

# Isolated Pulmonary Vein Stenosis Mimicking Chronic Lung Disease

S Chakrabarti, S Tsao, J Vettukattil, J Gnanapragasam

---

## Citation

S Chakrabarti, S Tsao, J Vettukattil, J Gnanapragasam. *Isolated Pulmonary Vein Stenosis Mimicking Chronic Lung Disease*. The Internet Journal of Pediatrics and Neonatology. 2003 Volume 4 Number 1.

## Abstract

Pulmonary vein stenosis is extremely rare, progressive and usually fatal, if left untreated. It is unsuspected during clinical examination and often overlooked during routine echocardiography and even cardiac catheter studies. This report describes a patient in whom the diagnosis was delayed as he had symptoms and signs, which are very frequently observed in chronic lung disease.

## CASE HISTORY

An 11-month-old female infant presented with history of poor feeding and shortness of breath for 4 weeks. Born at 24 weeks of gestation, she was ventilated for 5 weeks and was oxygen dependant for nearly 3 months. Apart from a ductus arteriosus (which had been treated medically during her neonatal period) she was known to have a structurally normal heart on echocardiography.

She had been having progressive wheezy symptoms requiring repeated hospital admissions with “wheezy episodes”, “upper respiratory tract infections” and “chest infections” in the next few months.

On examination, the infant was in severe respiratory distress with oxygen saturations of 50% in air. A third heart sound and prominent hepatomegaly was present. A large cavernous haemangioma was seen on her head.

There was evidence of right atrial and right ventricular enlargement on electrocardiogram. A significantly increased cardio-thoracic ratio was observed on X Ray (Figures 1 & 2) and a clinical diagnosis of right heart failure secondary to chronic lung disease was made and treatment started with diuretics.

Trans-thoracic echo confirmed a grossly dilated right heart with poorly functioning right ventricle. Marked tricuspid regurgitation and right to left shunting across the patent foramen ovale suggested severe pulmonary hypertension. Doppler studies showed a high velocity pulmonary venous flow, which was suggestive of pulmonary venous

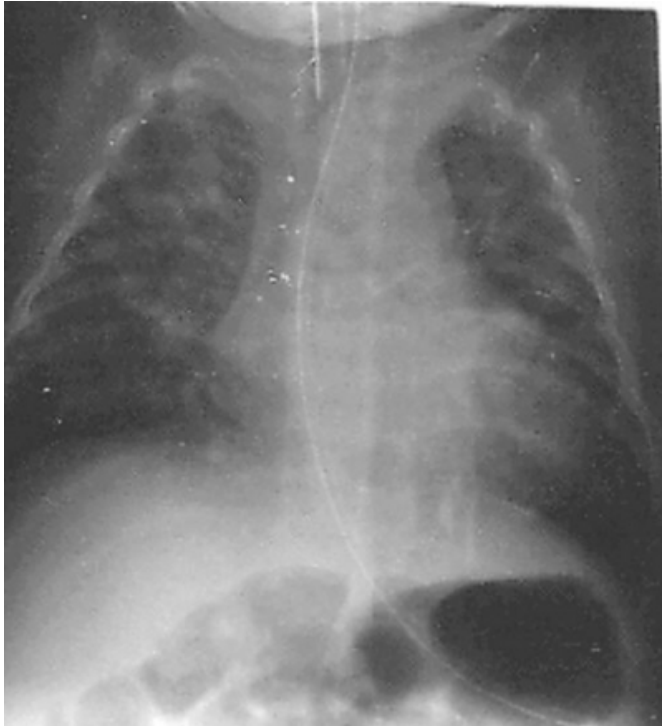
obstruction.

This was confirmed on transoesophageal echocardiography, which showed high velocity continuous inflow through both right and left pulmonary veins. Cardiac catheter studies revealed severe left sided pulmonary vein insertion stenosis. There was an abnormal hypertensive pulmonary arterial tree.

She was operated for her pulmonary vein stenosis subsequently, but died due to re-stenosis after a few months.

