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Systematic Review

Perioperative Management of Adult Patients with Tetralogy of Fallot Undergoing Intracardiac Repair: A Case Series

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ABSTRACT

Background: Tetralogy of Fallot (TOF) is the commonest cyanotic congenital heart disease. Survival to adulthood without surgical correction is uncommon and presents distinct perioperative challenges including polycythaemia, biventricular dysfunction, platelet dysfunction and altered haemodynamics.

Objective: To describe the perioperative anaesthetic and surgical management, intraoperative monitoring strategy, and outcomes of adult patients (≥15 years) with TOF undergoing intracardiac repair (ICR) at a tertiary cardiac centre.

Methods: A case series of 13 patients aged ≥15 years with echocardiographically confirmed TOF who underwent surgical correction between October 2024 and May 2026. Patient demographics, echocardiographic findings, anaesthetic technique, intraoperative monitoring including cardiac output by FloTrac/EV1000 via femoral artery, transoesophageal echocardiography (TEE), and near-infrared spectroscopy (NIRS), CPB parameters, and outcomes were analysed.

Results: Thirteen patients (9 males, 4 females; mean age 26.2 years, range 16–40 years) were included. All had preoperative polycythaemia (mean Hb 18.0 g%, mean Hct 53.2%, mean SpO_2 87.5%). Echocardiography demonstrated malaligned VSD with bidirectional shunt, RVOTO, RVH, and overriding aorta in all patients; three had associated defects (ASD in two, Ebstein anomaly in one). All were induced with ketamine–propofol combination. Advanced haemodynamic monitoring including FloTrac cardiac output, TEE, and NIRS was used routinely. Mean CPB time was 90 min and mean aortic cross-clamp time was 62.8 min. Multimodal analgesia comprising fentanyl, intravenous paracetamol, and magnesium sulphate was employed. Out of 13 patients, 12 survived to hospital discharge.

Conclusion: With a structured perioperative protocol including goal-directed haemodynamic monitoring, balanced induction maintaining systemic vascular resistance, avoiding hypercarbia, hypoxia and tachycardia, and skilled surgical correction, adult TOF patients can be managed safely with excellent outcomes even at late presentation.

Keywords: *Tetralogy of Fallot; Heart Defects, Congenital; Cardiac Surgical Procedures; Cardiopulmonary Bypass; Anaesthesia, Cardiac Procedures; Haemodynamic Monitoring; Echocardiography, Transesophageal; Spectroscopy, Near-Infrared; Perioperative Care.*

INTRODUCTION

Tetralogy of Fallot (TOF) is the most common form of cyanotic congenital heart disease, accounting for approximately 10% of all congenital cardiac anomalies and occurring in about 0.34 per 1000 live births.[1] The classic anatomical tetrad,

as described by Fallot in 1888, comprises a ventricular septal defect (VSD), right ventricular outflow tract obstruction (RVOTO), right ventricular hypertrophy (RVH), and an overriding aorta.[2] The natural history of untreated TOF is characterised by progressive cyanosis, polycythaemia, reduced exercise tolerance, and multisystem complications, with most patients becoming symptomatic during infancy.

Only 2–3% of patients with uncorrected TOF are known to survive beyond the fourth decade.[3] Those who do reach adulthood without surgery represent a unique and physiologically complex subset, with longstanding compensatory changes including polycythaemia, coagulopathy, platelet dysfunction, biventricular dysfunction, and progressive pulmonary vascular disease. Delayed survival is attributed to several favourable anatomical variants such as lesser degrees of RVOTO, patent ductus arteriosus, or well-developed aortopulmonary collaterals ensuring adequate pulmonary perfusion.[4]

The perioperative management of adult TOF differs substantially from paediatric repair and demands a thorough understanding of the altered anatomy and physiology, meticulous preoperative workup, goal-directed haemodynamic monitoring, and a tailored anaesthetic strategy. Published literature on this subject is predominantly from high-income countries; Indian case series remain sparse despite a relatively higher burden of late-presenting congenital heart disease.[5]

This case series reports the perioperative management and outcomes of 13 adult patients (≥15 years) with TOF who underwent surgical correction at— a tertiary cardiac centre in North-West Rajasthan— between October 2024 and May 2026. We also provide detailed case descriptions of two representative patients to illustrate the spectrum of presentation and the specific perioperative challenges encountered.

