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Diagnostic Roentgenology in Congenital Heart Disease

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ROENTGENOGRAPHIC and fluoroscopic observations, always important to the patient and physician when heart disease is suspected, gain significance only by correlation with information gained by other methods. Isolation or over-emphasis of roentgenologic observations is as a sentence read out of context. The recognition of a pattern might well be called interpretation. The "coeur en sabot" heart, fondly thought of by many as diagnostic of the tetralogy of Fallot, assumes an entirely different significance in the absence of cyanosis, or in the presence of left ventricular dominance as shown by an electrocardiogram. Should we discard a medical finding because of lack of specificity when, basically, it is the recognition of its relation to the pattern that actually makes it significant?

Certain roentgenographic technics, such as angiocardiography, kymography, and retrograde aortography, have not enjoyed as broad a sphere of usefulness as the heart film or fluoroscopy. The importance of these technics lies in the very specific, though limited, information they yield. At times they constitute the key to the differentiation of one entity from the other. We should not like to argue the question of whether the already assembled or the missing pieces of a jigsaw puzzle are the more important.

THE GROWING HEART

The size, shape, and beat of the heart and its vessels are products of its basic embryologic design and of its load, past and present. Radiologic examination of these features yields information on the structure of the heart and its load. In acquired heart disease of older children or adults, the recognition and interpretation of these changes is a relatively simple process. One assumes that the patient began life with a normal heart already well developed into four chambers, that this was followed by the development of normal left ventricular dominance, and that this pre-existing familiar picture was altered by disease. Whether there be valvular stenosis, insufficiency, or altered extracardiac work load (hypertension, traumatic arteriovenous fistula, and so forth), the heart responds by adding hypertrophy or enlargement to the known pre-existing pattern. Occasionally, one may see diminution in size (Addison's disease), but in all instances of acquired disease (as opposed to congenital malformations) there is an alteration of the normal adult four-chambered heart. Selective chamber enlargement is relatively easily recognized, and criteria are well established.

The course of events in the development of the normal heart is well documented. In utero, as at birth, the heart reflects the load of the fetal circulation, resulting in right and left ventricles of comparable size with right and left myocardium of equal thickness. Most normal hearts at birth will measure and weigh approximately the same; in fact, the majority of congenitally defective ones will also show little variation in size in the first few weeks of life. The commonest congenital malformations will normally sustain fetal circulation. If the developmental malformation of the heart grossly
fails to sustain fetal circulation, intrauterine death intervenes. As a result, most infants begin life with a heart of approximately the same external dimensions and configuration. Only those having primary myocardial disease, aortic atresia, and congenital arteriovenous aneurysms are the exceptions. Thus, roentgenograms will fail to distinguish the normal heart from the majority of hearts having congenital lesions at, or shortly after, birth.

When the lungs become aerated shortly after birth, after the foramen ovale ceases to function, and after the ductus closes, the pulmonary and systemic circulations are separated. The left ventricle gradually hypertrophies, paralleling the increased pressure of the systemic circulation. The left ventricle thus becomes heavier and larger, and thereby the dominant chamber of the heart. Normally, this trend probably persists throughout life, the left ventricle continuing to hypertrophy relatively more than the right until myocardial degenerative changes set in due to impaired circulation, fibrosis, or other factors, or until the load balance is altered by acquired valvular or pulmonary disease. Hypertension in the young adult simply aggravates this trend. It is evident that the size and configuration of the heart, despite a probable plateau in the middle span of life when it is relatively stable, is not static.

Cardiac Configuration

The shape of the heart is determined not only by its position, component chambers, and content, but by its own elastic resistance or muscle tone.

Criteria for enlargement of a chamber in acquired heart disease are readily available in radiologic texts. This is not true in congenital cardiac disease. In congenital malformations of the heart, development may be so abnormal as to bear no resemblance to the normal adult four-chambered organ. At times, one may be dealing with only one ventricle. Examples of this are a congenitally single ventricle with truncus communis, and tricuspid atresia or severe tetralogy of Fallot, where the substance of the heart is predominantly one ventricle. In none of these is the enlarged ventricle superimposed upon a normal heart or even a normal opposite ventricle. In the tricuspid atresias, the right ventricle is congenitally rudimentary or is completely absent, while in the extreme types of tetralogy of Fallot, the left ventricle may remain essentially infantile in development as a result of the diminished load. In both of these examples the substance of the heart may be so overwhelmingly one ventricle, without a fully developed opposite ventricle, that the ventricular configuration loses its roentgenographic distinguishing characteristics. Thus, the roentgenologic appearance of the heart with these lesions approaches a common form. In this case, there are no universally applicable roentgenologic criteria of left and right ventricular enlargement.

In clinical medicine, one sees a patient more often who has a congenital heart defect which altered the extraterine load on the heart and, thereby, its normal development. Here again, the usually accepted roentgenographic criteria of selective chamber enlargement are not applicable. In contrast to acquired heart disease, where chamber enlargement alters a pre-existing pattern, the heart fails to develop the adult chamber relationship.

The commonest stumbling block in evaluation of the heart shape in congenital heart disease is the recognition of right-sided enlargement. What part the right ventricle normally plays in forming the right border of the heart in the posteroanterior silhouette on
Fig. 1. Right ventricular hypertrophy with only slight chamber enlargement as seen in valvular pulmonic stenosis. Here the left ventricle is of normal size and is not displaced by the right ventricle, which is hypertrophied but only slightly enlarged by external dimension. a. Posteroanterior projection. Note normal transverse diameter with a configuration not specifically abnormal. b. Right anterior oblique projection. There is a slight increase in anterior-posterior dimension. c. Left anterior oblique projection. The slight right ventricular enlargement is recognized by the slight increased convexity of the anterior border and slight upward displacement of the left ventricle, resulting in a poorly defined apex and an almost round shape. d. Left anterior oblique projection with contrast media in right side of heart showing how the right ventricle lies anteriorly and in contact with the diaphragm. Enlargement of this chamber will cause counter-clockwise rotation of the apex in this view.

The roentgenogram has been a controversial subject since the earliest writings on cardiac roentgenology. The effect of selective hypertrophy and enlargement of the right ventricular chamber is now well understood but not widely recognized. Ordinarily, the right ventricle does not add appreciably to the transverse diameter of the heart when it is only slightly
or moderately enlarged. However, it may alter the contour of the heart in the posteroanterior view by elevating the left ventricle in a counterclockwise direction, thus raising the apex of the heart and giving it a rounded appearance. As the increase in size of the right ventricle is normally toward front and back, it will be demonstrated optimally in the oblique projections where the increase in horizontal dimensions becomes obvious in the right anterior oblique, and its alteration of the configuration is most striking in the left anterior oblique, in that the apex is elevated and poorly defined, and the heart appears almost round (fig. 1).

These changes take place in the presence of a normal left ventricle. If the hypertrophy of the right ventricle is associated with moderate enlargement but is unbalanced by a normal left ventricle, the classic "coeur en sabot" shape results. Here, the left ventricle remains infantile or hypoplastic in development as the right is sharing a large part of the work necessary to sustain the systemic pressure. The small left ventricle is cocked up high as the right ventricle becomes the increasingly dominant chamber (fig. 2).

