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Nursing care in patients with Tetralogy of Fallot: a systematic review of the literature

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ABSTRACT

Background: Tetralogy of Fallot (ToF) is the most common cyanotic congenital heart disease, with an incidence of approximately 1 in 3,500 live births, accounting for 7–10% of all congenital heart defects. Advances in cardiac surgery and intensive care have increased survival into adulthood to over 85%, resulting in a growing population of adults with repaired congenital heart disease and, in resource-limited settings, adults with untreated or previously undiagnosed disease. In this context, specialist nursing care plays a pivotal role throughout the continuum of care.

Objective: To systematically review the available scientific evidence on nursing care for patients with tetralogy of Fallot, with particular emphasis on nursing interventions during the preoperative and postoperative phases and on the competencies required across different clinical settings.

Materials and Methods: A systematic literature review was conducted between November 2025 and March 2026 using PubMed, the Cochrane Library, and CINAHL, following the PRISMA methodology. The search strategy was: care AND tetralogy of Fallot AND nurs*. Of the 56 records initially identified, 33 articles were screened after duplicate removal. Following the application of the eligibility criteria and methodological quality assessment using the Joanna Briggs Institute (JBI) Critical Appraisal Checklist, with an inclusion threshold of ≥75%, four studies were included in the review. The selected studies comprised two case reports and two narrative reviews.

Results: The four included studies collectively addressed the major stages of the care pathway for patients with ToF: neonatal preoperative stabilization (Tran et al., 2022), postoperative management in the pediatric intensive care unit (Forman et al., 2019), nursing management of adults with unrepaired ToF (Araújo et al., 2017), and peri-anesthetic assessment of an adult patient with previously undiagnosed ToF (Edwards et al., 2025). Four principal themes emerged: systematic clinical monitoring as a strategy for preventing complications; management of hypercyanotic (Tet) spells; implementation of

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the nursing process using the standardized NANDA-I, NIC, and NOC taxonomies; and family support as an integral component of comprehensive nursing care.

Discussion: Comparison of the included studies demonstrated strong consistency regarding the core principles of nursing care for patients with ToF, including systematic monitoring, prevention of complications, individualized care, and family-centered support. These principles are adapted according to the stage of care, patient age, and clinical setting. Particular attention emerged regarding the management of adults with ToF, whether surgically repaired or previously undiagnosed, representing an increasingly important challenge for specialist nursing practice. The expanding population of adults with congenital heart disease requires nurses with advanced competencies in clinical assessment, critical thinking, and cultural sensitivity.

Conclusions: Nurses play a key and irreplaceable role in the care pathway of patients with tetralogy of Fallot. Specialist clinical expertise, an evidence-based systematic approach, early clinical assessment, and family-centered care constitute the cornerstones of high-quality nursing care for this complex condition. The development of specialized educational pathways in adult congenital cardiology nursing should be considered a priority to ensure safe, equitable, and high-quality care for this continuously growing patient population.

INTRODUCTION

Congenital heart disease (CHD) represents the most common group of congenital malformations, with an estimated global incidence ranging from 6 to 13 cases per 1,000 live births, corresponding to approximately 1.35 million new cases annually worldwide [1]. Among these conditions, tetralogy of Fallot (ToF) is of particular clinical significance. It is the most common cyanotic congenital heart disease, accounting for 7–10% of all congenital heart defects and approximately 75% of cyanotic congenital heart diseases. ToF is characterized by the coexistence of four anatomical abnormalities: right ventricular outflow tract obstruction, a large ventricular septal defect, overriding of the aorta, and right ventricular hypertrophy. In patients with significant right ventricular outflow tract obstruction, these abnormalities result in reduced pulmonary blood flow and systemic hypoxemia, which clinically manifests as cyanosis, reduced exercise tolerance, and, in the most severe cases, paroxysmal episodes of acute desaturation known as hypercyanotic spells or "Tet spells" [2]. Since Étienne Fallot first systematically described the condition in 1888, through the therapeutic breakthrough of the Blalock–Taussig shunt introduced in 1944 [3], and subsequently the development of modern techniques for complete intracardiac repair under cardiopulmonary bypass, clinical outcomes have improved dramatically. Operative mortality, which historically reached 25%, has now approached 0% for patients with typical anatomical forms treated in specialized centers, with 30-year survival rates exceeding 95% [4,11]. These remarkable advances have transformed ToF from a lethal disease of early childhood into a manageable chronic condition, resulting in a steadily growing population of adults living with this diagnosis for decades. In this context, surgical or transcatheter pulmonary valve replacement (PVR) has become a cornerstone of long-term follow-up. Recent evidence demonstrates that timely PVR is associated with a significant reduction in the risk of sudden cardiac death and sustained ventricular tachycardia compared with conservative management [12]. Within this evolving clinical landscape, the role of nurses has expanded substantially, encompassing neonatal intensive care units, where infants diagnosed prenatally are stabilized; pediatric

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cardiac intensive care units, where complex postoperative complications are managed; and adult cardiology departments and preoperative assessment clinics, where adults with repaired, unrepaired, or previously undiagnosed ToF require increasingly specialized nursing expertise [5,6]. Moreover, the growing prevalence of adults living with congenital heart disease—which now exceeds that of pediatric patients in high-income countries [13] —poses new challenges that nursing professionals must address through advanced education and specialized training. Despite its clinical relevance, the nursing literature specifically addressing ToF remains relatively limited compared with the medical and surgical literature. Systematic reviews comprehensively evaluating nursing interventions throughout the different phases of the care pathway are scarce. This gap provides the rationale for the present systematic review.

