# Innovation in Pulmonary Atresia Repair Using Right Atrial Appendage as Pulmonary Valve: A Case Report

Waleed Shahzad Ali[1], Muhammad Zaid Ali[1], Nohela Rehman[2], Hafsah Alim Ur Rahman[2] and Muneer Amanullah[1]

[1]*Department of Cardiothoracic Sciences, Sindh Institute of Urology and Transplantation, Karachi, Pakistan*
[2]*Department of Internal Medicine, Dow University of Health Sciences, Karachi, Pakistan*

## ABSTRACT

A four-year girl with a ventricular septal defect (VSD) and pulmonary valve atresia (PVA) underwent a total correction surgery. Preoperative findings revealed a severe right-sided dilation, a right ventricular hypertrophy (RVH), a large conoventricular VSD, a slightly hypoplastic main pulmonary artery (MPA), and appropriately-sized pulmonary arteries. The surgery began with a midline sternotomy, patent ductus arteriosus (PDA) ligation, and initiation of cardiopulmonary bypass (CPB). The pericardium was harvested, and the right atrial appendage (RAA) was shaped into a bi-leaflet valve. A myomectomy was performed, and the VSD was closed with a continuous Dacron patch. Continuity between the right ventricular outflow tract (RVOT) and pulmonary artery (PA) was achieved using a transannular patch from autologous pericardium, and the RAA valve was implanted. A 15 mm Hegar dilator confirmed smooth blood flow. The patient showed satisfactory intraoperative transesophageal echocardiography (TEE) findings, including good ventricular function, an RVOT gradient of 20-25 mmHg, and no residual defects. The procedure achieved successful outcomes.

**Key Words:** *Tetralogy of Fallot, Pulmonary atresia, Ventricular septal defect, Atrial appendage, Right ventricular outflow tract obstruction, Cardiac surgery, Congenital heart defect repair*.

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## INTRODUCTION

Tetralogy of Fallot (TOF) is the leading cyanotic congenital heart disease, accounting for 3-5% of cases.[1] It involves four defects: overriding aorta, right ventricular outflow tract (RVOT) obstruction, ventricular septal defect (VSD), and right ventricular hypertrophy (RVH). Pulmonary valve atresia (PVA) with VSD, its most severe form, is marked by discontinuity between the RVOT and the main pulmonary artery (MPA). Complete surgical repair connects the right ventricle to the pulmonary artery, often *via* the Rastelli procedure using synthetic conduits.[2] However, these conduits frequently fail, causing complications such as restenosis.[3,4]

This report emphasises the use of the right atrial appendage (RAA) as a neo-pulmonary valve, a technique that could represent a promising alternative in specific clinical contexts, improving long-term outcomes.

*Correspondence to: Dr. Waleed Shahzad Ali, Department of Cardiothoracic Sciences, Sindh Institute of Urology and Transplantation, Karachi, Pakistan*
*E-mail: waleedshahzad221@gmail.com*

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## CASE REPORT

A four-year, 15-kg girl with VSD and PVA required a small-sized conduit. However, due to a higher risk of stenosis, a patent ductus arteriosus (PDA) stent was placed as a temporising measure or palliative measure to maintain stable blood flow till she achieved an appropriate weight threshold for complete corrective surgery. At the age of four, she presented to the outpatient department for an elective surgery consultation with cyanosis (72% oxygen saturation), clubbing, and a continuous murmur. A recent echocardiogram revealed severe right-sided dilatation and RVH, a large conoventricular VSD, a slightly hypoplastic MPA (8 mm, Z score -3.13), and a VSD with PVA. The branched pulmonary arteries were of appropriate size: RPA (9 mm, Z score +0.29) and LPA (6 mm, Z score -2.07).

The patient underwent a total correction surgery *via* midline sternotomy. The pericardium was harvested, and the MPA was dissected to the hilum, exposing the PDA. After achieving an ACT >480 seconds with heparin, cardiopulmonary bypass (CPB) machine was initiated *via* aortic and bicaval cannulation, and the PDA was ligated.

The patient was cooled to 30°C to reduce metabolic demand, and the superior vena cava (SVC) and inferior vena cava (IVC) were snared. The aorta was cross-clamped, and cold blood (DEL NIDO) cardioplegia was administered into the aortic root, resulting in prompt diastolic arrest. The RAA was harvested (Figure 1), and the trabeculation was excised and shaped into a

bi-leaflet valve. The RA was opened parallel to the atrioventricular (AV) groove. A ventricular catheter was inserted into the left atrium (LA) through the patent foramen ovale (PFO). The RVOT was opened, a myomectomy was performed, and the VSD was identified through the RVOT incision and then closed with a Dacron patch in a continuous manner.

