ORIGINAL ARTICLE

Vol. 10 No. 4 October-December 2025

doi: 10.35366/121355

Follow-up of pediatric patients with total correction of tetralogy of Fallot over a 10-year period

Seguimiento de pacientes pediátricos con corrección total de tetralogía de Fallot en un periodo de 10 años

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ABSTRACT

Introduction: tetralogy of Fallot is a common pathology in infancy. Surgical correction has excellent results although it still presents long term complications. **Material:** this retrospective study examined the outcomes of 70 patients with tetralogy of Fallot who underwent surgical correction. The patients' ages at surgery ranged from 3 to 13 years, with 67% undergoing correction before the age of five. Postoperative follow-up was conducted between 1 and 8 years after surgery. **Results:** the results showed that 25% of patients developed signs of septic shock within the first 36 hours postoperatively, and 22.9% experienced respiratory complications. Conversely, 20% of patients had an uncomplicated postoperative course and were discharged within 10 days. During follow-up, 63% of patients were asymptomatic, while 34% exhibited NYHA functional class II symptoms. Notably, 35.7% of patients had residual ventricular septal defects. Right ventricular dilatation was observed in 50% of cases, and pulmonary insufficiency was classified as severe in 23.3% of cases. **Conclusions:** the study concluded that surgical correction of TF yields satisfactory long-term outcomes, but regular follow-up is crucial to detect complications and facilitate timely interventions. The findings underscore the importance of ongoing monitoring and management to optimize patient outcomes and mitigate potential long-term sequelae.

RESUMEN

Introducción: *la tetralogía de Fallot es una patología común en la infancia. La corrección quirúrgica tiene excelentes resultados aunque todavía presenta complicaciones a largo plazo.* ***Material:*** *este estudio retrospectivo examinó los resultados de 70 pacientes con tetralogía de Fallot que se sometieron a corrección quirúrgica. Las edades de los pacientes en el momento de la cirugía oscilaron entre 3 y 13 años, y el 67% se sometió a corrección antes de los cinco años. El seguimiento postoperatorio se realizó entre 1 y 8 años después de la cirugía.* ***Resultados:*** *los resultados mostraron que el 25% de los pacientes desarrollaron signos de choque séptico dentro de las primeras 36 horas después de la operación y el 22.9% experimentó complicaciones respiratorias. Por el contrario, el 20% de los pacientes tuvo una evolución postoperatoria sin complicaciones y fueron dados de alta dentro de los 10 días. Durante el seguimiento, el 63% de los pacientes estaban asintomáticos, mientras que el 34% presentó síntomas de clase funcional II de la NYHA. Cabe destacar que el 35.7% de los pacientes tenían defectos septales ventriculares residuales. Se observó dilatación del ventrículo derecho en el 50% de los casos, y la insuficiencia pulmonar se clasificó como grave en el 23.3%.* ***Conclusiones:*** *el estudio concluyó que la corrección quirúrgica de la TF produce resultados satisfactorios a largo plazo, pero el seguimiento regular es crucial para detectar complicaciones*

How to cite: López-Taylor J, Ramírez-Cedillo D, Jiménez-Fernández C, Masini-Aguilera I, Medina-Andrade MÁ, Contreras-Godínez A, et al. Follow-up of pediatric patients with total correction of tetralogy of Fallot over a 10-year period. Cir Card Mex. 2025; 10 (4): 105-109. https://dx.doi.org/10.35366/121355

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Received: 07-01-2025. Accepted: 07-16-2025.

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y facilitar intervenciones oportunas. Los hallazgos subrayan la importancia del seguimiento y el manejo continuos para optimizar los resultados del paciente y mitigar las posibles secuelas a largo plazo.

Keywords: cardiac surgery, congenital heart disease, long-term outcomes, pulmonary insufficiency, surgical correction, tetralogy of Fallot.

