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Re-do Pulmonary Valve Replacement with Concomitant Aortic Valve Replacement in a Post-surgical Patient of Tetralogy of Fallot- A Case Report

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Abstracts:

The management of Tetralogy of Fallot (TOF), one of the commonest congenital anomalies, has advanced rapidly and excellent long-term survival were observed. Residual defects like pulmonary valve regurgitations are identified in an increased number as long-term survival increased and requirement for intervention increased worldwide. Although rare in our setting, we performed aortic valve replacement (AVR) with re-do pulmonary valve replacement surgery on a 21-year-old, non-diabetic, hypertensive young male with the diagnosis of moderate to severe aortic regurgitation with stuck pulmonary (mechanical) valve, with status-post intracardiac repair (ICR) for Tetralogy of Fallot (TOF) with aortic valve repair (AVRe) with pulmonary valve replacement (PVR) with closure of ventricular septal defect (VSD). His postoperative recovery was satisfactory and he was discharged symptom free on 6th postoperative day.

Keywords: Pulmonary valve replacement (PVR), re-do Pulmonary valve replacement (re-do PVR), re-do on Tetralogy of Fallot repair

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Introduction:

Tetralogy of Fallot (TOF), is one of the commonest of cyanotic congenital heart disease (CHD), having a frequency rate of 3.4 per 10000 live births[1,2]. Even though, classical clinical characteristics of TOF, was explained in 1673 by Bishop and Nicolas Steno (anatomist), the precise anatomy was extensively described by the French physician Etienne-Louis Fallot in 1888[3]. Subsequently, first surgical repair was introduced in 1954 and since then treatment has improved exponentially which results in excellent long-term outcomes (30 years survival ranging from 68.5% to 90.5%)[4]. Although, residual defects like pulmonary regurgitation (PR) were initially thought as a benign hemodynamic lesion, but PR afterwards leads to RV dilatation and RV dysfunction. This RV pathology consequently leads to the ventricular arrhythmia and biventricular dysfunction[5] which requires further surgical intervention.

Fortunately, as many more patients are thrived to the adulthood and more pulmonary regurgitation are diagnosed, need for pulmonary valve replacement (PVR) has been increased too[5]. Hence, PVR is effective in reducing RV dilatation and the consequences including the LV function. PVR is performed in different ways; transcatheter and surgical[7]. For surgical PVR both biological and mechanical prosthesis are being used. Although, there is very little data available about the long-term outcome of the surgical PVR, homograft or the bio-prosthesis are being preferred[6]. Study shows, 10-year re-PVR–free survival of TOF patients undergoing homograft PVR ranges from 74% to 89%[8].

Long-term outcomes of TOF mostly depends on the understanding of the RV pathophysiology and the timely intervention, selecting the candidates and re-intervention required for the PVR which still remains challenging[6].

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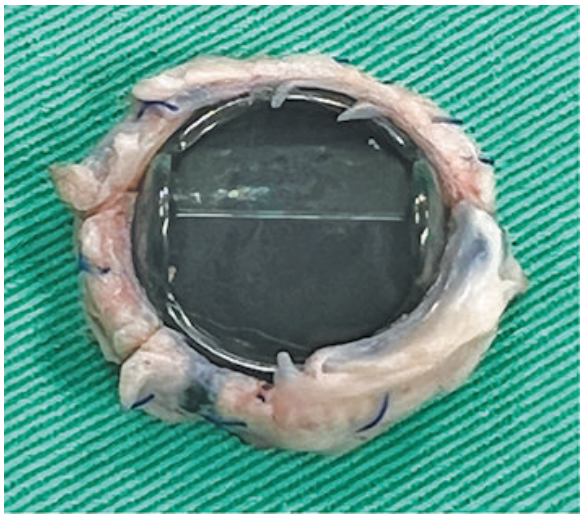

Figure 1: *Extracted old Pulmonary (mechanical) valve,*

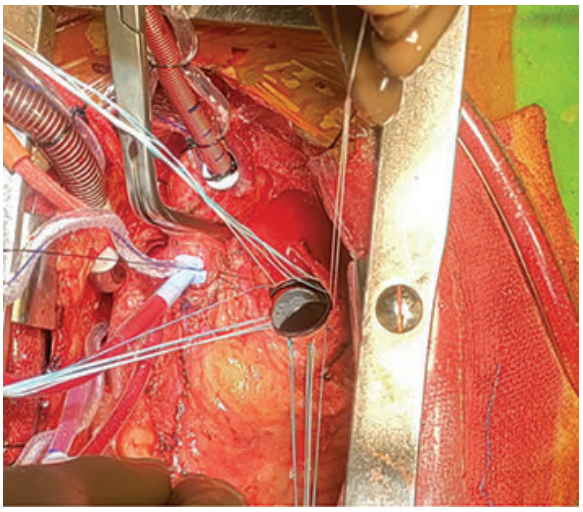

Figure 2: New pulmonary (mechanical) valve in situ,

Case presentation:

Mr. X, 21-year-old, non-diabetic, hypertensive young male got admitted to our institute as a diagnosed case of moderate to severe aortic regurgitation with stuck pulmonary (mechanical) valve, with status-post intracardiac repair (ICR) for Tetralogy of Fallot (TOF) with aortic valve repair (AVRe) with pulmonary valve replacement (PVR) with closure of ventricular septal defect (VSD) in 2011. He was diagnosed as a patient of TOF with congenital aortic regurgitation with absent pulmonary valve and underwent ICR with aortic valve repair & pulmonary valve replacement surgery in 2011. But, for the last 2 years, he is experiencing exertional chest pain with breathlessness and generalized weakness. His transthoracic echocardiogram revealed prosthetic pulmonary valve dysfunction with severe stenotic gradient (PPG-90mmHg) & mild-moderate pulmonary regurgitation, with bicuspid aortic valve and mild to moderate aortic regurgitation with mildly dilated aortic root with mild TR (PPG 80 mm of Hg) with good bi-ventricular function. He also underwent CT angiogram which revealed normal coronaries. So, he got admitted at our center for further better management.

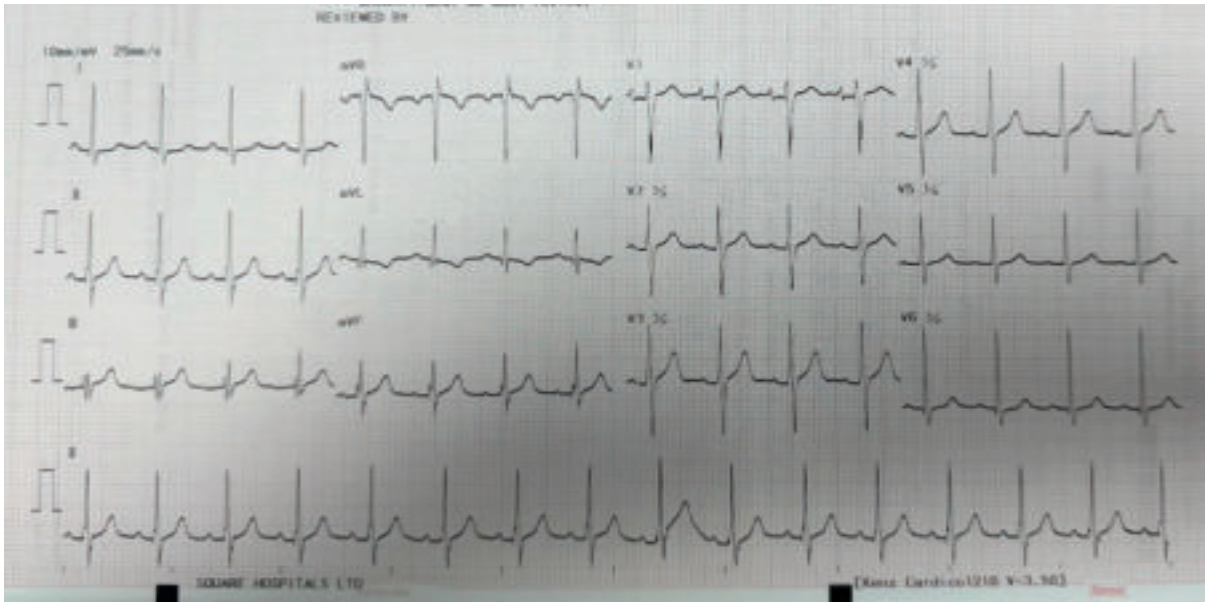

Figure 3: Post-operative ECG showing normal sinus rhythm.

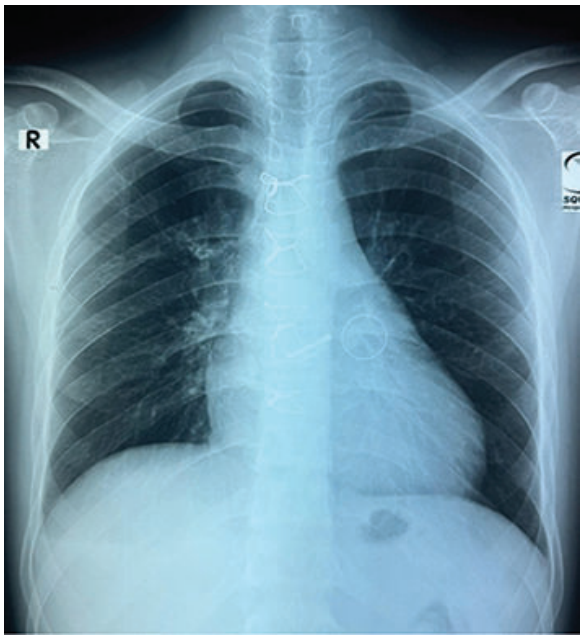

Figure 4: *Post-operative x-ray showing both the mechanical valves in-situ*

Operative Procedure:

Under all aseptic precautions after proper painting and draping, median re-sternotomy was done with oscillating saw. Careful adhesiolysis was done. CPB was established with Aortic and bicaval venous cannulation after heparinization. Aortic X-clamp was applied and cardioplegia was delivered. Heart was arrested and Aortotomy was done. Aortic valve was inspected, was found bicuspid and severely regurgitant. Valve replaced with 23mm SJM™ Masters Series Mechanical Heart Valve. Aortotomy was closed. Pulmonary prosthesis was approached through pulmonary arteriotomy and found non-functional. Valve explanted (Fig.-1) and replaced with 21mm SJM Regent™ Mechanical Heart Valve (Fig.-2). Pulmonary arteriotomy was closed. Aortic X-clamp was released after proper de-airing. Heart was weaned from CPB to normal sinus rhythm. Decannulation was done and Protamine was given. After proper hemostasis, chest wound was closed in layers keeping RV pacing wire and chest drain tubes in situ. Following the replacement of double valves (aortic and pulmonary), patient was sent to the intensive care unit where he was managed according to the usual valve surgery protocol. ECG showed normal sinus rhythm (Fig.-3) and inotropic support with pulmonary vasodilator was used with diuretics. As there was no sign of endocarditis usual antibiotics protocol followed.

Discussion:

Our patient underwent re-do pulmonary valve replacement along with Aortic valve replacement due to stuck mechanical pulmonary valve which was replaced in 13 years back, when he went through intra-cardiac repair for Tetralogy of Fallot. His Aortic valve was repaired then, which was a bicuspid valve. That repaired bicuspid valve was found to be moderate to severely incompetent after long 13 years. His primary surgery was performed as an intracardiac repair that included repair of VSD, pulmonary valve replacement with mechanical valve (due to pulmonary stenosis) and repair of the aortic valve.

Replacement of pulmonary valve is not very common in our circumstances especially in the younger population let alone re-do replacement. Proper clinical assessment and performing surgery to get the most benefit possible in the immediate and long term is crucial particularly in complex congenital cardiac diseases like TOF. Sometimes, when integrity of the pulmonary valve is left disrupted after relieving the RVOT obstruction might results in pulmonary valve regurgitation. This ensuing RV dilatation, RV dysfunction with tricuspid regurgitation, electrophysiological abnormalities and cardiomyopathies.[1] Early and timely replacement of pulmonary valve can prevent all these adverse outcomes. Although benefits of early replacement have already been established by several groups of surgeons worldwide, we are not very fortunate to do such procedures often. This case was one of the exceptional attempts of pulmonary valve replacement in TOF. Replacement of the both semilunar valve is very rare as well especially when one of them was replaced about a decade ago.

INR was kept within 3.0 in this patient by adjusting the dose of warfarin and low dose of aspirin. Patient was shifted to the cabin on 3rd POD as his postoperative period was uneventful. His postoperative recovery was satisfactory and he was discharged symptom free on 6th postoperative day. His first follow-up was 7 days later and he was doing better. Dose of the diuretics were reduced and dose of warfarin was adjusted accordingly. On his one-month follow-up Chest-X-ray showed nothing abnormal but both the valves in situ (Fig.-4) and transthoracic echocardiogram showed- Tetralogy physiology; S/P ICR, VSD closure with RVOT resection; S/P AVRe, PVR; Postop AVR-Redo PVR- i) Well seated

and functioning Aortic and Pulmonary prosthesis with MPG of 10 & 6 mmHg with trivial central AR & PR but no paravalvular leakage; ii) Patch intact with no residual shunt; iii) Laminar flow in RVOT; iv) No gross regional wall motion abnormality; v) Normal bi-ventricular function (EF-55%); vi) Mild concentric LVH; vii) No pericardial effusion/ intracardiac thrombus seen.

Conclusion:

Performing a re-do surgery is difficult in terms of adhesion, bleeding, tissue damage, retrosternal anatomical deformities, etc. But, in appropriate cases a re-do is a definitive life saver. Successful completion of this surgery and speedy patient recovery encourages the surgeons to perform these types of challenging task to alleviate patient sufferings. Replacement of pulmonary valve with homograft may result in interesting result in our population.

Limitation and Recommendations: This is a single case report and more cases like this can help us for opting the optimized management protocol.

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Informed Consent: Patient was informed about publication of this report, necessity of such publication.

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