

Congenital Atresia of Unilateral Pulmonary Veins Associated With a Single Ventricle

— A Rare Case Report and Literature Review —

Hsing-Yuan Lee, MD^{*,‡,§}; Betau Hwang, MD^{†,‡}; Pi-Chang Lee, MD^{*,‡,†};
Sheng-Ling Jan, MD^{†,†}; C.C. Laura Meng, MD^{*,†}

Congenital atresia or extreme hypoplasia of individual pulmonary veins is a rare condition that is usually asymptomatic if it only involves 1 or 2 segments of the lungs. It may be fatal if it occurs in combination with other complex cardiac defects. The patients often present with recurrent pulmonary infections, hemoptysis, or cyanosis in the latter case. A definitive diagnosis can be made by cardiac catheterization with selective pulmonary wedge angiography. The treatment of this condition is a challenge and is controversial, and the prognosis is usually poor. We describe the case of a male infant who suffered from shortness of breath and cyanosis since birth. On echocardiography and cardiac catheterization, he was found to have a complex congenital heart disease with a single ventricle (right ventricle morphology), mitral atresia, large atrial septal defect, and atresia of the right pulmonary veins; severe pulmonary hypertension was also observed. He underwent pulmonary artery banding at the age of 2.5 months, but he died because of a pulmonary infection at the age of 6 months. In this report, we present this rare case and review the previous literature. (*Circ J* 2008; 72: 1544–1546)

Key Words: Congenital heart disease; Pulmonary vein atresia; Single ventricle

Congenital atresia or extreme hypoplasia of the individual pulmonary veins is a rare condition. It is usually asymptomatic if it involved only 1 or 2 lung segments, but it might be fatal if it occurs in combination with other complex cardiac defects!^{1–4} In congenital pulmonary vein atresia, patients often present with recurrent pulmonary infections, hemoptysis, or cyanosis if they have a complex heart disease. A definitive diagnosis can be made by cardiac catheterization with selective pulmonary wedge angiography. Treatment is a challenge and the method is controversial, and the prognosis is usually poor. Only 25 cases have been reported previously, and one-third of the patients in these cases had other congenital heart anomalies. The patient in our case not only had an associated complex congenital heart disease, which has not been previously reported with pulmonary vein atresia, but also demonstrated the progressive development of a reticular pattern on the lung in a chest X-ray (CXR).

Case Report

A 1-month-old male infant was transferred to our pedi-

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*Department of Pediatrics, Taoyuan Armed Force General Hospital, Taoyuan, **Division of Pediatric Cardiology, Taipei Veterans General Hospital, Taipei, †National Yang-Ming University, Taipei, ††Division of Pediatric Cardiology, Taichung Veterans General Hospital, Taichung, ‡Department of Pediatrics, Taipei City Hospital, Zhongxiao Branch, Taipei and ‡‡Department of Biological Science and Technology, National Chia Tung University, Hsinchu, Taiwan, ROC

Mailing address: Betau Hwang, MD, Department of Pediatrics, Taipei City Hospital, Zhongxiao Branch, No. 87, Tongde Rd. Nangnag District, Taipei City 11556 Taiwan, R.O.C. E-mail: bthwang@vghtpe.gov.tw All rights are reserved to the Japanese Circulation Society. For permissions, please e-mail: cj@j-circ.or.jp

atric cardiology department because of shortness of breath and abnormalities in a CXR and echocardiograms since early birth. On fetal echocardiography, he had been found to have an abnormal cardiac structure. He was born via an elective Caesarean section because congenital cardiac defect with possible respiratory distress after birth was suspected. After delivery, shortness of breath and cyanosis were noted, but the CXR appeared relatively normal (Fig 1A). On the second day, his symptoms worsened and he was sent to a general hospital where another CXR revealed a diffuse ground-glass pattern on the right lung (Fig 1B). After serial examination, including echocardiography and cardiac catheterization, the infant was diagnosed with a complex congenital heart disease. He had a single ventricle (SV), mitral atresia (MA), atrial septal defect (ASD), patent ductus arteriosus (PDA), hypoplasia of the right pulmonary artery (RPA) with pulmonary hypertension (HTN), right-sided pulmonary edema, and pleural effusion. The shortness of breath did not improve despite the administration of anti-congestive therapy and spontaneous PDA closure. After a respiratory infection caused by Enterobacter aerogenes was controlled, the infant was transferred to our hospital for further evaluation.

