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and
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Roentgenology of Congenital Heart Disease

Volume XIX | Friday, June 11, 1948 | Number 30

BULLETIN OF THE
UNIVERSITY OF MINNESOTA HOSPITALS
and
MINNESOTA MEDICAL FOUNDATION

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UNIVERSITY OF MINNESOTA MEDICAL SCHOOL
CALENDAR OF EVENTS

Visitors Welcome

June 14 - June 19, 1948

No. 207

Monday, June 14

9:00 - 9:50 Roentgenology-Medicine Conference; L. G. Rigler, C. J. Watson and Staff; Todd Amphitheater, U. H.

9:00 - 10:50 Obstetrics and Gynecology Conference; J. L. McKelvey and Staff; Interns' Quarters, U. H.

9:15 - Fracture Rounds; A. A. Zierold and Staff; Ward A, Minneapolis General Hospital.

10:00 - 12:00 Neurology Ward Rounds; A. B. Baker and Staff; Station 50, U. H.

11:00 - 11:50 Roentgenology-Medicine Conference; Staff; Veterans' Hospital.

11:00 - 12:00 Cancer Clinic; K. Stenstrom and A. Kremen; Eustis Amphitheater, U. H.

12:15 - 1:20 Obstetrics and Gynecology Journal Club; M-435, U. H.

12:30 - 1:50 Surgery Grand Rounds; A. A. Zierold, Clarence Dennis and Staff; Minneapolis General Hospital.

1:30 - 2:30 Pediatric-Neurological Rounds; R. Jensen, A. B. Baker and Staff; U. H.

2:00 - 3:00 Surgery Problem Case Conference; C. Dennis and Staff; Small Class Room, General Hospital.

5:00 - 6:00 Urology-Reontgenology Conference; D. Creevy and H. M. Stauffer and Staffs; M-109, U. H.

Tuesday, June 15

8:30 - 10:20 Surgery Reading Conference; Lyle Hay; Small Conference Room, Bldg. I, Veterans' Hospital.

9:00 - 9:50 Roentgenology Pediatrics Conference; L. G. Rigler, I. MdQuarrie and Staffl Eustis Amphitheater, U. H.

10:30 - 11:50 Surgical Pathological Conference Lyle Hay and Robert Hebbel; Veterans' Hospital.

12:30 - 1:20 Pathology Conference; Autopsies; Pathology Staff; 102 I. A.

2:00 - 2:50 Dermatology and Syphilology Conference; H. E. Michelson and Staff; Bldg. III, Veterans' Hospital.

3:15 - 4:20 Gynecology Chart Conference; J. L. McKelvey and Staff; Station 54, U. H.

3:30 - 4:20 Clinical Pathological Conference; Staff; Veterans' Hospital.

4:00 - 5:30 Surgery-Physiology Conference; O. H. Wangensteen and M. B. Visscher; Eustis Amphitheater, U. H.

4:00 - 5:00 Pediatric Rounds on Wards; I. McQuarrie and Staff; U. H.

5:00 - 5:50 Urology Pathological Conference; C. D. Creevy and Staff; Todd Amphitheater, U. H.

Wednesday, June 16

8:00 - 8:50 Surgery Journal Club; O. H. Wangensteen and Staff; M-515, U. H.

8:30 - 12:00 Neurology Rehabilitation and Case Conference; A. B. Baker and Joe R. Brown; Veterans' Hospital.

11:00 - 11:50 Pathology-Medicine-Surgery Conference; Aneurysm of Abdominal Aorta; O. H. Wangensteen, C. J. Watson and Staff; Todd Amphitheater, U. H.

4:00 - 5:00 Infectious Disease Rounds; Todd Amphitheater, General Hospital, Veterans' Hospital.

Thursday, June 17

8:15 - 9:00 Roentgenology-Surgical-Pathology Conference; Walter Walker and H. M. Stauffer; M-109, U. H.

8:30 - 10:20 Surgery Grand Rounds; Lyle Hay and Staff; Veterans' Hospital.

9:00 - 9:50 Medicine Case Presentation; C. J. Watson and Staff; Todd Amphitheater, U. H.

10:00 - 11:50 Medicine Ward Rounds; C. J. Watson and Staff; E-221, U. H.

10:30 - 11:50 Surgery-Radiology Conference; Daniel Fink and Lyle Hay; Veterans' Hospital.

11:00 - 12:00 Cancer Clinic; K. Stenstrom and A. Kremen; Eustis Amphitheater, U. H.

11:30 - 12:30 Clinical Pathology Conference; Steven Barron, C. Dennis, George Fahr, A. V. Stoesser and Staffs; Large Class Room, General Hospital.

1:00 - 1:50 Fracture Conference; A. A. Zierold and Staff; Minneapolis General Hospital.

4:30 - 5:20 Ophthalmology Ward Rounds; Erling W. Hansen and Staff; E-534, U. H.

Friday, June 18

8:30 - 10:00 Neurology Grand Rounds; A. B. Baker and Staff; Station 50, U. H.

9:00 - 9:50 Medicine Grand Rounds; C. J. Watson and Staff; Todd Amphitheater, U. H.

10:00 - 11:50 Medicine Ward Rounds; C. J. Watson and Staff; E-221, U. H.

10:30 - 11:20 Medicine Grand Rounds; Staff; Veterans' Hospital.

10:30 - 11:50 Otolaryngology Case Studies; L. R. Boies and Staff; Out-Patient Department, U. H.

11:00 - 12:00 Surgery-Pediatric Conference; C. Dennis, A. V. Stoesser and Staffs; Minneapolis General Hospital.

12:00 - 1:00 Surgery Literature Conference; Clarence Dennis and Staff; Minneapolis General Hospital, Small Class Room.

1:00 - 1:50 Dermatology and Syphilology; Presentation of Selected Cases of the Week; H. E. Michelson and Staff; W-312, U. H.

1:00 - 2:50 Neurosurgery-Roentgenology Conference; W. T. Peyton, Harold O. Peterson and Staff; Todd Amphitheater, U. H.

Saturday, June 19

7:45 - 8:50 Orthopedics Conference; Wallace H. Cole and Staff; Station 21, U. H.

8:00 - 9:00 Pediatric Psychiatric Rounds; Reynold Jensen; 6th Floor West Wing, U. H.

9:00 - 10:30 Pediatric Grand Rounds; I. McQuarrie and Staff; Eustis Amphitheater, U. H.

9:00 - 9:50 Surgery-Roentgenology Conference; O. H. Wangensteen, L. G. Rigler and Staff; Todd Amphitheater, U. H.

9:00 - 9:50 Medicine Case Presentation; C. J. Watson and Staff; E-101, U. H.

10:00 - 11:50 Medicine Ward Rounds; C. J. Watson and Staff; E-221, U. H.

10:00 - 12:50 Obstetrics and Gynecology Grand Rounds; J. L. McKelvey and Staff; Station 44, U. H.

II. ROENTGENOLOGY OF CONGENITAL HEART DISEASE

Herbert M. Stauffer

Fundamental advances in the diagnosis and treatment of congenital heart disease in the past ten years have resulted in a tremendous intensification of interest in this field. The development of the methods of angiocardiography and right heart catheterization have made possible accurate anatomical and often physiological diagnosis in a field where the label "congenital heart" was formerly very often the final clincial diagnosis. At the same time, knowledge gained from these more complex methods of examination has aided greatly in our ability to interpret the usual and simpler methods of examination, particularly the x-ray examination. The development of methods of surgical attack for patent ductus arteriosus, tetralogy of Fallot, coarctation of the aorta and certain of the great vessel anamalies has at once created the necessity for accurate anatomical diagnosis and has provided in the larger centers a stream of patients with all sorts of congenital heart disease to be evaluated as possible candidates for surgery. Roentgenology has both contributed greatly to and profited greatly from this revival of interest in congenital heart disease. Angiocardiography and heart catheterization make possible accurate anatomical diagnosis where that possibility never existed before, short of autopsy. It has thus become possible in the larger clinics for those interested in this field to observe in a relatively short period of time large numbers of well authenticated cases of congenital heart disease, proved examples of which were regarded as curiosities a few years ago.

