x-ray and orthodiagrma. In many cases the abnormal bulging is more marked than in any other condition, except a large interauricular septal defect.

If the patency is of large degree, the signs associated with aortic regurgitation are present, namely Corrigan pulse, wide pulse pressure

(systolic pressure usually being normal), capillary pulse, Duroziez's sign and hypertrophy of the left ventricle. Gross has noted that "one of the interesting features about the low diastolic pressure is the fact that it becomes lower still during exercise." The heart is usually overactive. There may also be engorgement and pulsation of the pulmonary vessels at the hilus of the lung. Children with a wide patency usually show impairment of growth and development. The electrocardiogram is frequently normal, but may show left axis deviation and occasionally right axis deviation, depending upon the size of the opening.

As this is an arterio-venous shunt, cyanosis is rare, transient when it does occur during life and is most frequently seen only as a terminal event.

Cor Triloculare. There are two types of three-chambered hearts; in one the interauricular septum is entirely absent and in the other the interventricular septum is missing. The former type is the least serious, and unfortunately the more rare. The shunt is arterio-venous for the most part, so cyanosis is of only moderate degree if it occurs at all - except as a terminal event. There are no murmurs or thrills in this anomaly and the heart may be but little enlarged, making the diagnosis difficult or impossible. Abbott reports that one case lived to be 31 years old and the mean age in five cases was six years.

In the cor triloculare biatriatum type, there is free mixture of venous and arterial blood in the common ventricle and cyanosis is usually present, although varying in degree from slight to moderate, with clubbing absent or slight. These patients have a fair expectation of life until the third or fourth decade.

Anomalies of the Coronary Arteries. Most of these anomalies are of no clinical importance and so will not be discussed, but there is one serious anomaly, namely origin of the left coronary artery from the pulmonary artery. With venous blood in the coronary artery, the result is hypertrophy and dilatation of the left ventricle, myocardial necrosis and fibrosis, and death is a matter of days or months. White reports of one baby who suffered attacks closely resembling angina pectoris and whose electrocardiogram showed inverted "coronary" T waves in Leads I and II.

DATA FROM WISCONSIN GENERAL HOSPITAL.

Hospital Number	Age	Sex	Symptoms	Cardiac Signs	Associated Signs	Electrocardiogram	O
200975	21 mo.	M	Tachycardia. Occasional dyspnea.	Harsh to and fro murmur maximum along left sternal border but heard over entire chest. P_2 louder than A_2. Continuous grating thrill in 3rd left interspace, widely transmitted.*	Cyanosis of nails. Early clubbing. Retarded development and malnutrition. B.P. 130/?		Ri si en fl vi ar pr
89607	22	M	Precordial pain. Dyspnea on moderate exertion. Palpitation.	Harsh systolic murmur heard over entire precordium, loudest over 3rd left interspace, transmitted to left interscapular region.	None. B.P. 132/72 R.B.C. 4,780,000	Right axis deviation.	Pr mo ca la fr / co co he
78158	5 mo.	M		Loud, systolic, machinery murmur best heard over pulmonic area, with a systolic thrill present there. Murmur widely transmitted.*	None.		Ma en th to
97607	37	M	Dyspnea, Palpitation and weakness.	To and fro murmur at 4th left interspace. Systolic murmur at base. P_2 accentuated. Systolic thrill at tricuspid area. Systolic and diastolic thrill at pulmonic area.	Cherry lips since childhood. No clubbing. R.B.C. 5,060,000	Right axis deviation. Auricular flutter. Moderate myocardial changes	Ma la ch ri en ti ar se ov au ta
97290	2 mo.	F	Heaving of precordium. Dyspnea.	Harsh systolic murmur over precordium. Systolid thrill at apex.	Cyanosis. Shallow respirations.		Ri si en x
95248	47	M	Palpitation, dyspnea, orthopnea and precordial pain.	Diastolic and harsh systolic murmur at aortic area. Extrasystolic arrhythmia and cardiac decompensation. Presystolic gallop rhythm. Systolic and diastolic murmurs at apex.	Subcyanosis of nails and mucous membranes. B.P. 160/70.	Left ventricular strain.	Ma ve la eu of ac di ac ar er no
3761	2 wks.	M	Marked dyspnea.	Cardiac enlargement. No cardiac murmurs.	Very cyanotic.		
4532	4 mo.	F	Dyspnea and weakness	Cardiac enlargement. No cardiac murmurs.	Cyanosis, marked.		Ma er x-
44476	18 mos.	F		Continuous thrill in 1st and 2nd left interspace. Systolic thrill at apex. Loud continuous murmur in 1st and 2nd left interspace. Loud harsh systolic murmur in 4th left interspace, transmitted laterally.*	Blood pressure 90/20. Pulse almost Corrigan. Capillary pulse.		Le er Di mo Ma er mo Fr †
65904	1 yr.	M		Systolic thrill and murmur and diastolic murmur greatest over pulmonic area. Two years later, a precordial systolic murmur.	Retarded development.		
87631	3 mo.	F		Continuous "humming" murmur over entire precordium, at times being a harsh systolic murmur.	Dark purplish-red lips.		Ca en x-
87701	5 yrs.	M	Retarded development and growth.	Systolic murmur at 1st and 2nd left interspace. P_2 louder than A_2.*	B.P. 90/70. Prominent veins of head and neck, and upper chest.	Possible pericardial involvement.	Di pu te pe ca at

iogram	Orthodiascopy and X-ray	Other Congenital Defects	Birth History	Clinical Diagnosis	Anatomical Diagnosis
	Right and left sides of heart enlarged to fluoroscopic view. Pulmonary arch not prominent.	Phimosis	Negative	Interventricular septal defect.	Living
	Prominent pulmonic arch. No cardiac enlargement, frontal area ≠ 12%. Shape consistent with congenital heart disease.	None	Negative	Interventricular septal defect.	Living.
	Marked cardiac enlargement to the left side to x-ray.	None	Negative	Interventricular septal defect, with a mixture of lesions quite likely.	1. Defect in pars membranacea. 2. Right cardiac enlargement. 3. Dilatation of first part of pulmonic artery.
deviaular derate changes	Markedly enlarged heart, chiefly to the right. Prominent, overactive pulmonary arch and vessels. Markedly overactive left auricle. Frontal area ≠60%.	Bilateral pes cavus. Deformed bones of right wrist and forearm, with atrophy.	Blue baby.	Interventricular septal defect. Cardiac enlargement. Possible pulmonary lesion.	Living.
	Right and left sides of heart enlarged to x-ray.		Cyanotic since birth.	Pulmonary stenosis. Interventricular septal defect.	Died at 2 months.
cular	Marked left ventricular enlargement. Aneurysm at base of ascending aorta. Marked dilatation of aorta. Frontal area ≠88.7%. No erosion of ribs noted on x-ray.	None.	Negative	Syphilitic heart disease, with aneurysm of aorta, cardiac enlargement, aortic and mitral insufficiency, F.C. III.	Coarctation of aorta infantile type, cardiac hypertrophy and dilatation. Aneurysm of ascending aorta. Aortic insufficiency.
		Cleft palate. Deformed ears, Bilateral absent radius. Hare lip. Congenital atelectasis. Supernumery spleens.	Negative	No cardiac diagnosis.	Transposition of great vessels. Hypertrophy of right ventricle (great). Absence of inter-auricular septum. Malformed tricuspid valve.
	Marked cardiac enlargement to x-ray.	Cleft palate.	Negative	Congenital heart disease.	Died at 4 months.
	Left cardiac enlargement. Dilated pulmonary artery. Marked prominence of pulmonic arch. Frontal area + 28.8%.	None.	Negative	Patent ductus arteriosus. Interventricular septal defect. Questionable coarctation of aorta, and pulmonary stenosis.	Living.
		Idiocy, possibly due to birth injury. Symmetrical internal hydrocephalus.	Instrumental birth with trauma.	Congenital heart disease.	Living.
	Cardiac enlargement to x-ray.	Partial cleft palate. Micrognathia.	Negative	Congenital heart disease.	Living.
icarment.	Dilatation of pulmonary artery and superior vena cava is moderately dilated.	Lorraine type of infantilism.	Negative	Pulmonary stenosis, possibly. Dilatation of of pulmonary artery and superior vena cava.	Living.

								x-ray.
44476	18 mos.	F		Continuous thrill in 1st and 2nd left interspace. Systolic thrill at apex. Loud continuous murmur in 1st and 2nd left interspace. Loud harsh systolic murmur in 4th left interspace, transmitted laterally.*	Blood pressure 90/20. Pulse almost Corrigan. Capillary pulse.			Left cardi enlargeme Dilated p monary ar Marked pr ence of p monic arc Frontal a + 28.8%.
65904	1 yr.	M		Systolic thrill and murmur and diastolic murmur greatest over pulmonic area. Two years later, a precordial systolic murmur.	Retarded development.			
87631	3 mo.	F		Continuous "humming" murmur over entire precordium, at times being a harsh systolic murmur.	Dark purplish-red lips.			Cardiac e enlargeme x-ray.
87701	5 yrs.	M	Retarded development and growth.	Systolic murmur at 1st and 2nd left interspace. P_2 louder than A_2.*	B.P. 90/70. Prominent veins of head and neck, and upper chest.	Possible pericardial involvement.		Dilatatio pulmonary tery and perior ve cava is m ately dil Frontal a + 25.3%.
209472	7 wks.	F		Harsh systolic murmur at 3rd and 4th left interspace. Tachycardia.	Cyanosis. B.P.59/? Marked cyanosis when crying. Dusky skin.			No cardia largement x-ray.
109976	11 yrs.	M	Dyspnea on exertion. Occasional precordial pain. Palpitation and vertigo.	Systolic thrill and harsh murmur over entire precordium, maximum in 3rd and 4th left interspace, widely transmitted.*	Clubbing of fingers. Cyanosis. B.P. 100/85. Watch glass nails.	Right axis deviation; suggests a congenital lesion.		No cardia largement orthodiag
214447	4	F	Convulsions, apparently brought on by effort.	Continuous thrill and a harsh, continuous blowing murmur at pulmonic area, widely transmitted. Systolic thrill at apex. Precordial heaving.	B.P. 90/50. Retarded development and growth.			Left card Enlargeme with prom pulmonic Frontal a + 20.9%.
42451-A	5 days	F	Dyspnea.	Systolic murmur along left border of sternum.	Subcyanosis since birth.			Marked ca enlargeme x-ray.
94274	14	F	Tachycardia; dyspnea, paroxysmal, nocturnal dyspnea.	Systolic thrill and murmur and short diastolic murmur at pulmonic area. Murmurs loud and rasping. Systolic murmur in 3rd and 4th left interspace, transmitted laterally.*	Hirsutism. B.P. 100/80. R.B.C. 6,310,000. Clubbing. Cyanosis of lips, hands and feet. Watch glass nails.	Right axis deviation and right heart strain.		Right car enlargeme Frontal minus 12 Aortic a markedly active.
202711	16 mos.	M		Systolic blowing murmur over entire precordium.	Pulse rate 140. Retarded development.			
204581	28	F	Headaches. Dyspnea, palpitation and arrhythmia on moderate exertion.	Loud, harsh, blowing systolic murmur at pulmonic area, widely transmitted. Short early diastolic blow along left sternal border. Both murmurs heard posteriorly.*	B.P. Rt. Lt. 160/90 150/90 Arms 130/100 125/105 Legs	Definite myocardial involvement. High 'P' waves.		No cardi largemen minent p ic arch. tionable of contr of desce portion aorta. F area -7. x-ray ev of erosi

