

## Specific echocardiographic findings useful for the diagnosis of common pulmonary vein atresia

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### Abstract

In this paper, we report a case of common pulmonary vein atresia, which is a very rare disease characterized by cyanosis, heart failure and pulmonary hypertension. Reverse flow in the pulmonary artery at end-diastole as well as in the isthmus of the aorta from early systole to end-diastole detected by echocardiography were found to be specific features useful in diagnosing the disease.

### Case Report

The patient was a Japanese male born after a full-term pregnancy (40 weeks and 0 days; weight: 3142 g, height: 48.5 cm). The mother had a normal gestational history, no familial history of congenital malformations, and a non-consanguineous marriage. The mother did not smoke or consume alcohol during pregnancy. The Apgar scores were 7 and 8 at 1 and 5 min after birth, respectively. The patient experienced severe dyspnea and cyanosis. No heart murmur or liver enlargement was detected. Radiographic examination showed pneumothorax, a slight accumulation of meconium, and no cardiac dilatation. The presence of the pneumothorax made it challenging to precisely identify the cause of cyanosis by echocardiography. Findings at admission were small left atrium with a possible lack of pulmonary venous flow to it, as well as no common chamber like structure. Administration of a vasodilator and nitric oxide therapy resulted in no effects. The alveolar-arterial oxygen difference was 641 mmHg, and extracorporeal membranous oxygenation therapy was started at the age of 1 day to treat hypoxia. At day 3 after birth, echocardiographic examination revealed: i) total anomalous pulmonary venous connection, ii) a small common chamber without discrete stenosis, iii) a small unobstructed vertical vein connected to the innominate vein, and iv) severe tricuspid valve regurgitation (flow speed, 6.0 m/s). Abnormal echocardiographic findings included a retrograde flow in the pulmonary artery at end-diastole and a

reverse flow in the isthmus of the aorta from early systole to end-diastole (Figure 1). These findings remained unchanged throughout the study period.

Despite intensive treatment, the patient died of hypoxia and heart failure on day 7 after birth. An autopsy was performed after obtaining parental consent, which revealed: i) no venous connections to the left atrium, ii) four pulmonary veins of similar size with no stenosis that formed a small common chamber, and iii) a small but unobstructed vertical vein connecting the common chamber and the innominate vein (Figure 2). No other defects were detected in the heart or other organs.

### Discussion

Common pulmonary vein atresia is a rare congenital heart disease characterized by the lack of direct communication of the common chamber of the pulmonary veins to the heart or systemic venous system. It was first reported by Lucas *et al.* in 1962.<sup>1</sup> Despite recent medical advances, the prognosis for this condition is poor. In the absence of surgical intervention, early survival may depend on the collateral circulation from the pulmonary veins to the systemic venous systems.<sup>2</sup> Although angiocardiology would be helpful for visualizing venous structures and flows as well as for evaluating the feasibility of operation, the condition of our patient and the necessity of intensive care therapy made it impossible to perform. Under such circumstances, echocardiography remained the next most feasible examination technique. Khonsari *et al.* reported that immediate recognition and surgical intervention are important for successful treatment of this condition.<sup>3</sup> However, echocardiographic examination was found to be difficult in patients with pneumothorax.<sup>4,5</sup> In addition, definitive criteria that would allow establishing a diagnosis using this approach have not been reported. Nevertheless, in this study we successfully used echocardiography to observe a reverse flow in the pulmonary artery at end-diastole as well as in the isthmus of the aorta from early systole to end-diastole. Thus far, no such findings have not been reported in patients with total anomalous pulmonary venous connection. Interestingly, no endothelial thickness of the pulmonary arteries or pulmonary vein obstruction was seen on macroscopic examination, although the results of echocardiography indicated high pressure in the pulmonary artery, suggesting such an obstruction. These findings may facilitate establishing a diagnosis of common pulmonary vein atresia using echocardiography. Further studies will determine the progress in the diagnosis of this condition.

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### Conclusions

Reverse flow in the pulmonary artery at end-diastole as well as in the isthmus of the aorta from early systole to end-diastole detected by echocardiography were found to be specific features useful in diagnosing the disease.

### References

1. Lucas RV, Woolfrey BF, Anderson RC, et al. Atresia of the common pulmonary vein. *Pediatrics* 1962;29:729-39.
2. Shimazaki Y, Yagihara T, Nakada T, et al. Common pulmonary vein atresia: a successfully corrected case. *J Cardiocasc Surg* 1987;28:395-7.
3. Khonsari S, Saunders PW, Lees MH, Starr A. Common pulmonary vein atresia: importance of immediate recognition and surgical intervention. *J Thorac Cardiovasc Surg* 1982;83:443-8.
4. Sharda JK, Kurlandsky LE, Lacina SJ, Radecki LL. Spontaneous pneumothorax in common pulmonary vein atresia. *J Perinatol* 1990;10:70-4.
5. Dominguez Garcia O, Granados Ruiz MA, Sanchez-Redondo MD, et al. A difficult emergency surgical diagnosis: atresia of the common pulmonary vein. *Pediatr Cardiol* 2009;30:989-91.