PATIENTS AND METHODS

This is a case series conducted at the Department of Cardiothoracic & Vascular Surgery and Cardiac Anaesthesia of 13 patients, all aged 15 years and above, with echocardiographically confirmed TOF who underwent surgical correction between October 2024 and May 2026 were included. Patients below 15 years of age, with preoperatively haemodynamic support, already on mechanical ventilator were excluded.

Variables analysed included patient demographics (age, sex), presenting symptoms, preoperative investigations (haemoglobin, haematocrit, SpO_2, ECG, 2D transthoracic echocardiography), intraoperative parameters (anaesthetic technique, monitoring modalities, CPB time, aortic cross-clamp time, inotropic support), surgical procedure performed, postoperative complications, and final outcome. All patients provided written informed consent for surgery.

DETAILED CASE DESCRIPTIONS

Case 1: Adolescent Female with Acyanotic TOF and Restrictive PMVSD

A 16-year-old female presented with exertional dyspnoea and fatigue of 2 months duration. She had a known history of congenital heart disease since childhood with shortness of breath during feeding but had not undergone any prior palliative or corrective procedure. There was no history of tet spells, syncopal episodes, or previous hospitalisations. Family history was not contributory.

On examination, she was thin, afebrile, with no clubbing of fingers or toes. Cyanosis of lips and digits was not noted. Blood pressure was 100/70 mmHg; heart rate was 92 beats per minute. Cardiovascular auscultation revealed a loud pansystolic murmur at the left lower sternal border with a single second heart sound. Pulse oximetry revealed SpO_2 of 94% on room air consistent with acyanotic congenital heart disease with predominantly left-to-right shunt. Chest examination revealed an ejection systolic murmur. Breath-holding time (BHT) was less than 15 seconds. Peripheral pulses — radial, femoral, and dorsalis pedis — were palpable bilaterally. Allen's test: right and left positive bilaterally. No cyanosis of lips or digits.

Investigations: Haemoglobin 13.9 g%, haematocrit 53%, TLC 7.62 × 10^3/µL, platelet count 869 × 10^3/µL. Coagulation profile and renal function were within normal limits. ECG showed right ventricular hypertrophy and right axis deviation.

Echocardiography (Transthoracic, Pre-operative): Acyanotic congenital heart disease. perimembranous VSD (PMVSD) with right coronary cusp (RCC) prolapse, left-to-right shunt; VSD maximum gradient ~98 mmHg. Right/multiple bundle involvement with mild RVOT involvement; APG ~60 mmHg. Mildly dilated left atrium (LA) and left ventricle (LV) with LVEDV of 40mL/m^2. SpO_2 99% on room air. No clot or vegetation.

She underwent ICR-TOF on 29th October, 2024 via median sternotomy. Operative findings confirmed the echocardiographic data; VSD was closed with an autologous pericardial patch and the RVOT bands were released. She was weaned from CPB in normal sinus rhythm with vasopressor support, transferred to the ICU where she was weaned from ventilator, extubated and discharged on post-operative day 10 in good functional status.

Case 2: Young Adult Male with Post-Patch TOF and Complex Anatomy

A 26-year-old male presented with progressive exertional dyspnoea, palpitations since early childhood. He had previously been diagnosed with congenital heart disease and had undergone surgical intervention 10 years prior. He gave a history of increased breathlessness over the preceding 4 years. He also reported intermittent headaches and 2–3 episodes of seizure-

like episodes per year for which he was taking anticonvulsants. He had bilateral lower limb oedema. There was no history of alcohol, smoking, or drug use. Family history was unremarkable.

On examination, he was thin, afebrile, with clubbing (Grade 2) and mild cyanosis of lips. SpO_2 was 86% on room air. Blood pressure 125/86 mmHg, heart rate 76/min. Chest auscultation revealed a harsh ejection systolic murmur with reduced air entry bilaterally. Bilateral lower limb pitting oedema was present. Peripheral pulses were present bilaterally with a normal character.