MATERIALS AND METHODS

This systematic literature review was conducted between November 2025 and March 2026. The literature search was performed using three biomedical databases: MEDLINE, the Cochrane Library, and CINAHL. The PICO framework was employed to formulate the research question and identify the most appropriate search terms to achieve the objective of this systematic review. The following search strategy was applied across all three databases: care AND tetralogy of Fallot AND nurs *

Inclusion and Exclusion Criteria

Inclusion criteria were as follows: studies published within the previous 10 years; studies involving human subjects; articles published in English, Italian, or Spanish; studies addressing nursing care in patients with tetralogy of Fallot; and articles available in full text.

Exclusion criteria included studies not relevant to the scope of the review (n = 3); studies outside the thematic focus (n = 10); articles exclusively related to medical practice (n = 5); studies involving populations from healthcare settings not consistent with the objectives of the review (n = 10); and articles unavailable in full text (n = 1).

Study Selection Process

The search strategy yielded a total of 56 articles: 19 from PubMed, 4 from the Cochrane Library, and 33 from CINAHL. After duplicate removal, 33 articles underwent title and abstract screening. Application of the predefined inclusion and exclusion criteria resulted in the exclusion of 28 articles. Of the five potentially eligible studies, only four were available in full text and were therefore included in the review.

Methodological Quality Assessment

The four full-text articles were critically appraised using the Joanna Briggs Institute (JBI) Critical Appraisal Checklists, applying the versions appropriate to each study design: the JBI Critical Appraisal Checklist for Case Reports and the JBI Critical Appraisal Checklist for Text and Opinion Papers. An inclusion threshold corresponding to an acceptable methodological quality score of at least 75% was established. All four studies met or exceeded this threshold and were therefore included in the systematic review. The review was conducted according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines. The study selection process is illustrated in the PRISMA flow diagram (Figure 1).

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RESULTS

The studies included in the review are presented in Table 1, which summarizes their main characteristics and findings. The table includes the following information: authors, year of publication, study design, and bibliographic reference; article title; journal; study setting; study summary; and main findings.

DISCUSSION

Clinical Context and Study Populations

The four studies included in this review encompass clinically heterogeneous populations spanning the entire lifespan of patients with tetralogy of Fallot (ToF)—from the neonatal period to adulthood—and reflect markedly different healthcare settings in terms of clinical environment, available resources, and complexity of care. Tran et al. (2022) conducted their study in the neonatal intensive care unit (NICU) of Children's Hospital Los Angeles, focusing on neonates with complex congenital heart disease requiring preoperative stabilization. Approximately 40,000 newborns are diagnosed with congenital heart disease annually in the United States [8]. When available, prenatal diagnosis plays a pivotal role in reducing preoperative mortality by enabling timely care planning and referral to highly specialized cardiac centers. Current American Heart Association guidelines recommend structured Individualized Family-Centered Developmental Care (IFDC) programs for neonates with complex CHD admitted to NICUs, emphasizing that developmentally supportive nursing practices remain inconsistently implemented across centers and that specialized nursing education is associated with greater adoption of these practices [14]. Forman et al. (2019), working at Kentucky Children's Hospital and Cincinnati Children's Hospital, focused on pediatric patients during the immediate postoperative period in the pediatric intensive care unit (PICU). Tetralogy of Fallot accounts for 7–10% of all congenital heart diseases. Owing to advances in congenital cardiac surgery and intensive care, early operative mortality has declined from the historical rate of 25% to approximately 2%, with survival into adulthood exceeding 85% [4]. This epidemiological transition has created a new clinical reality: an expanding population of adults with repaired congenital heart disease requiring lifelong specialized follow-up. Marelli et al. demonstrated that, since the early 2000s, the prevalence of adults living with congenital heart disease has surpassed that of children in high-income countries [13]. Araújo et al. (2017) described the case of a 23-year-old woman with unrepaired ToF treated in the cardiology department of a Brazilian university hospital. Although the diagnosis had been established during childhood, the patient had never received specialist follow-up or undergone corrective surgery because of limited access to healthcare services. This case reflects the systemic challenges faced by low- and middle-income countries, where 47–63% of congenital heart diseases are diagnosed only after irreversible complications have developed and where limited access to echocardiography, shortages of specialized personnel, and prohibitive healthcare costs constitute major barriers to care [15]. Finally, Edwards et al. (2025) reported the case of a 25-year-old migrant patient who presented for elective orthopedic surgery with previously undiagnosed ToF. This case illustrates the vulnerability of migrant populations, who often experience fragmented access to healthcare services [9]. International evidence indicates that migrants living in high-income countries encounter multiple barriers to healthcare—including lack of health insurance, transportation difficulties, and unfamiliarity with healthcare systems—which frequently result in delayed medical attention, more advanced

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disease, and poorer clinical outcomes [16]. Overall, the diversity of patient populations represented in these studies confirms that nursing care for ToF extends far beyond pediatric cardiac surgery units and encompasses the entire continuum of care across the patient's lifespan.

