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CASE REPORT

Surgical Management of Aortopulmonary Window with Tetralogy of Fallot

Amitabh Satsangi[1], B. Kanmaniyan[2], Pratik Jha[3]

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ABSTRACT

Background: Aortopulmonary window with Tetralogy of Fallot is a rare cardiac defect with very few reported cases in literature.

Case: We report surgical management of incidentally detected type 1 aortopulmonary window in a case of tetralogy of Fallot during routine intraoperative transesophageal echocardiography.

Conclusion: Aortopulmonary window with Tetralogy of Fallot is a rare entity and the two pathologies mask the signs and symptoms of each other. Surgical correction is feasible and associated with good outcome.

KEYWORDS

• Transesophageal echocardiography • Aortopulmonary window • Tetralogy of fallot • Congenital heart disease

AUTHOR'S AFFILIATION:

[1] Assistant Professor, Department of Cardiothoracic and Vascular Surgery, All India Institute of Medical Sciences, New Delhi, India.

[2] Senior Resident, Department of Cardiothoracic and Vascular Surgery, All India Institute of Medical Sciences, New Delhi, India.

[3] Senior Resident, Department of Cardiothoracic and Vascular Surgery, All India Institute of Medical Sciences, New Delhi, India.

CORRESPONDING AUTHOR:

Amitabh Satsangi, Assistant Professor, Department of Cardiothoracic and Vascular Surgery, All India Institute of Medical Sciences, New Delhi, India.

E-mail: amitabhs@aiims.edu

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INTRODUCTION

Tetralogy of Fallot (TOF) is one of the most prevalent cyanotic congenital heart illnesses, constituting approximately 10% of all congenital cardiac anomalies.[1] Associated anomalies with TOF include ASD, patent ductus arteriosus, pulmonary artery variations, atrioventricular canal defects, coronary artery anomalies, aortic arch anomalies. Another rare association of TOF is with aortopulmonary window (APW) with very few reported cases so far.[2,3] We present a rare case of TOF with APW and its successful surgical management.

CASE DETAILS

A 11-month male child, weighing 7kg presented with feeding diaphoresis, suck-rest-suck cycle and recurrent lower respiratory tract infections since birth. The parents give history of multiple hospital admissions for lower respiratory tract infections, with history of poor feeding and poor weight gain. The child was diagnosed to have a congenital heart disease and was referred to our centre for further evaluation and management. Transthoracic echocardiography (TTE) revealed situs solitus, levocardia, concordant atrio-ventricular and ventriculo arterial connections, normal relation of great arteries. The TTE also revealed a single, large mal-aligned ventricular septal defect (VSD) with bidirectional shunt flow and features of left ventricular volume overload. Deviation of conal septum was also noted without any pressure gradient. The pulmonary valve was dysplastic with dilated and confluent branch pulmonary arteries. The child was planned for VSD closure and right ventricular outflow tract (RVOT) inspection. Room air saturation was 78%.

Patient was taken up for elective intracardiac repair for maligned ventricular septal defect and pulmonary valvotomy. Intraoperative transoesophageal echocardiography (TEE) was performed which revealed a single sub-aortic VSD, 1.1cm in size, shunting from left to right (Figure 1) with aortic override. A 0.3cm ostium secundum atrial septal defect was noted. The pulmonary valve appeared dysplastic, with annulus measuring 1cm. Mild tricuspid regurgitation was noted. Biventricular function was normal. As part of comprehensive intraoperative TEE, a communication between the ascending aorta and the main pulmonary artery was noted in the mid-oesophageal ascending aorta short axis view which was not reported in the TTE. This measured around 1 cm with shunting of blood from left to right (Figure 2). This finding was communicated to the operating surgeon. Pulmonary artery acceleration time of 65ms was calculated.

