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Brief Report

Operable patent ductus arteriosus even with differential cyanosis: a case of patent ductus arteriosus and mitral stenosis

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Abstract Patients with patent ductus arteriosus and significant left-to-right shunt develop Eisenmenger syndrome at an early age and are not operable after development of irreversible pulmonary artery hypertension. Patients with mitral stenosis, however, are treatable even with suprasystemic pulmonary artery pressures. A combination of these two lesions is rare. We document a patient with differential cyanosis who improved after corrective surgery of both the lesions. The importance of post-capillary pulmonary artery hypertension in shunt lesions needs to be better appreciated.

Keywords: Patent ductus arteriosus; mitral stenosis; pulmonary artery hypertension

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Patients with large patent ductus arteriosus develop irreversible pulmonary vascular changes early in life. Once the changes are severe enough to cause right-to-left shunt and differential cyanosis, they are deemed inoperable.[1] Patients with mitral stenosis with severe pulmonary artery hypertension, however, are treatable even with suprasystemic pulmonary artery pressures and markedly raised pulmonary vascular resistance.[2] A combination of patent ductus arteriosus and mitral stenosis, therefore, poses an interesting and unusual therapeutic dilemma. In this article, we report one such case with a view to document the successful outcome.

Case report

An 8-year-old child presented with a history of recurrent lower respiratory tract infection since infancy. There was no history of cyanosis. On examination, there was differential cyanosis with upper-limb saturation of 94% and lower-limb saturation of 68%. His heart rate was 89/minute, and his blood pressure was 104/60 mmHg. There was a mid-diastolic murmur at the apex along with a narrow split second heart sound and a loud P2 component. Clinically, he did not experience congestive heart failure. There was no past history of rheumatic fever. His electrocardiogram showed right-axis deviation and right ventricular hypertrophy. His chest X-ray showed left atrial enlargement with pulmonary venous hypertension (Fig 1). An echocardiogram showed thickened mitral leaflets with commissural fusion and subvalvular deformity. The mitral valve area was 0.5 cm^2. There was a large patent ductus arteriosus with bidirectional shunt. A cardiac catheterisation was performed under conscious sedation (Table 1). His pulmonary artery pressure and wedge pressures were markedly elevated. His calculated pulmonary vascular resistive index fell from 26 to 19 Woods units $\times$ m^2 after 10 minute of oxygen. A left ventricle angiography showed large patent ductus arteriosus with bidirectional shunting (Supplementary video 1). There were no posterior aortic shelf or withdrawal gradients to suggest coarctation of the aorta.

Even though the baseline pulmonary vascular resistive index was high and there was no significant fall after oxygen administration, suggesting irreversible pulmonary artery hypertension, it was decided to operate the patient (discussed later). The patient underwent open mitral commisurotomy and ligation of patent ductus arteriosus. His postoperative stay was uneventful. His mitral valve increased to

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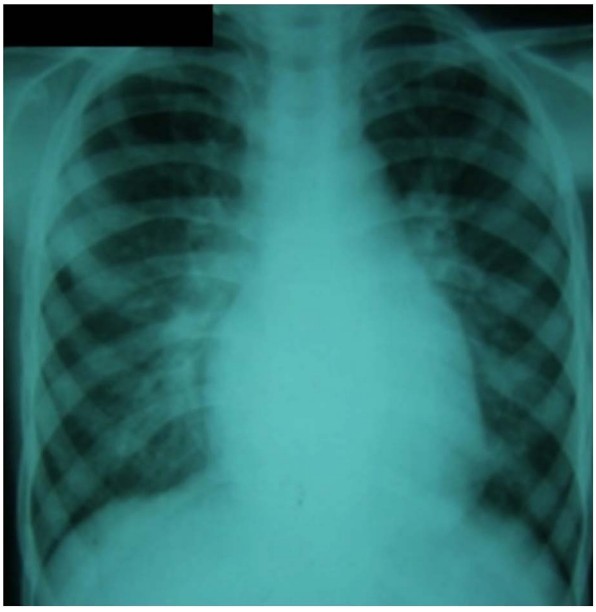

Figure 1.
Chest X-ray in the posteroanterior view showing left atrial enlargement and pulmonary venous hypertension.

