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Postoperative Pulmonary Hypertension After Complete Tetralogy of Fallot Repair: Mechanistic Determinants and Prognostic Implications for Early Clinical Outcomes

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ABSTRACT

Introduction: Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease, with an incidence of approximately three per 10,000 live births. Postoperative pulmonary hypertension following complete repair is a rare but life-threatening complication that may lead to weaning failure from cardiopulmonary bypass. This condition is driven by acute increases in pulmonary vascular resistance and impaired right ventricular–pulmonary arterial coupling. Recognized risk factors include chronic preoperative hypoxia, younger age at surgery, and severe right ventricular dysfunction.

Case Description: A three-year-old child with stunted growth and longstanding cyanosis since infancy underwent evaluation, revealing a large malaligned ventricular septal defect, 50% overriding aorta, and severe pulmonary stenosis with a pressure gradient of 85 mmHg. Intraoperative findings confirmed a double-outlet right ventricle with multiple atrial septal defects. Following total correction, the patient was not weaned from cardiopulmonary bypass because of acute right ventricular failure and severe pulmonary hypertension. Atrial septal defect creation for decompression and maximal inotropic support with dobutamine, adrenaline, and milrinone were performed. Despite aggressive management, the patient progressed to refractory cardiogenic shock with an arterial pressure of 25/19 mmHg and died within 24 hours postoperatively.

Conclusion: This case underscores the catastrophic impact of postoperative pulmonary hypertensive crises in patients with complex congenital heart disease. The failure of weaning from cardiopulmonary bypass due to acute right ventricular failure represents a critical inflection point associated with extremely high mortality. Early risk stratification, perioperative optimization, and timely consideration of advanced mechanical circulatory support are essential. The integration of targeted pulmonary vasodilator therapy with vigilant hemodynamic monitoring may improve right ventricular adaptation and clinical outcomes in high-risk patients.

Keywords

Tetralogy of Fallot, Pulmonary Hypertension, Cardiopulmonary Bypass, Cardiogenic Shock, Right Ventricular Failure, Congenital Heart Disease

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INTRODUCTION

Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease, with an estimated incidence of approximately three per 10,000 live births, accounting for 7–10% of all congenital cardiac malformations worldwide [1,2]. In developing countries, including Indonesia, TOF continues to represent a substantial proportion of pediatric congenital heart diseases. Advances in surgical techniques and perioperative management have markedly reduced operative mortality to < 3% in the modern era; however, postoperative

complications remain a major determinant of early morbidity and long-term clinical outcomes [3]. Postoperative pulmonary hypertension following complete TOF repair is an uncommon but potentially catastrophic complication, particularly in patients who fail to wean from cardiopulmonary bypass (CPB). Prolonged CPB duration has consistently been identified as a key predictor of adverse postoperative events, with cardiac complications occurring in up to one-quarter of patients undergoing complete repair. The underlying pathophysiology is complex, involving acute elevation of pulmonary vascular resistance, endothelial dysfunction, and disruption of right ventricular–pulmonary arterial coupling, ultimately leading to profound hemodynamic instability [4].

Several perioperative factors contribute to the development of postoperative pulmonary hypertension, including chronic preoperative hypoxemia, delayed surgical correction, and severe right ventricular hypertrophy or dysfunction [5]. Patients with preoperative oxygen saturation levels of < 90% are particularly susceptible to prolonged mechanical ventilation and pulmonary complications. Furthermore, the CPB-induced systemic inflammatory response and ischemia–reperfusion injury exacerbate pulmonary vascular reactivity and impair cardiopulmonary function, further increasing the risk of perioperative deterioration. Failure to wean patients from CPB following TOF repair is most frequently associated with acute right ventricular failure driven by an abrupt mismatch between pulmonary and systemic vascular resistance [6]. This condition represents a critical perioperative inflection point and often necessitates rapid escalation of support, including vasoactive therapy, targeted pulmonary vasodilators, and mechanical circulatory support in selected cases. Despite these interventions, outcomes remain poor once severe right ventricular dysfunction is established. Therefore, a comprehensive understanding of the mechanisms, risk stratification, and management strategies of postoperative pulmonary hypertension is essential for improving clinical outcomes. This case report aims to highlight the clinical course and perioperative challenges associated with pulmonary hypertensive crisis following complete TOF repair, emphasizing its prognostic implications and the importance of early, targeted intervention [7].

CASE DESCRIPTION

A three-year-old boy with a history of cyanotic congenital heart disease was admitted for elective complete repair of Tetralogy of Fallot. The patient had experienced persistent cyanosis since the age of three months, accompanied by failure to thrive and recurrent upper respiratory tract infections occurring approximately every two months. Anthropometric assessment revealed severe stunting, with a body weight of 10 kg and height of 78 cm (weight-for-age z-score −3.24 SD; height-for-age z-score −5.19 SD). On admission, the patient was conscious but irritable, with a heart rate of 128 beats per minute, respiratory rate of 25 breaths per minute, temperature of 36.5°C, and oxygen saturation of 79–80% on room air. Physical examination revealed digital clubbing, systolic cardiac murmur, and dysmorphic facial features suggestive of Down syndrome. Pulmonary auscultation revealed vesicular breath sounds without additional adventitious sounds (Figure 1).

