

A Case Report on Anaesthetic Management of Lower Limb Fracture in an Adult with Uncorrected Tetralogy of Fallot

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Abstract

Tetralogy of Fallot is one of the commonest forms of cyanotic congenital heart disease. The treatment strategies currently used in the treatment of ToF result in excellent long-term survival (30 year survival ranges from 68.5% to 90.5%). Unrepaired or nonpalliated, approximately 25% of patients survive to adolescence, after which the mortality is 6.6% per year. Only 3% survive to age 40. Adults with uncorrected ToF pose a challenge for anaesthesia because perioperative morbidity and mortality is greater. The most important aspect of the perioperative care is the involvement of a multidisciplinary team with a detailed understanding of the patient's cardiac defect, their functional status and anticipation of the perioperative stresses. We report a case of middle-aged woman who was asymptomatic during her life and diagnosed with uncorrected ToF during her 32nd year of age now presenting with lower limb fracture after RTA for surgical fixation. The surgery was successfully conducted under graded epidural anaesthesia. This anaesthetic technique can be considered as an alternative technique to general anaesthesia, in selected patients with uncorrected ToF presenting for lower limb fractures, especially in cases where the risks of administering a general anaesthetic are deemed high.

Keywords: Congenital heart disease, Right ventricular outflow tract obstruction (RVOTO), Epidural anaesthesia.

Introduction

French physician Etienne-Louis Fallot recognised a series of malformations and reported the first case series in 1888 with anatomical and pathological descriptions of the four key features [1], namely VSD, RVOTO, Overriding aorta, RVH. ToF accounting for 7–10% of all congenital cardiac malformations with an incidence of one in 3,500 live births [2]. Uncorrected, 70% of children with ToF die before their tenth birthday, although survival into adulthood is possible [3].

ToF may present with different phenotypes ranging from mild to severe cases such as Fallot-type double outlet right ventricle or ToF with pulmonary atresia [4]. Almost all tetralogy of fallot patients present with cyanosis. This is due to right-to-left shunting through the large VSD. Some other conditions are associated with ToF; 15% are part of syndromes, including Down, DiGeorge, and Alagille syndromes; vertebral defects, anal atresia, cardiac defects, tracheo-oesophageal fistula, renal anomalies, and limb abnormalities (VACTERL association); and velocardiofacial abnormalities [5].

Chronic hypoxemia and right-to-left shunting cause significant alterations in pathophysiology which makes these patients more vulnerable to perioperative complications such as hemodynamic instability, congestive heart failure, arrhythmias, cyanotic spells,

acid–base imbalance, coagulation defects.

Case Report

A 35-year-old female admitted with alleged history of self fall from two wheeler and sustained right inter trochanteric fracture of femur. She gave a history of heart disease diagnosed during her 32nd year of life when she was in second trimester of pregnancy. Her pregnancy was terminated due to uncorrected heart disease and breathlessness. The details of those treatment were lost during course of time. Currently the patient is able to do her regular household activities and able to walk a distance of approximately of 1 km to her workplace. On general examination patient had cyanosis with grade II clubbing (Fig. 2). She had pan systolic murmur with palpable thrill over precordium. Her room air saturation was 58% (Fig. 3). hemoglobin was 15 g% with coagulation parameters like PT/INR, Aptt within normal limits. Airway examination revealed MMC 2 with adequate mouth opening. Spine examination was normal. ECG showing right axis deviation with RVH (Fig. 1) and echocardiography suggestive of TOF with subaortic VSD, RVH, aortic override, normal pulmonary artery pressure and ejection fraction 67% (Fig. 9).