Clinical Monitoring and Prevention of Complications

All four studies consistently identify continuous and systematic assessment of vital signs as the cornerstone of nursing care for patients with ToF. Tran et al. [8] describe comprehensive physical assessment including capillary refill, skin temperature, peripheral oxygen saturation, heart rate and rhythm, and neurological status. Forman et al. [4] further expand postoperative monitoring to include central venous pressure, mixed venous oxygen saturation, mediastinal drainage output, and bedside echocardiographic assessment. Araújo et al. [7] identify heart rate and rhythm monitoring as the primary intervention addressing the patient's priority nursing diagnosis. Edwards et al. [9] demonstrate how comprehensive preoperative physical assessment, as an autonomous nursing responsibility with high diagnostic value, can identify previously undiagnosed ToF in an adult patient, thereby preventing potentially life-threatening perioperative complications. Management of hypercyanotic ("Tet") spells emerged as another key nursing intervention across multiple studies. Tran et al. [8] describe a standardized emergency management protocol consisting of immediate knee–chest positioning, supplemental oxygen administration, and sedation. Edwards et al. [9] reported that the adult patient spontaneously relieved symptoms by assuming a posture analogous to squatting—the physiological equivalent of the knee–chest position—highlighting how careful interpretation of this historical clue may raise clinical suspicion even in the absence of a previous diagnosis. An increasingly important aspect concerns neurodevelopmental outcomes. Recent evidence indicates that more than half of infants with complex congenital heart disease undergoing cardiac surgery during the first year of life will experience some degree of cognitive, motor, or language impairment [17], making preoperative and postoperative neurological assessment an essential component of specialist nursing care.

Postoperative Nursing Management

Forman et al. [4] provide the most comprehensive and detailed description of postoperative management in ToF, outlining the principal complications encountered by pediatric intensive care nurses during the first hours and days following surgical repair. Regarding hemodynamic monitoring, the authors identify continuous rhythm monitoring using temporary epicardial pacing wires, central venous pressure measurement, peripheral perfusion assessment, hourly urine output, mixed venous oxygen saturation, and bedside echocardiography as indispensable monitoring tools in the PICU. Knowledge of residual postoperative lesions—including pulmonary regurgitation, residual right ventricular outflow tract obstruction (RVOTO), residual ventricular septal defect (VSD), or atrial septal defect (ASD)—is fundamental for defining individualized oxygen saturation targets and guiding therapeutic decisions [4]. Among postoperative arrhythmias, junctional ectopic tachycardia (JET) is the most common complication, occurring in up to 15% of patients and being associated with increased postoperative morbidity and mortality [4]. Right ventricular diastolic dysfunction affects nearly half of all patients and is a major contributor to low cardiac output syndrome (LCOS), which occurs in approximately 25% of cases. High-dose milrinone prophylaxis has been shown to reduce the incidence of LCOS by 64% compared with placebo [4]. Long-term follow-up has increasingly focused

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on residual pulmonary regurgitation, which is nearly universal after transannular patch repair. Bokma et al. (JACC, 2023) demonstrated that early pulmonary valve replacement is associated with a significantly lower risk of death and ventricular arrhythmias than delayed intervention, reinforcing the importance of longitudinal surveillance by specialized nursing teams [12]. Tran et al. [8] further emphasize that the quality of preoperative stabilization directly influences postoperative outcomes, suggesting continuity between the two phases of care that nurses are uniquely positioned to coordinate. Similarly, Cooper et al. (Pediatrics, 2022) reported that significant residual hemodynamic lesions following surgery are associated with increased morbidity, prolonged intensive care unit stay, and poorer neurodevelopmental outcomes [18], further supporting the critical role of nursing monitoring throughout both the preoperative and postoperative periods.

Methodological Approaches to Nursing Care

A major distinction among the included studies lies in the methodological approach adopted for planning and documenting nursing care. Araújo et al. [7] were the only authors to explicitly describe the systematic implementation of the nursing process using the standardized NANDA-I, Nursing Interventions Classification (NIC), and Nursing Outcomes Classification (NOC) taxonomies. The identified nursing problems—particularly activity intolerance related to the imbalance between oxygen supply and demand—closely reflect the pathophysiological mechanisms of ToF described by the remaining studies, suggesting that activity intolerance represents a common nursing concern across all stages of the care continuum. Comparison of the different approaches highlights an unresolved challenge within international nursing practice: balancing the benefits of standardized nursing taxonomies—which promote consistency, documentation, and standardized communication—with the flexibility required to address the complexity of high-acuity clinical settings. Standardization of nursing care for patients with ToF through the widespread adoption of validated nursing classification systems should therefore be regarded as a priority for the future development of specialist nursing practice.

