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# TETRALOGY OF FALLOT ASSOCIATED WITH A RESTRICTIVE VENTRICULAR SEPTAL DEFECT IN AN INFANT: A CASE REPORT

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## ABSTRACT

***Background:*** Tetralogy of Fallot (TOF) is one of the most common cyanotic congenital heart defects. In most patients the ventricular septal defect (VSD) is large and non-restrictive, allowing equalization of ventricular pressures. TOF associated with a restrictive VSD is a rare variant in which the interventricular pressure gradient elevates right ventricular pressure, altering the hemodynamic profile and complicating both diagnosis and surgical management.

***Case presentation:*** A 3-month-old female infant (4.8 kg) presented with progressive cyanosis and oxygen saturation of 70–72% in room air. Echocardiography and cardiac computed tomography demonstrated TOF with a restrictive perimembranous VSD (approximately 3 mm; interventricular gradient 52 mmHg) and severe right ventricular outflow tract (RVOT) obstruction (peak gradient 77 mmHg). Complete repair was performed through a minimally invasive lower partial sternotomy. The early postoperative course was complicated by right ventricular dysfunction with myocardial edema and fluid overload, requiring delayed sternal closure, multiple inotropes, and peritoneal dialysis. The patient recovered gradually; at 3-month follow-up she was thriving, with good biventricular function, no residual shunt, and only mild residual RVOT obstruction.

***Conclusion:*** TOF with a restrictive VSD is a rare variant whose distinct hemodynamics predispose to postoperative right ventricular dysfunction. Accurate preoperative diagnosis, an individualized surgical strategy, and anticipatory management of right heart failure are essential for a favorable outcome, even in young infants.

**Keywords:** *Tetralogy of Fallot; Restrictive ventricular septal defect; Infant; Right ventricular dysfunction; Minimally invasive cardiac surgery.*

## 1. INTRODUCTION

Tetralogy of Fallot (TOF) is one of the most common cyanotic congenital heart defects and is characterized by a ventricular septal defect, overriding aorta, right ventricular outflow tract obstruction, and right ventricular hypertrophy [6,7,8]. In most cases, the VSD is large and non-restrictive, resulting in equalization of right and left ventricular pressures [1,2]. However, TOF associated with a restrictive VSD is a rare anatomical variant that substantially alters the hemodynamic profile and clinical presentation of the disease. The presence of a restrictive VSD may lead to increased right ventricular pressure, resulting in cyanosis that is disproportionate to the degree of RVOT obstruction and complicating preoperative assessment.

Complete surgical repair remains the definitive treatment for TOF and generally provides excellent long-term outcomes [9,11]. However, in patients with a restrictive VSD, surgical planning requires special consideration because of the increased risk of postoperative hemodynamic instability and right ventricular dysfunction. Furthermore, reports describing this rare entity remain limited, particularly in infants.

## 2. CASE PRESENTATION

A 3-month-old female infant weighing 4.8 kg was admitted because of progressive cyanosis. She was the first child of a mother who underwent cesarean delivery at 40 weeks of gestation. Birth weight was 2.8 kg. Congenital heart disease had been detected prenatally at 26 weeks of gestation, and the patient had been followed regularly at the Cardiovascular Center, E Hospital.

Physical examination revealed moderate cyanosis involving the lips and extremities, with oxygen saturation ranging from 70% to 72% in room air. A grade 3/6 systolic murmur was audible along the left sternal border. Respiratory rate was 45 breaths per minute, heart rate was 140 beats per minute, and feeding was poor.

Transthoracic echocardiography demonstrated TOF with a perimembranous VSD extending beneath the aortic valve. The VSD was markedly restrictive, measuring approximately 3mm, with complete right-to-left shunting. The aorta overrode approximately 50% of the ventricular septum. Severe RVOT obstruction was present, with a peak pressure gradient of 77mmHg, accompanied by right ventricular hypertrophy. Doppler interrogation across the VSD demonstrated a peak pressure gradient of 52mmHg, confirming its restrictive nature. The pulmonary valve annulus measured 5.5mm (Z-score −3.2), while the left and right pulmonary arteries measured 4.2 mm and 4.8 mm, respectively. Biventricular systolic function was preserved, with a left ventricular ejection fraction of 81%.