It is the purpose of this paper to discuss briefly the methods available today for the roentgenologic study of congenital heart disease. The roentgen findings in patent ductus, tetralogy of Fallot, coarctation of the aorta and the great vessel anomalies will be presented together with some consideration of the differential diagnostic possibilities. Work with angiocardiography and heart catheterization in congenital heart disease at the university of Minnesota Hospitals is just beginning so that this report is based on the use of the ordinary roentgenological methods of examination. It should be emphasized at the outset that the roentgenological examination is but a part, albeit an important one, in the total diagnostic evaluation of cases of congenital heart disease. In a great many instances the roentgen findings are not specific for any particular anomaly or combination of anomalies and they will serve mainly to corroborate the clinical impression or to suggest the possibility of some other process.

Roentgenological Methods[35,39,42]

Fluoroscopy is by far the simplest and most essential part of the examination in congenital heart disease as in any other type of heart disease. The fluoroscopic examination permits the study of the heart under physiological conditions of respiration. This desirable feature is often not achieved in the film studies since only one phase of respiration is caught and extreme changes in the cardiac contour result in the extremes of inspiration and expiration. The phases of respiration also produce extreme variations in the filling of the pulmonary vessels which is an important diagnostic feature in many of the cases. The standard position for fluoroscopic observation are the postero-anterior and the right and left anterior oblique positions with approximately 50 degrees of rotation, and the lateral position. Fluoroscopy permits an attempt at evaluation of the relative sizes of cardiac chambers on the basis of their pulsations as well as their contour and, of course, an attempt is made to evaluate the amplitude of pulsations of both the heart borders and the great vessels. Along the left border in the postero-anterior view of pivotal point of opposite pulsation can commonly be recognized which may be elevated in the event of left ventricular enlargement or depressed to a lower point along the left border in right ventricular enlargement. Similarly, in the left anterior oblique view, anterior to the left lower pole of the heart along the diaphragmatic surface of the left ventricle a small notch may

be observed representing the interventricular septum. Displacement of this point anteriorly may indicate left ventricular enlargement; or posteriorly may indicate right ventricular enlargement. These points have not proved of very great help to us in the analysis of the congenital heart, and Sussman[39] states on the basis of angiocardiography that the apparent position of the interventricular notch does not correlate well with changes in the relative sizes of the ventricles.

An attempt to evaluate the amplitude of pulsations is of some value. The amplitude of the cardiac pulsations is apt to be increased in the presence of a significant shunt and this is particularly noteworthy in the cases of patent ductus. When the pulmonary vessels are dilated they ordinarily show increased pulsations, as in the cases of patent ductus and interatrial septal defect. When the hilar and pulmonsry vessels are normal in size or somewhat reduced the evaluation of pulsations becomes much more difficult and open to considerable variations in individual interpretation. A great deal of importance has been attached to evaluation of these pulsations in the diagnosis of tetralogy of Fallot where they are ordinarily diminished. In our experience it is often extremely difficult to differentiate with any certainty the diminished pulsations in this condition from the normal and particularly to distinguish transmitted from actual expansile pulsations of the vessels themselves.

Visualization of the relationship of the opacified esophagus is of the greatest importance. The deviation of the esophagus about the enlarged left atrium, which ordinarily displaces the esophagus to the right and posteriorly is well known and may suggest involvement of the mitral valve, either by congenital or acquired mitral stenosis. In dealing with congenital heart disease the relationships at the level of the arch of the aorta are perhaps even more important. The normal left-sided aortic arch produces an indentation on the left side of the esophagus in the postero-anterior view and on its anterior surface in the right oblique and lateral views. Similarly, immediately below this indentation in the right oblique and lateral views a second indentation is commonly noted at the level of the right pulmonary artery, but probably produced by structures adjacent to the artery including the left main bronchus and adjacent lymph nodes, etc. It is immediately below this second indentation that the broader indentation of the left atrium will be noted if enlargement is present. In dealing with young infants, particularly those with stridor or dysphagia where compression upon the esophagus is to be determined, the administration of barium mixture may be dangerous because of the possibility of aspiration. For this reason it has become good practice to visualize the upper esophagus by injection of several ccs. of lipiodol through a small catheter, enough to well distend the esophagus, particularly at the level of the great vessels.

Roentgenography

The roentgenographic examination is principally important in providing a record for permanent study. It also has the advantage that because of the increased target distance the magnification and distortion which is inherent in the fluoroscopic method is minimized. The film studies are made in the standard positions as outlined under the fluoroscopic procedure. A film should be made with barium in the esophagus particularly in the postero-anterior and lateral views, but also wherever possible in the oblique views also.

Angiocardiography

The history of the development of methods of visualizing the intracardiac chambers by the injection of opaque contrast material begins with the demonstration by Forssman[18] of the possibility of passing a ureteral catheter through one of the arm veins, entering at the elbow, into the right side of the heart. Subsequently the Portugese school of Egas Moniz[24] injected sodium iodide into the heart through such catheters in aminals, calling the method angiopneumography, since they succeeded in visualizing the pulmonary vascular pattern in this manner. Later other European workers

visualized the lung vascular pattern in man using sounding of the right atrium and concentrated sodium iodide solution. The severe reactions attending the use of sodium iodide in this manner discouraged further work until the pyridine derivatives, notably Per-Abrodil and Uro-Selectan B became available. Castellanos and Pereiras[10] of Havana were the first to do extensive studies using these substances by injection rapidly into the antecubital veins of infants. Castellanos and Pereiras reported a number of clinical case studies in infants using their method with demonstration of intracardiac shunts, pulmonic stenosis, and transposition of the great vessels. Their monograph on angiocardiography in the infant appeared in 1938. They reported that they had never seen troublesome symptoms or accidents of any kind despite application of the method to serious cases with severe heart failure, orthopnea or deep cyanosis. The 35 per cent solutions of radio-opaque material which they employed were not sufficiently opaque to yield satisfactory results in older children.

The technique of angiocardiography as perfected by Robb and Steinberg[32] and first reported in 1938, employing 70 per cent diodrast is the one generally used in this country today. These authors injected 25 to 45 ccs. of 70 per cent diodrast through a 12 gauge needle into an antecubital vein with the arm raised to an angle approximately 45 degrees above the horizontal. For visualization of the right ventricle and the pulmonary artery a film is exposed one or two seconds less than the arm to lung circulation time, usually three seconds, while the time of exposure for the left ventricle varies between six to nine seconds, one to two seconds less than the arm to head circulation time. The cassette changer used for stereoscopic chest films in x-ray departments has been widely used to obtain two successive exposures for angiocardiography. However, it is desirable to secure at least six to eight serial exposures over a period averaging ten seconds following completion of the injection, and for this purpose various cassette changers have been devised. Rotary wheel and vertical magazine types have been employed. One of the most effective devices in use at present is that employed by Sussman[39] in which the lead backed cassettes stand vertically on an inclined plane in a magazine with the one in front being dropped in a lower magazine by an electrically operated catch after the x-ray exposure has been made. The exposure mechanism can also be integrated electronically with electrocardiographic leads from the patient so that the exposures can be made in any predetermined sequence of cardiac cycles and can even be made in systole or diastole employing the R wave of the electrocardiogram to "fire" the exposure mechanism. The x-ray equipment should permit exposure times of one-twentieth of a second or less.

Robb and Steinberg and Sussman have advocated carrying out angiocardiography in the upright position. Others, particularly those dealing principally with infants and children, use the horizontal position. In a preliminary trial of angiocardiography on a case of coarctation of the aorta in an 11 year old boy, we have used a cassette changer of the type described by Neuhauser[25]. This is a very simple device consisting of a box in which six lead backed cassettes rest on a platform supported on four bedsprings. The cassettes are ejected partially by an assistant on one side of the table and pulled out by a second assistant on the opposite side of the table with the next cassette below springing immediately into place upon which a third assistant closes the circuit making the x-ray exposure. It was found feasible to make six exposures in this fashion in ten seconds. With the rapid injection of 20 ccs. of 70 per cent diodrast, visualization of the right heart and pulmonary arterial tree was thoroughly satisfactory in the postero-anterior view and to some extent in the lateral, for which purpose the injection was repeated a few minutes after the initial injection for the postero-anterior view. Visualization of the left side of the heart and the aorta was unsatisfactory in this one case.