Family Support and Therapeutic Communication

Family support and therapeutic communication emerged consistently across all four studies as essential components of high-quality nursing care for patients with ToF. Tran et al. [8] identify NICU nurses as key facilitators of bidirectional communication between the medical team and the family. Forman et al. [4] advocate the Family-Centered Care model as the recommended approach within pediatric cardiac intensive care units. Araújo et al. [7] demonstrate that therapeutic communication assumes a different role in adult patients, incorporating educational interventions aimed at improving autonomy and self-management. Edwards et al. [9] introduce the additional dimension of cultural competence, illustrating how caring for migrant patients requires nurses to obtain reliable clinical histories despite linguistic and cultural barriers. These findings are supported by recent literature. Ferentzi et al. (Frontiers in Pediatrics, 2021) demonstrated that the quality of Family-Centered Care in pediatric cardiac intensive care units is significantly associated with parental well-being and their ability to actively participate in their child's care [19]. Likewise, Lisanti et al. (Nursing in Critical Care, 2024) reported a direct association between parents' perceptions of family-centered care and reduced post-traumatic stress symptoms during the first weeks following their infant's admission to a cardiac intensive care unit [20]. Similarly, the integrative review by Cavalcanti et al.

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(Revista Brasileira de Enfermagem, 2023) on nursing guidance for caregivers of children with congenital heart disease after hospital discharge emphasized that healthcare professionals should establish trusting relationships with parents, provide continuous support, and ensure clear, consistent communication throughout the care pathway [21]. The convergence of these principles across markedly different geographical and clinical settings—including Brazil, neonatal intensive care units in the United States, pediatric cardiac intensive care, and perianesthesia care—demonstrates that family support and therapeutic communication are universally recognized as fundamental elements of high-quality nursing care for patients with ToF.

Adults with Tetralogy of Fallot

One of the most significant findings emerging from this review is the growing recognition of adults with ToF as an increasingly important population within specialist nursing practice. Three distinct adult patient profiles are represented in the included studies: adults with repaired ToF and long-term residual lesions [4]; adults with known but unrepaired ToF presenting with advanced chronic disease characterized by severe polycythemia, chronic cyanosis, and neurocognitive impairment [7]; and adults with previously undiagnosed ToF originating from countries without neonatal screening programs [9]. Regarding the first group, current evidence indicates that 30-year survival exceeds 95% among patients who undergo early surgical repair [11]. Nevertheless, residual lesions—particularly progressive pulmonary regurgitation—remain major contributors to reduced functional capacity, late arrhythmias, and right ventricular failure. Khairy et al. (Journal of the American College of Cardiology, 2010) documented a continuous decline in mortality among patients with congenital heart disease, largely attributable to improvements in specialized follow-up; however, long-term survival remains lower than that observed in the general population [22]. For the latter two groups—adults with untreated or previously undiagnosed ToF—the Global Burden and Diagnostic Challenges Review (2025) demonstrated that 47–63% of congenital heart diseases in low- and middle-income countries are diagnosed only after irreversible complications have developed, with congestive heart failure already present at initial evaluation in 49.4% of cases [15]. These findings underscore that advanced nursing competence in recognizing the clinical manifestations of previously undiagnosed congenital heart disease—as illustrated by Edwards et al. [9]—should not be regarded as an exceptional circumstance but rather as an increasingly necessary competency in healthcare systems serving highly mobile and multicultural populations. Finally, Marelli et al. (Circulation, 2014) estimated that the lifetime prevalence of congenital heart disease reached 6.1 per 1,000 individuals by 2010, representing an almost 30% increase compared with 2000 [13]. This trend is expected to continue, posing increasingly important challenges for specialist nursing education, workforce development, and clinical practice.

CONCLUSIONS

This systematic review analyzed and synthesized the available scientific evidence on nursing care for patients with tetralogy of Fallot (ToF), identifying four fundamental principles that are consistently applicable across all clinical settings and throughout the entire continuum of care. The first principle is specialized clinical expertise. Comprehensive knowledge of the pathophysiology of ToF, proficiency in advanced hemodynamic monitoring, and the early recognition of complications have a direct and measurable impact on patient outcomes. The findings reported by Forman et al. [4]—

