

Isolated Unilateral Pulmonary Vein Atresia in Adult Patients: A Case Report and Literature Review

Min Zhang, Qing-Chen Wu, Cheng Zhang, Qiang Li, Ying-Jiu Jiang, Dan Chen

Department of Cardiothoracic Surgery, the First Affiliated Hospital, Chongqing Medical University, Chongqing, China

ABSTRACT

Unilateral pulmonary vein atresia is a rare abnormality that usually presents in infants with recurrent hemoptysis and pneumonia. Presentation in adulthood without additional congenital heart disease is rare but does occur. Anatomic variations in the pulmonary vessels that supply and drain the affected lung can explain the mildly symptomatic process. The diagnosis of isolated unilateral pulmonary vein atresia is usually made angiographically. Pneumonectomy is indicated once symptoms or complications are present so that irreversible pulmonary hypertension can be avoided.

INTRODUCTION

Congenital unilateral pulmonary vein atresia frequently occurs in patients with total anomalous pulmonary venous drainage and associated structural abnormalities of the heart [Kingston 1983]. Patients usually present in infancy or childhood with recurrent episodes of pneumonia or hemoptysis [Mataciunas 2009]. Presentation in adulthood without additional congenital heart disease is rare but does occur [Heyneman 2001]. We describe an adult patient with isolated right-sided pulmonary venous atresia. This case brings the total number of cases in the literature to 3.

CASE REPORT

A 22-year-old female patient was admitted for investigation of recurrent hemoptysis and a grave episode in 2008. Chest radiography revealed a hypoaerated right lung with significant consolidation of the right middle and lower lobe. There was a mediastinal shift to the right with an asymmetric vascular pattern. Turbulent reversed flow in the right pulmonary artery was found during an echocardiography examination, and visualization of the right pulmonary veins was difficult. There were no signs of pulmonary hypertension or right ventricular overload. She had no history of surgical procedures.

A chest computed tomography scan performed to exclude a mediastinal mass revealed the absence of right pulmonary veins (Figure 1). A cardiac catheterization evaluation showed

no intracardiac anatomic abnormality. An injection into the main pulmonary artery revealed that all of the pulmonary blood flow was directed to the left lung. The right pulmonary artery was hypoplastic with no contrast reaching beyond the first segmental pulmonary artery branches. There was significant collateral blood flow from large bronchial and subclavian vessels into the right lung, with drainage occurring through the right pulmonary artery (Figure 2). No pulmonary veins were observed on the right side. A bronchoscopy examination showed a hyperemic right bronchial mucosa with increased secretions, but the left side was normal.

At surgery, the affected lung appeared hypoplastic and inflammatory. The macroscopic pulmonary veins were hypoplastic and markedly thickened. The luminal continuity of the right pulmonary venous pathway was completely obliterated and replaced by 3 cords of fibrosis, which extended to the site of entrance into the right atrium. A right pneumonectomy was performed. A pathologic examination confirmed the hypoplasia of the macroscopic veins and revealed medial hypertrophy with significant intimal fibrosis of the pulmonary veins.

Nine days later, after an uneventful recovery, the patient was discharged home.

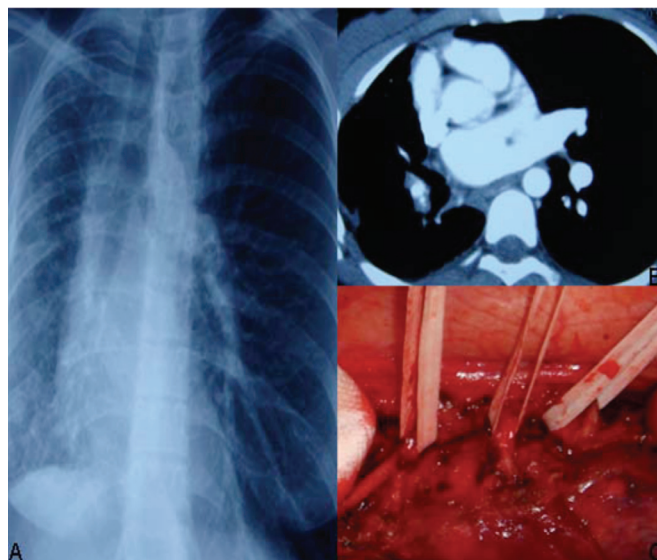


Figure 1. A, Chest x-ray showing the hypoplasia of the right lung. B, Chest computed tomography scan showing the absence of right pulmonary veins. C, Fibrosis of the right pulmonary veins in the operative field.

