Health Sciences & Disease
The Journal of Medicine and Biomedical Sciences

Original Research

Repair of Tetralogy of Fallot in Cameroonian Children: Report of 105 cases

La Réparation de la Tétralogie de Fallot chez les Enfants Camerounais : à Propos de 105 Cas

[1,4,5]Mve Mvondo Charles, [4,5]Ngo Yon Laurence Carole, [4]Kobe Fokalbo Zephanie, [4,5]Tsague Kengni Hermann Nestor, [3]Banga Nkomo Douglas, [4]Lowe Kuissu Sepolin, [5]Metogo Bengono Junette, [2]Ndobo Nkoe Valerie, [2]Ela Bella Amos, [1]Tantchou Tchoumi Cabral, [4]Yepmo Yannick, [5]William Djomo Ngatchou and [5]Ngowe Ngowe Marcelin

Affiliations
[1]St Elizabeth Catholic General Hospital Shisong, Cardiac Centre Shisong, Kumbo, Cameroon
[2]Faculty of Medicine and Biomedical Sciences, Yaoundé, Cameroon
[3]Faculty of Medicine and Pharmaceutical Sciences, Université de Dschang, Dschang, Cameroon
[4]Department of Cardiothoracic and Vascular Surgery, Yaoundé General Hospital, Yaoundé, Cameroon
[5]Department of Surgery, Faculty of Medicine and Pharmaceutical Sciences, Douala, Cameroon

Corresponding author: Charles Mve Mvondo, MD
mmvondocarlo@yahoo.fr; +237-655633474

Key words: Tetralogy of Fallot; cardiac surgery; Cameroon

ABSTRACT

Background. Delayed diagnosis of congenital heart diseases is common in children living in low- to middle-income countries due to limited access to cardiovascular care. Late diagnosis of tetralogy of Fallot (ToF) often leads to the presentation of complex clinical features, significantly affecting surgical outcomes. This paper aims to review the operative mortality following surgical treatment of African children with ToF over a 12-year period. **Materials and Methods.** Between November 2009 and December 2021, a total of 105 children diagnosed with ToF underwent surgical intervention at our institution. Clinical, echocardiographic, and surgical data were retrospectively reviewed. **Results.** The median age at surgery was 36 months (IQR: 20–72 months). The sex ratio was 1.39 (male to female). The primary symptoms reported were fatigue (16.2%, n = 17/105), cyanosis (7.6%, n = 8/105), or a combination of both (76.2%, n = 80/105). The vast majority (95.2%, n = 100/105) underwent a total ToF correction. The transatrial-approach for ventricular defect (VSD) closure was performed in 92.3% of cases (n = 97/105). Transanular and/or infundibular patches were used in 89 (84.8%) patients. Postoperative complications included respiratory distress in 5 (4.8%) patients and reexploration for bleeding in 6 (5.75%) patients. The operative mortality rate was 5.7% (n = 6/105). **Conclusion**. Despite the increased age and more complex pathophysiological features, surgical treatment of children with ToF in our setting is associated with acceptable early outcomes. Timely screening and surgical correction before the onset of associated complications are advocated as crucial measures to mitigate surgical risks.