		Idiocy, possibly due to birth injury. Symmetrical internal hydrocephalus.	Instrumental birth with trauma.	Congenital heart disease.	Living.
	Cardiac enenlargement to x-ray.	Partial cleft palate. Micrognathia.	Negative	Congenital heart disease.	Living.
pericarlvement.	Dilatation of pulmonary artery and superior vena cava is moderately dilated. Frontal area + 25.3%.	Lorraine type of infantilism.	Negative	Pulmonary stenosis, possibly. Dilatation of of pulmonary artery and superior vena cava.	Living.
	No cardiac enlargement to x-ray.	Cleft palate. Hare lip.	Negative.	Interventricular septal defect.	Died at 10½ months.
is deviagests a l lesion.	No cardiac enlargement to orthodiagram.	Cryptorchidism. Epilepsy	Negative.	Interventricular septal defect.	Died at 12 years.
	Left cardiac Enlargement with prominent pulmonic arch. Frontal area + 20.9%.	None.	Cyanotic at birth	Patent ductus arteriosus	Living.
	Marked cardiac enlargement to x-ray.	Partial transposition of abdominal viscera.	Jaundice Subcyanosis.	Congenital heart disease.	Triloculare biatriatum heart. Multiple defects; see complete description in test.
s deviaright ain.	Right cardiac enlargement. Frontal area minus 12.3%. Aortic area markedly overactive.	Malformation of proximal end of umbilical cord at birth.	Negative.	Tetrology of Fallot. Low cardiac reserve. F.C.III.	Still living at age of 20 years.
		Mongolian Idiocy.	Negative.	Congenital heart disease.	Living.
myocardial nt. High .	No cardiac enlargement. Prominent pulmonic arch. Questionable area of contriction of descending portion of aorta. Frontal area -7.3%. No x-ray evidence of erosion of ribs.	None.	Negative	Coarctation of aorta. F. C. II.	Living.

Age	Sex	Symptoms	Cardiac Signs	Associated Signs	Electrocardiogram	Orthodiascopy and x-ray
2 mos.	F	Tachycardia Dyspnea	Harsh systolic and diastolic murmurs at pulmonic area, transmitted to neck vessels.*	Cyanotic since birth. Early clubbing of fingers. Weakness. R.B.C. 5,100,000.		Normal heart to x-ray
17	F	Shortness of breath; palpitation. Occasional vertigo and G-I symptoms.	Systolic thrill in 3rd and 4th left interspace. A_2 louder than P_2. Loud, harsh systolic murmur, loudest in 4th interspace, transmitted to abdomen and back and neck.*	B. P. 110/85. Subcyanosis of lips and fingernails. R.B.C. 5,002,000	Right axis deviation. Slight myocardial change.	Right cardiac enlargem and dilated left pulmo ary artery by orthodia scopy. Questionable de traposition of aorta. Frontal area + 22.3%
24	F	Precordial pain. Dyspnea on moderate exertion.	Continuous murmur in 2nd and 3rd left interspace. Diastolic murmur along left sternal border. P_2 faint, Systolic thrill alongleft sternal border.*	B. P. 132/48 Capillary pulse, pistol shot and Durosiez's sign present.		Moderate cardiac enlar ment. Prominent pulmon arch. Overactive dilat aorta. Increased vascu lar markings throughou both lung fields. Fron area + 33.0%.
15	M	Shortness of breath.	Systolic thrill and murmur at pulmonic area. P_2 soft. Systolic murmur at apex.	B.P. 122/58.	Right axis deviation.	No cardiac enlargement Prominent pulmonic arc Frontal area -6.4%. S prominence of right a cular salient.
3 mos.	F	Dyspnea since birth.	Systolic murmur maximum at 3rd and 4th left interspace, widely transmitted. P_2 louder than A_2.*	Cyanosis.		
17	F	Dyspnea on moderate exertion. Arrhythmia and palpitation.	Systolic thrill and diastolic shock at pulmonic area. Systolic and early soft diastolic murmur at pulmonic area.	Subcyanosis. B. P. 115/85. R.B.C. 5,920,000	Right axis deviation	No cardiac enlargemen Prominent pulmonary a and sharp pulsation o main branches of pulm ary artery, poorly su tained.
3 yrs 11 mos.	M	Dyspnea, orthopnea on mild exertion Frequent low sternal pain	Systolic and diastolic thrill and murmur at pulmonic area. Systolic murmur at tricuspid area. Systolic thrill and murmur at apex.	Clubbed toes. Cyanosis. Tachycardia. B. P. 110/60. Watch-glass nails.	Right axis deviation and strain. Auricular fibrillation. Moderately advanced myocardial involvement.	Cardiac enlargement. minent pulmonic arch. Frontal area + 7.6%.
6 wks.	F	Difficulty in breathing	Protruding precordium. No murmurs.	Attacks of cyanosis.		Cardiac enlargement t x-ray, especially the left side.
11	F	Dyspnea and precordial pain on much exertion.	Harsh systolic thrill in 3rd right interspace. Systolic murmurs in 4th left interspace, near sternum: widely transmitted. Early diastolic murmur in 2nd and 3rd right interspace.*	Clubbing and cyanosis of nails and subcyanosis of lips. B.P. 105/65.	Dextrocardia. Definite myocardial involvement.	Marked cardiac enlarg ment. Dextrocardia.
19 days	M	Dyspnea with periods of apnea.	Systolic murmur over precordium.	Mild cyanosis, that becomes severe when crying.		Normal heart to x-ra
19	F	Precordial pain. Slight dyspnea and palpitation.	Double thrill and continuous machinery murmur in 2nd left interspace. Functional systolic murmur at pulmonic area since surgery.*	Pulsations of neck vessels. B.P. 135/80.	Definite myocardial involvement.	No cardiac enlargemen Prominent pulmonic a Overactive hilar vesse Frontal area -7.9%.
2	F	Dyspnea and tachycardia	Coarse, rasping systolic thrill and harsh loud-pitched systolic murmur at apex, widely transmitted. Short, rough early diastolic murmur at apex.*	B. P. 130/90. Retarded development and growth.		Cardiac enlargement to x-ray, both to left an right.
10	F	Dyspnea, marked on exertion. Orthopnea	Systolic murmur in 2nd left interspace, widely transmitted.*	Cyanosis on exertion. B.P.148/95 Dilated veins over left anterior	Right axis deviation, may be due to a congenital	Normal heart to ortho diascopy. Absent aort knob. Frontal area -7.

Orthodiascopy and x-ray	Other Congenital Defects	Birth History	Clinical Diagnosis	Anatomical Diagnosis
Normal heart to x-ray	Accessory spleen.	Cyanotic since birth.	Patent ductus arteriosus. Interventricular septal defect.	Dextroposition of aorta. Atresia of pulmonary orifice and artery. I-V. septal defect. Patent foraman ovale. Patent ductus arteriosus with anomalous position of ductus, origin in ascending arch of aorta.
Right cardiac enlargement and dilated left pulmonary artery by orthodiascopy. Questionable dextraposition of aorta. Frontal area + 22.3%	None	Blue baby	Interventricular septal defect. Pulmonic stenosis. Possible aortic stenosis. Cardiac enlargement. F.C. III. Possible tetrology of Fallot.	Living.
Moderate cardiac enlargement. Prominent pulmonic arch. Overactive dilated aorta. Increased vascular markings throughout both lung fields. Frontal area + 33.0%.	None.	Negative	Patent ductus arteriosus. Interventricular septal defect; possible rudimentary pulmonary valve cusps. Dilated pulmonary artery. F.C. III. Possible subacute bacterial endocarditis. Questionable aortic regurgitation.	Living.
No cardiac enlargement. Prominent pulmonic arch. Frontal area -6.4%. Some prominence of right auricular salient.	None.	Negative.	Pulmonic stenosis. Interventricular septal defect.	Living.
	Pilonidal cyst and sinus.	Dyspnea since birth.	Interventricular septal defect.	Living.
No cardiac enlargement. Prominent pulmonary arch, and sharp pulsation of main branches of pulmonary artery, poorly sustained.	None.	Negative.	Pulmonic stenosis. Pulmonic regurgitation. F.C. II. Possible patent ductus arteriosus.	Living.
Cardiac enlargement. Prominent pulmonic arch. Frontal area + 7.6%.	None.	Negative.	Pulmonic stenosis and insufficiency. I-V. septal defect. Tricuspid insufficiency.	Living.
Cardiac enlargement to x-ray, especially the left side.	None.	Negative.	Congenital heart disease.	Coarctation of aorta Adult type. Myocardial hypertrophy. Patent foramen ovale. Aortic stenosis. Bicuspid aortic valve. Congenital degenerative arteritis.
Marked cardiac enlargement. Dextrocardia.	None.	Cyanotic for 15 minutes after birth.	Dextrocardia. I-V. septal defect. Cardiac enlargement. Questionable pulmonic stenosis and insufficiency.	Living.
Normal heart to x-ray.	None.	Negative.	Congenital heart disease.	Patent ductus arteriosus. Patent foramen ovale.
No cardiac enlargement. Prominent pulmonic arch. Overactive hilar vessels. Frontal area -7.9%.	None.	Negative.	Patent ductus arteriosus. Possible interventricular septal defect.	Patent ductus arteriosus found at surgery and ligated successfully. Apparent complete recovery.
Cardiac enlargement to x-ray, both to left and right.	Microophthalmia.	Negative.	Interventricular septal defect.	Died at 2 years.
Normal heart to orthodiascopy. Absent aortic knob. Frontal area -7.1%	None.	Negative.	Interventricular septal defect. Pulmonic stenosis. Some lesion of aorta or	Living.

