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Double Barrel Repair in a Child with Tetralogy of Fallot : The Role of Intraoperative Trans-esophageal Echocardiography- A Case Report

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Case Report

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Abstract

Introduction:

Double barrel repair is a surgical technique used to avoid injury to the coronary artery crossing the right ventricular outflow tract in individuals suffering from Tetralogy of Fallot. Though the surgical technique is widely known, the echocardiographic images during the procedure is scanty in the literature.

Case Presentation:

We describe the clinical details and the intraoperative evaluation of an eight year old boy with Tetralogy of Fallot and an anomalous coronary artery crossing the right ventricular outflow tract who underwent the double barrel repair surgery. Intraoperative TEE confirmed the TTE findings, and detected tricuspid valve hypoplasia (Annulus – 21 mm ; 16.4 mm / m2 ; - 1.8 z) and right ventricular diastolic dysfunction.

Conclusion:

Intraoperative TEE identified the hypoplastic pulmonary valve, right ventricular diastolic dysfunction and the anterior barrel flow-related gradient in our patient, in addition to confirming the TTE findings. It was instrumental in surgical planning, avoiding the BDG takedown and an unremarkable post-operative course.

Introduction

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart defect, with an incidence of three out of every 10,000 live births and a prevalence rate of 5–10% among all congenital heart defects [1]. 4–6% of the patients with TOF can have anomalous coronary arteries. In 72% of these patients, the anomalous coronary artery can cross the RVOT [2]. A variety of modified surgical techniques are developed to account for these anomalies during the intracardiac repair. We report a case of double barrel repair done for Tetralogy of Fallot in a child with an anomalous coronary artery crossing the RVOT. There are very few case reports including echocardiographic images of double barrel repair. This case report is submitted after written informed consent from the child's close family and does not require ethics committee approval.

Case Report

An eight year old boy was planned for Intracardiac repair for Tetralogy of Fallot. The child was diagnosed on day 4 of life as a case of TOF, after being evaluated for a murmur. He was scheduled for intracardiac repair at 2.5 years of age. Intraoperatively, the surgical plan was changed in view of inadequate calibre of the pulmonary arteries to bidirectional Glenn shunt (BDG) and bilateral pulmonary artery (PA) plasty. During follow up at 5 years of age, the child developed dyspnoea on exertion. Subsequent evaluation revealed the presence of major aortopulmonary collaterals (MAPCA) and MAPCA coiling was done. Two

years later he had recurrence of dyspnoea and was diagnosed to have left pulmonary artery(LPA) stenosis and LPA stenting was done. Following this he was electively planned for intracardiac repair.

Preoperative Transthoracic Echocardiography (TTE), showed a peri-membranous Ventricular Septal defect (VSD) with 50% aortic override, hypoplasia of the pulmonary annulus (5mm, -7 z) and main pulmonary artery (8.2mm, -6 z), confluent branch PAs, dilated aortic annulus (22.5mm, 2.3 z) and ascending aorta with mild aortic regurgitation. Computed Tomography showed the left anterior descending artery (LAD) arising from the Right Coronary artery (RCA) and crossing the RVOT (Fig. 1). The retrosternal anatomy was good with preserved fat planes posterior to all the five sternal wires.

Intraoperative TEE confirmed the TTE findings, and detected tricuspid valve hypoplasia (Annulus – 21 mm ; 16.4 mm / m2 ; – 1.8 z) and right ventricular diastolic dysfunction. After an uneventful sternotomy, cardiopulmonary bypass was instituted after systemic heparinization and aorto bicaval cannulation. VSD was closed through the right atrium. In view of the unfavourable coronary anatomy, enlargement of the pulmonary annulus by a trans annular patch was not a feasible option. To prevent further surgeries for potential conduit or homograft stenosis, a double barrel RVOT repair was done. RVOT was opened longitudinally below the anomalous coronary, with an incision parallel to the vessel. Septal and parietal bands were resected. MPA was then opened and a flat patch was placed between RVOT and MPA and then an additional patch was placed on top of it forming its roof, thus creating two barrels or outflows i.e., an anterior and posterior barrel (Fig. 2). The anterior barrel is the newly constructed outflow tract using patch and the posterior barrel is the native RVOT with the native pulmonary valve. The native valve was preserved by this technique as it had the potential to grow unlike the conduit which will likely develop stenosis over a period of time. After intracardiac repair, patient was gradually weaned from cardiopulmonary bypass, with inotropic supports of adrenaline 0.05 mics/kg/min, noradrenaline 0.05 mics/kg/min and milrinone 0.5 mics/kg/min.

