



Anaesthetic Challenges and Transesophageal Echocardiography-Guided Perioperative Management in a Patient with Uncorrected Adult Congenital Heart Disease Presenting for Emergency Laparoscopic Hysterectomy

Niranjan Dilip Waje¹ , Nilay Chatterjee² , Nishant Ram Arora³ , Vijay Kumar⁴

¹Cardiothoracic and Liver Transplant Unit, Reliance Hospital, Kopar Khairane, Navi Mumbai, India

²Department of Anaesthetics, Yeovil District Hospital, Higher Kingston, Yeovil, United Kingdom

³Department of Cardiac Anaesthesia, National Heart Centre, The Royal Hospital, Muscat, Oman

⁴Clinical Fellow in Anaesthesia, The Walton Centre NHS Foundation Trust, Lower Lane, Fazakerley, Liverpool, United Kingdom

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Abstract

Anaesthetic management of a patient with adult congenital heart disease with a single ventricle physiology presenting for an emergency laparoscopic surgery is challenging. The importance of a multidisciplinary approach, astute understanding of the pathophysiology and optimisation of intraoperative hemodynamic goals cannot be overemphasised. The present report describes the anaesthetic challenges and the role of transoesophageal echocardiography in perioperative management of a patient with uncorrected tetralogy of Fallot with pulmonary atresia, who successfully underwent an emergency laparoscopic hysterectomy under general anaesthesia.

Keywords: Anaesthesia, congenital heart disease, echocardiography, hemodynamics, laparoscopic surgery, perioperative care, transesophageal

Introduction

Congenital heart disease (CHD) occurs in 0.4%–5% of all live births (1). Tetralogy of Fallot (TOF) with pulmonary atresia (PA) comprises 1%–2% of all CHD. Less than 3% of patients with adult congenital heart disease (ACHD) with uncorrected TOF and PA survive until the fourth decade of life (2).

In this case report, authors describe the anaesthetic challenges and role of transoesophageal echocardiography (TEE) in the perioperative management of a patient of uncorrected ACHD with unique pathophysiology, who successfully underwent an emergency laparoscopic hysterectomy under general anaesthesia. This manuscript adheres to applicable Enhancing the QUALity and Transparency Of health Research guidelines (EQUATOR). Written consent was obtained from the patient before writing this report.

Case Presentation

A 44-year-old lady presented to the emergency department (ED) with complaints of uncontrolled menorrhagia. She was haemodynamically unstable with a heart rate of 105/min, blood pressure of 95/55 mmHg and room air oxygen saturation by pulse oximetry (SpO₂) of 78%. She was a known case of ACHD (uncorrected TOF and PA with pulmonary flows supported by multiple major aorto-pulmonary collateral arteries (MAPCAs) in a single ventricle physiology). Previous cardiac catheterization reported 50% overriding of aorta, atretic origin of main pulmonary