Table 1. Cardiac catheterisation data.

	Pressure (mmHg)		
	Systolic/diastolic	Mean	Saturation (%)
RA	a8/v10	7	58
RV	106/8		
PA	108/76	90	62
Ao	116/76	92	66
PAWP	a45/v46	38	92

Ao = aorta; PA = pulmonary artery; PAWP = pulmonary artery wedge pressure; RA = right atrium; RV = right ventricle

2.47 cm^2 after surgery. He showed remarkable clinical improvement and was discharged in stable condition. He was lost to follow-up and reported 4 years later. A repeat catheterisation after 4 years showed remarkable fall in pulmonary artery pressures (Fig 2). The pulmonary artery pressures were 54/18 mmHg. The calculated pulmonary vascular resistive index dropped to 8 Wood units × m^2.

Discussion

Pulmonary artery hypertension in patients with mitral stenosis is well described and regresses after successful relief of mitral stenosis.[2,3] Even suprasystemic pulmonary artery hypertension regresses significantly after relief of obstruction in mitral stenosis. The pulmonary venous hypertension in mitral stenosis apart from passive transmission also triggers vasoactive and structural re-modelling changes in the pulmonary vasculature.[2] The advanced structural changes of plexiform lesions seen in pre-capillary hypertension are not seen, and pulmonary artery hypertension regresses as a rule.[4] A rapid regression is described following Balloon Mitral Valvotomy in the initial weeks, although it may continue for months.[5] In a minority of patients with pulmonary artery hypertension, the regression is suboptimal, and this may be due to inadequate relief of mitral stenosis, or long-standing structural changes; however, the cause is not clearly delineated.[6] The pulmonary artery hypertension from mitral stenosis may cause reversal of shunt in patients with coexisting patent ductus arteriosus or ventricular septal defects even if the changes in lung vasculature are not irreversible. It is therefore conceivable, as in this patient, to have right-to-left shunt from patent ductus arteriosus that does not mean irreversible or inoperable status. Post-capillary pulmonary artery hypertension therefore may be protective in patients with shunt lesions, and patients may be operable despite significantly high pulmonary vascular resistive index. Conventionally, a pulmonary vascular resistive index of 8 Wood units × m^2 or more is considered inoperable in lesions such as patent ductus arteriosus or ventricular septal defect;[7] however, as shown by the clinical course of the patient, the patient could be successfully operated, and showed mildly elevated pulmonary vascular resistance on follow-up.

The first case of this unusual combination was described by Mackinnon and Briggs in 1958.[8] Talwar et al[9] described a similar case of large patent ductus arteriosus and mitral stenosis with severe pulmonary artery hypertension, with pulmonary vascular resistive index in operable range due to high pulmonary capillary wedge pressure. Kalantre et al discussed similar issues of pulmonary venous hypertension in cases with operable complex congenital cyanotic heart disease and univentricular hearts. It was also shown that pulmonary venous hypertension was protective and allowed delayed cavopulmonary shunt.[10]

Whether mitral stenosis in our patient was congenital or was acquired is difficult to ascertain, but that was not the point of this report. We decided to operate on this patient because of the presence of severe mitral stenosis and familiarity with the concept of reversibility in this setting. There are no data available regarding the combination of shunt lesion with post-capillary pulmonary artery hypertension. This case is reported to enhance the understanding of the protective effects of post-capillary pulmonary artery hypertension in shunt lesions and operability. Although pulmonary vascular resistive index was not normal on follow-up, remarkable regression has been documented. The precise time course of regression was not well clarified as the patient