As has been stated, the original in-

vestigations using this method employed heart catheterization with ureteral catheters and injection of opaque material through the catheters. The disadvantage of this method was the narrow bore of the catheters which hindered the extremely rapid injection which is necessary for opacification. Recently Chavez[11] has published beautiful illustrations of opacification of both the right and left sides of the heart using a 16 F. ureteral catheter in the adult. The catheters is passed into the right atrium directly through the external jugular vein entered through a small incision in the lower part of the neck. That this method has considerably more hazard than the injection into the antecubital vein is indicated by the fact that Chavez in a small group of cases reported one death, and a death was reported following injection of 70 per cent diodrast through an ordinary ureteral type of heart catheter.

Sussman[39] states that the right auricle, right atrium and ventricle are clearly seen immediately after injection. The pulmonary valve is regularly visualized when several exposures are made early following injection and the pulmonary veins and left auricle are quite regularly visualized. On the other hand, the left ventricle and aortic valve are not constantly outlined although the aorta, in the normal individual, is visualized about five to seven seconds after injection and the branches of the arch of the aorta often can also be visualized. With a right to left interventricular shunt or transposition of the great vessels opacification of the aorta occurs within two seconds. In this case the aorta together with its major branches are visualized including the superior mesentreric artery.

Angiocardiography has thrown new light on the relationship of various parts of the cardiac contour to the underlying heart chambers and great vessels in the various roentgenologic positions. One of the most important concepts arising from this work is that the pulmonary conus actually has a deep location within the cardiac silhouette as does the origin of the pulmonary artery. The middle left cardiac silhouette in the postero-anterior view is regularly formed in the normal individual by the pulmonary trunk and the left pulmonary artery. This point has been re-emphasized recently by Chavez et al. The right ventricle forms no part of the cardiac contour in the postero-anterior view. While the outline of the left atrium is within the cardiac shadow, the left auricular appendage approaches the contour on the left side between the pulmonary artery segment and the border of the left ventricle. In the right oblique view with moderate degrees of rotation, the left ventricle forms the lower anterior border while with more marked rotation the right ventricle forms the lower anterior contour and the pulmonary artery the upper anterior contour. In the left oblique view with moderate rotation the lower anterior contour is formed by the right atrium and right auricular appendage with the ascending aorta above, while with more marked rotation the anterior surface of the right ventricle forms the lower contour. The lower posterior contour is formed by the left ventricle and the upper posterior contour of the heart by the left atrium. The vascular portion of the silhouette in the left oblique view shows the aortic arch forming the so-called aortic window across which the left branch of the pulmonary artery courses. In the lateral view the right ventricle forms the anterior surface of the heart while the left ventricle forms the lower posterior contour with the left atrium forming the upper posterior contour.

Kymography

Roentgen kymography by the multiple slit method, although it has proved interesting in the recording of the increased amplitude of pulsations over the heart borders and the pulmonary arteries in cases of congenital heart disease of certain types, has not provided information of specific practical diagnostic value so that it has not become one of the essential methods in the roentgenologic study of these cases. The method of Keys and Friedell[23] for measuring the stroke volume of the heart with the roentgenkymogram has been applied in cases of patent ductus arteriosus with

demonstration the the flow through the ductus can be very large, up to 50 per cent of the kymographically estimated stroke output, with the "leak" roughly proportional to the arterial pulse pressure.

Electrokymography[36,40], likewise, although it represents a more refined method of recording cardiac and vascular pulsations and permits correlation of these with the events of the cardiac cycle, has not as yet yielded any practical information in congenital heart disease. In fact, our records in a considerable number of cases of various types of congenital heart disease have shown tracings remarkably similar to those obtained from normal individuals. Due to the electrical character of the recording the amplitude of tracings obtained from the borders of structures is not of particular significance. Some suggestive results have been obtained by recording the density changes over the pulmonary parenchyma at a distance from visible vessels of any size with comparison of the amplitude of these pulsations with the deflection produced by introducing during the recording one millimeter of aluminum into the x-ray beam. Comparison between individuals would ony be possible where x-ray kilovoltage and intensity factors and chest thickness are the same. Records in several cases up to date suggest reduced or absent pulsations over the parenchyma in cases of tetralogy of Fallot.

Heart Catheterization

Heart catheterization has been very fully considered in an earlier Staff Meeting presentation this year by Doctors Borden and Ebert[5]. Its importance in the roentgenology of congenital heart disease lies not only in the fact that fluoroscopy is ordinarily an essential part of the procedure, but also because the roentgenologist obtains from a successful catheterization positive diagnostic information in cases which would otherwise remain obscure. While the method primarily provides physiologic information, thus complementing the angiocardiographic anatomical studies, the catheter in some cases may actually be visualized passing through intracardiac septal defects or from the right ventricle directly into the aorta in the case of dextroposition or transposition of the aorta. As the catheter is passed from the superior vena cava successively into the right atrium, the right ventricle and the pulmonary artery, comparison of the position of the tip of the catheter as noted fluoroscopically with the pressure and oxygen saturation determinations provides an accurate index of the position of the catheter. Spot films made with the fluoroscope during the catheterization provide a record of the catheter position for correlation with the oxygen saturation and pressure studies.

Congenital Heart Lesions of Infancy

The cardiac silhouette in the infant is usually of relatively little help in diagnosis. The cardiac silhouette may be within normal limits in the presence of the most severe anomalies. If definite changes are present, these usually consist of enlargement of one or more of the chambers which usually cannot be definitely identified, the heart assuming a more or less globular character as it enlarges. The presence of pulmonary stenosis may be suggested by a deeply concave pulmonary artery segment along the left heart border with possibly a sabot type of cardiac silhouette as a result of lifting of the cardiac apex on the left accompanying right ventricular enlargement. Additional evidence suggesting pulmonic stenosis is the increased radiobility of the lung fields with diminution in the size of the hilar vascular shadows. The pulmonary vessels may be enlarged in the case of left to right shunts, in the case of certain rare examples of unequal division of the truncus arteriosus, or in the presence of cardiac failure. Congenital idiopathic hypertrophy causes heart enlargement in the absence of any recognizable defect in the formation of the cardiac chambers, valves or great vessels.

The following case illustrates the difficulty of making a definite roentgenologic diagnosis in this age group when angiocardiography is not employed.

____, U.H., male infant, 11 months

of age, had had persistent cyanosis from birth. A harsh systolic murmur over the entire precordium had been a constant finding. The child was followed in the Outpatient Clinic and was developing normally. On the day of admission the patient had a tonic convulsion and following the convulsion had been comatose and rigid. Roentgenograms made in the Outpatient Department had shown no characteristic changes in the cardiac silhouette. The EKG showed right axis deviations in clinical diagnosis of tetralogy of Fallot was made. The child was critically ill on admission and expired the same day. Autopsy showed complete transposition of the great vessels with aorta arising from the right ventricle and the pulmonary artery from the left. There was a high interventricular septal defect 6 mm. in diameter. The right ventricle was markedly hypertrophied and the right auricle markedly dilated. The pulmonary arteries and aorta appeared normal.

Patent Ductus Arteriosus

The appearance of the heart and great vessels may be entirely normal in cases of isolated patent ductus arteriosus. Thus in nine of 38 cases in which the roentgenograms were reviewed, the heart size was within normal limits. These 38 cases represent part of the series which have been operated upon at the University of Minnesota Hospitals in the past several years. The ages of the patients whose films were reviewed ranged from 19 months to 33 years. In only four of the cases showing cardiac enlargement was the enlargement extreme. In two of these, very large ducts were found at the time of the surgical procedure. In 35 cases the pulmonary artery segment was abnormally prominent and in 22 there was increase in the size of the hilar and parenchymal lung vessels. There was evidence of left ventricular enlargement in 24 cases, but in only a few cases could evidence of right ventricular enlargement be recognized in the films. Of 25 cases in which films with barium in the esophagus were available for study of the left atrium, 15 showed some degree of enlargement, usually slight.

These findings are in general agreement with those recorded in the literature. The commonest changes, namely enlargement of the pulmonary artery segment and the pulmonary artery branches with varying degrees of left ventricular enlargement and a normal sized aorta present the average roentgenological picture. Any marked deviation from this average type of silhouette, including marked cardiac enlargement, definite evidence of predominant right ventricular enlargement or other variations must make one think of some other anomaly or of associated anomalies if a typical machinery murmur is present.

In many of the cases decrease in the size of the heart and decrease in the size of the pulmonary artery was notable following ligation or severing of the ductus.

