Surgical Correction of Absence of Proximal Segment of Left Pulmonary Artery

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SUMMARY
In a 14-month-old patient who had been repeatedly hospitalized for recurrent pulmonary infections, cardiac angiography found “absence” of the left pulmonary artery, but thoracic aortography demonstrated that a rudimentary patent ductus filled a distal patent pulmonary artery. There was a coexistent vascular ring with a retroesophageal right subclavian artery.

At surgical exploration it was possible to mobilize the distal left pulmonary artery and perform a direct anastomosis between the left pulmonary artery and the main pulmonary artery. This is perhaps the first patient in whom such an operation has been performed. It is emphasized that the absence of a pulmonary artery on pulmonary angiography does not indicate that the entire pulmonary artery is absent. The presence of a patent and surgically reconstructable distal pulmonary artery can be determined only by aortography or possibly by surgical exploration.

Additional Indexing Words:
Vascular ring Thoracic aortography Selective pulmonary angiography
Retroesophageal subclavian artery

INCREASING NUMBERS of reports of unilateral absence of a pulmonary artery with hypovascular lung field as an isolated congenital anomaly1-8 have appeared. In 1958 Anderson and associates2 advised operation to restore normal pulmonary arterial flow. This is the first report of such an operation.

Report of Case
The patient born on October 3, 1965, was the product of an uncomplicated pregnancy. The infant weighed 7 lb 3 oz at birth. Bilateral club feet, poor muscle tone, and right facial paresis were noted at birth. Weight gain was poor. During the first 3 months of life the child had frequent episodes of respiratory distress and was hospitalized for treatment of pneumonia.

Following 6 weeks of therapy the physical and x-ray findings resolved and he was discharged.

The ensuing 9 months were marked by frequent and severe episodes of respiratory distress that were not associated with feedings. He was treated with antibiotics and expectorants at home. In November 1966 he was again hospitalized for treatment of right middle and lower lobe pneumonia. Clinical response was poor and the resolution of physical and x-ray signs was slow. On November 14, 1966, the patient was admitted to the New York University Hospital (no. 147946) for the first time in a critically ill condition.

On admission his respirations were labored and he was cyanotic. His weight was 16 lb, 12 oz., and his height 30 in. The left hemithorax was smaller than the right. There were marked rhonchi and wheezes in both lung fields. Cardiac sounds were normal.

X-ray examination (fig. 1) revealed an enlarged right hemithorax, an asymmetrically small left hemithorax, and a shift of the heart and mediastinum to the left. The pulmonary vascular markings of the right lung were normal while those of the left lung were markedly diminished.

Electrocardiogram was suggestive of right ventricular hypertrophy. Blood count, urinalysis, and blood chemistries were within normal limits.

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ANOMALY OF LEFT PULMONARY ARTERY

Figure 1
The chest x-ray showed a small left hemithorax with diminished vascular markings and displacement of the cardiac shadow to the left.

Figure 2
An esophageal x-ray with barium shows anterior displacement from a retroesophageal structure that subsequently proved to be a subclavian artery.

Barium swallow showed anterior displacement of the esophagus at the level of the lower part of the trachea (fig. 2). During bronchoscopy on November 23, 1966, expiration and cough caused unusual narrowing of the lower trachea. Bronchography did not show any abnormalities (fig. 3).

On December 6, 1966, right-heart catheterization revealed the pressure in the right ventricle to be 55/0-10 mm Hg. Left atrial sampling, via the foramen ovale, revealed oxygen saturation of 89%, pH 7.28, pO₂ 74, and pCO₂ 54. The elevated pressures were attributed to hypoxia inadvertently induced by general anesthesia. The catheter was easily passed into both the main and

Figure 3
A bronchogram performed because of the recurrent respiratory infections did not show any abnormalities.

Figure 4
Selective angiography of the pulmonary artery showed a normal main and right pulmonary artery but no trace of a left pulmonary artery.
right pulmonary arteries but could not be manipulated into the left pulmonary artery. Injection of contrast material into the main pulmonary artery outlined it and the right pulmonary artery but revealed no evidence of a left pulmonary artery (fig. 4). Subsequent injection into the right ventricle revealed a large main pulmonary artery and right pulmonary artery, but again no left pulmonary artery could be demonstrated.

On December 19, 1966, left-heart catheterization and aortography were performed. The distal left pulmonary artery opacified via a small left ductus (fig. 5). An enlarged aberrant right subclavian artery arose from the descending aorta and passed behind the esophagus at the level of the lower trachea (fig. 6).