Cardiac computed tomography confirmed the diagnosis of TOF with a perimembranous VSD measuring approximately 3.6mm, an overriding aorta, and severe RVOT narrowing, with the narrowest diameter measuring approximately 1.6 mm. The main pulmonary artery was hypoplastic, measuring 5.6mm in diameter. The pulmonary arteries remained confluent, with diameters of 4.5mm and 6.6mm for the right and left branches, respectively. No patent ductus arteriosus or major aortopulmonary collateral arteries were identified. The aortic arch was left-sided without coarctation. Right ventricular hypertrophy was present, and the coronary arteries appeared normal. An anomalous retroaortic left brachiocephalic vein draining into the superior vena cava was also noted.

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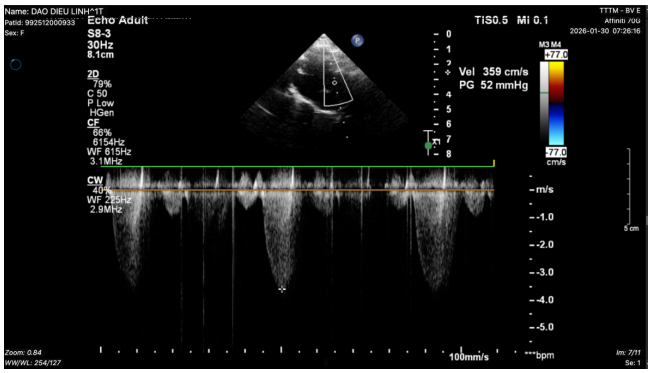


**Figure 1. Preoperative Doppler echocardiography demonstrating a restrictive ventricular septal defect with a peak pressure gradient of 52 mmHg**

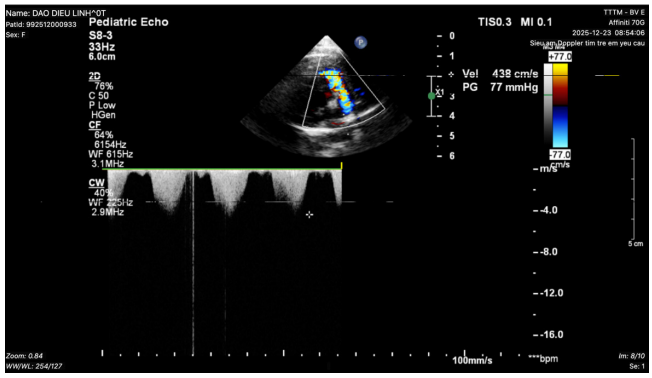


**Figure 2. Preoperative Doppler echocardiography showing severe right ventricular outflow tract obstruction with a peak velocity of 4.38 m/s and a peak pressure gradient of 77 mmHg**

The patient was diagnosed with TOF associated with a restrictive VSD and was scheduled for complete surgical repair. The procedure was performed through a minimally invasive lower partial sternotomy using cardiopulmonary bypass. Following institution of cardiopulmonary bypass and aortic cross-clamping, myocardial protection was achieved with Custodiol cardioplegia.

A right atriotomy was performed. Intraoperative inspection revealed a 3-mm perimembranous VSD that was largely covered by tricuspid valve tissue, producing a significant interventricular pressure gradient. The right ventricular outflow tract was severely obstructed, and the pulmonary valve was bicuspid with mild commissural fusion.

The VSD was closed with a bovine pericardial patch using continuous 6-0 polypropylene sutures. A valve-sparing repair was undertaken: The fused pulmonary commissures were incised (commissurotomy), and the outflow tract was enlarged through combined transatrial and transpulmonary approaches, working across the tricuspid and pulmonary valves. After enlargement, the pulmonary valve annulus freely admitted a 7–8 mm Hegar dilator, and both branch pulmonary arteries admitted a 5–6 mm dilator. The main pulmonary artery was augmented with an autologous pericardial patch, and the pulmonary annulus was preserved. Intraoperative echocardiography demonstrated satisfactory relief of RVOT obstruction and complete closure of the VSD without residual shunting.

Cardiopulmonary bypass and aortic cross-clamp times were 116 minutes and 87 minutes, respectively. Intraoperative transesophageal echocardiography confirmed complete VSD closure, no residual shunt, and adequate RVOT enlargement with a residual peak gradient of approximately 35 mmHg. Mild residual pulmonary stenosis and mild pulmonary regurgitation were present. Left ventricular systolic function remained normal, and right ventricular contractility was preserved. No pericardial or pleural effusion was identified.

