THE SURGICAL TREATMENT OF TRANSPOSITION OF THE PULMONARY VEINS*

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ANOMALOUS DRAINAGE of the pulmonary veins into the right side of the heart has been thought to occur rarely. Brody[1] reported that the literature before 1942 disclosed 106 cases. In each of these the condition was discovered at postmortem examination. Cases diagnosed during life have been reported since the addition of angiocardiography and cardiac catheterization to routine procedures for the diagnosis of congenital cardiovascular deformities. In 1949, Dotter, Hardisty and Steinberg[2] found reports of 27 cases in the literature, in addition to those reviewed by Brody in 1942. To these they added two cases which they had diagnosed with the aid of angiocardiography and cardiac catheterization. More recently Friedlich, Bing and Blount[3] reported 18 cases in which pulmonary veins drained partially or totally into the right side of the heart.

In this report we are presenting a case of complete transposition of the pulmonary veins and a discussion of a surgical procedure employed in the treatment of this condition.

The developmental aspects of this anomaly are not completely understood. Brody,[1] in his excellent review of available information, reported general agreement among investigators that the initial position of the opening of the common pulmonary stem in the dorsal wall of the sinus venosus in some laboratory animals is central and that this stem shifts to the left when the interauricular septum forms. It has been postulated that the shift of this pulmonary stem is due to formation of the "pulmonary fold." This fold is an invagination of the right wall of the common pulmonary trunk into the sinus venosus. Its developmental function is to divide the primitive atrium into a right and left part. It has been suggested further that the anomalous drainage of pulmonary veins ino the right atrium is the result of the failure of the pulmonary fold to develop properly. This hypothesis, however, as well as various others, fails to explain the communication of some pulmonary veins with tributaries of the right side of the heart.

In the development of human pulmonary veins, the pulmonary venous plexus is assumed to be derived from the splanchnic venous plexus. The common pulmonary vein forms from the pulmonary venous plexus and joins the left auricle directly. The four pulmonary veins are thought to arise as vessels which drain different parts of the lung bed and form a common trunk which enters the dorsal aspect of the left atrium. As the heart continues to grow, this vessel is gradually absorbed into the wall of the auricle until its four original branches open separately into this chamber when development is complete.

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According to a theory proposed by Patten, anomalous connections of the pulmonary veins originate in extremely early embryonic stages in which the developing foregut, trachea and lung beds are supplied by a common plexus of small channels. These channels pass in all directions through the mesenchyma and join the primitive cardinal veins in many places. When certain of the channels enlarge to form pulmonary veins leading to the left atrium, the primitive connections with the cardinal veins usually disappear. Patten considered that the unusually strong development of one of these early channels and the persistence of its embryonic venous connection are responsible for the formation of these abnormal communications. The unusually well-developed channels would then explain the wide variation in points of drainage which the pulmonary veins may have. Brantigan[4] stated that abnormal sites of drainage of pulmonary blood on first consideration would seem unpredictable but that their existence might be considered reasonable from an embryonic point of view. The blood will always drain into some structure which originates from the primitive venous system.

Partial anomalous drainage of the pulmonary veins occurs more frequently than total anomalous drainage into the right side of the heart or into some tributary thereof. The latter condition was present in 37 of the 106 cases reported by Brody, the ratio being approximately two to one. The right pulmonary veins are about twice as frequently affected as the left. The superior vena cava is the most common point of anomalous entry. Other points of entry in order of frequency are the right auricle and the left innominate vein. Less commonly, anomalous pulmonary veins drain into the coronary sinus, the inferior vena cava, the hepatic veins, the azygous vein, the left subclavian vein, and the anomalous left superior vena cava.

The usual anatomic structure of the completely transposed pulmonary venous system is as follows: The superior and inferior pulmonary veins on each side form a common trunk. This trunk in turn unites with a similar trunk on the opposite side to form a common vessel, which enters the right auricle or one of its tributaries (Fig. 1).

Partial drainage of the pulmonary veins into the right side of the heart is entirely compatible with life. Several patients in Brody's series were in the seventh and eighth decades of life, and one was in the ninth decade. Total anomalous drainage, however, is not tolerated so well. In all but eight of the 37 cases of total anomalous drainage which were reported by Brody, death occurred before the age of six months. The oldest patient of this group was 27. Brody suggesed that life may be possible, at least until adulthood, so long as less than half the pulmonary veins are shunted into the right side of the heart.