Informed written and high risk consent taken. In the operating room intravenous line secured with 18 G cannula and standard monitors

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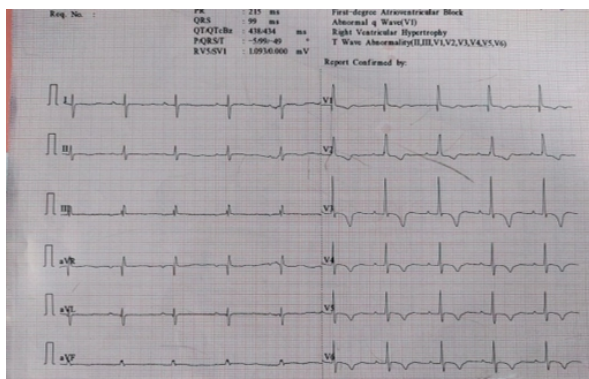


Figure 1: ECG showing RVH



Figure 2: grade - II clubbing

ECG, NIBP, pulse oximeter, were attached. Defibrillator was kept ready in case of arrhythmias. Under local anaesthesia right internal jugular and left radial artery cannulated. Antibiotic coverage given for infective endocarditis prophylaxis. Preoperative ABG findings were pH 7.44, PaO₂ -37, PaCO₂-32.9, (image 4) SpO₂-73% at room air and preoperative pulse rate was 70/min and blood pressure 92/58 mm Hg. With non rebreathing bag and mask 10 L of O₂, SpO₂ raised to 82%. Midazolam 1 mg and fentanyl 50 micro gram given intravenously to relieve pain and anxiety. Ultrasound guided supra inguinal fascia iliaca block (Fig. 6) given for analgesia and positioning. In sitting position Tuohy needle inserted at L3-L4 interspace through para median approach and epidural space identified by loss of resistance to air and catheter kept at 10 cm skin level (Fig. 7). Ringer lactate 500 ml given as bolus to counteract vasodilation. 0.75% ropivacaine 10 ml was given in increments of 5 ml to achieve adequate motor and sensory blockade. Sensory level upto T10 was achieved. Epidural top up of 0.75% ropivacaine 10 ml in increments of 5 ml was repeated after one hour of initial dose.

There were two episodes of hypotension (80/42 mm Hg) requiring 20 mics of phenylephrine. The lowest blood pressure recorded throughout the procedure was systolic of 80 mm hg and diastolic of 40 mmHg. Total duration of surgery after positioning on fracture table was 50 minutes. Intra operative blood loss was around 300 ml .Total of 1litre ringer lactate was given. Urine output was 250 ml at the end of procedure. Central venous pressure ranged between 6-10 during the surgical procedure.

Thereafter patient hemodynamics (Fig. 8) remained stable throughout the procedure. Postoperatively she was started on 0.2% ropivacaine infusion 5 ml/hr for pain relief. She received enoxaparin

40 mg S/C od as deep vein thrombosis prophylaxis 12 hours after surgery. Epidural catheter was removed on second Post-operative day.

Discussion

Anaesthetic management of a patient with uncorrected TOF for noncardiac surgery is a challenge to anaesthesiologists and requires a thorough understanding of pathophysiology, events and effects of medications which can alter the magnitude of right to left shunting.

The usual description of tetralogy of Fallot includes (1) a large, nonrestrictive malaligned ventricular septal defect (VSD), with (2) an overriding aorta, (3) infundibular pulmonic stenosis, and (4) consequent right ventricular hypertrophy, all these result from an embryonic anterocephalad deviation of the outlet septum. At times, there is spectrum of disease with more severe defects, including stenosis of the pulmonary valve, stenosis of the pulmonary valve annulus, or stenosis and hypoplasia of the pulmonary arteries in the most severe cases. Pentalogy of Fallot refers to ToF with addition of an ASD.

It is uncommon to encounter an adolescent or adult with unrepaired ToF. Pulmonic stenosis results in obstruction to right ventricular outflow. Right-to-left shunting is caused by obstruction at the level of the right ventricular outflow tract and is unaffected by altering PVR. Systemic hypertension developing in adult life thrust an increased load on both ventricles because of unrestrictive VSD.