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demonstrating a 64% reduction in the incidence of low cardiac output syndrome through prophylactic milrinone administration managed in the pediatric intensive care setting—illustrate how high-quality specialist nursing care translates into tangible improvements in morbidity and survival. The second principle is an evidence-based, systematic approach to nursing care. The study by Araújo et al. [7] demonstrates that the structured implementation of the nursing process is a practical and effective strategy for identifying priority nursing problems, planning targeted interventions, and objectively evaluating patient outcomes, resulting in measurable clinical improvements even in resource-limited settings. The third principle is the timeliness of assessment and intervention. Tran et al. [8] emphasize that prenatal diagnosis and optimal preoperative stabilization are independent predictors of both surgical and neurodevelopmental outcomes. Similarly, Edwards et al. [9] demonstrate that a comprehensive preoperative assessment performed by a perianesthesia nurse can identify previously undiagnosed congenital heart disease, thereby preventing potentially life-threatening perioperative complications. The fourth principle is family support and patient-centered care. All four included studies recognize psychological support, effective therapeutic communication, and active family involvement as essential components of nursing care for patients with ToF. These findings are further supported by recent evidence on Family-Centered Care in pediatric cardiology, which consistently demonstrates improved parental well-being, greater family engagement, and better overall care experiences when families are actively involved in the care process [19–21]. The inclusion of only four studies—two case reports and two narrative reviews—reflects a structural limitation of the nursing literature specifically addressing ToF. The rarity of the condition, the ethical challenges associated with conducting controlled studies in neonatal populations, and the inherent difficulty of standardizing nursing interventions limit the availability of high-level evidence according to traditional Evidence-Based Medicine hierarchies. Nevertheless, these limitations do not diminish the clinical relevance of the available evidence, which is consistent with the recommendations of major international scientific societies. In conclusion, nurses play a key and irreplaceable role throughout the continuum of care for patients with tetralogy of Fallot, from preoperative neonatal stabilization to lifelong follow-up in adulthood. The rapidly expanding population of adults with repaired congenital heart disease, the increasing number of patients with previously undiagnosed ToF presenting to healthcare systems in high-income countries after migrating from resource-limited settings, and the growing body of evidence documenting the long-term neurodevelopmental impact of congenital heart disease [17] pose new and increasingly complex challenges for the nursing profession. Investment in specialist education, the development of dedicated care pathways for adults with congenital heart disease, the promotion of original nursing research, and the strengthening of cultural competence among nurses working in multicultural healthcare environments should be regarded as essential priorities to ensure safe, equitable, evidence-based, and high-quality care for this complex and continuously growing patient population.

REFERENCES

[1] Van der Linde D, Konings EEM, Slager MA, et al. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. J Am Coll Cardiol. 2011;58(21):2241-2247. doi:10.1016/j.jacc.2011.08.025.

[2] Villafañe J, Feinstein JA, Jenkins KJ, et al. Hot topics in tetralogy of Fallot. J Am Coll Cardiol. 2013;62(23):2155-2166. doi:10.1016/j.jacc.2013.07.100

[3] Lillehei CW, Cohen M, Warden HE, et al. Direct vision intracardiac surgical correction of the tetralogy of Fallot, pentalogy of Fallot, and pulmonary atresia defects. Ann Surg. 1955;142(3):418-445.

[4] Forman J, Beech R, Slugantz L, Donnellan A. A review of tetralogy of Fallot and postoperative management. Crit Care Nurs Clin North Am. 2019;31(3):315-328. doi:10.1016/j.cnc.2019.05.003.

[5] Baumgartner H, De Backer J, Babu-Narayan SV, et al. 2020 ESC Guidelines for the management of adult congenital heart disease. Eur Heart J. 2021;42(6):563-645.

[6] Bhatt AB, Foster E, Kuehl K, et al. Congenital heart disease in the older adult: a scientific statement from the American Heart Association. Circulation. 2015;131(21):1884-1931.

[7] Araújo JNM, Gomes ATL, Medeiros Costa RA, Fernandes APNL, Santos VEP, Vitor AF. Proceso de enfermería aplicado al paciente con tetralogía de Fallot [Nursing process applied to the patient with tetralogy of Fallot]. Cult Cuid. 2017;21(47):165-174. doi:10.7184/cuid.2017.47.14. (in Spanish).

[8] Tran NN, Tran M, Lemus RE, et al. Preoperative care of neonates with congenital heart disease. Neonatal Netw. 2022;41(4):200-210. doi:10.1891/NN-2021-0028.

[9] Edwards S, Bochenek JM, Taylor T, McGhee S. The crucial role of the perianesthesia nurse in preadmission assessment with unexpected findings: a case report. J Perianesth Nurs. 2025;40(6):1427-1430. doi:10.1016/j.jopan.2025.03.008.

[10] Marelli AJ, Ionescu-Ittu R, Mackie AS, et al. Lifetime prevalence of congenital heart disease in the general population from 2000 to 2010. Circulation. 2014;130(9):749-756. doi:10.1161/CIRCULATIONAHA.113.008396.

[11] Moons P, Daelman B, Marelli A. The aging patient with tetralogy of Fallot: out of the blue and into the pink. CJC Pediatr Congenit Heart Dis. 2023;2(6):335-338. doi:10.1016/j.cjcpc.2023.08.004.

[12] Bokma JP, Geva T, Sleeper LA, et al. Improved outcomes after pulmonary valve replacement in repaired tetralogy of Fallot. J Am Coll Cardiol. 2023;81(21):2075-2085. doi:10.1016/j.jacc.2023.02.052.

[13] Marelli AJ, Mackie AS, Ionescu-Ittu R, Rahme E, Pilote L. Congenital heart disease in the general population: changing prevalence and age distribution. Circulation. 2007;115(2):163-172. doi:10.1161/CIRCULATIONAHA.106.627224.

[14] Butler SC, Rofeberg V, Smith-Parrish M, et al. Individualized family-centered developmental care: an essential model to address the unique needs of infants with congenital heart disease. Front Pediatr. 2024;12:1384615. doi:10.3389/fped.2024.1384615.

[15] Al-Shawki Y. The global burden and diagnostic challenges of undiagnosed congenital heart disease in resource-limited settings: a systematic review. Res Rep Clin Cardiol. 2026;17:1-20. doi:10.2147/RRCC.S558007.