Received June 9, 2010; accepted July 7, 2010.

Correspondence: Prof. Qing-Chen Wu, Department of Cardiothoracic Surgery, the First Affiliated Hospital, Chongqing Medical University, 1, Youyi Rd, Yuzhong District, Chongqing, China (e-mail: doctor1984@sina.com).

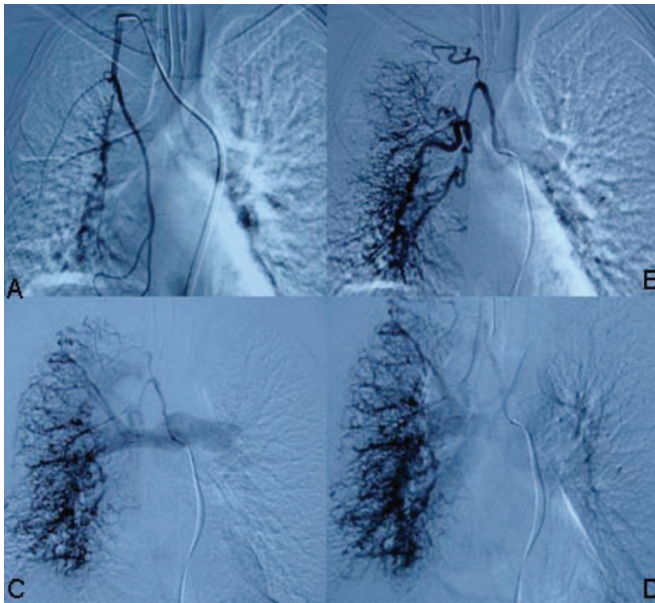


Figure 2. Anteroposterior view of the systemic-to-pulmonary circulation with collateral angiograms from the subclavian artery (A) and from the bronchial artery (B). This circulation caused a shunt from the aorta to the right pulmonary artery (C) and then, with reversed flow, to the left pulmonary circulation (D).

DISCUSSION

Including our patient, 3 cases of isolated unilateral pulmonary vein atresia in adult patients have been reported in the medical literature. The median age was 23 years. All patients underwent pulmonary resection and were alive at the end of the follow-up period. The clinical data are summarized in the Table. Such atresia may occur in either lung, with no right- or left-sided predominance, and it usually presents in infants. The majority of cases are thought to be congenital and caused by late failure of incorporation of the common pulmonary vein into the left atrium [Cabrera 1985]. Abnormal intimal proliferation of spindle-shaped cells identified as myofibroblasts, without any evidence of thrombosis, inflammation, or fibrosis, may be an additional mechanism [Pourmoghadam 2003]. Some other acquired causes of pulmonary vein atresia include myxoma of the left atrium, mediastinal neoplasm, mediastinal fibrosis, diaphragm completely occluding the venoatrial junction, and so forth [Cabrera 1985; Heyneman 2001; Pourmoghadam 2003].

The clinical presentation consists of recurrent pulmonary infections, hemoptysis, and exercise intolerance in infancy

[Cabrera 1985]. Patients may also remain mildly symptomatic until adulthood. This finding can be explained by anatomic variations in the pulmonary vessels that supply and drain the affected lung. In our patient, for example, significant collateral blood flow from large bronchial and subclavian vessels into the right lung, with drainage occurring through the right pulmonary artery, was well developed (Figure 2). This anatomy may explain the benign course in our patient. Close follow-up is suggested for asymptomatic or mildly symptomatic patients with this pathology [Pourmoghadam 2003; Tissot 2008]. Pulmonary artery hypertension is also frequently associated with this disorder. The increased blood flow through the bronchial arteries, which normally carry <5% of the cardiac output and communicate with the pulmonary branches via precapillary anastomoses, is a possible explanation [Cullen 1990].

The diagnosis of isolated unilateral pulmonary vein atresia is usually made angiographically. Selective injections into the pulmonary arteries revealed the slow passage of contrast into the affected lung with an absence of venous opacity on venous-phase imaging. The presence of systemic-to-pulmonary arterial collaterals is suggested both by the reversal of blood flow in the pulmonary artery and by an increase in oxygenation in the ipsilateral pulmonary artery. This systemic-to-pulmonary circulation can also be confirmed directly by a collateral angiogram, as in our patient. Noninvasive diagnosis by computed tomographic angiography, cardiac magnetic resonance imaging, or bronchoscopy has been described in recent publications [Heyneman 2001; Artero 2008; Tissot 2008] and has the advantages of avoiding unnecessary repetition of intracardiac investigations and guiding further assessment and treatment options [Mataciunas 2009].