Hughes and Rumore[5] devised a method of calculating the amount of shunted blood by comparing the actual cross-sectional area of the anomalous vessel with that of the pulmonary veins draining normally into the left auricle. They reported two cases in which the shunted blood was calculated by this method and found that 26.1 and 20.1 per cent of oxygenated blood respectively were returned to the right side of the heart. They believed, as Brody had previously suggested, that decompensation ensues only when more than 50 per cent of the pulmonary venous blood drains into the right side of the heart.

Partial transposition is likely to be associated with a patent foramen ovale, and complete transposition obviously must be accompanied by a patent foramen ovale or by a common auricle if life is to exist. Other major congenital cardiac anomalies frequently co-exist with pulmonary transposition.

The diagnostic features of these anomalies will be reported in a separate publication by Dr. George C. Griffith[6] and his coworkers.

A review of the literature has revealed no case in which successful surgical treatment has been accomplished for transposed pulmonary veins. In the series reported by Friedlich, Bing, and Blunt two cases of complete transposition were operated upon unsuccessfully.

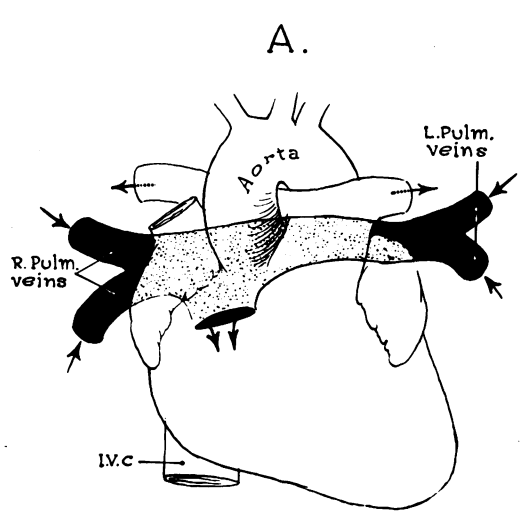


FIG. 1.—Diagrammatic illustration of the usual anatomic configuration of the completely transposed pulmonary venous system.

REPORT OF CASES

We have operated on two patients with this deformity. The diagnostic studies in both of these cases were performed by Dr. George C. Griffith, Dr. Richard Cosby, Dr. Willard Zinn, and Dr. David Levinson.

Case 1. The first patient was a 4-year-old white female who was admitted to the Los Angeles County Harbor General Hospital on April 5, 1950, with a chief complaint of heart trouble since birth. Shortly after her birth the parents were told that the patient had a heart murmur. This murmur has persisted until the present time. She has had frequent upper respiratory infections and several episodes of pneumonitis. These infections have been accompanied by cyanosis and congestive heart failure, which necessitated hospitalization. Her exercise tolerance was diminished so that she could walk only about one block before becoming fatigued. On each admission, bedrest, digitalis, and other cardiac medications alleviated her cardiac failure. At first the episodes of failure were infrequent, but during the past year or so they have occurred every 6 weeks to 2 months.

Before her admission on April 5, she had been treated in the Cardiac Clinic at the Los Angeles County General Hospital, where she underwent cardiac catheterization. During the procedure the catheter entered a pulmonary vein (Fig. 3). Following the catheterization, angiocardiography disclosed an anomalous inferior vena cava, which entered the right auricle superiorly.

Physical examination showed that the patient was a moderately well-developed, poorly-nourished child who appeared chronically ill. Her mucous membranes were slightly dusky. There was a bulging of the anterior aspect of the chest on the left. No thrills could be felt. There was a blowing systolic apical and basal murmur, as well as a diastolic apical murmur. Percussion showed the heart to be enlarged on both the right and the left. Fine crepitant râles could be heard at the base of both lungs. The liver edge was about 6 cm. below the right costal border. It did not seem to pulsate. The spleen was not palpable. There was moderate clubbing of the fingers and toes and faint cyanosis of the nailbeds.

The red blood cell count was 4.8 million, the hemoglobin 13 Gm., and the hematocrit 39. The white blood cell count was 7000, and the differential count was essentially normal. The oxygen content of the blood in the right auricle, right ventricle, and femoral artery was slightly over 14 vols. per 100, a finding which supported the diagnosis of complete transposition of the pulmonary veins.