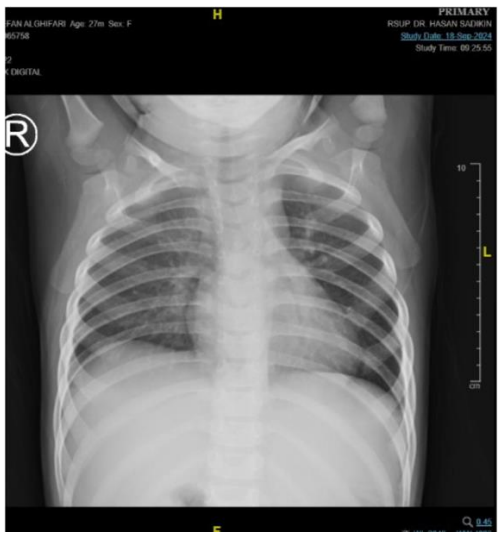

Figure 1. Chest Radiograph Demonstrating Features Consistent with Cyanotic Congenital Heart Disease

Transthoracic echocardiography revealed a large malaligned ventricular septal defect (10 mm) with bidirectional shunting, approximately 50% aortic override, and severe multilevel pulmonary stenosis involving the infundibular, valvular, and supravalvular segments, with a peak pressure gradient of 85 mmHg. Intraoperative transesophageal echocardiography revealed a more complex anatomy, consistent with a double outlet right ventricle, a large ventricular septal defect (15 mm), and multiple atrial septal defects measuring up to 2.5 cm (Figure 2).

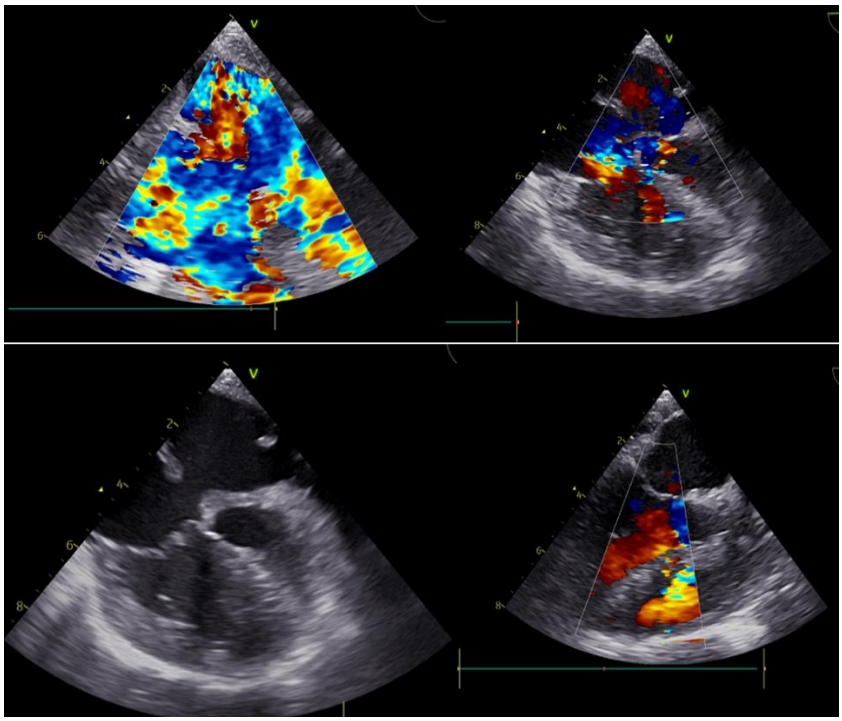

Figure 2. Intraoperative Transesophageal Echocardiography Showing Double Outlet Right Ventricle with Ventricular Septal Defect and Multiple Atrial Septal Defects

The pulmonary valve was bicuspid with significant valvular stenosis. Complete repair was performed over 8 h, with a cardiopulmonary bypass duration of 49 min and an aortic cross-clamp time of 32 min. During the initial attempt to wean from cardiopulmonary bypass, the patient developed acute hemodynamic instability, characterized by cardiac distension, suggestive of severe right ventricular failure and pulmonary hypertension. Hemodynamic measurements demonstrated a marked increase in pulmonary artery pressure following repair (RVOT: 50/38 mmHg; MPA: 48/31 mmHg), indicating acute pulmonary vascular overload. The patient required reinstitution of cardiopulmonary bypass and surgical creation of an atrial septal defect to facilitate right-sided decompression. The patient was transferred to the cardiac intensive care unit in a critically unstable condition, with profound hypotension, bradycardia, absent spontaneous respiration, and signs of systemic hypoperfusion. Despite maximal inotropic support with dobutamine (10 mcg/kg/min), adrenaline (0.3 mcg/kg/min), and milrinone (0.375 mcg/kg/min), adequate cardiac output could not be maintained. The patient rapidly deteriorated into refractory cardiogenic shock, with a blood pressure of 25/19 mmHg, heart rate of 58 beats per minute, and unmeasurable oxygen saturation. Mechanical ventilation was initiated using a pressure-controlled assist-control mode (PEEP 6 cmH_2O, PIP 25 cmH_2O, FiO_2 100%). However, progressive hemodynamic collapse ensued, culminating in cardiac arrest. Advanced cardiopulmonary resuscitation, including repeated administration of epinephrine, failed to achieve a sustained return of spontaneous circulation. Neurological assessment revealed fixed dilated pupils with absent light reflexes, consistent with brainstem death. The patient died within 24 h of the operation. The critical turning point occurred during the initial weaning from cardiopulmonary bypass, when an acute pulmonary hypertensive crisis precipitated right ventricular failure and irreversible circulatory collapse, despite aggressive surgical and pharmacological interventions.