			:apnea.					
215814	19	F	:Precordial pain. Slight dyspnea and palpitation.	:Double thrill and continuous machinery murmur in 2nd left interspace. Functional systolic murmur at pulmonic area since surgery.*	:Pulsations of neck vessels. B.P. 135/80.	: Definite myocardial involvement.	:No cardiac enla :Prominent pulmo :Overactive hilar :Frontal area -7.	
209923	2	F	:Dyspnea and tachycardia	:Coarse, rasping systolic thrill and harsh loud-pitched systolic murmur at apex, widely transmitted. Short, rough early diastolic murmur at apex.*	: B. P. 130/90. Retarded development and growth.		:Cardiac enlargem :x-ray, both to l :right.	
217049	10	F	:Dyspnea, marked on exertion. Orthopnea and palpitation.	:Systolic murmur in 2nd left interspace, widely transmitted.*	: Cyanosis on exertion. B.P.148/95 :Dilated veins over left anterior chest.	:Right axis deviation, may be due to a congenital anomaly.	: Normal heart to : diascopy. Absen :knob. Frontal ar	
217048	15	F	:Precordial pain and dyspnea on mild exertion. Palpitation.	:Rough, low-pitched harsh systolic murmur maximum in 2nd left interspace, transmitted laterally, and upward. Possible diastolic element below left clavicle.*	: B. P. 105/70.	:Right axis deviation. consistent with congenital heart changes.	:Sub-normal cardi :Prominent pulmon :Frontal area -1	
223340	6 mos.	F	:Dyspnea :Tachycardia :- - - - - - :Patient is now 1 yr. old and has no symptoms.	:Loud, harsh, precordial systolic murmur, maximum in 3rd left interspace well heard to the right (Roger's sign) with a localizing area of to and fro murmur over the left nipple.*	:Cyanosis when crying. Sub-cyanosis constantly present.	:Definite myocardial involvement.	:Marked cardiac :ment in all dir :predominantly o :left side to x-	
224483	20	F	:Dyspnea on exertion. :Precordial pain.	:Loud continuous machinery murmur at pulmonic area, with a systolic thrill. Widening of pulmonary conus to percussion.*	: B. P. 110/55.	:Slight myocardial changes.		
82150	42	M	:Moderate dyspnea. :Precordial pain. Arrhythmia.	:Systolic thrill in 3rd and 4th left interspace. Prolonged high-pitched systolic murmur over precordium; blowing diastolic murmur at base of heart. A_2 is accentuated and tambour.	: B.P. Arms 150/70 Legs 0 Corrigan pulse	:Advanced myocardial degeneration and possible coronary involvement.	:Sabot shaped he :ked enlargement :area + 70% to o :gram. To x-ray: :of left ventric :cending aorta. :scalloping of l :gins of posteri :ribs 3-9th.	
66520	1½ mos.	M	:Rapid, deep respirations	:No murmurs heard.	: R.B.C., 8,000,000		:Cardiac enlarge :x-ray.	
65134	2 mos.	M	:Marked dyspnea.	:Loud harsh systolic murmur maximum at base and transmitted up the right side of sternum under the clavicle:	:Marked cyanosis since U.R.I. five weeks ago.		:Moderate cardia :ment to x-ray, :on the right si	

	;Frontal area -7.9%.				:cessful- complete recovery.
	:Cardiac enlargement to x-ray, both to left and right.	: Micro-ophthalmia.	:Negative.	: Interventricular septal defect.	: Died at 2 years.
de- ay a l	: Normal heart to orthodiascopy. Absent aortic knob. Frontal area -7.1%	None.	:Negative.	: Interventricular septal defect. Pulmonic stenosis. Some lesion of aorta or aortic valve, not coarctation.	: Living.
s . t eni-	:Sub-normal cardiac size. Prominent pulmonic arch. Frontal area -13.7%.	None.	:Negative.	:Possible interventricular septal defect or pulmonic stenosis. Probable patent ductus arteriosus.	: Living.
l nt.	:Marked cardiac enlargement in all directions, predominantly on the left side to x-ray.	None.	:Negative.	:Interventricular septal defect. Possible patent ductus arteriosus.	: Living.
ro-		None.	:Negative.	:Patent ductus arteriosus. Subacute bacterial endocarditis.	:Attempt to ligate ductus was unsuccessful due to severe hemorrhage. Patient died several days following surgery, At autopsy, ductus arteriosus was found patent.
myo- de- on ible in- t.	:Sabot shaped heart; marked enlargement; frontal area + 70% to orthodiagram. To x-ray: prominence of left ventricle and ascending aorta. Definite scalloping of lower margins of posterior arcs of ribs 3-9th.	None.	:Negative.	:Coarctation of aorta with aortic insufficiency, enlargement, coronary sclerosis, F.C. III. Relative mitral insufficiency.	: Living.
-	:Cardiac enlargement to x-ray.	None.	:Cyanotic since birth.	:Congenital heart disease. Possible patent ductus arteriosus.	:Patent foramen ovale. I-V septal defect. Ductus arteriosus was closed.
	:Moderate cardiac enlargement to x-ray, especially on the right side.	Hare lip.	:Negative.	:Tetrology of Fallot.	:Died at 5 months.

Hospital Number	Age	Sex	Symptoms	Cardiac Signs	Associated Signs	Electrocardiogram	Orthodiascopy X-ray
220577	11	M	Dyspnea	Systolic thrill in 1st, 2nd, 3rd, 4th left interspaces. P_2 louder than A_2. Systolic murmur maximum in 1st and 2nd left interspace, widely transmitted.*	B. P. 90/50	Right ventricular strain and is consistent with a congenital defect.	Moderate cardia ment to x-ray v prominence of t ary conus.
224460	9	F		Systolic thrill and murmur at apex.	Cyanotic and clubbing of fingers and toes.		
218918	21	F	Dyspnea, palpitation and arrhythmia on moderate exertion.	Double thrill and continuous machinery murmur at pulmonic area, poorly transmitted to left clavicle.*	B. P. 125/90 R.B.C. 5,180,000	Slight myocardial involvement.	Dilated overact arch. Pulmonary mal. Frontal ar
208926	28	M	Weakness, dyspnea, palpitation on exertion.	Systolic thrill at aortic area. Systolic and presystolic thrills at apex. P_2 louder than A_2. Mid-diastolic short, low-pitched murmur at apex. Long, high-pitched diastolic murmur at pulmonic area. Harsh, low-pitched systolic murmurs at aortic and pulmonic areas.*	B.P.Arm 140/60 Leg 70/50 Internal mammary and intercostal arteries pulsating visibly. Leg pulses are feeble.	Diffuse myocardial changes involving the conduction system.	Marked cardiac ment. Frontal a Marked prominen monic arch. Dis notching of the borders of some ribs.
117698	19	M	Palpitation, usually after eating. Dyspnea on exertion. Occasional dull precordial pain.	Slight precordial heaving. Presystolic and systolic thrills at apex and continuous thrill at 2nd left interspace. Loud machinery murmur at pulmonic area, widely transmitted. Double murmur at apex.*	B. P. 150/70	Within normal limits.	Ascending aorta widened. Pulmon mildly prominen tal area + 29.2 cardiac space n
116034	8	F	Dyspnea on mild exertion.	Systolic thrill at pulmonic area. Precordial systolic murmur, most prominent at pulmonic area.*	Cyanosis, mild. B.P. 106/70	Advanced myocardial involvement. Compatible with congenital heart disease and chronic right ventricular strain.	Slight cardiac ment. Prominent ic arch. Fronta + 17.2%.
117528	35	F		Moderately loud blowing almost continuous murmur with systolic accentuation at pulmonic area, widely transmitted.*	B. P. 120/70	Within normal limits.	Moderate cardia ment. Frontal a +26.5%. Promine monic arch.
65657	50	M	Dyspnea and orthopnea on slight exertion. Arrhythmia, palpitation and precordial pain.	Late systolic murmur at apex. Loud systolic murmur at aortic area, transmitted to neck vessels. To and fro murmur over intercostal arteries posteriorly.	B.P. Left. Rt. Arm 248/110 240/102 Leg 132/100 120/108 Pulsations in intercostal and brachial arteries.		Moderate cardia ment, left vent Aortic knob not Frontal area + tinct notching ferior borders posterior ribs, ially on the ri
14793	45	F	Dyspnea, slight orthopnea, precordial pain.	Loud, harsh, systolic murmur at pulmonic area, transmitted to neck vessels. Soft systolic murmur over sternum at 3rd and 4th interspace. Soft blowing early diastolic murmur along left sternal border. Faint systolic thrill over apex and along left sternal border.*	Cyanosis, clubbing and watch-crystal nails of all extremities. B. P. 135/90 R.B.C. 7,900,000	Complete heart block. Marked right ventricular strain. Advanced myocardial degeneration.	Marked cardiac ment to right a Frontal area + Fairly marked d of pulmonic arc
218353	63	M	Frequent tachycardia, arrhythmia.	No cardiac murmurs.	B. P. 100/88	Early diffuse coronary sclerosis.	Heart size norm 2/3 to right of Aortic arch is left of the tra x-ray.
207013	2 days	F	Dyspnea and cyanosis upon crying.	Systolic murmur at 2nd and 4th right interspaces.	Cyanosis when crying.		Dextrocardia wi rather large gl appearing heart

Orthodiascopy and X-ray	Other Congenital Defects	Birth History	Clinical Diagnosis	Anatomical Diagnosis
Moderate cardiac enlargement to x-ray with marked prominence of the pulmonary conus.	None.	Negative.	Tetrology of Fallot with superimposed subacute bacterial endocarditis.	Died at 11 years.
	None.	Negative.	Congenital heart disease with superimposed acute bacterial endocarditis.	Died at 9 years.
Dilated overactive aortic arch. Pulmonary arch normal. Frontal area +8.1%.	None.	Negative.	Patent ductus arteriosus most logical. Diagnosis since surgery: Pulmonary stenosis with possible pulmonary insufficiency.	Patient was operated on and no patent ductus arteriosus was found.
Marked cardiac enlargement. Frontal area +60%. Marked prominence of pulmonic arch. Distinct wavy notching of the lower borders of some of the ribs.	None.	Negative.	Coarctation of aorta, with superimposed rheumatic heart disease, cardiac enlargement, mitral stenosis and insufficiency, aortic stenosis and insufficiency and partial heart block. F.C. III.	Living.
Ascending aorta somewhat widened. Pulmonic arch mildly prominent. Frontal area + 29.2%. Retrocardiac space narrowed.	None.	Negative.	Patent ductus arteriosus. Bifid aortic valve.	Living.
Slight cardiac enlargement. Prominent pulmonic arch. Frontal area + 17.2%.	None.	Negative.	Infundibular pulmonic stenosis, and interventricular septal defect.	Living.
Moderate cardiac enlargement. Frontal area +26.5%. Prominent pulmonic arch.	None.	Negative.	Patent ductus arteriosus	Living. Patient went through pregnancy without any complications.
Moderate cardiac enlargement, left ventricular. Aortic knob not prominent. Frontal area + 25.2%. Distinct notching of the inferior borders of the posterior ribs, especially on the right side.	None.	Negative.	Coarctation of the aorta, adult type. Cardiac hypertrophy.	Living.
Marked cardiac enlargement to right and left. Frontal area + 92.4%. Fairly marked dilatation of pulmonic arch.	None.	Negative.	Tetralogy of Fallot. Marked cardiac enlargement. Complete heart block. F. C. III.	I-V. septal defect, pulmonic stenosis corrected transposition of the great vessels. Right ventricular enlargement.
Heart size normal, but 2/3 to right of midline. Aortic arch is to the left of the trachea, to x-ray.	None.	Negative.	True dextrocardia with a normally situated aortic arch. Arterio-sclerotic heart disease. F.C. II	Living.
Dextrocardia with a rather large globular appearing heart to x-ray.	Absence of left ear, atresia of right auditory canal, left facial paralysis.	Negative.	Dextrocardia (multiple congenital anomalies).	Died at 43 days.

