Direct Communication Between the Left Pulmonary Artery and the Left Atrium

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A 17-year-old patient was found to have cyanosis. A right-to-left shunt was suspected clinically and confirmed by noninvasive techniques such as perfusion lung scan and contrast echocardiography. Angiography showed this shunt to be between the left pulmonary artery and the left atrium. We believe this to be the first report of a hitherto undescribed congenital anomaly. (Chest 1989; 96:937-39)

An abnormal communication between the pulmonary artery and the left atrium is a very rare congenital anomaly and has now been recognized as a specific diagnostic category. Only 25 cases had been reported in the English literature until 1986.1 All of them had a communication from the right pulmonary artery to the left atrium. We describe a young patient who was found to have cyanosis and a communication from the left pulmonary artery to the left atrium, an anomaly that we believe has never been reported before.

Case Report

A 17-year-old schoolgirl had cyanosis and was referred for further investigation. When first seen in the outpatient clinic in October 1986, the patient admitted that she had felt mild exertional dyspnea since childhood, and three years earlier had noticed persistent blue coloration of her fingernails but, being asymptomatic, had considered this to be "normal." She was a nonsmoker and had no history of diabetes, hypertension, or rheumatic fever.

The patient was born full-term after a normal vaginal delivery. Her mother had not taken any drugs or suffered from any illness during the pregnancy, and there were no specific problems in the perinatal period. None of the three siblings or parents has a problem similar to that of the patient.

This patient, whose weight was 50 kg, height 1.6 m, and body

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Figure 1. Echocardiography. A. Parasternal short-axis view. A dilated venous channel can be seen entering the left atrium behind the atrial appendage. B. Apical four-chamber view. Note dilated venous channel entering the medial part of the left atrium. C. Right atrium and right ventricle filled with contrast medium. The contrast can be seen entering the left atrium through the dilated venous channel. D. Contrast echocardiogram one year later, showing absence of right-to-left shunt. RA = right ventricle; LA = left atrium; RV = right ventricle; LV = left ventricle; RPV = right pulmonary vein; LPV = left pulmonary vein; AO = aorta; VC = venous channel; LAA = left atrial appendage.
was no thrill. The jugular venous pressure was normal. There were no signs of heart failure and apart from a soft systolic murmur in the upper left sternal edge, no abnormality was found on the remainder of the general physical examination. There were no telangiectasiae and no bruits were heard anywhere in the chest.

Her hemoglobin was 17 g/dl and packed cell volume 53.6. An ECG and results of spirometric study were normal. On room air, \( P_{O_2} \) was 7.37 kPa (54.86 mm Hg), and \( P_{C0_2} \) was 4.38 kPa (32.85 mm Hg). While she breathed 100 percent \( O_2 \) for 15 min, the values were 8.41 kPa (63.08 mm Hg) and 4.73 kPa (35.48 mm Hg), respectively, indicating that she had a right-to-left shunt at the cardiac or pulmonary level. Perfusion lung scan demonstrated right-to-left shunt. Perfusion to both lungs was normal. Two-dimensional echocardiography showed that the cardiac chambers, the valves, and the great vessels were normal but showed a greatly dilated venous channel, measuring 2.2 cm in diameter, entering the superomedial part of the left atrium (Fig 1, A and B).

Contrast medium injection from the right antecubital vein completely opacified the right atrium and ventricle, demonstrating intact atrial and ventricular septa. Within two cardiac cycles after its appearance in the right side of the heart, the contrast material appeared in the left atrium through the dilated venous channel (Fig 1, C). Soon the whole left heart was flooded by the contrast material, indicating a communication between the pulmonary artery and the pulmonary venous system. The diagnosis and location of this shunt were confirmed by an angiogram that showed an abnormal communication from the posterior aspect of the left pulmonary artery, close to its origin from the main trunk, to the left atrium (Fig 2).