Treatment of unilateral pulmonary vein atresia depends on the severity and extent of the anomaly. Percutaneous balloon dilation with stent implantation at the junction of the pulmonary venous confluence and the left atrium junction has been reported in the cases with localized stenosis, but the long-term prognosis for this procedure still has to be evaluated [Tissot 2008]. Resection of the obstructing membrane at the pulmonary vein and the left atrial junction or resection of the atretic segment of the pulmonary veins and their reanastomosis to the left atrium has been tried when a direct approach to the pulmonary veins has been feasible. Reconstitution of flow to the left atrium, however, does not allow the restoration of pulmonary function and remodeling of the pulmonary vasculature. The evolution toward subsequent involvement of the contralateral lung and development of pulmonary hypertension is irreversible [Ussia 2004]. Pneumonectomy has therefore been proposed to avoid irreversible pulmonary hypertension [van der Werf 1994]. The Table shows cases of satisfactory long-term results.

Cases of Isolated Unilateral Pulmonary Vein Atresia in Adult Patients

Reference	Symptoms	Age at Diagnosis, y	Sex	Side	Surgery	Outcome (Patient Age, y)
[Heyneman 2001]	Dyspnea and PAH*	21	Female	Right	Pneumonectomy	Alive (33)
[Harrison 1996]	Hemoptysis and hematemesis	27	Female	Left	Pneumonectomy	Alive (39)
Current study	Hemoptysis	22	Female	Right	Pneumonectomy	Alive

*PAH indicates pulmonary artery hypertension.

CONCLUSION

In summary, unilateral pulmonary vein atresia is a rare abnormality that usually presents in infants with recurrent hemoptysis and pneumonia. Presentation in adulthood without additional congenital heart disease is rare but does occur. Anatomic variations in the pulmonary vessels that supply and drain the affected lung can explain the mildly symptomatic process. The diagnosis of isolated unilateral pulmonary vein atresia is usually made angiographically. Pneumonectomy is indicated once symptoms or complications are present so that irreversible pulmonary hypertension can be avoided.

REFERENCES

- Artero MI, Serrano PF, Padin Marin MI, Serrano RF. 2008. Congenital unilateral pulmonary vein atresia: imaging findings [in Spanish]. *Radiologia* 50:82-5.
- Cabrera A, Vazquez C, Lekuona I. 1985. Isolated atresia of the left pulmonary veins. *Int J Cardiol* 7:298-302.
- Cullen S, Deasy PF, Tempany, Duff DF. 1990. Isolated pulmonary vein atresia. *Br Heart J* 63:350-4.
- Harrison JK, Hearne SE, Baker WA, et al. 1996. Esophageal varices in association with unilateral pulmonary vein atresia. *Cathet Cardiovasc Diagn* 38:387-92.
- Heyneman LE, Nolan RL, Harrison JK, McAdams HP. 2001. Congenital unilateral pulmonary vein atresia: radiologic findings in three adult patients. *AJR Am J Roentgenol* 177:681-5.
- Kingston HM, Patel RG, Watson GH. 1983. Unilateral absence or extreme hypoplasia of pulmonary veins. *Br Heart J* 49:148-53.
- Mataciunas M, Gumbiene L, Cibiras S, Tarutis V, Tamosiunas AE. 2009. CT angiography of mildly symptomatic, isolated, unilateral right pulmonary vein atresia. *Pediatr Radiol* 39:1087-90.
- Pourmoghadam KK, Moore JW, Khan M, et al. 2003. Congenital unilateral pulmonary venous atresia: definitive diagnosis and treatment. *Pediatr Cardiol* 24:73-9.
- Tissot C, Corbelli R, Aggoun Y, Beghetti M, da Cruz E. 2008. Bronchoscopic diagnosis of asymptomatic unilateral pulmonary vein atresia in an infant. *Pediatr Cardiol* 29:976-9.
- Ussia GP, Marasini M, Rimini A, Pongiglione G. 2004. Atresia of right pulmonary veins with intact atrial septum and major aorto-pulmonary collateral treated with percutaneous stent implantation and embolization. *J Interv Cardiol* 17:183-7.
- van der Werf TS, Mannes GP, Hew JM, et al. 1994. Improved gas exchange after pneumonectomy in an adult with incomplete pulmonary vein atresia. *Thorax* 49:723-5.