[16] Agyemang C, Bhopal R, Boon NA, et al. International migr15ation and cardiovascular health: unraveling the disease burden among migrants to North America and Europe. J Am Heart Assoc. 2024;13(9):e030228. doi:10.1161/JAHA.123.030228.

[17] Ortinau CM, Smyser CD, Arthur L, Gordon EE, Heydarian HC, Wolovits J, Nedrelow J, Marino BS, Levy VY. Optimizing Neurodevelopmental Outcomes in Neonates With Congenital Heart Disease. Pediatrics. 2022 Nov 1;150(Suppl

Igiene e Sanità Pubblica 2026; 99 (4)

2):e2022056415L. doi: 10.1542/peds.2022-056415L. PMID: 36317967; PMCID: PMC10435013.

[18] Cooper DS, Hill KD, Krishnamurthy G, Sen S, Costello JM, Lehenbauer D, Twite M, James L, Mah KE, Taylor C, McBride ME. Acute Cardiac Care for Neonatal Heart Disease. Pediatrics. 2022 Nov 1;150(Suppl 2):e2022056415J. doi: 10.1542/peds.2022-056415J. PMID: 36317971.

[19] Ferentzi H, Rippe RCA, Latour JM, et al. Family-centered care at pediatric cardiac intensive care units in Germany and the relationship with parent and infant well-being: a study protocol. Front Pediatr. 2021;9:666904. doi:10.3389/fped.2021.666904.

[20] Lisanti AJ, Min J, Golfenshtein N, et al. Perceived family-centered care and post-traumatic stress in parents of infants cared for in the paediatric cardiac intensive care unit. Nurs Crit Care. 2024;29(5):1059-1066. doi:10.1111/nicc.13094.

[21] Cavalcanti AMTS, Medeiros Lima LH, Santos Silva M, et al. Nursing guidelines for caregivers of children with congenital heart disease after discharge: integrative review. Rev Bras Enferm. 2023;76(6):e20230054. doi:10.1590/0034-7167-2023-0054.

[22] Khairy P, Ionescu-Ittu R, Mackie AS, Abrahamowicz M, Pilote L, Marelli AJ. Changing mortality in congenital heart disease. J Am Coll Cardiol. 2010;56(14):1149-1157. doi:10.1016/j.jacc.2010.03.085.

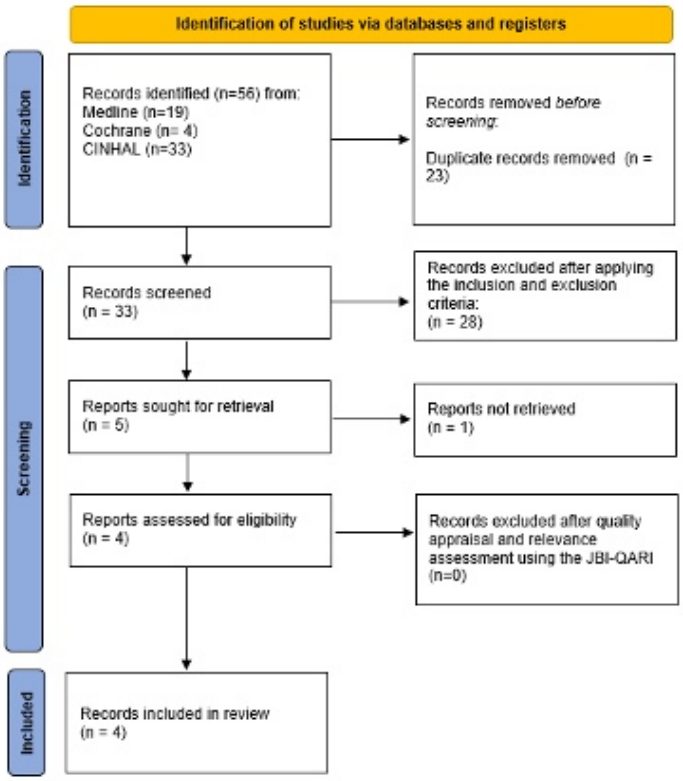

Figure 1. PRISMA flow diagram illustrating the methodology used for the identification, screening, eligibility assessment, and inclusion of studies on nursing care for patients with tetralogy of Fallot.

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Table 1. Literature review summary table