Increase in systemic vascular resistance decreases the right to left shunting and diminishes cyanosis but at the cost of right ventricular or biventricular failure. Increases in the inotropic state of the heart increase the dynamic obstruction at the right ventricular infundibulum and worsen right-to-left shunting. β -Blockers are often



Figure 3: room air saturation of patient

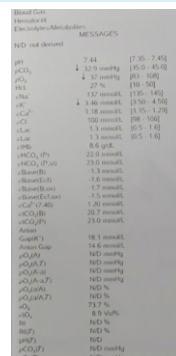


Figure 4: ABG in Room Air



Figure 5: Right Lower limb in everted and comfortable position after SIFI block

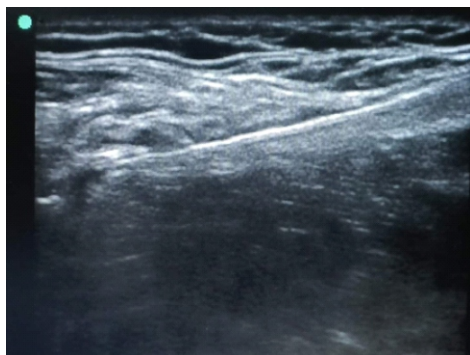


Figure 6: SIFI Block with Needle below Circumflex artery



Figure 7: epidural catheter at L2-L3 space

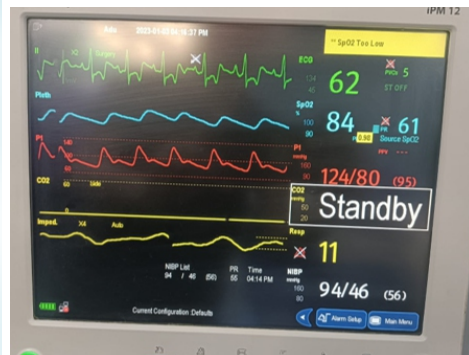


Figure 8: Intra op hemodynamics

used to decrease inotropy. The anaesthetic goals in management of these patients include maintenance of systemic blood pressure, increasing SVR, decreasing PVR, avoidance of hypovolemia, tachycardia, dehydration, hypoxia, hypercarbia, and acidosis.

The reason for delayed presentation with uncorrected lesion in this patient is balanced pulmonary and systemic circulation and may remain relatively asymptomatic until the balance between pulmonary and systemic circulation is disturbed. 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, [6]
- 2) a hypoplastic pulmonary artery with the slow development of sub-pulmonary obstruction,
- 3) left ventricular hypertrophy which acts by delaying the right to left ventricular shunt.

However, the uncorrected group present a severe challenge, in issues related to long term effects of chronic hypoxia [7]. These patients develop severe clubbing and learn to “squat” to increase pulmonary blood flow. They may be polycythemic and suffer complications of long standing polycythemia: Coagulopathy, intracranial abscess, stroke, hyperuricemia, and neuro developmental delay. They have reduced exercise tolerance and are vulnerable to ventricular arrhythmia or sudden death secondary to chronic pulmonary

egurgitation, problems related to abnormalities of RV physiology.

The severity of obstruction to pulmonary blood flow determines the presenting feature of tetralogy of fallot. Most patients will present in the neonatal period with mild-to-moderate cyanosis, but typically without respiratory distress.

Cyanosis occurs due to de-oxygenated blood entering the systemic circulation through right to left shunt and mixing of blood at ventricular septal defect. The relative pressure gradient between the right ventricle and left ventricle determines the degree of shunt through VSD. The amount of pulmonary blood flow (the RV stroke volume) is determined by the degree of the right ventricular outflow tract obstruction (RVOTO). Frequently, the degree of RVOTO has both fixed anatomical and variable physiological components [8].

RVOTO is often caused by sub-pulmonary valve muscle bundles. In addition to any fixed RVOTO, infundibular muscle spasm (often during times of distress) causes dynamic RVOTO thereby increasing the right-to-left-shunt fraction. This occurrence leads to profound hypoxaemia. Such episodes of sudden oxygen desaturation are often referred to as ‘cyanotic spells’ [9].

Uncorrected ToF survival is uncommon, it has been reported that around 10% of affected persons can survive to adulthood, and only 5% reach 40 years of age [10]. Mechanisms that explain longevity in patients who remained undiagnosed and survived to their adulthood may be attributed to having a small pulmonary artery and slow development of subpulmonary obstruction, left ventricular hypertrophy, and extra cardiac shunting or systemic to pulmonary shunt.