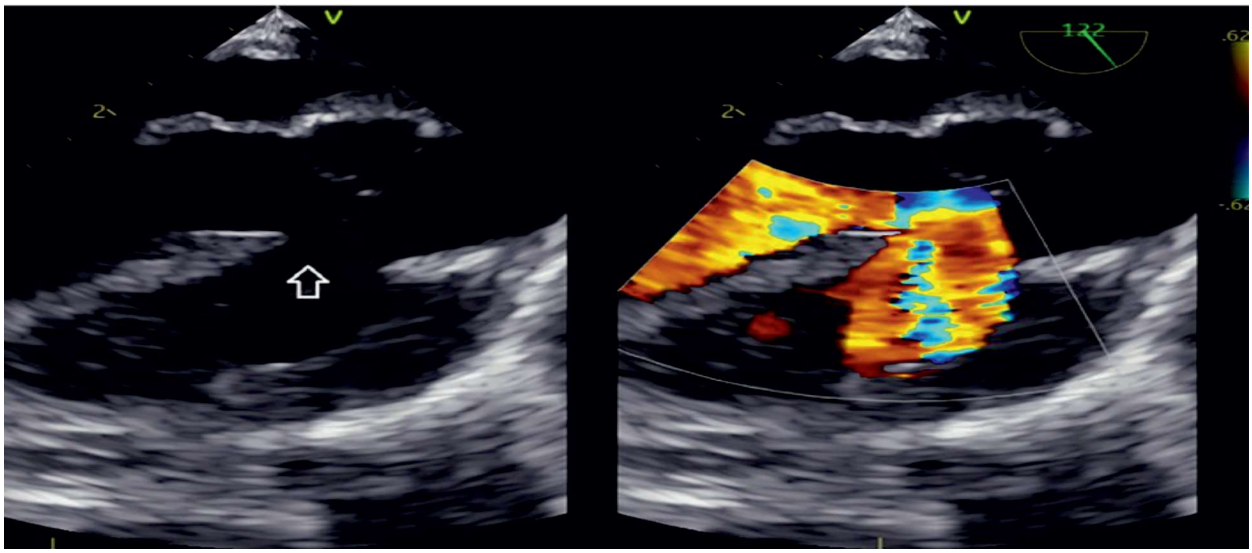

Figure 1: A simultaneous color doppler image showing mid-esophageal aortic valve long axis view, with the arrow pointing toward the sub-aortic VSD and shunt flow

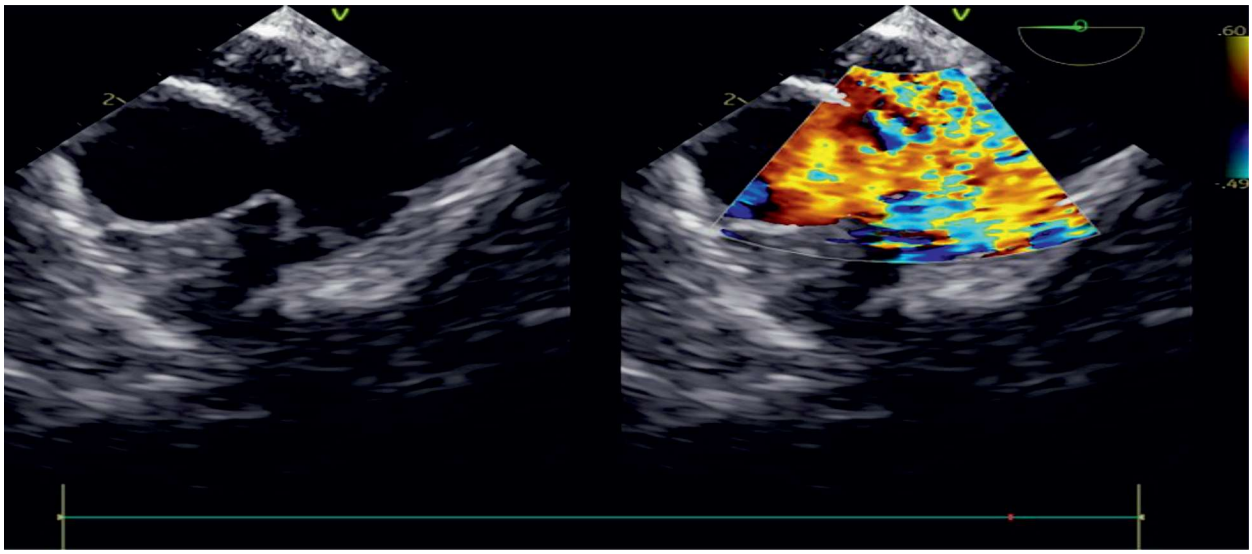

Figure 2: Mid-esophageal ascending aorta short axis view showing the communication between the aorta and the main pulmonary artery with color doppler showing the flow across it

