Two Cases of Total Anomalous Pulmonary Venous Return of the Supracardiac Type with Stenosis Simulating Infradiaphragmatic Drainage

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Total anomalous pulmonary venous return has been classified into four types: cardiac, supracardiac, infracardiac, and mixed, depending on the route by which blood is conveyed by the anomalous vein to the right side of the heart. A simplified classification has recently been suggested based on the known physiologic effect of infradiaphragmatic drainage, namely pulmonary venous obstruction. On this basis only two types of drainage are given, the supradiaphragmatic, without obstruction, and the infradiaphragmatic with obstruction. The association of obstruction with infradiaphragmatic drainage appears to be constant, as pulmonary veins entering below the diaphragm join either the ductus venosus or portal vein, and in both cases the venous blood will meet hindrance to flow.

The statement has been made that total anomalous pulmonary drainage below the diaphragm forms a distinct clinical entity with characteristic x-ray findings. Although all instances of infradiaphragmatic drainage have been associated with pulmonary vein obstruction, there may also be obstruction in some patients belonging to other groups. We have recently seen two patients who presented with symptoms of such obstruction and were found at autopsy to have stenosis of an anomalous vein at the supracardiac level.

Case Reports

Case 1
A 3-day-old white male infant was admitted to the Babies Hospital on August 16, 1960, because of cyanosis and bilateral pneumothorax.

The infant was born to a 28-year-old, gravida II, para 1, woman after an uncomplicated full-term pregnancy. At birth he was cyanotic, and chest x-rays taken within the first 2 days showed bilateral pneumothorax. Chest tubes were inserted and later chest films showed expanded lungs. The infant could not tolerate oral feedings and vomited brownish mucoid material several times before admission.

Physical examination showed a well-developed infant who was cyanotic and had substernal retractions. His breath sounds were poor, and ronchi were heard throughout both lung fields. The heart rate was 120 per minute and regular. A grade-II, low-pitched systolic murmur was best heard at the apex with the infant lying on his back. The pulmonic second sound was louder than the aortic second sound. The liver was 2.5 to 3.0 cm. below the right costal margin; the spleen was not palpable. The remainder of the physical examination was normal.

Chest x-rays (fig. 1) taken on the third and fourth days of life in lateral and frontal projections revealed a partial pneumothorax on the right side. A thoracostomy tube was present on the left and the left lung appeared expanded. The heart was of normal size. Each lung showed severe reticular densities suggesting a marked degree of pulmonary vascular congestion with pulmonary edema. A film taken 24 hours later, revealed reexpansion of the right lung. The appearance of marked pulmonary venous congestion was again noted.

An electrocardiogram consisting of six extremity leads and three precordial leads (V1, V2, and V3) was obtained on the fourth day of life. There were normal sinus rhythm, rate 125, a P-R interval of 0.12 second, and right axis deviation. The P wave in lead II measured 1 mm. The right precordial leads showed tall R waves, and small S waves associated with upright T waves. The electrocardiogram was considered within normal limits for the age of the patient. The upright T waves were equivocal evidence of abnormality. Respiratory distress and cyanosis continued.
ANOMALOUS PULMONARY VENOUS RETURN

Cultures of the nose, throat, and blood were negative. The infant was treated with penicillin and chloromycetin. His condition continued to deteriorate and he died on the second hospital day at 5 days of age. Digitalis had not been administered. The clinical diagnosis was congenital heart disease, pneumonitis, and question of sepsis.

Postmortem examination: The infant was well developed and poorly nourished, weighed 2450 Gm., and measured 46 cm. in length. The lips and nailbeds were cyanotic. There was no peripheral edema or effusion into the body cavities.

