

CASE SERIES

ANESTHESIA & CONCURRENT DISEASE

Anesthetic management of adult patients with tetralogy of Fallot: a case series

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ABSTRACT

Tetralogy of Fallot (ToF) is the most common form of cyanotic congenital heart disease, occurring predominantly in the childhood with a rare late presentation in the adults. the prognosis OF ToF remains poor, especially in those who do not undergo surgical correction. However, in the recent times, there is a significant improvement in the prognosis of congenital heart diseases and almost 85% patients are expected to survive till adulthood due to early diagnosis and appropriate medical and surgical management. These adult patients with congenital heart disease exhibit specific and complex anatomic and physiological changes, which can affect the perioperative morbidity and mortality to a great extent. We present this case series to discuss the relevant clinical and successful anaesthetic conduct of adult patients with ToF. before and after surgical repair, with good perioperative outcome.

Key words: Tetralogy of Fallot; Congenital heart disease; Anesthesia; Cyanosis; Pulmonary stenosis; Pulmonary atresia.

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1. INTRODUCTION

Tetralogy of Fallot (ToF) is a severe form of cyanotic congenital heart disease, occurring in around 0.34 per 1000 live births,¹ while the prevalence in adult population is roughly 2.4 per 1000 individuals.² It is usually diagnosed before adolescence and most of the patients are symptomatic during infancy and mandate early treatment. There are few reports of survival in late diagnosis of middle-aged patients.³ The classic anatomical defect includes a tetrad of Right Ventricular Outflow Tract Obstruction (RVOTO), Ventricular Septal Defect (VSD), Right Ventricular Hypertrophy (RVH) and an overriding of aorta. The clinical presentation in adult ToF is difficulty on exertion, palpitations and chest pain; whereas, tet spells are

typically not seen beyond adolescence. Babies with ToF have episodes called tet spells, when they suddenly turn bluish and may faint. These spells are serious. The degree of severity is determined by the relative pressure gradient between the right and left ventricles and the degree of RVOTO. The treatment is generally based on the severity, and the prognosis after surgical correction is improved, making surgical intervention, the treatment of choice. However, delay in the diagnosis and late intervention are highly associated with poor outcomes. Anesthetic management of an uncorrected ToF is challenging, which requires a thorough understanding of complex anatomic and physiological changes, anticipation of perioperative events and their management, and warrants a diligent anesthetic as well

as surgical planning and its successful execution. Here, this case series will discuss the relevant clinical and successful anesthetic conduct of adult ToF before and after surgical repair with good outcome.

2. CASE DESCRIPTION

2.1. CASE 1: ToF with double SVC

An 18-year-old female presented with dyspnea on exertion for two months. She was a known case of hypothyroidism and was on tab thyroxine 50 µg OD. There was no history of discoloration of hands/feet or tet spells since childhood. On examination, she was thin, afebrile, not cyanosed with grade III clubbing. Vital signs were; SpO₂ 80% on room air, NIBP 130/80 mmHg, HR 78/min. Airway examination was within normal limits. On auscultation, S₁S₂ heard along with pansystolic murmur over left sternal border at fourth intercostal space. Hemogram showed hemoglobin 18 g% and hematocrit of 57%. Other investigations were within normal limits. ECG showed right ventricular hypertrophy and right axis deviation. Chest X-ray was suggestive of boot shaped heart.

2D Echo revealed a large sub-aortic VSD with bidirectional shunt, right ventricular hypertrophy, overriding of aorta, valvular and subvalvular pulmonary stenosis with a gradient of 70 mmHg. Normal carotid Doppler study. High-Resolution Computed Tomography (HRCT) thorax showed double SVC possibly draining into coronary sinus and e/o fibrobronchiectatic and atelectatic changes in her right lung and tiny sub-pleural nodules in left lower lobe.