Postural drainage and aminophylline only slightly diminished the intensity of the baby’s daily episodes of respiratory distress. The abnormal left pulmonary artery, seen on aortography, was considered to be surgically reconstructable. Also, although x-rays did not conclusively demonstrate a vascular ring compressing the trachea, surgical exploration was planned in the hope of finding and relieving a mechanical constriction.

On December 21, 1966, left posterolateral thoracotomy was performed through the fourth interspace. A 2 by 0.5 cm patch of myxomatous material was present in the anterior wall of the right ventricle, the significance of which could not be determined. The left vagus nerve was dissected and mobilized distally to identify the left recurrent nerve and the ductus arteriosus. The recurrent nerve encircled the ductus arteriosus which arose in the normal location from the aorta just distal to the subclavian artery. Near the aorta the ductus was a cordlike structure about 2 mm in diameter. Mobilization of the ductus disclosed that it coursed distally toward the hilum of the left lung for about 2 cm, where it gradually enlarged into a normal left pulmonary artery a short distance proximal to the origin of the left upper lobe branch. The left pulmonary artery collapsed easily on palpation but was clearly patent.

Dissection of the aorta beyond the ductus exposed a 2 cm aortic diverticulum arising from the right lateral wall of the aorta and passing behind the esophagus. Mobilization of this structure disclosed that a 5 mm right subclavian artery arose to the right of the esophagus and went into the superior mediastinum. A short distance below the origin of the right subclavian artery, a large bronchial artery of similar size originated. These two vessels were ligated, after which the diverticulum was transected and oversewn. The sutured margin of the diverticulum retracted to the right of the esophagus and completely relieved the retroesophageal constriction.

Figure 5
An aortogram performed by injecting dye through a catheter in the aorta opacified a threadlike patent ductus which distally entered the left pulmonary artery (L P A).

Figure 6
Aortogram showing the aberrant right subclavian artery (R S A) which, on surgical exploration, was found to have a retroesophageal location.
The pericardium was then opened and the main pulmonary artery and right pulmonary artery were widely mobilized by dividing their pericardial attachments. There was no trace of the origin of the left pulmonary artery, as the main pulmonary artery had a smooth contour at the site of the origin of the right pulmonary artery. The left pulmonary artery was then widely mobilized distally beyond the branches to the left upper lobe. Although the initial gap between the left pulmonary artery and the main pulmonary artery was between 4 and 5 cm, wide mobilization of these structures made it apparent that a direct anastomosis could be performed. Earlier it was thought that a vascular graft would be required. The left pulmonary artery was accordingly divided beyond the ductus arteriosus and the orifice was enlarged with a longitudinal incision along the margin. A partly occluding clamp was applied to the main pulmonary artery and a short arteriotomy was made. An end-to-side anastomosis was then constructed between the end of the left pulmonary artery and the side of the main pulmonary artery, using interrupted sutures of 6-0 silk (fig. 7). Upon release of the occluding clamp, there was good pulsation in the left pulmonary artery without significant tension on the structures. The inferior pulmonary ligament was divided to displace the lung upward toward the main pulmonary artery.

The subsequent postoperative course was uncomplicated. Fifteen days following operation, right-heart catheterization found the right ventricular pressure to be 28/8 mm Hg. A right ventricular angiogram showed prompt filling of the left pulmonary artery from the main pulmonary artery (fig. 8).

The patient was discharged from the hospital 3 weeks after operation. His subsequent course has been most gratifying. No episodes of respiratory distress have occurred. Wheezing has ceased, and sleep, which had been continually interrupted by paroxysms of coughing, is now normal. Weight gain has been rapid, with a total gain of 6½ lb in the first 7 months following operation. The mother states, “He is a new baby.”

**Figure 7**

Schematic drawing illustrating pathology as seen in cineangiograms and at operation. The drawing at the lower right illustrates the site of ligation of the ductus and the corrective anastomosis accomplished at operation.

*ANOMALY OF LEFT PULMONARY ARTERY*

65
complete absence by carefully performed aortography. The available literature does not clearly indicate what percentage of apparently “absent” pulmonary arteries involve merely absence of a primary division and could be reconstructed surgically.

