A Case of Severe Bilateral Peripheral Pulmonary Stenosis

Shalima Gautam+, Deepak Kumar Mishra***, BC Kalmath*, Ramesh Kawar*, Anil Sharma*, BK Goyal**

Abstract

Isolated PPS is known in children. Mild/Moderate PPS remains stable over time. Severe PPS is progressive and patients are symptomatic but catastrophic events are not known in children. This is a rare case of severe PPS in a child with sudden cardiac death.

Introduction

Stenosis of pulmonary artery isolated or in association with other cardiac defects occur in 2-3% of all cases of congenital heart disease. The stenosis may be single, involving the main pulmonary artery or its branches or multiple involving both the main and the smaller peripheral pulmonary artery branches. We report a case of severe bilateral peripheral pulmonary artery stenosis associated with patent ductus arteriosus and intact ventricular septum.

Case Report

This 7 year old boy presented with complaints of breathlessness of 6 months duration.

Examination revealed normal pulse and blood pressure. General physical examination was inconspicuous. Cardiovascular system examination revealed normal S1, second heart sound was narrowly split, and a continuous murmur heard in left infraclavicular region. An ejection systolic murmur grade 3/6 in intensity was heard in pulmonary area radiating to back, interscapular region and axilla. Other system examination was within normal limit. There was no history of antenatal Rubella and the developmental history was normal. ECG showed right ventricular hypertrophy with right axis deviation. Angiocardiography revealed severe bilateral pulmonary artery stenosis close to the bifurcation. The gradient across the RPA and LPA was 90 mm Hg and 110 mm Hg respectively. Percutaneous balloon angioplasty and stenting was planned in subsequent course of follow up but before it could be done, the child had sudden cardiac death.

Discussion

Isolated peripheral pulmonary artery stenosis (PPS) in children has been well described.

It is characterised by diffuse arterial involvement generally affecting the main and the lobar arteries and occasionally produces symptom of right ventricular failure when severe. The stenosis may be confined to trunk or main PA, multiple peripheral involving segmental pulmonary arteries, central and peripheral stenosis (Gays classification).

These lesions are frequently seen in association with other CHD or as a part of well defined syndromes (Williams, alagilles, noonans, congenital rubella, cutis laxa, ehlers danlos and silvers). The haemodynamic effects of these narrowings may be mild dyspnoea on exertion as presenting complaint as the RV pressure is suprasystemic.

Patients with mild to moderate stenosis
remain stable and rarely progress. This is particularly true for PPS associated with syndromes mentioned above.

PPS of severe degree may be progressive and complications include RV failure, PA thrombosis, post stenotic aneurysmal dilatation with pulmonary artery haemorrhage.

Although death in childhood is rare, severe PPS deserves management in the form of angioplasty or surgery.

This case is a rare presentation of severe pulmonary artery stenosis, rapid progression of symptoms and subsequent sudden cardiac death.

References

PALLIATIVE CARE
Three-quarters of patients wish to die at home. Liaison between the primary care team, patient, relatives and carers, outside agencies and secondary care is essential to the success of domiciliary palliative care.