The timing of intervention is affected by delayed diagnosis. Usually patients with ToF repair tend to have excellent results but ToF repair during adulthood carries a higher risk of developing arrhythmias, heart failure, and sudden cardiac arrest [11].

Faraoni et al. demonstrated in a large-scale registry that preoperative markers of critical illness like (emergency procedure, inotropic support, mechanical ventilation, preoperative cardiopulmonary resuscitation, and acute or chronic kidney injury), the type of lesion (i.e., single-ventricle physiology), and the functional severity of the heart disease were strong predictors of in-hospital mortality in children undergoing noncardiac surgery, criteria that can probably apply also to adults with uncorrected CHD [12]. Older age at repair, severe left ventricular dysfunction, postoperative right ventricular hypertension from residual or recurrent outflow tract obstruction, wide-open pulmonary insufficiency, and prolongation of the QRS (to

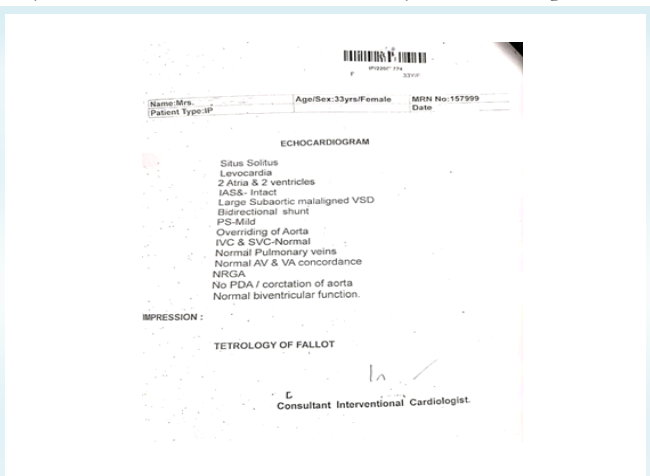


Figure 9: Echocardiogram report

>180 milliseconds) are all predictors of sudden death.

Cardiac catheterization plays an important role in the management of ToF in adults, to characterize the pulmonary arteries anatomically and also to define unanticipated anomalies such as aortopulmonary collateral arteries which are found in 15% of ToF patients.

Pain management is a critical factor during intra- or postoperative management. Opioid infusion or patient-controlled analgesia for major operations has been the primary postoperative intervention for pain in patients with CHD. The use of regional Anaesthesia for well-compensated patients with CHD was reported with almost no complications. Regional block avoids all the above complications and hence it was our choice. Regional (epidural and spinal) anaesthesia preserves patient baseline ventilation and perfusion state. Epidural anaesthesia also allows for continuous anaesthetic and analgesic infusion.

In our patient we chose graded epidural anaesthesia because of good effort tolerance preoperatively and mild pulmonic stenosis and also for the benefits from good postoperative analgesia, and avoidance of activation of the sympathetic nervous system with consequent catecholamine release.

Conclusion

We presented a case of a patient who was diagnosed to have ToF during her third decade of life without any symptoms. She had breathlessness during her second trimester of antenatal period at 32nd years of age and conception was terminated in view of uncorrected heart disease. It is uncommon to see adults with uncorrected ToF. These patients may be seen by Anaesthesiologist for primary repair of cardiac lesion or for other common ailments of ageing or trauma requiring surgical intervention. This case report highlight that patients with primary congenital diseases may remain undiagnosed until an older age despite medical advances and present with lot of anatomical and physiological complexities. These adult patients with uncorrected ToF require the best care for both cardiac and noncardiac surgery in a center with a multidisciplinary team experienced in the care of adults with CHD with meticulous Anaesthetic plan and post operative care. However, our patient had an smooth post-operative course with six months follow-up profile. She was able to do her day to day activities like cooking, thorough physical examination of newborns and a screening echocardiogram in the early life may aid in detecting the disease earlier.

Declaration of patient consent: The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his/her consent for his/her images and other clinical information to be reported in the Journal. The patient understands that his/her name and initials will not be published, and due efforts will be made to conceal his/her identity, but anonymity cannot be guaranteed.

Conflict of interest: Nil **Source of support:** None

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