Total Correction of Tetralogy of Fallot: Do We Still Need Trans annular Patch

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ABSTRACT	
Objective	To evaluate early results after surgical repair for tetralogy of Fallot; to find out the trends of repair technique and outcomes at NICVD.
Study design	Descriptive case series.
<i>Place & Duration of study</i>	Department of Paediatric Cardiac Surgery, National Institute of Cardiovascular Diseases Karachi, from January 2016 to December 2016.
Methodology	All patients operated for total correction of tetralogy of Fallot were included in the study. Each patient was followed for three months post repair and echocardiography was used for final assessment of gradient across right ventricular outflow tract.
Results	A total of 74 patients who underwent surgical repair of tetralogy of Fallot were included in the study. There were 45 (61%) males and 29 (39%) females with an age range from 2 year to 13 year. Mean age was 6.3 year. Repair was done through ventriculotomy with trans annular patch (TAP) in seven patients. Sixty-seven patients were operated without ventriculotomy. Patients in whom TAP was used had more severe pulmonary regurgitation, delayed extubation and long ICU stay than patients where repair was performed without ventriculotomy. Follow up echocardiography showed residual right ventricular outflow obstruction in patients operated without ventriculotomy and trans annular patch placement with mild obstruction in 29 (43%) patients while moderate and severe right ventricular outflow tract obstruction found in 36 (56%) and 2 (3%) patients respectively. All patients who underwent trans annular patch placement had moderate right ventricular outflow tract obstruction.
Conclusion	Despite concern about residual right ventricular outflow tract obstruction after correction of tetralogy of Fallot, our data showed moderate obstruction is well tolerated postoperatively.
Key words	Tetralogy of Fallot, Trans annular patch, Echocardiography.

INTRODUCTION:

Tetralogy of Fallot is the most common cyanotic cardiac defect and found in 3.5% of all infants born with a congenital heart disease. It is one of the common cardiac defects in which surgical intervention is required. Both palliative intervention as well as

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Correspondence: Dr. Iqbal Hussain Pathan Department of Paediatric Cardiac Surgery National Institute of Cardiovascular Diseases Karachi Email: driqbalnicvd@gmail.com definitive operations are reported for this condition. Natural history of these children shows that only 50% survive for the first years of life and very few patients reach beyond adolescence.¹

First intracardiac correction of tetralogy of Fallot was performed in 1955.² Despite the poor natural history of these children, now with timely surgical intervention, a good outcome is reported in many studies.³⁻⁸ Despite excellent long term results of a greater than 20 years survival with timely surgical intervention, there is growing concern for residual hemodynamic impairments like residual pulmonary insufficiency or pulmonary stenosis. Long standing severe pulmonary regurgitation has a well established association with right ventricular dilatation and dysfunction with functional impairment and arrhythmias; even sudden death secondary to ventricular fibrillation.

Right ventricular outflow tract enlargement was carried out by a large right ventriculotomy and trans annular patch applied for the complete relief of obstruction with acceptance of resultant severe pulmonary regurgitation. For a long period pulmonary regurgitation was thought to be an innocent condition which was supported by different studies.^{9,10} Later on it became evident that long term pulmonary insufficiency results in right ventricular dysfunction secondary to chronic volume overload with an increase in long-term morbidity and mortality.¹¹⁻¹³

It was also observed that many patients with right ventricular dysfunction require prosthetic pulmonary valve replacement for correction of the resultant pulmonary regurgitation secondary to tetralogy of Fallot repair with trans annular patch . This generated strong debate when to plan replacement prior to irreversible changes in right ventricle. Currently most surgeons try to adopt strategies that attempt to preserve the pulmonary valve complex; thereafter the residual pulmonary stenosis is confronted and the amount of residual pulmonary stenosis is now a matter of concern. This study was conducted to present the trends and outcomes of total correction of tetralogy of Fallot in our department.