The electrocardiogram showed a sinus rhythm with bradycardia, the rate being approximately 58 to 60, and there was marked right axis deviation. Roentgen-ray and fluoroscopic findings revealed considerable generalized cardiac enlargement displacing the esophagus posteriorly and to the right in a wide arc. There was considerable prominence of the left upper cardiac border, presumably due in part to the pulmonary vascular enlargement. There was enlargement of the hilar vessels with vigorous expansile pulsations (Fig. 2). The films confirmed the fluoroscopic findings. A diagnosis of complete transposition of the pulmonary veins was made.

Before operating upon this patient we were fortunate in having an opportunity to discuss this problem briefly with Dr. Alfred Blalock,[7] and

this discussion was extremely valuable to us in planning the operative procedure which we used.

On May 16, 1950, an exploratory thoracotomy was carried out through a posterolateral incision. A large portion of the fifth rib was removed, and the chest was entered through this rib bed. The apical segment of the lower lobe was found to be adherent to the parietal pleura. These adhesions were divided, and the hilus of the lung was exposed. What appeared to be a large superior pulmonary vein could be easily seen (Fig. 4). The phrenic nerve and pericardiophrenic vessels were dissected from the pericardium over a distance of several centimeters in the region of the pulmonary hilus. About 10 cc. of 2 per cent Novocain was placed in the pericardial cavity. The pulmonary vein was then dissected free from the hilus; it was found to drain both lobes of the lung. The pericardium was entered. It contained about 30 cc. of straw-colored fluid. The large pulmonary vein did not enter the pericardium in the usual manner and did not drain normally into the left auricle. It appeared to pass between the layers of the pericardium over to the right side. It was traced as far as possible, but, because of fear of displacing the heart and initiating severe arrhythmia or cardiac arrest, it could not be traced to its entry into the right side of the heart. The pulmonary artery was then freed for a short distance and an umbilical tape passed about it. It was approximately three times its normal size. Temporary ligatures were then placed on the pulmonary vein distally, and a Blalock partial occlusion clamp was placed on the left pulmonary vein as far medially as possible. A similar clamp was placed on the left auricular appendage, and its tip was excised. A large transverse incision was then made in the pulmonary vein, and an anastomosis was carried out between the end of the auricular appendage and the side of the pulmonary vein (Fig. 5). The patency of the anastomosis was at least 1 cm. in diameter. The clamps were then removed from the various structures, and the temporarily placed ligatures were likewise removed. Although a thrill could not be felt, the anastomosis appeared to be widely patent. The pericardium was closed loosely. A de Pezzer catheter was placed in the mid-axillary line in the seventh intercostal space. Five hundred thousand units of penicillin were placed in the chest cavity, and the chest was closed in layers with silk sutures. With the exception of an occasional extra systole, the patient withstood the procedure well and was returned to the ward in good condition.

Her postoperative course was entirely uneventful. She was discharged from the hospital on May 30, 1950, at which time she seemed improved, although it was too early to make any definite conclusions. Her progress was followed in the out-