The sternum was initially closed at the completion of surgery because hemodynamics were stable. However, during the early postoperative period, the patient developed significant myocardial edema and fluid overload associated with hemodynamic instability. Therefore, delayed sternal closure was performed to reduce cardiac compression and facilitate hemodynamic recovery. Intensive postoperative

support included adrenaline, noradrenaline, and milrinone infusions, prolonged mechanical ventilation, and continuous peritoneal dialysis.

During the first postoperative days, the patient developed generalized edema, anuria, and dependence on high-dose vasoactive support. Nevertheless, cardiac function and hemodynamic status gradually improved with intensive care management. Definitive sternal closure was successfully performed on postoperative day 7 after resolution of myocardial edema and stabilization of hemodynamics. Subsequently, vasoactive support was weaned, respiratory function improved, and extubation was achieved successfully.

Postoperative echocardiography demonstrated complete VSD closure without residual shunting and satisfactory RVOT flow. At 1-month follow-up, the right ventricle remained mildly dilated with mildly reduced systolic function, and no intracardiac thrombus was identified. At 2 months, right ventricular size and function had improved substantially. Mild residual supravalvular pulmonary stenosis, mild-to-moderate pulmonary regurgitation, and mild-to-moderate tricuspid regurgitation were present.

At 3-month follow-up, echocardiography demonstrated good right ventricular function, complete VSD closure without residual shunting, and satisfactory RVOT flow with a peak gradient of approximately 37 mmHg. Only mild residual pulmonary stenosis, mild pulmonary regurgitation, and mild tricuspid regurgitation remained. Clinically, the patient was thriving, with stable oxygen saturation in room air and complete resolution of cyanosis.

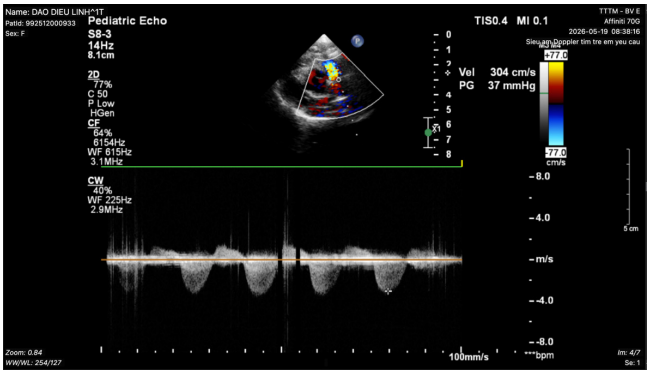


**Figure 3. Doppler echocardiography at 3-month follow-up demonstrating satisfactory right ventricular outflow tract flow with only mild residual obstruction (peak gradient 37 mmHg)**

## 3. DISCUSSION

- Tetralogy of Fallot is a common cyanotic congenital heart defect in which the VSD is typically large and non-restrictive, allowing equalization of ventricular pressures. Consequently, right ventricular pressure in classical TOF is usually systemic, and the degree of cyanosis depends primarily on the severity of RVOT obstruction. In contrast, TOF associated with a restrictive VSD is a rare variant with distinct hemodynamic characteristics. Flanagan et al. reported only four cases of restrictive VSD among 269 patients undergoing surgical repair for TOF, highlighting the rarity of this condition [1].

- The mechanism of VSD restriction in TOF is commonly related to accessory tricuspid valve tissue, hypertrophied right ventricular muscle bundles, or aneurysmal membranous septal tissue partially occluding the defect [3,4]. Soni et al. identified accessory tricuspid valve tissue as one of the most frequent causes of VSD restriction in these patients [3]. Restriction of the VSD creates a significant interventricular pressure gradient, resulting in true elevation of right ventricular pressure rather than the pressure equalization typically observed in classical TOF. McBrien et al. reported that right ventricular

pressure may reach systemic or even suprasystemic levels in patients with restrictive VSD, increasing the risk of right ventricular dysfunction and affecting surgical outcomes [2,5].

- Clinically, patients with TOF and a restrictive VSD may present with cyanosis that appears disproportionate to the degree of RVOT obstruction, making preoperative assessment challenging. Doppler echocardiography plays a crucial role in diagnosis by demonstrating a measurable pressure gradient across the VSD, unlike classical TOF in which little or no gradient is present [1]. In our patient, the combination of a small restrictive VSD and severe RVOT obstruction was consistent with previously reported hemodynamic findings.

