Two Adult Cases of Unilateral Absence of the Right Pulmonary Artery
With Markedly Different Clinical Presentations

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Two adult cases of unilateral absence of the right pulmonary artery with markedly different clinical presentations are reported. One patient was a 21-year-old female without any known history of a coexisting congenital anomaly. An abnormal chest roentgenogram (small right hemithorax, deviation of the mediastinum toward the right side and a dilated left pulmonary artery) was noted and prompted further evaluation. No pulmonary hypertension was noted and the patient remained asymptomatic. The other patient was a 42-year-old male who had unilateral absence of the right pulmonary artery and a peripheral stenosis of the left pulmonary artery. The clinical course of this patient had been complicated by impaired exercise tolerance and occasional hemoptysis since adolescence. At the age of 29 years, a cardiac catheterization revealed pulmonary hypertension, but no left-to-right shunt. Progressive respiratory failure resulted in a premature death at the age of 42 years.

The prognosis of patients with unilateral absence of the pulmonary artery largely depends on the coexisting cardiac anomaly (left-to-right shunt) and pulmonary hypertension. A combination of unilateral absence of the pulmonary artery and contralateral peripheral pulmonary arterial stenosis is very rare, but is an important cause of pulmonary hypertension and gives a worse prognosis for this entity.

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Congenital unilateral absence of the pulmonary artery (UAPA) is an uncommon anomaly that is frequently associated with other congenital cardiovascular diseases. The diagnosis of this disease is usually made during childhood, but the patient may present later in life. The clinical presentation in adult patients with UAPA may vary from coincidental abnormal chest roentgenogram to severe life-threatening symptoms such as congestive heart failure, recurrent pneumonia, massive hemoptysis and so forth. The prognosis largely depends on coexisting cardiovascular anomalies (mainly left-to-right shunt) and pulmonary hypertension. We present here two adult cases of UAPA with markedly different clinical presentations. Special emphasis was placed on coexisting contralateral peripheral pulmonary arterial stenosis as a factor for a poor prognosis.

CASE REPORT

Case 1
A 21-year-old female receptionist was referred to our hospital for further evaluation of an abnormal chest roentgenogram. She had never been aware of any cardiopulmonary symptoms since childhood. On physical examination, she was 150 cm tall and weighed 48.6 kg. Blood pressure was...
120/80 mmHg, respiratory rate was 18/min and pulse rate was 85 beats/min. Auscultation of the heart revealed grade 2/6 systolic ejection-type murmur at the second left sternal border, and a mildly accentuated pulmonary component of the second heart sound. The jugular vein was not distended and there was no peripheral edema. An electrocardiogram was normal. A chest roentgenogram revealed a deviation of the mediastinum toward the right side and a dilated left pulmonary artery (Fig. 1). The volume and vascularity of the right lung appeared to be diminished. The diagnosis of right UAPA was confirmed by a computer tomographic examination of the chest.

A pulmonary function test revealed a vital capacity of 2.59 l (88% of predicted value) and a forced expiratory volume at 1 sec of 2.23 l (84.5%). An arterial blood gas analysis on room air showed PaO$_2$ 83.2 mmHg, PaCO$_2$ 38.8 mmHg and pH 7.42. Hemoglobin was 12.6 g/dl and the hematocrit was 39.3%. Cardiac catheterization revealed aortic pressure, 105/67 mmHg (mean 83), mean pulmonary arterial wedge pressure, 11 mmHg, pulmonary arterial pressure, 32/12 (20) mmHg, mean right atrial pressure, 5 mmHg, and cardiac index, 3.52 L/min per m$^2$. A pulmonary arteriogram revealed the absence of the right pulmonary artery and a dilated left pulmonary artery (Fig. 2). The right lung was supplied by the transeptal collateral circulation via the right internal thoracic artery and the lower right intercostal artery. The bronchial artery was normal. The upper and lower pulmonary veins were visualized at the levophase, and were normal. There was no left-to-right shunt or other congenital heart disease.