At admission, physical examination under endotracheal intubation revealed generalized cyanosis, a grade II/VI systolic murmur over the left upper sternal border, and coarse breath sounds with rales over the right lung. A CXR revealed a reticular pattern over the right lung. Echocardiography and cardiac catheterization revealed the situs solitus, SV with a right ventricular morphology, MA, a large ASD, a hypoplastic RPA, and severe pulmonary HTN. Main pulmonary artery angiography revealed a relatively small RPA, and it was observed that the flow from the RPA and its branch reversed to the left side after the contrast medium in the left pulmonary artery (LPA) had drained into the left

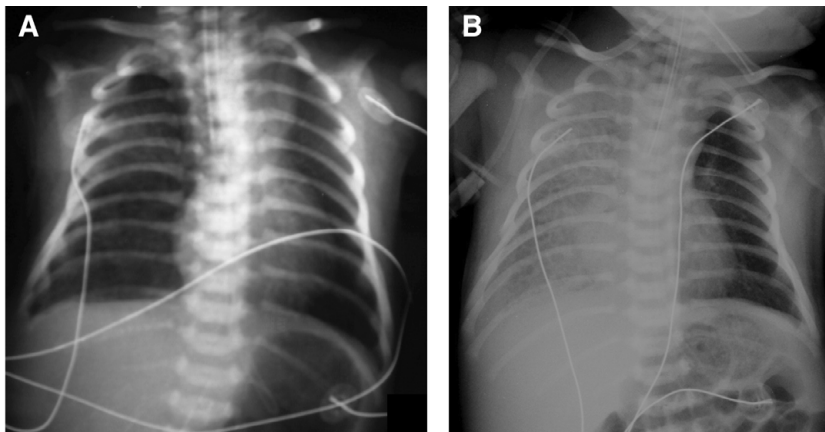


Fig 1. A chest X-ray on day 1 (A) revealed a relatively normal lung field and heart size, but that on day 2 (B) revealed a reticular pattern on the right lung.

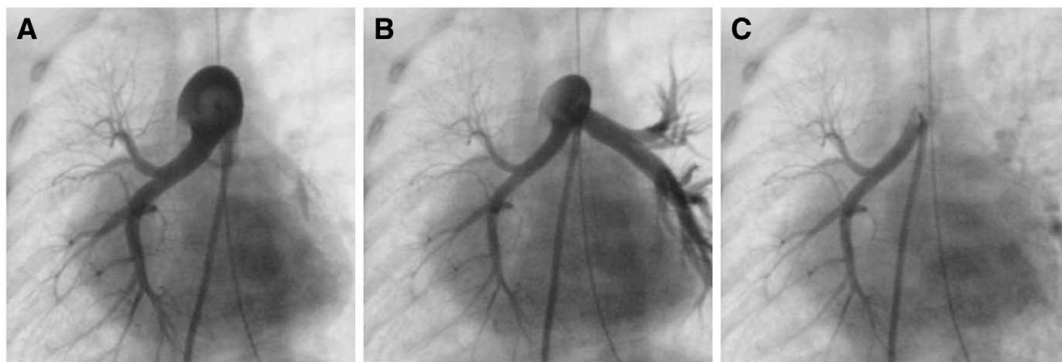


Fig 2. Balloon occlusion angiography of the pulmonary artery. (A) During balloon inflation to occlude the left pulmonary artery (LPA), the contrast medium filled the right pulmonary artery (RPA) and its branches. After 30 heartbeats, the RPA could still be visualized and showed the steady movement of the contrast medium. (B) After the balloon was released, the contrast medium drained into the LPA and then drained back into the left atrium through the left pulmonary vein. (C) Even after the contrast medium in the left lung had disappeared, that in the RPA revealed steady movement.

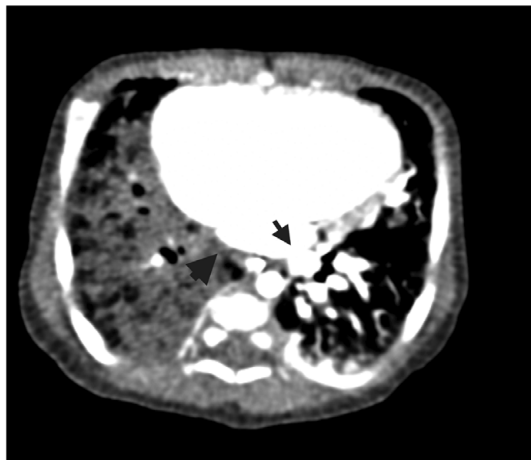


Fig 3. An axial CT scan with contrast medium injection showed diffuse right pulmonary edema and the absence of a junction between the right pulmonary vein and left atrium (large arrow). The tortuous and dilated left pulmonary veins drained into the left atrium on the left side (small arrow).