If the right ventricular enlargement is balanced by a well-developed or enlarged left ventricle, the effect on the cardiac configuration is of the type more familiar in acquired heart disease, as seen in mitral insufficiency with stenosis. The left ventricle is not so readily displaced by the addition of an enlarged right. In this case, right ventricular enlargement is almost immediately reflected by increase in the maximum transverse diameter of the heart, including the right border, with some increased convexity on this side. The forward bulge of the anterior contour of the heart in both obliques, so commonly described in the literature, is recognized under these conditions only when the well-developed or thick left ventricle is no longer readily displaced posteriorly and upward. This type of right ventricular enlargement is seen in the high ventricular septal defect where pulmonary hypertension gradually may add right ventricular enlargement to an already heavy, hard working left ventricle, resulting in a balanced enlargement of the heart (fig. 3). Similar examples, for which there is no place here, may be given for each of the cardiac chambers. Thumb rules of chamber enlargement of acquired heart disease are not generally applicable in many developmental lesions, and, again, the part is evaluated in light of the whole.

Localization of the interventricular notch or measurements of the left anterior oblique dimensions of the heart, based on an estimate of the position of the septum, as recently

![Fig. 2. Right ventricular enlargement as seen in a moderately severe tetralogy in which the right ventricle is the dominant chamber of the heart. a. Posteroanterior projection showing normal transverse diameter but a distinct elevation of the apex of the heart with an almost round right and lower border. b. Left anterior oblique projection with contrast material outlining the right side of the heart. c. Same projection with contrast material outlining the left ventricle. This heart illustrates how the enlarged right ventricle displaces a relatively small nonhypertrophied left ventricle upward in a counter-clockwise rotation. This results in the "coeur en sabot" configuration in the posteroanterior projection and a round heart in the left anterior oblique projection.](http://circ.ahajournals.org/content/1/1/465)
advocated,2 have failed to increase accuracy in evaluating selective ventricular enlargements in the hands of the authors.

Hypertrophy Versus Enlargement. Cardiac chamber enlargement means disease. Disease may, of course, exist without enlargement. The heart does not necessarily respond to an abnormal load by enlargement in its external dimensions. The increased load may be met by hypertrophy of muscle fibers which in time may produce a thickened myocardium. This commonly takes place without increase in the external dimensions of the chamber. In fact, the internal dimensions of the chamber may be slightly diminished in the presence of increased “elastic resistance” of the wall and increased output of the heart. The fact that there may be no change in heart size in the presence of long-standing increased load met by cardiac hypertrophy has recently been reemphasized by Kleinfield and Redish.3 In their study, 18 of 45 patients with sustained or progressive essential hypertension showed no change in heart size by teleoentgenogram for periods of 5 to 20 years! That this is also a common experience in aortic stenosis and coarctation of the aorta has not been as readily recognized. In pulmonic stenosis of mild to moderate degree, absence of significant right ventricular enlargement appears to be common (fig. 1). In general, if a ventricle is working against increased resistance or pressure, it would appear that hypertrophy, with little change in size, is common. Increase in “flow work,” that is, an increase in the volume of blood handled by the chamber per unit of time, such as is seen in extracardiac shunts or septal defects, appears to be more commonly associated with chamber enlargement (probably also with hypertrophy) early in life. These observations do not apply as well to the atriums which appear to be more distensible chambers.

Cardiac Beat

Observation of the amplitude of excursion of the cardiac border, main pulmonary artery, and aortic arch during fluoroscopy yields information on the state of the myocardium. A poor excursion of the cardiac border indicates either pericardial fluid or myocardial weakness, or both, and often preclinical failure may be recognized in this manner. Increased amplitude of pulsation of the pulmonary artery and aorta together indicates an extracardiac shunt, although a high septal defect with aortic insufficiency may give the same.

Interestingly enough, hypertrophy of the ventricle, as seen in hypertension, may be associated with a diminished amplitude of pulsation. However, this should cause no confusion, as in these instances there is relatively little chamber enlargement.

Actual thinning of the wall of a vessel may
be reflected by an increase in amplitude of pulsation even in the presence of low pressure and diminished flow. This is true in localized poststenotic dilatation of the pulmonary artery. Though the cause remains obscure, the dilated segment to which this increased amplitude of pulsation is limited is observed to be thinwalled at operation.

PULMONARY VASCULATURE

The authors are among those who have long emphasized the value of the pulmonary vasculature, as seen on roentgenograms, as an aid in the diagnosis of congenital heart disease. Excellent research on the relation of the pulmonary to the systemic circulations in congenital heart disease⁰ and the correlation of this with roentgenoscopic observations⁰ has added to and clarified what was previously clinical empiricism.

A film on full inspiration will give the best definition of the size and course of the pulmonary vascular tree. Fluoroscopy will supplement this information by revealing the presence or degree of pulsation of the vessels. Judging vasculature simply by the fluoroscopic screen “brightness” is a hazardous substitute for detailed examination of individual vessels on the film.

The normal vascular pattern observed on the roentgenogram is almost entirely the pulmonary arterial tree. The pulmonary veins, although adding density to the chest, normally are not recognized as a distinct pattern, probably because they are only approximately half the size of the pulmonary arteries for any given distance from the hilum. In addition, the pulmonary veins do not emerge from single trunks in the hilums, but emanate from the usually four pulmonary veins which lie behind the heart in the posteroanterior projection.

Pulmonary Vascular Engorgement. An increase in size, or more specifically, in caliber, of the normal arteries of the lung represents an increase in content of these vessels. This is vascular engorgement, pulmonary hyperemia, or the pleonemic lungs of Campbell.⁵ This change is not to be confused with those loosely termed as “prominent markings,” a nondescript term commonly used to denote increased radial density of the lung without specification as to whether it be intravascular or extravascular in origin. The increased caliber of vascular engorgement of the arteries is best recognized in the middle third of their course as they traverse the lung field, and not in the hilum. The increased volume of blood in the pulmonary arterial tree may be the result of increased amount of blood flowing through these vessels, an active vascular engorgement; or it may be relatively static; a passive vascular engorgement.

Active or Passive Pulmonary Vascular Engorgement: The differentiation of active from passive engorgement cannot be made by examining individual vessels on the films. Supplementary information is necessary, which, however, often can be found on the films. Intrapulmonary congestion (extravascular fluid evidenced by an ill-defined increase in density of the bronchial or interstitial supporting tissues) suggests passive engorgement. Active expansile pulsation of the vessels at fluoroscopy is almost incontrovertible evidence that the engorgement is active and secondary to left-to-right shunt.

The recognition of active vascular engorgement is, of course, the sine qua non for radiologic evidence of left-to-right shunt. Without it, one may be at a loss as to whether the chamber enlargement is based on a stenotic lesion or an arteriopulmonary shunt. This is often the radiologist’s dilemma in trying to recognize small shunts. In general, if chamber enlargement is unequivocally recognized, it may not be attributable to a left-to-right shunt unless active vascular engorgement is demonstrable, such is the parallelism in these lesions. Prominence of the main pulmonary artery is an exceedingly poor index of increased pulmonary blood flow, and it should not be attributed to this unless the increased flow is reflected by active pulmonary vascular engorgement in the lung.

