

Pulmonary hypertension secondary to pulmonary veno occlusive disease: Catastrophe in the catheterization laboratory

Sir,

Pulmonary veno occlusive disease (PVOD) is rare form of pulmonary hypertension (PH) and classified in group 1.6. PVOD carries worse prognosis due to limited response to pulmonary vasodilators. We present a case of PVOD, who was misdiagnosed as idiopathic pulmonary artery hypertension (PAH) and had a catastrophic

episode of pulmonary oedema during acute vasodilator testing (AVT).

A 5-year-old child presented with easy fatigability and had one episode of syncope. Clinically the child had severe PAH and the baseline oxygen saturation was 95%. Echocardiogram confirmed the diagnosis of PAH with moderate right ventricular systolic dysfunction. Initial

high resolution computerized tomography (HRCT) was reported to be unremarkable. The child was on diuretics and sildenafil, and was referred in view of unsatisfactory clinical response. Cardiac catheterization with AVT was performed at our centre under conscious sedation and local anaesthesia. Baseline catheterization data is mentioned in Table 1. On initiation of inhaled nitric oxide (NO) @ 20 ppm for AVT, pulmonary artery pressure increased, and oxygen saturations decreased requiring intubation. Bilateral extensive crepitations with frothy sputum was noted, suggesting flash pulmonary oedema. Inhaled NO was stopped, and the child was stabilized with intravenous diuretics and inotropes. Chest X-ray was suggestive of pulmonary oedema which cleared on follow up X-ray [Figure 1a -c]. Repeat HRCT chest revealed multifocal ground glass opacities, thickening of interlobar fissures, prominent pulmonary arteries and normal caliber pulmonary veins, highly suggestive of PVOD [Figures 2 and 3]. Patient was discharged on diuretics and referred for lung/heart lung transplant.

PVOD accounts for 3%–12% of patients labelled as “idiopathic PAH.”^[1-3] It could be inherited as autosomal recessive or acquired secondary to exposure to inhaled toxins and chemotherapeutic agents.^[1] No history suggestive of either hereditary PAH or toxin exposure was noted in our patient. Apart from mild baseline desaturation which increases on exercise, clinical features and baseline catheterization data of PVOD are indistinguishable from other causes of PAH. Although the anatomical obstruction in PVOD is in the postcapillary vascular bed, the pulmonary artery wedge pressure is often normal.^[4] AVT must be avoided in cases of suspected PVOD due to high incidence of pulmonary oedema.^[5] On HRCT, a triad of mediastinal lymphadenopathy, centrilobular ground-glass opacities, and thickening of the interlobular septa is the hallmark of PVOD.^[1] Although reduction in diffusion capacity of the lung for carbon monoxide and pulmonary haemorrhage in broncho-alveolar lavage have been described, they are nonspecific and difficult to perform in children.^[4] Lung biopsy is the gold standard for diagnosis of PVOD,

which is limited by high risk involved.^[5] Data on specific PAH therapies in PVOD are weak and conflicting, pulmonary oedema has been reported with use of pulmonary vasodilators.^[4] Although, Lung or heart-lung transplantation is the only definitive therapy it has guarded long term prognosis.^[5]

Our patient’s clinical symptoms, echocardiography, and baseline cardiac catheterization finding were consistent with that of PAH. Development of flash pulmonary oedema on AVT and HRCT was characteristic of PVOD. This case emphasises the importance of HRCT in an experienced centre prior to AVT and use of short acting agents like inhaled NO.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Table 1: Baseline cardiac catheterization data

	Pressure (mmHg)	Saturation (%)
SVC		70.3
RA (a , v , mean)	20/18/15	
LVEdp	4	
PA	65/28/46	56.2
PAW	4	
Aorta	78/52/64	94.6
Qp (l/min/m ²)		3.96
Qs (l/min/m ²)		4.13
Qp/Qs		0.96
PVRi (woodsunit.m ²)		10.6
SVRI (woodsunit.m ²)		11.9
Rp/Rs		0.89

SVC: Superior venacava, RA: Right atrium, LVEdp: Left ventricular end diastolic pressure, PA: Pulmonary artery, PAW: Pulmonary artery wedge, Qp: Pulmonary blood flow indexed to body surface area, Qs: Cardiac index, PVRi: Pulmonary vascular resistance indexed to body surface area, SVRI: Systemic vascular resistance indexed to body surface area, Rp/Rs: Ratio of pulmonary to systemic vascular resistance

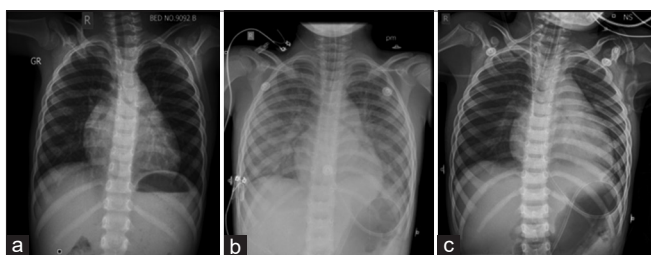


Figure 1: (a) Baseline chest X-ray done prior to cardiac catheterisation suggestive of mild cardiomegaly with dilated right atrium, (b) chest X-ray done immediately after cardiac catheterisation suggestive of pulmonary oedema, (c) chest X-ray done prior to extubating shows pulmonary oedema has resolved

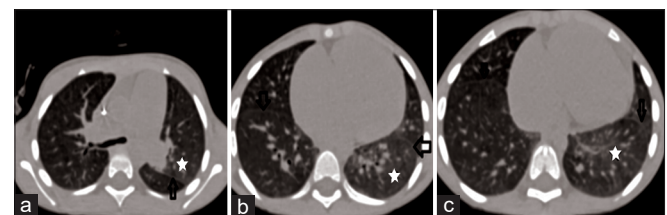


Figure 2: (a-c) Computerized tomography images showing multifocal ground glass opacities with thickening of interlobar fissures (Arrow: Interlobar fissure thickening, Asterisk: Ground glass opacity)

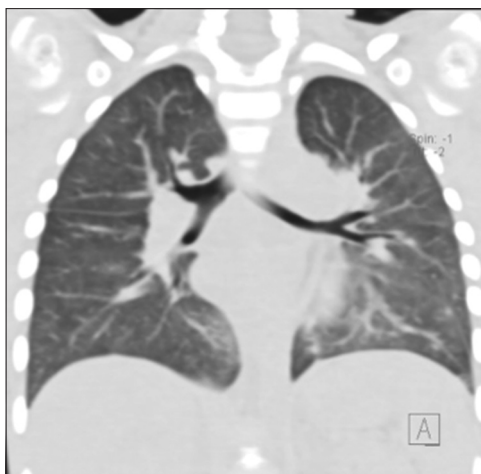


Figure 3: Coronal image obtained from computerized tomography showing ground glass appearance with prominent interlobar fissures

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Conflicts of interest

There are no conflicts of interest.

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