The heart was normally shaped and not enlarged; it weighed approximately 13 Gm. The right ventricle formed two thirds of the anterior surface of the heart and the apex was made up of both right and left ventricles. The right atrium and ventricle were dilated and hypertrophied, and had a capacity three or four times that of the left heart. The right atrium received the dilated superior vena cava, the inferior vena cava, and the coronary sinus. The foramen ovale measured 1.0 cm., the septum primum was absent. The right ventricle measured 3 mm. in thickness near the septum posteriorly. The tricuspid and pulmonary valves were normal and measured 3.6 cm. and 2.1 cm. respectively. No pulmonary veins entered the small left atrium. The mitral and aortic valves measured 2.7 cm. and 1.2 cm. in circumference respectively. The ductus arteriosus was patent and measured 1.1 cm. in circumference. The pulmonary veins joined posterior to the pericardium to form a common pulmonary vein (fig. 2). From the left superior aspect of this common chamber an anomalous vein arose and passed superiorly; it measured 1.1 cm. in circumference. One-half centimeter from its origin, the vein passed posterior to the left pulmonary artery, between it and the left main stem bronchus. At this point there was marked narrowing of its lumen to a circumference of 0.3 cm. The vein continued anteriorly and then upward as a persistent left superior vena cava, and joined the left subclavian and internal jugular veins to form the innominate vein. Between the point of constriction and the entrance into the innominate vein there was an aneurysmal dilatation of the vessel that measured 1.2 cm. in circumference.

The common pulmonary chamber from which the anomalous vein arose had a saccular dilatation on its superior surface measuring 0.8 by 0.7 cm.; this ended blindly on the posterior wall of the left atrium within 2 mm. of the atrial septum and 8 mm. above the coronary sinus.

The lungs showed congestion, patchy atelectasis, interstitial edema, and prominent, dilated, blood-filled lymphatics in the interlobular septa.

Figure 1

Case 1. Chest x-ray, age 4 days; showing a diffuse homogeneous infiltrate in both lung fields and residual pneumothorax on the right.

Figure 2

Case 1. Total anomalous pulmonary venous return. Left. The common pulmonary vein gives off a main trunk which is constricted between the left pulmonary artery and left main stem bronchus. The anomalous vessel passes anteriorly and forms a saccular dilatation before joining the innominate vein. Right. View with the heart elevated, showing the relation of the anomalous vein to the heart. A small saccular dilatation arising from the common pulmonary chamber ends blindly on the wall of the left atrium above the coronary sinus. The heart is normal in size.
Rubella in the first trimester. Although the infant appeared vigorous and normal at birth, 6 to 8 hours later she developed tachypnea.

On admission she was well developed and well nourished and had generalized moderate cyanosis. The pulse was 140 per minute and the respirations were 44 per minute with mild substernal retractions. A grade-III systolic murmur was heard along the left sternal border, loudest in the fourth left interspace without significant radiation. The second pulmonic sound was louder than the second aortic sound. The liver was palpable 3 to 4 cm. below the right costal margin, and the tip of the spleen was just felt.

Films of the chest on the day of admission (fig. 3) showed a heart of normal size. The vascular pattern in the lung was markedly abnormal, with prominent venous congestion. The diagnosis of anomalous pulmonary venous drainage below the diaphragm was suggested by the radiologist.

An electrocardiogram (fig. 4) was obtained on the fifth day of life, prior to digitalization. There were normal sinus rhythm, heart rate of 166 per minute, P-R interval of 0.10 second, and right axis deviation. The P wave in lead II measured 1.5 mm. The T waves were inverted in leads II, aV$_R$, V$_5$, and V$_6$. The precordial tracings showed tall R waves in right-sided leads and deep S waves in left-sided leads. The electrocardiogram was considered probably abnormal for the age of the patient, showing evidence of right ventricular enlargement.

On the second hospital day (age 6 days) the infant was considered to be in congestive failure, and digitalis was administered. That evening the infant was noted to tire during feedings. The following day she had an acute episode of cyanosis, restlessness, and sweating during a feeding. Penicillin therapy was begun. The infant appeared to be doing well until the fifth hospital day when the liver increased in size, pretibial edema developed, and the respiratory rate rose to 102 per minute. She died suddenly the same afternoon at the age of 9 days. The clinical diagnosis was congenital heart disease, cyanotic, probably total anomalous pulmonary return below the diaphragm.