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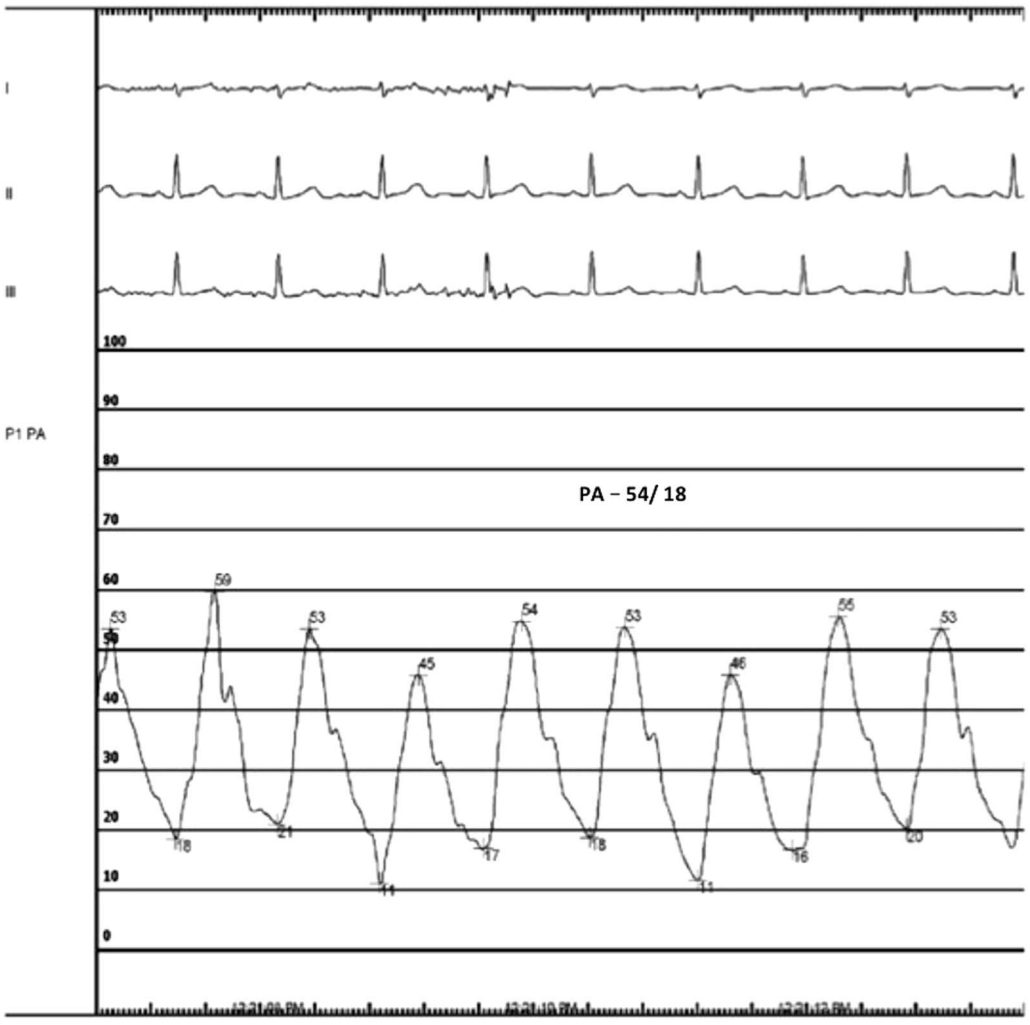

Figure 2.
Pressure tracings of the pulmonary artery (PA) 4 years after surgery. Pulmonary artery pressures dropped to 54/18 mmHg.

was lost to follow-up after operation. The pulmonary vascular resistive index after 4 years, however, was remarkably low compared with the preoperative status. The successful outcome of our patient adds to the belief of operability in such a scenario. The presence of mitral stenosis paradoxically benefitted the patient and allowed definitive treatment.

It is important to document such cases, as there is a lack of adequate information in the literature about the optimum management in such conditions. The patient could have been denied curative surgery if the treating physicians were not aware of this expected response. Similar response is expected with other post-capillary obstruction conditions such as cortriatriatum or supramitral ring in combination with left-to-right shunt lesions. We hope that this report will simulate further studies on the importance of post-capillary hypertension in patients with shunt lesions.

In conclusion, we documented an unusual case of patent ductus arteriosus with differential cyanosis and severe mitral stenosis who improved after corrective surgery.

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Conflicts of Interest

None.

Supplementary material

To view supplementary material for this article, please visit https://doi.org/10.1017/S1047951117001275

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