Median sternotomy was performed, thymus tissue was excised and a right vertical pericardiotomy was done. The heart was inspected with a communication noted between the great vessels (Figure 3). After systemic heparinisation, routine cardiopulmonary bypass (CPB) was instituted with aorto-bicaval cannulation. Intraoperatively, type 1 aortopulmonary window was diagnosed and pre-CPB TEE finding of the aorto-pulmonary communication was confirmed. The aorta and pulmonary artery were dissected and the patent ductus arteriosus was clipped. Antegrade root cardioplegia was administered after aortic cross clamping and snaring of the branch pulmonary arteries. After administration of cardioplegia pulmonary artery was opened and the aorto-pulmonary window was examined. The surgical correction consisted of trans right atrial maligned VSD closure with a PTFE patch, pulmonary valvotomy and trans pulmonary patch closure of the aortopulmonary window (Figure 4). Total aortic cross clamp time was 57 mins and CPB time was 87 minutes. Nitro-glycerine (NTG) was started at 0.5mcg/kg/min and dobutamine at 5mcg/kg/min as inotropic support at termination of CPB. Post-CPB TEE revealed an intact patch with no residual VSD. There was mild pulmonary regurgitation. A *cul-de-sac* with no flow was noted at the place where aortopulmonary connection was present in the pre-CPB examination.

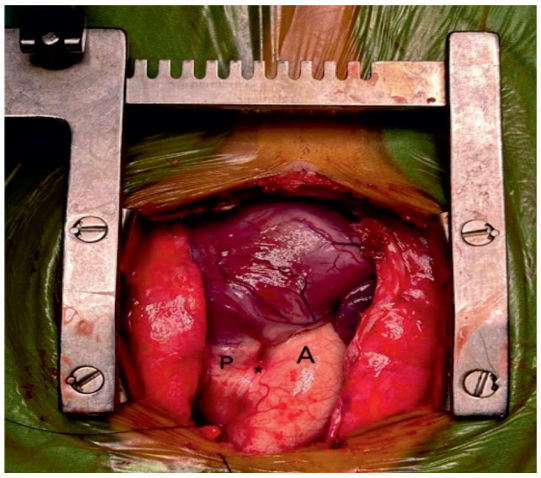

Figure 3: Intra-operative image showing the aortopulmonary communication. *, communication between the aorta and pulmonary artery; A, aorta; P, main pulmonary artery

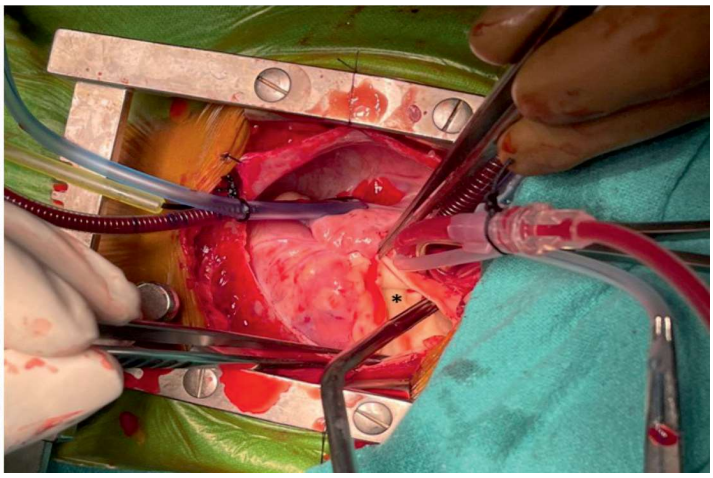

Figure 4: Transpulmonary artery view of aorto-pulmonary artery window (*)

DISCUSSION

The uncommon correlations of TOF and APW render it distinctive in its clinical manifestation. A left-to-right shunt caused by the APW will obscure the symptoms and clinical manifestations of pulmonary stenosis. The symptoms in our case indicated elevated pulmonary blood flow. Pulmonary hypertension resulting from elevated pulmonary flow can complicate perioperative care and outcomes. Management aims to reduce right ventricular afterload and attain pulmonary vasodilation. Administration of pulmonary vasodilators such as intravenous milrinone and sildenafil may be contemplated. A thorough TEE examination is essential to validate the preoperative TTE findings and to identify any potentially critical findings that may have been overlooked, which could impact patient outcomes. Over the years, surgical repair procedures for the aortopulmonary window have progressed from basic ligation, division, and suturing, to transpulmonary artery closure, anterior sandwich patch closure, transaortic direct closure, and ultimately transaortic patch closure.[4] In instances of related TOF, the surgical sequence involves the closure of the AP window, succeeded by intracardiac repair. In instances of related TOF, the pulmonary arteries are typically well-developed, allowing for the possibility of defect closure via the pulmonary artery approach to avoid the additional surgical step of aortotomy.

CONCLUSION

Aortopulmonary window with tetralogy of Fallot is a rare entity and can be successfully surgically corrected with good outcomes if patient presents early.

Conflicts of interest: None

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Ethics Declaration: Patient consent obtained from legal guardian and is available with the authors.

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