Postmortem examination: The infant weighed 3,540 Gm. and measured 47 cm. in length. The lips and fingertips were cyanotic, and there was generalized subcutaneous edema. The right pleural cavity contained 15 ml. and the left 30 ml. of serosanguineous fluid. The mesentery and retroperitoneal tissues were edematous, and the peritoneal cavity contained 15 ml. of free serous fluid.

The heart was normal in contour and weighed 20 Gm. Approximately 75 per cent of the anterior surface was occupied by the right ventricle, and the apex consisted of both right and left ven-

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Case 2

A 5-day-old female infant was admitted to the Babies Hospital on October 7, 1960, because of poor color and rapid respiration since 6 to 8 hours of age.

The infant was born to a 36-year-old, gravida IV, para III, woman whose pregnancy was uncomplicated. There was a history of exposure to

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Figure 3

Case 2. Chest x-ray, age 5 days; showing the diffuse homogeneous infiltration of the lungs similar to that seen in case 1. The heart is normal in size.
Figure 4

Case 2. Electrocardiogram (see text for description). Standard and unipolar limb leads at half standardization. Precordial leads at half standardization.

cases

tricus. The right atrium and ventricle were hypertrophied and dilated, whereas the left atrium and ventricle were hypoplastic. The left heart was estimated to have less than 25 per cent of the volume of the right heart. The greatly dilated right atrium received the superior and inferior venae cavae and the coronary sinus. The superior vena cava was dilated and had an unusually wide orifice. The foramen ovale was partially covered by the septum primum but admitted a probe of 1.2 cm. in diameter. Both the trabecular muscles and the crista supraventricularis were hypertrophied. The tricuspid and pulmonary valves were normal and measured 4.5 cm. and 2.3 cm. respectively. No pulmonary veins entered the left atrium. The mitral and aortic valves were normal and measured 3.5 cm. and 1.5 cm. respectively. The ductus arteriosus was patent and had a circumference of 0.6 cm.

The pulmonary veins from each lung joined posterior to the pericardium to form a common pulmonary vein (fig. 5). A large vein arose from the left superior wall of this chamber and almost immediately gave off a small branch to be described later. The main anomalous vein measured 1.0 cm. in circumference at its origin. Six millimeters distal to its origin it passed between the left pulmonary artery and the left main stem bronchus and was narrowed to 3 mm. in circumference at this point. It then passed upward to join the innominate vein. Between its emergence from behind the pulmonary artery and its entrance into the innominate vein, there was an aneurysmal dilatation of the vessel that

Figure 5

Case 2. Left. Anterior view of the great vessels. The main anomalous pulmonary vein with its aneurysmal dilatation is shown. In addition, a smaller anomalous vein courses diagonally from the common pulmonary vein to the right superior vena cava. Right. View of the specimen from the left side shows the marked narrowing of the anomalous pulmonary vein as it passes between the left pulmonary artery and left main stem bronchus.
Figure 6
Case 2. Postmortem angiogram. Contrast media outlines the aneurysmal dilatation of the persistent left superior vena cava, the constricted segment, and pulmonary veins. The right atrium and portal system are filled through an accessory anomalous vein.

measured 2.0 cm. in circumference. The small branch given off the main anomalous vein near its origin measured 2 mm. in circumference and passed diagonally upward and posterior to the pulmonary artery and ascending aorta to empty into the superior vena cava below the entrance of the innominate vein and at the level of the orifice of the azygos.

The lungs were strikingly congested and many lobules were outlined by large dilated lymphatics containing blood. Microscopically the interlobular veins as well as lymphatics were congested and alveolar walls were thickened because of edema and capillary engorgement. The liver, spleen, kidneys, and gastrointestinal tract were congested. No other malformations or other significant changes were present.

A postmortem angiogram is shown in figure 6. A clamp has been placed on the innominate vein and the left jugular and left subclavian veins have been tied. A needle is inserted into the aneurysmal dilatation of the persistent left superior vena cava and contrast material injected. This has outlined the dilatation, the constricted segment, and the pulmonary veins. In addition, the accessory anomalous vein leading from the main anomalous trunk is seen. Through it, the contrast media has spread to the right superior vena cava, azygos vein, right atrium, and portal system.

