

Research Article

Clinical profile and surgical outcomes of children presenting with teratology of Fallot

Josephat M Chinawa^{1*}, Agarwal Vijay², Sarang Gaikwad³, Bhadra Trivedi⁴ and Bartholomew Chukwu⁵

¹Associate Professor and Pediatric Cardiologist, College of Medicine, Department of pediatrics, University of Nigeria/University of Nigeria Teaching Hospital (UNTH), Ituku, Ozalla, Enugu, Nigeria

²Chief Pediatric Cardiac Surgeon, Fortis Hospital limited, Mulund goregaon Link road, Bhandup (West), Mumbai, India

³Pediatric Cardiac Surgeon, Fortis Hospital limited, Mulund goregaon Link road, Bhandup (West), Mumbai, India

⁴Pediatric Cardiologist, Fortis Hospital limited, Mulund goregaon Link road, Bhandup (West), Mumbai, India

⁵Senior Lecturer, College of Medicine, Department of pediatrics, University of Nigeria/University of Nigeria Teaching Hospital (UNTH), Ituku, Ozalla, Enugu, Nigeria

More Information

***Address for Correspondence:** Dr. Josephat M Chinawa, Associate Professor and Pediatric Cardiologist, College of Medicine, Department of pediatrics, University of Nigeria/University of Nigeria Teaching Hospital (UNTH), Ituku, Ozalla, Enugu, Nigeria, Email: Josephat.chinawa@unn.edu.ng

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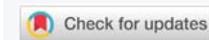
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Keywords: Teratology of fallot; Children; Surgical repair; Death; Sequel



Abstract

Background: Tetralogy of Fallot (TOF) is a very common cyanotic congenital heart disease presenting early at birth with various degrees of cyanosis. If left uncorrected surgically, can lead to death.

Objectives: This study is aimed at determining pattern and surgical outcome of children with teratology of Fallot in a budding health facility in India over a year period.

Result: A total of 51 children were diagnosed of TOF over the period, of which 66.7% were males with mean age of 48.14 ± 45.36 months.

The surgical outcome showed only 3.9% mortality. The death was among children >1 to 5 years. The mean number of days in intensive care unit (ICU) was 5.8 ± 11.2 days. 82.4% of the patients were off-pump post-operatively, compared to 17.6% with re-pump. Among those who had re-pump, 77.8% were males and among those without re-pump, 64.3% were likewise males ($\chi^2 = 0.6, p = 0.41$). About 92.2% (47/51) of patients had pulmonary regurgitation post-op, ranging from mild to moderate regurgitation. 51.1% of the regurgitations were mild while 25.5% and 23.4% were moderate and severe regurgitations respectively.

Post-operative VSD was detected in 51% (26/51) of the patients. The post-op right ventricular pressure (RVOT) was significantly lower than that of pre-op pressure, 10.8 ± 1.5 mmHg vs. 31.7 ± 4.5 mmHg (pair t test = 8.7, $p < 0.001$).

Conclusion: Timely surgical repair is crucial in alleviating several morbidity and mortality associated with teratology of fallot. Pulmonary regurgitation is a very common sequel after surgery and can result in death.

Introduction

Tetralogy of Fallot (TOF) is the most occurring cyanotic congenital heart disease (CHD), with prevalence of 34 per 100000 live births [1]. Children presenting with teratology of fallot have varying degrees of cyanosis depending on the level and severity of right ventricular outflow

tract (RVOT) and architecture of the pulmonary artery. death is related to right ventricular hypertrophy or dilation and mortality increases if intra-cardiac repair is not instituted early. The timing of surgery is therefore very important so as to avoid complications [2]. It is important to note that residual lesions are common sequel and could result in late morbidity and mortality [3-5]. The commonest sequel



is pulmonary regurgitation (PR); this was documented in majority of patients, especially in children with trans annular patch. Pulmonary regurgitation was seen initially as mild and innocuous, but this can cause right ventricular dilatation, arrhythmias and eventual dysfunction and even death [4-7].

This study is important because it is the first to be done in the hospital with the notion that early identification and appropriate intervention can significantly improve the quality of life of children with teratology of fallot. This study is aimed at determining the gender and age distribution of children with teratology of fallot, to ascertain the different levels and severity of pulmonary stenosis and right ventricular outlet obstruction (RVOT) to identify the different surgical route or technique of repair and to ascertain If there is any correlation between gender, body mass index, age, relationship between right ventricular to left ventricular volume ratio (RV/LV) ratio with surgical outcome .It is also aimed at determining post-operative complications seen among children with teratology of fallot.

Materials and methods

Study area

The study was conducted at the Pediatric outpatient and Pediatric intensive care unit of Fortis Hospital over a one-year period.

