REVIEW

Journal of Indonesian Thoracic Cardiac and Vascular Surgery *(JINATCVS)* 2025,
Volume 2, Number 2: 61-65

Staged repair versus primary repair strategy in a neonate with symptomatic tetralogy of Fallot: a systematic review

Published By :
The Indonesian Association of Thoracic and Vascular Surgeons

Herick Alvenus Willim[1*], Zhana Daisya Triani[1], Elva Katharina Simamora[1], Salomo Purba[1,2]

[1]Cardiothoracic and Vascular Surgery Division, Department of Surgery, Faculty of Medicine, Universitas Indonesia, Jakarta, Indonesia;
[2]Pediatric and Congenital Heart Surgery Department, National Cardiovascular Center Harapan Kita, Jakarta, Indonesia.

*Corresponding to:
Herick Alvenus Willim;
Cardiothoracic and Vascular Surgery Division, Department of Surgery, Faculty of Medicine, Universitas Indonesia, Jakarta, Indonesia;
dr.herickalvenuswillim@gmail.com

Received: 2025-06-25
Accepted: 2025-08-26
Published:2025-09-29

ABSTRACT

Introduction: The most prevalent cyanotic congenital cardiac condition is tetralogy of Fallot (TOF), which has a significant fatality rate in neonates who exhibit symptoms if treatment is not received. In this group, two surgical techniques are frequently employed: primary repair and staged repair. The best course of action is still up for dispute, though. The purpose of this study is to compare the clinical results of staged repair versus primary repair in order to assess the surgical strategy for symptomatic newborn TOF.

Methods: The PubMed, Cochrane, and ScienceDirect databases were searched for relevant material. Mortality, morbidity, duration of stay, and reintervention were among the postoperative outcomes evaluated. The Oxford Centre for Evidence-Based Medicine's recommendations were adhered to during the critical evaluation process.

Results: Seven studies were analyzed, and the results showed no discernible difference between staged and primary repair in terms of mortality or reintervention rates. Nonetheless, staged repair was linked to more expensive and prolonged hospitalizations, whereas primary repair carried a greater risk of complications.

Conclusion: It has not been demonstrated that any surgical technique is consistently better across all clinical outcomes. For high-risk newborns, such as those with low birth weight or pulmonary artery hypoplasia, staged repair seems more suitable. Individualized surgical decisions should take into account the anatomy of the patient, institutional knowledge, and the facilities that are available.

Keywords: clinical outcomes, neonatal tetralogy of Fallot, primary repair, staged repair.
Cite This Article: Willim, H.A., Triani, Z.D., Simamora, E.K., Purba, S. 2025. Staged repair versus primary repair strategy in a neonate with symptomatic tetralogy of Fallot: a systematic review. *Journal of Indonesian Thoracic Cardiac and Vascular Surgery* 2(2): 61-65

INTRODUCTION

The most common cyanotic congenital heart disease (CHD) is tetralogy of Fallot (TOF), which affects around 3 out of 10,000 live infants and makes up 6–7% of all CHD cases.[1,2] In around 3% of cases, genetic factors are implicated in the multifactorial etiology.[2] Non-restrictive ventricular septal defect (VSD), overriding aorta, right ventricular outflow tract (RVOT) blockage (infundibular, valvular, or supravalvular pulmonary stenosis), and right ventricular hypertrophy are the four characteristic cardiac abnormalities that characterize TOF.[3] The clinical presentation of TOF can vary, encompassing milder forms to more complex variants like TOF with pulmonary atresia or double outlet right ventricle.[3] In untreated TOF, the primary causes of death include hypoxic or tet spells (68%), stroke (17%), and brain abscess (13%). Without surgical correction, the prognosis is poor; around 25% of patients with severe RVOT obstruction die within the first year, 40% by age 3, 70% by age 10, and 95% by age 40.[4]

For patients who are asymptomatic or just slightly symptomatic, final corrective surgery within the first year—ideally within three to six months—is the best course of treatment for TOF. Patients who are highly symptomatic, however, should have surgery right away.[5] Staged repair and primary repair are the two main techniques that are being debated as the best strategies for newborns with symptoms.[6] Palliative measures like the Blalock-Taussig-Thomas (BTT) shunt, RVOT stenting, or patent ductus arteriosus (PDA) stenting are used prior to staged repair in order to boost pulmonary blood flow and encourage vascular development before complete correction is performed later. A transannular patch (TAP) is frequently necessary for primary repair, a one-stage operation that has an early mortality rate of around 7%, increases the risk of arrhythmias and reintervention, and lengthens the length of time in the intensive care unit.[7]

