Case Report

Critical decision of operability in congenital heart disease patient with severe pulmonary hypertension

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ABSTRACT

Repair of congenital heart disease in the presence of high pulmonary pressure has always been a contentious issue. Pulmonary vascular resistance (PVR) is considered important for establishing operability in these patients. However, PVR estimation is not always accurate and cannot solely be relied upon to make critical decision of operability. Clinical examination, chest X-ray, and echocardiography are also important indicators of pulmonary vascular disease. Knowledge of pits and falls of each investigation is important for appropriate management in these patients. We present a case report of successfully operated, 6-year-old child with anomalous origin of the right pulmonary artery from aorta, deemed inoperable on the basis of PVR estimation.

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INTRODUCTION

Patients with severe pulmonary arterial (PA) hypertension need to be evaluated thoroughly before being taken up for surgery. Pulmonary vascular resistance (PVR) is often considered to be gold standard for making such decisions. Therefore, it is important to realize the downside of it and role of other investigation in making critical decision of operability in these patients.

CASE REPORT

A 6-year-old female child presented with recurrent chest infections and failure to thrive. Examination revealed, resting saturation in the right upper limb and lower limb was 94% and 92%, respectively, apex beat in 4th intercostal space 2 cm lateral to the midclavicular line, Grade II parasternal heave with P2 palpable. Other organ system examination was otherwise normal. Chest X-ray showed cardiothoracic ratio of 52% with normal pulmonary vascularity. Electrocardiogram had signs of left ventricular volume overload. Echocardiography revealed anomalous origin of the right PA (RPA) from posterior aspect of aorta with right ventricular outflow tract continuing as left PA (LPA). There was a large patent ductus arteriosus (PDA) with bidirectional flow along with flow reversal in arch [Figures 1 and 2]. Cardiac catheterization done outside reported very high PVR of the left lung and right lung as 14.21 Wood units and 37.22 Wood units, respectively.

Review of literature revealed that PVR estimation is not always reliable. Therefore, plan was made to connect RPA to main PA and to assess LPA pressure after snugging of PDA. During surgery, temporary occlusion of PDA lead to fall in LPA pressure, which indicated the reversibility of high LPA pressure. It convinced

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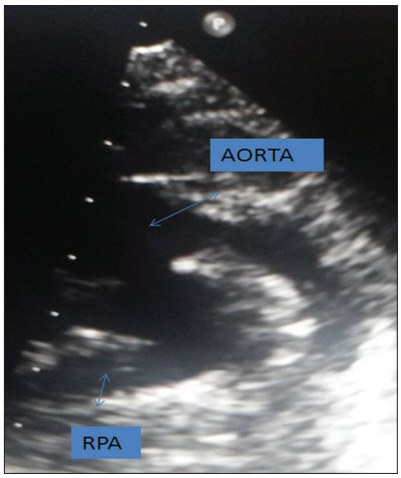

Figure 1: Anomalous origin of right pulmonary artery from aorta

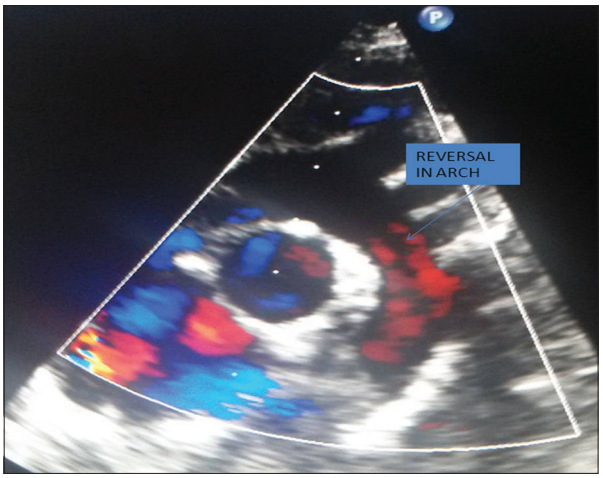

Figure 2: Flow reversal in arch

that both lungs will benefit from repair. Transesophageal echocardiography in the operation theater showed a tricuspid regurgitation of 25 mmHg (central venous pressure of 7 mmHg). The patient had a stable postoperative course and was discharged on the 5th postoperative day. The patient is doing well in follow-up presently, 6 months after surgery with mean PA pressure half of systemic pressure.

DISCUSSION

Operability in shunt lesions in the presence of high PA pressures is a contentious issue. Clinical assessment, chest X-ray, echocardiography, and catheterization are often used to make the final decision of operability. Catheterization study is often considered of utmost importance to make the decision of operability in the presence of severe PAH.

Catheterization study gives an accurate measure of PA pressures and PVR beside anatomical details. PVR and its reversibility on the administration of oxygen or NO is often considered as sign of operability.[1,2] PVR is calculated as (mean PA pressures-mean left atrial pressures/pulmonary blood flow, i.e. Qp) where Qp: Oxygen consumption (VO_2)/pulmonary vein O_2 content (ml/L) – PA O_2 content (ml/L). Anatomical substrate like truncus/hemitruncus/transposition of great arteries/aortopulmonary window often contains highly oxygenated blood in PAs. The narrow pulmonary venous - PA difference with respect to oxygen content, makes "oxygen" a poor substrate, to estimate pulmonary blood flow.[3] Therefore, inaccurate determination of Qp does not allow for true estimation of PVR in these cases. In our patient, RPA was originating from aorta, containing highly oxygenated blood. Therefore, PVR of the right lung was not accurate.

Furthermore, large shunts like PDA can also lead to false estimation of PVR of left lung.[4] Transient balloon occlusion of PDA has been described for true estimation of PA pressures and PVR calculations. PDA was not occluded to determine PVR of the left lung while doing catheterization study, leading to falsely high PVR of left lung. Intraoperative decrease in LPA pressure on occluding PDA confirmed our suspicion of false high estimation of PVR during catheterization. Therefore, correlation of catheterization study with other investigations becomes of paramount to determine operability.

In the present case, the patient was referred to our center as inoperable on the basis of catheterization study. However, absence of differential cyanosis, the presence of cardiomegaly on chest X-ray, continuous flow in RPA and flow reversal in arch (due to continuous flow in RPA from ascending aorta) convinced us that patient has reversible high RPA and can undergo at least single lung repair. The fall of LPA pressure on ligation of PDA during surgery further convinced us that both lungs would benefit from repair.

CONCLUSION

Catheterization study is not reliable to calculate PVR in congenital heart diseases where oxygen content in pulmonary vein and PA is similar. Inaccurate determination of pulmonary blood flow does in such cases does not allow for true estimation of PVR. The decision of operability should be made on basis of multiple investigation modalities in such case.

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Conflicts of interest

There are no conflicts of interest.

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