

GREAT ARTERY RATIO: DOES IT REALLY MATTERS IN TOTAL CORRECTION OF TETRALOGY OF FALLOT?

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Contribution

IHP conceived the idea, planned the study and drafted the manuscript. SKB, AMK helped in acquisition of data and did statistical analysis. IHP drafted and critically revised manuscript. All authors contributed significantly to the submitted manuscript.

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ABSTRACT

Objective: To investigate the significance of Great artery ratio, for decision of right ventricular outflow tract management in patients with Tetralogy Of Fallot.

Methodology: A retrospective cross sectional study including surgical record of patients operated for total correction of Tetralogy Of Fallot from January 2015 to December 2015 in Department of Paediatric cardiac surgery NICVD, was done. Patients whose aortic annulus were documented in record were included in study. A relation of great artery ratio with cut off value of 0.55 with Trans Annular Patch Enlargement Repair and patient outcome as defined by time of extubation lesser or greater than six hours and survival after surgery was evaluated. Data was analysis on SPSS 21.

Results: Of total 66 patients, male were 66.7%. Great artery ratio of 0.55 or less was calculated in 39.4% of patients, while trans annular patch enlargement repair of Tetralogy Of Fallot (TOF) was observed in 15% of patients with 6 patients in GA ratio 0.55 or less required trans annular patch enlargement compared to 4 patients in GA ratio of >0.55 ($p=0.14$). However a significant association of mortality was observed with 15.4% of patients expired in group with low GA Ratio compared to no mortalities in patients with GA ratio of >0.55 ($p<0.05$).

Conclusion: Although Trans annular patch enlargement with low GA ratio was required in very few patients but it was associated with decreased survival.

Key Word: Tetralogy Of Fallot, Trans Annular Patch Enlargement, Great Artery Ratio

INTRODUCTION

Tetralogy Of Fallot is a congenital cardiac malformation that is consisted of right ventricular outflow tract stenosis and ventricular septal defect, each with specific morphological characteristics and secondary involvement of other structures. Stenson in 1671 first described the morphology of this cardiac malformation, later Fallot published his series of cases in 1988.¹ Van Praagh considered insufficient growth of distal pulmonary conus as main event leading resultant hypertrophy of proximal conus along with inadequate dorsal growth of the crista supraventricularis leaving a junctional ventricular septal defect.² Surgical intervention for Tetralogy Of Fallot was performed by Blalock with the idea of Taussing in the 1940's by subclavian artery-pulmonary artery anastomosis known as classical Blalock and Taussing shunt, a ray of hope for blue babies³. Today it is about 76 years since the the surgical intervention for Tetralogy Of Fallot thanks to pioneer work by Blalock, Lillehei, Kirklin, and many more in treating this congenital heart defect.⁴⁻⁷ Despite of this all still Controversy persists regarding best management strategy. Surgeons' s choices range from those who prefer of neonatal complete correction in every patient to those favor for delaying total correction until suitable age and weight achieved later in life, using initial palliation with modified Blalock Taussing Shunt, utilizing conduit to rout blood from subclavian artery to pulmonary artery. The first surgical correction of TOF involved ventricular septal defect closure via right ventriculotomy and right ventricular outflow tract reconstruction with trans annular patch enlargement of the right ventricular out flow tract was reported in 1954.⁷ Persistent right ventricle volume loading due to pulmonary regurgitation secondary to trans annular patch enlargement (TAPE) leads to right ventricular dysfunction, dilation, and arrhythmias.⁸ These complications led to several modification in classical technique like transatrial repair without ventriculotomy and transatrial-transpulmonary repair with the aim of preservation of right ventricular function by avoiding or minimizing ventriculotomy.⁹⁻¹¹ Till today, the z score is used to guide for trans annular patch enlargement repair if desired diameter of right ventricular out flow tract is not achieved despite of adequate infundibular myomectomy and valvotomy.¹²⁻¹³ Calculation of z score has many intrinsic limitations¹⁴⁻¹⁵. When operating on small pulmonary annulus generally transannular patch is considered for the patients with Z-scores of less than - 2. Nevertheless there are reports of successful transatrial repair in patients with Z-scores of as low as - 3 and even - 4.¹³ Recently an interest has been observed regarding ratio of great arteries annulus (pulmonary valvular annulus size to aortic valve annulus size [GA Ratio]) in decision making regarding use of trans annular patch enlargement.¹⁶

We conducted a retrospective review of record to find out relation of great artery ratio with patch enlargement of right ventricular outflow tract in patients with TOF. Our secondary objective was to look out its association with patient outcome like extubation time and survival.