METHODOLOGY:

This was a case series of patients of tetralogy of Fallot who were operated in the Department of Paediatric Cardiac Surgery, National Institute of Cardiovascular Diseases Karachi, from January 2016 to December 2016. Patients with absent pulmonary valve and pulmonary atresia were excluded from study. Patients who expired as well as those with missing data were also excluded. All the patients after surgery were shifted to PICU and put on mechanical ventilation with single or double inotrope support that included epinephrine and if required further supported with dopamine. Postoperative course was recorded and echocardiography advised in follow up. Patients were followed up to three month after surgery.

During surgery, trans annular patch was avoided whenever possible, even when a Hegar size of 2 to 4 mm less than desired was passed. When needed, the smallest posssible trans annular patch was used for right ventricular outflow tract reconstruction to minimize pulmonary regurgitation.

Moderate systemic hypothermia (28° C esophageal temperature), cardiopulmonary bypass with superior vena cava (SVC) and inferior vena cava (IVC) canulation and trans septal vent were used routinely. The ventricular septal defect was approached via the tricuspid valve and closed using a 0.2% glutaraldehyde-fixed autologous pericardial patch and the infundibular myomectomy was done and if required pulmonary valvotomy added. Adequacy of right ventricular outflow tract was assessed by Hegar dilators and if found to be less than 4mm according to normative Rowlat table, a longitudinal pulmonary arteriotomy across the pulmonary annulus was performed. A trans-annular patch of untreated pericardium was placed across the right ventricular outflow tract. Patulous trans annular patch was avoided. Same patch was also used to augment main or branch pulmonary artery if any narrowing was encountered. Valve leaflets were preserved to limit postoperative pulmonary regurgitation.

PICU course, extubation time and total PICU stay were noted. Severity of residual pulmonary stenosis or pulmonary regurgitation was assessed with echocardiography. Any additional surgery performed was also recorded. Data was presented as number, mean, range and percentage where appropriate.

RESULTS:

A total 74 patients were selected for the study. Patients' characteristics are shown in table I. Right ventricular outflow tract reconstruction was achieved without ventriculotomy in 67 (91%) patients. Seven (9%) patients required transannular patch placement of the right ventricle outflow tract. In the PICU, 92% patients had fast tract extubation with average PICU stay of 90 hours (range: 64 - 160 hours). Re do intervention was needed in one patient with moderate pulmonary stenosis with a gradient of 60 mmHg and with failure to wean off from ventilator. This patient underwent right ventricular outflow tract reconstruction with trans annular patch placement. The postoperative gradient in this patient was 28 mmHg with moderate pulmonary regurgitation were noted. Despite careful measuring of the outflow tract, two patients with severe pulmonary stenosis had a gradient of 65 mmHg and 75 mmHg. Both patients had a smooth postoperative course.

One of the patients was advised for redo surgery but the parents refused. In another patient, large conal artery was crossing right ventricular outflow tract and thus considered, unsuitable for re do surgery. Two patients required reexploration for tamponade. Both had an unremarkable course. Four

Table I: Patients' Information	
Gender	Male 45 (61%) Female 29 (39%)
Age	2 - 13 Year Mean 6.3 Year
Body Surface Area (BSA)	0.4 - 1.2 (m2) Mean 0.65 (m2)
Modified Blalock Taussing Shunt (n%)	6 (8%)

Table II: Outcome Between Two Group of Patients Operated With or Without TAP				
Variable	Without TAP	With TAP		
Extubation (n%)	Fast track 64 (95.5%) Delayed 3 (4.5%)	Fast track 4 (57%) Delayed 3 (43%)		
Residual Pulmonary Stenosis (n%)	Moderate 36 (56%): Severe 2 (3%)	Moderate 7 (100%)		
Pulmonary Regurgitation (n%)	Moderate 6 (5%)	Moderate 4 (57%) Severe 3 (43%)		

patients developed junctional tachycardia and were treated with amiodarone infusion. Fast tract extubation was achieved in 68 (92%) patients. Postoperative right ventricular out flow tract gradient was moderate in 43 (58%) patients while two had severe obstruction. Three (4%) patients had severe pulmonary regurgitation. Outcome difference of the two groups of patients with or without trans annular patch is shown in table II.