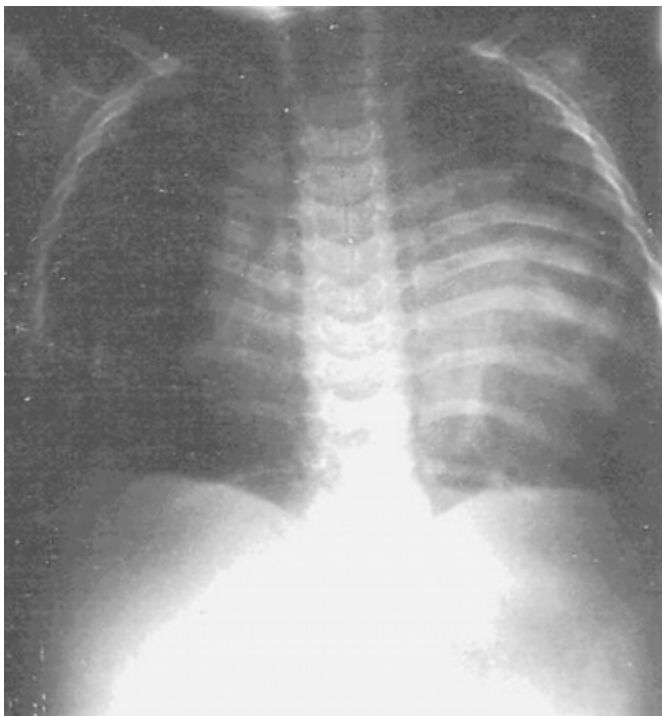
**Figure 1**

Figure 1: X Ray in the neonatal period



**Figure 2**

Figure 2: X Ray at presentation



### DISCUSSION

Isolated stenosis of pulmonary veins, as they enter into the left atrium, is a rare congenital cardiac abnormality. It is a

serious lesion that has significant impact on clinical outcome. Significant and progressive pulmonary hypertension leads to rapid deterioration and death often before infancy, hence early recognition is vital for optimum management.

In our case, right heart failure was initially attributed to pulmonary hypertension secondary to chronic lung disease. With a background of early prematurity, extended periods of ventilation and prolonged oxygen support, a similar clinical scenario is not uncommon in current paediatric practice. A history of wheezing also prompts clinicians to think primarily of chronic lung disease.

Symptoms of pulmonary vein stenosis are also quite similar, i.e., respiratory distress and evolving cardiac failure. In the early neonatal period, the condition has increasingly been recognized as a cause of persistent pulmonary hypertension of the newborn,<sup>1</sup> in the more acute setting.

Symptoms and clinical signs of pulmonary vein stenosis are very similar to those of chronic lung disease and are very difficult to differentiate. Diagnosis is difficult to suspect, especially in the absence of other cardiac abnormalities. There are reports when pulmonary vein stenosis was not suspected clinically and was detected only on autopsy.<sup>2</sup> Persistent respiratory symptoms, no response to antibiotic therapy and presence of differential radiological changes on chest X-Ray may give a clue. Confirmation is by cardiac catheter studies documenting pulmonary hypertension and elevated pulmonary capillary wedge pressure in the absence of left ventricular inflow and outflow obstruction. Angiography also helps to clinch the diagnosis.

It is important to be aware of the condition as newer modalities of management have emerged, including transcatheter interventions and newer surgical techniques. Having a “normal” heart scan initially does not rule out pulmonary vein stenosis presenting later on.<sup>3</sup> However, the lesion is usually associated with other heart defects,<sup>4</sup> (most common lesions are atrial septal defect and persistent ductus arteriosus) and a very strong degree of suspicion must be maintained on all cases of pulmonary hypertension. All infants with suspected pulmonary hypertension must be evaluated echocardiographically to exclude the lesion and prompt referral to the cardiologists is desirable.

Prognosis of pulmonary vein stenosis is guarded. Both mortality and morbidity are related to the number of stenosed vessels.<sup>4</sup> Recurrence of stenosis, which is a dreaded

complication after surgery, is notoriously common. It is still not clear why it happens, but neoproliferation is thought to be a factor<sup>5</sup>, suggesting antiproliferative therapy might have a role in the management to prevent recurrence in the future.

### **References**

1. Holcomb RG, Tyson RW, Ivy DD, et al. Congenital pulmonary venous stenosis presenting as persistent pulmonary hypertension of the newborn. *Pediatr Pulmonol.* 1999; 28(4): 301-6.
2. Bini RM, Cleveland DC, Ceballos R, et al. Congenital pulmonary vein stenosis. *Am J Cardiol.* 1984 ; 54(3): 369-75.
3. Minich LL, Tani LY, Breinholt JP, et al. Complete follow-up echocardiograms are needed to detect stenosis of normally connecting pulmonary veins. *Echocardiography* 2001;18(7):589-92.
4. Breinholt JP, Hawkins JA, Minich LA, et al. Pulmonary vein stenosis with normal connection: associated cardiac abnormalities and variable outcome. *Ann Thorac Surg.* 1999; 68(1): 164-8.
5. Sadr IM, Tan PE, Kieran MW, et al. Mechanism of pulmonary vein stenosis in infants with normally connected veins. *Am J Cardiol.* 2000; 86(5):577-9.

**Author Information**

**Santabhanu Chakrabarti**

Department of Paediatric Cardiology, Wessex Cardiothoracic Centre, Southampton General Hospital

**Sabrina Tsao**

Department of Paediatric Cardiology, Wessex Cardiothoracic Centre, Southampton General Hospital

**Joseph J. Vettukattil**

Department of Paediatric Cardiology, Wessex Cardiothoracic Centre, Southampton General Hospital

**James P. Gnanapragasam**

Department of Paediatric Cardiology, Wessex Cardiothoracic Centre, Southampton General Hospital