After weaning from CPB, TEE confirmed the closure of VSD, pulsatile non-turbulent flow in the posterior barrel (native pulmonary annulus)(Video 1 B) with no PR, and turbulent flow was noted in the anterior barrel (pericardial patch)(Video 1 C) with a peak gradient of 60 mm Hg and free PR(Fig. 3A and B). RV needle pressures were consistently within 2/3rd of the systemic pressures (RV needle pressure 63/17 ; SBP 97 / 64), hence the anterior barrel was not revised. The initial plan of taking down the BDG was abandoned due to high gradients at the conduit, hypoplastic tricuspid valve and RV diastolic dysfunction, thus utilizing the BDG as a method to offload the RV. He was shifted to ICU with inotropic supports of adrenaline and noradrenaline at 0.05 mics/kg/min and milrinone at 0.5 mics/kg/min.

Discussion

Coronary artery anomalies (CAAs) or anomalous coronary artery (ACA) includes congenital conditions characterized by abnormal origin or course of any of the three main epicardial coronary arteries. They can be classified as anomalies of origin, of course, and of termination [3]. Prevalence of anomalous coronaries is more common in individuals with congenital heart disease than in individuals with a

structurally normal heart. During development, vascular sinusoids develops within the embryonic myocardium which develops into network of veins, arteries and capillaries. Endothelial buds appear at the base of truncus arteriosus which joins with the coronary artery network forming the definitive coronary artery circulation. Abnormal development or fusion of these endothelial buds or abnormal septation of truncus arteriosus leads to abnormal origin of coronary arteries [4]. Our patient had LAD arising from RCA and was crossing the RVOT which was a unique problem considering the hypoplastic pulmonary annulus which cannot be enlarged with standard techniques such as a transannular patch [5].

Another option is to create a RV – PA conduit to bypass the annular stenosis. It carries the risk of reoperations due to potential conduit stenosis or calcific degeneration. Double-barrel repair was formulated initially to address the anomalous coronary crossing infundibulum in a setting where there is limited availability of RV – PA conduits. This technique preserves the growth potential of the native annulus, as the annulus is not incised. Other advantages include delaying the pulmonary valve replacement, avoiding patient – conduit mismatch and lesser limitation of the retrosternal space than a conduit [6].

The anterior barrel will however, have free pulmonary regurgitation as it does not have a valve, unlike a valved conduit. This carries the risk of right ventricular failure as in the case of an ICR with a transannular patch, but since the annulus still has growth potential, volume of regurgitation may decrease over time. Another limitation described for this procedure is the potential for pulmonary artery distortion [6].

The Bidirectional Glenn shunt can help in unloading the right ventricle by diverting superior vena caval blood directly into the pulmonary artery, preventing RV volume overload. The use of BDG in cases of residual outflow obstruction after TOF repair has been reported in literature. In these situations, BDG significantly reduced transpulmonary gradients avoiding revision. RV to LV pressure ratios were significantly reduced after BDG and no complications related to BDG were noted [7].

Conclusion

Intraoperative TEE identified the hypoplastic pulmonary valve, right ventricular diastolic dysfunction and the anterior barrel flow-related gradient in our patient, in addition to confirming the TTE findings. It was instrumental in surgical planning, avoiding the BDG takedown and an unremarkable post-operative course.

Declarations

We have no conflicts of interest to declare. We received no specific grant from public, commercial or not-for-profit funding agencies. This communication is not currently submitted in any other journal in any other form for publication. The manuscript is read and accepted by both the authors and the requirements of authorship are fully met.