Anatomical circumstances, mortality rates, the chance of reintervention, and the danger of postoperative complications all influence the choice of TOF repair approach.[6] For infants weighing more than 3 kg and having a suitable pulmonary artery size, some specialists recommend primary repair. On the other hand, high-risk neonates—those with low birth weight, preterm delivery, pulmonary artery hypoplasia, or other comorbidities—are advised to undergo staged repair. The best

surgical approach is still up for discussion, though.[7] This systematic review study aims to evaluate the optimal surgical approach for managing a neonate with symptomatic TOF by critically appraising current evidence in a clinical scenario.

METHODS

Search strategy

A systematic literature search was conducted using PubMed, Cochrane, and ScienceDirect databases. The keywords used were a combination of: "Primary Repair," "Primary Approach," "Staged Repair," "Staged Approach," "Neonates," "Newborn," "Tetralogy of Fallot," "Fallot's Tetralogy," "TOF," "Outcomes," "Morbidity," "Mortality," "Complication," "Reintervention," and "Reoperation." The keywords were connected using "AND" or "OR." The study selection was limited to research published within the last 10 years (2016–2025). No language or geographical restrictions were applied.

Inclusion and exclusion criteria

Inclusion criteria consisted of original studies involving symptomatic neonates with TOF that compared postoperative outcomes between staged and primary repair, with study designs including systematic reviews, meta-analyses, randomized controlled trials (RCTs), or cohort studies. Exclusion criteria comprised case reports, literature reviews, letters, editorials, conference abstracts, studies unrelated to TOF surgery, or those lacking precise postoperative outcome data.

Critical appraisal

Articles meeting the above criteria were assessed for their level of evidence and critically appraised using the Oxford Centre for Evidence-Based Medicine (CEBM) Critical Appraisal Tools. The assessment was conducted based on three aspects: validity, importance, and applicability.[8]

RESULTS

Literature search

A total of 371 possible publications were found in the first search, including 340 from ScienceDirect, 3 from Cochrane, and 28 from PubMed. 310 papers were screened using their titles and abstracts after duplicate articles were eliminated. Twenty-eight papers were chosen for full-text examination after the screening procedure. Seven publications were finally included for study. Figure 1 displays the flow chart for the literature search.