Anatomic diagnoses: Total anomalous pulmonary venous return, supracardia, via two routes: A. Anomalous vein posterior to left pulmonary artery, leading to left superior vena cava and left innominate vein; B. Accessory anomalous vein leading from "A" to the right superior vena cava; stenosis of anomalous vein "A," localized, between left pulmonary artery and left main stem bronchus; poststenotic dilatation of left superior vena cava; patent foramen ovale; patent ductus arteriosus; congestion and edema of lungs; congestion of abdominal viscera; anasarca.

Discussion
Harris, Neuhauer, and Giedion mentioned two patients in their group who, by clinical and x-ray signs, were thought to have infra-diaphragmatic drainage, but at autopsy were
found to have supracardiac drainage. Since the complete autopsy findings were not given, it is not clear whether there was stenosis in the supracardiac circuit as in our cases.

Only one case similar to ours has been found in the literature. Didion (case 2) described a 4-month-old boy who had dyspnea beginning in the third week of life. He developed cyanosis on slight provocation and died suddenly with bronchopneumonia. On postmortem examination, a common pulmonary vein 3.2 cm. in circumference was seen to pass between the left pulmonary artery and the left main-stem bronchus. Distal to this, it entered a saccular dilatation that joined the innominate vein. No other anomalies of the heart or vessels were described.

Appreciation of the existence of a group of cases with obstruction at the supracardiac level, has led us to review the literature in search of anatomic variants that occur in pulmonary venous return via the persistent left superior vena cava.

Illustrations from specimens usually show the left superior vena cava anterior to the left pulmonary artery. Occasionally, a diagrammatic sketch or a reconstruction from an angiocardiogram indicates the vein posterior to the left pulmonary artery. Though the latter arrangement may prove to be more common than is now apparent, in the great majority of cases the anomalous vein lies anterior to the left pulmonary artery.

An explanation for the common anterior position of the persistent left superior vena cava is given by the embryologic origin of the veins. Figure 7 shows the embryologic relationships of the splanchnic plexus, cardinal and common pulmonary veins. The forerunner of the left superior vena cava, the left anterior cardinal vein, is always anterior to the left pulmonary artery while only the veins of the splanchnic plexus are posterior to it. Consequently, we believe that a true persistent left superior vena cava is always anterior to the left pulmonary artery. In our cases, the proximal part of the anomalous channel was probably derived from the primitive esophageal plexus and not the anterior caval vein. The point of stenosis, where the left pulmonary artery crosses the anomalous vein, represents the point of communication between the channel derived from the splanchnic plexus and the cephalic portion of the left anterior cardinal vein (fig. 7). Thereafter, the anomalous vein assumes an anterior position as a persistent left superior vena cava.

An additional point of interest is the presence, in case 1, of a dilatation of the common pulmonary vein that is attached by a cord to the left atrium in the area of the embryologic site of budding of the atrial component of the common pulmonary vein. Occlusion of the vein at its exit from the atrial wall at an early stage of development may be inferred.

Summary

Two patients who died during the neonatal period with congestive heart failure and marked pulmonary congestion were found at autopsy to have total anomalous pulmonary venous return of the supracardiac type with stenosis of the pulmonary vein. The narrowing was the result of an unusual course of the vein, which passed between the left pulmonary artery and left main bronchus and was stenosed at this point. It is emphasized that the clinical and x-ray findings were indistinguishable from those of total anomalous pulmonary venous drainage below the diaphragm.

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References


As to your method of work, I have a single bit of advice, which I give with the earnest conviction of its paramount influence in any success which may have attended my efforts in life—Take no thought for the morrow. Live neither in the past nor in the future, but let each day's work absorb your entire energies, and satisfy your widest ambition. That was a singular but very wise answer which Cromwell gave to Bellevire—"No one rises so high as he who knows not whither he is going," and there is much truth in it. The student who is worrying about his future, anxious over the examinations, doubting his fitness for the profession, is certain not to do so well as the man who cares for nothing but the matter in hand, and who knows not whither he is going!—Sir William Osler, Aphorisms From His Bedside Teachings and Writings. Edited by William Bennett Bean, M.D. New York, Henry Schuman, Inc., 1950, p. 71.
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