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Outcomes Following Primary Pulmonary Valve Replacement in Adult Tetralogy Of Fallot

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Abstract:
Introduction: *Adult Tetralogy of Fallot is a burnt-out disease and has higher morbidity and mortality due to volume over load from multiple collaterals, dysplastic pulmonary valve and severe right ventricular hypertrophy. Primary Pulmonary Valve Replacement at the time of surgery addresses these problems and decreases the morbidity and mortality associated with repair in adult Tetralogy of Fallot.* ***Objective:*** *The objective of this study is to analyze the outcomes of primary pulmonary valve replacement in adult Tetralogy of Fallot patients.* ***Materials and Methods:*** *This is a retrospective study conducted over a period of five years from January 2014 to January 2019 in all adult patients who presented with uncomplicated Tetralogy of Fallot at our institution. Their initial presentations, previous procedures if any, detailed anatomy based on 2d echo and CT scan, intra operative findings and post op course have been studied. An average follow up for two years was noted to enquire about their symptoms and quality of life.* ***Results:*** *To avoid complications of volume overload, free pulmonary regurgitation and right ventricular dysfunction, we have offered primary pulmonary valve replacement as a part ofcomplete correction of Tetralogy of Fallot. There has been a total of eighteen patients who underwent this procedure along with complete Tetralogy repair. After ventricular septal defect closure and right ventricular out flow tract obstruction resection, pulmonary valve is excised. A prosthetic pulmonary valve appropriate for the size of the patient is implanted using the continuous suture technique. We have chosen mechanical valves in men and bioprosthetic valves in women of child bearing age who wish to continue their family. There has been no mortality and minimal use of inotropes during weaning and early extubation within 6-10hrsalong with reduced ICU and hospital stay in all these patients. There has been an improvement infunctional activity and quality of life.*
Conclusion: *Surgery for adult Tetralogy of Fallot can be made safe by reducing the post operative morbidityand mortality that is associated with free pulmonary valve regurgitation and right ventricular dysfunction. This is achieved by replacing the dysfunctional pulmonary valve at the time for surgery.*

Keywords: *Adult Tetralogy of Fallot, Pulmonary Valve Replacement, Right Ventricular Dysfunction*

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I. Introduction:

Less than 2% of patients born with Tetralogy of Fallot reach adulthood. These patients present with the complications of long standing cyanosis and chronic right ventricular out flow obstruction in the form of right ventricular dysfunction, multiple collaterals and volume over load and arrhythmias.[1] Patients in whom the pulmonary valve has not been addressed, there are long term complications of free pulmonary valve regurgitation, right ventricular distention and dysfunction, tricuspid regurgitation and right heart failure.[2] The development of arrythmias post correction still remains a controversy whether it's a result of chronic right ventricular fibrosis and dilatation or due to resection of the right ventricular outflow tract bundle.[3] In patients who present with Tetralogy of Fallot in adulthood we find that Primary Pulmonary Valve Replacement along with total correction addresses all problems and decreases the morbidity and mortality associated with this pathology.

II. Materials And Methods:

This is a retrospective study conducted over a period of 5 years with an average 2 year follow up in all adult patients who presented with uncomplicated Tetralogy of Fallot. From 2014 to 2019, 18 patients underwent primary pulmonary valve replacement as a part of complete correction for adult Tetralogy of Fallot. The data used for the study were operative presentation, intra operative procedure and post-operative course. All the patients were on periodic follow up for 2yrs (once in 6 months), and during follow up, further data were collected with respect to symptoms and echocardiographic findings.

The diagnosis of the condition was made by echocardiography. All patients in addition underwent contrast enhanced computed topography scan for evaluation of the peripheral branches of pulmonary arteries. Our institute protocol of coronary angiography was performed in patients aged more than 40 years.