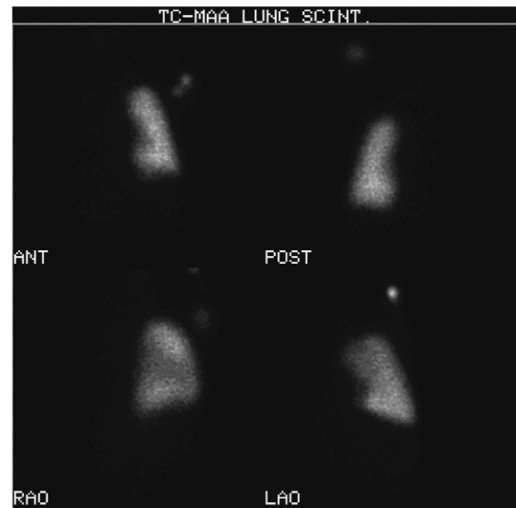


Fig 4. On the Tc-99m lung perfusion scan, the right lung could not be visualized, indicating the absence of perfusion. RAO, right anterior oblique; LAO, left anterior oblique.

atrium. Balloon occlusion angiography of the pulmonary artery, in which a Berman catheter occluded the LPA, showed that the contrast medium persisted in the RPA and its branches for dozens of heartbeats, and none of the right pulmonary veins was opacified. After deflation of the bal-

loon, the contrast medium refluxed sequentially into the LPA, left pulmonary veins, and left atrium (Fig 2). A contrast axial chest CT revealed that the left pulmonary veins were dilated and tortuous, but no right pulmonary veins were visualized (Fig 3). A Tc-99m lung perfusion scan dem-

onstrated the absence of right lung perfusion (Fig4). To treat the congestive heart failure and preserve lung function, pulmonary artery banding was performed at the age of 2.5 months. After surgery, the patient was ventilator dependent because of severe hypoxia, and he finally died of Enterobacter bacteremia and massive hemoptysis at the age of 6 months.

Discussion

The pulmonary veins communicate with the vitello-umbilical and cardinal veins during the embryonic stage. An outpouching from the left atrium then establishes a communication with this plexus to form the common pulmonary vein. The communication with the fetal systemic veins disappears later, and the common pulmonary vein regresses into the left atrium.⁵ In our case, the pulmonary vein atresia might have occurred after the disappearance of the communication between the fetal pulmonary vein and the systemic venous system, resulting in the absence of anomalous pulmonary vein connections. Sade et al reported that the obstruction between the pulmonary vein and left atrium might be a diaphragm flap or segmental intimal fibrosis.⁵ Although bronchial-to-pulmonary circulation allows for ipsilateral lung growth, it usually causes hypoplasia during fetal and perinatal life.^{5,6}

The symptoms and signs become severe and appear earlier if the patients exhibit pulmonary vein atresia along with a complex heart disease. Pulmonary venous congestion develops after birth. Severe shortness of breath and cyanosis occur during early infant life because of ventilation-perfusion mismatch, cyanotic congenital heart disease, and congestive heart failure. The reticular pattern on the lung also develops early, and it is obviously caused by massive pulmonary blood flow and pulmonary HTN. The lungs are easily infected, and the hemoptysis occurs earlier.⁷

Physical examination and electrocardiograms are of no value if the pulmonary vein atresia is combined with other cardiac abnormalities.

A definitive diagnosis could only be obtained by pulmonary arterial wedge angiography, which usually shows blind pulmonary venous confluence or the atresic segments of the pulmonary veins.^{1,8} Correct diagnosis is important, especially when the pulmonary arterial wedge angiography is unsuccessful. Some imaging studies are helpful and suggestive. A CXR often reveals a unilateral reticular vascular pattern on the involved lung.^{1,2,4,9} A chest axial CT scan showed a smooth ipsilateral atrial wall at the expected location of the involved pulmonary vein.¹⁰ Tc-99m lung perfusion scans often show a markedly decreased or absent blood flow to the involved lung.^{4,10,11} Pulmonary angiography frequently demonstrates preferential flow to the uninvolved lung and the distension of the contralateral pulmonary veins that have normal transit time. A very important clue in the diagnosis of unilateral pulmonary vein atresia is the absence or rather the reversal of pulmonary blood flow in the affected lung.⁶ This particular finding of pulmonary flow reversal is strongly suggestive of unilateral pulmonary vein atresia.

The hemodynamic findings generally exhibit elevated

unilateral or bilateral pulmonary artery and pulmonary capillary wedge pressures! Unfortunately, the elevated pulmonary artery pressure makes it impossible for the Fontan operation to be performed. Oxygen saturation in the left atrium is reduced because of the ventilation-perfusion mismatch, which is not easily seen in SV with MA.

The treatment of unilateral pulmonary vein atresia is challenging and controversial. Usually, the aim of the treatment is to preserve lung function. Theoretically, treatment would involve a surgical anastomosis between the pulmonary vein and left atrium;⁴ however, it is often difficult to preoperatively determine the position of obstruction. If pulmonary venous drainage cannot be re-established, the involved lung sections should be removed to prevent reflex vasoconstriction secondary to the ventilation-perfusion mismatch.⁷ Pulmonary artery banding is inadequate because in our case, capillary constriction by the involved lung persisted and the hypoxia worsened. Ipsilateral pneumectomy has been used to treat simple unilateral pulmonary vein atresia, and in most cases, the patient survived; however, most patients who also had congenital heart disease died before the decision to perform surgery was made!¹ Heart-lung transplantation might be considered for patients with pulmonary vein atresia combined with other uncorrectable congenital cardiac defects, but it has not been reported thus far.

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