			slight or-	murmur at pulmonic	and watch-crystal	heart block.	ment to right an
			thopnea,	area, transmitted to	nails of all ex-	Marked right	Frontal area + 9
			precordial	neck vessels. Soft	tremities.	ventricular	Fairly marked di
			pain.	systolic murmur over	B. P. 135/90	strain. Ad-	of pulmonic arch
				sternum at 3rd and 4th	R.B.C. 7,900,000	vanced myo-	
				interspace. Soft blow-		cardial de-	
				ing early diastolic		generation.	
				murmur along left			
				sternal border. Faint			
				systolic thrill over			
				apex and along left			
				sternal border.*			
218353	63	M	Frequent	No cardiac murmurs.	B. P. 100/88	Early diffuse	Heart size norma
			tachycardia,			coronary	2/3 to right of
			arrhythmia.			sclerosis.	Aortic arch is t
							left of the trac
							x-ray.
207013	2 days	F	Dyspnea and	Systolic murmur at 2nd	Cyanosis when		Dextrocardia wit
			cyanosis	and 4th right inter-	crying.		rather large glo
			upon crying.	spaces.			appearing heart
99330	59	M		No cardiac murmurs.	B. P. 130/75.		Heart is on the
				Apex beat on the right			side with transp
				side.			of abdominal vis
							x-ray.
64351	2 mos.	M	Dyspnea.	Loud, harsh systolic	Marked cyanosis.		
				murmur over precordium	R.B.C. 5,430,000		
66600	6	F	Tires very	Marked systolic thrill	Subcyanosis of	Slight myo-	Cardiac shape su
			easily.	at apex, faint syste-	lips and nails.	cardial	left ventricular
				lic thrill at pulmon-	B. P. 100/65.	change.	ment. Frontal ar
				ic area. Systolic			Slight prominenc
				thrill in suprasternal			pulmonic arch.
				notch. Loud, blowing			
				systolic murmur at 4th			
				left interspace, wide-			
				ly transmitted. Harsh			
				systolic murmur over			
				carotids, subclavians			
				and axillary arteries.*			
				F. C. III.			
83826	20	F	Some dyspnea	Short systolic thrill	B. P. 110/70	Right axis	Slight cardiac e
			tires easily	at apex. Loud blowing		deviation.	ment. Markeddila
			and some	systolic murmur in 5th		Tachycardia.	pulmonic arch. F
			palpitation	interspace (left),			area + 17.6%. Le
			on exertion.	widely transmitted.			cle normal.
				Short diastolic murmur			
				at 3rd rib.*			
60943	7 mos.	F	Attacks of	Loud, harsh systolic	Dusky hue to face.		Displacement of
			cyanosis and	murmur in 3rd and 4th	No cyanosis.		left, by x-ray.
			convulsions.	left interspace, trans-	R.B.C. 3,220,000		
				mitted laterally to	Mental retardation		
				the left. Murmur is			
				loud at pulmonic area			
				also.*			
57540	7 mos.	F		Very loud systolic	Retarded develop-		
				murmur over apex.	ment.		
55853	24	F	Dyspnea, pal-	Loud, harsh, systolic	Clubbing and cy-	Right axis	Questionable sli
			pitation and	murmur at 5th left in-	anosis of fingers.	deviation.	troposition of a
			weakness.	terspace, widely	Slight cyanosis of	Bradycardia.	Frontal area -7%
				transmitted.*	face. B.P.120/90.		
					Watch-crystal nails:		
					R.B.C. 6,320,000.		

				tricular enlargement.	
use	Heart size normal, but 2/3 to right of midline. Aortic arch is to the left of the trachea, to x-ray.	None.	Negative.	True dextrocardia with a normally situated aortic arch. Arterio-sclerotic heart disease. F.C. II	Living.
	Dextrocardia with a rather large globular appearing heart to x-ray.	Absence of left ear, atresia of right auditory canal, left facial paralysis.	Negative.	Dextrocardia (multiple congenital anomalies).	Died at 43 days.
	Heart is on the right side with transposition of abdominal viscera to x-ray.	Complete transposition of abdominal viscera.	Negative.	Dextrocardia	Living.
		Cleft palate, stubtoe.	Blue baby	Congenital heart disease	Triloculare biatriatum heart. Absent I-V septum, lack of mitral valve, and mitral orifice, pulmonary artery arising from left ventricle, patent foramen ovale, patent ductus arteriosus.
-	Cardiac shape suggests left ventricular enlargement. Frontal area 0.%. Slight prominence of pulmonic arch.	Cleft palate.	Blue baby.	I-V. septal defect, probable dextro-position of aorta.	Living.
	Slight cardiac enlargement. Markeddilatation of pulmonic arch. Frontal area + 17.6%. Left auricle normal.	None.	Negative.	Small I.-V. septal defect. Dilated pulmonary artery. F. C. II. Believe defect located in posterior portion of septum.	Living.
	Displacement of heart to left, by x-ray.	None.	Negative.	Possible interventricular septal defect or pulmonmonary stenosis.	Living.
		Mongolian idiocy.	Negative.	Congenital heart disease	Interventricular septal defect.
	Questionable slight dextroposition of aorta. Frontal area -7%	None.	Blue baby	Interventricular septal defect and dextroposition of aorta.	Living.

Hospital Number	Age	Sex	Symptoms	Cardiac Signs	Associated Signs	Electrocardiogram	Orthodiascopy X-ray
37106	13	F		Systolic thrill and loud roaring continuous murmur at pulmonic area. Systolic murmur at 4th left interspace.*		Right axis deviation.	Pulmonic arch a prominent and s pulsating. Retr space obliterat tal area + 35.8
13238	13	M	Dyspnea on exertion.	Systolic thrill and long rough systolic murmur at 4th left interspace, widely transmitted.*	Marked cyanosis of face. Clubbing and cyanosis of fingers. B. P. 88/60. Hb.105.%. R.B.C. 7,900,000.	Extreme right ventricular preponderance.	Frontal area +
12013	1 mo.	F		Loud systolic murmur at apex.	Moderate cyanosis		Cardiac enlarge x-ray.
86141	15	F	Marked palpitation. Dyspnea on slight exertion.	Marked systolic thrill at aortic area, suprasternal notch, entire sternum and at apex. Fine diastolic thrill at aortic area. Loud, harsh systolic murmur over upper part of sternum, widely transmitted. Loud, blowing early diastolic murmur at 2nd right interspace. High-pitched, musical diastolic blow at upper part of sternum. Marked pre-systolic thrill and murmur at apex.*	B.P.Arms 205/50 Legs 80-90/? Increased arterial pulsations in neck over subclavians, both scapulae and intercostals of chest posteriorly. Marked capillary pulse, brachial pistol shot. Weak femoral pulsation. No abdominal aorta pulsation palpable.	Moderate myocardial changes. Slight right axis deviation.	Marked cardiac e ment, especially ventricle. Front + 72.3%. Pronou sation of ascend which ends abru level of the tra arch. Unexplaine bulging of upper cardiac border pands during ca tole, but may be ing of base of a aorta. Slight de notching of the surface of some upper ribs close spine.
85189	2 mos.	F	Tachycardia.	Faint systolic thrill over precordium. Loud harsh systolic murmur at pulmonic area. Another systolic murmur along left sternal border.*	Cyanosis when crying.		Marked cardiac e ment in the tra diameter, of bo with bulging of monary arch, to
87632	35	F	Palpitation and dyspnea on moderate exertion.	Systolic thrill at apex. Loud, harsh systolic murmur at 4th left interspace.*	B. P. 110/80.	Left axis deviation.	Moderate dilatat pulmonary artery displaced to lef tal area + 4.3%.
92338	17	F	Precordial pain, palpitation, dyspnea and vertigo upon exertion.	Diastolic shock in 2nd left interspace. Systolic murmur at 3rd left interspace. Continuous machinery murmur at pulmonic area.*	B. P. 120/80. R.B.C. 4,980,000.	Slight myocardial involvement.	Prominent and ov pulmonic arch co with congenital Frontal area -26
94526	3 mos.	M		Loud, harsh systolic murmur over entire precordium. Loud booming sound at apex. First or second sounds indistinguishable.	Cyanosis and occasional convulsions.		
119458	21	M	Weakness and dyspnea on exertion.	Soft systolic thrill and harsh, systolic murmur at pulmonic area, poorly transmitted. Palpable pulmonic valve closure.	Marked plethoric cyanosis. Marked clubbing of all extremities. B.P. 140/100. Hb. 22.8 grams. R.B.C. 8,270,000.	Advanced myocardial degeneration. Marked left axis deviation. Delayed I-V conduction.	Enlarged right v Frontal area +8. Slight fulness c ic arch, very sm knob, prominent vena cava by x-r
217687	4	M		Moderately loud, harsh systolic murmur over 3rd and 4th left interspace and has a distinct leathery quality.*		Within normal limits.	Normal heart.
217584	7	M	Palpitation with exertion.	Moderately loud, harsh systolic murmur along left sternal border, widely transmitted.*		Within normal limits.	Slight cardiac ment. Frontal a + 13.4%.
208921	19	F	Frequent palpitation and arrhythmia.	Systolic thrill at apex. Systolic murmur at pulmonic area.	B.P. 140/70 R.B.C. 4,100,000	Slight myocardial changes.	Prominent pulmon tery. Frontal ar
46234-A	3	F		Loud, harsh, systolic murmur at apex.	Clubbing of fingers and toes.		
110404	9	M	Dyspnea and cyanosis on moderate exertion.	Systolic thrill at base. Harsh blowing systolic murmur at base, widely transmitted. Less harsh systolic blowing murmur over bottom of sternum*	B.P. 100/70. Watch-crystal nails.	Suggests cardiac anomaly, and progressive myocardial changes.	Slight pulsation of sternum. Fron + 4.8%.
200745	19	F	Precordial pain.	Thrill at apex, chiefly systolic but with a short diastolic element. Short harsh sys-	B. P. 115/90. R.B.C. 5,250,000.	Right axis deviation. Slight myocardial in-	Cardiac enlargem Frontal area + 3 Prominent pulmon

Orthodiascopy and X-ray	Other Congenital Defects	Birth History	Clinical Diagnosis	Anatomical Diagnosis
Pulmonic arch abnormally prominent and strongly pulsating. Retrocardiac space obliterated. Frontal area + 35.8%.	None.	Negative.	Patent ductus arteriosus: Interventricular septal defect. F. C. I.	Living.
ht: Frontal area + 10%.	None.	Blue baby	Interventricular septal defect.	Living.
Cardiac enlargement to x-ray.	Cleft palate.	Negative.	Interventricular septal defect.	Living.
o-: Marked cardiac enlargement, especially the left ventricle. Frontal area + 72.3%. Pronounced pulsation of ascending aorta which ends abruptly at level of the transverse arch. Unexplained marked bulging of upper right cardiac border which expands during cardiac systole, but may be a widening of base of ascending aorta. Slight definite notching of the inferior surface of some of the upper ribs close to the spine.	None.	Negative.	Coarctation of aorta. Congenital aortic insufficiency. F. C. II.	Living.
Marked cardiac enlargement in the transverse diameter, of box shape with bulging of the pulmonary arch, to x-ray.	None.	Negative.	Tetrology of Fallot.	Living.
Moderate dilatation of pulmonary artery. Heart displaced to left. Frontal area + 4.3%.	None.	Negative.	Interventricular septal defect.	Living.
Prominent and overactive pulmonic arch consistent with congenital lesion. Frontal area -26.7%.	None.	Blue baby.	Patent ductus arteriosus. Probable interventricular septal defect.	Living.
	Cleft palate. Bilateral equinovavus.	Negative.	Pulmonary stenosis with interventricular septal defect.	Interventricular septal defect. Patent foramen ovale. Malformation of pulmonvalve (2 leaflets).
o-: Enlarged right ventricle. gen- Frontal area +8.1%. r-: Slight fulness of pulmon- is: ic arch, very small aortic knob, prominent superior vena cava by x-ray.	None.	Negative.	Interventricular septal defect. Some atresia of pulmonary artery.	Living.
al: Normal heart.	None.	Negative.	Interventricular septal defect.	Living.
al: Slight cardiac enlargement. Frontal area + 13.4%.	petit mal epilepsy.	Negative.	Interventricular septal defect.	Living.
Prominent pulmonic artery. Frontal area -12.2%.	None.	Negative.	Pulmonary stenosis.	Living.
	Congenital obliteration of bile duct:	Negative.	Congenital heart disease.	Living.
r-: Slight pulsation to right y, of sternum. Frontal area s- + 4.8%. es.	None.	Negative.	Possible tetralogy of Fallot.	Living.
Cardiac enlargement. Frontal area + 30.3%. Prominent pulmonic arch.	None.	Negative.	Patent ductus arteriosus.	Living.