Study population

Diagnosis of teratology of fallot was made among subjects who are between the age of 6 months and 18 years. This was made with Echocardiography. Computerized axial tomography (CT) scan was also used in children with difficult anatomy, cardiac catheterization was used to ascertain pulmonary vascular resistance for patients with major aorto-pulmonary collaterals (MAPCAS) and to access operability.

Study design

This is a retrospective descriptive study among in patients attending Fortis Hospital and who had surgery for TOF repair January 2014 to December 2014. All patients underwent cardiopulmonary bypass. Children who had teratology of fallot and had surgical repair and who gave consent were included in the study while those who did not give consent were excluded.

Consent

This was obtained from the parents and/or caregivers in the course of admission to the ward.

Data analysis

Categorical variables such as gender, age categories, and type of VSD were summarized in form of percentages and proportions while continuous variables such as age, number of days in intensive care unit (ICU), right ventricular pressure

were summarized as means and standard deviations. Difference in means was tested by Student's *t* test and difference in proportion by Chi-square test. Relationship between continuous variables was analyzed using Pearson correlation coefficient. Significant difference was taken as $p < 0.05$.

Results

Table 1 shows the age and sex distribution of the subjects with TOF. A total of 51 children were diagnosed of TOF over the period, of which 66.7% were males. Their age ranged from 9 to 192 months with mean of 48.14 ± 45.36 months. Distribution of the age groups shows infants (≤ 12 months) made up 29.4% of the population.

The types of VSD diagnosed were 76.5% sub-aortic while 15.7% and 7.8% were pulmonary and sub-arterial respectively. A review of the levels of pulmonary stenosis among the patients revealed that 47.1% were valvular, 35.3% were supravalvular and 17.6% at the infundibulum.

Blood flow within the ventricles was bidirectional in 45.1% of the patients, 33.3% had left to right and 15.7% right to left flow. Direction of blood flow was not recorded in 5.9% of the patients.

The different surgical routes used in VSD repair were as follows: trans-atrial (74.5%), RVOT resection (19.6%) and intra-cardiac (5.9%) routes. Of the total number of patients, 88.2% had their infundibulum assessed intra-operatively, of which 33.3% had long and hypoplastic infundibulum, and 28.9%, 17.8, 15.5%, had short, hypoplastic; and short and narrow infundibulum respectively while roomy and long infundibulum were seen in 2.2% of the patients each. 19.6% of the patients had transannular patch.

The surgical outcome showed only 3.9% mortality. The death was among children >1 to 5 years. The total number of days on hospital admission ranged from 4-75 days with mean of 14 ± 13.3 days. The mean number of days in intensive care unit (ICU) was 5.8 ± 11.2 days. The gender or age of patients did not influence the number of days spent in ICU as shown in table 2. The mean number of days spent in ICU by males (6.2 ± 13.1) was comparable to that of females (4.8 ± 4.6), $p = 0.67$. However, infants, ≤ 12 months spent longer days in ICU compared to those 2-5 years and those ≥ 11 years.

Table 1: Age and gender distribution of children with TOF.

Sex	N	%
Male	34	66.7
Female	17	33.3
Total	51	100
Age (months)	N	%
≤ 12	15	29.4
13-60	22	43.1
61-120	10	19.6
121-192	4	7.8
Total	51	100

Table 2: Mean number of days in ICU in respect of gender and age.

Sex	N	Mean	p
Male	34	6.2 ± 13.1	0.67
Female	17	4.8 ± 4.6	
Age	N	Mean	p
≤12	15	11.5 ± 19.0	0.9
13-60	22	3.5 ± 3.6	
61-120	10	3.6 ± 4.6	
≥121	4	2.25 ± 1.3	

82.4% of the patients were off-pump post-operatively, compared to 17.6% with re-pump. Among those who had re-pump, 77.8% were males and among those without re-pump, 64.3% were likewise males ($\chi^2 = 0.6, p = 0.41$). The mean body mass index (BMI) of those with and without re-pump were comparable, 16.26 ± 2.2 vs. 14.07 ± 4.8 ($t = -1.33, p = 0.2$).

About 92.2% (47/51) of patients had pulmonary regurgitation post-op, ranging from mild to moderate regurgitation. 51.1% of the regurgitations were mild while 25.5% and 23.4% were moderate and severe regurgitations respectively.

Post-operative VSD was detected in 51% (26/51) of the patients. Further analysis showed 57.7% of the VSDs were tiny, 37.6% and 7.7% were small and moderate post-op VSD respectively. The mean Post-operative right ventricular pressure (RVOTP) was 23.67 mmHg. Mean RVOTP in males was comparable to that of females (23.97 ± 10.38 vs. $22.94 \pm 11.31, t = 0.32, p = 0.75$). Majority of the patients (86.3%) had normal follow-up right ventricular systolic and diastolic pressure, unlike 11.7% and 1.9% with mild to moderate dysfunction of right ventricular pressure. The post-op right ventricular pressure (RVOT) was significantly lower than that of pre-op pressure, 10.8 ± 1.5 mmHg vs. 31.7 ± 4.5 mmHg (pair t test = 8.7, $p < 0.001$). Also the post-op right ventricular volume was significantly lower than pre-op volume, 0.4 ± 0.08 vs. 0.7 ± 0.2 (paired test = 8.5, $p < 0.001$).