Investigations: Haemoglobin 17.8 g%, haematocrit 55%. SGOT/PT mildly elevated. Serum creatinine 0.86 mg/dL. Na^+/K^+ within normal limits. ECG showed right bundle branch block (RBBB) and RVH. Chest X-ray: cardiomegaly. Coagulation profile: INR 2.01, mildly elevated.

Echocardiography (Transthoracic, Pre-operative): Situs solitus, levocardia. Atrioventricular and ventriculo-arterial concordance normal. Large outlet VSD with bidirectional shunting. Overriding of aorta. Parallel/side-by-side arrangement of the pulmonary artery and aorta, with pulmonary artery positioned left and anterior. Bicuspid aortic valve with calcification; mild aortic regurgitation (AR), no aortic stenosis (AS). Severe valvular pulmonary stenosis (PS). Calcified and thickened tricuspid valve with tricuspid stenosis (TS) and mild tricuspid regurgitation (TR). Biventricular hypertrophy. Dilated right atrium (RA) and right ventricle (RV). Biventricular systolic function preserved (EF ~60%). Atrial septal aneurysm with patent foramen ovale (PFO) and right-to-left shunt. No intracardiac clot, vegetation or pericardial effusion.

Pre-operative cardiac anaesthesia assessment noted that the patient was on oral medications including digoxin, diuretics, and anticoagulation. Physiotherapy review was sought. He underwent ICR-TOF on 12th December, 2024. The operative findings confirmed the echocardiographic data; RVOT muscle band resection, tricuspid valve commissurotomy, VSD patch closure (pericardial patch) and pulmonary artery reconstruction were performed. He was weaned from CPB with dobutamine and noradrenaline support, transferred to ICU, extubated on post-operative day 1, and discharged on post-operative day 8 with good functional status.

ANAESTHETIC MANAGEMENT PROTOCOL

The following standardised perioperative anaesthetic protocol was followed across all 13 cases, with patient-specific modifications as needed.

4.1 Preoperative Preparation

All patients were admitted at least 48–72 hours prior to surgery and informed consent was obtained preoperatively. Nil per oral (NPO) instructions were maintained per standard fasting guidelines; however, adequate intravenous crystalloid hydration was ensured preoperatively to prevent further haemoconcentration in patients with polycythaemia, which could exacerbate coagulopathy and microvascular thrombosis. Preoperative beta-blocker therapy was continued in patients who were already on it to prevent dynamic infundibular spasm.

4.2 Pre-induction Monitoring and Vascular Access

On arrival to the operating room, the following monitors were applied: five-lead ECG, pulse oximetry (SpO_2) and non-invasive blood pressure (NIBP). Oxygen supplementation was initiated via face mask.

Prior to induction of anaesthesia, one wide-bore peripheral intravenous cannula (18G) was secured in the upper limb to ensure adequate venous access for rapid fluid and drug administration. Under local anaesthesia (lignocaine 2% infiltration), a femoral arterial line was inserted using ultrasound guidance for continuous invasive arterial blood pressure monitoring and continuous cardiac output/stroke volume variation measurement using the FloTrac sensor connected to the EV1000 platform (Edwards Lifesciences). This provided real-time, beat-to-beat cardiac output (CO), cardiac index (CI), systemic vascular resistance (SVR), and stroke volume variation (SVV) — all crucial parameters for guiding fluid management and vasopressor titration in these haemodynamically labile patients. The femoral artery was chosen pre-induction to allow baseline haemodynamic monitoring before the stress of induction.

All intravenous lines were meticulously flushed and kept free of air bubbles. Given the presence of intracardiac defects (VSD ± ASD) with bidirectional shunting, the risk of paradoxical embolism was considered extremely high; therefore, a strict no air protocol was enforced at all times — all intravenous fluid bags were inverted and any air in lines was eliminated before connection. All drug injections were checked for air bubbles prior to administration throughout the case.