				lity.*				
17584	7	M	Palpitation with exertion.	Moderately loud, harsh systolic murmur along left sternal border, widely transmitted.*			Within normal limits.	Slight cardiac ment. Frontal a + 13.4%.
08921	19	F	Frequent palpitation and arrhythmia.	Systolic thrill at apex. Systolic murmur at pulmonic area.	B.P. 140/70 R.B.C. 4,100,000		Slight myocardial changes.	Prominent pulmon tery. Frontal ar
46234-A	3	F		Loud, harsh, systolic murmur at apex.	Clubbing of fingers and toes.			
110404	9	M	Dyspnea and cyanosis on moderate exertion.	Systolic thrill at base. Harsh blowing systolic murmur at base, widely transmitted. Less harsh systolic blowing murmur over bottom of sternum.*	B.P. 100/70. Watch-crystal nails.		Suggests cardiac anomaly, and progressive myocardial changes.	Slight pulsation of sternum. Fron + 4.8%.
200745	19	F	Precordial pain.	Thrill at apex, chiefly systolic but with a short diastolic element. Short harsh systolic murmur at pulmonic area, transmitted to apex. Short early diastolic murmur along left sternal border.*	B. P. 115/90. R.B.C. 5,250,000.		Right axis deviation. Slight myocardial involvement.	Cardiac enlargem Frontal area + 3 Prominent pulmon
201338	31	M	Palpitation, dyspnea and arrhythmia on moderate exertion.	Systolic thrill in 2nd and 3rd right interspaces. Loud, blowing, harsh systolic murmur at 2nd right interspace, widely transmitted. Diastolic gallop rhythm.*	B. P. 150/100. R.B.C. 5,230,000. Marked cyanosis of lips and nail beds		Left axis deviation. Congenital lesion.	Huge cardiac enl mainly right-sid aorta with displ to right. Fronta + 98%.
201696	15	F		Loud rough systolic murmur at base.	R.B.C. 5,280,000		Advanced myocardial involvement. Right bundle branch block.	
211717	1	F		Moderately loud blowing systolic murmur at pulmonic area widely transmitted.*	R.B.C. 3,900,000.			
218333	5 mos.	F		Systolic murmur at 4th left interspace, widely transmitted.*				Cardiac size no x-ray.
224750	18	F	Occasional palpitation.	Systolic murmur at 3rd and 4th left interspace.*			Within normal limits.	Marked prominenc monary conus. Fr area + 4%.
81526	20	M	Some dyspnea.	Systolic thrill in 3rd and 4th left interspace. Loud, blowing, harsh systolic murmur and early diastolic murmur at pulmonic area.*	R.B.C. 5,360,000. B. P. 126/74.		Moderate myocardial degeneration. Right axis deviation.	Dilated pulmonar Moderate cardiac ment to right an Frontal area + 3
98006	10	F	Marked palpitation on exertion.	Faint systolic thrill and loud harsh systolic murmur at pulmonic area.*			Definite myocardial involvement. Slight right axis deviation	Prominent pulmon

-	Prominent pulmonic artery. Frontal area -12.2%.	None.	Negative.	Pulmonary stenosis.	Living.
		Congenital obliteration of bile duct.	Negative.	Congenital heart disease.	Living.
ar- ly, s- r- es.	Slight pulsation to right of sternum. Frontal area + 4.8%.	None.	Negative.	Possible tetralogy of Fallot.	Living.
-	Cardiac enlargement. Frontal area + 30.3%. Prominent pulmonic arch.	None.	Negative.	Patent ductus arteriosus.	Living.
l	Huge cardiac enlargement, mainly right-sided. Small aorta with displacement to right. Frontal area + 98%.	Complete transposi- of abdom- inal vis- cera.	Negative.	Dextro-cardia. Probable pulmonic stenosis. F. C. III.	Dextrocardia. Marked myocardial hypertrophy of left side. Marked pulmonary stenosis. Aortic stenosis.
yo- - le ck.		None.	Negative.	Pulmonary stenosis.	Living.
		Cavernous hemangioma of lip.	Negative.	Interventricular septal defect.	Living.
	Cardiac size normal to x-ray.	Congenital malformation of genitalia.	Negative.	Interventricular septal defect.	Living.
nal	Marked prominence of pulmonary conus. Frontal area + 4%.	Congenital absence of vagina and uterus.	Negative.	Pulmonary stenosis.	Living.
yo- gen-	Dilated pulmonary artery. Moderate cardiac enlargement to right and left. Frontal area + 39%.	None.	Negative.	Patent ductus arteriosus. Possible interventricular septal defect. F.C. I.	Living.
yo- - t tion	Prominent pulmonic arch.	None.	Negative.	Patent ductus arteriosus. of I.-A. septal defect or both.	Living.

Hospital Number	Age	Sex	Symptoms	Cardiac Signs	Associated Signs	Electrocardiogram	Orthodias X-
95004	24	F	Dyspnea on slight exertion.	Moderately loud blowing systolic murmur and high-pitched blowing diastolic murmur at pulmonic area, transmitted to apex. Murmurs become continuous when recumbent. Short early diastolic murmur in 1st and 2nd right interspace.*	Cyanosis of finger-nails.	Slight myocardial involvement.	Aorta disp Pulmonary dilated. M enlargemen area + 29.
214495	50	M		Systolic thrill and soft blowing systolic murmur at 2nd and 3rd left interspace. Short early diastolic blow heart during inspiration only along left sternal border.*	B. P. 150/100 R.B.C. 5,030,000.	Changes of right ventricular strain.	Dilatation aorta. Pro arch with pulmonic v Frontal ar
225304	18	F	Some dyspnea on moderate exertion. Pain in chest.	Slight systolic apical thrill. To and fro thrill and loud, continuous machinery-like murmur at pulmonic area, both with systolic accentuation. Since surgery the thrill and diastolic murmur at the pulmonic area have disappeared.*	B.P. 110/50. Capillary pulse and Traube's pistol shot. R.B.C. 5,310,000	Within normal limits.	Prominent Thrusting area + 10% there was lar enlarg latation o ary pulmon branches. there is p prominent and decrea size. Fron
117861	24	F	Occasional palpitation. Dyspnea on moderate exertion. Occasional precordial pain.	Diastolic and presystolic thrill and loud, early, high-pitched, blowing diastolic murmur at 3rd left interspace.*	Cyanosis and clubbing of fingernails. B.P. 110/85. R.B.C. 5,630,000.	Right axis deviation. Slight myocardial involvement.	Pulmonary prominent. cardiac spa literated. + 13.9%.
109123	34	F	Recent dyspnea on moderate exertion.	Systolic thrill and systolic murmur at apex.*	B. P. 120/90. R.B.C. 5,490,000.	Moderate myocardial degeneration. Right axis deviation.	Prominent Overactive culation. + 16.4%.
114918A	2½ hrs.	F					
225327	21	F	Dyspnea on moderate exertion, arrhythmia, palpitation and severe headaches.	Systolic murmur at apex, widely transmitted.	B.P. Arms 160/100 Legs 100/? Abdominal and femoral arteries difficult to palpate.	Within normal limits.	Absent aor erior ribs wavy notch undersurfa Frontal ar

* Denotes cardiology examination and diagnosis.

Orthodiascopy and X-ray	Other Congenital Defects	Birth History	Clinical Diagnosis	Anatomical Diagnosis
Aorta displaced to right. Pulmonary artery markedly dilated. Moderate cardiac enlargement. Frontal area + 29.5%	None	Negative.	Patent ductus arteriosus. I.-V. septal defect. Aortic regurgitation. Questionable pulmonic stenosis. F. C. III.	Living.
Dilatation of ascending aorta. Prominent pulmonic arch with pulsation of pulmonic vessels. Frontal area -5.5%.	None.	Negative.	Pulmonic stenosis.	Living.
al Prominent pulmonic arch. Thrusting aorta. Frontal area + 10%. On x-ray there was left ventricular enlargement and dilatation of the secondary pulmonary vascular branches. Since surgery there is persistence of prominent pulmonic arch and decrease in cardiac size. Frontal area +4%.	None.	Negative.	Patent ductus arteriosus	Patent ductus arteriosus was successfully ligated by surgery.
Pulmonary arch markedly prominent. Upper retrocardiac space almost obliterated. Frontal area + 13.9%.	None.	Negative.	Pulmonary regurgitation with questionable absence of pulmonary valve leaflet. F. C. II.	Living.
b- Prominent pulmonic arch. Overactive pulmonary circulation. Frontal area + 16.4%.	None.	Negative.	Infundibular pulmonic stenosis.	Living.
	Congenital atelectasis. Accessory spleen. Single fused right kidney.	Cyanotic since birth.	Congenital heart disease	Patent ductus arteriosus with malposition.
al Absent aortic knob. Posterior ribs show distinct wavy notchings along the undersurfaces. Frontal area + 5.2%.	None.	Negative.	Coarctation of the aorta.	Living.

ANALYSIS OF CASES AT WISCONSIN GENERAL HOSPITAL AND COMPARISON WITH ABBOTT'S SERIES

There are many ways of analyzing a series of cases and false conclusions may easily be drawn. Therefore I have included all the data available; this data to be considered a primary analysis and all the facts and figures included so that the reader may make additions, revisions, or repudiations justifiable by his personal experiences. It may be erroneous to compare this series with Abbott's series of 1,000 cases, because of the relatively small number of our cases and because of the low percentage of anatomical diagnoses, since Abbott's cases have all been proven by autopsy. However, the accuracy of the clinical diagnoses has been quite high. All diagnoses that are labeled "possible" or "questionable" are either not included, or if included are considered separately. This series of 85 cases is necessarily limited to hospital patients and does not include the outpatient department or student health cases. The series as a whole will be analyzed first and then the commoner lesions will be considered separately.