Tables 3 reveals the correlation between patients' body surface area and some cardiac parameters while table 4 demonstrates also the correlation between number of days in intensive care unit (ICU) and some cardiac parameters. The mean Z-score RPA was significantly higher in males compared with females, 0.3 ± 0.9 vs. -0.5 ± 1.2 ($t = 2.3, p = 0.02$). However,

Table 3: Correlation between body surface area and some cardiac parameters.

Parameter	Pearson coefficient	p value
RPA	-0.09	0.5
LPA	-0.2	0.2
RcVol	-0.1	0.4
RVOT	0.1	0.5

Table 4: Correlation between number of days in ICU and some cardiac parameters.

Parameter	Pearson coefficient	p value
RPAZ	0.2	0.2
LPAZ	0.1	0.4
RVOTZ	-0.03	0.8
PVZ	0.2	0.2

the mean Z-score LPA was comparable between the two groups, 0.4 ± 1.4 vs. -0.3 ± 1.3 ($t = 1.6, p = 0.1$). There was a negative correlation between RPA, LPA and body surface area, although the correlation was not significant.

Discussion

Intra-cardiac repair of Fallot's tetralogy is the standard mechanism of repair for majority of children with Teratology of fallot (TOF) [8]. We noted male preponderance in our study. This was also similar to studies by Mahmoud, et al. [9] and Chinawa, et al. in Nigeria, Imran, et al. [11] in in Brazil who all noted male preponderance. The male dominance can be explained by the fact that congenital heart defect from conotruncal origin usually occur more in males [12].

We noted the mean age of presentation for children with teratology of fallot in our study, as 48.14 ± 45.36 months. Our study is similar to that of Glen et al who noted a mean age of 50.9 ± 45.9 in months (4.2 ± 3.8 in years). This is also similar to that reported by Kennedy, et al. [13] in Malawi. Due to increase mortality and morbidity outcomes, the best age for elective repair of tetralogy of fallot is 3 to 11 months of age. It is important to note that right ventricular hypertrophy increases with age and this is a bad prognostic sign for surgical outcome.

Late presentation is the norm in our study. Though diagnosis can be made in-utero in developed countries, however, it is important to note that diagnosis of teratology of fallot (TOF) and other congenital heart diseases are normally made between 1-5 years of age in developing countries [14-17]. This late diagnosis is basically due to late presentation, poor health care facilities or lack of awareness [14-18].

The commonest route used for the repair in this study is trans-atrial (74.5%) and the least is trans pulmonary route. On assessment of infundibulum intraoperatively, majority of our cases had a long and hypoplastic infundibulum. This is contrary to a study were trans atrial- trans-pulmonary were used in majority of their cases [19]. Arch, et al. [19] also noted that majority of his series present with long and elongated infundibulum necessitating trans annular patch operation [19].

Sub-aortic ventricular-septal defect (VSD) is the commonest type of VSD seen in our study. This is in keeping with other studies. [20,21].

A review of the levels of pulmonary stenosis among the patients in our study showed that majority were valvar. This is at variance with the known fact that infundibular stenosis is the commonest level of obstruction. The sub-pulmonic, or infundibular, obstruction in TOF is characterized by anterior and cephalad deviation of the outlet, or infundibular, septum. This deviation of the outlet septum results in muscular sub-valvular narrowing [21]. It is important to note that due to



a very elongated and narrow infundibulum, 19.6% of our patients had trans-annular patch. Several studies have used the approach of this technique in their repair. For instance, correction with trans-annular patch was performed in 80%, as reported by Seddio, et al. [22]. In addition, the European Association for Cardio-Thoracic Surgery [23] report repair with trans-annular patch as the most frequently used technique (57% of their cases), while, Tirilomis, et al. [24] report its use in 27% of their study.

There is an increased risk of pulmonary regurgitation and long ICU stay that were noted in this study, especially in children with trans-annular patch. Pekka, et al. [25] also opined that trans-annular patch in tetralogy of Fallot surgery carries a higher risk of reoperation with attendant pulmonary regurgitation.

Pulmonary regurgitation was noted as an immediate complication in almost all our series. Indeed, one out of every ten patients presented with pulmonary regurgitation in our study.

It is important to stress that pulmonary regurgitation following repair of tetralogy of Fallot is a very common postoperative complication associated with right ventricular dilatation and eventual dysfunction. It also predicts late morbidity and mortality [26].