AUTHORS, YEAR, STUDY DESIGN, AND REFERENCE	TITLE	JOURNAL	SETTING
Araújo JNM, Gomes ATL, Medeiros RAC, Fernandes APNL, Santos VEP, Vitor AF 2017 – Case report Cultura de los Cuidados (Ed. digital). 2017;21(47):165-174. doi: 10.7184/cuid.2017.47.14	"Proceso de enfermería aplicado al paciente con tetralogia de Fallot"	Cultura de los Cuidados	Department of Cardiology, University Hospital, Northeastern Brazil
STUDY SUMMARY AND MAIN FINDINGS			
Objective: To describe the nursing process applied to a 23-year-old woman with unrepaired tetralogy of Fallot, admitted to the cardiology department of a Brazilian university hospital in 2012. Population: A 23-year-old woman with a diagnosis of ToF established during childhood, who had never undergone specialist follow-up or surgical correction. On admission, she presented with severe dyspnea on minimal exertion, cardiac arrhythmia, peripheral cyanosis, digital clubbing, marked polycythemia (red blood cell count: 8.39 million/mm³; hemoglobin: 22 g/dL; hematocrit: 66%), thrombocytopenia (142,000/mm³), a grade IV systolic murmur over the pulmonary area, a grade III systolic murmur over the tricuspid area, and cognitive impairment as assessed by the Mini-Mental State Examination (MMSE; score 16/30). Methods: A structured nursing process based on the NANDA-I taxonomy, the Nursing Interventions Classification (NIC), and the Nursing Outcomes Classification (NOC), with daily patient assessment over a 10-day period. Results: Four nursing diagnoses were identified: activity intolerance (priority diagnosis), impaired dentition, decreased cardiac output, and ineffective impulse control. The nursing care plan focused primarily on activity intolerance through interventions including heart rate and cardiac rhythm monitoring, maintenance of a rest-promoting environment, stress reduction, and gradual encouragement of ambulation. The overall NOC score improved from 12 to 15 out of 18. The concentration score reached the expected outcome (4/5) following targeted cognitive exercises, while walking distance and independence in activities of daily living (ADLs) improved to level 3. Hematocrit remained unchanged, reflecting a compensatory physiological response to chronic cyanosis. Conclusions and Nursing Implications: The systematic implementation of the nursing process contributed to improvements in the patient's health status, functional independence, and self-confidence, highlighting the value of individualized nursing care in the management of adults with unrepaired tetralogy of Fallot.			

AUTHORS, YEAR, STUDY DESIGN, AND REFERENCE	TITLE	JOURNAL	SETTING
Tran NN, Tran M, Lemus RE, Woon J, Lopez J, Dang R, Votava-Smith JK 2022 – Narrative Review - Neonatal Network. 2022;41(4):200-210. doi: 10.1891/NN-2021-0028	"Preoperative Care of Neonates With Congenital Heart Disease"	Neonatal Network	Neonatal Intensive Care Unit (NICU), Children's Hospital Los Angeles, USA
STUDY SUMMARY AND MAIN FINDINGS			
Objective: To describe the clinical presentation, nursing implications, and preoperative management of four complex neonatal congenital heart diseases, including tetralogy of Fallot (ToF). Population: Neonates with complex congenital heart disease (hypoplastic left heart syndrome [HLHS], dextro-transposition of the great arteries [D-TGA], tetralogy of Fallot [ToF], and pulmonary atresia with intact ventricular septum [PA/IVS]) admitted to the neonatal intensive care unit (NICU). Approximately 40,000 newborns are diagnosed with congenital heart disease each year in the United States. Preoperative Management of ToF: Oxygen saturation may decrease to 80% or lower. Preoperative nursing care includes continuous pulse oximetry monitoring, administration of prostaglandin E1 (PGE1) infusion in cases of severe pulmonary stenosis or pulmonary atresia to maintain ductal patency, and prompt recognition and management of hypercyanotic ("Tet") spells through the knee–chest position, supplemental oxygen, and sedation. Infants with mild-to-moderate disease are managed on an outpatient basis with regular clinical follow-up until elective surgical repair, typically performed between 3 and 6 months of age. Results: Prenatal diagnosis plays a crucial role in reducing preoperative morbidity and mortality by enabling timely care planning and neonatal stabilization in specialized cardiac centers before surgery. Infants with congenital heart disease undergoing cardiac surgery during the first year of life are at increased risk of neurodevelopmental impairment—including cognitive, motor, and language delays—which may already be detectable during the preschool years and are largely attributable to reduced cerebral perfusion during the perioperative period. Conclusions and Nursing Implications: NICU nurses play a pivotal role in facilitating effective communication between the multidisciplinary team and the family, performing continuous and systematic clinical assessment—including capillary refill, skin temperature, oxygen saturation, heart rate and rhythm, and neurological status—and supporting preoperative hemodynamic stabilization. Comprehensive knowledge of the anatomy and pathophysiology of congenital heart disease, including tetralogy of Fallot, is essential for delivering safe, evidence-based care, anticipating complications, and improving both neurological and long-term clinical outcomes.			

