Original Article

Tetralogy of Fallot

Surgical Correction of Grown Up Tetralogy of Fallot

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ABSTRACT

Objective: To get an audit of outcome of our patients operated for total correction for grown up Tetralogy of Fallot
Study Design: Observational / analytic study.
Place and Duration of Study: This study was conducted at the Department of Cardiac Surgery, NICVD, Karachi from January 2015 to August 2016.
Materials and Methods: We reviewed our surgical record and collected the data of patients with age 18 years and beyond, who underwent for total correction in Tetralogy of Fallot. We had included the patients whose prospective record of their surgical as well as socioeconomic outcome.
Results: Total 35 patients were identified , out of 35 patients 19 were selected as final cohort of patients for our study they included 11(48%) females and 8 (42%) males with age range of 18 to 28 years. Procedures for TOF repair included trans-annular patch (n=7), trans-ventricular (n=5) , trans-atrial (n=2). While the remaining patients (n=3) had combined approaches (tran-atrial with trans-pulmoary or trans-ventricular with trans-pulmonary). The 30-day mortality rate was 16% (right ventricular failure n=1; tamponade n=1; low cardiac output with pulmonary edema as a result of residual ventricular septal defect n=1) . 3 patients were re-explored due to mediastinal bleeding with one of them had developed cardiac tamponade.
Follow-up of minimum 3 month to maximum of 15 months was feasible in 16 out of 19 survivors, improvement in functional class (NYHA) was observed in 11 patients.
Conclusion: Complete repair of TOF in patients 18 years or older is possible but carries increased operative risk. Survivors have improvement in their functional class as well as social status however it is difficult to commit on economical productivity of patients.
Key Words: Grown Up, Cyanotic Heart Defects, Tetralog of Fallot, Total Correction

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INTRODUCTION

Congenital heart defects consists of about 1/3 of all congenital anomalies . Broadly congenital heart defects are classified in cyanotic and acyanotic defects. Tetralogy of Fallot is considered most common cyanotic heart defect presented for total correction in paediatric population[1] . Tetralogy of Fallot was first described by Etienne-Louis Fallot in 1888; it consists of a ventricular septal defect, right ventricular outflow tract obstruction, overriding of aorta, and right ventricular hypertrophy. First successful correction of this lesion was reported by Leheli in 1955[2] . Ever since it's identification, more and more efforts have been made for early age total correction. Currently definitive repair beyond childhood is extremely uncommon.

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However in the developing countries the practice of delayed repair is relatively common due to delayed diagnosis or lack of cardiac surgical facilities responsible for late surgical repair in adult hood or even beyond . These patients are often those who are either palliated with systemic to pulmonary artery shunt or they have less severe variety of Tetralogy of Fallot (Pink Tetralogy). As suggestive of poor natural history', indicates that hemodynamic impairment in the survivors to adulthood is less sever . It is still controversial whether surgery after long-standing cyanosis in adult patients with Tetralogy of Fallot has how much impact on survival[3,4,5] and socioeconomic status. Neverthless Nollert and colleagues showed normal life expectancy after definitive repair of Tetralogy of Fallot in adults of 18 years or beyond[6] . Despite of strong recommendation we still received patients diagnosed with Tetralogy of Fallot from cardiology departments for total correction beyond the childhood who have either had only a palliative procedure or not undergone any surgical intervention and first time presented or ignored past advice for surgical intervention due to social taboos . Few studies describe the outcome of such patients after total correction . However, the long standing hypoxia that is

major feature of patients with Tetralogy of Fallot may results in variable cerebral complications, compromised myocardial function, and an increased occurrence of .ventricular and atrial arrhythmias [6,7,8].

This study was conducted to analyze the early and intermediate results of patients operated for total correction of TOF over the period of 20 months.

MATERIALS AND METHODS

From January 2015 to April 2016, total 35 adults with Tetralogy Of Fallot referred to surgery from the cardiology department. Among these 19 were included in study because their complete E-record and recent follow up was available. Following Data was collected pre-admission (NYHA class), hospital course (pre and post op rhythm, intervention and complications) and socioeconomic status by review of record and interview on post-op follow up.

Statistical analysis: Variables are presented as number ,percentage and mean with range as appropriate .. Analyses were performed using SPSS 21.

RESULTS

Preoperative Findings: There were 11 (48%) female and 8 (42%) male with median age of **20** (18-28) years. All patients were cyanotic, with oxygen saturation from From 70% to 91%, and hemoglobin from 10 to 21 g/dl. 5 patients had undergone previous modified Blalock-Tussig shunt in their childhood with two patients having functional shunts. All patients had sinus rhythm. Out of 11 female patients, 1 was married compared to 2 male patients out of 8. However none of them had any children. Among 19 patients, 3 pateints were functional class NYHA 2, 11 patients were NYHA 3 and rest of NYHA 3+ status.

