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Results of repair of Tetralogy of Fallot in Adults: analysis of 27 cases

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Abstract:
Introduction*: Tetralogy of Fallot (TOF) is the commonest congenital cyanotic heart disease in children affecting 3 out of 10000 live birth (7-8% of all congenital cardiac diseases). Intracardiac repair is performed before one year of age but in India we still get adult patients with TOF.*
Materials and Methods*: From January 2012- December 2018, 27 adults had repair of TOF in our institutute. We excluded double outlet right ventricle (DORV), TOF with pulmonary atresia, TOF with atrioventricular septal defect and other complex anomalies. Follow up was upto 1 year.*
Results*: Mean hospital stay was 10.84 ± 4.08 days. 4 patients (14.81%) had re-exploration, 5 patients (18.51%) had pleural effusion, 7 patients (25.92%) had right ventricular dysfunction, 7 patients (25.92%) had tricuspid regurgitation grade 2+, 3 patients (11.11%) had junctional ectopic tachycardia with 3.70% mortality; within first 30 days. Analysis of 1 year follow up revealed: pulmonary regurgitation in 6 patients (23.07%), tricuspid regurgitation in 8 patients (30.76%), Residual VSD in 1 patient (3.84%), residual gradient across right ventricular outflow tract (more than 40 mmHg) in 1 patient (3.84%), NYHA Class II in 7 patients (26.92%%), RBBB in 20 patients (76.92%), no deaths (0) or Atrial fibrillation.*
Conclusion: *From our study we found adult TOF surgery causes more complications except post- op JET than in paediatric counterparts. However, our results are comparable with other centres.*
Keywords: *Adult Tetralogy of Fallot, Adult Congenital Surgery.*

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I. Introduction

Niels Seensen first described Tetralogy of Fallot(TOF) but it was Etienne Louis Arthur Fallot [1] in 1888 who correctly described the four components of TOF. It has an incidence of 3 per 10,000 live-births and constitutes (7 -8)% of all congenital cardiac diseases. TOF is the commonest congenital cyanotic heart disease.[2]Unrepaired TOF has 25% ten year survival, it is around 10% by third decade and only 3% beyond fourth decade. [3] Though it is called tetralogy the basic defect is antero- lateral displacement of infundibular septum giving rise to large malaligned sub- aortic ventricular septal defect(VSD), right ventricular outflow tract obstruction(RVOTO) which may be sub- infundibular, infundibular, valvular, pulmonary arterial(PA) or at branch PA level, overriding of aorta and right ventricular hypertrophy (RVH). [4,5,6]

Effort dyspnoea , cyanosis, poor exercise, tolerance, headache, haemoptysis, clubbing, squatting and cyanotic spells are the most common presenting features of TOF.[7] TOF is associated with various cardiac anomalies such as bicuspid pulmonary valve (60%), right sided aortic arch (25%), ASD, PDA, AVSD, coronary anomalies or part of other more complex congenital disease. [8] Most of the cardiac centres in India are adopting single- stage repair over two- stage repair [palliative Blalock Taussig (BT)- shunt followed by Intra-cardiac repair] preferably in first year of life. [9]

However absence of paediatric cardiac centres in different parts of India, especially in eastern part and problems of accessibility and affordability caused adult patients requiring repair of TOF as the primary operation or as a secondary operation having one palliative shunt (BT shunt) in childhood. We have analyzed the post operative results (30 days) and results of follow- up upto 1 year retrospectively.

II. Materials And Methods

This is a retrospective observational study. We have collected data from hospital records for all adult TOF patients operated for total correction from January 2012- December 2018. Due permission was taken for this study from the hospital ethical committee. We have assessed patients upto 30 days post- op and 1 year after operation in terms of complications. TOF with pulmonary atresia, [10] DORV[11] and associated with other complex defects are excluded from study.

All operations were done by the same set of surgeons in the same right atrial approach with pulmonary valvotomy when required. [12]

Closure of VSD was done with Dacron patch, right ventricular outflow tract (RVOT) was cleared with excision of muscle bands, pulmonary valvotomy was done adequately, and pulmonary arterial (PA) or transannular patch (TAP) was placed with glutaraldehyde treated pericardium as per individual requirement [13]. (figure 1 and 2) Two TOF patients required polytetrafluoroethylene (PTFE) monocusp valve repair for absent pulmonary valve .[14]

We have used Del Nido cardioplegia [15] in all the cases and the procedures were carried out in usual fashion.