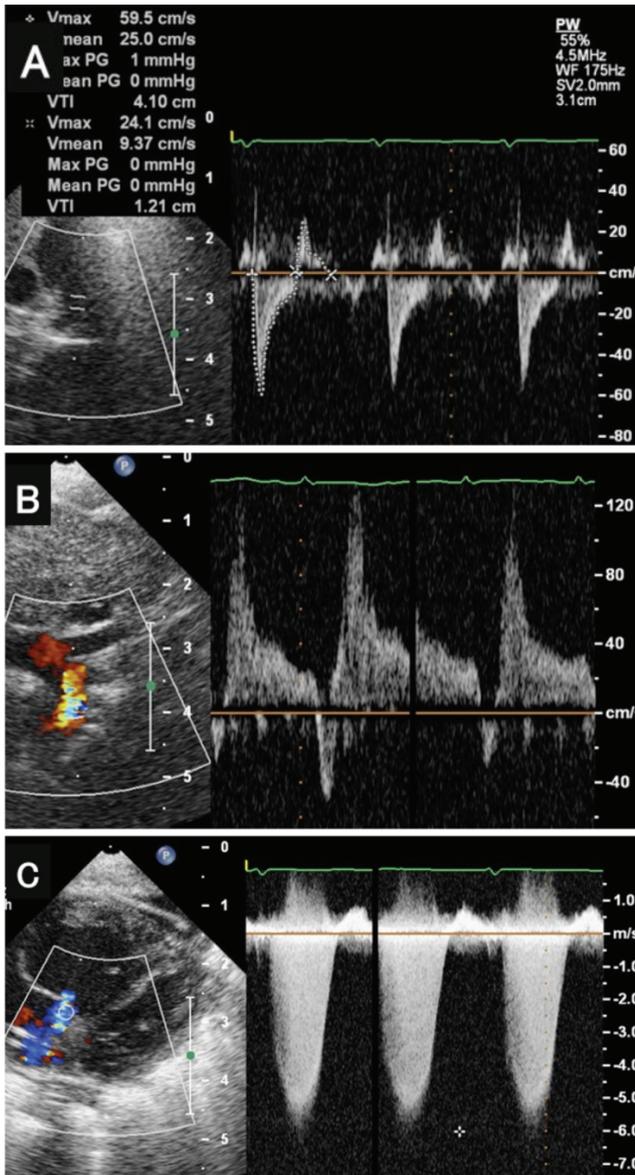


Figure 1. Echocardiographic findings. A) Pulmonary flow, a reverse flow was detected at end-diastole; B) Aortic isthmus flow, a reverse flow through the ductus arteriosus from early systole to end-diastole; C) remarkable tricuspid valve regurgitation at 3 days old.

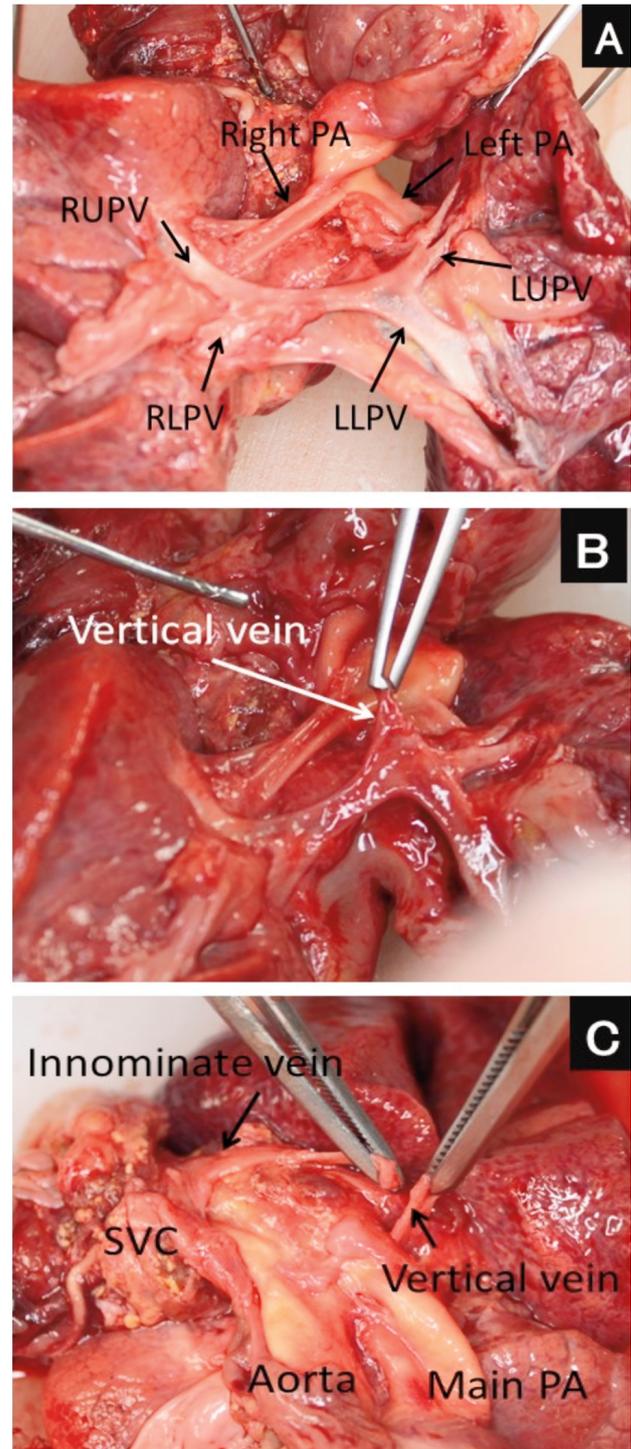


Figure 2. Macroscopic findings in the patient's heart. Dorsal side of the heart. A) No connective veins to the left atrium, with all four pulmonary veins forming a small common chamber; B) Very narrow vertical vein found at the dorsal side of the left pulmonary artery; C) This vertical vein connected to the innominate vein. Abbreviations: LLPV, left lower pulmonary vein; LUPV, left upper pulmonary vein; PA, pulmonary artery; RLPV, right lower pulmonary vein; RUPV, right upper pulmonary vein; SVC, superior vena cava.