Surgical Techniques and Results: All surgeries were performed under moderate hypothermic cardiopulmonary bypass using cold blood cardioplegia antegrade for myocardial protection. Mean Cardiopulmonary bypass time was 105 (60-164) min with mean aortic cross-clamp time of 73 (26-123) min. Different approaches were required for total correction in which trans-annular patch repair in 7 patients (26%) ,while trans-ventricular and trans-atrial approaches for total correction were used in 5 and 2 patients respectively. Rest of the patients required combined approach either trans-atrial with trans-pulmonary or trans-ventricular with trans-pulmonary.

Perioperative Management and Complications: hn All patients were weaned off from Cardio pulmonary bypass on ionotropic support in which 4 (21%) patient required double ionotrope support, 8 (42%) patients were successfully followed by fast tract protocol. Early postoperative course of 9(47%) patient remained unremarkable while 6(31%) patient have mediastinal bleeding in which 3(16%) required re-exploration and delayed extubation, while 2 patients (10%) developed multi-focal arrhythmias. Out of 19, 16 (84.2%) survived while 3(15.8%)expired during same hospital admission. Among the non survivors, one had sever Right ventricular dysfunction; who had preoperative enlarged right ventricle with severe tricuspid regurgitation, other developed pulmonary edema and hemodynamic instability secondary to residual ventricular septal defect and the last of those three had cardiac arrest secondary to cardiac tamponade.

Mid- and Long-Term Follow-Up: Follow-up consisted of minimum 3 month and maximum 13months between surgery and the last clinical examination or telephonic contact. After successful surgery NYHA class improvement was observed in 11(58%) patients with 5(26%) had equivocal reply while social improvement as asked by family members 12(63%) patients had improve social attitude than 4(21%) had equivocal outcome while one women had a successful marriage . However non of patient has an obvious improvement in economic productivity.

DISCUSSION

Review of our surgical audit suggested that Tetralogy Of Fallot can be operated in adult patients but with significant risk of postoperative morbidity and mortality as compared to paediatric patients[9,10,11,].

Nevertheless the fact of higher age as a risk factor for total correction of TOF for morbidity and mortality in long term follow up is also documented in many studies.[12,13,14] Despite of significant advances in surgical techniques, myocardial protection, postoperative care and understanding pump physiology in the past 30 years the operative mortality in adult patients operated for TOF remains high. This high mortality may be responsible of number of reasons due to long-standing cyanosis, which have independent effect on perioperative mortality and morbidity[15]. The long standing hypoxia results in right ventricular dysfunction secondary to myocardial fibrosis[16,17]. There is no obvious evidence for the support of two-stage repair with improvement of oxygen saturation before correction may improve surgical outcome in adult patients with Tetralogy of Fallot. Currently, results for total correction are reported as better as less than 1% operative mortality for Tetralogy of Fallot [18] in younger patients. However, operative mortality is age dependent; in experienced centers, it is less than 1% for patients under one year and 4.4% for older patients[19] . In another study 5.1% early mortality rate for older patients was observed[20]. Considering these results, our study population appears to have high mortality, but we should understand the fact this population was out of 35

patients operated during that period; if we calculate the mortality percentage out of total population than mortality will be 9%. Postoperative mediastinal bleeding is a frequent observe complication requiring multiple transfusion of blood and its products.

Postoperative ventricular and supraventricular dysrhythmias caused significant morbidity in postoperative period in adult patients. There are several studies suggestive of an association between right ventricular functional status and the occurrence of dysrhythmia[21,22,23] and demonstrated that early intervention may protect form some of these dysrhythmias[22,24]. We observed sinus tachycardia in one patient and atrial fibrillation in other patient. Both of these patient were treated with amiodarone infusion. There was both objective and subjective improvement in the well being of patients and was observed by patients themselves and by their close associate, which confirms the observations in previous studies [4,6,11]. And this remains a concern regarding long term outcome of such patients. Nevertheless it is reported in one study showing long-term survival up to 35 years after correction of grown up Tetralogy of Fallot, which is very similar with the general life expectancy[25] .Though social improvement was observed we could not commit on employability or socioeconomic contribution of our individual patients.

CONCLUSION

It is concluded that Hormonal contraceptive method is the most commonly used method in females and condoms in the male clients.

Conflict of Interest: The study has no conflict of interest to declare by any author.

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