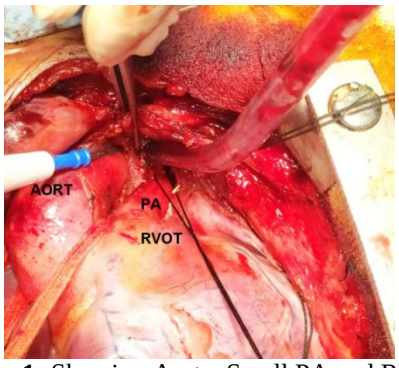

Figure 1: Showing Aorta, Small PA and RVOT

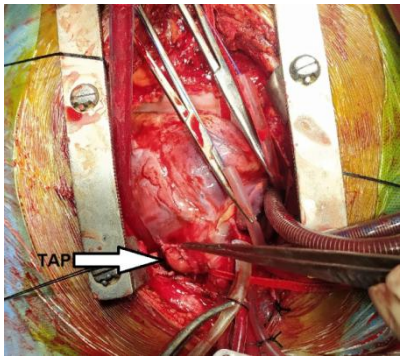

Figure 2: Showing trans-annular patch (TAP)

III. Results

Of the total 27 patients 19 were males and 8 were females. The mean age of patients were 22.84 ± 3.35 years. Mean hospital stay was 10.84 ± 4.08 days. All patients were monitored in ITU, ward and then upto 30 days post- op in outpatient department after discharge. All data collected from hospital and analyzed in this study for outcome of surgery upto 30 days; and data collected upto 1 year for analysis of results from 30 days post- op period upto 1 year. Major complications observed upto 30 days of post- operative period are (figure 3)

- Re- exploration- 4 patients (14.81%)
- Pleural effusion- 5 patients (18.51%)
- RV dysfunction- 7 patients (25.92%)
- TR (Gr II) - 7 patients (25.92%)
- PR mod- 5 patients (18.51%)
- JET – 3 (patients 11.11%)
- Mortality- 1 patients (3.70%)

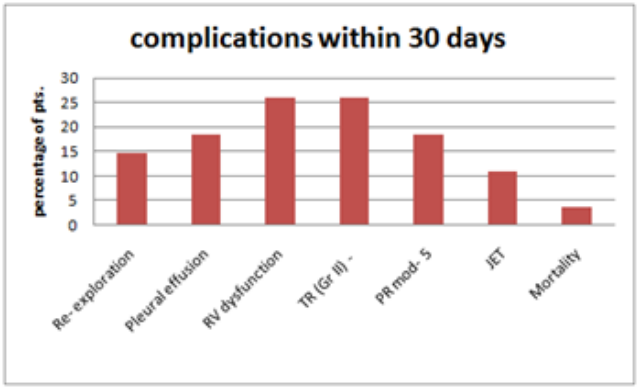

Figure 3: Complications within 30 days

Analysis of 1 year follow up revealed- (taking a total of 26 patients and excluding that one death within 30 days)

- PR- 6 (23.07%)
- TR (mod) – 8(30.76%)
- Residual VSD- 1 (3.84%)
- RVOTO- 1 (3.84%)
- NYHA Class II or more- 7 (26.92%)
- RBBB- 20 (76.92%)
- AF- 0
- Death- 0

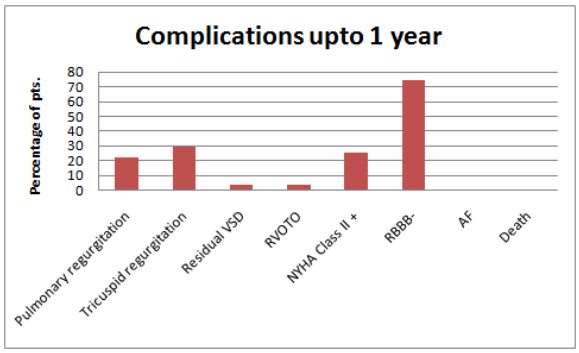

Figure 4: Complications upto 1 year

IV. Discussion

Nowadays TOF is repaired within first year of life with very low complication rates. However we still continue to get TOF in adult age (> 18 years in this study) for lack of paediatric cardiac centres in various parts of India and financial constraints of patients. Currently some centres report mortality of about 2-3% [16] in paediatric TOF repair. However we report a mortality of 3.7%. This result may be due to long standing cyanosis causing RV dysfunction and cerebral complications. Palliative shunt (Blalock-Taussig) was performed in initial days in many cardiac centres for lack of infrastructure or lack of trained healthcare provider. In our study 5 patients (18.51)% had B-T shunt during definitive repair for TOF conducted as second operation in adult life.

We have used transannular patch (TAP) for relief of RVOTO and significant gradient (> 40 mmHg). [17]For prevention of PR and its subsequent problems often we have created PTFE monocusp valve with good results observed subsequently followed upto 1 year.

We have observed that TOF repair in adults causes comparatively more bleeding and re- exploration than paediatric counterparts. RV dysfunction is also more than paediatric patients. However, Junctional Ectopic Tachycardia (JET) is more common in paediatric group.[18]

V. Conclusion

From our study we found adult TOF surgery causes more complications except post- op JET than paediatric counterpart. However the results are still encouraging and provide better relief of symptoms than continuing life without surgery and previously done B-T shunts with often suboptimal flow.

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