In this series 21 per cent of the cases are anatomically proven, 14 by autopsy and 4 by surgery. The sex ratio differs markedly from Abbott's series; 53 (62.3 per cent) were female and 32 (37.7 per cent) were male in the Wisconsin General Hospital series, while the males were in preponderance in Abbott's cases, 58 to 42 per cent of females. Our incidence of associated congenital defects located outside the cardio-vascular system is almost double that in Abbott's series. Thirty cases (35 per cent) had other congenital anomalies while Abbott reported only 18.8 per cent. This high incidence gives added weight to the opinion of many that when a con-

genital anomaly is found, the heart should always be examined to be sure that no cardiac anomaly coexists. Of these associated congenital defects, cleft palate is by far the most commonly encountered, being more than twice as frequent as harelip, deformed bones, accessory spleens, transposed abdominal viscera or idiocy. While only 49 cases (57.6 per cent) were seen by the Cardiology Department, which has special equipment that may be of aid in making the diagnosis, the majority of them have been seen in recent years thus showing a trend towards cardiac consultation. Every possible advantage should be given these patients because of the heavy load they already carry. Of special interest is the accuracy of diagnosis in the 18 cases that have an anatomical diagnosis. In 7 cases the diagnosis was simply congenital heart disease, no attempt to name the type of lesion being made. In all fairness to the departments involved, however, it should be noted that the age in these 7 cases ranged from a maximum of 7 months to a minimum of $2\frac{1}{2}$ hours, with a mean average of $1\frac{1}{2}$ months. It is well known that accurate diagnosis in early infancy is extremely difficult and, therefore, a differentiation is very frequently not possible. Eight cases were examined by a cardiologist and the clinical diagnosis was ultimately proven to be correct in seven of them. The clinical diagnosis in the eighth one was patent ductus arteriosus, and when the patient was operated on, no patent ductus was found. The cardiologist on the case hesitated in making the diagnosis because of some atypical signs, but finally agreed that a patent ductus was the most likely diagnosis. While Abbott's notes show that the combined congenital cardiac defects are more common than cases with individual or isolated defects alone, the reverse is true in our series. Of 85 cases, 44 of them have isolated defects, 35 have combined lesions, and 6

cases are undiagnosed. In 4 of the 44 cases of isolated defects, the diagnosis is uncertain as to the type of defect that exists, but it is believed that only one defect is present, whatever it may be.

In considering the commoner types of cardiac anomalies that occurred in this series, it was thought best to divide each anomaly into two types, the isolated defect and the combined cardiac anomaly. A more accurate picture can be attained by this division. Before considering the individual defects, it is necessary to explain that the percentage of incidence of the isolated defects is obtained from the total number (44) of cases of isolated defects and the incidence percentage for combined lesions is obtained from the total number (35) of cases of combined defects. The percentage of incidence for each cardiac anomaly, whether it be isolated or combined, is determined from the total number (85) of cases in this series.

In our series the interventricular septal defect was the most commonly encountered lesion. There are 17 isolated cases of this type (38.6 per cent), ranging in age from 1 month to 37 years. Nine are females and eight are males. Three cases give a history of cyanosis at birth. Twelve of the cases are still living and eleven have congenital defects elsewhere in the body, a surprisingly high incidence. Cyanosis is present in six and clubbing of the extremities in three cases. Only one person has an elevated red blood cell count, it being 7,900,000 per cubic millimeter. There is at the present no case with a complicating bacterial endocarditis.

In contrast, Abbott reports that only 62 of her 1,000 cases were primary interventricular septal defects, just 6.2 per cent. The sexes were also equal, 29 male to 29 female and 4 were not stated. Only 9 had other

congenital defects; cyanosis was present in 5 cases during life and occurred as a terminal event in 13.

The age range was from foetus to 49 years, with a mean age of 14½ years. There were 13 cases of a complicating bacterial endocarditis. Abbott also reports 228 cases of this defect combined with some other cardiac anomaly. Seven of these had but a dextroposition of the aorta as the additional defect, while 51 had pulmonary stenosis with aortic dextroposition in 32. With a grand total of 290 cases of interventricular defects, the incidence was 29 per cent in her 1,000 cases.

There are 25 cases (71.4 per cent) of combined interventricular septal defects in our series, varying from 2 months to 45 years of age. The sex ratio is 17 to 8 in favor the females. Other congenital defects were present in 5 cases, while 8 give a birth history of cyanosis. Eighteen are still living. There is an increased red cell count in 7, the highest one being 8,270,000 per cubic millimeter, with a hemoglobin of 22.8 grams per cent. Cyanosis is present in 17, clubbing in 8 cases and one patient has developed subacute bacterial endocarditis. The associated cardiac anomaly is pulmonary stenosis in 7 cases, and dextroposition of the aorta in 2 cases, while both of these anomalies were present in 6 more cases and in one case there was dextroposition of the aorta, atresia of the pulmonary orifice and artery, patent foramen ovale and patent ductus arteriosus. Other cases having but one associated cardiac anomaly include 5 cases of patent ductus arteriosis and 2 of patent foramen ovale. The total number of cases with interventricular septal defects is 42 (49.4 per cent).

Pulmonary stenosis is the next most frequent anomaly in our series,

totaling 21 cases (24.7 per cent). There are 6 cases (13.6 per cent) of isolated stenosis, 4 women and 2 men. Two have other congenital defects, all are living and there is no birth history of cyanosis in any. One-half of them have slightly increased red blood cell counts, cyanosis is present in one and one has clubbing. The age varies from 5 to 50 years. Again there is no complicating bacterial endocarditis.

Abbott has only 9 cases of primary pulmonary stenosis and 101 cases complicated by defective septa giving a total incidence of 11 per cent. Of the 9 cases, 5 were male and 4 female, and only one had anomalies elsewhere. The age varied from $10\frac{1}{2}$ to 45 years with a mean age of 22 years. Three patients were cyanotic, one had clubbing and two developed bacterial endocarditis. In the 101 complicated cases, 53 were male and 43 female, 15 had anomalies elsewhere, 82 had cyanosis (usually marked) and 51 had clubbing. The most frequent associated cardiac anomalies were patent ventricular septum (51), patent foramen ovale (16), patent foramen ovale and ventricular septum (34) and patent ductus arteriosus (12) in conjunction with the septal defects.

There are 15 cases (42.8 per cent) of combined pulmonary stenosis in our series, varying in age from 2 months to 35 years. Sex ratio is 9 women to 6 males, 3 have other congenital defects, 2 have birth histories of cyanosis and 10 are still living. Eleven have some degree of constant cyanosis, 4 have clubbing and there are 6 elevated red blood cell counts, the highest being 7,400,000 per cubic millimeter. The other cardiac anomalies are ventricular septal defects (13), dextroposition of the aorta (5), pulmonary insufficiency (2) and one case with patent ductus arteriosus.

Next in order of frequency is the 19 cases (22.3 per cent) of patent ductus arteriosus, 7 of which are isolated (15.9 per cent) and 12 are combined (34.2 per cent). In the isolated cases the age varies from 2½ hours to 35 years, all are women patients, only 1 has associated anomalies, 2 have birth histories of cyanosis, 5 are living and 3 have had surgical ligation of the patent ductus. One has subacute bacterial endocarditis.

In the combined cases, 8 are females and 4 are males, 2 have anomalies elsewhere, 3 were cyanotic at birth and 9 are living. There is one case of a possible subacute bacterial endocarditis and no surgical ligations have been performed on this group. The age varies from 19 days to 24 years. It is interesting to note that the 4 patients that have died came to autopsy and the patent ductus was found to be in an anomalous position in two of them. Most of the associated cardiac anomalies have already been mentioned, except for 2 cases in which the foramen ovale was also patent.

In Abbott's series there is a total of 242 cases (24.2 per cent) of patent ductus, of which 92 are primary lesions and 150 are combined, a ratio closely approximating ours. In the 92 cases the females were in preponderance, 55 to 29. Ten cases had anomalies elsewhere and 22 developed subacute bacterial endocarditis. The age range was 2 weeks to 66 years, with a mean age of 24 years. In the 150 combined cases, the patent ductus was associated with coarctation of the aorta 13 times, with pulmonary atresia 28 times, with transposition of the great vessels 33 times and with pulmonary stenosis in 12 cases. The cause of death in the 92 isolated cases was by heart failure in 40 cases, by bacterial endocarditis or endo-

carditis in 21, by a cerebral lesion in 3 and by broncho pneumonia in 3 cases.

There is a total of 8 cases (9.4 per cent) of coarctation of the aorta in our series, 6 of which are isolated defects (13.6 per cent) and 2 are combined (5.7 per cent). In the isolated cases, the age varies from 21 to 50 years, there are 4 males and 2 females, 5 are still living and the 1 that died at the age of 47 years had an infantile type of coarctation. To my knowledge, there is no other case of infantile coarctation that has been reported that lived beyond infancy. The other 5 cases had the adult type and there are no anomalies elsewhere nor birth histories of cyanosis. Erosion of the ribs is evident to x-ray in 4 cases, and 1 case has superimposed rheumatic heart disease. In the 2 cases of combined defects, both are in females, ages are 6 weeks and 15 years, the latter one is still living, although the defect appears to be in the transverse arch of the aorta. There is slight erosion of the upper ribs to x-ray. The associated cardiac defects are congenital aortic insufficiency in the living case and aortic stenosis, patent foramen ovale and bicuspid aortic valve in the case that died.

Abbott reported 70 cases of adult type of coarctation, ranging from about 3 years to 92 years, with a mean age of 33 years and 9 cases of the infantile type, ranging from 8 hours to 9 months, with a mean age of 1 3/4 months. In the adult type 53 were males and 15 females and 8 had anomalies elsewhere. There were 7 cases of bacterial endocarditis. In the infantile type 4 were men and 2 women and 4 had anomalies elsewhere. The bicuspid aortic valve was the most commonly associated defect, occurring in 50 out of 183 cases. The ductus arteriosus was patent in 6 cases of

the adult type and 7 cases of the infantile type.

There are 5 cases (5.8 per cent) of dextrocardia, 3 being isolated (6.8 per cent) and 2 are combined (5.7 per cent). In the isolated cases 2 are male and 1 female, one has complete transposition of abdominal viscera and one has other anomalies elsewhere and two are living. The one that died was 43 days old and the oldest is 63 years. In the 2 combined cases the sexes are evenly divided, the ages are 11 and 31 years, 1 is living and has a birth history of cyanosis, the other is dead and had complete transposition of the abdominal viscera.

In Abbott's series, there were 18 cases of isolated dextrocardia ranging in age from birth to 49 years, 10 of which were males and 6 females and 7 had anomalies elsewhere. There were also 11 cases of dextrocardia with situs inversus, ranging from foetus to 58 years, 3 males and 4 females and 11 had anomalies elsewhere. Incomplete heterotaxia occurred in 5 of Abbott's cases, all of which had anomalies elsewhere. Four were males and 1 female. In almost all of these cases of dextrocardia, there were other congenital defects in the heart, usually septal defects or a patent ductus arteriosus.

Our series is fortunate enough to include 3 cases (3.5 per cent) of congenital pulmonary regurgitation, a truly rare anomaly. In 1 case, a 24 year old girl, there are no other defects, cardiac or elsewhere, and she is still living. Her red blood cell count is 5,030,000 per cubic millimeter and cyanosis occurs only on exertion. The other 2 cases are combined with pulmonary stenosis, both are living and both have cyanosis. One is a 17 year old female and the other a 4 year old male, who also has clubbing of

the extremities.