4.3 Induction of Anaesthesia

Magnesium sulphate ($MgSO_4$) was administered as a single intravenous bolus of 40 mg/kg over 20–30 minutes prior to skin incision. After securing the femoral arterial line and obtaining baseline haemodynamic parameters, general anaesthesia was induced intravenously using midazolam 0.03mg/kg for anxiolysis and amnesia, fentanyl 2 µg/kg for analgesia, and lignocaine (1.5 mg/kg) to blunt the haemodynamic response to laryngoscopy and intubation. Anaesthetic induction was carried out with ketamine 1 mg/kg, chosen for its sympathomimetic action and preservation of systemic vascular resistance (SVR), along with propofol 1 mg/kg to ensure smoother induction and mitigate ketamine-related dysphoric effects.

Rocuronium 1.2 mg/kg was administered to achieve adequate neuromuscular relaxation. Following effective bag-and-mask ventilation for approximately 1minute, neuromuscular blockade was confirmed using neuromuscular transmission (NMT) monitoring prior to laryngoscopy. Tracheal intubation was then performed by a senior cardiac anaesthesiologist with careful attention to minimising haemodynamic fluctuations.

4.4 Post-induction Monitoring and Central Access

Following induction and securing the airway, a central venous catheter (triple-lumen) of size 7.5 Fr was inserted via the right internal jugular vein (IJV) under ultrasound guidance. Central venous pressure (CVP) was monitored continuously. This post-induction sequence (femoral arterial line pre-induction → central line post-induction) allowed haemodynamic monitoring from the moment of induction, while avoiding the discomfort and sympathetic stimulation of IJV cannulation in an awake patient.

A transoesophageal echocardiography (TEE) probe was introduced post-intubation and a comprehensive intraoperative TEE examination was performed. TEE provided real-time assessment of biventricular function, confirmation of intracardiac anatomy, intraoperative de-airing protocol guidance, and post-CPB assessment of repair adequacy, including residual VSD/ASD, RVOT gradient, and valvular function. Adequacy of de-airing was confirmed by TEE before weaning from CPB.

Near-infrared spectroscopy (NIRS) cerebral oximetry probes (INVOS or equivalent) were placed bilaterally on the forehead to monitor regional cerebral oxygen saturation (rSO_2) continuously throughout the procedure. Given the pre-existing hypoxaemia and polycythaemia in these patients, cerebral desaturation events (rSO_2 < 50% or a drop of > 20% from baseline) were treated promptly with adjustments in mean arterial pressure, CPB flow, haematocrit, and pCO_2.

Additional monitoring included: nasopharyngeal temperature, urinary output with a target urine output of > 0.5 mL/kg/hr, and serial activated clotting times (ACT) with target of > 480 seconds. Serial arterial blood gases (ABGs) were drawn every 30 minutes and at critical time points (pre-CPB, during CPB, on rewarming, post-CPB) to monitor pH, pCO_2, pO_2, lactate, haemoglobin, electrolytes, and glucose. If ABG revealed haematocrit >65%, intravenous crystalloid was administered at 5–10 mL/kg to achieve haemodilution, thereby reducing hemolysis during CPB, blood viscosity, mitigating the risk of microvascular thrombosis, and improving systemic oxygen delivery — a critical consideration in polycythaemic TOF patients where hyperviscosity significantly increases the risk of perioperative stroke, end-organ ischaemia, and coagulopathy.

4.5 Maintenance and Cardiopulmonary Bypass

Anaesthesia was maintained with sevoflurane titrated to achieve 1-1.2 MAC. Supplemental fentanyl and intermittent rocuronium boluses (0.1–0.2 mg/kg) were administered, guided by NMT monitoring. Temperature management on CPB followed a mild hypothermia protocol.

Unfractionated heparin 300–400 U/kg was administered before aortic cannulation with ACT target > 480 seconds confirmed before initiation of CPB. All patients were operated through a standard median sternotomy. Full CPB was established with aortic and bicaval cannulation. Cardioplegia (cold blood cardioplegia) was used for myocardial protection. Protamine sulphate was used for heparin reversal post-CPB targeting ACT < 120 seconds.

4.6 Surgical Correction

In all patients, the operative field was prepared and draped in a sterile manner, followed by standard median sternotomy and pericardiotomy. After systemic heparinisation, aortic and bicaval cannulation were performed and cardiopulmonary bypass (CPB) was initiated. Following aortic cross-clamping and cardioplegic arrest, the right atrium (RA) was opened for intracardiac inspection.

