

# Challenges Faced in Managing an Adult Uncorrected Tetralogy of Fallot Patient with Pneumonia and Septic Shock in the Intensive Care Unit

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## ABSTRACT

Tetralogy of Fallot (TOF) is the most common form of cyanotic congenital heart disease. We present a 61-year-old uncorrected TOF patient who presented with sepsis and acute respiratory failure. At presentation, the patient had tachypnea, tachycardia, and oxygen saturation (SpO<sub>2</sub>) 64%. Chest X-ray (CXR) showed bilateral basal opacities. 2D echo revealed left ventricular dysfunction, infundibulum stenosis, pulmonary atresia, overriding of the aorta, and ventricular septal defect. A provisional diagnosis of community-acquired pneumonia with septic shock with uncorrected TOF was made. He received empirical antibiotics, ventilatory support, and supportive care. Ventilator weaning was done accepting an arbitrary baseline SpO<sub>2</sub> of 70–75% and lactate-guided volume resuscitation led to a successful outcome. Coronary angiogram showed collaterals. This case report illustrates the dilemmas faced in treating a critically ill uncorrected TOF. To the best of our knowledge, this case could be the oldest surviving uncorrected TOF patient in the Indian population.

**Keywords:** Cyanotic congenital heart disease, Fluid resuscitation, Sepsis, Uncorrected adult tetralogy of Fallot, Ventilation.

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## INTRODUCTION

The average life expectancy of a newborn with uncorrected tetralogy of Fallot (TOF) is only 12 years. Ten percent of TOF patients reach their 30s and it is only 3% for the age group above 40s.<sup>1</sup> Tetralogy of Fallot comprises ventricular outflow tract obstruction, overriding of the aorta, interventricular septal defect, and right ventricular hypertrophy.<sup>2</sup> Pulmonary infections in patients with cyanotic heart disease are rare as compared to acyanotic heart disease. Tetralogy of Fallot patients have high hematocrit and polycythemia with low oxygen saturations. We present an uncorrected TOF with pneumonia, septic shock, and the dilemmas in ventilation and volume resuscitation.

## CASE DESCRIPTION

A 61-year-old male uncorrected TOF patient presented with a history of dry cough and shortness of breath (NYHA grade III) for 2 days. At a presentation in the emergency room (ER), he was conscious, coherent with a respiratory rate of 45/minute, heart rate (HR) 110/minute, and blood pressure (BP) 180/110 mm Hg. Room air SpO<sub>2</sub> was 64 and 72% with 6 L/minute O<sub>2</sub>. On examination, central cyanosis was seen with bilateral digital clubbing and on auscultation bilateral rhonchi were heard. Arterial blood gas (ABG) report showed pH 6.8, PCO<sub>2</sub> 61.2 mm Hg, PaO<sub>2</sub> 57 mm Hg, bicarbonate of 9 mmol/L, and elevated lactate of 5.2 mmol/L. He was intubated in the ER. Received empirical antibiotics after sending appropriate cultures.

During the intensive care unit (ICU) stay, he was ventilated with pressure control of 16 cm H<sub>2</sub>O, rate of 28/minute, 10 positive end expiratory pressure (PEEP), and 100% FiO<sub>2</sub>. Chest X-ray (CXR) showed bilateral basal opacities and cardiomegaly. Lung ultrasound revealed bilateral basal dynamic air bronchogram suggestive of consolidation. Electrocardiogram showed right axis deviation, right ventricular hypertrophy, and incomplete right

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bundle branch block. 2D echo revealed severe left ventricular dysfunction, severe infundibular stenosis, pulmonary atresia, overriding of the aorta, and ventricular septal defect (VSD). Lab parameters revealed hemoglobin 18.8 g/dL, hematocrit 66.8, creatinine 1.2 mg/dL, normal electrolytes, and 17,700 cells/mm<sup>3</sup> leukocytes. The patient received vasopressor and inotropic support to reduce shunt and treat septic shock. We could not use a hemodynamic monitor to guide volume resuscitation because of his financial constraint.

Post-resuscitation ABG showed pH 7.17, PCO<sub>2</sub> 51.4 mm Hg, PaO<sub>2</sub> 57.2 mm Hg, and bicarbonate 18.3 mmol/L with a decrease in both lactates and hematocrit. Based on his SpO<sub>2</sub> which was targeted between 70 and 75% ventilator was weaned.

Endotracheal (ET) culture grew multi drug resistant (MDR) *Klebsiella* and appropriate antibiotics were continued. On the 4th day of minimal ventilatory support, ABG showed pH 7.41, PCO<sub>2</sub> 31 mm Hg, PaO<sub>2</sub> 43.9 mm Hg, bicarbonate of 19.5 mmol/L, lactates 1.91 mmol/L, and hematocrit 54%. So, the patient was extubated and received non invasive ventilation (NIV) support to avoid reintubation. The next day, he was weaned off NIV support,

**Table 1:** Factors varying from pulmonary vascular resistance (PVR) and systemic vascular resistance (SVR)

Increase in PVR	Decrease in SVR
Hypercarbia	Vasodilatation—sepsis, anaphylaxis
Acidosis	Anemia
Hypoxia	Cirrhosis
Hypothermia	Hyperthermia

vasopressor, inotropic supports, and was shifted to room on the 6th day of admission. A coronary angiogram revealed extensive collaterals. He was discharged on the 8th day of admission.