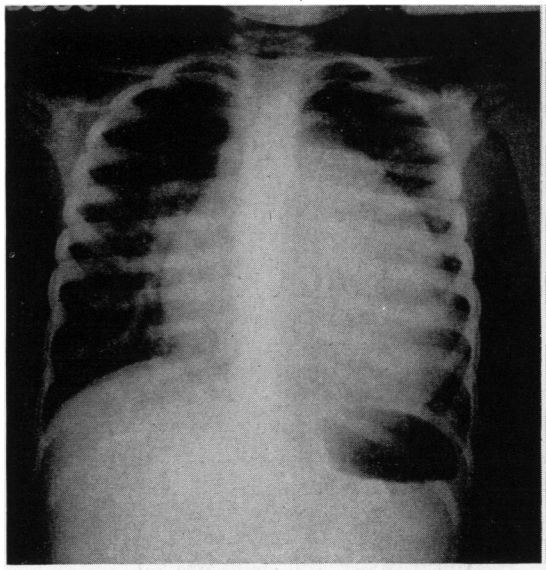


FIG. 2

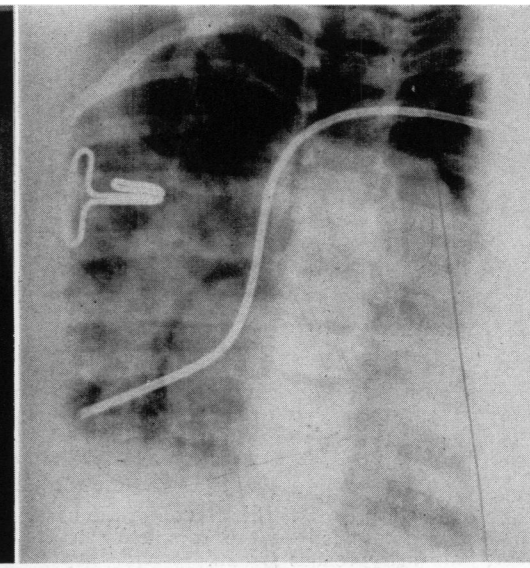
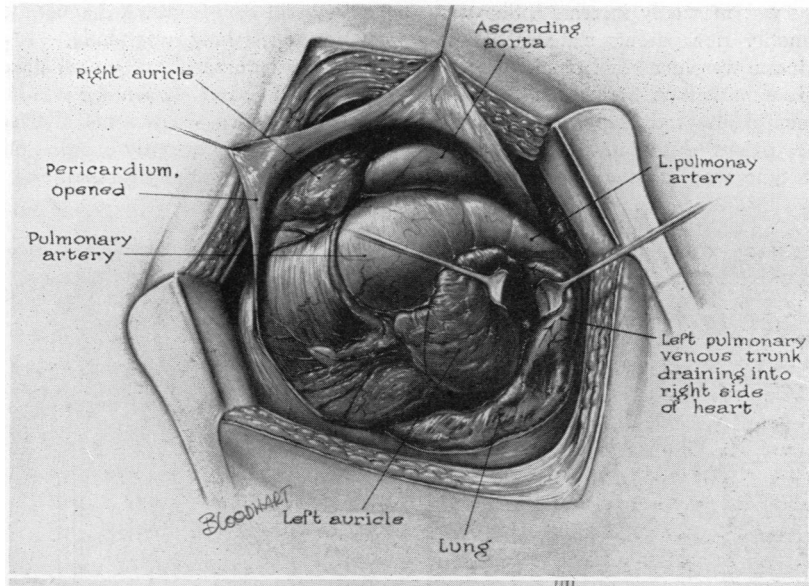


FIG. 3

FIG. 2.—Roentgen-ray of the chest of the first patient with complete transposition. The picture shows generalized cardiac enlargement and prominent vascular lung field markings.

FIG. 3.—Roentgen-ray of chest taken during cardiac catheterization of first patient. The catheter is shown entering the right lung field directly from the right auricle.

FIG. 4



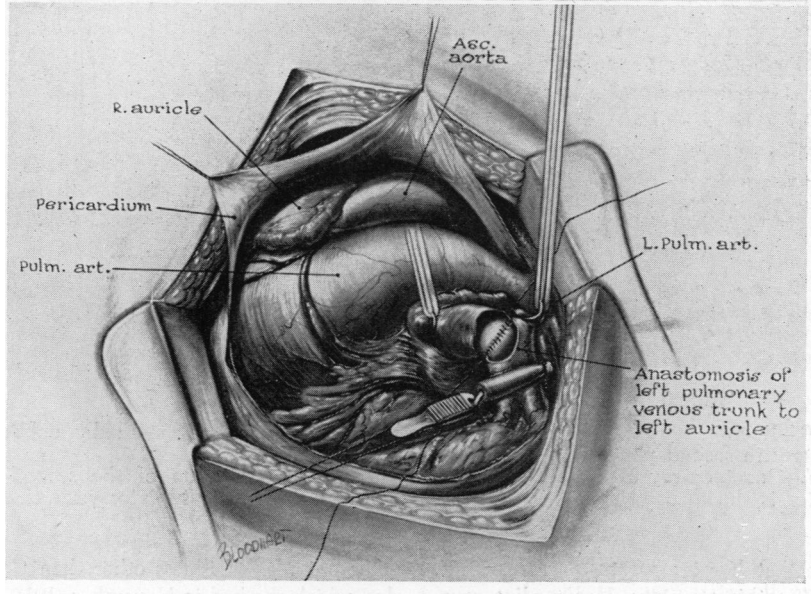


FIG. 5

FIG. 4.—Drawing of heart of first patient showing operative findings as seen through the posterolateral incision in the bed of the fifth rib. The entire pulmonary venous blood flow of the left lung is through a common venous trunk which passes along the posterior pericardium to the right auricle or one of its tributaries.

FIG. 5.—Drawing of operative procedure performed on first patient. The end of the left auricular appendage has been excised, and an anastomosis is being carried out between the end of the left auricular appendage and the side of the left common venous trunk.

patient department and, when seen on several occasions, she was thought to be improved. Her exercise tolerance was markedly increased. She was no longer cyanotic. Her parents stated that she could play as if she were a normal child.