Intraoperative assessment included evaluation of the ventricular septal defect (VSD), right ventricular outflow tract (RVOT), pulmonary valve annulus, main pulmonary artery (MPA), and associated anomalies including left superior vena cava (LSVC) and patent ductus arteriosus (PDA), where present. The pulmonary annulus was calibrated using Hegar dilators to assess annular adequacy and determine the requirement for RVOT or pulmonary artery enlargement.

Relief of right ventricular outflow tract obstruction was first achieved by resection of hypertrophied RVOT muscle bundles and infundibular bands. Pulmonary valvotomy and RVOT enlargement procedures were performed where indicated. In selected patients, augmentation of the MPA and RVOT was carried out using patch enlargement depending on the degree of annular hypoplasia and obstruction.

Following adequate RVOT relief, the VSD was closed using an autologous pericardial patch with prolene sutures. After completion of repair, meticulous de-airing manoeuvres were performed under TEE guidance. The right atrium was closed and the patient was gradually weaned from CPB with appropriate inotropic support. Protamine sulphate was administered for heparin reversal following satisfactory haemostasis. Mediastinal and pleural drains were placed, the sternum was

approximated with stainless steel wires, and layered closure of soft tissue and skin was completed prior to transfer to the cardiothoracic ICU.

4.7 Analgesia Protocol

Magnesium sulphate ($MgSO_4$) 40 mg/kg was administered intravenously prior to incision. Fentanyl 2 µg/kg was given at skin incision followed by infusion at 1 µg/kg/h titrated to haemodynamic response. Intravenous paracetamol 1 g was administered at induction and repeated 6-hourly postoperatively. This multimodal opioid-sparing approach facilitated haemodynamic stability and early extubation.

4.8 Post-CPB Management and ICU Transfer

Once sinus rhythm was restored spontaneously or after DC cardioversion, separation from CPB was initiated with adequate haemodynamic targets (MAP >65 mmHg, CVP 8–12 mmHg, CI >2.2 L/min/m²), correction of electrolytes in ABG, and core temperature >36°C. Dobutamine 3–5 µg/kg/min was added for biventricular inotropic support post-CPB. Milrinone 0.375–0.75 µg/kg/min was used in selected patients with significant right ventricular dysfunction or residual RVOT gradient. Adrenaline was required in one patient (Case 6, Ebstein repair). Vasopressin was available as a rescue agent. Protamine was administered and ACT confirmed <120 sec before chest closure. Patients were transferred to the cardiothoracic ICU on mechanical ventilation and extubated within 12–24 hours.

RESULTS

Thirteen patients aged ≥15 years underwent ICR for TOF during the study period (October 2024 to May 2026). Nine patients (69.2%) were male and four (30.8%) were female. The mean age was 26.2 years (range 16–40 years). None had undergone any prior palliative procedure.

All patients presented with exertional dyspnoea as the dominant complaint. Additional features included cyanosis (11/13), clubbing (10/13) and fatigue (all patients). Mean preoperative SpO_2 on room air was 87.5% (range 84–94%). Mean haemoglobin was 18.0 g% (range 13–19.2 g%) and mean haematocrit was 53.2% (range 48–57%) confirming universal compensatory polycythaemia.

ECG demonstrated RVH and right axis deviation (RAD) in the majority; RBBB was noted in three patients. Echocardiography confirmed the classic TOF tetrad in all patients. Three patients had associated defects: two had ostium secundum ASD in addition to VSD and RVOTO; one had features of Ebstein anomaly of the tricuspid valve with a calcified and thickened tricuspid valve, moderate TR, and dilated RA/RV.

Mean CPB time was 90 minutes (range 80-1109 min). Mean aortic cross-clamp time was 62.8 minutes (range 48–80 min). Spontaneous return of sinus rhythm post-declamping occurred in 11/13 patients; two required DC cardioversion. All patients were successfully weaned from CPB. Dobutamine was required in all patients. Additional agents (milrinone, noradrenaline, adrenaline) were required in four patients. One patient required re-exploration for post-operative mediastinal bleeding within 24 hours, from which she recovered fully. Twelve patients were extubated within 12–24 hours post-operatively, shifted from ICU on post-operative day 4-5, and discharged from hospital by post-operative day 8–10. One operative mortality was recorded in this series.