RÉSUMÉ

Introduction. Le retard de diagnostic des cardiopathies congénitales est fréquent chez les enfants vivant dans les pays à revenus faibles ou moyens, en raison de l'accès limité aux soins cardiovasculaires. Le diagnostic tardif de la tétralogie de Fallot (TdF) conduit souvent à l'apparition de caractéristiques cliniques complexes, ce qui affecte de manière significative les résultats chirurgicaux. Cet article a pour but d'examiner la mortalité opératoire suite au traitement chirurgical des enfants africains atteints de ToF sur une période de 12 ans. **Matériels et méthodes.** Entre novembre 2009 et décembre 2021, un total de 105 enfants diagnostiqués avec une TdF ont subi une intervention chirurgicale dans notre institution. Les données cliniques, échocardiographiques et chirurgicales ont été revues rétrospectivement. **Résultats.** L'âge médian au moment de l'intervention chirurgicale était de 36 mois (IQR : 20-72). Le sex-ratio était de 1,39 (hommes/femmes). Les principaux symptômes signalés étaient la fatigue (16,2 %, n = 17/105), la cyanose (7,6 %, n = 8/105) ou une combinaison des deux (76,2 %, n = 80/105). La grande majorité (95,2 %, n = 100/105) a subi une correction totale de la ToF. L'approche transatriale pour la fermeture du défaut ventriculaire a été réalisée dans 92,3 % des cas (n = 97/105). Des patchs transannulaires et/ou infundibulaires ont été utilisés chez 89 patients (84,8 %). Les complications postopératoires comprenaient une détresse respiratoire chez 5 (4,8 %) patients et une réexploration pour hémorragie chez 6 (5,75 %) patients. Le taux de mortalité opératoire était de 5,7 % (n = 6/105). **Conclusion.** Malgré l'âge élevé et les caractéristiques physiopathologiques plus complexes, le traitement chirurgical des enfants atteints de TdF dans notre contexte est associé à des résultats précoces acceptables. Un dépistage opportun et une correction chirurgicale avant l'apparition des complications associées sont préconisés comme des mesures cruciales pour réduire les risques chirurgicaux.

INTRODUCTION

Tetralogy of Fallot (ToF), a cyanotic congenital heart disease (CHD), is typically characterized by four morphological features: 1) a ventricular defect, 2) overriding aorta, 3) right ventricular outflow tract obstruction, and 4) right ventricular hypertrophy [1]. Although the defect was first described by Steensen in 1671, the term ToF was only established in 1888 by Etienne Fallot [2]. As the most prevalent cyanotic CHD, its incidence is approximately 3 per 10,000 live births, and it accounts for about 5% to 7% of all CHD cases [3, 4]. A better understanding of ToF physiopathology patterns and advances in diagnostic and surgical management have significantly improved the life expectancy of operated patients over the past five decades. Presently, complete surgical correction within the first year of life has emerged as the preferred therapeutic approach [5], given the potential for increased morbidity associated with both neonatal [6] and delay repair.

In developing regions such as sub-Saharan Africa, delayed diagnosis of CHD is remains prevalent, primarily due to limited access to cardiovascular care [7]. Consequently, the majority of ToF patients present with advanced manifestations, including severe right ventricular hypertrophy, polycythemia, and left ventricular dysfunction, which render surgical repair more challenging. However, few data exist on the outcomes of delayed repair of ToF in this region [8, 9], largely attributable to the insufficient availability of pediatric cardiosurgical programs.

This paper reviews the early surgical outcomes following surgical treatment of ToF over a 12-year period.

MATERIALS AND METHODS

Between November 2009 and December 2021, a total of 105 children with ToF underwent surgical intervention at the Division of Cardiac Surgery within the Cardiac Centre of Shisong. Their clinical files and operative records were retrospectively reviewed.

Indications for implementing the palliative technique, specifically the Blalock–Taussig Shunt, included an age below 6 months, a weight below 6 kg and the presence of insufficiently developed pulmonary branches. Early postoperative events (mortality and morbidities) were defined as occurrences within 30 days following the surgery. The institutional review board approved the study, and the requirement for consent was waived given the retrospective nature of the investigation.

Surgical Technique

All patients underwent preoperative transthoracic echocardiography, which included an accurate review of morphological features such as the location of the defect and the degree of right ventricular tract obstruction, measurements of the diameters of the pulmonary trunk and branches, and examination of the anatomy of the coronary arteries. Figure 1 illustrates a preoperative echocardiogram of a child with ToF.

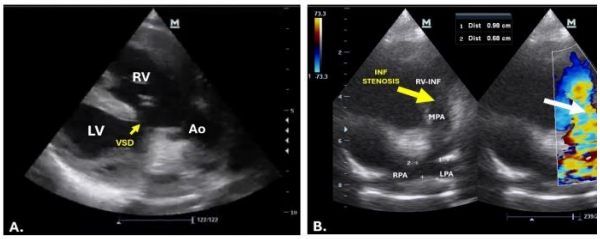

Figure.1. Transthoracic echocardiogram of a 2-year old child with ToF : A) illustration of the overriding aorta and the ventricular defect (VSD) ; B) infundibular stenosis (yellow arrow) and flow acceleration from the stenotic RVOT (white arrow).