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2. Conflicts of interest/Competing interests: The authors have no conflicts of interest to disclose

3. Ethics approval: Being a case report, The ethics committee approval is not applicable for our manuscript.

4. Consent to participate: Not applicable

5. Written Consent for publication: The child's immediate relative has given consent for publication of the case report

6. Availability of data and material: The data is available with the corresponding author, on request.

7. Code availability (software application or custom code): Not applicable

8. Authors' contributions: Dr MS has written the manuscript and was part of the clinical management. Dr MM has edited and reviewed the manuscript and was part of the case management. Dr AM was part of the clinical management and have provided resources.

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Figures

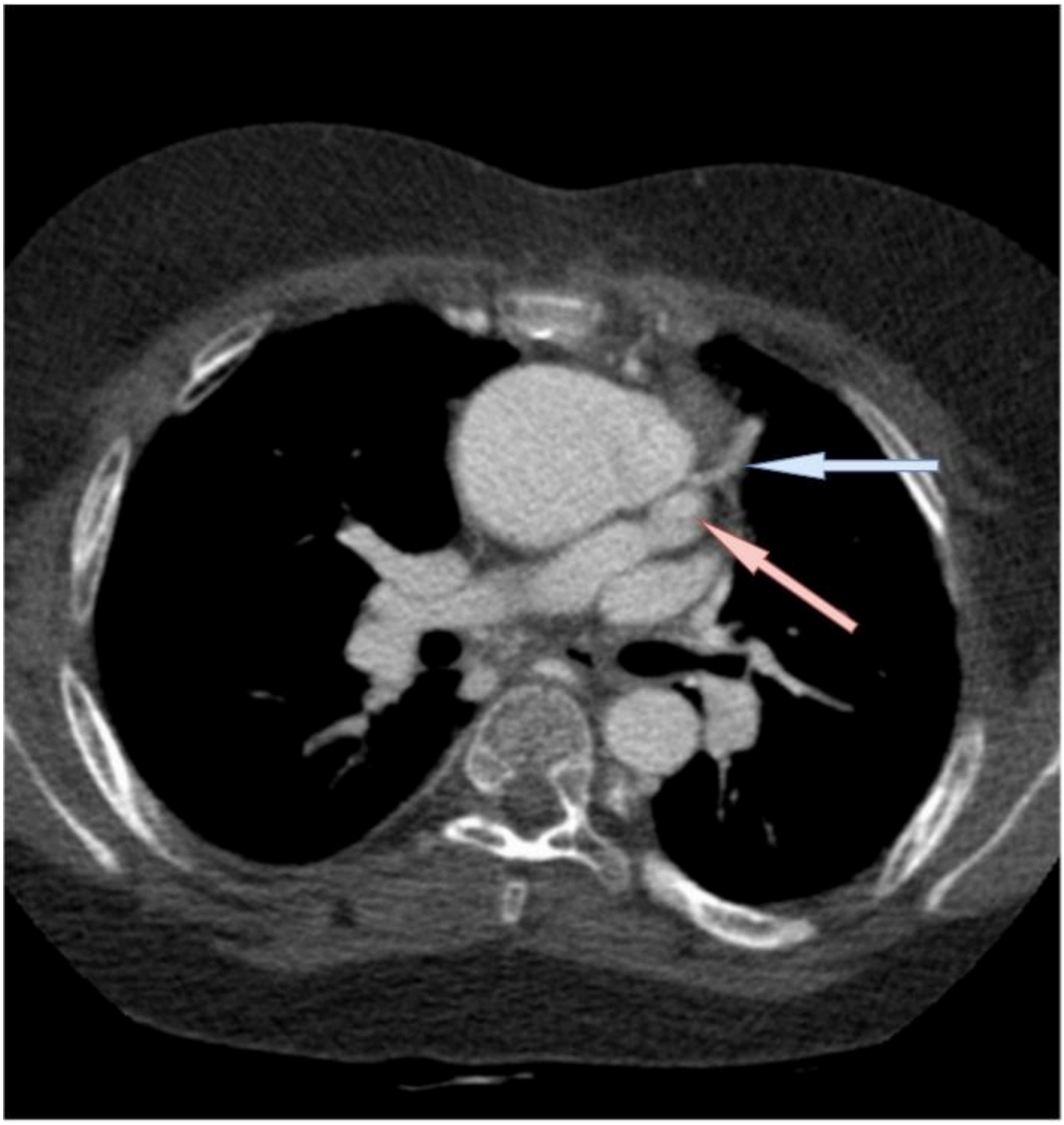


Figure 1

Image from Computed Tomography showing an anomalous coronary artery (blue arrow) coursing anterior to the Right Ventricular outflow tract / Proximal Main Pulmonary artery (pink arrow)