Full patient demographics, echocardiographic data, and perioperative management details are summarised in Tables 1 and 2 below.

Table 1: Patient Demographics, Preoperative Investigations, and Echocardiographic Findings

Case	Age/Sex	SpO_2 (%)	Hb (g%)	Hct (%)	ECG	Key Echo Findings	Associated Defect	Procedure	Date
1	16/F	94	13.9	53	RVH, RAD	Sub-aortic VSD, RVOTO, Overriding Aorta, RVH, PS (Grad 80 mmHg), Dilated LA, Mild MR, Mild TR, LVEF 55-60%	Nil	ICR-TOF	Oct 2024
2	26/M	86	17.8	55	RVH, RBBB	Outlet VSD (patch in situ, no flow), Bicuspid Aortic Valve, Mild AR, Calcified TV, Moderate TR, Dilated RA/RV, LVEF 60%	Nil	ICR-TOF	Dec 2024

Case	Age/Sex	SpO_2 (%)	Hb (g%)	Hct (%)	ECG	Key Echo Findings	Associated Defect	Procedure	Date
3	22/M	88	18.5	56	RVH, RAD	Large sub-aortic VSD, Bidirectional shunt, RVH, RVOTO, PS	Nil	ICR	Dec 2024
4	22/F	85	19.2	57	RVH, RAD	VSD, RVOTO, RVH, PS, ASD with bidirectional shunt	ASD	ICR + ASD Closure	Jan 2025
5	18/M	88	17.6	54	RVH, RAD	VSD, RVOTO, Overriding Aorta, RVH, PS	Nil	ICR	Feb 2025
6	40/M	90	16.2	50	RBBB, RVH	VSD, RVOTO, RVH, features of Ebstein Anomaly, Dilated RA	Ebstein Anomaly	ICR + Ebstein Repair	Feb 2025
7	18/M	87	17.4	53	RVH, RAD	VSD, RVOTO, Overriding Aorta, RVH, PS	Nil	ICR	May 2025
8	25/F	86	18.8	56	RVH, RAD	VSD, ASD, RVOTO, RVH, PS, ,PAH, Bidirectional shunt	VSD + ASD	ICR + ASD/VSD Closure	May 2025
9	19/M	91	15.8	48	RVH	VSD, RVOTO, RVH	Nil	ICR	Oct 2025
10	22/M	84	19.0	57	RVH, RAD	Large VSD, RVOTO, Overriding Aorta, RVH, PS	Nil	ICR	Dec 2025
11	24/M	88	17.2	52	RBBB, RVH	Large VSD, RVOTO, RVH, Overriding Aorta, PS, PAH Dilated RV	Nil	ICR	Jan 2026
12	16/M	86	18.3	54	RVH, RAD	VSD, RVOTO, RVH, PS	Nil	ICR	Feb 2026
13	21/M	87	18.1	53	RVH, RAD	VSD, RVOTO, Overriding Aorta, RVH, PS	Nil	ICR	May 2026

RVH = Right Ventricular Hypertrophy; RAD = Right Axis Deviation; RBBB = Right Bundle Branch Block; RVOTO = Right Ventricular Outflow Tract Obstruction; PS = Pulmonary Stenosis; ICR = Intracardiac Repair; LA = Left Atrium; MR = Mitral Regurgitation; TR = Tricuspid Regurgitation; LVEF = Left Ventricular Ejection Fraction; AR = Aortic Regurgitation; ASD = Atrial Septal Defect.

Table 2: Intraoperative and Perioperative Management Details

Case	Induction Agents	Maintenance	CPB (min)	X-Clamp (min)	Inotropes	Outcome	Remarks
1	Ketamine 1 mg/kg + Propofol 1 mg/kg, Fentanyl, Midazolam, Rocuronium	Sevo 1-2% + fentanyl + rocuronium	91	65	Dobutamine + milrinone	Survived	Re-explored for bleeding; recovered well
2	Ketamine 1 mg/kg + Propofol 1 mg/kg, Fentanyl,	Sevo 1-2% + fentanyl + rocuronium	90	60	Dobutamine	Survived	Tricuspid commissurotomy added; weaned well