Diminished Pulmonary Vasculature. Diminution in the caliber of the intrapulmonary vessels, as recognized on the roentgenogram, is indicative of diminished content in the vessels and is practically pathognomonic of diminished pulmonary blood flow. This is seen most
commonly in pulmonic stenosis of moderate or marked degree, with or without associated lesions of overriding aorta, atrial septal defect, and so forth. Diminished blood flow through the lung also occurs in tricuspid atresia or stenosis, Ebstein's disease, and anomalous insertion of a vena cava into the left atrium. By catheterization, we are recognizing an increasing number of cases of primary pulmonary hypertension of undetermined cause in which the obstructive phenomena appear to take place in the periphery of the pulmonary arterial tree (perhaps the congenitally high "elasticity resistance" of Deuchar and Knebel). This group of patients also appears to have roentgenographic evidence of diminished pulmonary blood flow, at times making the x-ray differentiation between this condition and pulmonic stenosis exceedingly difficult.

**Expansile Pulsation of Pulmonary Vessels.** This is an actual increase in the dimensions or transverse diameter of the vascular shadow observed during systole at fluoroscopy. It is to be distinguished from the mere rhythmic movement of the vessel, secondary to respiratory activity, or transmission of pulsation by proximity to the heart or great vessels.

Normally, expansile pulsation may be seen in the main pulmonary artery and its primary divisions. Slight expansile pulsation of the secondary and, rarely, tertiary branches in the hilar shadow or lung root may be observed in hyperthyroidism, a hyperactive heart of excitement, or anemia. Technically, this is observed best while the patient is holding his breath, with the screen coned down to a square four to 5 cm. over the right hilum. This eliminates the transmitted pulsation due to the proximity of the main pulmonary artery so commonly seen in the left hilum. Individual vascular shadows should be fixed in the field of vision, preferably a vessel on end. If expansile pulsation is picked up in the right hilum, it should be followed out peripherally, and if it can be observed distal to the secondary and tertiary divisions of the hilum as one approaches the mid-lung fields, it can usually be considered abnormal. Almost invariably it means increased pulmonary blood flow. The expansile pulsation is most commonly observed in atrial septal defects, high or large ventricular septal defects, and truncus communis. It is less commonly observed in patent ductus arteriosus, and is characteristically lost in the older age group if and when pulmonary hypertension intervenes. The mechanism involved in the production of increased amplitude of pulsation of the main pulmonary artery and expansile pulsation of the pulmonary vascular tree has recently been investigated.

Evidence suggests that increased pulmonary flow is the dominant factor in production of the vascular prominence, and that it is also a factor in expansile pulsation. The stroke output of the right ventricle is probably more important in the latter. The increase in the pulmonary vascular expansile pulsation of the atrial septal defect and ventricular septal defect over the patent ductus arteriosus is explained by the fact that, in the former, cardiac output is forced into the arteries suddenly during systole, instead of more gradually during systole and diastole, as in patent ductus arteriosus. Pulmonary pressure probably plays a less significant role than flow in producing these pulmonary vascular changes.

"Hilar Dance" and "Hilar Sling". Campbell pointed out that the "dance of the hilum" as originally described in 1925 was presented as being pathognomonic of pulmonary regurgitation, although review of the case notes today suggests that the patient had an atrial septal defect. Up to this point, we have avoided using this highly descriptive term because of the variable interpretations placed upon it throughout the literature. Many use it synonymously with any hyperactive beat or pulsatile main or primary division of the pulmonary arteries. When used in this sense, it does not necessarily imply the presence of increased pulmonary blood flow or shunt. When used subsequently, it will mean an intrinsic vascular expansile pulsation observed lateral to the right hilar shadow.

**Collateral Pulmonary Circulation.** In many patients with deficient pulmonary blood flow, a collateral circulation develops through the bronchial arteries which is recognizable on roentgenograms of the chest. The diagnostic clue lies in the alteration of the vascular pat-
tern. The bronchial arteries pursue a considerably more tortuous course, lacking the linear radial distribution of the pulmonary arteries, and they fail to emanate from a central point in the hila (fig. 4 and 5). This appearance has recently been well described.6

It takes at least two to three years for this picture to develop, but it is common in patients having tetralogy of Fallot and in some having tricuspid atresia, particularly if they are doing well. It has never been observed in uncomplicated pulmonic stenosis.

Classification of Congenital Heart Anomalies

A practical clinical working classification of congenital heart anomalies cannot rest on a single yardstick or diagnostic instrument. A straight physiologic classification suffers the disadvantage of grouping diverse anatomic lesions under a single physiologic group. A completely anatomic classification lends itself only to adequate application at the postmortem table. A roentgenologic classification suffers the severe handicap of using predominantly anatomic criteria in a technic which falls far short in its revelations of gross anatomy. The radiologist is likely to overemphasize, in his necessarily simple classification, certain roentgenographically conspicuous anatomic features, particularly prominence of the pulmonary artery, just as the pathologist for years trivialized the physiologically important pulmonic stenoses and hypertension, simply because the structures appeared grossly normal, and he failed to appreciate the physiologic implications. Any classification, if it is to be helpful, must contain clinical, physiologic, and anatomic data. It is a relatively simple matter to classify the majority of cardiac abnormalities into similar physiologic groups. This can be done in approximately 85 per cent of the cardiac malformations in patients more than 2 years of age, using routine diagnostic methods. In the remaining patients it may be necessary to call upon the more complicated techniques of angiocardiography and cardiovascular catheterization. A recent "correlation of the physiologic and clinical findings of the more common congenital malformations of the heart" has been published by Bing and his associates.7 A major division of the cardiac malformations usually is made on a clinically recognizable physiologic feature: The presence, absence, or late appearance of cyanosis (table 1).

Malformations of the acyanotic type are subdivided into those presenting evidence of
congenital stenosis as opposed to those presenting evidence of a defect between the pulmonary and systemic circulation permitting an abnormal shunting of blood from left to right.

If cardiac cyanosis is present, one may assume a flow defect between the pulmonary and systemic circulations in the opposite direction; namely, a right-to-left shunt.

If cyanosis is inconstantly or has appeared later in life (usually fourth to sixth year) it is reasonable to assume, in the case of a congenital cardiac malformation, that a left-to-right shunt has reversed itself, with blood now flowing in the opposite direction, from right to left. This reversal is usually associated with pulmonary hypertension (table 2).

Acyanotic Group

Coarctation of the Aorta. Usually, the diagnosis of coarctation of the aorta is readily established by clinical and roentgenologic examination. In the barium-filled esophagus, the site of the coarctation is often indicated by an indentation in the esophagus on the left, just below the level of the aortic arch. This indentation usually indicates a poststenotic dilatation of the aorta; multiple indentations

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Left-to-right shunts, whether intracardiac or extracardiac may be theoretically reversed if the pressure in the pulmonary arterial system reaches or exceeds that of the systemic system. With this shift, the load on the heart is altered, the configuration reflects the altered load by x-ray, and cyanosis is usually recognized clinically. The table illustrates the lesions in which this mechanism of cyanosis has been observed; the thickness of the arrows indicates the frequency with which this shift from the one group to the other occurs. It becomes obvious that in this shift, the heart in each instance assumes a more balanced enlargement and loses its more distinctive selective features. Once the hearts have shifted to the right, they assume a uniform appearance and become essentially indistinguishable from one another. (See text under Eisenmenger's Complex.)

or ring shadows of calcium in this area usually indicate intercostal aneurysms. Both of these significant complications of coarctation may now be dealt with surgically in the majority of cases by replacement of the coarcted segment by grafts.8, 9, 10

Abnormal pulsation of the left ventricle in the presence of coarctation usually suggests aortic insufficiency which is commonly the result of bicuspid valves.