2.2. CASE 2: ToF with absent pulmonary valve

A 43-year-old male presented with dyspnea on exertion and fatigue since childhood, gradually progressive, aggravated since last two years and was started on tab metoprolol 25 mg BID. There were no other complaints or known co-morbidities. On examination, he was thin, afebrile and his vital signs were; SpO₂ 97% on room air, NIBP 102/62 mmHg, HR 67/min. On auscultation, S₁S₂ were heard along with systolic murmur at apex and pulmonary area. Airway examination and all blood investigations were within normal limits (hemoglobin 13.2 g%, hematocrit 41.9%). ECG showed a right bundle branch

block (RBBB) and right ventricular hypertrophy. Chest X-ray was suggestive of cardiomegaly. (Figure 1).

2D Echo revealed a large sub-aortic VSD with bidirectional shunt (predominantly left to right shunt), right ventricular hypertrophy, overriding of aorta and a rudimentary or absent pulmonary valve with a small annulus, severe PR, dilated MPA with dilated branches. Carotid Doppler study was normal.

2.3. CASE 3: ToF with severe pulmonary stenosis

A 29-year-old male presented with dyspnea and atypical chest pain for two months. No other complaints/known co-morbidities. On examination, he was thin, afebrile and vitals were within normal limits. On auscultation, S₁S₂ heard along with a harsh ejection systolic murmur best heard at the left upper sternal border. Airway examination and all blood investigations were within normal limits (hemoglobin- 15.8g%, hematocrit – 43.5%). ECG showed RBBB and right ventricular hypertrophy. Chest X-ray revealed a normal heart appearance.

2D Echo revealed a membranous ventricular septal defect with a right to left shunt, severe right ventricular hypertrophy with severe tricuspid regurgitation (TR), severe pulmonary valve stenosis and overriding aorta. Carotid Doppler study was normal.

2.4. CASE 4: Pentalogy of Fallot for ToF repair

A 23-year-old female presented with dyspnea on exertion for two years. She was evaluated for dyspnea and diagnosed to have pentalogy of Fallot and was started on tab. metoprolol 12.5 mg OD, tab. furosemide and tab. spironolactone. She was a known case of

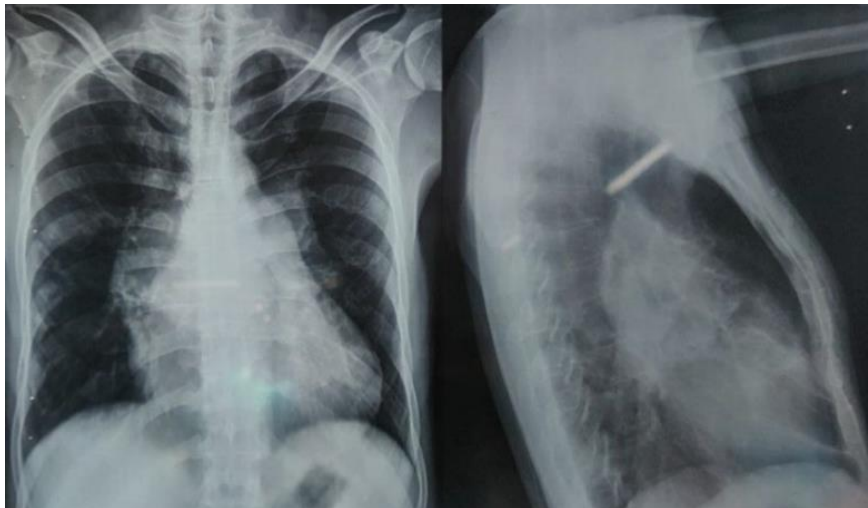


Figure 1: Chest X-ray PA and lateral views

pulmonary Koch's and had completed anti-tuberculosis therapy (ATT) for six months. She was also found to have a liver abscess one month back and was on conservative management. On examination, she was thin, afebrile, not cyanosed with grade IV clubbing. Her SpO₂ was 88% on room air, NIBP 100/80 mmHg, and HR 90/min. Airway examination was within normal limits. On auscultation, S₁S₂ were heard along with pansystolic murmur over left sternal border at second intercostal space. Hemogram showed hemoglobin 20 g% and hematocrit 57%. Other investigations were within normal limits. ECG showed right ventricular hypertrophy and right axis deviation. Chest X-ray was suggestive of boot shaped heart.