She was re-admitted on September 9, 1950, with a chief complaint of difficulty in breathing and fever of two days' duration. Physical examination at that time was essentially the same as before except that her temperature was 102° F. and she was breathing rapidly. Her cardiac murmurs were still present, and there was an area of dullness at the left base posteriorly. Râles could be heard in both lung fields posteriorly. Roentgenograms revealed a grossly enlarged heart and an extremely prominent pulmonary segment of the left cardiac border. Vascular markings were obscured in the left lung root but were prominent in the right lung root. There was a homogeneous density in the lower portion of the left hemithorax, which was thought to be due to a small pleural effusion. The patient was given aureomycin, and her digitalis was increased to the point of intolerance. On this medication she improved and was discharged from the hospital on September 28, 1950, without complaints. A diagnosis of bronchial pneumonia, left lung, was made.

She has progressed very well since discharge, and on April 5, 1951, when she was last seen, her parents stated that she was not allowed to run as much as she had previously but that otherwise her activities were essentially normal. She has had no other episodes of illness other than German measles, which she had in November, 1950. No further cyanosis has been noted.

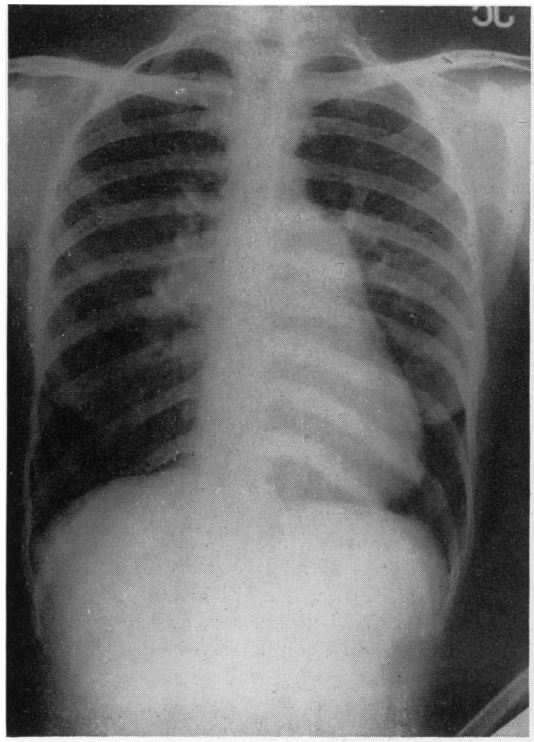


FIG. 6

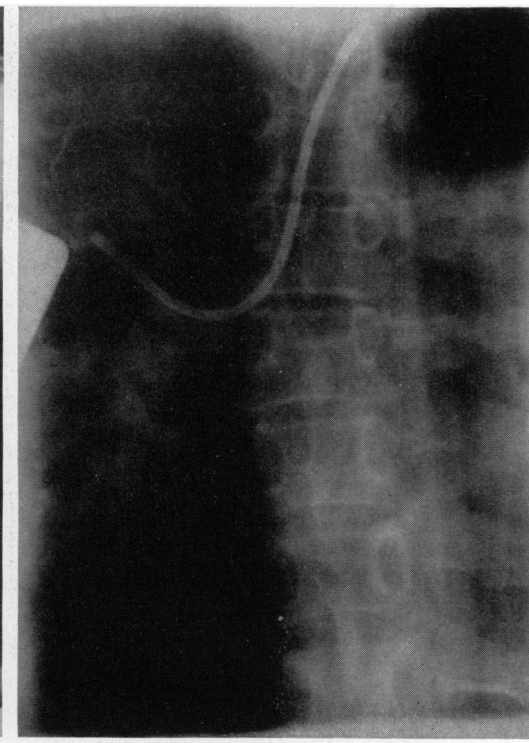


FIG. 7

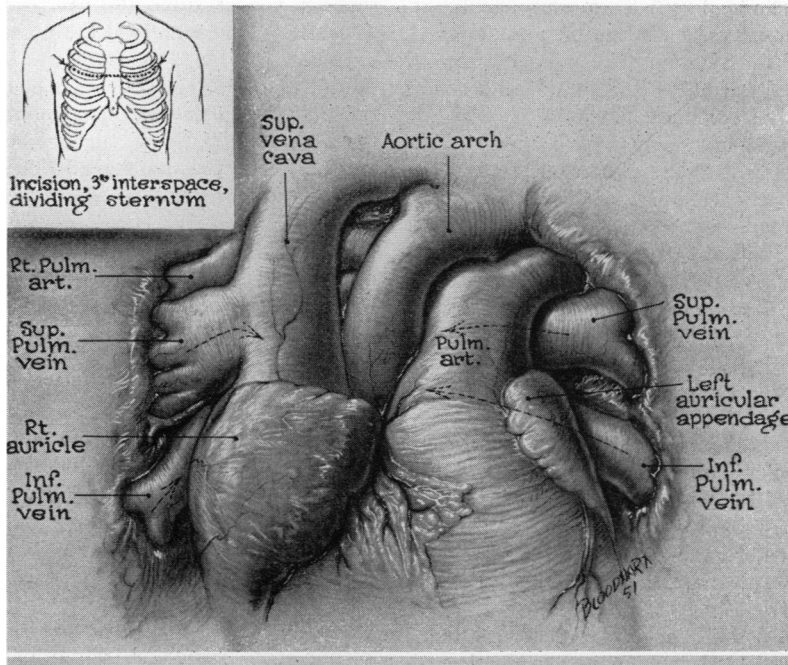
FIG. 6.—Roentgen-ray of chest showing cardiac enlargement and markedly enlarged pulmonary artery in second patient.