A full median sternotomy was performed in all patients. Cardiopulmonary bypass (CPB) was established through cannulation of the ascending aorta and both caval veins in patients undergoing total correction. The mode of administration and the type of cardioplegic solution, either crystalloid or hematic, were defined by the senior surgeon. The ventricular septal defect (VSD) was approached mainly through a standard right atriotomy and closed with a pericardial heterologous patch. The size of the pulmonary tract (annulus, main trunk, and branches) was measured intra-operatively using various Hegar dilatators to assess the need for a transannular patch. Right ventricular outflow obstruction was relieved by muscular band resection followed by pericardial patch enlargement of the infundibular tract and/or transannular patch enlargement and extracardiac valved conduit. Closure of the patent ductus arteriosus (PDA) and atrial septal defect (ASD) was performed if present.

The modified B-T shunt, interposing an armed expanded polytetrafluoroethylene (PTFE) graft between the right subclavian artery and the right pulmonary artery, was performed through a median sternotomy without CPB. The pulmonary artery was dissected as for the classic B-T shunt [9]. A side-biting clamp was placed at the origin of the dissected subclavian artery to facilitate end-to-side anastomosis between the subclavian artery and the right pulmonary branch. The heparin dose was given intravenously before the prosthetic conduit was anastomosed.

Statistical Analysis

Statistical analysis was performed with R software version 4.1.1 (10). We carried out a descriptive statistical analysis in which the qualitative variables were expressed in terms of numbers and proportions in brackets, and the quantitative variables, being non-Gaussian, were expressed in terms of median (interquartile range, IQR).

RESULTS

There was male predominance, with a sex ratio (male to female) of 1.39. The median age at surgery was 36 months (IQR: 20–72). The median weight at surgery was 14 kg (IQR: 10–17 kg). Table 1 displays the preoperative characteristics of the patients.

Table 1: Patient's characteristics: preoperative and postoperative data

Variables	Value
Age (years), median (IQR)	3 (1.66 - 6)
Sex ratio (M/F)	1.39
Body surface area (m^2), median (IQR)	0.60 (0.46-0.70)
Down's syndrome	12 (11.4)
Haematocrit	
ToF, n (%)	96 (91.4)
ToF + PDA, n (%)	5 (4.8)
ToF + ASD, n (%)	4 (3.8)
Symptoms	
Cyanosis, n (%)	8 (7.6)
Dyspnea, n (%)	17 (16.2)
Both n (%)	80 (76.2)
Operative and postoperative data	
Total correction n (%)	100 (95.2)
Palliative procedure (BT Shunt), n (%)	5 (4.8)
Cardiopulmonary bypass time (min) median (IQR)	110 (86-142)
Cross clamping time (min), median (IQR)	63 (53.5-87.7)
Mechanical ventilation (hours), median (IQR)	11 (6-19)
Hospital length of stay (days), median (IQR)	7 (6-7)
Hospital death (30-d mortality), n (%)	4 (3.8)

IQR, interquartile range

Associated lesions were PDA and ASD in 4.8% (5/105) and 3.8% (4/105) of the patients, respectively. Genetic syndromes were found in 15% of the cases. Both cyanosis and dyspnea were reported in 76.2% (80/105) of the patients; only 7.6% (8/105) and 16.2% (17/105) of the children had isolated cyanosis or dyspnea, respectively. Eleven (10.5%) patients presented with severe digital hippocratism, as shown in Figure 2.