DISCUSSION:

Contradicting the traditional concept that post total correction of tetralogy of Fallot, free pulmonary regurgitation not only supports early recovery but is also well tolerated on long-term basis. There are several reports suggesting adverse outcome associated with long-term pulmonary insufficiency. Our study also demonstrated that patients with trans annular patch with resultant severe pulmonary regurgitation had prolonged PICU course. Functional testing has demonstrated that functional status is less optimal in tetralogy of Fallot patients with pulmonary regurgitation.¹⁴⁻¹⁷ Imaging studies of patients late after surgery has shown that pulmonary regurgitation severity is closely associated with trans annular patch implantation and is associated with right ventricular dilation and dysfunction, even in clinically asymptomatic patients.^{16,17} Association of right ventricular dilation with fatal arrhythmias is well established.¹⁸ Functional improvement and ventricular remodeling is reported in many papers following pulmonary valve insertion late after tetralogy of Fallot repair, which further support the adverse effects

of TAP and pulmonary regurgitation.^{19,20} Considering these reports, several surgeons began to preserve the pulmonary valve or at least the annulus.²²

Many surgeons are in favor of the complete primary repair of tetralogy of Fallot as early as possible even in neonates.^{22,23} Nevertheless there are many surgeons who showed concerns about the use of a large trans annular patch during the neonatal period and very early correction with resultant free pulmonary regurgitation and late re operation for pulmonary valve replacement. These concerns lead others to advocate not to operate in neonates.

In this study, we adopted a simple right atrial approach for trans tricuspid valve excision or division of muscle bands and if required pulmonary valvotomy and accepting a restrictive right venricular outflow tract at the time of correction on the basis of normative data of the pulmonary annulus. This helped in limiting the use of trans annular patch. We accepted sizing of the pulmonary valve even up to three times less than the required diameter. Furthermore, limiting the pulmonary insufficiency, has obvious positive impact on long-term outcome. For this reason majority of patients in this study had moderate pulmonary stenosis at end of repair which was acceptable.

Only three patients were admitted for re do surgery during the study period. In these, we have implanted bioprosthetic valve with hood of bovine pericardial patch. In our series two patients had residual severe pulmonary stenosis. A standard approach in these patients is provision of homograft conduit.

CONCLUSIONS:

For total correction avoidance of trans annular patch enlargement by aggressive infundibular myomectomy and repair with preservation of the pulmonary annulus integrity; if required pulmonary valvotomy, has worked well. Moderate pulmonary stenosis in our patient population was not of major concern as better early recovery coupled with a preserved annulus and a moderate gradient were achieved. Failure of regression of gradient was noted at three months follow up on echocardiography which requires a long-term follow up.

REFERENCES:

- 1. Bertranou EG, Blackstone EH, Hazelrig JB, Turner ME, Kirklin JW. Life expectancy without surgery in tetralogy of Fallot. Am J Cardiol. 1978;42:458-66.
- 2. Lillehei CW, Cohen M, Warden HE, Read RC, Aust JB, Dewall RA et al. Direct vision intracardiac surgical correction of the tetralogy of Fallot, pentalogy of Fallot, and pulmonary atresia defects; report of first ten cases. Ann Surg. 1955;142:418-42.
- Kirklin JW, Ellis FH Jr, McGoon DC, DuShane JW, Swan HFC. Surgical treatment for the tetralogy of Fallot by open intracardiac repair. J Thorac Surg 1959;37:22-51.
- 4. Shumway NE. Total surgical correction of tetralogy of Fallot. Arizona Med 1966;S106.
- Castaneda AR, Freed MD, Williams RG, Norwood WI. Repair of tetralogy of Fallot in infancy. Early and late results. J Thorac Cardiovasc Surg. 1977;74:372-81.
- Reddy VM, Liddicoat JR, McElhinney DB, Brook MM, Stanger P, Hanley FL. Routine primary repair of tetralogy of Fallot in neonates and infants less than three months of age. Ann Thorac Surg. 1995;60:S592-6.
- Pigula FA, Khalil PN, Mayer JE, del Nido PJ, Jonas RA. Repair of tetralogy of Fallot in neonates and young infants. Circulation. 1999;100:II157-61.
- 8. Hirsch JC, Mosca RS, Bove EL. Complete

repair of tetralogy of Fallot in the neonate: results in the modern era. Ann Surg. 2000;232:508-14.