AUTHORS, YEAR, STUDY DESIGN, AND REFERENCE	TITLE	JOURNAL	SETTING
Edwards S, Bochenek JM, Taylor T, McGhee S 2025 – Case report Journal of PeriAnesthesia Nursing. 2025;40:1427-1430. doi: 10.1016/j.jopan.2025.03.008	"The Crucial Role of the Perianesthesia Nurse in Preadmission Assessment With Unexpected Findings: A Case Report"	Journal of PeriAnesthesia Nursing	Preoperative Assessment Center, The Ohio State University, USA
STUDY SUMMARY AND MAIN FINDINGS			
Objective: To illustrate the role of the perianesthesia nurse in identifying previously undiagnosed tetralogy of Fallot during a routine preoperative assessment. Case Presentation: A 25-year-old male migrant worker from Nicaragua presented to a preoperative assessment center one week before scheduled right meniscal repair surgery. He reported chronic fatigue, a 6-day history of burning chest pain, dyspnea on moderate exertion, and symptom relief by assuming a posture involving compression of the chest, analogous to the squatting position classically associated with hypercyanotic ("Tet") spells. He had no documented medical history and no regular access to healthcare, having originated from a country without standardized neonatal screening. Clinical Findings: Physical examination revealed digital clubbing with pallor, capillary refill time >3 seconds, oxygen saturation (SpO_2) of 90%, bilateral blood pressure of 148/90 mmHg with elevated jugular venous pressure, a left parasternal systolic murmur, and an irregular respiratory rate of 26 breaths/min. Laboratory investigations showed a troponin T level of 0.12 ng/mL and a PaO_2 of 70 mmHg. Electrocardiography demonstrated right ventricular hypertrophy with right bundle branch block, while chest radiography revealed the characteristic boot-shaped heart. The diagnosis of previously unrecognized tetralogy of Fallot was subsequently confirmed by transthoracic echocardiography and cardiac magnetic resonance imaging. Results: The perianesthesia nurse promptly notified the orthopedic surgeon, leading to cancellation of the planned procedure and immediate referral for cardiology consultation. The patient was started on antihypertensive and diuretic therapy and scheduled for cardiology follow-up. This case highlights that tetralogy of Fallot may remain undiagnosed until adulthood in individuals from countries lacking comprehensive neonatal screening programs and underscores the vulnerability of migrant populations with fragmented access to healthcare. Conclusions and Nursing Implications: Critical thinking, advanced clinical assessment skills, and cultural competence are essential competencies for perianesthesia nurses to identify previously undiagnosed serious cardiac conditions and prevent potentially life-threatening perioperative and anesthetic complications.			

AUTHORS, YEAR, STUDY DESIGN, AND REFERENCE	TITLE	JOURNAL	SETTING
Forman J, Beech R, Slugantz L, Donnellan A 2019 – Narrative Review Critical Care Nursing Clinics of North America. 2019;31:315-328. doi: 10.1016/j.cnc.2019.05.003	"A Review of Tetralogy of Fallot and Postoperative Management"	Critical Care Nursing Clinics of North America	PICU / Cardiac ICU, Kentucky Children's Hospital e Cincinnati Children's Hospital, USA
STUDY SUMMARY AND MAIN FINDINGS			
Objective: To provide a comprehensive overview of tetralogy of Fallot (ToF), encompassing its pathophysiology, anatomical variants, surgical indications, and postoperative nursing management in the pediatric intensive care unit (PICU). Epidemiology and Pathophysiology: Tetralogy of Fallot accounts for 7–10% of all congenital heart diseases, with an incidence of approximately 1 in 3,500 live births (approximately 1,660 newborns annually in the United States). Early mortality has declined from the historical rate of 25% to approximately 2%, with survival into adulthood now exceeding 85%. The condition consists of four structural cardiac defects associated with a variable spectrum of right ventricular outflow tract obstruction (RVOTO), ranging from acyanotic patients ("pink Tets") to critically cyanotic neonates with pulmonary atresia who are dependent on a patent ductus arteriosus (PDA) for pulmonary blood flow. Approximately 25% of patients have associated chromosomal abnormalities, including 22q11.2 deletion syndrome and trisomy 21. Surgical Management: Complete surgical repair includes transatrial closure of the ventricular septal defect (VSD), infundibular muscle resection, and placement of a transannular patch in patients with an inadequate pulmonary valve annulus (z-score < −2 SD). Approximately 25% of neonates require early palliative intervention with a modified Blalock–Taussig shunt or ductal artery stenting before definitive repair. Thirty-day postoperative mortality approaches zero in specialized cardiac centers. Postoperative Complications and Nursing Management: Right bundle branch block (RBBB) is nearly universal after repair and is generally asymptomatic. Junctional ectopic tachycardia (JET) occurs in approximately 15% of cases and is managed through normothermia, correction of electrolyte imbalances, reduction of catecholamine administration, temporary atrial pacing, and pharmacological therapy with dexmedetomidine or amiodarone. Complete atrioventricular block (AV block) occurs in 1–3% of patients and requires permanent pacemaker implantation if it persists beyond two weeks. Right ventricular diastolic dysfunction affects nearly 50% of patients and is managed with early extubation, minimization of positive-pressure ventilation, and milrinone therapy. Low cardiac output syndrome (LCOS) develops in approximately 25% of patients and can be prevented by high-dose milrinone, which has been shown to reduce its incidence by 64% compared with placebo. Conclusions and Nursing Implications: Continuous, systematic monitoring in the PICU includes assessment of heart rate and rhythm through temporary epicardial pacing wires, central venous pressure, peripheral perfusion, urine output, mixed venous oxygen saturation, and bedside echocardiographic evaluation. Knowledge of residual postoperative lesions—including pulmonary regurgitation, residual RVOTO, residual ventricular septal defect (VSD), or atrial septal defect (ASD)—guides individualized oxygen saturation targets and therapeutic decision-making. The integration of advanced nursing expertise, comprehensive hemodynamic monitoring, and multidisciplinary collaboration represents the cornerstone of postoperative management in patients with tetralogy of Fallot, with a direct impact on short-, medium-, and long-term clinical outcomes.			