Cardiology in the Young

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Operable ventricular septal defect despite severe pulmonary hypertension and cyanosis!

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Abstract

Patients with a significant left-to-right shunt at ventricular level may become inoperable at an early age due to irreversible pulmonary vascular disease. On the other hand, even suprasystemic pulmonary hypertension due to mitral stenosis remains treatable. We report a 24-year-old patient with large ventricular septal defect, severe mitral stenosis and cyanosis who improved after surgical correction of both the lesions. This emphasises the importance of additional post-capillary pulmonary hypertension in Eisenmenger syndrome.

Brief Report

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Brief report

Patients with ventricular septal defect and a right-to-left shunt due to pulmonary hypertension are inoperable. The impact of additional post-capillary hypertension in such patients has not been well appreciated. We previously reported a patient with large patent ductus arteriosus with severe mitral stenosis, severe pulmonary arterial hypertension and differential cyanosis, who was successfully operated.[1] We report a similar patient with ventricular septal defect, severe mitral stenosis, severe pulmonary arterial hypertension with cyanosis to re-emphasise the importance of additional post-capillary pulmonary hypertension in Eisenmenger syndrome.

Case report

A 24-year-old female presented with complaints of dyspnea on exertion since childhood and palpitations on exertion. On examination, patient had cyanosis and clubbing with oxygen saturation of 81%. On auscultation, there was a loud pulmonic component of second heart sound and a mid-diastolic murmur at apex. Chest X-ray (Fig 1a) showed a dilated left atrium and evidence of pulmonary venous hypertension. Two-dimensional echo revealed large mid-muscular ventricular septal defect shunting bidirectionally with two additional small mid-muscular ventricular septal defects, severe mitral stenosis due to parachute mitral valve with a mean diastolic gradient of 11 mmHg, mitral valve area of 0.5 cm^2 by pressure half time method, associated mild mitral regurgitation and an enlarged left atrium. Catheterisation with vasoreactivity testing, under conscious sedation, had been done at another hospital just prior to patient's first visit to our centre. Baseline pulmonary artery pressure was 112/84 mmHg against a systemic pressure of 126/60 mmHg with an indexed pulmonary vascular resistance of 25.2 Woods units × m^2 and ratio of pulmonary to systemic vascular resistance of 1.0. Post-oxygen administration, there was no significant change in pressures or systemic saturation (81–86%) but calculated pulmonary vascular resistance index decreased to 16.8 Woods units × m^2 with a ratio of pulmonary to systemic vascular resistance of 0.6. Patient had been denied corrective surgery due to the pulmonary hypertension being apparently irreversible. Based on our familiarity with similar situation, we suggested corrective operation.[1] Patient underwent ventricular septal defect closure and implantation of a mechanical bileaflet mitral valve (#25 mm St. Jude Medical; St. Paul, Minn, USA). The two additional small mid-muscular ventricular septal defects were not closed in view of severe pulmonary hypertension (Supplementary video 1). She had a usual perioperative course with no episodes of pulmonary hypertensive crisis.

Patient had a remarkable symptomatic relief and reduction in pulmonary arterial pressures (Fig 2). The post-operative X-ray is shown in Fig 1b. A repeat hemodynamic study was done after 1 year of operation (Table 1), and the patient showed significant decrease in pulmonary arterial hypertension, although indexed pulmonary vascular resistance was still elevated. Patient did receive Bosentan, only in the post-operative period for a year and later declined. The further characterisation of pulmonary vascular resistance in future will be of interest, but the dramatic decrease in pulmonary vascular resistance and the clinical benefits in the patient are quite similar to our previous report.[1] At the latest follow-up, almost 18 months after operation, she is in NYHA class 1, and the echocardiogram shows small streaks of ventricular septal defect (VSD) flow and mild tricuspid regurgitation with velocity of 3.47 m/s (Supplementary material 1) with normal right ventricular size and function.

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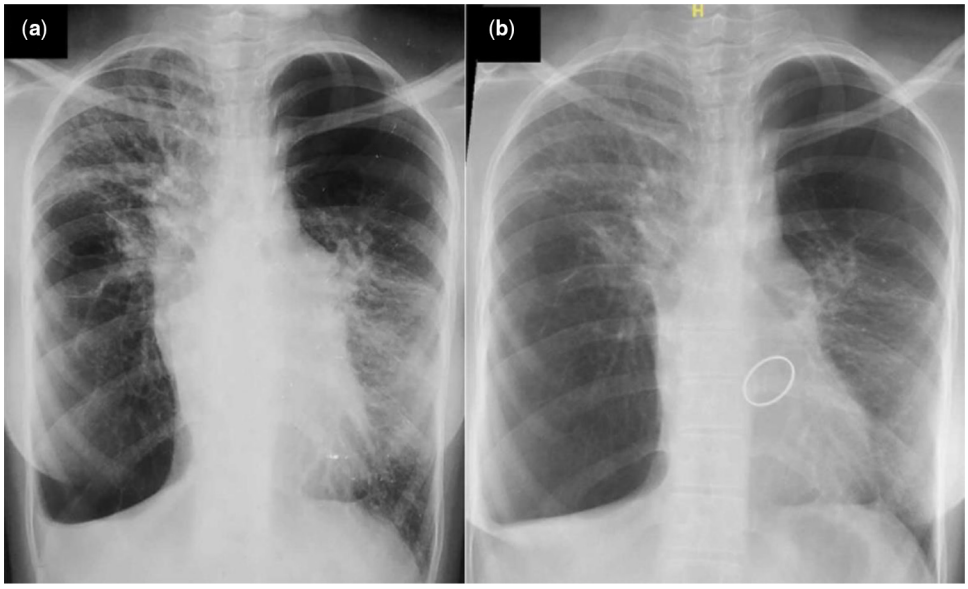