She underwent surgery on May 19, 1987. There was a thrill at the main pulmonary artery. A communication was found between the base of the left pulmonary artery and the left atrium. The channel was 1.5 cm in diameter at the pulmonary end but was difficult to identify at the atrial end—in fact, it was acting as an access to the left atrial appendage. The communication was double ligated. The thrill disappeared, and \( O_2 \) saturation rose to 99.8 percent. Her cyanosis has disappeared, she does not feel dyspneic on exertion, she has gained weight, and her hemoglobin level is normal. Contrast echocardiogram one year after surgery did not show any shunting of blood (Fig 1, D).

**COMMENT**

The inability to correct the hypoxemia with 100 percent \( O_2 \) suggested that the patient described here had a right-to-left shunt.\(^4\) Shunting can occur at the cardiac level, in the lungs, and has been reported to occur in the liver in patients with cirrhosis. In view of the absence of any cardiac or liver disease, the most likely site of a shunt in this patient was the lung. Pulmonary venous fistula of the lung is an unusual abnormality, and in 60 percent of cases it is associated with hereditary hemorrhagic telangiectasia (Rendu-Osler-Weber syndrome). Studies have established the effective prevention of morbidity and relief of symptoms by surgical excision.\(^5\)

Various authors have described an abnormal communication between the right pulmonary artery and the left atrium.\(^1,4,5\) Literature search, however, did not reveal a single case report of a communication between the left pulmonary artery and the left atrium, and we believe this to be the first report of a hitherto undescribed congenital anomaly. The patient's right-to-left shunt was diagnosed clinically, substantiated by noninvasive techniques such as perfusion lung scan and contrast echocardiography,\(^4\) and confirmed and localized preoperatively by angiography. Similar to other abnormal communications, this shunt can also be expected to have high morbidity and mortality but is curable by

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**FIGURE 2.** Pulmonary angiography: injection of the dye into the shunt with spillover into the left atrium. A (upper) Right antero-oblique view. B (lower) Left antero-oblique view. a = shunt; b = left atrium; c = catheter in the pulmonary artery.

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mass index 19.53, had central cyanosis and clubbing of her finger and toenails. Her blood pressure was 110/60 mm Hg and pulse rate 70 beats/min and regular. The apex beat was normal, situated medial to the left midclavicular line in the fifth intercostal space. There
surgery if suspected and diagnosed accurately.

ACKNOWLEDGMENT: We thank Mr. George Varughese, Mrs. Augustilia G. Pinto, and Miss Aida Adomian for their secretarial assistance; Mr. Milan Sieber for his help with the figures; and Prof. Lars Bjork for his comments regarding the angiography.

REFERENCES
1 Lekuona I, Cabrera A, Inguzzo R, Gid C, Agosti J. Direct communication between the right pulmonary artery and the left atrium. Thorax 1986; 41:78-79

Rapid Resolution of Acute Cor Pulmonale with Recombinant Tissue Plasminogen Activator*

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A case of massive pulmonary embolism complicated by acute cor pulmonale and shock is presented. The IV administration of recombinant tissue-type plasminogen activator (rt-PA) was associated with prompt reversal of clinical, ECG, and nuclear radiographic findings. The role of thrombolytic therapy with rt-PA for massive pulmonary embolism associated with hemodynamic decompensation deserves further investigation. (Chest 1989; 96:939-41)

Acute cor pulmonale following massive pulmonary embolism is frequently a catastrophic event, resulting in significant morbidity and mortality. Early management is dictated by the degree of pulmonary vascular obstruction and directed toward rapid resolution of the obstruction and correction of accompanying hemodynamic instability. For patients in shock, two modalities are currently available that may achieve this end point: thrombolytic therapy and surgical embolectomy. The latter treatment is useful in selected cases: however, the procedure requires cardiopulmonary bypass and is associated with an unacceptable high mortality. Experience with streptokinase* and urokinase* has shown that mortality rates tend to be lower in patients treated with thrombolytic agents compared with heparin, although morbidity is higher. The availability of recombinant tissue plasminogen activator (rt-PA) has led to the reexamination of thrombolytic therapy in the treatment of venous thromboembolism. rt-PA is fibrin specific and, in addition, is equally effective in lysing both new and aged clots, whereas streptokinase is most effective in lysing recently formed clots. We present a case of acute cor pulmonale rapidly reversed with the IV administration of rt-PA.