FIG. 7.—Roentgen-ray of chest taken during cardiac catheterization of the second patient. The catheter is shown entering the right lung field directly from the superior vena cava.

Case 2. The second patient was a 25-year-old white woman who was initially thought to have a partial transposition of the pulmonary veins. At the age of 6 she was found to have a heart murmur, but she had no symptoms at that time. Cyanosis had never been present. During the past 4 years she has had exertional dyspnea which progressed to the point that it occurred on even slight exertion.

Physical examination revealed that the patient was poorly developed and poorly nourished. Per-

FIG. 8



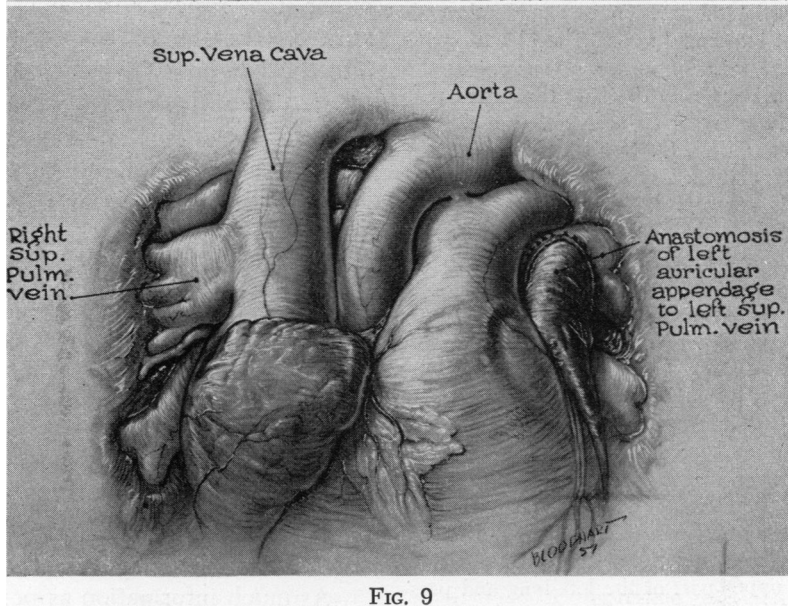


FIG. 9

FIG. 8.—Drawing of heart of second patient which shows operative findings as seen through an incision transecting the sternum and entering both sides of the chest through the third intercostal space. A large right superior pulmonary vein enters the superior vena cava, and a smaller inferior vein enters the right auricle. A large superior pulmonary vein and a smaller inferior one on the left enter the right auricle posteriorly. The right auricle and auricular appendage are markedly enlarged while those on the left are small.

FIG. 9.—Drawing of the operative procedure performed on the second patient. An anastomosis has been carried out between the end of the right auricular appendage and the side of the anomalous left superior pulmonary vein.

cussion showed the heart to be enlarged on the left. A diastolic and systolic murmur could be heard over the second and third intercostal spaces to the left of the sternum. The pulmonic second sound was loud and snapping. The blood pressure was 100/60 and the pulse rate 80.