Fluoroscopically increase in the pulsations of the left ventricle, the pulmonary artery segment, and of the hilar vessels were observed and in many cases roentgenkymograms were obtained demonstrating the increased amplitude of these pulsations.

The presence of left atrial enlargement in patent ductus arteriosus is an interesting phenomenon and experience, even in large series of cases, is varied with respect to this finding. In the series reported by Donovan, Neuhauser and Sosman[16], of 45 cases in which the status of the left auricle was recorded, 32 or 71 per cent showed enlargement of the left auricle. On the other hand, Steinberg, Grishman and Sussman[38] reported on 27 cases, some of which had been studied with angiocardiography in addition to the routine methods, found no instances of enlargement of the left atrium. Eppinger and Burwell[17] demonstrated by analysis of blood samples from the aorta and pulmonary artery during operations for ductal closure that the left ventricle had to pump from two to four times the amount of blood put out during the same time by the right ventricle. This accounts for the roentgenological findings and these authors further suggest that if the normal mitral valve is not sufficiently wide to transmit the large additional volume of blood without an elevated left auricular pres-

sure visible dilatation of the left atrium may result.

Steinberg et al[38] report an angiocardiographic finding which they believe to be thoroughly specific for patent ductus. This consists of a localized dilatation extending anteriorly from the distal portion of the arch of the aorta at or near the site of origin of the ductus. This localized dilatation just beyond the isthmus has been described anatomically. All but one of 27 cases studied angiocardiographically showed this abnormality.

The lesions most apt to be confused roentgenologically with patent ductus are those showing marked enlargement of the pulmonary artery. These include interatrial septal defect, Eisenmenger's complex, and idiopathic dilatation of the pulmonary artery. In all of these the pulmonary artery dilatation is apt to be considerably greater than seen in isolated patent ductus arteriosus.

Thetralogy of Fallot

In 1888, Fallot analyzed 55 collected cases of cyanotic congenital heart disease including three of his own. Seventy-four per cent of these cases were examples of the tetralogy which now bears this author's name[9]. This tetrad of malformations includes stenosis, hypoplasia or atresia of the pulmonary artery of developmental type, high interventricular septal defect, dextroposition of the aorta, and hypertrophy of the right ventricle. Fallot held that a physician might well diagnose this tetralogy on being confronted with a case of cyanotic congenital heart disease. In a general way this statement still holds good today for cases surviving beyond infancy. The roentgenological findings aid chiefly in weeding out examples of other less common malformations where the x-ray picture may deviate from the average seen with the tetralogy. In addition to other less frequent associated anomalies it was early noted that a persistent right-sided aortic arch occurred frequently in cyanotic heart disease, and in particularly, with the tetralogy of Fallot. This additional anomaly, which is not to be confused with the dextroposition of the aorta which is part of the basic tetrad, is ordinarily readily demonstrated roentgenologically.

The typical or textbook roentgen picture of tetralogy of Fallot consists of the sabot-shaped contour of the heart itself with prominence of the left lower pole of the silhouette which is blunt and somewhat elevated from the leaf of the diaphragm. This finding results principally from predominance of the right ventricle in size over the left ventricle and is not in itself specific for the tetralogy of Fallot since it can be present in any condition giving rise to this type of right ventricular enlargement. In addition, there is ordinarily a deep concavity of the pulmonary artery segment which further accentuates the sabot shape of the cardiac contour. The hilar shadows are commonly small and the lung fields appear more radiable than normal due to the diminutive size of the pulmonary arteries. If, as is true in about 20 per cent of the cases[42], a right sided aortic arch is present, there is an absence of the aortic knob prominence on the left upper segment of the cardiac silhouette and in its place there is a somewhat similar prominence on the right side at the same level. In such a case when barium is administered the stream is deviated slightly to the left at the level of the aortic arch instead of its normal deviation to the right. Ordinarily, however, there will be no anterior displacement of the esophagus as it is visualized in the right anterior oblique and lateral views, such as would be the case in the usual example of persistent right-sided aortic arch occurring as an isolated anomaly. In occasional cases there may be sufficient dilatation of the pulmonary conus to produce a prominent convexity of the pulmonary artery segment instead of the much more common concavity.

The development of the Blalock procedure[4] for anastomosing the innominate or subclavian artery to one of the pulmonary arteries has not only made available a large number of cases for study, but also has made it necessary to critically evaluate the ordinary methods of x-ray

examination and to determine the need and indications for more elaborate methods of x-ray diagnosis. A number of observers including Roesler[35] have noted that in certain cases of tetralogy of Fallot the cardiac silhouette may be remarkably normal and they actually exhibit no characteristic features whatever. In infancy the heart appears small and usually not at all characteristic, and with growth, while a characteristic silhouette may take shape, the heart usually remains relatively small.

In reviewing the cases which have been submitted to surgery at the University of Minnesota Hospitals, the striking thing roentgenologically has been the relatively small number of cases which have appeared to present the classic or textbook picture of tetralogy of Fallot. It seems important to reemphasize the fact that variations from this classic picture are apparently so frequent. Potts and Gibson[29], reviewing the results of the Potts procedure for anastomosing the pulmonary artery to the aorta, have recently emphasized the variations in the roentgen picture of their cases.

The roentgenologic findings were reviewed in 42 cases submitted to surgery with the aim of performing the Blalock procedure. The fact that a high percentage of the cases in whom the anastomosis could be performed showed fair to excellent clinical results in corroborative evidence of the correctness of the diagnosis, at least as far as the pulmonic stenosis is concerned. The cases roentgenologically seem to fall very naturally into three large groups. In Group I are included the cases showing a fairly classical silhouette. There were nine cases in this group, one proved at autopsy to be tetralogy of Fallot. In Group II are collected those cases with rather varying appearances with a more globular silhouette than is ordinarily considered characteristic of the tetralogy, but all showing a somewhat elevated blunt cardiac apex. In these cases there was often a straight or convex conus or pulmonary artery segment. However, in these cases there was usually preserved some evidence of a pulmonic notch immediately below the outline of the arch of the aorta or the aortic knob. This notch very commonly reveals the left hilar shadow more markedly than is usual in the normal cardiac silhouette and could well give rise to an erroneous impression that these pulmonary vessels are somewhat larger and more prominent than is actually the case. In this group there were 24 cases, two of which were proved to be instances of the tetralogy at autopsy. In Group III the cardiac silhouette was much more nearly normal and in this group of cases the pulmonic segment was convex. There were nine cases in this group of which two were proved at autopsy to be tetralogies. Two of these cases, despite the absence of any definite abnormality in the silhouette in any view, derived good results from performance of the Blalock anastomosis and in all probability represent examples of tetralogy of Fallot with essentially normal cardiac silhouettes.

An attempt was made to evaluate the appearance of the lung vessels in the x-ray films. These appeared to be within normal limits in 11 of our cases. In 22 cases the hilar vessel shadows or the intrapulmonic smaller branches or both were decreased. In nine cases with small or normal hilar branches there were increased small intrapulmonic vessels visible. This latter finding is obviously suggestive of development of the well known collateral arterial supply to the lungs in pulmonic stenosis by way of anomalous bronchial arteries.

A right aortic arch was present in nine cases. As is usual in the tetralogy of Fallot, there was no evidence of pressure by the aortic arch or associated vessels from behind, although the esophagus was displaced toward the left by the right-sided aortic arch. It is extremely important for the surgeon to know the side on which the aortic arch lies and also to be aware of any additional vascular anomaly which he may encounter. For example, Brean and Neuhauser[7] have recently pointed out the association of an anomalous left-sided origin of the right subclavian artery from a left-sided aortic arch in a case of tetralogy of Fallot. Further, two cases reported by Paul[27] have shown an

aortic arch anomaly in which there is a crossing over of the aorta to the right side after arching normally on the left.

In summary, while practically all of our cases gave some evidence of right ventricular enlargement in the lateral and right anterior oblique view, there was a substantial number of cases which showed a convexity instead of the classical concavity in the pulmonary artery region. A relatively small group of cases showed what might be considered the classical coeur-en-sabot contour. The presence of a rather prominent pattern of finer vessels in the lung fields often is an indication of collateral circulation and does not exclude pulmonic stenosis. These points should be borne in mind so that the diagnosis of tetralogy of Fallot is not erroneously excluded on the basis of roentgen findings deviating from the classical picture. This series bears witness to the fact that the diagnosis can be satisfactorily made in the vast majority of cases without catheterization and angiocardiography. Gross variations from the cardiac silhouette described above demand these more elaborate studies to determine whether there is a possibility of improvement through surgery. The roentgen findings in three cases, in addition to those described above, were atypical and these cases proved not to be instances of the tetralogy of Fallot. These cases will be discussed subsequently.