Figure 1. Chest X-ray in the posteroanterior view. (*a*) Pre-operative film showing pulmonary venous hypertension. (*b*) Post-operative film with mechanical prosthesis in the mitral position.

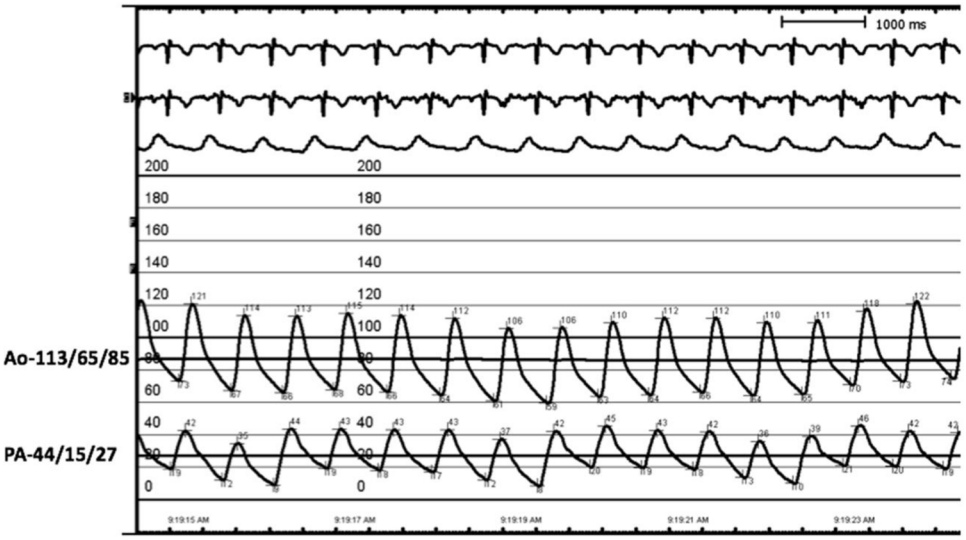

Figure 2. Simultaneous pressure tracings of the pulmonary artery and aorta 1 year after surgery. Pulmonary artery pressures dropped to 44/15 mmHg.

Table 1. Cardiac catheterisation data before and after surgery

	Before	After
RV (Sys/edp) (in mmHg)	126/6	48/6
PA (Sys/dia/mean) (in mmHg)	112/84/98	44/15/27
PA saturation (%)	63	62
PAWP (mean) (in mmHg)	28	8
Ao (Sys/dia/mean) (in mmHg)	126/60/82	113/65/85
Ao saturation (%)	81	99
PVRI (Woods units × m^2)	25.2	10.2
PVRI/SVRI	1.0	0.2

Ao = aorta; dia = diastolic pressure; edp = end diastolic pressure; PA = pulmonary artery; PAWP = pulmonary artery wedge pressure; PVRI = Pulmonary Vascular Resistance Index; RV = right ventricle; SVRI = Systemic Vascular Resistance Index; Sys = systolic pressure.

Pulmonary arterial hypertension in patients with mitral stenosis is secondary to passive transmission of elevated left atrial pressure as well as a reactive component.[2,3] However, it is generally reversible on treating mitral stenosis and is not accompanied by higher grades of structural changes in the lungs. [4–6] The presence of pulmonary arterial hypertension due to mitral stenosis in patients with shunt lesion leads to reversal of shunt – Eisenmenger syndrome – even when the changes are not irreversible. In that sense, mitral stenosis somehow is protective for the patients with shunt lesions and as is unequivocally documented by these cases. Patients are usually operable despite severe pulmonary arterial hypertension and right-to-left shunt. Even though the PA pressures may not completely normalise, the remarkable decrease in PA pressures would alter the clinical course favourably. Whether the pulmonary hypertension would recur on long-term follow-up remains to be determined.

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Although we do not have reports of lung biopsy in this case, the clinical course of events is the proof of concept of the protective effect of post-capillary pulmonary hypertension. The documentation of such cases is important to draw attention towards the knowledge gaps in this area. Further studies on the impact of additional post-capillary pulmonary hypertension in Eisenmenger syndrome are warranted.

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Conflicts of Interest. None.

Ethical Standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant Indian guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2013, and a need for formal approval was waived off by the Ethics Committee of All India Institute of Medical Sciences, New Delhi, India.

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