DISCUSSION

Postoperative pulmonary hypertensive crisis represents a life-threatening complication following complete repair of Tetralogy of Fallot, characterized by an abrupt increase in pulmonary vascular resistance, acute right ventricular failure, and severe hemodynamic instability [8,9]. In the present case, failure to wean from cardiopulmonary bypass (CPB) marked a critical inflection point, reflecting a profound mismatch between right ventricular contractility and pulmonary vascular load, ultimately leading to refractory cardiogenic shock. Effective perioperative management requires a comprehensive multidisciplinary strategy encompassing preoperative optimization, intraoperative stabilization, and intensive postoperative care. Preoperative pharmacological conditioning with endothelin receptor antagonists and phosphodiesterase type 5 inhibitors has been shown to reduce pulmonary vascular resistance and improve right ventricular performance, particularly in patients with chronic hypoxemia [10]. These strategies may be crucial in high-risk populations undergoing complex congenital cardiac repair. Intraoperatively, prevention of pulmonary hypertensive crises focuses on minimizing triggers such as hypoxia, hypercarbia, acidosis, and inadequate anesthetic depth. Selective pulmonary vasodilation using inhaled nitric oxide or milrinone has demonstrated efficacy in reducing pulmonary artery pressure without inducing systemic hypotension [11,12]. In this case, the marked increase in pulmonary artery pressure following repair suggested an exaggerated pulmonary vascular response that exceeded the compensatory capacity of the right ventricle.

When a pulmonary hypertensive crisis occurs, rapid escalation of therapy is essential. Combined inotropic support using dobutamine and milrinone enhances myocardial contractility while providing pulmonary vasodilation, whereas epinephrine serves as a rescue therapy in refractory cardiogenic shock [13-21]. Despite maximal triple inotropic therapy, the patient progressed to irreversible circulatory collapse, indicating severe right ventricular failure that was unresponsive to pharmacological intervention. Adjunctive surgical and interventional strategies, including atrial septal defect creation or balloon atrial septostomy, may provide right ventricular decompression and temporary hemodynamic stabilization [15,16]. In the present case, surgical atrial septal defect creation was performed; however, it failed to reverse progressive hemodynamic deterioration, underscoring the severity of the underlying pathophysiological derangement. Advanced hemodynamic monitoring plays a pivotal role in guiding therapy and assessing responses. Continuous assessment of arterial pressure, central venous pressure, lactate levels, and mixed venous oxygen saturation provides critical insights into tissue perfusion and circulatory adequacy [22,23]. Persistent elevation of pulmonary artery pressure combined with profound systemic hypotension in this patient reflected refractory cardiogenic shock with inadequate systemic oxygen delivery.

Emerging evidence highlights the role of advanced monitoring techniques and predictive hemodynamic analytics in identifying early deterioration following pediatric cardiac surgery [24-26]. In addition, comparative studies suggest that milrinone-based strategies may offer advantages in reducing pulmonary artery pressure and improving right ventricular performance compared with conventional inotropic regimens. Importantly, this case demonstrates that once acute right ventricular failure is established in the setting of severe pulmonary hypertension, outcomes remain poor despite aggressive intervention. Early identification of high-risk patients, proactive perioperative pulmonary vasodilator therapy, and timely consideration of mechanical circulatory support such as extracorporeal membrane oxygenation may represent critical strategies to improve survival in similar clinical scenarios [9].

CONCLUSION

This case highlights the high mortality associated with postoperative pulmonary hypertensive crises in patients with complex congenital heart disease who undergo complete repair of Tetralogy of Fallot. Failure to wean from cardiopulmonary bypass due to acute pulmonary hypertension and right ventricular failure represents a critical prognostic inflection point. Despite aggressive surgical and pharmacological interventions, the patient progressed to refractory cardiogenic shock. These findings underscore the need for early risk stratification, meticulous perioperative optimization, and timely consideration of advanced mechanical circulatory support in high-risk patients.

DECLARATIONS
None

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AUTHORS' CONTRIBUTIONS
E.D. was responsible for patient management, data acquisition, and drafting the manuscript. D.T. contributed to clinical supervision, conceptual guidance, and critical revision of the manuscript for important intellectual content. Both authors reviewed and approved the final version of the manuscript and agreed to be accountable for all aspects of this work.

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