Palabras clave: *cirugía cardíaca, cardiopatía congénita, resultados a largo plazo, insuficiencia pulmonar, corrección quirúrgica, tetralogía de Fallot.*

Abbreviations:
CI = confidence intervals
HR = hazard ratios
PI = pulmonary insufficiency
PS = pulmonary stenosis
RV = right ventricular
RVEF = right ventricular ejection fraction
TF = tetralogy of Fallot
VSD = ventricular septal defects

Tetralogy of Fallot (TF) is a prevalent congenital heart defect in infancy. Although the first successful repair was performed in the 1950s, with some authors reporting excellent long-term outcomes,[1,2] the long-term survival rate in patients who underwent repair remains lower than that of the age- and gender-matched general population. Furthermore, late complications such as arrhythmias, sudden death, decreased functional capacity, and re-operations, primarily attributed to the right ventricular (RV) outflow tract reconstruction technique and the approach to addressing the interventricular defect, continue to be a subject of debate. This study investigated the follow-up outcomes after total TF repair, focusing on echocardiographic alterations and demographic and surgical variables in these patients.

MATERIAL

A retrospective, descriptive study was conducted, reviewing the records of patients diagnosed with TF over a 10-year period. Patients with pulmonary atresia and those referred to the adult cardiology service were excluded. Statistical analysis was performed using SPSS version 15. Quantitative variables were summarized using measures of central tendency (mean or median) and dispersion (standard deviation or interquartile range), depending on normality assumptions assessed via the Shapiro-Wilk test. Categorical variables were summarized as absolute frequencies and percentages. Event-free survival probabilities were estimated using the Kaplan-Meier method and compared using the log-rank test. A bivariate Cox regression model was fitted for all variables, and hazard ratios (HR) with 95% confidence intervals (CI) were reported. Statistical significance was set at $p < 0.05$.

RESULTS

Demographic data

The age at surgical correction ranged from 3 to 13 years. Patients were divided into two groups based on age at surgery: group 1 (under five years, 67%, n = 47) and group 2 (over five years, 32.8%, n = 23). Of the 70 patients, 33 (47.1%) were male. Postoperative follow-up time was categorized into three groups: under two years (54.2%), 3-5 years (15.7%), and over five years (30%) *(Table 1)*.

Surgical treatment

In this section, patients were divided into two groups

1. Palliative treatment

Fourteen patients (20%) underwent palliative surgery (systemic-pulmonary shunt) between 4 and 10 days of life (mean six days). The indications for palliative surgery were: (1) mild to moderate hypoxemia secondary to severe pulmonary branch hypoplasia (Z value < -2.5) in eight cases, and (2) pulmonary annulus hypoplasia (Z value -2) in six cases. Corrective surgery was performed in all patients with a previous systemic-pulmonary shunt, with a mean interval of 18 months (range 12-21 months) after palliative surgery.

Table 1: Demographic data of patients with follow-up for tetralogy of Fallot correction (N = 70).

Variable	n (%)
Age at time of correction (mean, six years)	3-13
Age group at time of correction (years)	
< 5	47 (67.0)
> 5	23 (33.0)
Gender	
Male	33 (47.0)
Female	37 (53.0)
Group based on postoperative time (years)	
< 2	38 (54.2)
3-5	11 (15.7)
> 5	21 (30.0)

Table 2: Surgical data of patients operated of tetralogy of Fallot (N = 70).

Variable	n (%)
Palliative surgery (PSF)	14 (20)
PSF indication	
Hypoplastic branches	8
Pulmonary annulus hypoplasia	6
Time from palliative to corrective surgery (mean, 18 months)	12 to 21
Coronary alterations	10 (7.0)
Patent ductus arteriosus	6 (8.5)

2. Total correction

All surgical cases were performed via median sternotomy. Conventional extracorporeal circulation with moderate hypothermia (25-30 °C) and anterograde cardioplegia were used in all cases, with additional myocardial protection measures including saline at 4 °C in the pericardial sac every 15-20 minutes. A longitudinal ventriculotomy was performed, extending towards the annulus, trunk, or branches when hypoplasia was present. The ventricular septal defect was closed with a patch and interrupted sutures on the posterior, inferior, and superior edges, and continuous sutures on the remaining areas. A patch was required in the right ventricular outflow tract in all cases. Coronary circulation abnormalities were found in 10 cases, including anomalous origin of the left anterior descending artery and left main coronary artery from the right coronary artery. Associated patent ductus arteriosus was present in six patients and was ligated prior to cardiopulmonary bypass. Systemic-pulmonary shunts were also addressed prior to bypass *(Table 2)*. Notably, surgical technique details were unavailable for five cases due to the procedure being performed over five years ago.