Operative Technique:

All patients underwent standardized techniques of anesthesia and monitoring, and cardiopulmonary bypass. After Median sternotomy, pericardium was opened to the right so as to preserve as much as possible for reconstruction of right ventricular outflow tract. After adequate heparinization to achieve the desired activated clotting time of greater than 480 seconds, standard ascending aortic and bicaval cannulation was done, and patient was put on the cardiopulmonary bypass. In patients with previous shunt procedures, the shunt was dissected and looped prior to commencement of the bypass. In case of difficulty in looping the shunt, on low flows, the main pulmonary artery was opened and incision extended to the left pulmonary artery and the pulmonary end of the shunt was closed using a pledgeted 4-0 monofilament polypropylene suture. The entire procedure was carried out at moderate systemic hypothermia (27 degrees centigrade) with antegrade Del Nido cardioplegia as our preferred method of myocardial protection. The left side of the heart was vented through a right superior pulmonary vein vent.

Once arrested in diastole, the patient was put on total bypass with the snaring of the superior and inferior vena cava and flows confirmed. The anatomy was inspected both from the right atrium and pulmonary arterial end. The outflow tract bundles were first incised or excised to try and create a roomy tract. After this the ventricular septal defect was identified, and that was closed using a knitted double velour polyester patch using a continuous suture technique with a 4-0 mono filament polypropylene pledgeted suture. After closure of the septal defect, the right ventricular outflow tract was reconstructed. The pulmonary valve was inspected by extending the main pulmonary artery incision to the pulmonary annulus and pulmonary valve abnormalities of tethering, commissural fusion, bicuspid valve were noted. The incision was distally extended till the bifurcation of the main pulmonary artery or beyond the stenosis of the branch pulmonary arteries if present. An adequately sized pericardium was harvested, a reconstruction of the main pulmonary artery was done from distally to proximally up till the pulmonary annulus using a 4-0 mono filament polypropylene suture. The main pulmonary artery incision was extended then to one thirds of the right ventricular outflow tract parallel to the left anterior descending artery.

Adequately sized aortic valve prosthesis for the patients' body surface area was then taken and reinforcedto the pulmonary annulus in a continuous suture fashion using a 3-0 17.5 mm needle mono filament polypropylene suture starting from laterally to the medial part (Fig.1). After securing the posterior annulus, the anterior and lateral parts of the prosthetic valve were secured with a 3-0 mono filament polypropylene suture by creating a neo annulus with the pericardium that was used for the augmentation of the main pulmonary artery (Fig.3). In some cases, the valve was implanted using an intermittent suture technique using double arm 2-0 17.5 mm needle pledgeted polyester suture (Fig.2). After securing the valve, the remaining portion of the pericardium was used to augment the right ventricular outflow tract using a 3-0 mono filament polypropylene suture (Fig.4). In case there was pericardial patch deficiency, a polyester graft was used to augment the right ventricular outflow tract and secure to the neo-pulmonary annulus.

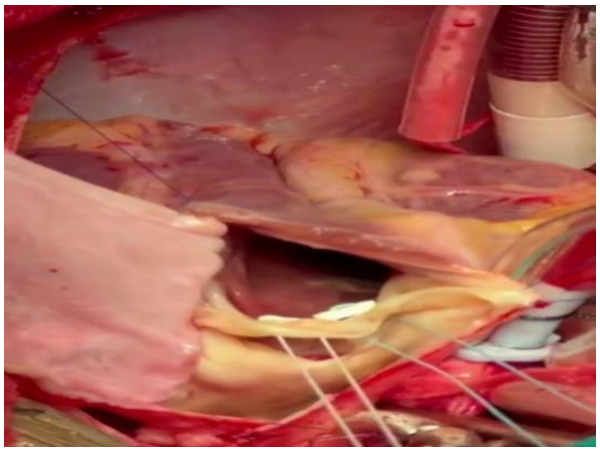

Figure 1: After fixing the pericardium on the lateral wall of the main pulmonary artery, valve sutures are taken on the pulmonary annulus.

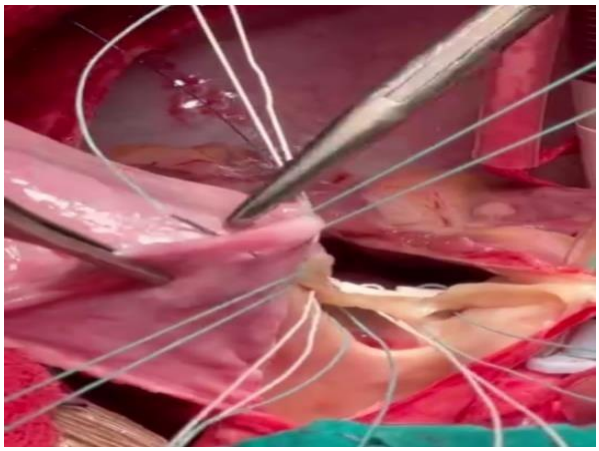

Figure 2: Creation of neo-pulmonary annulus on the pericardial patch using the intermittent pledgeted suture technique.