2D Echo revealed a 13 mm ostium secundum atrial septal defect (OS-ASD) with bidirectional flow, pulmonary valvular stenosis with a gradient of 105 mmHg. Grade II TR, right ventricular hypertrophy, severe pulmonary arterial hypertension, large sub-aortic VSD with bidirectional shunt, ejection fraction (EF)–60%. HRCT thorax showed thin walled cavitary lesion in superior segment of left lower lobe with few adjacent cavities. Centrilobular nodules in superior segment of left lower lobe.

2.5. CASE 5: Sub-aortic VSD for ToF repair

An 18-year-old female presented with breathlessness for eight months which aggravated on exertion. There was a history of bluish discoloration of lips and fingers for eight months. No other co-morbidities were noted. On examination, she was thin, afebrile, and had cyanosis of fingers and lips with grade III clubbing. SpO₂ was 89% on room air, NIBP 100/70 mmHg, and HR 100/min. Airway examination was within normal limits. On auscultation, S₁S₂ were heard along with pansystolic murmur over left sternal border at second intercostal space. Hemogram showed hemoglobin 19.2 g% and hematocrit 53%. Other investigations were within normal limits. ECG showed right axis deviation. Chest X-ray revealed a normal heart.

2D Echo revealed a large sub-aortic VSD with bidirectional flow, pulmonary valvular and subvalvular stenosis with a gradient of 80 mmHg, overriding of aorta and right ventricular hypertrophy. Carotid Doppler and HRCT thorax studies were normal.

2.6. Anesthetic Management

Patients were admitted a week prior to surgery and necessary preoperative evaluation was done. On the day of surgery, starvation was ensured and informed high-risk consent was obtained. An 18G or 20G IV cannula was inserted in a peripheral vein and sedation was given in the operating room (OR) with inj. midazolam

0.02–0.03 mg/kg and inj. fentanyl 1–2 µg/kg. A central venous line was inserted in the right internal jugular vein. A right femoral arterial line was passed under USG guidance under local anesthesia. Vital sign monitoring included electrocardiography, pulse oximetry, invasive blood pressure, central venous pressure, capnography, NMT, entropy, temperature and urine output. Activated clotting time (ACT), hemogram, ionogram, blood glucose and serial arterial blood gases were measured. Infective endocarditis prophylactic was achieved with inj. ceftriaxone 1g IV, given prior to the surgery.

Anesthetic induction was performed with inj. midazolam 0.03–0.05 mg/kg, inj. fentanyl 5–8 mcg/kg, inj. ketamine 1 mg/kg, inj. etomidate 0.3–0.5 mg/kg, and relaxation was achieved with inj. rocuronium 1–1.5 mg/kg. Maintenance was done with sevoflurane 1–2% in oxygen:air in a ratio of 60:40. Intraoperatively, inj. propofol and inj. atracurium were titrated as per NMT and Entropy monitors and continued during cardiopulmonary bypass (CPB). Necessary precautions were taken against the entry of air bubbles into the circulation. Unfractionated heparin 300 U/kg was injected before cannulation and ACT was kept above 400 sec.

Patients were operated by median sternotomy. The surgical correction was done by the closure of VSD with a patch and pulmonary valvulotomy in CASE 1; VSD closure, pulmonary pericardial patch repair and pulmonary valve replacement in CASE 2; VSD closure and pulmonary valve replacement in CASE 3; closure of ASD and VSD, and pulmonary valve replacement was done in CASE 4; and closure of VSD was done in CASE 5. Right ventricular outlet tract coring was done in all patients, in addition, to decompressing the right ventricle (RV).

Once sinus rhythm was restored, separation from CPB was initiated and ACT was targeted below 120 sec by giving inj. protamine. Inj. dopamine 3–5 µg/kg/min was started in CASE 3 and 4. Inj. nitroglycerine 0.3–0.5 µg/kg/min was started during weaning in all patients. Heart rate, rhythm and mean arterial pressure (MAP) were maintained. The patients were transferred to ICU for further weaning and management, and were discharged from the ICU on the third or fourth postoperative day and discharged successfully from the hospital by eighth day.