Roentgen-ray and fluoroscopic examination revealed enlargement of the right ventricle and the pulmonary artery (Fig. 6). Prominent vascular markings were present in the lung fields. Angiocardiography showed tremendous dilatation of the pulmonary artery. During cardiac catheterization the catheter passed directly from the superior vena cava into the right lung field (Fig. 7). It was visualized in 3 positions and was thought to have entered either three pulmonary veins or smaller tributaries of a large vein.

The red blood cell count was 4.1 million with a hemoglobin of 12 Gm. The white blood cell count was 14,700 with a normal differential. Because of the progressive nature of her symptoms and the degree of cardiac enlargement, it was decided to attempt surgical correction of the deformity.

On April 2, 1951, an exploration was carried out. An anterolateral incision was made through the third right intercostal space. The pleural space was obliterated by adhesions. Upon exposing the hilar region, a large vein draining the upper part of the right lung could be seen entering the superior vena cava (Fig. 8). It was decided to free the right pulmonary artery first. After some difficulty this was accomplished, and the vessel was found to be several times its normal size and to have a markedly increased pressure in it. An inferior pulmonary vein was also present, but one could not be sure whether or not it drained normally into the left auricle. The sternum was divided transversely, and the incision was extended well into the left side of the chest. The pericardium was then divided transversely. The aorta was small, and the pulmonary artery was several times its normal size. The right auricular appendage was enlarged, but the left auricular appendage was smaller than normal and was located far to the posterior. A large superior vein could be seen draining the upper part of the left lung and passing posteriorly into what appeared to be the right auricle (Fig. 8). Just before the large vein entered, a smaller vein from the lower part of the left lung joined it. A probe could be passed from the upper vein on the left across into the lower vein on the right. As these findings revealed that the transposition was essentially complete, it was, therefore, decided to anastomose the left auricular appendage to the superior venous trunk on the left. Thus the end of the auricular appendage was excised and anastomosed to the side of the left superior venous trunk (Fig. 9).

The patient withstood the procedure well except that her blood pressure was somewhat low during the time that both sides of the chest were open. Although she responded promptly after the operation, her pulse remained from 130 to 160 and her blood pressure from 70/60 to 85/60. Her immediate postoperative course appeared to be entirely satisfactory. However, she expired suddenly about 10 hours after the completion of the operation. Permission for a postmortem examination could not be obtained.

DISCUSSION

In partial transposition the diagnosis may be extremely difficult, while in complete transposition it may present fewer problems. We believe that transposition of the veins on the right side is present if during cardiac catheterization the catheter passes directly into the right lung field from either the superior vena cava or right auricle. Passage of the catheter directly into the left lung field does not confirm a diagnosis of transposed pulmonary veins because the catheter may have passed through a patent foramen ovale into a normal left pulmonary vein.

Complete transposition may be suspected when the oxygen content of the blood in the right auricle, right ventricle, and a peripheral artery are approximately the same and when other laboratory studies and clinical findings support such a diagnosis. Angiocardiography may be of value in demonstrating other deformities such as septal defects. This procedure, however, does not usually disclose the anomalous veins themselves.

As much information as possible should be obtained regarding the anatomic structure of the anomalous pulmonary veins before performing an operative procedure for the correction of the deformity. If anomalous venous drainage occurs only on the left, the anatomic relationship of the left auricle to the pulmonary veins on that side makes an anastomosis feasible be-

tween these two structures. When complete anomalous drainage occurs, the same relationship is present between the left anomalous venous trunk and the left auricle, and a similar anastomosis may be carried out. At a subsequent exploration on the opposite side one might be able to close the common point of anomalous entry and direct the entire pulmonary venous return to the left auricle, as Gerbode[9] has suggested. If the condition of the patient permitted it, one might be able to carry out the procedure in one stage through an incision which would divide the sternum transversely and enter the pleural cavity on either side. When the veins are transposed only on the right, a more difficult problem arises. At the present time we feel that the initial exploration should be performed on the left regardless of which veins are thought to be transposed, because one cannot be sure of the venous configuration on the left. If transposition of the veins on the right is demonstrated before the operation, it would seem that at least partial transposition of those on the left must be present to produce severe symptoms. An anastomosis between anomalous right pulmonary veins and the posterior aspect of the left auricle does not seem technically feasible. A homologous graft of either vein or aorta would probably not remain patent in a venous system. A vein graft from the patient's own internal jugular, azygos, or saphenous vein might be utilized, or the defect might be bridged with a tube made of pericardium or parietal pleura. When only one vein enters the right auricle or its tributaries, surgical intervention is not necessary; however, if one transposed vein should be encountered during a left thorocotomy which is being performed for another reason, it should be anastomosed to the left auricle if the patient's condition permits.