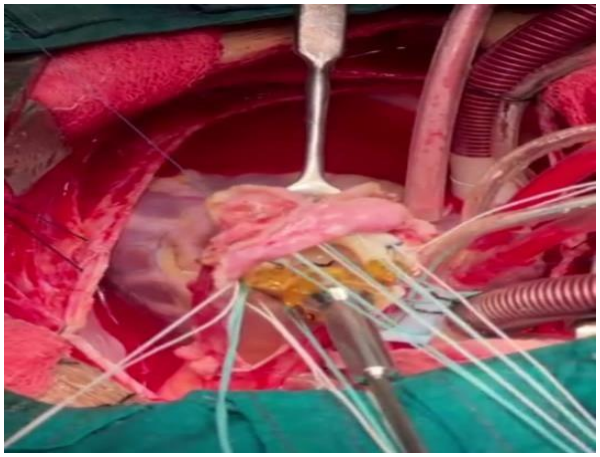

Figure 3: Securing the pulmonary valve along the native and neo-Pulmonary annulus.

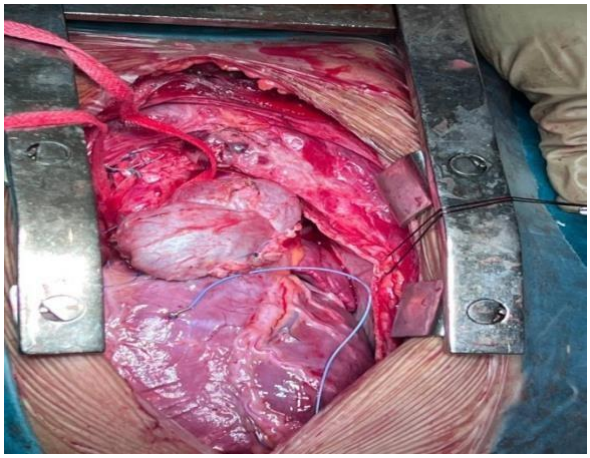

Figure 4: Completed picture demonstrating the neo pulmonary artery with annulus with right ventricular outflow tract reconstruction.

After reconstruction of the right ventricular outflow tract, the tricuspid valve was checked to look for competence. A tiny patent foramen ovale was left. Rewarming was initiated at right atrium closure. Standardized practices of weaning and closure were performed. All patients underwent primary closure of the sternum.

Patients were shifted to the intensive care unit with low dose dopamine and adrenaline as per our institute protocol and placed on ventilator support. They were shifted out of intensive care unit after Extubation and after weaning of inotropes. They were discharged from step-down ward. At time of discharge echocardiographic findings were noted with respect to status of ventricular septal defect patch, right ventricular and pulmonary artery gradients. Patients were followed up initially after one month and 6 months thereafter for a period of two years with symptoms enquiry and echocardiographic findings. All patients were on anticoagulation with ecosprin 75mg and warfarin with INR range of 2-3.

Statistical Analysis: Statistical analysis of the data was done using Microsoft excel sheet where results were calculated and expressed as mean and standard deviation.

Results: Table 1 summarizes the main pre-operative characteristics of the patients who underwent this procedure. The average age at presentation was 27.44 years (17-55 years). There were 7 women (38.88%) and 11 men (61.12%). 2 patients (11.11%) were having NYHA class IV symptoms, 11 patients (61.12%) had NYHA class III and the remaining 5 patients (27.77%) had class II symptoms. Most common mode of presentation was dyspnea on exertion (72.2%) followed by fatigue on exertion (44.4%). The mean saturation at presentation was 79.4±9 %

and hematocrit recorded was 60.8±8.7%. 6 patients (33.33%) underwent BT shunt procedure an average 27.6 years (18-54years) before the definitive surgery. Associated anomalies of branched pulmonary artery stenosis were found in 6 patients (33.33%). No other systemic abnormalities were noted.