- 9. 9.Nollert G, Fischlein T, Bouterwek S, B[^]hmer C, Klinner W, Reichart B. Long-term survival in patients with repair of tetralogy of Fallot: 36-year follow-up of 490 survivors of the fi rst year after surgical repair. J Am Coll Cardiol. 1997;30:1374-83.
- Lillehei CW, Varco RL, Cohen M, Warden HE, Gott VL, DeWall RA, et al. The first open heart corrections of tetralogy of Fallot: A 26-31 year follow-up of 106 patients. Ann Surg. 1986;204:490-502.
- Wessel HU, Cunningham WJ, Paul MH, Bastanier CK, Muster AJ, Idriss FS. Exercise performance in tetralogy of Fallot after intracardiac repair. J Thorac Cardiovasc Surg. 1980;80:582-93.
- 12. Carvalho JS, Shinebourne EA, Busst C, Rigby ML, Redington AN. Exercise capacity after complete repair of tetralogy of Fallot: deleterious effects of residual pulmonary regurgitation. Br Heart J. 1992;67:470-3.
- Gatzoulis MA, Balaji S, Webber SA, Siu SC, Hokanson JS, Poile C, et al. Risk factors for arrhythmia and sudden cardiac death late after repair of tetralogy of Fallot: a multicentre study. Lancet. 2000;356:975-81
- Singh GK, Greenberg SB, Yap YS, Delany DP, Keeton BR, Monro JL. Right ventricular function and exercise performance late after primary repair of tetralogy of Fallot with the transannular patch in infancy. Am J Cardiol. 1998;81:1378-82.
- 15. Schamberger MS, Hurwitz RA. Course of right and left ventricular function in patients with pulmonary insufficiency after repair of tetralogy of Fallot. Pediatr Cardiol. 2000;21:244-8.
- Nollert GD, Dabritz SH, Schmoeckel M, Vicol C, Reichart B. Risk factors for sudden death after repair of tetralogy of Fallot. Ann Thorac Surg. 2003;76:1901-5.
- 17. d'Udekem d'Acoz Y, Pasquet A, Lebreux L, Ovaert C, Mascart F, Robert A, Rubay JE. Does right ventricular outflow tract damage

play a role in the genesis of late right ventricular dilatation after tetralogy of Fallot repair? Ann Thorac Surg. 2003;76:555-61.

- Therrien J, Siu SC, Harris L, Dore A, Niwa K, Janousek J, Williams WG, Webb G, Gatzoulis MA. Impact of pulmonary valve replacement on arrhythmia propensity after repair of tetralogy of Fallot. Circulation. 2001;103:2489-94.
- 19. Naito Y, Fujita T, Manabe H, Kawashima Y. The criteria for reconstruction of the right ventricular outflow tract in total correction of tetralogy of Fallot. J Thorac Cardiovasc Surg. 1980;80:574-81.
- Katz NM, Blackstone EH, Kirklin JW, Pacifico AD, Bargeron Jr LM. Late survival and symptoms after repair of tetralogy of Fallot. Circulation. 1982;65:403-10.

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Conflict of Interest:

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- Eyskens B, Reybrouck T, Bogaert J, Dymarkowsky S, Daenen W, Dumoulin M, Gewillig M. Homograft insertion for pulmonary regurgitation after repair of tetralogy of Fallot improves cardiorespiratory exercise performance. Am J Cardiol. 2000;85:221-5.
- Stellin G, Milanesi O, Rubino M, Michielon G, Moreolo GS, Boneva R et al. Repair of tetralogy of Fallot in the first six months of life: transatrial versus transventricular approach. Ann Thorac Surg. 1995;60:S588-91.
- Kanter KR, Kogon BE, Kirshbom PM, Carlock PR. Symptomatic neonatal tetralogy of Fallot: repair or shunt? Ann Thorac Surg. 2010;89:858–63.