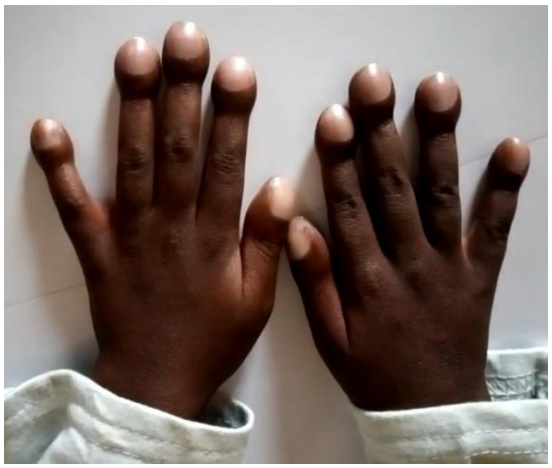

Figure 2: Severe digital hippocratism in a 5 year old boy with tetralogy of Fallot.

One hundred patients (95.2%) underwent a total correction, whereas five (4.8%) had palliative BT shunt. The median CPB and aortic cross clamp durations were 110 (IQR 86-142) and 63 minutes (IQR 53,5-87,75), respectively. A transannular pericardial patch and/or infundibular patch (Figure 3) were performed in 84,8% (n=89/105) patients and only 10.5% (n=11/105) had right ventricular outflow tract (RVOT) conduit.

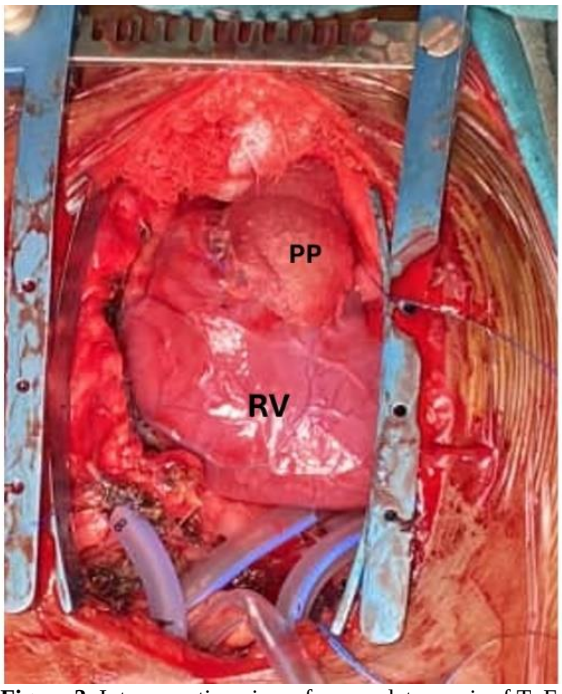

Figure 3: Intraoperative view of a complete repair of ToF with a transannular patch (pp).

The median intubation time was 11 hours (IQR 6-19). The most common postoperative complications were re-exploration for bleeding in 5.75% (6/105) patients, respiratory distress in 4.7% (5/105), right heart failure in 4,8% (5/105). Patient's characteristics, surgical data, and postoperative complications are summarized in Tables 1 and 2.

Table 2: Postoperative complications

Complications	n (%)
Re-exploration for bleeding	6 (5.7)
Respiratory distress	5 (4.8)
Ventricular dysfunction (left or/and right)	5 (4.8)
Postoperative arrythmias	15 (14.2)
Renal failure	1 (0.9)
Low cardiac output syndrome	4 (3.8)

The median hospitalization after surgery was 7 days (IQR: 6–7), and the overall 30-day operative mortality was 5.7% (6/105). The causes of deaths were low cardiac output syndrome, renal failure, and respiratory distress.

DISCUSSION

The current study reports our experience with 105 sub-Saharan children who underwent surgical repair of ToF over a 12-year period. Patients in our series were referred for surgery at a later stage and had a higher median age (3 years) than those in western series [10, 11]. Our findings were comparable with other developing country studies where ages ranged between 4.4 and 7.4 years in children who had undergone ToF repair [8, 12]. Indeed, a late diagnosis of CHD remains a major concern in our area due

mainly to the limited availability of pediatric cardiovascular specialists and cardiosurgical units, in addition to some reluctance to surgery due to sociocultural behaviors. Despite the growing number of cardiologists in the country, very few have developed interest in CHD over the last few years. Moreover, the establishment of a coordinated multidisciplinary approach involving gynecologists, neonatologists, pediatricians, and cardiologists to pioneer protocols for prenatal and perinatal diagnosis of CHD remains an unmet goal.