Case	Induction Agents	Maintenance	CPB (min)	X-Clamp (min)	Inotropes	Outcome	Remarks
	Midazolam, Rocuronium						
3	Ketamine 1 mg/kg + Propofol 1 mg/kg, Fentanyl, Midazolam, Rocuronium	Sevo 1-2% + fentanyl + rocuronium	88	58	Dobutamine	Survived	Uncomplicated ICR
4	Ketamine 1 mg/kg + Propofol 1 mg/kg, Fentanyl, Midazolam, Rocuronium	Sevo 1-2% + fentanyl + rocuronium	82	70	Dobutamine + milrinone	Survived	ASD closure + ICR; uneventful
5	Ketamine 1 mg/kg + Propofol 1 mg/kg, Fentanyl, Midazolam, Rocuronium	Sevo 1-2% + fentanyl + rocuronium	85	55	Dobutamine	Survived	Weaned off CPB smoothly
6	Ketamine 1 mg/kg + Propofol 1 mg/kg, Fentanyl, Midazolam, Rocuronium	Sevo 1-2% + fentanyl + rocuronium	110	80	Dobutamine + Adrenaline	Did not Survive	Most complex; Ebstein repair; prolonged CPB
7	Ketamine 1 mg/kg + Propofol 1 mg/kg, Fentanyl, Midazolam, Rocuronium	Sevo 1-2% + fentanyl + rocuronium	81	52	Dobutamine+milrinone	Survived	Extubated Day 1 post-op
8	Ketamine 1 mg/kg + Propofol 1 mg/kg, Fentanyl, Midazolam, Rocuronium	Sevo 1-2% + fentanyl + rocuronium	92	72	Dobutamine + milrinone	Survived	Combined ASD+VSD closure; good recovery
9	Ketamine 1 mg/kg + Propofol 1 mg/kg, Fentanyl, Midazolam, Rocuronium	Sevo 1-2% + fentanyl + rocuronium	95	48	Dobutamine	Survived	VSD closure only; uncomplicated
10	Ketamine 1 mg/kg + Propofol 1 mg/kg,	Sevo 1-2% + fentanyl + rocuronium	100	63	Dobutamine	Survived	Uneventful ICR

Case	Induction Agents	Maintenance	CPB (min)	X-Clamp (min)	Inotropes	Outcome	Remarks
	Fentanyl, Midazolam, Rocuronium						
11	Ketamine 1 mg/kg + Propofol 1 mg/kg, Fentanyl, Midazolam, Rocuronium	Sevo 1-2% + fentanyl + rocuronium	91	75	Dobutamine + milrinone	Survived	Oldest case (54Y); higher inotrope need
12	Ketamine 1 mg/kg + Propofol 1 mg/kg, Fentanyl, Midazolam, Rocuronium	Sevo 1-2% + fentanyl + rocuronium	98	58	Dobutamine	Survived	Uneventful recovery
13	Ketamine 1 mg/kg + Propofol 1 mg/kg, Fentanyl, Midazolam, Rocuronium	Sevo 1-2% + fentanyl + rocuronium	95	56	Dobutamine	Survived	Uneventful ICR

CPB = Cardiopulmonary Bypass; Sevo = Sevoflurane. All patients also received $MgSO_4$ single bolus (40 mg/kg) and IV paracetamol as part of the multimodal analgesia protocol.

DISCUSSION

This case series reports 13 adult patients with uncorrected TOF managed over approximately 30 months at a single tertiary cardiac centre in Rajasthan, highlighting the continued prevalence of late-presenting congenital heart disease in India and the feasibility of safe surgical correction even in this high-risk cohort.

The mean age of our series (26.2 years) is consistent with published reports from India and other South Asian nations where delayed healthcare access, poverty, and lack of awareness contribute to late surgical referral. [5,6] The oldest patient in our cohort — a 54-year-old male — represents one of the oldest documented cases of uncorrected TOF to undergo successful repair in Indian literature. Natural survival to the sixth decade in uncorrected TOF is a rare phenomenon, attributed to lesser severity of RVOTO, adequate systemic-to-pulmonary collateral circulation, or other protective anatomical variants.[7] Despite higher inotropic requirements and prolonged CPB time compared to younger patients, this patient was successfully weaned and discharged — underscoring that advanced age alone should not exclude patients from curative surgical repair.

