

## CLINICAL OBSERVATIONS

## Pulmonary vein stenosis mimicking chronic lung disease

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The presence of recurrent respiratory symptoms and right heart enlargement in an ex-premature infant is suggestive of chronic lung disease. Pulmonary vein stenosis is a rare, progressive disorder that produces similar symptoms and signs. A case is reported in which pulmonary vein stenosis was revealed by Doppler echocardiography, and this application is recommended in similar cases. Pulmonary vein stenosis is an extremely rare and progressive disorder, which, if left untreated, is usually fatal. The possibility of this diagnosis may not be considered during clinical examination and may be overlooked during routine echocardiography.

**Conclusion:** This report describes a patient with pulmonary vein stenosis in whom the diagnosis was delayed as she had symptoms and signs more commonly associated with chronic lung disease.

**Key words:** *Chronic lung disease, pulmonary vein stenosis*

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## Case report

An 11-month-old female infant presented with poor feeding and shortness of breath of 4 week duration. Born at 24 weeks of gestation, the infant had been ventilated for 5 weeks and had been oxygen dependent for nearly 3 months. Apart from a ductus arteriosus (which had been treated medically during the neonatal period), the infant appeared to have a structurally normal heart on echocardiography. However, she had frequent respiratory symptoms requiring repeated hospital admissions from the age of 4 months.

On admission at 11 months of age, the infant was in severe respiratory distress, with oxygen saturations of 50% in air. A third heart sound and prominent hepatomegaly were present. There was evidence of right atrial and right ventricular enlargement on the electrocardiogram. A significantly increased cardiothoracic ratio was observed on X-ray (Figs 1 and 2) and a clinical diagnosis of right heart failure secondary to chronic lung disease was made. The patient was treated with diuretics.

Severe right atrial and right ventricular dilatation with poor right ventricular function was confirmed by echocardiography. Marked tricuspid regurgitation and right to left shunting across the patent foramen ovale suggested significant pulmonary hypertension. Colour Doppler studies showed a high velocity pulmonary venous flow through the pulmonary veins, on both sides.

In the absence of an intra-cardiac shunt to increase the pulmonary blood flow, this was highly suggestive of pulmonary venous obstruction.

Cardiac catheter studies revealed severe pulmonary vein insertion stenosis, particularly on the left side, with resultant significant pulmonary hypertension. Surgical repair of the stenotic pulmonary veins was carried out, but the stenosis recurred. Following repeat surgery, the vein stenosis recurred and the patient died a few months later.

## Discussion

Isolated stenosis of the pulmonary veins, at the site of entry into the left atrium, is a rare congenital cardiac abnormality. It is a serious lesion that has a significant impact on quality of life and life expectancy. Significant and progressive pulmonary hypertension leads to rapid deterioration and, in many cases, death occurs during infancy.

In the case described in this report, right heart failure was initially attributed to pulmonary hypertension secondary to chronic lung disease. With a background of extreme prematurity, extended periods of ventilation and prolonged oxygen support, it is not uncommon for infants to present with similar symptoms due to underlying respiratory disease. Recurrent wheezing is compatible with chronic lung disease in the ex-premature

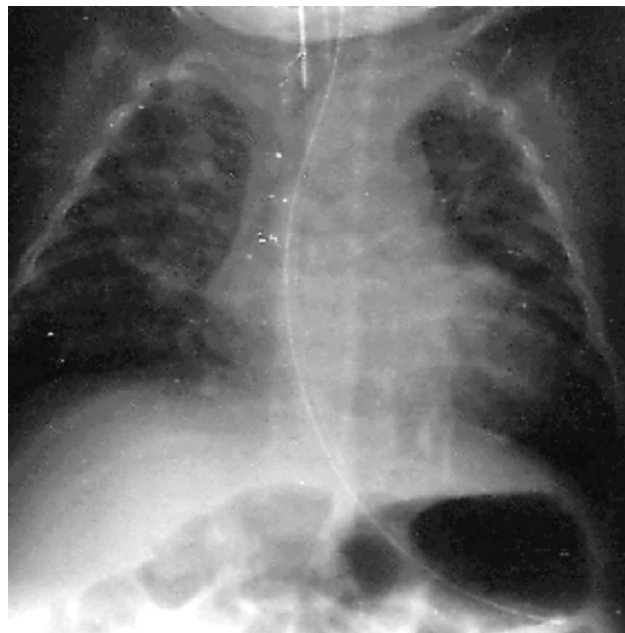


Fig. 1. X-ray in the neonatal period.

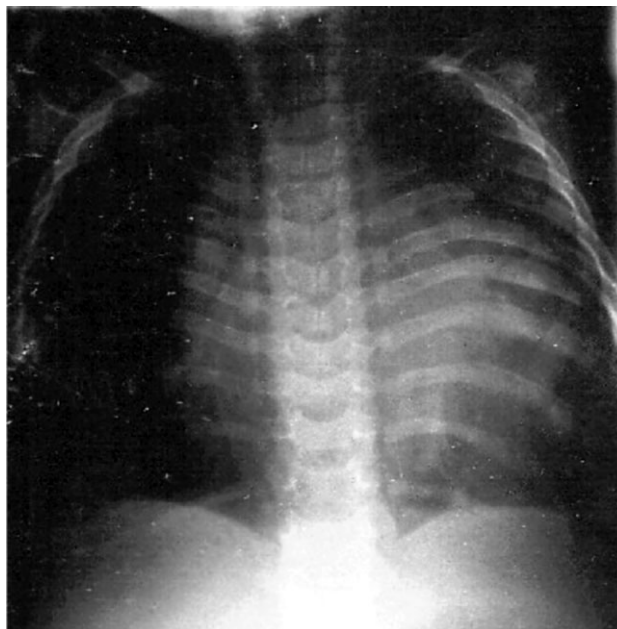


Fig. 2. X-ray at 11 months of age.

infant. Severe chronic lung disease can also lead to pulmonary hypertension and right heart dilatation.

Symptoms and signs of pulmonary vein stenosis are very similar to those of chronic lung disease and it may be very difficult to differentiate them. The possibility of a primary cardiac pathology may not be suspected, as the infant may not have a heart murmur on auscultation. Pulmonary vein stenosis has also been reported as a rare cause of persistent pulmonary hypertension of the newborn (1) in a more acute setting. In some cases pulmonary vein stenosis may not be suspected clinically and may be detected only on autopsy (2).

Persistent respiratory symptoms, no response to antibiotic therapy and presence of cardiomegaly and pulmonary venous congestion (either focal or lobar changes) on chest X-ray may provide a clue to the diagnosis. Confirmation is by cardiac catheter studies demonstrating pulmonary hypertension, elevated pulmonary capillary wedge pressures and stenotic veins on angiography.

It is important to be aware of the condition, as newer modalities of management have emerged, including transcatheter interventions and newer surgical techniques. Previous reports have shown that a normal heart scan in the neonatal period does not rule out pulmonary vein stenosis presenting later (3). The lesion is usually associated with other heart defects (4) such as atrial septal defect and persistent ductus arteriosus. As our case shows, it is essential to consider the diagnostic possibility in all cases of pulmonary hypertension in

infancy. We suggest that all infants with features of pulmonary hypertension must be evaluated echocardiographically, with documentation of the pulmonary venous inflow patterns.

Prognosis of pulmonary vein stenosis is guarded. Both mortality and morbidity are related to the number of stenosed vessels (4). Recurrence of stenosis is unfortunately very common after surgical repair. The reason for this is not clear, but intimal neoproliferation is thought to be a factor (5), suggesting that anti-proliferative therapy may have a role in the management to prevent recurrence in the future.

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