When compared with younger patients, patients with late diagnosis of ToF are more likely to have severe chronic hypoxia, severe right ventricular hypertrophy, decreased LV contractility, and polycythemia that are associated with adverse outcomes. While postoperative RV restrictive physiology (partly related to the severity of RV hypertrophy) has been associated with increased need and duration of inotrope support, as well as increased susceptibility to low cardiac output syndrome and extended hospital stays [13–15]. Conversely, severe polycythemia and severe hypoxia pose an elevated risk of postoperative blood loss and short-term mortality [16].

The operative mortality was 5.7%, representing a notable improvement compared to our previous experience where the operative mortality stood at 9% This suggests a progressive improvement in clinical outcomes over the past years [9]. It is worth noting that, during earlier periods, surgeries were carried out by foreign experts within relatively short-duration surgical missions (<10 days). Additionally, factors such as the poorer clinical condition of patients (e.g., malnutrition, severe chronic hypoxia, and polycythemia) and the lack of multidisciplinary support services (including neonatology, pediatrics, and imaging services) have potentially impacted our earlier outcomes. Other sub-Saharan African groups have reported an operative mortality rate ranging between 6.4% and 15.2% in locally repaired ToF [8,17,18]. Indeed, better outcomes in late repair of ToF were reported when SSA patients were transferred to experienced western institutes. In a cohort of 125 African children with a late diagnosis of ToF (mean age 4.4 years) transferred in a Swiss hospital through a humanitarian program, Mottier et al. reported no mortality following complete surgical correction [19]. Similar outcomes were described by Schaffner and colleagues in a group of 165 children (0% mortality rate) from developing countries who underwent late repair of ToF in Switzerland [20]. To some extent, clinical features of late-diagnosed ToF seem to not influence the operative outcomes, which are more related to the therapeutic environment. This was emphasized by Benbrick et al., who did not find any significant differences in terms of operative mortality between late repair of ToF in children from developing countries and their younger Swiss counterpart who had timely repair, despite more risk factors (growth retardation, low ventricular ejection fraction, polycythemia, etc.) reported in African children [21].

The transannular patch (TAP) emerged as the most frequently employed technique (58.3%) in the current series. This prevalence was higher than in reports from similar studies on late ToF repair reporting TAP in 16% to 38.2% of patients, respectively [19,20]. Although TAP is potentially associated with an increased risk of early and late morbidity as compared to valve sparing, it remains an acceptable compromise in hypoplastic pulmonary artery or in the presence of valvular lesions. Indeed, there is a significant variation in TAP (0%–100%) among studies as described by Romeo et al. in a meta-analysis with a nearly 50% rate of TAP in 21,427 patients who underwent ToF repair [22]. Although various modifications of the TAP techniques, such as the monocusp insertion, seem to provide a significant decrease in pulmonary regurgitation with consequent reduction in the intensive care stay [23] and reoperation rates, conflicting findings reporting no advantages of such technique in the immediate postoperative period have been described by several groups [24–26].

Study Limitations

The limitations of the present study are inherent to its retrospective design. Moreover, the lack of comparative outcomes between surgical sub-groups (TAP vs valve sparing or monocuspid patch) could not permit further analysis of factors that impact the early outcomes.

CONCLUSION

Despite the late diagnosis and more complex pathophysiological features, the postsurgical outcomes of children with ToF in our context are associated with acceptable early outcomes. Early diagnosis and surgical correction before the onset of right ventricular hypertrophy or other complications are warranted in order to reduce surgical risk.