There were only 2 cases of isolated pulmonary insufficiency in Abbott's series and in 10 other cases it complicated other cardiac defects. Both of the isolated cases were females, ages 22 and 64 years; one had cyanosis and there were no anomalies elsewhere.

The remaining cases in our series all have combined cardiac lesions. Dextroposition of the aorta is present in 8 cases, 5 women and 3 men. Four have anomalies elsewhere, 3 have a birth history of cyanosis, clubbing is present in 2 and cyanosis in 4. Five are still living. The age ranges from 2 months to 45 years. In 7 of these cases there is an interventricular septal defect, 5 of which also have pulmonary stenosis.

In Abbott's series there were 10 cases of primary dextroposition and 101 cases in which it complicated other defects, especially pulmonary stenosis and (or) ventricular septal defects.

Although patent foramen ovale is generally considered to be the most common of all congenital cardiac lesions, there are only 6 cases in our series, all combined with other cardiac defects. Perhaps the most logical explanation for this discrepancy is that this defect is rarely diagnosed during life, but is frequently encountered at autopsy. All 6 of these cases were diagnosed at post mortem, not during life. The age varies from 19 days to 3 months and in all cases it was the associated cardiac defect that was mainly responsible for the death. There are 4 males and 2 females, 3 have defects elsewhere and 3 have a birth history of cyanosis.

Auricular septal defects complicated 326 cases of Abbott's series and

there were only 32 cases of primary patency of the foramen ovale, of which 8 had anomalies elsewhere.

Although the tetralogy of Fallot is composed of a specific combination of cardiac anomalies, it is clinically recognized as a distinct entity and so our cases are presented here as a group. There are 6 cases of this type, varying in age from 2 months to 45 years. There are 3 males and 3 females, two have anomalies elsewhere and 3 are still living. 5 cases have cyanosis, 3 have clubbing and the red blood cell count is elevated in 2. One case has subacute bacterial endocarditis. In 85 cases reported by Abbott, the maximum age was 59 years 9 months and the mean age was 12 years. One of our cases lived to be 45 years old, thereby being the second oldest to be recorded, but the fact that she had corrected transposition of the great vessels was undoubtedly a contributing factor to her long life.

The remaining cases in our series have defects that are too few to be analyzed and so are mentioned for the sake of completeness only. All of these have other cardiac anomalies. Atresia of the pulmonary valve and artery 2, aortic insufficiency 2, aortic stenosis 2, tricuspid insufficiency 1, bicuspid aortic valve 2, rudimentary pulmonary valve cusps 1, interauricular septal defect 1, transposition of the great vessels 1, malformed tricuspid valve, 1, and lack of mitral valve and orifice 1.

In addition there are 3 cases of cor triloculare in our series, all of which are dead. Two of these had a common ventricle and two auricles. The female died when 5 days old and the male when 2 months old. Other anomalies are present in both, the female having a partial transposition of

the abdominal viscera. The case of the common auricle and two ventricles is in a male that died when 2 weeks old, and he had many anomalies elsewhere. There is also a true transposition of the great vessels in the latter case.

Abbott reported 5 cases of triloculare biventriculare, with ages varying from 17 days to 31 years, the mean age being 6 years. Two were males and 2 females and 3 had anomalies elsewhere. She also reported 13 cases of triloculare biatriatum, with ages varying from birth to 35 years, the mean age being almost 8 years. Seven were males, 3 females and only one had other anomalies.

One case of cor triloculare presented such an unusual picture that its full description is included for those who may be interested. The two great arteries side by side, the apparent aorta on the right and the apparent pulmonary artery on the left. The right auricle and tricuspid valve are normal. There is complete absence of the ventricular septum. The great artery on the right, apparently the aorta, has a normal semilunar valve, gives off the right and left pulmonary branches and the right subclavian artery and curves to the left and downward to form the thoracic and abdominal aorta. The great vessel on the left, apparently the pulmonary artery, gives off two coronary arteries and the common carotid arteries and goes on to form the left subclavian artery in its terminal branch. Between the right subclavian and the right common carotid arteries is a communicating artery about 1.5 cms. long. It opens into these two arteries just distal to where they leave their respective arterial trunks. The left auricle empties into the common ventricle by an opening about 1.3 cms. in diameter just to the

left of the tricuspid valve. The margins of this opening are smooth, rounded but not thickened and there is no trace of true valve formation. To my knowledge no similar combination of anomalies has ever been reported.

There are six cases in our series which are diagnosed just as congenital heart disease. The age varies from 1 month to 9 years. Four are females and 2 are males, 5 have anomalies elsewhere and 4 are still living. One case died from a complicating acute bacterial endocarditis.

PROGNOSIS AND COMPLICATIONS

The prognosis in congenital heart disease depends for the most part upon the type and degree of congenital defect. Some anomalies, such as ectopia cordis abdominalis, uncomplicated transposition of the great vessels and pulmonary or aortic atresia with closed ventricular septum, are incompatible with life of more than a few days or weeks at the most. Other defects, like abnormal chordae tendinae and valve cusps, simple dextrocardia, pericardial anomalies, etc. place very little, if any, burden upon the heart and consequently in these the prognosis is excellent. Using Abbott's classification it may be said that in general those defects belonging to the acyanotic and cyanotic tardive groups bear a better prognosis than those of the cyanotic group, although there are certain obvious exceptions to this rule. In the arterio-venous shunt group, the degree of the shunt is very important in determining not only the life expectancy but also the amount of activity the patient may be allowed. In the morbus caeruleus group the gravity of the prognosis depends upon the degree of anoxemia as well as on the amount of direct strain upon the heart. Anoxemia and cyanosis are correlated to a certain extent, but it must be remembered that if a polycythemia exists, there may be sufficient oxygen in the blood for the tissues and yet enough reduced hemoglobin to cause cyanosis. This explains why many cyanotic individuals are not dyspneic. Any anomaly, regardless of the group in which it is classified, that places undue strain upon one or more chambers of the heart has a serious prognosis, because hypertrophy, dilatation and eventual loss of cardiac reserve are inevitable; the physician must analyze as accurately as possible the amount of undue strain imposed before deciding upon the extent of limitation of activity and

the prognosis. Loud murmurs may mean very little if there is no associated cardiac strain.

Almost all cardiac anomalies are subject to infection - in the form of either rheumatic or bacterial endocarditis. It is this hazard that leads many physicians into making a more serious prognosis than they would otherwise do if just the defect itself were to be considered; a glance at the statistics offers some justification for such a pessimistic outlook. The margins of or adjacent to a cardiac defect seem to supply a nidus for the lodgement and accumulation of microorganisms from the circulating blood, with resultant bacterial endocarditis. Rheumatic infection shows a predilection for localized areas of strain, being fairly frequent in cases of cardiac anomalies that reach adult life. Naturally the dangers of such complications makes one hesitate to give a good prognosis, even though there may be no cardiac strain resulting from the defect itself.

Small interventricular septal defects may allow a person to lead a fairly normal, active life, but Abbott and Bland have shown that about 25 per cent of these defects have a fatal complication of secondary bacterial endocarditis. In contrast, patent foramen ovale rarely has a secondary infection, congestive failure being the usual mode of exitus. Bicuspid aortic valves are also frequently the sites of infection.

The prognosis in coarctation of the aorta depends upon the degree of collateral circulation that can be built up, and upon the degree of stenosis, an inverse proportion existing between the amount of constriction and the life expectancy. Other complications, in order of relative frequency, which add to the weight of the prognosis are: Congestive heart failure, aortic

aneurysm or rupture, cerebral complications and bacterial endocarditis.

Patent ductus arteriosus is another anomaly that is difficult to prognosticate. If the patency is large, the signs of congestive failure appear early. If the patency is small the individual may live a normal life for many years. There is a high incidence of subacute bacterial endocarditis in these cases, which consequently increases the gravity of the prognosis. However, the recent surgical treatment of this anomaly seems to have much to offer these cases and the prognosis in the future will probably be infinitely superior to that of the past. The possibilities of such will be more fully discussed under treatment.

In lesions involving the pulmonary infundibulum the incidence of secondary infection is about twenty per cent, according to Dry, and if the blood supply to the lungs is reduced, pulmonary tuberculosis is fairly common, thereby making the prognosis worse.

Frequently it is practically impossible to diagnose what kind of congenital defect exists in infants and children from physical signs alone, making an accurate prognosis difficult. However, the new science of angiocardiography is a valuable practical aid in the differential diagnosis of these cases. This consists of the injection of a radio-opaque substance into a peripheral vein immediately before roentgenography of the heart. The chambers of the heart and the great vessels and defects are thus outlined in the x-ray picture. Ten to thirty cubic centimeters of a thirty-five per cent solution of Per-Abrodil or Uroselecton B is used, and the procedure is said to be quite harmless. With the diagnosis established, the prognosis can then be better ascertained.

In short, in attempting to arrive at a conclusion in prognosticating these cases, the physician must consider not only the defect itself, but also the possible complications that may occur with their relative frequency. However, to assume that the majority of cases of congenital heart disease will acquire fatal infectious complications would be as erroneous as to ignore the possibility entirely. Optimism is preferred over pessimism.

TREATMENT

Treatment of cardiac anomalies is necessarily confined almost entirely to preventive and palliative measures. Preventive treatment may be divided into three fields. First, the prenatal care of the mother. Especially during the early weeks of pregnancy, she should avoid as much as possible any strains, infections or trauma that could affect the environment of the developing embryo. Second, the prevention from overstrain of the heart. Patients should limit their activity to whatever extent is necessary in order to avoid dyspnea and cyanosis or an increase of cyanosis. Any evidence of a failing cardiac reserve is an indication that the activity should be limited to a degree which the heart can maintain without undue strain. Third, the prevention from bacterial invasion at the site of the defect. Removal of foci of infection and the avoidance of colds and diseases involving the upper respiratory tract are of prime importance. Infected teeth or tonsils should be removed if the operative risk is not too great. Obstructed sinuses should be drained. Patients should have an adequate diet to maintain a high degree of resistance to respiratory infections. These preventive measures are especially important for those cases in which cyanosis is absent or terminal, since in a relatively large number of these cases the expectation of life is good so long as they keep within the limits of their cardiac reserve and do not develop a secondary cardiac infection.

In the cyanotic type of cases, symptomatic measures may be necessary for relief. Venesection may aid an overloaded right heart; diffusible stimulants, etc. may ease the dyspneic attacks and sedatives or hypnotics usually make the patient more comfortable. Oxygen inhalation is of little

aid in these cases.

If congestive heart failure enters into the picture, the usual therapeutic measures are indicated: bed rest, digitalis, possibly other cardiac or diuretic drugs, cardiac diet, limited fluid intake, sedatives and possibly intravenous glucose.

While there is little enough to offer these cases of cardiac anomalies in respect to therapeutics, there has been one outstanding achievement in recent years which marks the beginning of a new era of treatment. This late advancement lies in the realm of surgery, namely the surgical ligation of a patent ductus arteriosus. The possibility of ligating the ductus was first suggested by Munro in 1907, although the idea was not investigated at that time. Strieder stimulated renewed interest in this field in 1938 by making the first attempt to ligate a patent ductus. His attempt was unsuccessful because of anatomical difficulties. He did manage to reduce somewhat the size of the patency, but the patient died four days later from acute dilatation of the stomach.