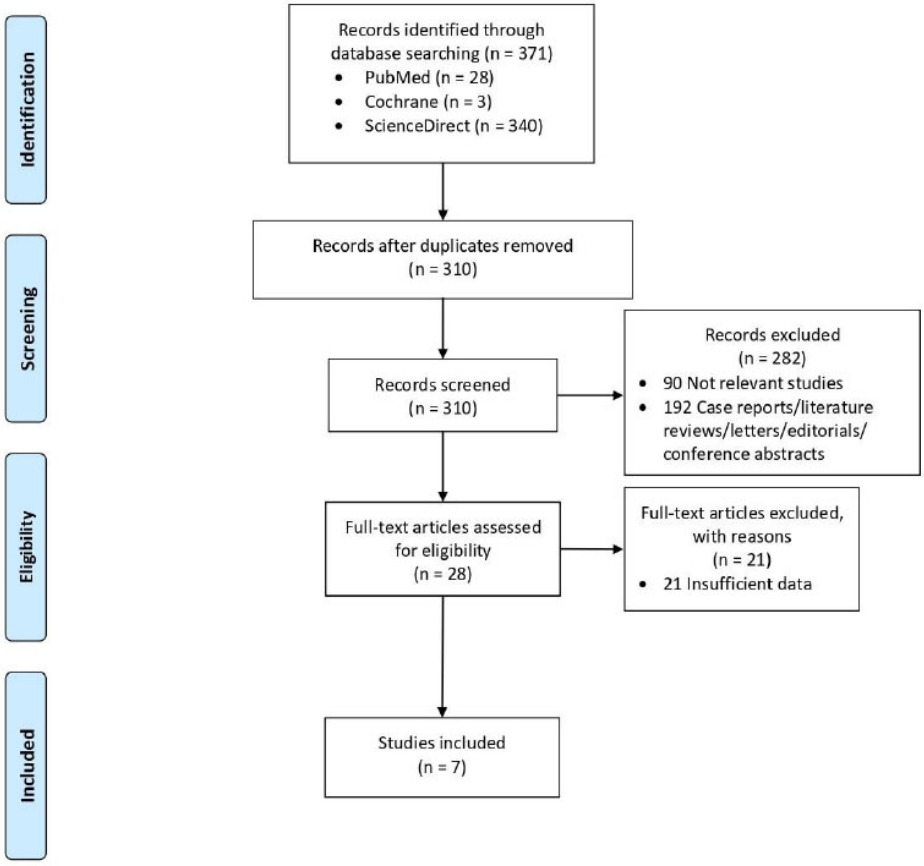

Figure 1. Literature search flow chart.

Study characteristics

A total of seven studies were selected. One study was a meta-analysis.[9] Six studies were retrospective cohort.[10-15] The outcomes reported in these studies included mortality, postoperative complications, duration of ICU and hospital stay, and reintervention. The characteristics of the selected studies are presented in Table 1. The largest study, a meta-analysis by Busro et al. (2024) involving 4,464 patients, provided Level 2a evidence, while the remaining six were retrospective cohort studies offering Level 2b evidence. Sample sizes across these cohorts ranged from 120 to 2,363 participants. All studies evaluated key clinical outcomes, including mortality, postoperative complications, length of hospital stay, and reintervention rates. Collectively, the data provide moderate-level evidence indicating that both surgical strategies yield comparable mortality and reintervention outcomes, though variations exist in complication rates and hospitalization duration.

Critical Appraisal

Critical appraisal was conducted on the seven selected studies, covering the aspects of validity, importance, and applicability. The results of the critical appraisal are presented in Table 2. Across all studies, methodological validity was strong, featuring clearly defined outcomes, representative samples, and appropriate statistical adjustments such as propensity score weighting or regression analysis. The meta-analysis by Busro et al. (2024) demonstrated robust validity through a comprehensive search and inclusion of moderate- to high-quality studies. In terms of importance, most studies consistently reported no significant difference in mortality or reintervention rates between the two approaches. However, primary repair was generally associated with a higher incidence of cardiac complications

Table 1. Characteristics of the included studies

Author	Study design	Sample size (n)	Intervention	Comparison	Outcomes	Level of Evidence
Busro et al., 2024[9]	Meta-analysis	4464	Staged repair	Primary repair	Mortality, complications, length of stay, reintervention	2a
Meadows et al., 2023[10]	Retrospective cohort	282	Staged repair	Primary repair	Mortality, complications, length of stay, reintervention	2b
Qureshi et al., 2022[11]	Retrospective cohort	120	Staged repair	Primary repair	Mortality, complications, length of stay, reintervention	2b
O'Byrne et al., 2022[12]	Retrospective cohort	324	Staged repair	Primary repair	Mortality, complications, length of stay, reintervention	2b
Goldstein et al., 2021[13]	Retrospective cohort	342	Staged repair	Primary repair	Mortality, complications, length of stay, reintervention	2b
Bailey et al., 2020[14]	Retrospective cohort	138	Staged repair	Primary repair	Mortality, complications, length of stay, reintervention	2b
Savla et al., 2019[15]	Retrospective cohort	2363	Staged repair	Primary repair	Mortality, complications	2b