Shallow bilateral notching of the lower margins of the ribs was formerly considered a pathognomonic sign of collateral circulation around an intrathoracic coarctation of the
aorta. Neurofibromatosis with multiple nodular lesions of the intercostal nerves, collateral circulation from bronchial vessels in tetralogy of Fallot, and, most recently, vascular notching produced by dilated tortuous intercostal veins resulting from collateral circulation of a long-standing obstruction of the superior vena cava, have been recognized as producing a similar appearance. The actual visualization of the tortuous internal mammary arteries projecting from below the sternum into the lung fields on the plain lateral or oblique films is helpful evidence of collateral circulation.

Coarctation as a cause of infantile cardiac failure has been increasingly recognized at this Clinic. Death during infancy, as a result of cardiac failure secondary to coarctation, is not an unfamiliar picture to the pathologist. Unfortunately, the clinician and radiologist have not been so aware of this occurrence. They usually see the child after myocardial failure has set in, and the clinical picture is one of cardiac decompensation and myocardial strain. Perhaps too often the findings are attributed to primary myocardial disease, as careful comparative blood pressure studies between the upper and lower extremities are not commonly done in this age group. Radiologic examination is of little help in this problem except that the examiner should constantly be suspicious of a possible coarctation when seeing an infant in cardiac failure without evidence of a large left-to-right shunt, cyanosis, or major malformation at the base of the heart.

The stark pessimism felt by clinicians in the past regarding the infant in cardiac failure secondary to coarctation of the aorta is probably not justified in light of recent experiences in which 10 or 12 of these infants in failure have been carried through the period of adjustment to the coarctation during the rapid infantile growth by strictly medical measures. Thus, they have been carried on to the age at which surgical correction may be carried out.

Aortic and Subaortic Stenosis. Congenital aortic and subaortic stenosis is not a roentgenologic diagnosis, and is more often missed than recognized by x-ray examination. Characteristic is isolated left ventricular hypertrophy identical to that seen in hypertension, aortic insufficiency, or coarctation of the aorta. In congenital aortic stenosis this is often recognized only late in life when hypertrophy is accompanied by considerable enlargement. Campbell recently emphasized that the delayed appearance of symptoms and signs accounts for many cases not being recognized until middle life. The congenital nature of the lesion is thus perhaps not suspected as often as it should be. Acquired myocardial fibrosis and valvular calcification add to the difficulty in the differential diagnosis of this lesion from acquired aortic valvular disease of rheumatic origin in older individuals. A roentgenologic clue to the diagnosis of these conditions may be found in the great vessels. The aorta is commonly dilated; apparently this is poststenotic dilatation with widening of the arch and increased caliber of the ascending aorta. In a rarer form, the aorta presents diffuse hypoplasia in that it is unusually small in caliber throughout.

Pulmonic Stenosis. This condition is characterized by congenital narrowing of the tract through which the blood flows from the right ventricle to the lungs. The resulting degree of obstruction is variable. It may be exceedingly mild or severe. The roentgenologic and clinical appearance parallels this same scale of severity, yet the correlation is not 100 per cent. The stenosis may be of such mild degree as to produce no recognizable structural changes. If changes exist, the one constant finding is right ventricular enlargement. If the intrapulmonary blood flow is considerably reduced as a result of the stenosis, the pulmonary markings throughout the lungs will be diminished in caliber on the x-ray films. In most moderate and severe pulmonic stenoses the right atrium is also slightly enlarged. In patients with pulmonic stenosis of sufficient severity to give rise to symptoms, the pulmonarv vasculature is always diminished, and in about half of those without symptoms the vasculature can also be recognized as diminished.

Recently, much emphasis has been placed on the actual location of the narrowing in the right ventricular outflow tract. The narrowing
may be in the infundibulum (as a generalized narrowed tract or small annulus), in the valve ring, in the pulmonary valve leaflets, in the pulmonary artery itself, or in a combination of these.

Valvular Stenosis With Poststenotic Dilatation: In uncomplicated ("pure") pulmonic valvular stenosis there is usually an associated poststenotic dilatation of the main pulmonary artery which may extend into the left and right branches for a short distance, but is poorly visualized on the right because of its deep retroaortic and prespinal location. This poststenotic dilatation is recognizable roentgenologically as a prominent convexity of the middle cardiac segment, along the left border of the cardiac silhouette below the aortic knob and above the left ventricle.

Fluoroscopically, one may observe a frank expansile or exaggerated systolic pulsation of this prominent pulmonary artery segment; however, the expansile pulsation is limited only to the main pulmonary artery segment. There is never an expansile pulmonary pulsation involving the secondary, tertiary, or smaller vessels. This prominent pulsation of the pulmonary artery probably represents the "pulmonary artery aneurysms" of the early radiologic literature. Recently, two patients with gross "idiopathic dilatation" of the pulmonary artery have been described who had normal pulmonary arterial and right ventricular pressures, but had pulmonic incompetence.\(^{14}\)

Valvular Stenosis Without Poststenotic Dilatation: That uncomplicated valvular stenosis can exist without poststenotic dilatation is now well established.\(^{14, 15}\) Patients with this condition present a normal-appearing pulmonary artery segment.

Infundibular Stenosis Without Poststenotic Dilatation: Uncomplicated ("pure") infundibular stenosis with normal valves is rare. Isolated examples have been reported.\(^{14, 16, 17, 18}\)

In most of these, the pulmonary artery may be recognized, but it is usually small. In the narrowed or annular type of infundibular stenosis, a poststenotic dilatation of the distal infundibulum may at times be seen, representing almost a separate infundibular chamber. This is one of the rare examples where one recognizes enlargement of the pulmonary conus. It presents itself roentgenoscopically as a prominence or bulge of the cardiac contour just below the origin of the pulmonary artery and is best seen in a mild right anterior oblique projection. It lies considerably anterior to the prominence, often seen at the same level, produced by the left atrium.

Angiocardiography in Pulmonic Stenosis: Angiocardiography adds little of value in differentiating infundibular and valvular pulmonic stenosis, except when carried out as in Sweden by direct intracardiac injection and rapid serialographic roentgenography (10 to 12 films per second).\(^{19, 20, 21}\)

**Septal Defects.** Interauricular Septal Defect: Exceedingly small defects, although they may give rise to significant murmurs, may produce no change in the appearance of the heart. If the shunt is of sufficient magnitude to produce roentgenographic changes, right ventricular enlargement will always be recognized. Larger shunts will sooner or later be reflected by increased prominence of the pulmonary vascular markings, and there may be prominence of the atria, especially the right. There is usually an increased amplitude of excursion of both the right and left ventricular walls and commonly an expansile pulsation of the intrapulmonary vessels. If roentgenographic changes are present, the electrocardiogram should show incomplete right bundle branch block.

Actual demonstration of the atrial defect by angiocardiography, using an extremely rapid injection technic and simultaneously recording an electrocardiogram with roentgenologic exposures of 10 to 12 films per second, has recently been reported.\(^{19}\) These studies suggest that the foramen ovale normally closes between birth and the sixth day of life, that its delayed closure may play a role in "asphyxia neonatorum," and that the congenital auricular defect varies in size during the cardiac cycle, being smallest at the height of auricular systole.

Lutembacher’s syndrome, the coexistence of an atrial septal defect and mitral stenosis, in the few confirmed cases, does not differ roentgenographically from a large atrial septal defect, as the effects of each lesion are similar.
and additive. Actually, it has been shown recently that this combination of lesions is exceedingly rare, and the association of a "mitral-like" diastolic murmur with congenital lesions is common in the absence of mitral disease. In our experience, many of the cases diagnosed clinically as Lutembacher's syndrome turn out, on further investigation, to be Eisenmenger's complex.