According to Sussman[39], angiocardiography in tetralogy of Fallot usually shows opacification of the aorta simultaneously with the pulmonary artery with both usually being visualized within two seconds of the end of the injection. The stenosed portion of the pulmonary artery usually can be made out. The right ventricle and auricle are enlarged, but their cavities ordinarily are not very greatly dilated.

Eisenmenger's Complex[9,42]

In this condition, which resembles tetralogy of Fallot in that there is dextroposition of the aorta which overrides a high interventricular septal defect, together with right ventricular hypertrophy, there is no pulmonic stenosis. The pulmonary artery may be normal in size or more characteristically is dilated and the intrapulmonic branches are usually more or less enlarged. The cyanosis in these cases of Eisenmenger's complex may result either simply from the overriding of the aorta with deviation of blood from the right ventricle directly into the aorta. Bing has demonstrated increased peripheral resistance in the pulmonary circuit in such cases which may contribute to the cyanosis.

The striking roentgenologic finding in these cases is the large size of the pulmonary artery and its branches in the presence of cyanosis. In pulmonic stenosis one may encounter, as will be noted below, enlargement of the pulmonary artery segment and occasionally of the main trunk of the pulmonary artery as a result of post-stenotic dilatation, but ordinarily in pulmonic stenosis the intrapulmonic branches are decreased in size rather than enlarged as in the usual example of Eisenmenger's complex.

The cardiac silhouette usually shows right ventricular enlargement in these cases ordinarily with not much overall increase in size of the silhouette. There is not necessarily a characteristic coeur-en-sabot contour.

One example of this condition studied recently occurred in an adult, T.V., U. H., who had been cyanotic from early life and who had considerable effort limitation. Fluoroscopic and roentgenkymographic studies indicated rather small amplitude of pulsations of the pulmonary arteries despite their increased size in this case. The patient was explored surgically with the idea of possibly attempting a Blalock anastomosis, but very high pressure in the pulmonary artery was encountered. The patient succumbed post-operatively and at autopsy showed new and old thrombi in the pulmonary arteries which presumably were at least in part responsible for the diminished pulsations observed roentgenologically.

Tricuspid Atresia and Stenosis[41,42]

In this group of cases of cyanotic congenital heart disease, underdevelop-

ment of the right ventricle is associated with the narrowing or atresia of the tricuspid orifice and there is also pulmonic stenosis or atresia. To be compatible with life there must be a right to left shunt, either an interatrial or interventricular septal defect, usually the former. In the event that there is pulmonic atresia a patent ductus arteriosus must also be present to sustain life past early infancy, unless the bronchial collateral circulation is uaually well developed. The undeveloped right ventricle is responsible for the fact that these cases are the only instances of cyanotic congenital heart disease regularly showing left axis deviation in the electrocardiogram.

Two of the cases explored for the Blalock procedure showed roentgenologically some evidence of left ventricular enlargement, but this was discounted in view of right axis deviation in the electrocardiograms.

The first, [redacted], U. H., female of 19 months, had been cyanotic since birth but with some improvement after six months of age. There were a number of severe respiratory infections with marked dyspnea and cyanosis at those times. A systolic murmur was heard to the left of the sternum in the third intercostal space. Fluoroscopically and in the roentgenograms there was evidence suggesting enlargement of both right and left ventricles with a concave pulmonary artery segment and diminution of the size of the pulmonary vessels bilaterally. The EKG showed right axis deviation. Thoracotomy was performed with the aim of performing a Blalock anastomosis, but the patient expired during the surgical procedure. Marked vascularity of the pleura, especially over the left hilum, was noted during the exploration.

Post-mortem examination of the heart disclosed a large interatrial septal defect, marked stenosis of the tricuspid valve, and a complete stenosis of the pulmonary valve. The interventricular septum was normal and although there was hypertrophy of the right ventricular wall, the cavity of the right ventricle was quite small and evidently would hold only a few cubic centimeters of blood during life. Presumably an extensive collateral circulation sustained life even in the presence of complete pulmonic stenosis and in the absence of a patent ductus.

The second patient, [redacted], U.H., a male, eight years of age, had been cyanotic since birth and was rather small for his age. Moderate cyanosis was present on examination and the heart was slightly enlarged to the left on physical examination. A palpable thrill could be felt over the precordium best along the left sternal border and there was a harsh systolic murmur over the same area. There was a loud continuous murmur over the base of the heart on the left side. There was marked clubbing of fingers and toes. The roentgenographic findings were regarded as somewhat atypical in that there was some evidence of left ventricular in addition to right ventricular enlargement. The EKG showed right axis deviation.

Thoracotomy was performed, but for technical reasons the anastomosis on the right side could not be carried out. Although the patient returned from the operating room in good condition, his condition deteriorated post-operatively and death occurred on the third post-operative day.

Post-mortem examination of the heart showed atresia of the tricuspid and pulmonary valves with an interatrial septal defect, patent ductus arteriosus, and rudimentary right ventricle.

Although in both of these cases there was some roentgenographic evidence that would have been suggestive of the correct diagnosis, electrocardiographic evidence favored the more conventional diagnosis of tetralogy of Fallot. It seems very probable that the right ventricular hypertrophy in the first of these cases accounted for the right axis deviation, but this exploration is not available in the second case.

The following case history illustrates a more typical situation warranting a clinical diagnosis of rudimentary

right ventricle with tricuspid atresia and pulmonic stenosis or atresia.

I___, U.H., 3½ year old female had been cyanotic since birth. The cyanosis has been increasing somewhat and there is dyspnea on exertion and easy fatiguability. The patient can walk a block slowly without dyspnea. There is marked clubbing of the fingers and suffusion of the conjunctivae. Some examiners reported no cardiac murmurs; others a short, soft systolic murmur just inside the left nipple line. The EKG showed a definite left axis deviation on two occasions. The roentgen findings were as follows: marked prominence of the left ventricle with absence of the pulmonic arch on the left side of the silhouette in the postero-anterior view. The pulmonary vessels, both the major hilar branches and the more peripheral branches, are diminished in size. All of the findings were regarded as suggestive of a tricuspid atresia and the patient is scheduled to have exploration for the Blalock procedure in the near future.

Isolated Pulmonic Stenosis[14,42]

Isolated pulmonic stenosis is one of the rarer of the congenital cardiac malformations. While the stenosis may either be at the pulmonic valve or in the pulmonic conus below the valve, it is much more unusual to have conus stenosis in the isolated form of the anomaly than when it is combined with other anomalies as in the tetralogy of Fallot where conus stenosis is extremely common. The occurrence of the stenosis at the valve site is apparently responsible for one of the roentgenologic findings which to some extent distinguishes this condition, namely, bulging of the lower portion of the pulmonary artery segment as a result of the conus enlargement. There may also be post-stenotic dilatation of the pulmonary artery which further increases the size of the pulmonary artery segment. These cases ordinarily do not show cyanosis early and those cases without an associated patent foramen ovale ordinarily show cyanosis only terminally. There is usually pallor and there may be mild cyanosis, apparently due to the slowing of the circulation with increased oxygen utilization in the systemic capillaries. A common clinical finding is dyspnea out of proportion to the cyanosis. With a patent foramen ovale there may be cyanosis due to reversal of the shunt, with venous blood passing from the right atrium into the left atrium.

Roentgenologically there is usually evidence of marked right ventricular enlargement and there may be evidence of right atrial enlargement as well. The prominence of the pulmonary artery segment is a common finding, as already mentioned. The pulmonary artery branches usually are small but the phenomenon of arterialization of the lungs by way of the bronchial arteries may cause some increase in the finer vascular pattern in the lungs.

The following cases illustrate pulmonic stenosis with and without interatrial septal defect.