Case 2

This patient was a 42-year-old male who had been previously employed as a taxi driver. He was 169 cm tall and weighed 55 kg. His first cardiopulmonary symptom was noted in his adolescence when he became aware of shortness of breath and was easily fatigued. At the ages of 19 and 25 years, he had experienced hemoptysis and was empirically treated for possible tuberculosis. At the age of 29, he presented at our hospital complaining of acute worsening of dyspnea and palpitation shortly after an episode of respiratory infection. In our emergency room, his blood pressure was 120/85 mmHg, pulse rate was 120 beats/min, and respiratory rate was 40/min. Auscultation revealed a widely split second heart sound, S4 gallop and grade 2/6 systolic ejection murmur at the
left sternal border. A chest roentgenogram (Fig. 3) revealed deviation of the mediastinum toward the right side and a markedly enlarged, tortuous left pulmonary artery. An electrocardiogram revealed a sinus rhythm, marked right axis deviation, a tall P wave in leads II, III, and aVF, a tall R wave in V1 and a persistently deep S wave in V5,6, suggesting right ventricular hypertrophy (Fig. 4). An arterial blood gas analysis showed pH 7.47, PaCO2 34 mmHg, PaO2 62 mmHg, and SaO2 93%, while breathing room air. Hemoglobin was 19.7 g/dl and the hematocrit was 58%. An oxygen inhalation test was not performed. A ventilation lung scan revealed bilateral uptake, but the uptake was diminished in the right lung. A perfusion scan revealed absent uptake in the right lung and decreased uptake in the left upper lobe. Cardiac catheterization revealed cardiac output 3.2 L/min (2.0 L/min per m²), aortic pressure 98/61 (mean 73) mmHg, mean pulmonary arterial wedge pressure 4 mmHg, pulmonary arterial pressure 56/32 (40) mmHg, right ventricular pressure 56/0–3 mmHg, and mean right atrial pressure 4 mmHg. The distal lower left pulmonary arterial pressure was 30/16 (25) mmHg, suggesting a peripheral pulmonary stenosis. A pulmonary angiogram revealed an absent right pulmonary artery and a markedly engorged left pulmonary artery (Fig. 5). Peripheral vessels supplying the left upper lobe appeared to be hypoplastic. The right lung was supplied from the right internal thoracic artery and intercostal artery. A stenosis at the left lower pulmonary artery was also confirmed. The final diagnosis included unilateral absence of the right pulmonary artery, pulmonary hypertension, hypoplastic left upper pulmonary artery and peripheral left pulmonary artery stenosis. Surgical intervention was not performed because there was no arterial system large enough for anastomosis in the right lung.

Over the next several years, his symptoms were stable with diuretics and he worked as a taxi driver, although he experienced occasional episodes of mild hemoptysis. However, since the age of 38 years, his symptoms gradually progressed and he stopped working because of worsening.

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in this group is an abnormal chest roentgenogram. It is clear that increased blood flow to the contralateral lung is the most important causative factor in the development of pulmonary hypertension in the presence of left-to-right shunt. However, it is poorly understood why pulmonary hypertension develops in cases of isolated UAPA, since usually one lung has the ability to accept a markedly increased blood flow with little or no change in pulmonary arterial pressure, as in case 1? Lang et al reported a case of late-onset pulmonary hypertension (25 years old) caused by pulmonary veno-occlusive disease, but this is not a universal finding.

The pathophysiology of pulmonary hypertension in case 2 is interesting. The combination of UAPA and contralateral peripheral pulmonary artery stenosis in this case is very rare, and we are aware of only 3 cases in the literature. Clearly, stenosis of the left lower pulmonary artery contributed to the development of pulmonary hypertension in the main and the left upper pulmonary artery. Chronic hypoxia caused by V/Q mismatch might have partly contributed to the development of pulmonary hypertension. Although chronic recurrent pulmonary emboli associated with secondary polycythemia might be another possibility, anticoagulation therapy was not administered due to recurrent hemoptysis.

There are several therapeutic options depending on the type and severity of the symptoms. Asymptomatic patients should be followed serially without intervention. In cases with recurrent pneumonia or hemoptysis, pneumonectomy should be considered. In cases of pulmonary hypertension and/or congestive heart failure, a surgical anastomosis should be considered, if adequate intrapulmonary branches of the pulmonary artery exist? However, it is frequently difficult to perform a corrective surgery and there have been only a limited number of reports regarding successful surgical treatment. This may be attributed to the abnormal vasculature in the affected lung, such as hypoplasia or arterio-venous fistula. Neither of our patients was treated surgically.

**Concluding Remarks**

Contralateral peripheral pulmonary arterio-
al stenosis is a rare coexisting anomaly in UAPA, but is an important cause of pulmonary hypertension and a poor prognosis similar to a left-to-right shunt. Physicians should be aware of the possibility of this rare combination in cases of adult UAPA with pulmonary hypertension.

REFERENCES