**Differential Diagnosis**

Differential diagnosis of the asymmetrically small hemithorax requires bronchography, pulmonary angiography, and thoracic aortography. Bronchography can exclude bronchectasis, agenesis, and bronchial obstruction. Pulmonary angiography may be helpful in distinguishing coarctation from absence of the pulmonary artery and can exclude primary emphysema of the contralateral lung.12 The critical decision arises, however, when pulmonary angiography discloses that one of the pulmonary arteries is absent. Whether or not a distal, surgically reconstructable, pulmonary artery is present can be determined only by aortography or by surgical exploration. Aortography should be carefully performed to determine whether a distal pulmonary artery can be opacified through bronchial channels or through a residual ductus arteriosus. Theoretically, a patent but nonvisualized distal pulmonary artery could only be detected by surgical exploration. How often aortography may fail to visualize a distal pulmonary artery is not known at this time.

Hypertrophy of the contralateral lung is always present. It is imperative not to mistake the hypertrophy of the normal lung for obstructive unilateral emphysema.10

**Clinical Course**

The clinical course of absence of a primary division of the pulmonary trunk as an isolated anomaly is variable. The diagnosis of “absent pulmonary artery” has been made in adults who were asymptomatic except for exertional dyspnea.1, 11, 13-16 However, three complicating states have been described.

1. **Pulmonary Hypertension**

Anderson and associates2 reported an autopsied case in which cardiac catheterization at 8 and 13 months of age demonstrated pulmonary

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**Figure 8**

Postoperative selective angiogram of the pulmonary artery following anastomosis of the left pulmonary artery to the side of the main pulmonary artery shows opacification of the left pulmonary artery (L P A).

When last seen, 7 months after operation, the child was normally active and well.

**Discussion**

**Nomenclature**

Cases similar to the one described in this report are reported in the literature under the headings: abnormal transradiancy of one lung,9 unilateral emphysema,10 absence of a pulmonary artery,6 atresia of a pulmonary artery,3 proximal interruption of a pulmonary arch,2 and absence of a primary division of the pulmonary trunk.11 In most of the cases, sufficient data are not reported to permit one to determine whether these are the same or different conditions. Although differential diagnosis will be discussed below, it should be noted that “absence” and “atresia” of a pulmonary artery should be restricted to cases of complete obliteration of the embryological sixth aortic arch, that is, lungs which have only a bronchial arterial supply. Outside of the operating room or the autopsy room, absence of a primary division of the pulmonary artery can best be distinguished from complete absence by carefully performed aortography. The available literature does not clearly indicate what percentage of apparently “absent” pulmonary arteries involve merely absence of a primary division and could be reconstructed surgically.

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1. **Pulmonary Hypertension**

Anderson and associates2 reported an autopsied case in which cardiac catheterization at 8 and 13 months of age demonstrated pulmonary...
hypertension that progressed to systemic levels. Death from congestive heart failure occurred at 14 months of age. The only cardiovascular anomaly was absence of the primary division of the right pulmonary artery. The right pulmonary artery was connected to the right innominate artery by an obliterated ductus. In the collected series of Pool and associates, there were 32 patients with "absent pulmonary artery" as an isolated anomaly. Pulmonary hypertension was present in five of these. Each of the five patients with pulmonary hypertension died by the age of 14 months. Microscopic examination revealed medial hypertrophy in the small pulmonary vessels of three of the five functioning lungs. It was not present in the lungs that lacked a pulmonary artery.

In the present case, preoperative right ventricular pressure was elevated. However, this may have been due to anoxia that was inadvertently caused by general anesthesia. The postoperative right ventricular pressure was normal despite electrocardiographic evidence 2 months postoperatively, suggestive of right ventricular hypertrophy.

2. Respiratory Infection

Pool and associates cited infection as a frequent finding; it was prominent in Macleod's nine cases, and certainly it was significant in our case. However, there is no satisfactory explanation for it. Intra-alveolar carbon dioxide is reduced on the affected side, and this can cause bronchoconstriction. However, the respiratory infections can, as in the present case, arise in the normal as well as the affected lung.

One of the main obstacles to evaluating recurrent infection in cases of "absent pulmonary artery" is distinguishing them from cases of bronchiectasis. Bronchiectasis can result in nonvisualization of a pulmonary artery after right-heart injection. This is presumably due to reversal of flow via bronchial arteries. Alley and co-workers were able to demonstrate a rise in oxygen saturation from the right ventricle to the pulmonary artery in such a case. The difficulty inherent in excluding primary bronchiectasis is illustrated by the case of Swyer and James and is discussed by Swyer and Belcher and associates. Bronchography is necessary to arrive at a clear diagnosis.