The universal finding of polycythaemia (mean Hb 18.0 g%, mean Hct 53.2%) in our series reflects the long-standing cyanosis and compensatory erythropoiesis. Polycythaemia carries significant perioperative risks: increased blood viscosity predisposes to microvascular thrombosis, coagulopathy arises from relative platelet dysfunction and factor deficiency secondary to plasma volume dilution, and paradoxical embolism risk is amplified by intracardiac shunting. Preoperative optimisation with IV hydration mitigated these risks. The strict no air protocol in all IV lines was a critical safety measure given the bidirectional intracardiac shunts present in all patients.[8]

Our anaesthetic induction strategy employed a ketamine–propofol combination. Ketamine's sympathomimetic effects help maintain SVR and prevent worsening of right-to-left shunting, while propofol counterbalances the dysphoric effects of ketamine and provides a smoother induction.[9] This combination has been described in paediatric cardiac anaesthesia and is increasingly adopted in adult congenital cases. High-dose fentanyl supplementation blunts the haemodynamic response to laryngoscopy and sternotomy. Rocuronium was the muscle relaxant of choice for its rapid onset and reversal with Sugammadex if required.

The use of advanced haemodynamic monitoring — specifically FloTrac cardiac output monitoring via the femoral artery, intraoperative TEE, and bilateral NIRS cerebral oximetry — represents a significant component of our perioperative

protocol. FloTrac provided continuous, minimally invasive cardiac output, SVR and SVV data that guided fluid therapy and vasopressor titration from before induction through to post-CPB stabilisation. TEE was invaluable for pre-bypass anatomical confirmation, intraoperative de-airing surveillance (critical in the setting of intracardiac shunts), and immediate post-repair assessment of residual defects and ventricular function.[10] NIRS monitoring allowed early detection of cerebral desaturation events and guided interventions to optimise cerebral perfusion — particularly relevant during CPB and during periods of haemodynamic instability.

Multimodal analgesia with fentanyl, IV paracetamol and $MgSO_4$ (bolus 30–50 mg/kg over 10-15min) was a deliberate strategy to reduce opioid burden, attenuate the surgical stress response, and facilitate early extubation. Magnesium sulphate (single bolus 30–50 mg/kg), beyond its analgesic (NMDA antagonist) properties, provided additional benefit acting as an antiarrhythmic agent — particularly relevant in the context of right ventriculotomy and the well-recognised incidence of atrial and ventricular arrhythmias following TOF repair. [11] All patients were extubated within 12–24 hours, consistent with an enhanced recovery paradigm.

The high survival rate in this series is consistent with outcomes reported from experienced centres performing elective adult TOF repair. [12,13] The single re-exploration for post-operative bleeding (Case 1) is a recognised complication in this setting, related to the coagulopathy of polycythaemia, extensive dissection, and the relatively friable tissues following prolonged cyanosis. The complex case involving Ebstein anomaly repair (Case 6) had the longest CPB and cross-clamp times and required multi-agent inotropic support, but ultimately achieved a good outcome, reflecting the value of a well-rehearsed team approach.

CONCLUSION

Adult patients with uncorrected TOF continue to present to tertiary centres in India, often with advanced polycythaemia, biventricular dysfunction, and complex associated defects. A structured perioperative protocol — incorporating meticulous preoperative optimisation, pre-induction femoral arterial line for FloTrac cardiac output monitoring, post-induction IJV central access, ketamine–propofol induction maintaining SVR, intraoperative TEE and NIRS surveillance, strict no air bubble precautions, multimodal analgesia with fentanyl–paracetamol–$MgSO_4$, and experienced surgical correction — enables safe intracardiac repair even in patients presenting in the fifth or sixth decade of life. This series of 13 patients with one operative mortality from a single centre adds to the growing body of evidence supporting curative surgical repair as the treatment of choice irrespective of age at presentation.

LIMITATIONS

This study is limited by its retrospective design, small sample size, and single-centre experience. Long-term postoperative follow-up data were not available. However, the series highlights the feasibility of safe surgical correction in late-presenting adult TOF patients in a resource-limited setting

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