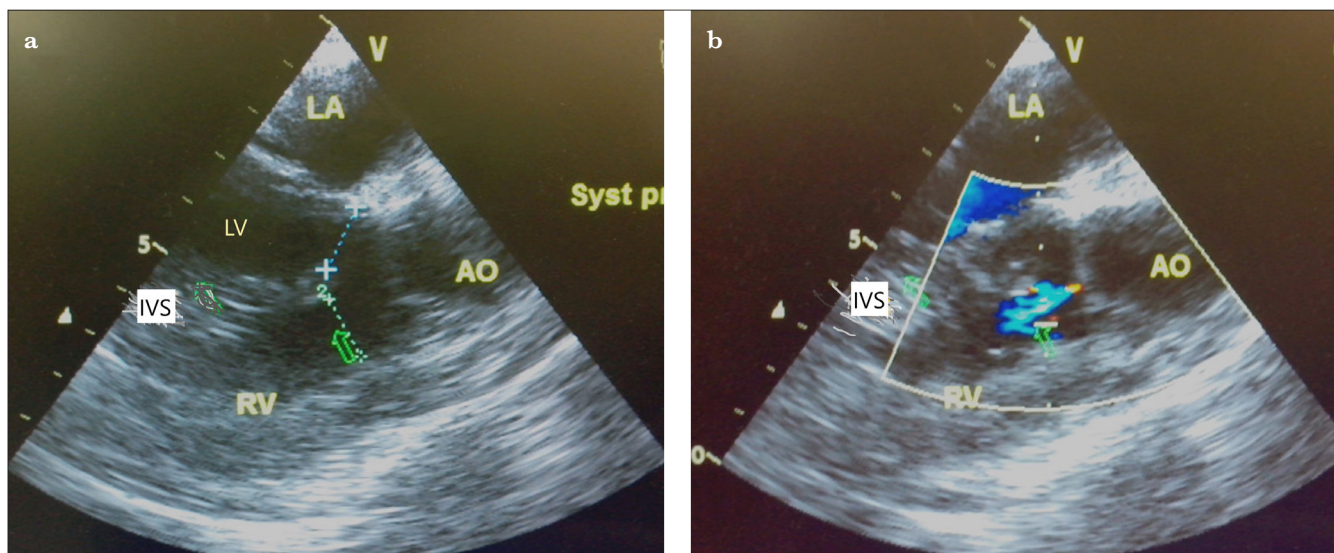


Figure 1. a, b. (a) Mid-oesophageal long axis view showing large ventricular septal defect, overriding of aorta and non-dilated left ventricle. (b) mid-oesophageal long axis with colour Doppler showing trivial atrial regurgitation
LA: left atrium; AO: aorta; LV: left ventricle; RV: right ventricle; IVS: inter-ventricular septum

artery (MPA), narrowed distal MPA, normal right pulmonary artery and hypoplastic left pulmonary artery. The gradient across the severely stenosed pulmonic valve was 140 mmHg. Pulmonary flow was achieved by a single right-sided and three large, tortuous and narrowed left-sided aorto-pulmonary collaterals. A thoracic echocardiography (TTE) showed situs solitus, atrio-ventricular and ventriculo-arterial concordance, infundibular stenosis, overriding of aorta, large sub-aortic ventricular septal defect (VSD), hypertrophied right ventricle (RV), normal left ventricle (LV) with good biventricular function, and normal systemic and pulmonary venous drainage. Electrocardiography (ECG) showed a normal sinus rhythm and right axis deviation. Previously, she displayed good exercise tolerance. One year ago, she successfully tolerated a full-term vaginal delivery and an uneventful postnatal course. These findings suggested a naturally developed compensated pathophysiology. Before her current presentation with men-

orrhagia and compromised haemodynamics, she was gradually developing a gradually worsening dyspnoea (New York Heart Association grade II); over the past 8 months. She was reviewed by a multidisciplinary team consisting of ACHD cardiologist, cardiac surgeon, cardiac intensivist and a cardiac anesthesiologist to offer her an elective total corrective surgery. The present episode occurred while she was waiting for her elective surgery. She was resuscitated and stabilized in the ED with fluids. Oxygen was administered through facemask, blood samples were sent for laboratory tests, grouping, cross matching. A comprehensive TTE was performed. Her SpO₂ improved with higher mean arterial pressures (MAP) during her stabilization. Successful management of laparoscopic surgery in a patient with uncorrected ACHD with single ventricle physiology has been described in the literature (3). Considering all the concerning factors, the multidisciplinary team concluded that laparoscopic hysterectomy would be safe for her to proceed (3), keeping a low threshold to convert to laparotomy based on the prevalent clinical conditions.

Main Points:

- It is extremely important to understand the pathophysiology of adult uncorrected congenital heart disease (ACHD) in order to make a robust peri operative plan for emergency non cardiac surgery.
- Setting realistic targets of adequate hemodynamic parameters in a critically balanced physiology, as in a case of an uncorrected ACHD.
- Peri Operative TEE serves as an important tool in management of cardiac cases for non cardiac surgery.
- A multidisciplinary approach is highly emphasized in the management of a critically balanced circulation in uncorrected ACHD, challenged by the risk of a laparoscopic surgery in an emergency setting.