Table 2. Critical appraisal of the included studies

Author	Validity	Importance	Applicability
Busro et al., 2024[9]	- This meta-analysis had a clearly defined PICO - Comprehensive search across 6 major databases - Appropriate inclusion criteria - Included studies rated moderate to high quality - Consistent outcomes reported across studies	- There was no discernible difference in the two groups' death rates (OR=1.26; p=0.42). - The primary repair group experienced 1.5 times as many cardiac complications as the staged repair group (OR=1.50; p=0.02). - The staged repair group experienced lengthier hospital and intensive care unit stays than the primary repair group (MD=3.06; p<0.001, and MD=11.84; p<0.001). - Transannular patches were used 2.62 times more frequently in the primary repair group than in the staged repair group (OR=2.62; p<0.001). - The two groups' reintervention rates did not differ substantially (OR=0.86; p=0.21).	Clinical decision-making in newborns with symptomatic TOF can benefit from the results.
Meadows et al., 2023[10]	- This retrospective cohort study included a representative sample - Follow-up duration was adequate (median 5.2 years) - The outcomes were clearly defined - Baseline differences were adjusted using propensity score weighting	- There was no discernible difference in mortality between the two groups (p=0.72). - There was no significant difference in complications between the two groups (p=0.895). - The staged repair group experienced lengthier hospital and intensive care unit stays than the primary repair group (p<0.001 and p<0.001). - The two groups' reintervention rates did not differ substantially (p=0.316).	Clinical decision-making in newborns with symptomatic TOF can benefit from the results.
Qureshi et al., 2022[11]	- This retrospective cohort study included a representative sample - Follow-up duration was adequate (median 5.3 years) - The outcomes were clearly defined - Baseline differences were adjusted using propensity score weighting	- There was no discernible difference in mortality between the two groups (p=0.214). - There was no significant difference in complications between the two groups (p=0.777). - The staged repair group experienced lengthier hospital and intensive care unit stays than the primary repair group (p<0.001 and p<0.001). - The two groups' reintervention rates did not differ substantially (p=0.072).	Clinical decision-making in newborns with symptomatic TOF can benefit from the results.
O'Byrne et al., 2022[12]	- This retrospective cohort study included a representative sample - Follow-up duration was adequate (median 1.5 years) - The outcomes were clearly defined - Baseline differences were adjusted using propensity score weighting	- There was no discernible difference in mortality between the two groups (p=0.18). - There was no significant difference in complications between the two groups (p=0.10). - The staged repair group experienced lengthier hospital and intensive care unit stays than the primary repair group (p<0.001 and p<0.001). - The two groups' reintervention rates did not differ substantially (p=0.22).	Clinical decision-making in newborns with symptomatic TOF can benefit from the results.

Author	Validity	Importance	Applicability
Goldstein et al., 2021[13]	- This retrospective cohort study included a representative sample - Follow-up duration was adequate (median 4.3 years) - The outcomes were clearly defined - Baseline differences were adjusted using propensity score weighting	- There was no discernible difference in mortality between the two groups (p=0.25). - There was no significant difference in complications between the two groups (p=0.20). - The staged repair group experienced lengthier hospital and intensive care unit stays than the primary repair group (p<0.001 and p<0.001). - The two groups' reintervention rates did not differ substantially (p=0.50).	Clinical decision-making in newborns with symptomatic TOF can benefit from the results.
Bailey et al., 2020[14]	- This retrospective cohort study included a representative sample - Follow-up duration was adequate (median 2 years) - The outcomes were clearly defined - Baseline differences were adjusted using linear and logistic regression	- There was no discernible difference in mortality between the two groups (p=0.19). - The primary repair group experienced more complications than the staged repair group (p=0.01). - The staged repair group experienced lengthier hospital and intensive care unit stays than the primary repair group (p<0.001 and p=0.016). - The two groups' reintervention rates did not differ substantially (p=0.63).	Clinical decision-making in newborns with symptomatic TOF can benefit from the results.
Savla et al., 2019[15]	- This retrospective cohort study included a representative sample - Follow-up duration was adequate (median 2 years) - The outcomes were clearly defined - Baseline differences were adjusted using propensity score weighting	- The primary repair group had a greater mortality rate than the staged repair group (p=0.024). - The primary repair group experienced more complications than the staged repair group (p<0.05).	Clinical decision-making in newborns with symptomatic TOF can benefit from the results.

and greater use of transannular patches, while staged repair resulted in longer hospital and ICU stays. Savla et al. (2019) uniquely found higher mortality and complication rates in the primary repair group. Overall, all studies concluded that the findings are clinically applicable to decision-making for neonates with symptomatic TOF, emphasizing the need for individualized surgical planning based on patient risk and institutional expertise.

DISCUSSION

The choice of surgical strategy for neonates with symptomatic TOF remains a topic of debate among pediatric cardiac surgeons. The two approaches to TOF correction, staged repair and primary repair, each have their own advantages and disadvantages. Based on the analysis of seven included studies, consisting of one meta-analysis and six retrospective cohort studies, no single strategy was found to be superior in all clinical aspects. However, differences in outcomes were observed across several parameters.[9-15]

In terms of mortality, only one retrospective study reported that primary repair was associated with a higher risk of mortality compared to staged repair, whereas five other retrospective studies found no significant difference in mortality between the two approaches.[10-15] Busro et al.'s meta-analysis, which likewise revealed no discernible difference in mortality between the two groups, lends more credence to this conclusion.[9] Postoperative outcomes for TOF correction, whether through a staged or primary approach, are generally favorable, with long-term survival rates remaining high. The survival rates at 1, 5, 10, 20, and 25 years are reported to be 98.6%, 97.8%, 97.1%, 95.5%, and 94.5%, respectively.[16]