Interventricular Septal Defect (Maladie de Roger): This section includes only the small or low interventricular defects traditionally known as maladie de Roger. Physiologically, they behave quite differently from the larger or high defects to be discussed later. These defects are commonly responsible for a loud murmur with no recognizable roentgenologic changes. Although this defect throws an abnormal load on both ventricles, the roentgenologic evidence of the lesion is first recognized as right ventricular enlargement. Both ventricles usually hypertrophy. However, the right ventricle is probably more distensible, and enlarges first. The left ventricle hypertrophies to a considerable degree before there is roentgenologic evidence of enlargement. As the hypertrophy of both ventricles may be such as not to disturb the relationship of the left to the right, the dominance of the left over the right may be similar to the normal on the electrocardiogram. Thus, in patients with this lesion, it is not uncommon to see prominence of the right ventricle on the roentgenogram and dominance of the left ventricle on the electrocardiogram. The lungs may reflect the increased blood flow by engorgement of the vessels, and "hilar dance" may be present, but is not common. If pulmonary vascular engorgement is present, the left atrium will reflect this increased blood flow by slight prominence.

Persistent Patent Ductus Arteriosus. Here again, as in the interventricular septal defects, the small shunts usually behave physiologically differently from the large shunts. The classic radiologic picture with five roentgenologic criteria has been well described as: left ventricular enlargement, prominence of the main pulmonary artery, left atrial prominence, pulmonary vascular engorgement (often with expansile pulsations) and a "hyperactive beat" along the entire left border of the heart with the pulmonary artery and aorta sharing in the hyperactivity. Any one or a combination of these changes may be present, but the experienced examiner will recognize in this pattern a parallelism of the size of the left ventricle, the degree of pulmonary vascular engorgement, and the prominence of the left atrium.

The characteristic machinery murmur with an essentially normal heart and the hemodynamic pattern just described was formerly accepted as the sine qua non of the diagnosis of persistent patency of the ductus arteriosus. An increasing number of patients in recent years have been found to have a patent ductus either without a machinery murmur, or presenting no murmur or one limited to systole. The risk of error in diagnosis in these cases has recently been emphasized. In our experience, patients lacking the classic murmur or the previously described x-ray appearance fall into two distinct categories: (1) patent ductus arteriosus with myocardial failure, and (2) patent ductus arteriosus with pulmonary hypertension. The patent ductus arteriosus in failure looks like any other heart in failure. There is generalized enlargement and pulmonary engorgement of a passive type, often associated with congestion. It is obvious that, as a result, it loses its radiologic distinctive features. In the exceedingly large patent ductus, with or without pulmonary hypertension, failure is not a rarity in infants, and we can add the patent ductus to the previously discussed coarctation and primary myocardial disease, as a diagnosis which must be entertained when an infant with acyanotic heart disease is presented in failure with a large heart, pulmonary engorgement, and congestion.

Patent Ductus Arteriosus With Pulmonary Hypertension: If pulmonary hypertension is associated with patent ductus arteriosus, the additional load on the right side of the heart will be reflected by right ventricular hypertrophy and often enlargement. If this is superimposed on the previously described characteristic changes of a patent ductus, the resulting cardiac configuration is one of a
balanced load with both right and left ventricular enlargement, a prominent pulmonary artery, pulmonary vascular engorgement, left atrial prominence, and often a hilar dance.

These variations in appearance of the ductus are often referred to as the "atypical patent ductus." Actually, we believe that the situation would be much clarified if these were thought of strictly in terms of complications of the ductus. Instead of referring to them as "atypical," they could be thought of as patent ductus complicated by cardiac failure or patent ductus complicated by or associated with pulmonary hypertension. These complications are expressed by an additional character in the configuration of the heart.

Thus, it becomes obvious that the patent ductus with either of these complications loses its distinctive features and is no longer readily recognizable by everyday diagnostic techniques. It is here that retrograde aortography becomes a definitive procedure. 30, 31 In fact, this procedure is definitely indicated for any acyanotic patient with nondescriptive murmurs in whom a patent ductus cannot be excluded with certainty, clinically, and who shows active pulmonary vascular engorgement for the more or less balanced right and left ventricular enlargement and a wide systemic pulse pressure. Only by this means can this important surgically correctible lesion be finally diagnosed (fig. 6). In the presence of patent ductus arteriosus, the aortogram shows prompt opacification of the pulmonary artery, the injected dye passing directly from the aorta to the pulmonary artery, as a result of a left-to-right shunt through the ductus (fig. 6).

Aortic Pulmonic Defect or Aortic Pulmonic Fenestration. This congenital malformation is characterized by fenestration between the ascending aorta and the main pulmonary artery, anatomically similar and only slightly removed from the location of the patent ductus arteriosus. In consequence, it places an identical hemodynamic load on the heart, resulting in a roentgenographic and roentgenoscopic

FIG. 6. Retrograde aortogram in the left anterior oblique position, demonstrating the contrast material injected in the left carotid artery passing down the aorta and simultaneously through a large ductus (arrow) into the pulmonary artery. The retrograde aortogram has specific application in the identification of the atypical patent ductus (as illustrated in this patient with no cardiac murmur) and in the differential diagnosis of the high, large ventricular septal defect, patent ductus arteriosus, and the aortic pulmonic defect (fig. 7).
appearance which may be identical with that of the patent ductus arteriosus. If the ductus-like murmur is slightly lower and to the right of its usual position, this diagnosis must be entertained.30, 32, 33

High or Large Interventricular Septal Defect. This intracardiac congenital defect resulting in a left-to-right shunt was not taken up with the "maladie de Roger" with which it is anatomically akin, but is discussed together with the patent ductus and the aortic pulmonary defect because it gives an identical hemodynamic disturbance and places the same type of load on the heart as the patent ductus and the aortic pulmonic defect.

The explanation is somewhat complex, but it may be simplified by pointing out that the intracardiac dynamics are such that the blood is shunted directly through the defect during systole from the outflow track of the left ventricle into the conus of the infundibulum of the right ventricle, and on to the main pulmonary artery. Thus, its physiology is similar to that of aortic fenestration in which the blood is directed in a left-to-right shunt from the base of the ascending aorta into the base of the pulmonary artery, or even to the patent ductus where the blood is shunted from the descending aorta directly into the pulmonary artery.34 The left half of figure 7 illustrates schematically the slight anatomic differences of these three lesions and their physiologic similarity. Here again is a definite indication for the retrograde aortogram. The differential diagnosis in these anatomically slightly dissimilar and physiologically similar lesions can often be made only by retrograde aortic injection (fig. 6).

The Shift from Acyanotic to Cyanotic Group

Pulmonary Hypertension. Pulmonary arterial hypertension as the result of increased pulmonary resistance may occur alone without cardiac lesions, or be associated with, or secondary to, congenital heart disease.35

Primary pulmonary hypertension is not necessarily associated with a cardiac murmur, but it is reflected in the heart by right ventricular enlargement, prominence of the main pulmonary artery, and, eventually, diminution in the caliber of the peripheral pulmonary vessels. The appearance is similar to that of cor pulmonale in the adult.