## DISCUSSION

The world estimate of congenital heart disease is around 8/1,000 live births, and TOF ranked fifth in the list.<sup>3</sup> The prevalence of TOF is 3.4/10,000 live births.<sup>4</sup> Adult TOF survivors showed adaptations like the hypoplastic pulmonary artery with the slow development of subpulmonary obstruction, left ventricular hypertrophy, extracardiac shunts, or systemic to pulmonary artery shunting via internal mammaries.<sup>5–10</sup>

The goals of management in such a patient are preventing hypoxia, preventing falls in systemic vascular resistance (SVR), avoiding an increase in pulmonary vascular resistance (PVR), prevention acidosis, hypothermia, tachycardia, and hypovolemia.

In TOF with septic shock, SVR is decreased leading to an increase in right to left (R→L) shunt and worsening hypoxia. Vasopressor supports help in the prevention of SVR fall, thereby preventing worsening of the shunt. Factors varying from PVR and SVR are to be avoided (Table 1). Acidosis in septic shock with TOF results in increased pulmonary vascular resistance and increasing R→L shunt. Henceforth, lactate clearance by volume resuscitation and respiratory acidosis by ventilator management is essential. Tachycardia causes increased myocardial oxygen demand. Sedation and paralysis prevent any stress or anxiety-related tachycardia. Hypovolemia not only causes cyanosis by decreasing preload which results in narrowing RVOT but also causes increase R→L shunt because of low systemic pressure. Hypothermia causes PVR to increase and worsening of R→L shunt.

The challenges faced in managing uncorrected adult TOF patients with pneumonia and sepsis are the prevention of hypoxia, determining the adequate PaO<sub>2</sub> levels for ventilating and weaning. The patient's PaO<sub>2</sub> never reached an acceptable range seven with 100% FiO<sub>2</sub> as TOF being cyanotic heart disease. Second, determining volume status and volume resuscitation in such a sepsis patient was difficult as these patients have high hematocrit and polycythemia.

FiO<sub>2</sub> was weaned gradually to a minimum of 60% over 24 hours based on his SpO<sub>2</sub> target of 70–75%. We slowly weaned ventilator support, extubated him on the 4th day of ventilation, and continued on NIV support for another day.

Volume resuscitation plays an important role in septic shock patients. As the patient had severe left ventricular dysfunction, volume resuscitation was challenging. Hematocrit could not be of much help as the patient had elevated values due to cyanotic heart disease. Arterial blood gas lactate levels at presentation were 5.2 mmol/L. We continued volume resuscitation basing on lactate clearance. Post-resuscitation ABG showed a fall in lactate to 4 mmol/L and was 1.91 mmol/L at the time of extubation. Vasopressor and inotropic supports were also weaned off.

A coronary angiogram revealed extensive collaterals. This could explain the underlying reason for the survival of our patients until this age.

## CONCLUSION

A good interpretation of altered physiology in TOF along with management of septic shock is essential for resuscitation. Such cases of uncorrected TOF with sepsis and acute respiratory failure pose a challenge for intensivist in determining parameters to guide ventilatory weaning and adequate volume resuscitation. In this case, lactate clearance as a fluid guidance parameter, and in ventilator management acceptance of target SpO<sub>2</sub>, prevent variations in SVR and PVR helped us for a successful outcome.

## REFERENCES

- Bertranou EG, Blackstone EH, Hazelrig JB, Turner ME, Kirklin JW. Life expectancy without surgery in tetralogy of Fallot. *Am J Cardiol* 1978;42(3):458–466. DOI: 10.1016/0002-9149(78)90941-4.
- Kirklin JW, Barratt-Boyes BG. Ventricular septal defect and pulmonary stenosis or atresia. In: Kirklin JW, Barratt-Boyes BG, ed. *Cardiac Surgery*. 2nd ed., New York: John Wiley and Sons; 1993. pp. 749–824.
- Van Der Linde D, Konings EEM, Slager MA, Witsenburg M, Helbing WA, Takkenberg JJM, 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.
- Gupta K, Bajaj B, Das B. Successful outcome of pregnancy in uncorrected tetralogy of Fallot. *Int J Reprod Contracept Obstet Gynecol* 2014;3:799–802. DOI: 10.5455/2320-1770.ijrcog20140935.
- Meindok H. Longevity in the tetralogy of Fallot. *Thorax* 1964;19(1):12–15. DOI: 10.1136/thx.19.1.12.
- Thomas SHL, Bass P, Pambakian H, Marigold JH. Cyanotic tetralogy of Fallot in a 77 year old man. *Postgrad Med J* 1987;63(739):361–362. DOI: 10.1136/pgmj.63.739.361.
- Bowie EA. Longevity in tetralogy and trisomy of Fallot. Discussion of cases in patients surviving 40 years and presentation of two further cases. *Am Heart J* 1961;62(1):125–132. DOI: 10.1016/0002-8703(61)90491-4.
- Chin J, Bashour T, Kabbani S. Tetralogy of Fallot in the elderly. *Clin Cardiol* 1984;7(8):453–456. DOI: 10.1002/clc.4960070807.
- Nottestad SY, Slife DM, Rubal BJ, Moody JM. Tetralogy of Fallot in a 71 year old patient with new onset hypoxemia. *Cathet Cardiovasc Diagn* 1993;28(4):335–338. DOI: 10.1002/ccd.1810280413.
- Liberthson RR, Miller SW, Drew F, Palacios I, Singh J, Singh J, et al. Congenital extracardiac shunts with tetralogy of Fallot. *Cardiovasc Intervent Radiol* 1981;4(2):131–135. DOI: 10.1007/BF02552393.