Postoperative cardiac complications in TOF patients can occur both early and long term, including residual VSD, residual RVOT obstruction, pulmonary regurgitation, right heart failure, endocarditis, and arrhythmias.[17] A meta-analysis by Busro et al. found that cardiac complications were 1.5 times more frequent in the primary repair group compared to the staged repair group.[9] One possible explanation is that staged repair allows better cardiovascular adaptation in neonates, particularly in supporting pulmonary artery maturation. Moreover, the higher use of transannular patches in the primary repair group, while effective in relieving RVOT obstruction, may increase the risk of pulmonary valve regurgitation, arrhythmias, and heart failure.[18]

All included studies reported that staged repair was associated with longer ICU and hospital stays compared to primary repair. It is likely due to the additional surgical procedures required in staged repair, which cumulatively extend the total treatment duration. According to the meta-analysis by Busro et al., ICU stay was approximately 3 days longer, and hospital stay up to 12 days longer in the staged group. As a result, staged repair tends to incur higher healthcare costs, which may influence surgical decision-making.[9]

The rate of reintervention is an important factor when comparing staged and primary repair strategies. All included studies reported no significant difference in reintervention rates between the two groups.[9-15] One retrospective study showed that TOF repair in neonates with confluent pulmonary arteries yields favorable outcomes if no primary non-cardiac conditions are present. Early neonatal surgery supports pulmonary artery growth with similar reintervention rates as later repairs. However, preoperative weight below 2.5 kg and small pulmonary arteries were associated with higher reintervention risk.[19]

This study has several limitations. First, the analysis was based on only seven studies, which limits the statistical power and generalizability of the findings. Considerable heterogeneity existed among the included studies in terms of patient selection, surgical techniques, institutional expertise, and perioperative management, potentially influencing the results. Moreover, reliance on published literature introduces a risk of publication bias, as studies with negative or inconclusive findings may be underrepresented. Most of the included studies were retrospective in nature, which restricts causal interpretation and increases susceptibility to confounding factors.

CONCLUSION

No single strategy is universally superior for managing neonates with symptomatic TOF. Primary repair offers shorter hospital stays but is associated with more postoperative cardiac complications. Staged repair allows better cardiovascular adaptation with fewer complications, but involves more extended and more costly care. In high-risk cases, such as low birth weight neonates with pulmonary artery hypoplasia, staged repair is more appropriate. Surgical decisions should be individualized based on anatomical characteristics, institutional experience, and available facilities.

FUNDING

None.

CONFLICT OF INTEREST

There are no disclosed conflicts of interest for the writers.

AUTHOR CONTRIBUTION

Each contributor made an equal contribution to the manuscript's composition.

ETHICAL CONSIDERATIONS

None.