Our reoperation cases were very few due to the fact that right ventricular/left ventricular pressure seen in our cases were less than two-thirds systemic pressure. Fraser, et al. [27] noted that right ventricular/left ventricular pressure that is more than two-thirds systemic pressure with residual obstruction of the right ventricular outflow tract is a strong and potent indicator for reoperation.

Reoperation rates are also noted to be more frequent in cases of trans annular patch. Pekka, et al. [25] elicited the fact that they could not exactly distinguish whether the higher reoperation rate in their patients with Trans annular patch was as a result of the surgical technique or the severity of the primary condition. They rather opted for the fact that trans-annular patch should be considered as a consequence of a more severe disease [25,28].

We documented extubation failure in one of our subjects. The patient had a reoperation with closure of residual ventricular septal defect, trans-annular patch was done with catheterization to check pulmonary vascular resistance. The patient had a long hospital stay after spending over 90 days on admission. Our study however showed that extubation failure is low. Andrew, et al. [29] also noted that extubation failure in patients after complete repair for tetralogy of Fallot is usually low with diverse etiology. The cause of failed extubation in our study is severe right ventricular outflow obstruction, precedence of residual ventricular septal defect and trans annular patch operation. Extubation failure is associated with a longer hospital stay as seen in our patient.

Very few of our series presented with arrhythmias especially Supra ventricular tachycardia (SVT). It is pertinent to note that the occurrence of supraventricular tachyarrhythmias is one of the major cause of morbidity especially in the adult population [29]. This SVT has been known to be associated with an increased incidence of reoperation and heart failure and even death [30].

The cause of SVT is known to arise from re-entrant circuits localized around the atriotomy scar and are often multiple and unstable, making catheter ablation difficult to be achieved [31,32].

We noted episodes of seizure in few of our series. Prolonged cardio pulmonary bypass (CPB) time, hypoxia and older age at presentation could all explain this. It is important to point out that the occurrence of a seizure in the early postoperative period after repair of teratology of fallot is a marker for a central nervous system (CNS) injury. This has been associated with adverse neurodevelopmental sequelae [32].

Bi-direction Glenn was instituted in one of our series with teratology of fallot. This is due to severe right ventricular dysfunction, very small z score for pulmonary valve annulus and narrow and elongated infundibulum. We finally lost the patient after spending 19 days in the ICU. It is noted in some studies that bidirectional Glenn shunt (BGS) can be used to decongest the pulmonary ventricle that is encountering the outflow obstruction [33-37]. This procedure will help avoid the need for insertion of a conduit.

Coronaries crossing the RVOT was noticed in two of our series. They had a conduit and was discharged after spending few days' post-operation. Marco, et al. [38] noted a preference of Blalock Taussigs (BT) shunt to conduit in his series. This is basically due to the fact that his subjects are neonates. Coronaries crossing the RVOT can occur with different coronary branches [39,40].

Brizard, et al. [40] opined that coronary crossing the infundibulum in TOF can be corrected by the transatrial-transpulmonary approach frequently without many changes in technique with a good outcome. Kalra, et al. [41] used Brizard method and affirmed that this is a suitable option with no need for an extra-cardiac conduit.

Two of our patients had a Blalock-Taussig (BT) shunt for right ventricular dysfunction. The patient had small branch pulmonary arteries, prolong intubation time and a younger age. The Blalock-Taussig shunt is seen as a palliative low-risk management option in management of tetralogy of Fallot, but this is not without morbidities and mortality [42]. Children with significantly smaller distal right pulmonary arteries and younger age were seen as indication for a BT shunt, as seen in our study [42].

Only a very minute fraction (7.7%) of our patient had a



significant residual VSD post operation. Most of these patients had re pump, prolong stay in hospital and one had a BT shunt.

Significant Residual ventricular septal defect (VSDs) are usually not well tolerated in children with tetralogy of Fallot. This is because of post-operative volume overload [43]. Over 90% of our children had small insignificant VSDs and were all discharged with little or no postoperative complications. It is known that small, residual VSDs are common occurrence after surgical repair and are always clinically insignificant [43].

We noted mortality from surgery to be 3.9%. Majority arising from younger ages and those that have longer perfusion time. This result is very commendable when compared with study by Fraser which showed mortality of about 10%. His larger sample size might have been the cause of this higher mortality rate [44]. Azari, et al. [45] also noted a mortality rate of 12.2% in his study, which was higher in younger patients, those with lower weight and those with longer perfusion time during cardiac surgery.

Conclusion

Timely surgical repair is crucial in alleviating several morbidity and mortality associated with teratology of fallot. Pulmonary regurgitation is a very common sequel after surgery and can result in death.

Author contributions

AV, JMC, BT and GS conceived and designed this study while JMC and BT helped in echo diagnosis .BC did the data analysis/interpretation of results and critical revision of the article.

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