To R. E. Gross goes the honor of performing the first successful patent ductus ligation on August 26, 1938. This case was a girl of $7\frac{1}{2}$ years manifesting the characteristic physical signs and evidence of incipient cardiac embarrassment. As more and more surgeons are becoming interested in the subject and are realizing the great advantages it offers to the patient, some thirty or forty more cases of successful ligation have appeared in the literature. The approach to the ductus is through the left pleural cavity anteroposteriorly above the compressed left lung. Most of the patients have little, if any, postoperative shock or complications, and the

characteristic signs of patent ductus are absent after the ligation. The mortality rate appears to be surprisingly low, although no definite figures are yet available.

With this new method of treatment the question arises: "should every case of patent ductus have a surgical ligation?" While there are differences of opinions most surgeons and cardiologists feel that surgery is not indicated if there is no undue strain on the heart and no evidence of complications. In other words, a conservative policy of let well enough alone. What, then, are the indications for surgery? Here again there is no full accord among the specialists. Some generally accepted indications are: Evidence that the child is not developing properly and no other explanation can be found, if the danger of bacterial endocarditis is high for some reason or other, indications that the patency of the ductus is enlarging and evidence that the heart is carrying an increased burden, and that congestive failure is a probability in the near future. If signs simulating aortic regurgitation are present in an uncomplicated cases of patent ductus, it means that there is a wide patency and ligation is strongly indicated. Results have proven that such patients are markedly improved following successful surgery. The diastolic pressure rises immediately after ligation, the thrill disappears and the pulmonary second sound is no longer accentuated. In respect to the murmur, there is frequently a soft systolic murmur remaining at the pulmonary area, but it has lost its "machinery" quality and there is no diastolic element. Those patients who were formerly on restricted activity, because of cardiac symptoms, were invariably able to return to more normal active lives following ligation, or at least be free of symptoms.

The big question at the present time concerns the complication of subacute bacterial endocarditis. How frequently occurs this complication in patent ductus arteriosus? At what age is it most apt to occur? Can surgery prevent it? In Abbott's series of 92 cases of patent ductus, 19 of them occurred in infants under two years of age, and as subacute bacterial endocarditis does not occur in infancy, these may be ruled out. Of the remaining 73 cases, 22 of them developed a secondary bacterial infection, an incidence of thirty per cent. Bullock, Jones and Dolley, in a study of 80 cases of patent ductus, found that 42 (53 per cent) of them died of bacterial endocarditis. Because of this high incidence some authors feel that ligation should be done as a prophylactic measure, even though the patency is small and the patient entirely asymptomatic. Hubbard, Emerson and Green analyzed 39 cases of secondarily infected patent ductus Botalli, noting that 71 per cent died before the age of 25 years, that 50 per cent died between the ages of 16 and 25 years, and that in all but one case the vegetations were located at the pulmonary orifice of the ductus or implanted on the walls of the pulmonary artery. Obviously, the patent ductus offers an excellent site for bacterial growths and removal of this site before infection occurs is a strong argument in favor of prophylactic surgical ligation, particularly as long as there is a wide difference between the apparently low surgical mortality rate and the high incidence (30-53 per cent) of bacterial invasion. However, there is at the present no proof that a ligated ductus does not offer just as favorable a site for bacterial growths as does a patent ductus. There is known one case of a ductus which obliterated normally during infancy but formed an aneurysmal sac on the pulmonary side, from which arose bacterial vegetations. The future course of those patients who have had surgical ligation will even-

tually clear up this point. The optimal age for surgery is during childhood before the second decade, when the incidence of bacterial endocarditis increases.

Another question receiving considerable discussion is whether or not a secondarily infected patent ductus should be ligated. Gross originally believed that bacterial endocarditis was a definite contraindication to surgery. The majority of investigators, however, disagree with this opinion. They feel that in view of the hopeless prognosis in infected cases, surgery is justified, hoping that the vegetations may be limited to the ductus itself and consequently be obliterated along with the ductus itself at ligation. Early diagnosis is desirable in these cases if ligation is to be attempted. Touroff has reported two cases of successful ligation when the ducts were infected, and the subsequent blood cultures remained positive in one case, but became negative and remained so to date in the other case. Bourne, Keele and Tubbs combined sulfapyridine with ligation in two cases. In one, the blood cultures showed Hemophilus influenzae before and after surgery, but were negative for the last nine months and the patient was asymptomatic for the past year and a half (up to the time of this publication). In the second case, blood cultures were positive but became negative under chemotherapy previous to surgery and have remained negative ever since. The patient has remained well for the past year. In a case recently operated on at Wisconsin General Hospital, blood cultures were positive before surgery and at operation the ductus was not identified. The aorta was ruptured with severe hemorrhage, but successfully closed. The patient died about two weeks later from the bacterial endocarditis. Autopsy will be discussed later.

An added danger in the surgery of these cases of infected ducts is that if the infection is not of recent inception, the walls of the ductus are frequently thinned, friable and adherent, and there may be an associated mycotic aneurysmal dilatation. All this greatly increases the operative risk. Touroff reported two cases of fatal hemorrhage that occurred because of the periarterial adhesions and fragility of the wall of the patent ductus. Gross, Jones, Humphries and Miangolarra have also torn the wall of the ductus, but have been able to control the subsequent hemorrhage without a fatality. I have seen one case in which the surgeon tore the wall of the aorta and a fatal hemorrhage was avoided only by the quick thinking, skill and dogged determination of the surgeon.

Much work has yet to be done on this new surgical treatment for patent ductus arteriosus. Its value as a prophylactic measure and the advisability of its use in cases where bacterial endocarditis already exists are questions which only future experimentation will answer.

CONCLUSIONS

While figures on the incidence of congenital heart disease are quite variable, depending upon what sections of the country or what age groups are involved, it is evident that these anomalies predominate in the younger age group and may easily reach an incidence of six per cent or slightly more in people under twenty years of age with heart disease. No doubt, with increasing interest in this field, better training of our younger physicians and increased laboratory facilities, the diagnosis of these defects is being more frequently made today than in the earlier years of this century, and incidence figures in the near future will be more accurate than those of the recent past.

At least a smattering of the embryology of the heart is essential before one can reasonably understand the formation of some of these cardiac anomalies. Many are explainable on an embryological basis, and in combined defects some anomalies undoubtedly arise as a compensatory change caused by another primary lesion. Fetal myocarditis and endocarditis are other important etiological factors.

Symptomatology is dependent upon the type of cardiac anomaly, but dyspnea, cough, palpitation and cerebral symptoms are the most common. White states that precordial pain is rare, but in our series it is relatively frequent.

Physical signs that are helpful in making the diagnosis, besides the cardiac findings in the specific lesions, are cyanosis, clubbing of the extremities, delayed development (either mental or physical), blood pressure and signs of congestive heart failure. Laboratory aids include the

electrocardiogram, x-ray and orthodiagram.

No attempt will be made to draw conclusions from the comparison of analysis of the various cardiac anomalies in our series with those in Abbott's series, primarily because all her cases have an anatomical diagnosis, while only 21 per cent of our cases are proven. However, there are a few interesting points worthy of reiterating. There is a marked difference in sex ratio between Abbott's series and ours. She had a predominance of males, while females were more frequent in ours. The incidence of congenital defects elsewhere in the body in our series almost doubles that in Abbptt's cases. The high degree of accuracy of diagnosis that has been attained proves that in many instances it is clinically possible to determine the type of cardiac anomaly a patient may have. It is surprising to note the few cases of subacute bacterial endocarditis that occurred in our series. In only three cases was this diagnosis definitely established and in one there is some question as to its presence. Other reports in the literature claim a much higher incidence of this infection. In regards to the controversy over the frequency of the "machinery" murmur in patent ductus arteriosus, six of our seven cases presented this typical sign and there was no report of a cardiac examination in the seventh case, a baby that died two and one-half hours after birth. This murmur is also present in most of our twelve cases that have a patent ductus combined with some other cardiac defect. But the presence of other defects may raise some question as to the validity of considering these cases of combined defects. The anomaly that is almost invariably missed by the interne, resident and staff physician on first examination is coarctation of the aorta. In only one case was the diagnosis made at the time of entrance. There are three unusual

cases in our series, the tetralogy of Fallot in a 45 year old woman, the infantile type of coarctation of the aorta in a 47 year old man, and the unusual course and branches of the great vessels in the 5 day old baby girl with cor triloculare biatriatum. The frequency of an anomalous position of the patent ductus arteriosus should be of interest to the surgeon. Almost half of our anatomically proven cases of patent ductus had an anomalous position, the aortic end of the ductus arising from the ascending arch.

While it is not always easy to make an accurate prognosis in cases of congenital heart disease, it is absolutely wrong for a physician to make just a diagnosis of "congenital cardiac disease" and assume that the patient is doomed to an inevitable early death. Each case is an individual problem and must be regarded as such. Abbott's classification is useful from the clinical standpoint. In determining the prognosis, the important things to consider are the type of anomaly that exists, the degree or size of that defect, the amount of added strain that it places on the heart, and the relative frequency or possibility of a superimposed endocarditis. Certain defects are much more susceptible to endocarditis than others, the important ones being patent ductus arteriosus, interventricular septal defects and infundibular pulmonary lesions. Prognosis in these defects consequently should be made with reservations for this possible complication.

Prophylaxis is an important measure in almost every field of medicine and congenital heart disease is no exception. It is useful in the prenatal care of the mother, important in the prevention of bacterial invasion at the site of the defect and imperative in preventing overstrain of the heart.

The rest of the treatment is, for the most part, similar to that used in acquired heart disease, with one outstanding exception.

Surgical ligation of the patent ductus arteriosus is still in the early stages of development and only careful observation of results in the future will determine what conclusion can be made. Four such operations have been attempted here. In two cases, the patent ductus has been successfully ligated, with subsequent marked improvement in the patient's cardiac status. In one case no patent ductus was found and so it was assumed that the diagnosis was inaccurate. In the fourth case, there was a complicating subacute bacterial endocarditis. During the operation the aortic wall was accidentally ruptured and was successfully repaired only after severe hemorrhage. The patent ductus was not ligated and the patient died about two weeks later from bacterial endocarditis. At autopsy vegetations were found at the pulmonary orifice of the ductus, on the wall of the pulmonary artery and a large growth on the pulmonary valve. In all probability, successful ligation would not have affected the subsequent course of the bacterial endocarditis, because of the widespread involvement.

At the present time, indications for surgery are conservative. In certain types of cases, the results appear to be excellent. Whether ligation does prevent the possibility of a future superimposed infection is a moot question. It seems logical that ligation would prevent it, but time alone will give the answer. To my knowledge, no case of successful ligation has developed this complication subsequent to surgery as yet, but the oldest case is only 3 years 8 months since ligation. Whether an operation is indicated in those cases already having a complicating infection is still

more debatable. The hazards of the operation are definitely increased and the results seem to be less promising. The next five or ten years will probably bring the answers to these questions. Nevertheless, the present uncertainty does not detract in any way from this brilliant advancement in cardiac surgery and the men responsible for it deserve whatever honors they have received. They have contributed an important milestone in the progress of American medicine.

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H H Shapiro

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