METHODOLOGY

A reterospective cross sectional study was conducted by reviewing record from January 2015 to December 2015 of patients operated in paediatric cardiac surgery department of NICVD. All the patients operated for total correction of Tetralogy Of Fallot were reviewed. Patients whose aortic annulus and surgical note were available included in study. Post operative course was traced from ICU record of patients. GA ratio of 0.55 was considered as cut off

All the patients underwent routine investigation preoperatively along with echocardiography imaging. In few cases CT angio record was available.

Intra-operative Course: All total corrections were carried out via median sternotomy using hypothermic cardiopulmonary bypass(28°C). After establishing bicaval cardiopulmonary bypass mobilization of the proximal pulmonary arteries with division of the patent ductus or ligamentum arteriosum was done. After aortic cross-clamping myocardial protection was achieved with antegrade intermittent cold blood hyperkalemic cardioplegia with added xylocaine and bicarbonate. Dose of cardioplegia was calculated as per weight. Cardioplegia was repeated in every 20 minutes till cross clamp removed. In all patients trans atrial approach was used for total correction. Right ventricular out flow tract was reconstructed by infundibular myomectomy and if required valvotomy. Adequacy of Right ventricular out flow tract was confirmed by passing Hegar dilator of required diameter, even one to two size smaller Hegar dilator passage accepted. In remaining case trans annular patch of untreated pericardium was used for right ventricular outflow tract reconstruction, trans annular patch extending onto the proximal left and/or right pulmonary artery if required. While ventricular septal defect closed with continuous suture technique with treated pericardium with 0.2% gluteraldehyde. Transoesophageal echocardiography was done after weaning from bypass to refine surgical repairs and to avoid significant residual defects and cardiac function access.

Post-operative course: All the patients were shifted to Pediatric ICU on mechanical ventilation and single ionotrope support and Nitroglycerine infusion. It is general policy of our institute to follow fast tract extubation; extubation within six hours unless any reservation likes bleeding, unsatisfactory hemodynamic and compromised blood gases were observed. Operative and postoperative features are mentioned in table 1

Variables were presented as number, percentage and mean with range as appropriate. The X-2 test was used to test for strength of association between the variables. The association between GA ratio with use of trans annular patch enlargement repair and patient outcome was analyzed. Statistical test was two-sided and type I error was controlled at 0.05. Analyses were performed using SPSS 21.

Table 1: Operative and Postoperative Variables of Study Population (n=66)

VARIABLES	VALUES
Pump time(min)	40m or less = 11(16.7%) 41 m to 50 m = 27(40.9%) 51 m to 60 m= 11(16.7%) >60 m= 6(24.7%)
X time (min)	<30 =41(62.1%) 30 to 50 =19(28.8%) >50 = 6(9.1%)
Trans annular patch enlargement	10(15.2%)
Extubation Fast track	31(47%)
Delayed	35(53%)

RESULTS

There were 66 patients with mean age and weight of 6.2 years and 12.08kg respectively. About 44(66.7%) were males. Demographic characters of patients with great artery ratio are shown in table 2. Great Artery ratio of 0.55 or less was used as cut off value. Great Artery ratio of 0.55 or less was documented in 26(39.4%) patients. While Right ventricular outflow tract reconstruction with trans annular patch repair to achieve desired area was done in 10(15.2%) patients. Out of 26 patient with GA Ratio of 0.55 or less 6(23.1%) patients requires trans annular patch compared to 4(10%) patients with GA Ratio of > 0.55, required trans annular patch enlargement out of 40 patients ($p = 0.148$). Further analysis to find any effect on patient outcome like extubation and survival showed that out of 26 patients with GA Ration 0.55 or less; 4 (15.4%) patients expired compared to no mortalities in group with GA Ratio >0.55 ($p < 0.05$) and delayed extubation was observed in 15(57.7%) patients with GA Ratio 0.55 or less as compare to 20 (50%) patients with GA Ratio >0.55 ($p = 0.541$). Interestingly

Table 2: Demographic Characters of Study Population (n=66)

VARIABLES	VALUES
Gender	M 44(66.7%), F 22(33.3%)
Age (years)	Mean age 6.2 years, (range 2 to 12 years)
Weight (kg)	Mean 12.08 kg, (range 2 to 37 kg)
Great artery ratio	
.55 or less	26(39.4%)
> .55	40(60.6%)

there was no significant association of trans annular patch enlargement repair with mortality of patient with 2(20%) mortality out of 10 patients requiring trans annular patch enlargement repair compare to 7(12%) patients out of 56 patients underwent total correction without trans annular patch enlargement ($p = 0.524$).