The clinical aspects and the role of pulmonary hypertension in congenital heart disease have been well evaluated recently by Nadas.36 Without entering the controversy of the mechanism involved in the pulmonary bed, resulting in increased pulmonary resistance and pulmonary hypertension, it can be said that there is evidence that prolonged pulmonary hypertension causes secondary changes in the vascular bed, which in turn may cause a progressive rise in pulmonary arterial pressure as well as an irreversible resistance.29, 36, 37 On the other hand, it is believed that under certain conditions the pulmonary vascular bed may retain its fetal characteristics.38, 39 Be that as it may, it can be said empirically that equalization of pressures between the systemic and pulmonary circulation is found in certain types of malformations and apparently under certain circumstances. In our experience, pulmonary hypertension of sufficient degree to cause a significant change in the cardiac silhouette is found in two general circumstances: in large interventricular septal defects or in large extracardiac shunts (patent ductus arteriosus, aortic-pulmonic defect). In the latter equalization of pressures between the pulmonary and systemic circulations takes place promptly in infancy. It could be speculated that this is possibly a reflex action of the complex neurocirculatory system which is made to sustain the systemic pressure and which is necessary to sustain life. It should be obvious that if the size of the defect shunting blood in the septum or aorta into the pulmonary artery approaches the caliber of the aorta itself in size, pulmonary pressure must rise to, or approach, systemic pressure in order to get enough blood through the systemic circulation to sustain life. If this did not take place, the child would, in effect, bleed to death through the hole in the side of the aorta into the pulmonary capillary bed (figs. 6 and 7).

Clinical empiricism also dictates that in certain congenital malformations of the heart, equalization of the pressures between the
systemic and pulmonic circulation takes place, or progresses slowly, considerably after infancy. These have been customarily thought of as an acquired secondary type of progressive pulmonary hypertension. It is in this group that the pathologic mechanism is still controversial. Be that as it may, clinical observation of the patients and the heart, recently documented by catheterization studies, indicate a gradual progressive increase in pulmonary pressures with resulting reversal of the shunt and onset of cyanosis. This picture occurs classically in the high or large ventricular septal defects, but the same mechanism and the same sequence of events may take place in other intracardiac and extracardiac left-to-right shunts (table 2). Thus, we make the transition from the cyanotic to the cyanotic group.

*Interventricular Septal Defect With Pulmonary Hypertension (Eisenmenger's Complex).* Anatomically, this malformation is characterized by high interventricular septal defect or large interventricular septal defect with a functionally overriding aorta and a potential left-to-right or right-to-left shunt, depending on the pulmonary vascular resistance. As a result, if the defect is huge, vascular pulmonary hypertension must take place promptly in order to sustain life, and the result is the infantile picture just described. If the defect is relatively small, there may be a left-to-right shunt in early life with increased blood flow through the pulmonary circulation. The resulting picture is identical to that previously described for patent ductus arteriosus. Later in life, if pulmonary hypertension supervenes or progresses, as often occurs between the ages of 6 to 12 years, there may be a reversal of the shunt from right to left, with the gradual onset of cyanosis. However, the group of cardiac malformations in which cyanosis appears relatively late in childhood is not made up entirely of the high ventricular septal defects (table 2). This equalization of the load on the right and left sides of the heart in the interventricular septal defect, patent ductus arteriosus, and aortic-pulmonic defect results in a picture which is roentgenographically, and at times clinically, identical, so that it is difficult to determine what the original lesion was. In fact, let us recognize that the physiologic changes in this group are the same; the anatomic site of the shunt is only slightly different. The development of this appearance, as a result of pulmonary hypertension in any intracardiac or extracardiac left-to-right shunt, with resulting reversal of the shunt and development of cyanosis, has loosely been called the "Eisenmenger physiology." This, of course, is based on a marked similarity in appearance and behavior. Because of the nonspecificity of the appearance, we would much prefer to indicate the change by that factor which is most important in bringing it about; namely, pulmonary hypertension. One might designate the condition, if the anatomic diagnosis is not known, as "cardiac enlargement secondary to left-to-right shunt, with pulmonary hypertension." The term "Eisenmenger's complex" could be dropped entirely, and in the cyanotic stage the lesion could be specified as "high ventricular septal defect with left-to-right shunt." When the peripheral blood in these cases becomes somewhat unsaturated, it might be specified as "high ventricular septal defect with pulmonary hypertension and right-to-left shunt." Such reversal of flow, long familiar in the high septal defects, has become recognized with increasing frequency in other lesions, especially in the patent ductus. 40, 41, 42 That recognition of this physiologic change is exceedingly important is testified to by the fact that once pulmonary hypertension has
PATENT DUCTUS ARTERIOSUS

VENTRICULAR SEPTAL DEFECT
HIGH or LARGE

AORTIC PULMONIC DEFECT

X-RAY APPEARANCE

INCREASED PULMONARY RESISTANCE + PULMONARY HYPERTENSION

"EISENMENGER" CONFIGURATION

FIG. 7

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set in, and particularly if it has been long standing, it may become irreversible. In this case, surgical closure of the patent ductus is an extremely hazardous procedure, and, in fact, of questionable therapeutic benefit.

Cyanotic Group

If we considered all the possible malformations causing cyanosis in the neonatal period in this discussion, we would be preoccupied with innumerable and sometimes monstrous curiosities of the obstetrical nursery and post-mortem room. In considering only those malformations which permit survival for 18 to 24 months or more, we find that the majority, perhaps 60 to 70 per cent, are made up of the tetralogy of Fallot. Thirty to 40 per cent are predominantly made up of the tricuspid atresias, truncales, and well-compensated transpositions. A minority of this latter group are composed of the rarer and exceedingly complex multiple lesions which challenge us constantly in differential diagnosis and call upon the maximum in diagnostic aids, including catheterization, angiography, and so forth.

The most rewarding single roentgenographic sign in subdividing the cyanotic group is abnormal pulmonary blood flow, as recognized by an increase or diminution in the caliber of the pulmonary vascular markings. Tetralogy of Fallot and pure tricuspid atresias usually present diminished pulmonary vascular markings as a result of the decreased pulmonary blood flow. Truncus communis invariably presents pulmonary vascular engorgement, evidenced by prominence of the vessels, often associated with an expansile pulsation or "hilar dance." The appearance of the transpositions is entirely dependent on the compensating associated lesion permitting the transfer of blood from the left to the right. However, the majority of the transpositions, unless some degree of pulmonary stenosis exists, present evidence of pulmonary vascular engorgement.

Tetralogy of Fallot. The roentgenologic appearance is determined by the degree of pulmonary stenosis and the functional degree of overriding of the interventricular septum by the aorta. Variations in the degree of the pulmonic stenosis or overriding of the aorta result in a wide range of appearances from that approaching a normal heart (in which the pulmonic stenosis is mild and the overriding minimal) to that of the extreme "coeur en sabot" (in which the pulmonary artery is atretic and the aortic overriding marked, that is, pseudotruncus). There are all intermediates.

As the ventricular enlargement is purely right and the left ventricle is either normal, or may be even somewhat hypoplastic, the enlargement is not reflected in the postero-anterior projection by an increase in the transverse diameter of the cardiac silhouette. In fact, in moderate to severe cases, the heart may appear smaller than normal. The heavy right ventricle dips into the diaphragm and elevates the apex of the heart, accounting for the "sabot" appearance. The cardiac waist is usually narrowed because of the absence or hypoplasia of the main pulmonary artery, and the beat in this middle cardiac segment on the left is either absent or diminished. In the left anterior oblique projection the heart appears almost round. Approximately one-fourth of the patients with this condition show evidence of a right aortic arch. The pulmonary vasculature, except in the exceedingly mild cases, is almost uniformly diminished.