Table 1: Pre-operative characteristics of patients in this study		
AVERAGE AGE		27.44 YRS (17-55)
SEX	Men	11 (61.12%)
	Women	7 (38.88%)
NYHA CLASS	II	5 (27.77%)
	III	11 (61.12%)
	IV	2 (11.11%)
Saturation %		79.4±9
Hematocrit		60.8±8.7
RV-PA Gradient (mm HG)		73.1±17.7
Previous Shunt		6 (33.33%)
Associated anomalies	Branch Pulmonary artery stenosis	6 (33.33%)

(NYHA-New York Heart Association, RV-PA-Right Ventricular- Pulmonary Artery)

The inotropic supports were as per our institute's protocol of low doses of dopamine and adrenaline. All needed ventilator support for 6-10 hours post-surgery. One patient was re-explored for bleeding within 12 hours and required prolonged ventilation and tracheostomy for 13 days post-surgery. The inotropic support was weaned in the next 36-48 hours. Except in the one patient mentioned above, the average intensive care unit stay was 3 days, and hospital stay was 5-7 days. All patients had a relatively uncomplicated course in the hospital.

Table 2: Summary of Surgical Outcomes		
Average ICU stay		3 days
Average hospital stay		5-7 days
Prosthetic valve	Bioprosthetic valve	13 (72.2%)
	Mechanical bileaflet valve	5 (27.8%)
Valve size	19mm	9 (50%)
	21mm	7 (39%)
	23mm	1 (5.5%)
	25mm	1 (5.5%)
Bypass time		137.2±13.8min
Cross clamp time		98.1±9.1min
Post-operative right ventricular pulmonary artery gradient		32.2±7.2mm Hg

The most common used valve was the 19mm valve in 9 patients (50%), followed by 21mm valve in 7 patients (39%) and 23mm valve in 1 patient (5.5%) and 25mm valve in 1 patient (5.5%). Bioprosthetic valve was the choice in 13 patients (72.2%) and the remaining 5 patients (27.8%) had mechanical bileaflet valve placed. Initially we had chosen mechanical valves in male patients and bioprosthetic valves in female patients of child bearing age who wish to continue their family. However, in the last 3 years, all the valves were replaced with a bioprosthetic valve irrespective of gender keeping in mind the future need for catheter-based interventions. Post-op echo demonstrated acceptable gradients across right ventricular outflow tract averaging 32.3±7.2mm Hg. 3 patients (16.7%) had mild RV dysfunction which was managed conservatively with diuretics. All patients were on anticoagulation with ecosprin 75mg and warfarin with INR range of 2-3. All patients were followed up every 6 months for a duration averaging 39 months (12-79 months) as shown in table 2. There has been no early or late mortality noted and 16 patients had NYHA class I symptoms and 2 patients had NYHA class II symptoms on follow up. Investigation with echocardiography demonstrated reduced gradients in all patients and no right or left ventricular dysfunction.

Statistical Analysis:
Statistical analysis of the data was done using Microsoft excel sheet where results were calculated and expressed as mean and standard deviation. The Z test was done to test the hypothesis that pulmonary valve replacement indeed reduced the right ventricular tract outflow obstruction and improved quality of life. The p-value was at 0.025 with a Z score at 1.96 and test statistic of 0.97.