In the present case, bronchiectasis was not present. To relate the recurrent episodes of respiratory distress and pulmonary infection to bronchial malfunction, to excessive blood flow through the right lung, or to deficient blood flow through the left lung is conjectural. However, it is certain that respiratory function dramatically improved following surgery.

3. Hemoptysis

In congenital absence of a pulmonary artery bronchial circulation is variable. At one extreme is the unique case in which bronchial circulation was so lacking that gangrene of the lung resulted while at the other extreme are cases in which excessive bronchial flow has resulted in hemoptysis. Experimentally, following pulmonary artery occlusion, marked augmentation of bronchial flow occurs. Clinically, measurements of bronchial flow in these cases have been in the range of 17%, 25%, and 30% of left-heart output. This is in contrast to the normal bronchial flow of 0.5 to 1% of left-heart output.

The anatomy as well as the volume of altered bronchial flow is variable. When bronchial arteries communicate with the alveolar capillary circulation, oxygen uptake is possible. When bronchial arteries bypass the alveolar capillary bed, oxygen uptake is not possible. The occurrence of hemoptysis probably is determined by both the volume and the anatomy of the bronchial flow. The incidence of hemoptysis has been estimated at less than 10%.

Surgery

Reports of surgical intervention in cases of absence of a primary division of the pulmonary artery have been limited to those situations in which there was a coexistent cardiac anomaly. Blalock was the first to report such a case. He anastomosed a right subclavian artery to the patent distal portion of
the right pulmonary artery in a child with tetralogy of Fallot and functional truncus arteriosus. Unfortunately the patient died as the anastomosis was being completed. Tetralogy of Fallot is present in about 20% of cases of “absent pulmonary artery.”

A contralateral patent ductus coexists in 15% of “absent pulmonary arteries.”7 Swan and co-workers25 attributed severe pulmonary hypertension to the contralateral ductus and reported ligation of the ductus in two such cases. Armer and associates26 were the first to report ligation of a ductus contralateral to an “absent” pulmonary artery and successful reconstruction of the “absent” artery. In their case, the “absent” right pulmonary artery arose from the ascending aorta, resulting in a plethoric lung field. The aortic origin of the pulmonary artery was divided and continuity reestablished with the main pulmonary artery with a prosthetic graft. Redo and associates27 reported a similar case.

The patient described in this report is unique in several respects. It may be the first such patient in whom surgical exploration found a patent distal pulmonary artery with atresia of the proximal portion of the artery, which could be surgically corrected by excising the atretic segment and performing a direct anastomosis. The patient is also unique in that the presence of the distal pulmonary artery was established by aortography, which has not been previously reported. Also, it is noteworthy that a large gap of 4 to 5 cm between the main pulmonary artery and the distal pulmonary artery could be overcome by appropriate mobilization of the involved arteries, thus avoiding the use of a vascular graft.

Finally, the etiology of the respiratory malfunction in this patient remains obscure but restoration of normal pulmonary blood flow resulted in immediate subsidence of recurrent respiratory symptoms which had required frequent hospitalization since birth.

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50 Years Ago

Spontaneous Closure of a Ventricular Septal Defect

... Some years ago I saw a small boy, 14 months old, and on examination of the heart there was a very loud systolic bruit, with its maximum intensity over the fourth intercostal space, close to the sternum, but audible also over the whole precordial region, and indeed over most of the chest, both back and front. It was accompanied by a thrill near the sternum, and those of you who have seen congenital heart cases in the wards will realise the kind of case it was when I say that the diagnosis made by myself and others was congenital perforation of the interventricular septum. The father of the boy was an officer in the Navy, and this was his only child. He had always made up his mind that any boy of his should follow him in the service, and it was a tremendous blow to him to find that the child had a bruit which was certain to cause his rejection at the medical examination. At any rate, in my ignorance I gave the opinion that it was useless to think of the boy's being some day eligible for the Navy or indeed for any service in which a medical examination had to be passed. Most clinicians would, I think, have given the same opinion as I did; but it was wrong; and that is why I want you to know about it.

I saw the boy again when he was two years old, and the bruit was about the same as before, very loud and universal. I saw him again when he was five, and there was then absolutely no bruit at all! I saw him again at the age of ten; and he was still without a bruit and had no objective evidence of any heart lesion. The father's keen desire that the boy should follow him in the service has since been gratified and the case has taught me that the bruit of a congenital malformation of the heart may disappear as a child grows up.—HERBERT FRENCH: A Series of Small Points. Three Clinical Lectures. Lecture II. Guy Hosp Gaz 32: 87, 1918.
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