3. DISCUSSION

ToF accounts for about 10% of all congenital heart diseases,⁴ which includes four anatomic components, e.g., ventricular septal defect, right ventricular outflow tract obstruction, right ventricular hypertrophy, and overriding of aorta, as described by Fallot in 1888. However, only 2% of all ToF patients survive till

4th decade and survival beyond the 7th decade is even rarer.⁵ Each of these components can vary in severity and this variation can significantly impact the disease presentation and further management. VSD can be of perimembranous (70%), muscular or sub-arterial types. The size of the defect, pressure difference and the associated anomalies determine the severity, but most of the cases have unrestricted bidirectional shunt. The presentation of RVOTO maybe pulmonary stenosis, pulmonary atresia or infundibular spasm (dynamic or fixed obstruction). Other variants associated with ToF include absent or rudimentary pulmonary valve (5%), double outlet right ventricle (DORV), atrioventricular septal defect (2%), anomalous origins of coronary arteries (16%) and a right aortic arch.⁶ Pentalogy of Fallot is also a ToF variant, associated with atrial septal defect, accounting for about 3.7% of all congenital heart diseases.⁶

The delayed presentation of ToF in the adulthood can be due to mild symptoms and lack of awareness. The unusual prolonged lifespan in these patients is probably related to many factors, including;

- 1) favorable anatomical changes resulting in an adequate pulmonary blood flow, better systemic to pulmonary collateral circulation or patent ductus arteriosus,⁷
- 2) a hypoplastic pulmonary artery with the slow development of sub-pulmonary obstruction, and
- 3) left ventricular hypertrophy which acts by delaying the right to left ventricular shunt.⁸

All of our patients had no shunt procedures during the childhood or adolescence, and presented with symptoms directly in the adulthood, which is possibly due to the given favorable anatomical-physiological changes.

The uncorrected ToF patients pose a major challenge due to the persistent changes. The clinical presentation can vary from reduced exercise tolerance, chest pain, clubbing, hypoxemia, ventricular dysrhythmias, endocarditis to polycythemia and its complications like coagulopathy, brain abscess, stroke and sudden death.⁹ Preoperatively, detailed assessment is required regarding the cardiac defect, altered physiology and its anesthetic implications. Cyanosis, pulmonary hypertension and congestive cardiac failure, especially right ventricle failure, should be evaluated properly as these are the common manifestations of congenital heart diseases. It is also necessary to rule out any other congenital systemic anomalies.

The main goals of anesthetic management in ToF patients are to maintain heart rate, contractility and preload to maintain cardiac output. Maintenance of euvolemic status is important to avoid decreased afterload, which in turn can aggravate reflex increase in

heart rate, right to left shunt and coagulopathy in the presence of polycythemia. It is essential to avoid prolonged starvation and maintain adequate intravenous hydration during fasting. Maintenance of normal or high SVR, low PVR and avoidance of tachycardia is required, so that the pulmonary blood flow is maintained. The drug combination preferred for our induction was fentanyl, ketamine and etomidate, with the aim to maintain SVR and oxygenation. Care was taken to avoid painful stimulus and air bubbles entering into the circulation.

Hypoxemia, hypercapnia, acidosis, high hematocrit and positive pressure ventilation can increase PVR and presenting a pulmonary hypertensive crisis. Simultaneously, hyperthermia, deeper planes of anesthesia and vasodilators can decrease SVR and worsen RVOT obstruction.³

Multiple anatomical variations of ToF have been documented in the literature. Prolonged survival can be related to a lesser degree of RVOT obstruction. Having said that, the stress on the anatomic areas, pulmonary circulation changes, reduced ventricular compliance and harmful effects of oxygen desaturation can compromise the survival in the long run, so most patients present in the early adulthood.⁷

4. CONCLUSION

With the advancements in the medical diagnostics and management, the adult patients with congenital heart disease are now often seen visiting medical institutions. A complete understanding of the disease, a meticulous planning and a multidisciplinary patient-centric approach are necessary for the successful management in the patients. More multi-center research with, large-scale studies is needed to offer definitive practice guidelines for the anesthetic management.

5. Conflict of interest

None declared by the authors.

6. Authors' contribution

All authors took part in the perioperative management of the documented patients as well as preparation of this manuscript.

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