___, U.H., male, aged 12. Congenital heart disease had first been diagnosed at the age of three years. There was exertional dyspnea limiting the capacity for exercise and there had been orthopnea since birth. History of recurrent winter cough each year. Examination showed slight cyanosis of the lips, ears and finger tips. The liver was two fingerbreadths below the costal margin and there was pulsation of the neck veins. The heart was enlarged with a systolic apical thrill and systolic apical murmur. The murmur was widely transmitted. EKG showed right axis deviation.

Roentgen Findings: Very marked enlargement of the heart with a globular silhouette extending predominantly into the left chest. A lateral view demonstrates that this enlargement is predominantly right-sided with marked filling out in the retrosternal region and no significant posterior extension. No evidence of enlargement of the left atrium. The pulmonary artery segment is moderately full above the protrusion representing the enlarged right ventricle. The pulmonary arteries and the lung fields are small in comparison with the marked cardiac enlargement. Roentgenologically a

diagnosis of pulmonic stenosis was entertained with the suggestion of probable additional cardiac anomalies including an interatrial septal defect.

The patient died the following year in another hospital. The pathologic findings with respect to the heart were isolated pulmonic stenosis and marked right ventricular hypertrophy.

____, U.H., a 22 year old white female, had had mild exertional dyspnea since infancy which had become marked in the three months prior to her first visit to the Outpatient Department. There was a history of cyanosis of the lips and finger tips on exposure to cold or marked exertion. There had been several episodes of precordial pain without radiation prior to coming to this Clinic.

Physical examination showed a fairly well developed white female with marked cyanosis of the lips and extremities and clubbing. The heart was enlarged to percussion and there was a loud systolic murmur heard over the entire precordium, loudest at the third left interspace 2 cm. lateral from the sternum. P_2 was greater than A_2. EKG showed a right bundle branch block. A year prior to admission here the patient had had an attack of what was diagnosed as spinal meningitis at another hospital from which she made a slow convalescence.

The patient was admitted to the hospital four days after her first visit to the Clinic with the clinical findings of meningitis, a positive blood culture, and a high white count in the spinal fluid. Patient's condition rapidly deteriorated and she expired two days after admission.

Roentgen Findings: Marked cardiac enlargement predominantly of the right ventricle with filling out of the retrosternal zone but on posterior projection of the cardiac silhouette in the lateral view. The waistline of the heart on the left side is filled out, with a straignt pulmonary artery segment. The pulmonary artery branches in the hilar regions and in the periphery are small. There was a small air cyst or bulla of the apex of the left lung.

Post-mortem exanination of the heart showed marked hypertrophy and dilatation of the right ventricle and dilatation of the right auricle. The left ventricle appeared normal in size. There was marked pulmonary stenosis with only a small diaphragm type of valve remaining with an opening so small that only the smallest probe would pass. There was a patent forament ovale approximately 4 cm. in diameter. A small patent ductus arteriosus was present which would admit only the tip of the fine probe. The post-mortem examination also showed a brain abscess and purulent meningitis.

Interatrial Septal Defect[1,34,43]

This is one of the common congenital anomalies which permit survival into adult life and very frequently individuals with this lesion lead surprisingly normal lives considering the usual marked cardiac enlargement until some terminal episode supervenes. The characteristic findings are marked dilatation and hypertrophy of the right heart with enlargement of the pulmonary artery and branches. There is characteristically marked disproportion between the size of the pulmonary artery and the aorta with the aorta being hypoplastic. A very frequent combination is an acquired mitral valvular lesion, the combination then being knwon as Lutembacker's syndrome. In this condition the most tremendous dilatation of the pulmonary artery occurs, so great that some of these cases have been operated upon with the suspicion of mediastinal and hilar masses. Taussig points out that the presence of the atrial septal defect may relieve the strain on the left atrium so that it is not enlarged or not as prominent as would be expected with the degree of mitral stenosis in the Lutembacher's cases.

Differentiation from patent ductus arteriosus roentgenologically which also gives enlargement of the pulmonary arteries and increased pulmonary vessel pulsations, is aided by the lack of left ventricular enlargement in atrial septal defect and the small size of the aorta in this lesion.

Isolated Interventricular Septal Defect

This malformation ordinarily produces no change in the cardiac silhouette. There may rarely be evidence of right ventricular and pulmonary artery enlargement.

Truncus Arteriosus

This anomaly is a rare cause of cyanotic congenital heart disease in childhood and young adult life. Most of the cases die in the neonatal period. In this anomaly a single large trunk is given off from the left and right ventricles and from it the pulmonary arteries arise in some cases, and if they do not the lungs are supplied by the bronchial arteries. With the single large arterial trunk overriding the interventricular septum, there is a defect in the membranous upper portion of the septum so that the arterial trunk receives blood from both left and right ventricles. Other additional anomalies are frequently present.

The roentgen findings may vary widely and no very characteristic appearance can be anticipated. Taussig[42] has described several cases in which there is marked right ventricular enlargement producing a shelf-like appearance of the heart in the left anterior oblique view in the retrosternal region as a result of the absence of the normal pulmonary trunk and the narrowness of the vascular pedicle. Danelius[15] has called attention to the absence of the normal hilar "comma" in this condition, but as Taussig points out, this may also occur in any condition in which the pulmonary arteries are deficient or absent, as in pulmonary atresia.

Our two cases, the histories of which are given below, showed nothing in common as far as the cardiac silhouette is concerned, but both showed evidence suggesting in retrospect an atypical origin for the pulmonary artery branches and evidence of this sort may be of some help in recognizing these cases. These cases may be aided by the Blalock procedure if a pulmonary artery adequate for the anastomosis exists.

____, U.H., male, aged 7 years. The child was admitted to the hospital because of left hemiplegia complicating cyanotic congenital heart disease. The child developed fairly normally until 14 months of age, at which time cyanosis appeared and the diagnosis of congenital heart disease was made. Cyanosis continued from that time with development of clubbing of the fingers and toes. After the age of three years he was unable to walk more than one or two blocks without squatting. Ten days prior to admission the patient developed fever with headache and facial twitching and subsequently paralysis of the left arm and leg. The positive physical finding with respect to the heart was a very unusual high pitched continuous whistling sound in the left upper chest transmitted well to the back bilaterally, but not to the right anterior chest. The clinical impression was cerebral thrombosis complicating congenital heart disease.

Roentgen Findings: Rather peculiar double contour of the left upper mediastimun with a marked buldge in the region of the aortic knob. Deep pulmonic notch with no evidence of the pulmonary trunk contour. The hilar pulmonary artery branches, however, are normal in size although somewhat atypical in appearance. Fluoroscopic examination suggested right ventricular enlargement and there is some suggestion of elevation of the apex of the heart which appears slightly enlarged in its transverse diameter.

The patient's condition progressed downhill with development of s left facial paralysis. Neurologic examination indicated a suppurative encephalitis or brain abscess. A blood culture was positive for staphylococcus. The patient expired four days after admission.

Examination of the heart showed marked hypertrophy of the right ventricle with a persistent truncus arteriosus overriding a defect in the membranous portion of the interventricular septum. The truncus showed three valve cusps. About four to five cms. from the valve the pulmonary arteries were given off. The post-mortem examination also showed

a brain abscess and purulent meningitis.

...., U.H., 16 year old female, admitted to the hospital on October 16, 1946, with a history of normal development until the age of two when she developed persistent cyanosis following an episode of pneumonia. Activity had not been limited, however, Two years before admission she had had an episode of subacute bacterial endocarditis treated at another hospital successfully with penicillin with no recurrence of fever subsequently. The patient had become more dyspneic and orthopneic in recent months. Examination showed a coarse systolic thrill palpable over the entire precordium and a harsh systolic murmur heard best at the lower end of the sternum which also transmitted over the entire precordium. There was a diastolic murmur localized over the pulmonic area. The lungs were hyperresonant throughout and numerous moist rales were present in both lungs.

The patient progressed downhill despite treatment and died two weeks after admission.

Roentgen Findings: A very bizarre cardiac silhouette with tremendous cardiac enlargement apparently involving both right and left ventricles. There was a tremendous enlargement in the pulmonary trunk region with no recognizatble aortic arch or knob shadow which could be differentiated from the pulmonary artery. The pulmonary arteries in the lungs appeared somewhat atypical but were somewhat larger than usual. Part of the peculiarity of the appearance of the pulmonary arteries is attributable to large air cysts which were recognizable particularly in the right base, causing some compression of the upper portion of the right lung.