REFERENCES:

1. Bailliard F, Anderson RH. Tetralogy of Fallot. Orphanet J Rare Dis. 2009 Jan 13;4:2.
2. Neill CA, Clark EB: Tetralogy of Fallot. The first 300 years. Tex Heart Inst J. 1994;21(4):272–9.
3. van der Linde D, Konings EE, Slager MA, et al.: Birth prevalence of congenital heart disease worldwide: A systematic review and meta-analysis. J Am Coll Cardiol. 2011;58(21):2241–7.
4. Postoev V. A., Talykova L. V., Vaktskjold A. (2014). Epidemiology of cardiovascular malformations among newborns in monchegorsk (North-West Russia): A register-based study. J. Public Health Res. ;3(2):270.
5. Martins IF, Doles IC, Bravo-Valenzuela NJM, Santos AORD, Varella MSP. When is the Best Time for Corrective Surgery in Patients with Tetralogy of Fallot between 0 and 12 Months of Age? Braz J Cardiovasc Surg. 2018 Sep-Oct;33(5):505-510.
6. van den Bosch E, Bogers AJJC, Roos-Hesselink JW, van Dijk APJ, van Wijngaarden MHEJ, Boersma E, Nijveld A, Luijten LWG, Tanke R, Koopman LP, Helbing WA. Long-term follow-up after transatrial-transpulmonary repair of tetralogy of Fallot: influence of timing on outcome. Eur J Cardiothorac Surg. 2020 Apr 1;57(4):635-643. doi: 10.1093/ejcts/ezz331. PMID: 31872208; PMCID: PMC7078865.
7. Yankah C, Fynn-Thompson F, Antunes M, Edwin F, Yuko-Jowi C, Mendis S, Thameur H, Urban A, Bolman R 3rd. Cardiac surgery capacity in sub-saharan Africa: quo vadis? Thorac Cardiovasc Surg. 2014 Aug;62(5):393-401.