Perioperatively, monitoring consisted of SpO₂, end tidal carbon dioxide (EtCO₂), ECG, arterial blood pressure, central venous pressure, central venous oxygen saturation (ScvO₂), temperature and TEE. General endotracheal anaesthesia was administered with 15 mg etomidate, 100 mg rocuronium, and 100 mcg fentanyl. Adequate anaesthesia and analgesia were maintained with volatile anaesthetic (sevoflurane), intermittent boluses of cisatracurium, fentanyl infusion, paracetamol and port-site skin infiltration with 0.25% bupivacaine. Prophylactic antibiotics, vitamin K and a bolus of 10 mg kg⁻¹ of tranexamic acid were administered before the skin incision. Intravenous line filters were applied to prevent air em-

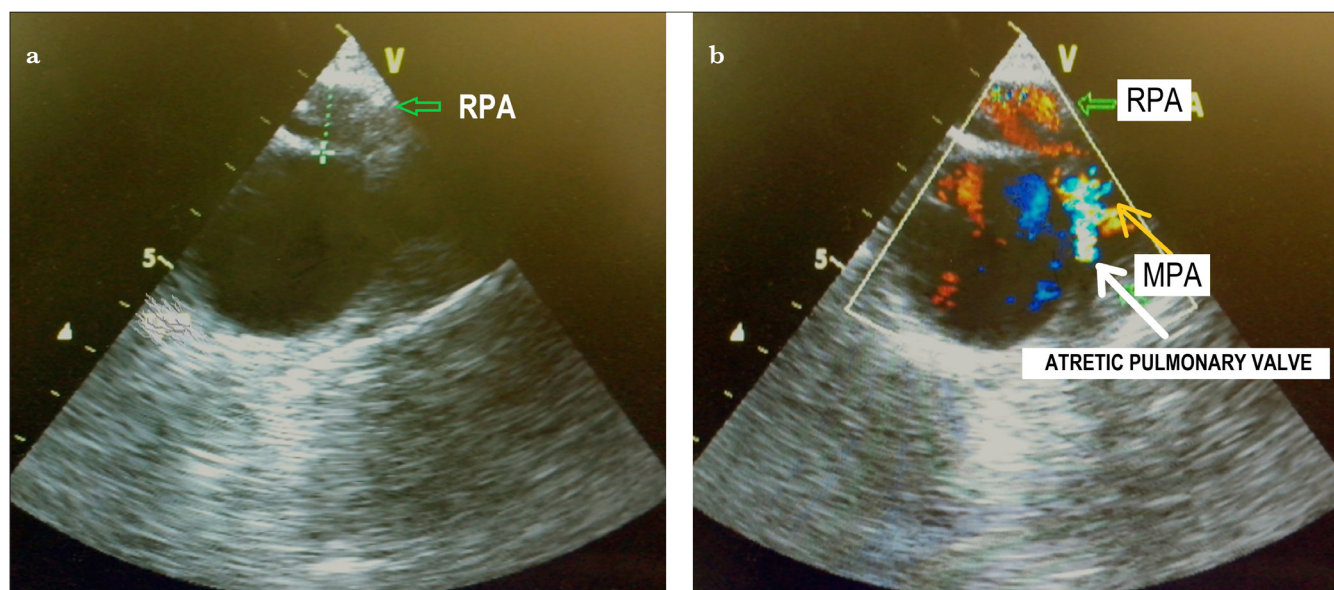


Figure 2. a, b. (a) Upper oesophageal ascending aorta short axis view showing well-formed RPA (RPA: right pulmonary artery, green open arrow); (b): upper oesophageal ascending aorta short axis view with colour Doppler showing atric pulmonic valve and proximal MPA, with trivial turbulent flow across atric pulmonic valve and good flow across RPA (MPA: main pulmonary artery, yellow solid arrow; atric pulmonary valve, white solid arrow)