- At the Cardiovascular Center, E Hospital, most TOF patients are currently treated through minimally invasive right axillary thoracotomy. However, in this infant, a lower partial sternotomy was selected to facilitate hemodynamic control and surgical exposure in the setting of a restrictive VSD and severe RVOT obstruction. Although complete repair successfully relieved RVOT obstruction and eliminated intracardiac shunting, postoperative myocardial edema and hemodynamic instability necessitated delayed sternal closure. This strategy is commonly employed in complex congenital heart surgery when there is concern regarding early postoperative right ventricular dysfunction or cardiac compression [10,11].

- The indications for and timing of complete repair in this infant merit particular consideration. Primary complete repair is currently the preferred strategy for symptomatic infants with TOF at experienced centers, and progressive cyanosis with declining oxygen saturation—as in our patient—is a recognized indication for early intervention [9,11]. Palliative procedures, such as a modified Blalock–Taussig shunt or right ventricular outflow tract stenting, are generally reserved for infants with markedly hypoplastic or discontinuous pulmonary arteries, an anomalous coronary artery crossing the outflow tract, or prohibitive comorbidities. In our patient the pulmonary arteries were confluent and of adequate caliber and the coronary anatomy was normal, so primary repair could be undertaken safely. A restrictive VSD does not change the fundamental indication for repair, but the associated suprasystemic right ventricular pressure and severe right ventricular hypertrophy increase the anticipated risk of postoperative right ventricular dysfunction and therefore influence perioperative planning. In this patient the outflow tract was managed with a valve-sparing strategy—pulmonary commissurotomy with combined transatrial and transpulmonary enlargement across the tricuspid and pulmonary valves, together with main pulmonary artery patch augmentation—rather than a transannular incision; after repair the pulmonary annulus admitted a 7–8 mm dilator and both branch pulmonary arteries admitted a 5–6 mm dilator. Preservation of pulmonary valve competence is particularly desirable in this setting, because the chronically pressure-loaded, hypertrophied right ventricle tolerates the additional volume load of free pulmonary regurgitation poorly [4]. A lower partial sternotomy was therefore chosen to optimize exposure and hemodynamic control and to preserve the option of delayed sternal closure.

- The most challenging aspect of the postoperative course was transient right ventricular dysfunction. In TOF with a restrictive VSD, the right ventricle is chronically exposed to systemic or suprasystemic pressure, producing marked hypertrophy and reduced diastolic compliance; after repair, this stiff, hypertrophied ventricle is prone to restrictive physiology and low cardiac output and tolerates volume loading poorly [6,12]. In our patient this was manifested by myocardial edema, generalized edema, anuria, and dependence on high-dose vasoactive support, and any residual outflow tract gradient or pulmonary regurgitation further increased the ventricular load. Management was therefore directed at supporting

right ventricular performance while minimizing its afterload and volume load. Milrinone was used for its inodilator and lusitropic effects and its ability to lower pulmonary vascular resistance, whereas adrenaline and noradrenaline maintained cardiac output and coronary perfusion pressure. Ventilation was managed to preserve oxygenation and to avoid the increases in pulmonary vascular resistance produced by hypoxemia, acidosis, and excessive mean airway pressure. Because a hypertrophied right ventricle depends on adequate but not excessive preload, fluid balance was carefully controlled, and peritoneal dialysis was instituted to treat fluid overload and anuria and to relieve edematous compression of the heart. Delayed sternal closure was the pivotal intervention: leaving the sternum open relieved compression by the edematous, poorly compliant heart and mediastinum and improved diastolic filling, and definitive closure was deferred to postoperative day 7, once myocardial edema had resolved and hemodynamics had stabilized [10,11]. In the most severe cases of postoperative right ventricular failure, additional measures such as an intentional atrial-level fenestration—allowing right-to-left shunting to sustain systemic output at the expense of transient desaturation—or mechanical circulatory support may be required. Consistent with the generally reversible nature of early restrictive right ventricular physiology, right ventricular size and function improved progressively over the following weeks, and by 3 months systolic function had normalized.

- Published reports describing TOF with a restrictive VSD remain scarce and are largely limited to isolated case reports [2–5]. To our knowledge, few such cases have been reported in Vietnam, particularly in infants. Therefore, this case contributes additional clinical, hemodynamic, and surgical data regarding a rare variant of TOF.

## 4. CONCLUSION

Tetralogy of Fallot associated with a restrictive ventricular septal defect represents a rare anatomical and hemodynamic variant. Accurate diagnosis and timely complete surgical repair can result in favorable outcomes even in young infants.

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