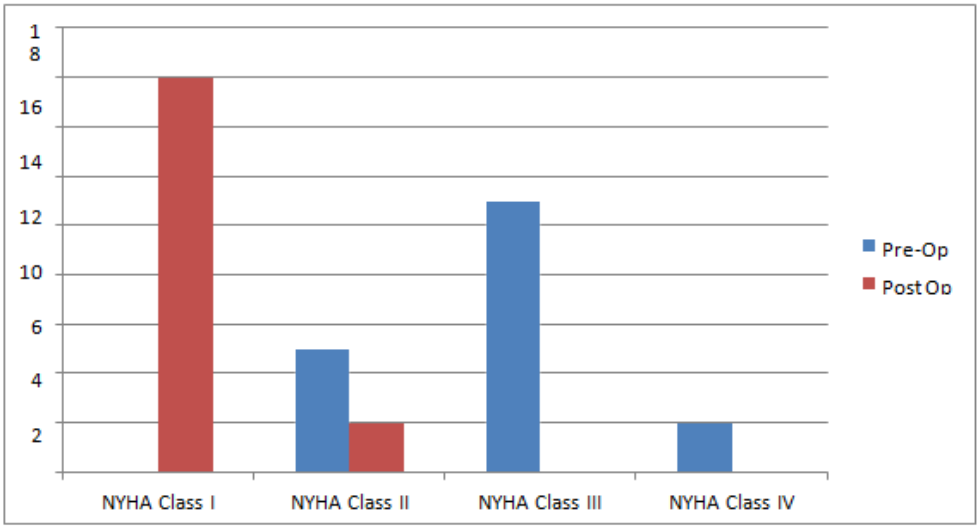

Figure 5: Improvement in NYHA class post operatively

NYHA-New York Heart Association

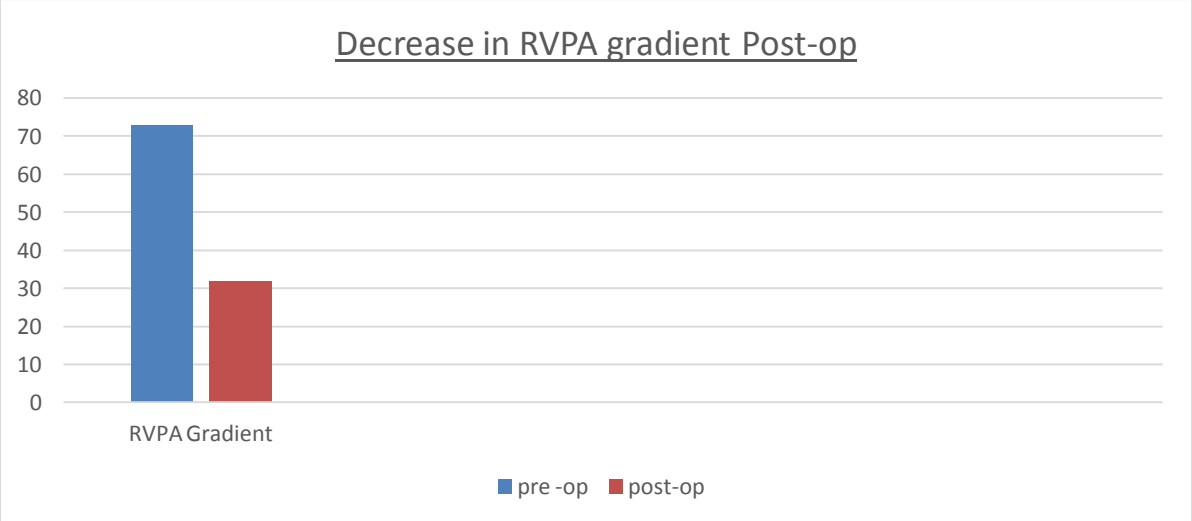

Figure 6: RVPA gradient – pre-op and post-op

(RVPA – Right ventricle pulmonary artery)

III. Discussion:

Etienne Louis Fallot first described this cyanotic congenital cardiac condition in one of the land mark papers in cardiac surgery titled *MALADIE de FALLOT* in the year 1888. [1,4,5] Since then and even before, this disease has interested both adult and pediatric physicians and surgeons. Multiple theories have been postulated about its origin and development. However, the end result is a large maligned nonrestrictive ventricular septal defect, varying degrees of aortic over ride, pulmonary outflow tract obstruction at different levels and progressive right ventricular hypertrophy. [1,4]

One of the initial attempts at palliation was attempted by *Alfred Blalock, Helen B. Taussig and Vivien Thomas* in 1944 at John Hopkins University.[6] There have been many modifications and alternatives to this procedure. The first attempt at total correction was made by Lillehei and colleagues. [5,7] The operative success

for repair is best when performed in the early age of life.