Post-mortem examination showed marked hypertrophy and dilatation of the heart which weighed 690 grams. There was a persistent truncus arteriosus with three valve cusps. The aorta and the pulmonary artery branched off the common truncus at a point 1.5 cm. above the valve leaflets. The pulmonary artery was very large, measuring 8 cm. in circumference at its origin from the truncus. There was hypertrophy and dilatation of both left and right ventricles with dilatation of the right auricle, the left auricle being very small in comparison. There was thickening of the truncal, mitral and tricuspid valve leaflets apparently due to an old hyaline rheumatic lesion. A patent foramen ovale 1.5 cm. in diameter was present. The membranous portion of the interventricular septum was absent. Several large emphysematous blebs were found in the right lower lobe and an apparent localized pneumothorax in the right base.

Coarctation of the Aorta

The third type of congenital cardiac anomaly for which surgical correction is now available is coarctation of the aorta. Gross[21] and Crafoord[13,14] have developed the surgical technique for excising the narrowed segment of the isthmus of the aorta distal to the origin of the left subclavian artery. This is one of the congenital anomalies of the heart and great vessels in which the roentgen examination is often highly specific and it may suggest this diagnosis where it has not even been suspected on the basis of a clinical examination. However, in almost every case in which the diagnosis is definite from the roentgen examination obvious clinical findings are present when they are looked for, i.e., evidence of thrills and bruits over the upper posterior thorax indicative of the collateral circulation and reduced blood pressure in the lower extremities.

The well known specific roentgenologic finding, notching or scalloping of the inferior aspects of the ribs, was described by Roesler[33] and by Railsback and Dock[31], apparently independently at about the same time. This rib notching is due to tortuosity of the enlarged intercostal vessels which carry the collateral circulation back to the aorta below the level of the construction. Bramwell and Jones[6] recently presented a very thorough post-mortem injection study of a case of coarctation and have shown very well how arterial loops in the

tortuous intercostals produce the rib notching, it thus being unnecessary to invoke the presence of small aneurysms of these vessels to explain the notches. Pereiras and Castellanos[28] have also recently demonstrated in vivo the tortuosity of the intercostals by the superior retrograde injection of the carotid artery with diodrast. This finding be calls a "secondary sign" of coarctation of the aorta.

The left ventricular hypertrophy and enlargement which occurs in this condition accompanying the hypertension in the upper part of the body is well known. Another roentgen finding which has been commonly described is the absence or inconspicuousness of the aortic knob. It has been difficult to explain this latter finding in the past in view of the fact that one would ordinarily expect the aortic arch to be somewhat enlarged in view of the hypertension and indeed very commonly the ascending aorta may be more prominent than normal in the presence of coarctation. Gladnikoff[12,19] has recently discussed some of the roentgen features of this condition in the light of several of the early cases operated upon by Crafoord. He feels that the lack of visibility of the aortic knob in the postero-anterior view and the decreased visibility of the aortic arch which is commonly observed in the left anterior oblique view is accounted for by shortening of the aortic arch which counteracts the tendency to elongation caused by the hypertension. The shortening is believed to be due to shortening of the ligamentum arteriosum displacing the aorta somewhat medially toward the mediastinum, and also shortening of the stenotic segment of the aorta. The arotic knob is thus pulled medially into the mediastinal shadow and not infrequently the only vascular projection above it along the left mediastinal contour will be formed by the left subclavian artery which is commonly dilated. The dilated origin of this vessel may closely stimulate the aortic knob. The low level of the arotic arch can frequently be recognized by the caudad displacement of the aortic arch impresseon on the left side of the esophagus.

A review of twelve cases of coarctation of the aorta in which roentgen studies done at the University of Minnesota Hospitals were available showed among these cases only one female, which is illustrative of the great predominance of males in this condition. The cases ranged in age from seven years to 67 years at the time the first x-ray examination was made here. All the patients showed evidence of left ventricular hypertrophy with variation from little or no increase in the transverse diameter of the heart to marked left ventricular enlargement. All but three of the cases showed obvious rib notching. In one of these, a boy of seven years, notching was suggested, but very questionable. The other two patients without rib notching were 11 years and 37 years of age respectively.

Three of our cases showed what was regarded as an essentially normal aortic knob with no indication from the roentgen studies available that this represented the dilated origin of the left subclavian artery. In eight cases, however, there was good roentgen evidence of enlargement of the left subclavian artery. Two of these cases showed a knob-like prominence in the postero-anterior view from which the aortic arch could be fairly well distinguished and it is presumed that this protrusion represents a localized dilatation of the proximal portion of the enlarged left subclavian artery. Six other cases showed a widening of the left upper mediastinal contour, but without localized protrusion. These cases are also believed to represent enlargement of the brachio-cephalic vessels, particularly the left subclavian artery. All the cases in which the left anterior oblique view was available for review showed an absence of the aortic window with no recognizable aortic arch. Gladnikoff, as has been mentioned above, suggests that this is due to the downward retraction and medial displacement of the aorta due to its shortening, with the result that its shadow blends in with that of the pulmonary artery so that no window is recognizable.

Three of our cases in which films were available with barium in the esophagus in the postero-anterior view showed a low aortic indentation very suggestive of the low situation of the aortic arch. The fourth case in which films were available with barium in the esophagus showed no evidence of an aortic indentation of the esophagus in the postero-anterior view. Five of the cases showed a double contour of the left mediastinum indicating aortic arch and descending aorta differentiated from the vascular shadow above, formed presumably principally by the left subclavian.

From the foregoing it will be seen that it is of considerable practical importance that the possibility that dilatation of the proximal portion of the left subclavian may simulate a prominent aortic knob in coarctation of the aorta be generally understood. It is of particular importance where rib notching is absent or is very minimal in which case coarctation might be excluded as a diagnostic possibility from the roentgenologic point of view owing to the apparent prominence of the aortic knob.

Despite the helpfulness of these diagnostic points in connection with conventional roentgenologic examination in coarctation of the aorta this method fails to provide accurate anatomical descriptive information of the lesion. (Our attempts at demonstrating the narrowed segment at the isthmus by planigraphy in several cases have proved unseccessful.) This can only be supplied by radio-opacification of the arterial structures involved. This can be accomplished by angiography by the method of Robb and Steinberg[20] in which case it may be difficult to secure adequate opacification of the aorta. Superior retrograde arteriography with retrograde injection of diodrast into the carotid artery has been advocated by Pereiras and Costellanos with demonstration of the intercostal collateral circulation, as mentioned above. Recently Broden, Hanson and Karnell[8], in Sweden, have suggested and employed in some cases catheterization of the radial artery with introduction of the catheter into the aorta very much in the same fashion that right heart catheterization is carried out through the veins. Diodrast is then injected and the involved segment of aorta may be demonstrated. This method permits, as do the other methods of opacification, demonstration of the enlarged arterial branches making up the collateral circulation especially the enlarged internal mammary arteries and the intercostals.

Anomalies of the Great Vessels

The fourth class of cardiovascular malformations for which surgical relief has recently become available is the group of anomalies of the great vessels. These abnormalities produce symptoms, not through their effect on the cardiovascular system itself, but because of compression of the esophagus and trachea with resulting dysphagia and stridor. The principal anomalies in this group are right aortic arch, left-sided origin of the right subclavian artery, and double aortic arch.

Left-sided origin of the right subclavian is the most frequent of the anomalies of the great vessels[26,37]. In a number of autopsy series the incidence of this anomaly has varied from .4 to 2 per cent. A review of the literature in 1945, showed that up until that time only five cases had been diagnosed roentgenologically and only one of these had been proved at autopsy. The rapidity with which experience with anomalies of this sort can be obtained in centers where vascular surgery is actively being pursued is indicated by the fact that Brean and Neuhauser[7] in December 1947, reported on a series of 15 cases in whom the diagnosis of aberrant right subclavian artery had been made. In three of these cases there was concomitant patency of the ductus arteriosus.

This anomaly may be entirely asymptomatic or it may lead to varying degrees of feeding diffuculty and dysphagia. This dysphagia was long ago termed dysphagia lusoria by Bayford of England. The term is today often used for any dysphagia attributible to compression of the esophagus by one of the great vessels.