Immediate postoperative

Of the 70 patients who underwent surgery, 25% developed signs of septic shock within the first 36 hours postoperatively. Additionally, 16 patients (22.9%) experienced other complications, all of which were respiratory in nature, with pneumonia being the primary diagnosis, requiring mechanical ventilation for a mean duration of six days. In contrast, 20% of patients had an uncomplicated postoperative course and were discharged within 10 days. Five patients (7.1%) developed complete atrioventricular block, with two requiring permanent pacemaker implantation. Chylothorax occurred in three patients (4.3%), two of whom responded to medical treatment, while one required surgical intervention. Re-operation was necessary in two cases (2.8%): one for chylothorax and diaphragmatic paralysis 14 days postoperatively, and another for permanent pacemaker placement 10 days postoperatively. Notably, no patients experienced postoperative hemorrhage or pericardial effusion requiring reintervention or evacuation *(Table 3)*.

Follow- up

Follow-up was conducted between 1 and 8 years after total correction of TF. Clinically, 63% of patients were asymptomatic, while 34% had NYHA functional class II symptoms, all of whom were in the postoperative group more than five years after surgery. Regarding medical treatment, 66% of patients did not require any medication, while 34% were receiving diuretic therapy. All patients were in sinus rhythm, except for one who had a permanent pacemaker and was being followed up by the electrophysiology department. Two patients presented with postoperative neurodevelopmental changes: one had gait disturbances and was undergoing rehabilitation with partial improvement, while the other had speech disturbances and neurodevelopmental delay *(Table 4)*.

Residual shunts

The 35.7% had residual ventricular septal defects (VSD); of this group 54% were in the intervention group under five years of age (p = 0.02).

Right ventricular assessment

Right ventricular (RV) dilatation was observed in 50% of cases, with no significant predominance in age at surgery (p = 0.025). However, there was a significant association between

Table 3: Immediate postoperative data.

Variable	n (%)
Septic shock in the first 36 hours post-surgery	18 (25)
Pneumonia	16 (23)
Days of mechanical ventilation (mean, 6 days)	4 to 12
Days of stay in the pediatric intensive care unit (mean, 8 days)	5 to 18
Complications	
Complete AV block	2 (3.0)
Chylothorax	3 (4.2)
Re-operations	
Pacemaker placement	1 (1.42)
Chylothorax	1 (1.42)

AV = atrioventricular

Table 4: Clinical data and follow-up patients with correction of tetralogy of Fallot.

Variable	n (%)
Postoperative follow up in years (mean, 4 years)	1 to 10
Functional class	
I	46 (63)
II	24 (34)
Management	
No management	46 (63)
Anti-congestive medical therapy	24 (34)
Postoperative neurodevelopmental alteration	
Gait and speech	2 (3)

postoperative time and RV dilatation, with 53% of patients over five years postoperatively and 26% of patients 3-5 years postoperatively affected. The mean transpulmonary gradient was 24 mmHg (range, 9-51 mmHg). Pulmonary stenosis (PS) was mild in 93.3% and moderate in 6.6% of cases. Pulmonary insufficiency (PI) was classified as mild in 40%, moderate in 36%, and severe in 23.3% of cases. There was no significant association between age at surgery and PI ($p = 0.15$), but a significant association was found between postoperative time and PI, with 66% of patients over five years postoperatively affected, compared to 20% in the 3- to 5-year postoperative group and none in the group under two years postoperatively ($p = 0.000$).

DISCUSSION

TF is the most prevalent congenital heart defect in infancy, accounting for approximately 5-6% of all congenital heart diseases.[1] Our findings are consistent with the literature, which reports no significant gender predominance, although a slight female preponderance of 53% was observed in our study. The clinical presentation is variable, with symptoms such as progressive cyanosis and decreased physical capacity directly related to right ventricular outflow tract obstruction.[2] TF is characterized by a spectrum of abnormalities, typically classified into two groups based on anatomy: favorable or poor, with a focus on the Z score of the pulmonary branches and annulus. This classification informs management strategies, particularly in the neonatal stage, where poor anatomy may necessitate a staged approach, including initial systemic-pulmonary shunt followed by complete correction. In our study, 20% of cases required systemic-pulmonary shunt, primarily due to pulmonary branch hypoplasia.