In our Indian scenario, due to multiple reasons, we still encounter these Tetralogy of Fallot patients who have grown into adult hood. This presents with its own set of additional problems. [5,10,11,13,14] Chronic cyanosis and its effects on the heart and other organs, increased hematocrit and coagulation abnormalities, volume over load from collaterals, progressive right ventricular dilatation and failure leading to arrhythmias, exercise intolerance and even premature death.

Complete pre operative evaluation of patients who present with this sub set of disease with echocardiography supported by cardiac catheterization is advised to identify associated anomalies and embolize collaterals when present to avoid intraoperative and postoperative increased pulmonary blood flow.[10,11,12,14] Long standing cyanosis, right ventricular fibrosis and dilatation, advanced age have been documented incremental risk factors in predicting the outcomes of surgery.[1,10,11,12] However considering all, outcomes are good whentotal correction is attempted in these patients.

In our study we noted that patients who underwent total correct of Tetralogy of Fallot even as adults responded well to surgery and the rate of complications is as per international reports. [10,11]

Not many studies are available in literature that advice pulmonary valve replacement in all patients who undergo surgery. Reports mention that these patients undergo a variety of treatment options including pulmonary valvotomy, transannular patch and in some replacement. These studies in which part of the patients underwent pulmonary valve replacement, fared better compared to other in whom replacement was not performed. Adult Tetralogy patients in whom pulmonary valve has not been replaced, still run the long-term risk of pulmonary valve dysfunction and regurgitation and its complications. [9,10,11]

Even after 68 years from the first surgical complete repair attempted, free pulmonary valve regurgitation and right ventricular volume overload is still a mystery unsolved. Right from attempting to fashion a mono-cusp valve to accepting severe pulmonary regurgitation, redo cardiac surgery for pulmonary valve replacement and/or homo-graft placement- there have been many trials to handle this common long term complication of progressive right ventricular failure.[5,8,9]There has been an interest to study the effect of pulmonary valve replacement to address these problems.[6,7,10] Pulmonary valve replacement, hopes to reduce pulmonary regurgitation and help in ventricular remodeling -thus preventing the long term complications.[10,11,12]

Studies indicate that bioprosthetic valves are more superior to mechanical valves or even homografts. They offer a freedom from repeated long-term anticoagulation and in case of valve degeneration, a catheter-based approach can be attempted. [10,13] The need for prolonged ventilation, duration of inotropesand hospital stay are considerable reduced when the pulmonary valve was replaced at the time to total correction.

But controversy still exists about the fibrosis that is already present in the ventricles due to the long-standing disease. The extensive resection and ventriculotomy may predispose to arrhythmias. Pulmonary valve replacement solves part of the long-standing complication of volume overload and progressive right ventricular dilatation and dysfunction. [13]

Limitations of the study:

This is a retrospective study for a specific subset of patients in whom this procedure is performed. The more complex spectrum of pulmonary atresia and absent pulmonary valve with ventricular septal defect, transposition of great arteries with ventricular septal defect and right ventricular outflow tract obstruction, double outlet right ventricle with right ventricular outflow tract obstruction and such spectrum have not been considered. Besides the main mode of diagnosis has been echocardiography supported by contrast enhanced computed tomography. The chronic effects of hypoxia and volume over load and right ventricular function are better studied on magnetic resonance imaging. [1,7]In a retrospective study, data available also has limitation in its study with respect to recording of vital information that impact the outcomes and all variables may not be considered-pre operatively and post operative ventricular volumes and measurements. Post operative errors may be operator specific in the setting of a patch in the outflow tract which alters the geometry and flow dynamics.

IV. Conclusion:

The biggest benefit that complete correction and pulmonary valve replacement provides is the drastic improvement in symptoms and alleviates the need for post operative re-intervention for pulmonary insufficiency. The volume over load and dilatation and progressive ventricular dysfunction is also managed effectively by adding a competent valve in the pulmonary position at the time of open-heart surgery. Adult Tetralogy of Fallot is that spectrum of disease where effects of long-term cyanosis and volume and pressure over load have detrimental effect on the outcome. Hence, we propose and prove with our study that Primary pulmonary valve replacement single solution is simple, safe and decreases the morbidity andmortality associated with complete correction in adult Tetralogy of Fallot.

Conflict of Interest:
The authors would like to state that there has been no conflict of interest what so ever in the present study.
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