The characteristic roentgenologic feature is a localized compression often

running obliquely from below on the left to above on the right across the posterior aspect of the esophagus just above the level of the aortic arch. It is also somewhat better visualized in the left anterior oblique view, but in practically all cases a porterior defect is recognizable. The author has had the opportunity of studying three cases, all of them observed as incidental findings in the course of routine gastro-intestinal or cardiac fluoroscopy. In all of these there was a defin te posterior defect slightly above the level of the arch of the aorta and in two the defect showed some obliquity. Two of the cases were observed in the course of gastro-intestinal examinations at the University of Minnesota Hospitals and the other case was observed elsewhere and has been reported previously[37].

Gross[22] has reported successful division of the anomalous vessel with subsequent relief of dysphagia.

Embryologically this anomaly results from persistence of the distal portion of the right fourth branchial arch which ordinarily disappears. The esophageal impression may be considerably larger than that expected from a normal sized left subclavina artery and in these cases is probably attributible to a diverticulum-like persistence of the old right aortic root.

____ U.H., male, six weeks of age. Developed cyanotic spells beginning 24 hours following birth on feeding. Because of these recurrent cyanotic spells coincident with feeding, he was admitted to another hospital where a chest x-ray suggested patchy atelectasis. Laryngoscopic examination indicated a tracheitis and absence of the arytenoid cartilages. Gastrostomy was resorted to for feeding on the sixth day and following that there was some improvement. Occasional episodes of cyanosis with regurgitation of feeding persisted and the infant was transferred to the University of Minnesota Hospitals. There was a mass in the left parietal region with the characteristics of a cephalhematoma. An esophagogram performed with lipiodol failed to demonstrate any compression of the esophagus. Barium injected into the stomach by gastrostomy tube was regurgitated into the esophagus and there was some aspiration into the bronchial tree. Patchy infiltration of the lungs followed thought to be due to aspiration pneumonia. The condition of the patient progressed downhill because of the pneumonia and death occurred three weeks after admission in the sixth week of life.

Post-mortem examination revealed no abnormalities of the heart, but an anomalous right subclavian artery was found arising from the posterior portion of the arch of the aorta in the region where the left subclavian artery is normally given off. The vessel passed posterior to the esophagus and trachea and gave off the vertebral artery on the right side. There was no innominate artery, but in its place the right common carotid artery is found and it forms a ring with the anomalous subclavian artery. The left common carotid artery was in its usual position.

Persistent Right Aortic Arch[2]

For practical purposes the persistent right aortic arch falls into two general categories. The first is the isolated variety occurring without any other evidence of heart disease and commonly found as an incidental finding in the x-ray examination of the adult. In these cases the aortic arch produces an indentation of the right side of the esophagus in the postero-anterior view and displaces the esophagus forward as visualized in the right anterior oblique or lateral view. This anterior displacement may be due either to a crossing of the esophagus from right to left at this level, or, frequently, to a diverticulum-like outpouching of the aorta at this level to which either the left subclavian artery or the ligamentum arteriosus may be attached. In the latter cases the aorta remains to the right of the midline and crosses at a lower level to pass through the diaphragm normally on the left side. These cases are only rarely associated with symptoms and when they develop they are

usually the result of widening and tortuosity of the aorta associated with age, resulting in esophageal and rarely tracheal compression. Variations in the first type of aortic arch in which a posterior defect in the esophagus is visualized have been described. In one of these the left subclanian artery produces a defect above the aortic arch defect as it passes from left to right. Neuhauser points out that in the rare instances when symptoms are produced in this type of aortic arch a vascular ring may be formed by the pulmonary artery and the ductus arteriosus or ligamentum arteriosus with the pulmonary artery then being pulled back against the anterior aspect of the trachea by the ductus with production of compression of the trachea visible in the lateral view. The common carotid may produce pressure on the trachea as it passes from right to left.

The second type of right aortic arch is commonly associated with the tetralogy of Fallot. There is displacement of the esophagus to the left at the level of the arch of the aorta, but no anterior displacement, the aorta descending on the right and crossing over at a lower level. As has been noted above, nine cases of this type of right-sided aortic arch were found in the series of 42 cases of the tetralogy submitted to surgery in this hospital in which the roentgenologic findings were reviewed. It is very important for the surgeon to know the side of the aortic arch and also to be aware of any possible additional anomalies of the great vessels insofar as these can be demonstrated roentgenologically, since such considerations may alter the surgical approach.

Double Aortic Arch

No cases of this variety have been diagnosed roentgenologically at the University of Minnesota Hospitals, but Neuhauser[26] presented illustrations of four such cases, two of them operated upon by Gross. When symptoms are present these patients usually present stridor which is worse on feeding and mild dysphagia. In all of Nuehauser's cases there was a rounded mass displacing the esophagus forward at the level of the aortic arch and representing the larger persistent right-sided aortic arch. In the postero-anterior view there is narrowing of the esophagus from both the right and the left, presumably due to pressure from the two sides of the vascular ring. Visualization of the trachea by means of lipiodol instillation shows similar deformities to those described for the esophagus. In the cases operated upon by Gross the anterior left arch was smaller than the posterior right, as was anticipated from the roentgenologic findings.

Summary

1. A review has been presented of the methods and results of the roentgenologic examination of cases of congenital anomalies of the heart and great vessels at the University of Minnesota Hospitals in recent years.

2. The results indicate that the conventional roentgenologic studies together with the complete clinical findings can lead to a proper diagnosis in a high percentage of cases, particularly in those for which surgical correction is now available.

3. It is highly desirable that these conventional methods be supplemented by angiocardiography and cardiac catheterization to permit diagnosis of the more complicated and obscure cases.

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III. MEDICAL SCHOOL NEWS

Visitors To The Campus

Dr. Paul C. Hodges, Professor or Radiology, University of Chicago, visited our Radiology Department this week. During his visit to Minneapolis he delivered the Russell D. Carman Memorial Lecture at the 95th annual meeting of the Minnesota State Medical Association. His subject was "The Role of X-Ray Pelvimetry in Obstetrics." He also spoke to the Minnesota Radiological Society.

Dr. Russell J. Blattner, Department of Pediatrics, Baylor University, Houston, Texas visited the Medical School this week. He addressed the Minnesota Medical Association on the subject, "St. Louis Encephalitis." Other guest speakers from outside of Minnesota included Doctors Dean McAllister Lierle, Professor of Otolaryngology, Iowa City and Michael L. Mason, Associate Professor of Surgery, Northwestern University Medical School

Staff Activities

Dr. O. H. Wangensteen, Dr. Richard Varco and Dr. Clarence Dennis have just returned from Quebec, Canada where they attended the meeting of the American Surgical Association. Dr. Dennis presented a paper, "Vagotomy in Ulcerative Colitis." Co-authors in this work which was presented by Dr. Dennis are Dr. Frank Eddy, Dr. Austin McCarthy, Dr. Darrell Westover and Dr. Howard Fryckman.

Dr. Cecil J. Watson will be in Chicago next week to take part in the examination given by the American Board of Internal Medicine. He will also give the Chairman's address of the Section on Internal Medicine of the American Medical Association.

Dr. David Seibel returned to the Department of Obstetrics and Gynecology on June 7 commencing his activities as a fellow in that department. He served as an intern in this hospital in 1945-46.

Continuation Course in Otolaryngology

The Sixth Biannual Continuation Course in Otolaryngology will be held on June 28-29-30 and July 1-2. This course is designed to bring to the practicing Otolaryngologist the newer concepts and develpoments in the specialty. The course will be under the direction of Dr. Lawrence Boies and associates of the University Medical School. Dr. Harold I. Lillie, Dr. Gordon B. New and others of the Graduage School faculty will participate in the instruction. Guests lecturers will include Dr. Kenneth Day, Pittsburgh; Dr. Gordon Hoople, Syracuse; Dr. C. Steward Nash, Rochester, N. Y.; Dr. Francis LaJeune, New Orleans. Fee for the course is $35.00. The enrollment i is limited. Application should be made at an early date to the Director, Center for Continuation Study, University of Minnesota.

The present number of the Bulletin concludes the series for the academic year 1947-48. The editorial staff extends best wishes to all the members of the staff of the Medical School, the Minnesota Medical Foundation and all the friends of the Medical School who are readers of the Bulletin. We look forward to October 1, 1948 when the weekly staff meetings and publication of the Bulletin will be resumed.

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