The optimal age for total correction of TF remains a topic of debate. Two prevailing schools of thought exist: one advocating for an age-based approach and the other for an anatomy- or physiology-based strategy. While early surgical mortality after TF repair should be rare, no single surgical strategy has proven superior in mitigating the need for late intervention.[3] Since the 1990s, surgical intervention in the first years of life has been preferred, with multiple studies supporting this approach. Barrat-Boyes[4] and Starr et al.[5] demonstrated in 1973 that TF could be corrected with extracorporeal circulation in early life with low morbidity and mortality. More recently, Arsdell, et al.[6] reported that the ideal time for correction is between 3 and 11 months. A systematic review by Martins, et al.[2] concluded that definitive correction in the neonatal stage is a viable option, with an ideal age of 3-6 months for children with mild to moderate symptoms and immediate surgery for those with severe symptoms. However, Castañeda, et al.[7] emphasize the importance of considering preoperative factors, such as branch size and coronary pattern, in determining the optimal surgical approach.

The age range for total repair in our series was 3-13 years, with older age at repair associated with increased risk of postoperative heart failure.[7] Delayed repair can lead to worse long-term prognosis due to prolonged cyanosis, resulting in ventricular fibrosis. Furthermore, delayed repair may necessitate more extensive muscle resection, potentially causing further RV damage. Given the relatively late age of repair in our series, continued follow-up into adulthood is crucial. The number of postoperative patients with TF surviving to adulthood is increasing, with over 90% reaching their third decade.[8] However, life expectancy remains lower compared to the general population.[9]

Surgical repair of TF has a complex history, marked by both successes and limitations. While surgery significantly improves prognosis, it is not curative, leaving residual anatomical and functional abnormalities such as PS, PI, and RV outflow tract obstruction.[1] These lesions require ongoing cardiological follow-up and may necessitate medical or interventional management. In our series, 25 cases had residual VSD, although none required re-operation.

Pulmonary valve competence is a critical factor in long-term outcomes, with PI being a common complication requiring re-operation. In our series, 23% of cases presented with severe PI, with a significant prevalence observed in patients who underwent surgery more than five years ago. Monitoring PI is essential, as it can contribute to RV dysfunction, exercise intolerance, and life-threatening arrhythmias. While magnetic resonance imaging is the gold standard for assessing these complications, transthoracic echocardiography is a more accessible and non-invasive alternative for monitoring patients.

The optimal surgical technique for TF repair remains a topic of debate, with various approaches aimed at preserving pulmonary valve function. In our series, mild PS was observed in 93% of patients, which may be protective and delay the

need for pulmonary valve replacement.[10] Further research is needed to determine the most effective surgical strategies and to improve long-term outcomes for patients with TF.

Pulmonary insufficiency

Severe PI is a common cause of progressive RV dilatation and dysfunction in patients with TF repair, and is associated with decreased functional class, arrhythmias, and sudden death. Progressive RV dilatation may be a primary sign of RV dysfunction and an indication for pulmonary valve replacement.[11] PI is a frequent complication after TF surgery, and its negative consequences on RV function have been well-established. The use of transannular patch techniques, as seen in our series, may contribute to the development of PI.

In our study, the frequency of severe PI was 23.3%, primarily affecting patients who underwent surgical correction more than five years ago. Notably, the degree of PI was not directly associated with RV function as assessed by echocardiogram. Only two patients had decreased RV ejection fraction (RVEF), which is consistent with some reports in the literature, such as the study by Frigiola et al.,[12] where the severity of PI was independent of RVEF. However, this finding contradicts other studies, like the one by Kjaergaard et al.,[13] which reported lower RVEF in patients with severe PI compared to those with moderate PI. Further research is needed to clarify the relationship between PI and RV function in patients with TF repair.

CONCLUSIONS

Surgical treatment of TF is a medical success story, yet it remains a topic of debate. Various management strategies have been proposed, including age-based and anatomy-based approaches. Over the years, surgical management of TF has evolved significantly, resulting in excellent early outcomes with near 100% survival for patients without genetic syndromes. The primary goal of current surgical therapy is to minimize late RV dysfunction by reducing the extent of surgical injury during initial repair. As surgical techniques continue to advance, outcomes are expected to improve.

Although long-term outcomes after TF repair are generally good, postoperative sequelae are common and can persist over time. Regular follow-up with adequate monitoring is crucial to avoid missing the optimal time for reintervention. MRI is the gold standard for assessing RV function, but echocardiography remains an essential tool for evaluation. A multidisciplinary approach is necessary to ensure optimal care and management of patients with TF, both in the short- and long-term.

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Funding: none.

Disclosure: the authors have no conflicts of interest to disclose.