bolism. Perioperative haemodynamic goals were maintained with the following: (i) maintenance of optimal MAP (close to the baseline) using appropriate vasopressors (phenylephrine at induction, intraoperative infusion of vasopressin IV; not exceeding 2.4 units hr^{-1}), (ii) maintenance of cardiac output by TEE-guided optimisation of LV preload and contractility, (iii) intra-abdominal pressures (IAP) not exceeding 12 mmHg, (iv) arterial blood gas-guided appropriate oxygenation and ventilation strategies to titrate pulmonary blood flow (Q_p) and systemic blood flow (Q_s), and (v) adequate haemoglobin (Hb) levels close to baseline (in her naturally compensated physiology). The haemodynamic goals were set to maintain arterial oxygen saturation (SaO_2) between 70% and 75%, ScvO_2 50%–55%, a SaO_2 - ScvO_2 gradient of less than 25, Hb not less than 15 g dL^{-1} and serum lactates not exceeding 2 mmol L^{-1} . Table 1 displays perioperative haemodynamic and laboratory data at various stages of surgery. Increase in the intraoperative level of lactates responded to TEE-guided preload augmentation, packed red blood cells (PRBC) transfusion and vasopressin-aided maintenance of MAP. Normothermia was maintained throughout the perioperative period using a heating mattress and fluid warming devices. Obligatory fluid and blood losses (300 mL) were replaced with a combination of crystalloids (plasmalyte), colloids and PRBC (ensuring a haematocrit close to the baseline) and an acceptable urine output (Table 1). At the end of surgery, the patient was successfully extubated in the theatre. Subsequently, she was transferred to the intensive care unit (ICU) for further observation and monitoring. ICU management included analgesia (paracetamol), aggressive spirometry and early ambulation. Her haemody-

namics remained favourable, and she was transferred to the ward on second postoperative day. She had an uneventful recovery.

Discussion

RV hypertrophy, unrestrictive VSD, overriding of aorta and RV outflow tract obstruction in form of PA characterizes TOF with PA. In such physiology, the intracardiac right to left shunt is fixed. The trivial flow across the pulmonary valve is independent of the relative changes in systemic vascular resistance (SVR): pulmonary vascular resistance (PVR) ratio. Pulmonary flow (Q_p) (primarily contributed by MAPCAs arising from the aorta) is affected by changes in PVR (4). The morphology and complexity of the pulmonary vasculature determines clinical presentation, and at times, this may not favour a surgical correction (5, 6).

TOF with PA is a classic example of functional single ventricle physiology. Large unrestricted VSD ensures uniform mixing of blood across well-formed left and right ventricles. The same level of oxygen saturation (SaO_2) is maintained in systemic (Q_s : exiting the aorta; determining the cardiac output (CO)) and pulmonary circulation (Q_p reaching the lungs through the MAPCAs). The thick-walled LV primarily responsible for aortic flows is considered as the systemic ventricle further in this report. The extent of Q_p and Q_s are governed by PVR and SVR, respectively (5). MAPCAs are notoriously known to develop ostial stenosis; thus severely compromising Q_p ($Q_p:Q_s < 1$) resulting in cyanosis. Rarely