If the patient survives a number of years, considerable collateral circulation through the bronchial arteries usually develops, and at first glance, this may mask the apparent diminution in the pulmonary blood flow. On closer examination, the distinctive pattern can be seen (fig. 4). These collaterals can make surgical dissection of the lung root exceedingly hazardous.10

It is widely agreed13 that in at least 90 per cent of cases of tetralogy of Fallot the obstruction is infundibular in position. Brock,18 on the other hand, feels that valvular stenosis in the tetralogy is considerably more frequent "if sought for carefully and properly." From the radiologic point of view it should be emphasized that in tetralogy of Fallot, it is exceedingly uncommon to see gross poststenotic dilatation of the pulmonary artery.

Tetralogy of Fallot With Unilateral Atresia
of the Pulmonary Artery: A rare variant of the tetralogy, which in addition to the usual features presents atresia of one of the main branches of the pulmonary arteries, has been reported.\textsuperscript{44} Roentgenographically, this may be recognized by a discrepancy in the vasculature of the two lungs. Instead of a uniform diminution of the pulmonary vascular pattern in both lung fields, the side with the atretic pulmonary artery will show considerable diminution of the pulmonary vasculature, whereas the opposite lung field will show a dilated main pulmonary artery, compensatory pulmonary vascular engorgement, and occasionally even a "hilar dance." Angiocardiography may confirm the absence of the pulmonary artery distal to the atresia on the affected side. This deformity considerably increases the risk of an operative shunt procedure. Atresia of one of the branches of the pulmonary artery in the absence of a tetralogy or any other abnormality has been recognized for some time.

\textit{Tricuspid Atresia}. This is one of the few entities which may present a distinctive, almost pathognomonic, cardiac silhouette; namely, absence of mass where one is accustomed to seeing the right ventricle.\textsuperscript{46} Unfortunately, less than 20 per cent of the tricuspid atresias present this appearance.\textsuperscript{46} The majority have a "coeur en sabot" configuration indistinguishable from tetralogy of Fallot, but they have a definite distinctive feature in that the electrocardiogram almost uniformly presents evidence of left ventricular hypertrophy instead of the characteristic right ventricular hypertrophy of the tetralogy of Fallot. In our experience it is the enlarged right auricle (probably a function of the size of the auricular septal defect) which fills in the space normally occupied by the right ventricle and accounts for the "coeur en sabot" silhouette. Thirty per cent of the patients with tricuspid atresia are found to have dextroposition or dextrocardia, and approximately 30 per cent, irrespective of the position of the heart, have reversal of the position of the aortic arch.

A new, and probably helpful radioscopic sign of tricuspid atresia has been reported.\textsuperscript{46} This is an asynchronous pulsation of the anterior and posterior borders of the heart in the left oblique position. It is attributed to the replacement of the right ventricle, normally the main component of the anterior border, by the enlarged right atrium.

\textit{Pulmonic Stenosis With Atrial Septal Defect or Foramen Ovale}. In this combination of lesions there are sufficient deviations from the classic signs of uncomplicated pulmonic stenosis to make it probable that in a given case a pattern will emerge which is incompatible with either pulmonic stenosis or auricular septal defect alone. If the patient is cyanotic, one must assume that the pulmonic stenosis is severe and dominant, thus reversing the septal shunt to a right-to-left flow. In the roentgenologic appearance of the heart, the pulmonic stenosis dominates, with prominence of the auricles. If cyanosis is present there will be right ventricular enlargement, auricular prominence, and pulmonary ischemia, as evidenced by diminution of the pulmonary vasculature. This conforms with our experience and recent reports of others.\textsuperscript{14, 15, 47}

It is important to recognize that this combination of lesions may give rise to cyanosis at birth, but in the majority of patients, the onset is later. In the vast majority there is poststenotic dilatation of the main pulmonary artery. This combination of lesions becomes exceedingly important in the differential diagnosis with tetralogy, as they do not respond well to shunt procedures, and valvulotomy appears a more rewarding procedure.

\textit{Ebstein's Disease}. This deformity, represented by a congenital downward displacement of the tricuspid valve into the right ventricle, has aroused considerable interest since Ebstein's original description in 1896. As it represents a distinct pathologic entity. In the presence of an auricular septal defect, which is the most commonly associated lesion, cyanosis always results.

In our experience the roentgenologic appearance of the heart is characterized by distinctive features which, however, are not necessarily specific for this entity. It presents a pure, exaggerated, right-sided enlargement with diminished prominence of the pulmonary vascular markings. Thus, the heart may be described
Transpositions of the Great Vessels. There are almost innumerable variations from the complete transposition in which the aorta arises from the right ventricle and the pulmonary artery from the left ventricle to variable degrees of overriding of the two ventricles by one or both of these vessels. Many of these malformations now travel in medical parlance under proper names which help to obscure their anatomic identity rather than emphasize their kinship based on the common denominator that in all these cases the basic malformation is the abnormal origin of the major vessels. The so-called “Taussig-Bing malformation” represents such a variation. Here the aorta arises from the right ventricle, while the pulmonary artery arises mainly from the right ventricle, but partially overrides the ventricular septum, receiving blood from both ventricles. Thus, there is cyanosis from birth. Roentgenograms show combined ventricular prominence with a large pulmonary artery, pulmonary vascular engorgement and “hilar dance.”

Generalizations on the clinical aspects, the course of the disease, the murmurs, the electrocardiographic findings, and particularly the roentgenographic appearance of the heart are not valid in transpositions. This is due to the fact that these will all be dependent on the associated defects which compensate for the transposition by permitting admixture of blood from the right and left sides of the heart. One common observation will be the abnormal relationship of the base of the aorta and pulmonary artery to the heart, usually characterized by a mediastinal shadow narrowed in the frontal projection and widened in the left anterior oblique view. To evaluate the remaining characteristics, one must think in terms of transposition compensated by atrial septal defect and patent ductus, or transposition with ventricular septal defect. Equally important is the size of these defects. Thus, there is much variation in this group.

Eighty-six per cent of the patients with transpositions die by the end of the first year, although patients have been reported to have lived to the age of 56 years without significant physical limitations. In the presence of an ab-
normal electrocardiogram, progressive cyanosis, and rapid increase in size of the heart, the diagnosis usually can be suspected in the first few weeks or months of life.

Anomalous Systemic Venous Return. Anomalous insertion of the superior or inferior vena cava or both into the left atrium is extremely rare, and clinically it is almost impossible to diagnose. Per se, this condition need not give rise to murmurs, but in most cases there is an associated congenital cardiac defect. Cyanosis is present, though not necessarily severe. Anatomic studies indicate that anomalous insertion of a persistent left superior vena cava is more likely than a right. Angiocardiography is probably the only method of making a definitive diagnosis.

As the curative potential of surgery should be so great in this type of deformity, one is constantly intrigued by the possibility of diagnosing this lesion, and it is probably a sound clinical policy to make all angiocardiograms for cyanotic children by means of a left-sided injection because of the greater chance of visualizing an abnormal insertion of a persistent left vena cava.