8. Bamigboye-Taiwo OT, Adeyefa B, Onakpoya UU, Ojo OO, Eyekpegha JO, Oguns A, Okeniyi JA. Tetralogy of Fallot in the nascent open-heart surgical era in a tertiary hospital in south-west Nigeria: lessons learnt. Cardiovasc J Afr. 2022 May-Jun 23;33(3):122-126.
9. Tchoumi JC, Ambassa JC, Giamberti A, Cirri S, Frogiola A, Butera G. Late surgical treatment of tetralogy of Fallot. Cardiovasc J Afr. 2011 Jul-Aug;22(4):179-81.
10. Smith CA, McCracken C, Thomas AS, Spector LG, St Louis JD, Oster ME, Moller JH, Kochilas L. Long-term Outcomes of Tetralogy of Fallot: A Study From the Pediatric Cardiac Care Consortium. JAMA Cardiol. 2019 Jan 1;4(1):34-41. doi: 10.1001/jamacardio.2018.4255. PMID: 30566184; PMCID: PMC6439686.
11. Athanasiadis DI, Mylonas KS, Kasparian K, Ziogas IA, Vlachopoulou D, Sfyridis PG, Schizas D, Spartalis E, Nikiteas N, Hemmati P, Kalangos A, Avgerinos DV. Surgical Outcomes in Syndromic Tetralogy of Fallot: A Systematic Review and Evidence Quality Assessment. Pediatr Cardiol. 2019 Aug;40(6):1105-1112. doi: 10.1007/s00246-019-02133-z. Epub 2019 Jun 18. PMID: 31214731.
12. Souaga KA, Bonny R, Katche EK, KiriouaKamenan AY, Amani AK, Degré JC, Niava RG, Kouamé J, Yapo P, Kendja FK. Transatrial-transpulmonary correction of tetralogy of Fallot: experience of a developing country. Kardiochir Torakochirurgia Pol. 2022 Sep;19(3):130-134.
13. Cullen S, Shore D, Redington A. Characterization of right ventricular diastolic performance after complete repair of tetralogy of Fallot. Restrictive physiology predicts slow postoperative recovery. Circulation. 1995 Mar 15;91(6):1782-9.
14. Rathore KS, Gupta N, Kapoor A, Modi N, Singh PK, Tewari P, Sinha N. Assessment of right ventricular diastolic function: does it predict post-operative course in tetralogy of Fallot. Indian Heart J. 2004 May-Jun;56(3):220-4.
15. Van den Eynde J, Derdeyn E, Schuermans A, Shivaram P, Budts W, Danford DA, Kutty S. End-Diastolic Forward Flow and Restrictive Physiology in Repaired Tetralogy of Fallot: A Systematic Review and Meta-Analysis. J Am Heart Assoc. 2022 Apr 5;11(7):e024036.
16. Saygi M, Ergul Y, Tola HT, Ozyilmaz I, Ozturk E, Onan IS, Haydin S, Erek E, Yeniterzi M, Guzeltas A, Odemis E, Bakir I. Factors affecting perioperative mortality in tetralogy of Fallot. Pediatr Int. 2015 Oct;57(5):832-9.
17. Yangni-Angate KH, Meneas C, Diby F, Diomande M, Adoubi A, Tanauh Y. Cardiac surgery in Africa: a thirty-five year experience on open heart surgery in Cote d'Ivoire. Cardiovasc Diagn Ther. 2016 Oct;6(Suppl 1):S44-S63.
18. IB Diop, ID Bindia, NH Mahamat Ahmat, G Ciss, M Kaya, K Ba, NA Sarr, S Manga, M Dioum, EA Tine, O Dieye, EM Sarr, M Seye, L Sy. Chirurgie des cardiopathies congénitales au Sénégal : Expérience du Centre Cardio-pédiatrique CUOMO de Dakar-CHU FANN. Cardiologie Tropicale. N° 160, Avril-Mai-Juin 2020.
19. Mottier V, Prsa M. Contemporary early results of late repair of tetralogy of Fallot in children: a single-centre case series. Swiss Med Wkly. 2021 Mar 29;151:w20491.
20. Damien Schaffner, Guillaume Maitre, SebastianoA.G. Lava, Yann Boegli, Mirko Dolci, Raymond Pfister, Nicole Sekarski, Perez Marie-Hélène, Stefano Di Bernardo. Outcome of humanitarian patients with late complete repair of tetralogy of Fallot: A 13-year long single-center experience. International Journal of Cardiology Congenital Heart Disease. Volume 10, December 2022, 100414.
21. Benbrik N, Romefort B, Le Gloan L, Warin K, Hauet Q, Guerin P, Baron O, Gournay V. Late repair of tetralogy of Fallot during childhood in patients from developing countries. Eur J Cardiothorac Surg. 2015 Mar;47(3):e113-7.
22. Romeo JLR, Etnel JRG, Takkenberg JJM, Roos-Hesselink JW, Helbing WA, van de Woestijne P, Bogers AJJC, Mokhles MM. Outcome after surgical repair of tetralogy of Fallot: A systematic review and meta-analysis. J Thorac Cardiovasc Surg. 2020 Jan;159 (1) :220-236.
23. Wei X, Li T, Ling Y, Chai Z, Cao Z, Chen K, Qian Y. Transannular patch repair of tetralogy of Fallot with or without monocusp valve reconstruction: a meta-analysis. BMC Surg. 2022 Jan 16;22(1):18.
24. Singh NM, Loomba RS, Gudausky TM, Mitchell ME. Monocusp valve placement in children with tetralogy of Fallot undergoing repair with transannular patch: A functioning pulmonary valve does not improve immediate postsurgical outcomes. Congenit Heart Dis. 2018 Nov;13(6):935-943.
25. Awori MN, Awori JA, Mehta NP, Makori O. Monocusp Valves Do Not Improve Early Operative Mortality in Tetralogy of Fallot: A Meta-Analysis. World J Pediatr Congenit Heart Surg. 2020 Sep;11(5):619-624.
26. Huang SW, Hsu WF, Li HY, Hwang B, Wu FY, Weng ZC, Chuang CM, Chen SJ, Wang CC, Wang DS, Lee PC. Implantation of monocusp valve prolongs the duration of chest tube drainage in children with tetralogy of fallot after corrective surgery. J Chin Med Assoc. 2022 Mar 1;85(3):364-368.