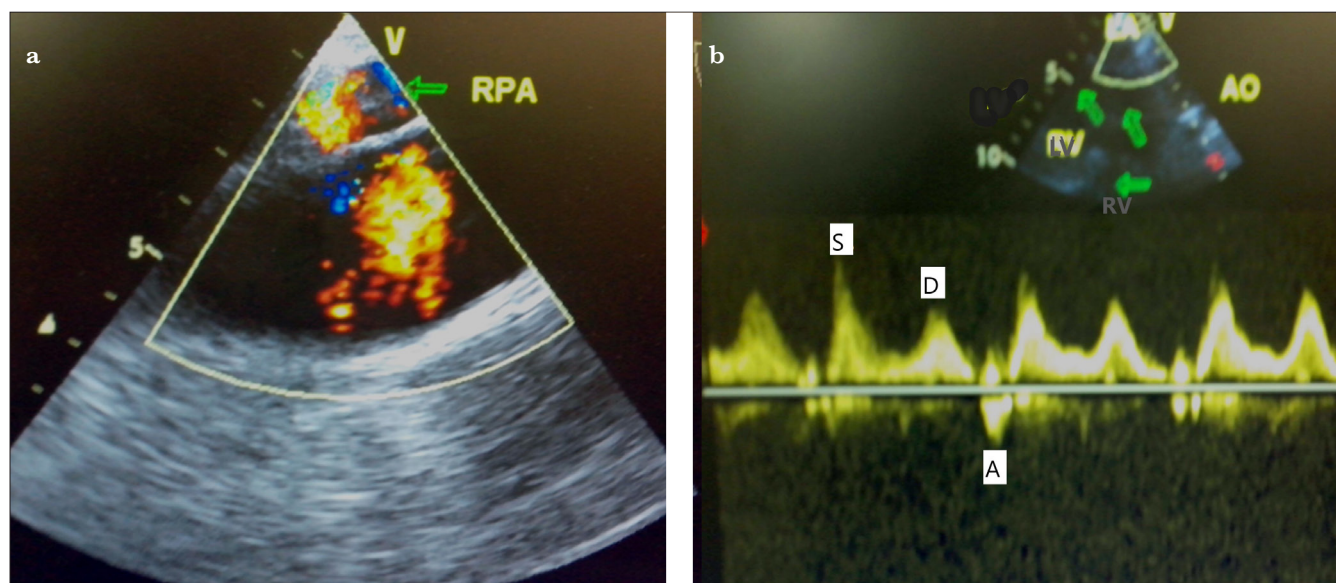


Figure 3. a, b. (a) Upper oesophageal ascending aorta long axis with colour Doppler showing good flow in right pulmonary artery (perioperative); (b) mid-oesophageal 2 chamber with pulsed wave Doppler across left upper pulmonary vein showing good intraoperative (left) pulmonary flow with normal patterns of 'S', 'D' and 'A' waves (S>D)
S: systolic; D: diastolic; A: atrial reversal waves of pulmonary venous flow

(as happened in this particular patient), mild ostial stenosis of MAPCAs may even protect the compliant pulmonary vasculature from uninterrupted high aortic flows, thereby potentially delaying the onset of pulmonary vascular occlusive disease and ensuring a balanced flow ($Q_p:Q_s$ close to unity). This might possibly explain the relatively longer life expectancy, symptom-free period and reasonable exercise tolerance in this patient. Similarly, the absence of severe aortic regurgitation (AR) ensured normal biventricular size and function (7). A normal QRS was an encouraging sign towards a favourable perioperative outcome (8).

Patients with cyanotic CHD are known to have altered clotting profile (increased fibrinolysis, deficiency of Von Willebrands multimers, factor V and vitamin K-dependent clotting factors) (6). This supports perioperative use of vitamin K, tranexamic acid and fresh frozen plasma (FFP), and avoidance of epidural analgesia. Principles of perioperative management for this patient with single ventricle physiology included optimization of systemic oxygen saturation (SaO_2) and tissue perfusion (CO), both of which were achieved by maintaining a $Q_p:Q_s$ balance close to unity. At $Q_p:Q_s > 1$, systemic tissue perfusion would have been compromised in spite of a relatively increased SaO_2 , whereas at $Q_p:Q_s < 1$, tissue perfusion might be adequate with a deficiency in the oxygen content of the blood (5). A brief period of higher oxygen saturations (SpO_2 94%) was achieved intraoperatively with higher MAP (increased Q_p). However, a $Q_p:Q_s$ imbalance was evidenced by raised lactates and acidosis. Acidosis improved by administering sodium bicarbonate and improving the oxygen delivery