It is difficult to predict what long-term benefits a patient with complete transposition of the pulmonary veins might derive from the procedure outlined above. It seems probable that the duration of life of such a patient might be similar to that of a patient with a large atrial septal defect. One would hope that the change produced in the auricular pressures from the reduced venous return to the right auricle and the concomitant increased venous return to the left auricle would cause the patent foramen ovale to become smaller. A patient with partial transposition who has undergone an operation which has directed all of the venous return to the left auricle might expect a normal duration of life.

Although transposition of the pulmonary veins has been considered a rare anomaly, an increased awareness of its occurrence and the availability of improved diagnostic adjuncts have promoted its more frequent diagnosis during the life of the patient. Certain other congenital cardiac lesions which were thought to be rare were discovered more frequently when interest was directed toward them. Cases of transposed pulmonary veins may likewise be detected more frequently as awareness of the existence of such anomalies increases and as the available special diagnostic technics are applied to patients in whom the presence of the deformity is suspected.

SUMMARY

We have been extremely interested in the problem of the treatment of transposed pulmonary veins and especially in devising methods to correct them. Thus in anticipation of a greater need for effective methods of treating patients with these anomalies, we have suggested certain surgical procedures which we feel might benefit such patients. These procedures are based upon our experience and observations in the two cases reported in this communication. As more cases of transposed pulmonary veins are diagnosed during the life of the patient, it will probably be necessary to modify the procedures we have mentioned or to devise

other operations for the correction of these abnormalities.

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DR. FRANK GERBODE, San Francisco: We have been interested in these anomalies for a number of years, and although we have not as yet operated upon a patient with one, we have studied a small number of patients, and contemplate doing some of these operations in the near future.

A number of years ago, with procedures such as these in mind, we tested the feasibility of anastomosing vessels to the atria; altogether, we did about 75 such shunting procedures. We believed that these atriovenous shunts might find a place not only in the treatment of congenital abnormal venous return to the heart but, also, in the treatment of certain types of superior venacaval obstruction.

In general, we found that small vein-to-auricle shunts invariably became fibrosed and shut off. In another series of about 25 animals, aorta homografts were used to bridge a gap between the superior venacava and the auricle. Thrombosis or fibrosis occurred, in time, in every instance. This demonstrated that arterial pressure and a free flow of blood are necessary to maintain the lumen in an arterial homograft.

On the other hand, large vein-to-atria shunts remained patent and functional in more than 80 per cent of instances. We learned that one could not crush the atrium with an occluding clamp during the suture, because thrombosis then occurred. One has to use narrow-bladed clamps which just approximate and hold the wall.

Furthermore, it was found necessary to evert a generous cuff of atrial wall so that no cut muscle protruded into the lumen. I will show you some of these experimental results.

[Slide] This is a diagram similar to the one shown by Dr. Miller. We suggested, following our dissections and studies of these patients, that one might anastomose the left pulmonary vein as an initial procedure, as done in Dr. Muller's and Dr. Longmire's case, using a side-biting clamp, so that the flow of blood to the right side would not be entirely shut off.

We also suggested that perhaps a side-to-side anastomosis, as they conducted, might be feasible. In all these dissections, these veins lay very close to the left auricular appendage.

[Slide] This is one of our experimental procedures in which, in order to obtain a large vein, we used the superior venacava, and anastomosed it to the end of the left auricular appendage. We did this because we found, in the animal, that the pulmonary veins were too small to stay open satisfactorily.

[Slide] This, then, produced a byproduct of the investigation, that of a cyanotic dog, because here we are mixing venous blood with arterial blood, and the oxygen saturation was not complete. This is an angiograph of one of those shunts, showing the blood coming into the superior venacava, the left side of the heart, and directly out of the aorta, showing that the anastomosis is patent.

[Slide] This is one of those shunts after a year. You can see, where the clamp has been placed, a little fibrosis occurs. This is a clean anastomosis, widely patent, and just as large as the vein which is anastomosed to the heart.

[Slide] Then we attempted to transpose the inferior venacava from the right side of the heart to the left, and did a series of experiments in which this atriacaval shunt was performed, likewise trying to test the feasibility of anastomosing a large vein to the left atrium.