DISCUSSION

Tetralogy Of Fallot is the one of the most common cyanotic heart defects presented for surgical intervention.¹⁷ Management of these patients significantly evaluated from the era of classical Blalock Tassung Shunt involving end to side anastomosis of subclavian artery to branched pulmonary artery to total correction in neonatal age. Despite of advances in technical management of these patients, ideal approach for reconstruction right ventricular outflow tract is still the area of controversy. Every paediatric cardiac surgeon desire to preserve pulmonary valve when we observe late effect of free Pulmonary regurgitation after trans annular patch enlargement repair of TOF despite some reports to the contrary results. Ideally we require total correction for Tetralogy of Fallot with no pulmonary regurgitation and no residual Right ventricular outflow tract stenosis. However this is possible in only ideal environment. Because in immediate post operative period on operation table it is difficult to decide how much residual pulmonary stenosis is acceptable and how much Pulmonary regurgitation is well tolerated over long period without developing right ventricular volume load keeping in mind that quantification of by transesophageal echocardiography is subjective and gives a gross idea only, in addition to the fact that Right Ventricular restrictive physiology, often present before and

after surgery, "minimizing" the severity of Pulmonary regurgitation. Despite of awareness of hazardous of trans annular patch for Right ventricular outflow tract reconstruction; ventriculotomy with trans annular patch placement remains the most common practice of Right ventricular outflow tract management with 52% of cases irrespective of primary repair and repair after previous palliation.¹⁸ Same trend was mentioned by Luijten and associates observed that more than 60% of Tetralogy Of Fallot patients were repaired with trans annular patch enlargement of right ventricular out flow tract.¹⁹ These and many more suggestive of high incidence of trans annular patch enlargement. Considering the hazardous of trans annular patch placement like ventriculotomy associated myocardial scarring and coronary artery damage that can result in arrhythmia and impair right ventricular function, right ventricular volume loading secondary to long standing free pulmonary regurgitation leading to ventricular dilation, dysfunction, heart failure, and cardiac death.²⁰⁻²¹ Our strategy for total correction is avoidance of trans annular patch enlargement by aggressive infundibular myomectomy and repair with preserving the pulmonary annulus integrity, if required pulmonary valvotomy was done. We tolerated 1 to 2 sizes down Hegar dilator if patient tolerate across the Right ventricular outflow tract. We tolerated gradient of 50 to 60mmHg if right ventricular contraction is comfortable. Because of all these measure we have trans annular patch enlargement repair incidence of 15% compare to above statics of as high as 60%. We used Rowlett table for sizing of Right ventricular outflow tract with Hegar dilator²¹. However measure calculated in Rowlett table were from western population. Many centers favoring calculating z score of individual patients. The z-score indicates how many standard deviations the case is from the mean. Nevertheless the z-score has its own limitations like does not correct any errors those results from non homogenous population distribution. Further z-score is complicated value influenced by height, body weight, body surface area and population group. Even different authors suggested different z scores for same body surface area for pulmonary annulus management having different blood flow suggestive of intrinsic limitation.²¹⁻²⁴ Considering these all limitation a new tool of great artery ratio is introduced. Kwang Ho Choi and other presented their results with strong association of trans annular patch enlargement with great artery ratio; GA ratio cutoff value for trans annular patch enlargement was 0.559 with a sensitivity of 82.5% and a specificity of 89.0%¹⁶. However our results failed to support its association with requirement of trans annular patch enlargement for total correction. This may be due to our practice of accepting high gradient after weaning from bypass if desired Hegar dilator is passed and clinically and visually patient as a whole maintaining hemodynamic with comfortable right ventricular contraction. This may be due to

our frequent observation that postoperative Transthoracic echocardiography detect low gradient at right ventricular outflow tract compare to post weaning in operation room. However a significant association was observed regarding survival of patients with low GA Ratio that require further evolution of this outcome. Considering retrospective study it is difficult to commit on cause of mortality of these patients

LIMITATIONS

Considering a retrospective study it has inherent limitations. In our study population we found very high mortality unexpectedly. Possible causes of these mortalities are difficult to elaborate due to lack of proper records.

CONCLUSION

For TOF patients Trans annular patch enlargement with low GA Ratio is required rarely. However this procedure is associated with decreased survival.

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