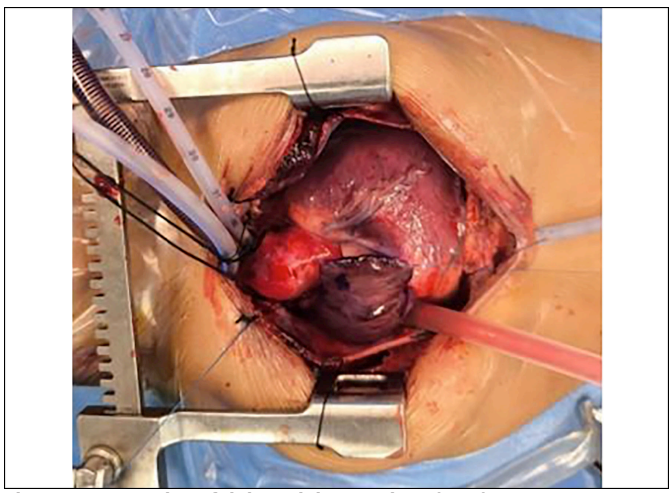

**Figure 1: Harvesting of right atrial appendage (RAA).**

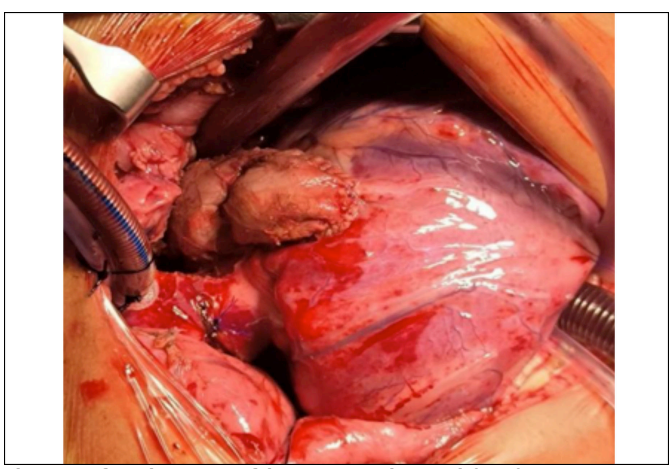

**Figure 2: After placement of the transannular patch (TAP).**

The MPA was opened from its blind pouch, and its posterior wall was attached to the RVOT using an autologous pericardial patch to create continuity between the RVOT and PA. The RAA was then implanted in the neo annulus, and RVOT-PA continuity was completed with an autologous pericardial transannular patch extending from the PA bifurcation to the RVOT, anteriorly at the level of the valve (Figure 2). Finally, a 15 mm Hegar dilator was passed smoothly through the RVOT valve and MPA to ensure unobstructed blood flow.

Rewarming was initiated, and the RA was closed in a double layer with Prolene 5/0. The aortic cross-clamp was removed after 138 minutes, the heart was de-aired, and the patient was weaned off CPB. Intraoperative transesophageal echocardiography (TEE) showed satisfactory ventricular function, a RVOT gradient of 20-25 mmHg, a PFO with left-to-right shunting, and no remaining VSD or valvular regurgitation. Protamine was administered. One pacing wire was placed in the right atrial and one in the right ventricle. Mediastinal and bilateral pleural drains were inserted and secured. Finally, the aortic and bicaval venous cannulas were removed.

Haemostasis was secured, and the pericardial cavity was washed with saline and transamine. The sternum was closed with steel wires in a figure-of-eight pattern, and the muscles and subcutaneous tissues were closed with Vicryl 3/0. Local anaesthetic was infiltrated into the wound, and an aseptic dressing was applied.

The patient was commenced on inotropic support of epinephrine (0.08 µg/kg/min) and milrinone (0.7 µg/kg/min). Postoperative status was assessed by echocardiography. Inotropes were discontinued on the first postoperative day, and she was ambulated and discharged on the second day. On follow-up, she has continued to progress.

## DISCUSSION

Managing complex congenital heart defects, such as VSD with PVA, in paediatric patients requiring RVOT reconstruction presents significant challenges. Traditional methods, such as prosthetic conduits or homografts, are effective but have limitations, including calcification risk, lack of growth potential, and the need for frequent reoperations due to somatic growth and conduit degeneration.

A novel surgical approach utilises autologous RAA to create a bi-leaflet pulmonary valve, combined with autologous pericardium for RVOT reconstruction. This method offers multiple advantages. Autologous tissues minimise immunologic reactions and eliminate the need for anticoagulation therapy, which is an important consideration in paediatric patients. Moreover, the growth potential of autologous pericardium reduces the risks of stenosis and reoperations, leading to better long-term outcomes.[4] Additionally, the technique reduces costs by avoiding commercial conduits and lowering reoperation frequency, making it particularly appealing in resource-limited settings.[4]

Recent studies reported promising results. Hosseinpour *et al*. demonstrated satisfactory short- and mid-term outcomes using RAA for pulmonary valve construction during TOF repairs.[5] Similarly, autologous pericardial patches have also demonstrated durability in RVOT reconstruction.[4] Collectively, these findings highlight the potential of autologous techniques to overcome limitations of prosthetic materials.

This paper demonstrates a promising technique for RAA valve reconstruction in PA-VSD, addressing a critical gap in the management of complex anatomies. The detailed intraoperative strategy and favourable short-term outcomes are encouraging; however, further investigation is needed to assess its long-term durability and broader applicability. Given its natural tissue properties such as flexibility, growth potential, and compatibility with right-sided pressures, the RAA may offer advantages over synthetic or xenograft conduits, which are prone to calcification and degeneration. Nonetheless, there are

still concerns regarding valve dysfunction, stenosis during growth, and technical complexity. Comparative studies with conventional conduits such as Contegra, combined with biomechanical evaluations and exploration of hybrid interventions, may help clarify its long-term benefits and potential for wider adoption. Careful surgical planning and expertise are essential for achieving successful outcomes.

In conclusion, this technique offers a compelling alternative in paediatric cardiac surgery due to its adaptability, reduced immunogenicity, and cost-effectiveness.

**PATIENT'S CONSENT:**
Informed consent was taken from the parents of the patient involved in this study for the publication of clinical data and images.

**COMPETING INTEREST:**
The authors declared no conflict of interest.

**AUTHORS' CONTRIBUTION:**
WSA: Contributed to the surgical procedure, manuscript review, and editing.
MZA, MA: Contributed to the surgical procedure.
NR, HAUR: Contributed to manuscript writing and editing.
All authors approved the final version of the manuscript to be published.

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