Anomalous Pulmonary Venous Return. As a result of a congenitally abnormal insertion, blood returning from the lungs via the pulmonary veins may drain into the superior vena cava, innominate, azygos, or subclavian veins, or into the right atrium. If there is complete drainage of pulmonary blood into the right side, an atrial septal defect usually exists to sustain life. More common is partial drainage of the pulmonary blood into one of these structures by way of abnormal insertion of one or more of the pulmonary veins. If more than 50 per cent of the pulmonary venous blood drains into the right atrium, important clinical signs may be expected. Conventional chest roentgenograms may present a characteristic appearance, as has been reported by Snellen and Albers.44 The abnormal vessels are then recognized low and lateral to the right border of the heart, with the upper part of the mediastinum widened, resulting in a "figure of 8" mediastinal silhouette44 (figs. 9 and 10).

Primary Myocardial Disease. In evaluating cardiac disease in children, particularly in infants, a number of patients will be found who have neither rheumatic nor congenital heart disease. This group recently has been well studied by Rosenbaum and his associates.55 and clinically, these patients are found to have certain features in common: cardiomegaly, absence of significant murmurs, electrocardiographic abnormalities, and normal blood pressure. Their patients all had primary myocardial disease.

By the time these patients are examined, they present almost uniformly similar roentgenologic appearances. The heart is generally enlarged, all chambers sharing. There is a diminished excursion of the beat. The main vessels are normally placed, and the pulmonary vasculature is either normal or shows passive engorgement with congestion. The picture is that of a failing myocardium. Although at least five underlying pathologic entities have been shown to be involved (glycogen storage disease, aberrant left coronary artery, medial necrosis of the coronary arteries, subendocardial sclerosis, and idiopathic myocarditis), once the myocardium fails, the radiologic picture becomes the same. It is of interest that the first three of these entities fail to respond to any form of therapy, while the latter two may respond.55

Angiocardiography

The value of angiocardiography in the research laboratory and as a teaching medium has been established. As a diagnostic clinical instrument it has a limited but specific place.

Clarification of the limitations of cardiac angiotherapy have only recently been stressed in the literature.56 Important among these are: a. A certain concentration of radiopaque substance is necessary at a given time in the circulation to be visualized on the films. Therefore, minute shunt or minimal overriding of the vessels will be overlooked by this method. b. Certain chambers or portions of chambers may be opaque for only a fraction of the cardiac cycle. Therefore, the speed and frequency of exposure become exceedingly important. The minimum practical requirement for a serialographic machine appears to be 10 to 12 films in eight seconds, although there
Figure 9. Anomalous pulmonary venous return with complete drainage of pulmonary blood into the right side of the heart via a persistent left-sided vena cava draining into the innominate vein and then into right vena cava resulting in the "figure of 8" configuration of the mediastinum.

Figure 10. Anomalous pulmonary venous return with only slight widening of the upper mediastinum on the left resulting from abnormal venous return but with marked enlargement of the right atrium resulting from the direct return of the major portion of the pulmonary blood into the right atrium.

appears to be some evidence that a faster rate of exposure may yield additional significant information.19 The literature is studded with reports in which angiocardiography either failed to give information or was actually misleading.10, 51, 57, 58 As a rule, angiocardiography may show right-to-left intracardiac shunts of moderate magni-
tude, while left-to-right intracardiac shunts are better demonstrated by the catheter. There has been an increased application of angiocardiography in the diagnosis of lesions in the neonatal period and infancy. Here, in the opinion of the authors, this diagnostic instrument has unique rewards to offer, in that it is possible to follow the course of the blood and to establish abnormalities prior to the time that adaptive changes take place in the heart which might make them recognizable by routine methods. This becomes important clinically in those infants suffering cardiac enlargement, failure, or other gross cardiac disturbances which cannot be diagnosed by other means.  

**Retrograde Aortography**

This specialized surgical procedure yields specific information in the localization of left-to-right shunts, in establishing whether they are intracardiac or extracardiac. This is a problem in which the catheter has not proved to be entirely accurate, and one which arises primarily in differentiating the inoperable high interventricular septal defects from the operable aortic-pulmonic defects and the patent ductus arteriosus. In coarctation of the aorta this procedure may yield specific information regarding the location of the coarcted segment. The retrograde injected dye follows the course of the blood instead of piling up above and below the coarcted segment, so that, as a rule, it demonstrates the entire collateral vascular channels better than the coarctation itself. The tighter the coarctation becomes, the more difficult it is to outline it accurately by dye.

This procedure should not be treated lightly, as deaths and cerebral complications are known to occur with considerable frequency, apparently the result of a high concentration of the contrast material reaching the brain. Complications probably can be minimized by checking the position of the catheter fluoroscopically prior to injection, if a catheter is used, to be certain that it has not passed up the carotid, by compression of the head and neck vessels cephalad to the site of the injection, and by avoidance of multiple injections.

**Vascular Rings and Aberrant Vessels**

There are many vascular malformations of the aortic arch and its branches, but the majority are of little importance. Those which produce actual compression of the trachea or the esophagus or both may give rise to difficulties in swallowing and stridor. The symptoms in these cases are almost invariably exaggerated during feeding. Fortunately, these anomalies can be readily recognized by roentgenoscopic and roentgenologic examination. Our cases have fallen into two general categories, the compression produced by complete vascular ring and compression by a single misplaced or aberrant artery. In the double aortic arch, the trachea and esophagus are trapped between the arches, one arch passing anterior to the trachea, the other posterior to the esophagus. This deformity produces a characteristic roentgenologic appearance.

Another form of ring is produced by the right aortic arch with a persistent ligamentum arteriosum passing posterior to the esophagus and connecting with the pulmonary artery, similarly compressing the trachea and esophagus, but producing a somewhat different roentgenographic appearance. Fortunately, both of these deformities are amenable to surgery.

A single anomalous artery may also produce compression deformities with symptoms. The anomalous innominate artery may arise further along on the aortic arch than normal and, thus, wind around the anterior surface of the trachea, compressing it. The symptoms are usually dyspnea, stridor, and multiple bouts of pneumonia.

The aberrant subclavian artery is usually the result of the right subclavian taking off independently from the distal part of the aortic arch rather than from its normal origin, the innominate. Then, in order to ascend to the right upper part of the chest, it must pass either between the trachea and esophagus, anterior to the trachea, or behind the esophagus, the latter being by far the most common course. In doing so, it produces a spiral defect in the posterior and left lateral aspect of the esophagus, often associated with obstructive symptoms and long known as the cause of so-
called “dysphagia lusoria.” This deformity is readily recognizable by barium swallow, and in infants there may be a temporary delay with momentary expansion of the upper part of the esophagus as a result of this deformity, particularly when solid or semisolid foods are taken. This often results in gagging and in distress during feeding and, at times, aspiration, although at no time has the obstruction been observed to be severe. In our series of aortic arch anomalies we have had approximately 40 patients within the past five years with symptoms sufficiently severe to justify surgical correction.69

CONCLUSION

As the scope of surgical corrective procedures for congenital heart disease widens, the burden of accurate diagnosis mounts. Correlation of all available data, not just roentgenographic, is the sine qua non for an understanding of the various types of congenital heart disease.

In our present state of knowledge, the simpler diagnostic procedures: History, physical examination, electrocardiogram, film and fluoroscopic observations, if properly correlated, should yield a working physiological diagnosis in 85 per cent of patients with congenital heart disease. The exact nature of the lesion in half of the remaining 15 per cent may become clear after angiography and/or catheter studies.

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MARTIN H. WITTENBORG and EDWARD B. D. NEUHAUSER

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