CASE REPORT
A 69-year-old woman was admitted with dyspnea of sudden onset. She was recovering from a motor vehicle accident that had occurred 5½ hours earlier during which she sustained fractures to her right femur, tibia, and fibula. The patient was a nonsmoker with no prior history of pulmonary or cardiac disease, except well-controlled systemic hypertension. She had an uneventful postoperative course, and received 5,000 units of heparin sodium subcutaneously every 8 h. She was discharged two weeks following her accident taking heparin sodium, 5,000 units given subcutaneously every 12 h.

On the afternoon of admission, she suddenly became short of breath. She presented to her primary hospital with blood pressure, 100/60 mm Hg; pulse rate, 130 beats/min; and respiratory rate, 45/min. On physical examination she was afebrile, tachycardic, and in acute distress, with clear lung fields and a rapid heart rate without murmurs or gallops. An arterial blood gas (ABG) analysis breathing room air showed a pH, 7.35; Pco2, 26 mm Hg; and Pao2, 35 mm Hg. She was placed on a 100 percent nonrebreathe face mask. Repeated ABG values were pH, 7.49; Pco2, 30 mm Hg; and Pao2, 40 mm Hg. A chest x-ray film showed clear lung fields and a normal cardiac silhouette. An ECG revealed sinus tachycardia at a rate of 150 beats/min, right axis deviation, an S wave in lead 1, and a Q wave and inverted T wave in lead 3. Acute pulmonary embolism was strongly suspected. She received a 7,500-unit IV bolus of heparin sodium followed by a continuous infusion at a rate of 1,000 units/h. She was transferred to our institution for further evaluation.

Upon arrival she was markedly dyspneic and cyanotic, with a respiratory rate of 44 breaths/min. She was immediately intubated. Her blood pressure following intubation was 100/60 mm Hg. The lung examination was unremarkable. Her neck veins were greatly distended. Cardiac examination showed a prominent right ventricular heave. The pulmonic and aortic components of the second heart sound were of equal intensity. Third and fourth heart sounds were not appreciated. There was unilateral swelling of the right lower extremity. An ECG revealed sinus tachycardia and an S1-Q3-T3 pattern (Fig 1A). An ABG analysis performed on 100 percent inspired oxygen revealed the following: pH, 7.34; Pco2, 36 mm Hg; and Pao2, 56 mm Hg. A nuclear perfusion scan showed complete absence of perfusion to the left lung with multiple segmental perfusion defects in the right lung (Fig 2).

Despite intubation, fluid administration, and blood pressure support, the patient remained hypotensive. The decision was made to treat her with thrombolytic therapy. Intravenous rt-PA was infused via peripheral vein at a rate of 20 mg for the first hour, followed by 10 mg/h for 6h, for a total dose of 100 mg. Four hours following initiation of therapy, an ABG study revealed pH, 7.49; Pco2, 24 mm Hg; and Pao2, 83 mm Hg on 100 percent Fio2. At the completion of rt-PA infusion, an ABG analysis revealed pH, 7.55; Pco2, 20 mm Hg; and Pao2, 45 mm Hg. An ECG performed at this time revealed sinus tachycardia at a rate of 107 beats/min but was otherwise normal (Fig 1B). Her blood pressure was 125/63 mm Hg; and respiratory rate was 13 breaths/min. Jugular venous distention was no longer appreciated. She was subsequently extubated, 10 h following initiation of treatment with rt-PA.

A repeated ventilation-perfusion scan showed normal ventilation and perfusion in the left lung and minor perfusion defects of the