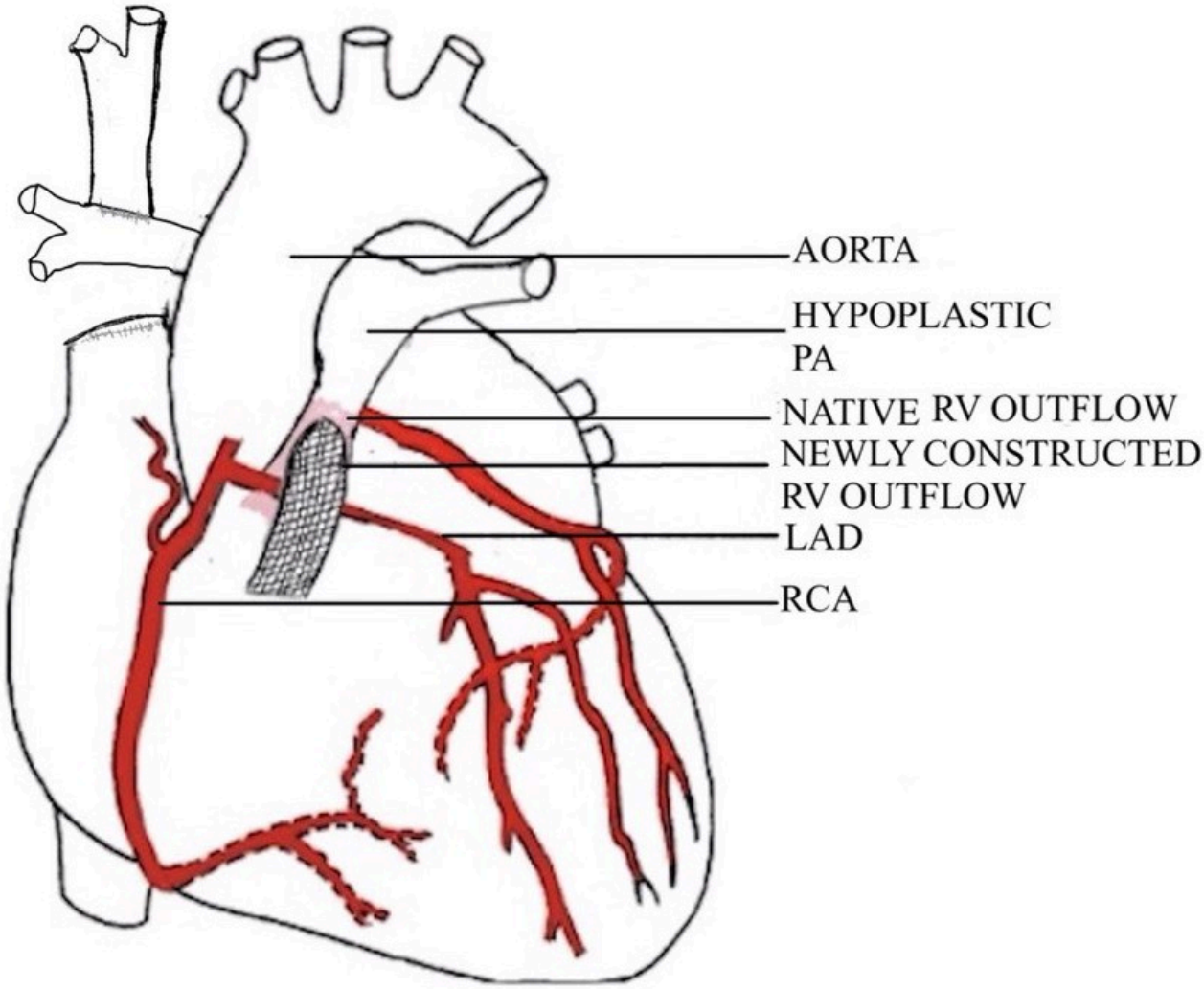


Figure 2

Diagrammatic representation of Double Barrel Outflow

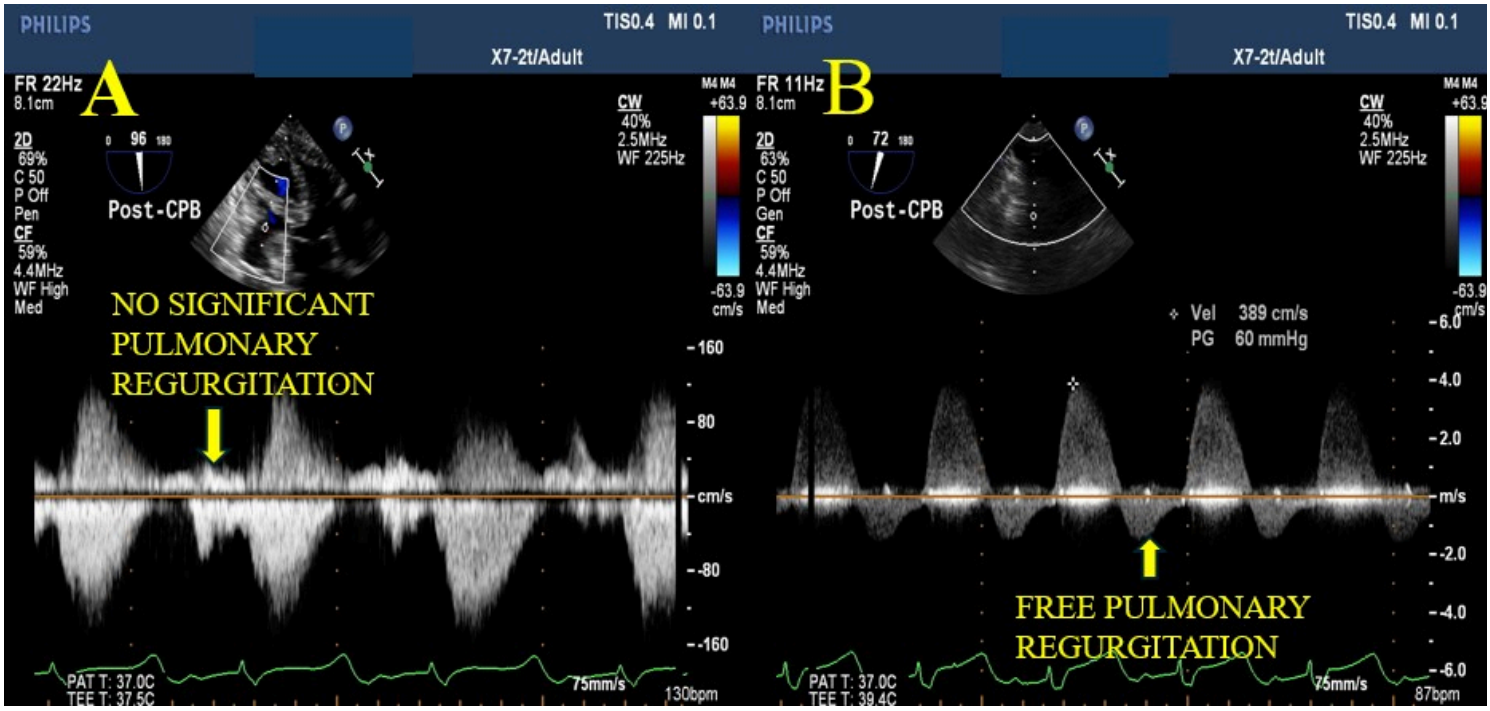


Figure 3

Transesophageal Echocardiography images showing

(A) Flow through native pulmonary valve

(B) Flow through newly constructed outflow with free PR.

Supplementary Files

This is a list of supplementary files associated with this preprint. Click to download.

- Doublebarrelvideo.mp4