REFERENCES

1. Love BA. Management of Infants With Tetralogy of Fallot: Questioning Conventional Wisdom. J Am Coll Cardiol. 2023;82(7):628–30. Available from: https://doi.org/10.1016/j.jacc.2023.06.008
2. Sagatov IY, Sapunov AV. Tetralogy of Fallot: Modern Data. Review of Literature. Bull Surg Kazakhstan. 2022;2:26–30. Available from: https://doi.org/10.35805/bsk2022ii026
3. Bassett AS, Reuter MS, Malecki S, Silversides C, Oechslin E. Clinically Relevant Genetic Considerations for Patients With Tetralogy of Fallot. CJC Pediatr Congenit Hear Dis. 2023;2(6):426–39. Available from: https://doi.org/10.1016/j.cjcpc.2023.10.002
4. Elizabeth Kaiser A, Husnain MA, Fakhare Alam L, Kumar Murugan S, Kumar R. Management of Fallot's Uncorrected Tetralogy in Adulthood: A Narrative Review. Cureus. 2024;16:8. Available from: https://doi.org/10.7759/cureus.67063
5. Martins IF, Doles IC, Bravo-Valenzuela NJM, Dos Santos AOR, Varella MSP. When is the best time for corrective surgery in patients with tetralogy of fallot between 0 and 12 months of age? Brazilian J Cardiovasc Surg. 2018;33(5):505–10. Available from: https://doi.org/10.21470/1678-9741-2018-0019
6. Juaneda I, Peirone A, Diaz J, Azar I, Molinas R, Guevara A, et al. Tetralogy of Fallot Repair After Neonatal Right Ventricular Outflow Tract Stenting: Initial Multicenter Experience in Argentina. World J Pediatr Congenit Hear Surg. 2023;14(2):222–6. Available from: https://doi.org/10.1177/21501351221140097
7. Vanderlaan RD, Barron DJ. Optimal Surgical Management of Tetralogy of Fallot. CJC Pediatr Congenit Hear Dis. 2023;2(6):352–60. Available from: https://doi.org/10.1016/j.cjcpc.2023.09.003
8. Center for Evidence-Based Medicine. Critical Appraisal tools [Internet]. 2025. Available from: https://www.cebm.ox.ac.uk/resources/ebm-tools/critical-appraisal-tools
9. Busro PW, Satria R, Safitri S, Sudirman AR, Bachmid MR, Fardhani DG, et al. Primary versus staged repair for tetralogy of Fallot in symptomatic neonates: systematic review and meta-analysis. Cardiol Young. 2024;34(10):2156–63. Available from: https://doi.org/10.1017/S1047951124025575
10. Meadows JJ, Bauser-Heaton H, Petit CJ, Goldstein BH, Qureshi AM, McCracken CE, et al. Comparison of treatment strategies for neonates with tetralogy of Fallot and pulmonary atresia. J Thorac Cardiovasc Surg. 2023;166(3):916-925.e6. Available from: https://doi.org/10.1016/j.jtcvs.2023.01.008
11. Savla JJ, Faerber JA, Huang YS, Zaoutis T, Goldmuntz E, Kawut SM M-RL. Two-year outcomes after complete or staged procedure for tetralogy of Fallot in neonates. J Am Coll Cardiol. 2019;74(12):1570–9. Available from: https://doi.org/10.1016/j.jacc.2019.05.057
12. Qureshi AM, Caldarone CA, Romano JC, Chai PJ, Mascio CE, Glatz AC, et al. Comparison of management strategies for neonates with symptomatic tetralogy of Fallot and weight <2.5 kg. J Thorac Cardiovasc Surg. 2022;163(1):192-207.e3. Available from: https://doi.org/10.1016/j.jtcvs.2021.01.100
13. O'Byrne ML, Glatz AC, Huang Y shung V., Kelleman MS, Petit CJ, Qureshi AM, et al. Comparative costs of management strategies for neonates with symptomatic tetralogy of Fallot. J Am Coll Cardiol. 2022;79(12):1170–80. Available from: https://doi.org/10.1016/j.jacc.2021.12.036
14. Goldstein BH, Petit CJ, Qureshi AM, McCracken CE, Kelleman MS, Nicholson GT, et al. Comparison of management strategies for neonates with symptomatic tetralogy of Fallot. J Am Coll Cardiol. 2021;77(8):1093–106. Available from: https://doi.org/10.1016/j.jacc.2020.12.048
15. Bailey J, Elci OU, Mascio CE, Mercer-Rosa L GE. Staged versus complete repair in the symptomatic neonate with tetralogy of Fallot. Ann Thorac Surg. 2020;109(3):802–8. Available from: https://doi.org/10.1016/j.athoracsur.2019.10.013.Staged
16. Smith CA, McCracken C, Thomas AS, Spector LG, St Louis JD, Oster ME, et al. Long-term outcomes of tetralogy of Fallot: A study from the Pediatric Cardiac Care Consortium. JAMA Cardiol. 2019;4(1):34–41. Available from: https://doi.org/10.1001/jamacardio.2018.4255
17. Wilson R, Ross O, Griksaitis MJ. Tetralogy of Fallot. BJA Educ. 2019;19(11):362–9. Available from: https://doi.org/10.1016/j.bjae.2019.07.003
18. van der Ven JPG, van den Bosch E, Bogers AJCC, Helbing WA. Current outcomes and treatment of tetralogy of fallot. F1000Research. 2019;8:1–15. Available from: https://doi.org/10.12688/f1000research.17174.1
19. Kolcz J, Pizarro C. Neonatal repair of tetralogy of Fallot results in improved pulmonary artery development without increased need for reintervention. Eur J Cardio-thoracic Surg. 2005;28(3):394–9. Available from: https://doi.org/10.1016/j.ejcts.2005.05.014