(by maintaining haematocrit closer to the baseline by PRBC transfusions and TEE-guided preload augmentation of CO in a well contracting LV, thus increasing Q_s). The target Hb was aimed at 15–16 g dL⁻¹ (guided by markers of tissue perfusion) for this patient with single ventricle ACHD physiology at risk of active haemorrhage during surgery. A balanced $Q_p:Q_s$ was achieved perioperatively by maintaining their surrogate markers within realistic limits of therapeutic targets, for example, SaO_2 between 70% and 75%, PaO_2 between 50 and 55 mmHg, $ScvO_2 > 50\%$ –55% (<30% critical threshold for anaerobic metabolism), SaO_2 - $ScvO_2 < 25\%$ –30% (5) and serum lactates not exceeding 2 mmol L⁻¹ (Table 1).

Intraoperative TEE confirmed bidirectional VSD, 50% overriding of aorta, pulmonary atresia and trivial AR (Figures 1, 2a, 2b). Mid-oesophageal long axis (ME LAX 2D; Figure 1a) and transgastric mid-papillary short axis (TG mid-papillary SAX 2D; video 1, supplementary digital content) views were used to monitor the preload and biventricular function. TEE-guided augmentation of CO and maintenance of appropriate MAP by using vasopressin (for optimum coronary, pulmonary and end-organ perfusion) (9) ensured optimal Q_p . Qualitative adequacy of bilateral pulmonary perfusion (Q_p) was reflected by right pulmonary arterial (Figures 2a and 3a) and left pulmonary venous flows (Figure 3b). Q_p was further titrated by maintaining PaO_2 , $PaCO_2$ (<45 mmHg) within an optimal range with appropriate ventilation strategies (peak airway pressure less than 30 cm of H_2O , positive end expiratory pressure (PEEP) 5 cm of H_2O , tidal volume 6–8 mL kg⁻¹ of ideal body weight and an appropriate respiratory rate

Table 1. Perioperative hemodynamic data

Time points (time from beginning)	In emergency department (0 min)	Immediate pre- induction (45 min)	Immediate post- induction (75 min)	At 30 mins of laparoscopy (130 min)	At 60 mins of laparoscopy (160 min)	Immediately before extubation (220 min)	Immediately after extubation (255 min)	In ICU (6 hr)
HR min ⁻¹	105	93	88	102	68	73	95	65
MAP (mmHg)	67	92	105	75	65	77	80	82
SpO ₂ (%)	78	90	93	86	82	84	88	90
EtCO ₂ (mmHg)			29	27	30	28		
CVP (mmHg)		5	5	10	7	9	8	7
SaO ₂ (%)	82	85	90	80	77	77	82	84
PaO ₂ (mmHg)	54	58	59	57	53	57	55	55
PaCO ₂ (mmHg)	30	33	38	43	48	32	39	41
pH	7.35	7.38	7.4	7.32	7.25	7.32	7.35	7.39
HCO ₃ (mmol L ⁻¹)	20	21	19	17	16	22		22
Hb (g dL ⁻¹)	13.5	13.8	14.3	15.1	14.7	15.3	16	15.6
INR	1.54							1.22
PT (seconds)	17							14
PTT (seconds)	75							62
ScvO ₂ (%)		64		56	50	60		67
(SaO ₂ -ScvO ₂)		21		24	27	17		17
S lactate (mmol L ⁻¹)	1.9	1.8	2	2.9	4.0	2.5	2.2	1.8
Urine output (mL)		70		100		100	40	70
PAW (cm of H ₂ O)			17	25	28	18		
TV (mL)			450	350	350	450		
RR	18 (spont)		14	14	18	15		15 (spont)
PEEP (cm of H ₂ O)			5	5	5	5		
LV contractility	Good (TTE)		Good (TEE)	Good (TEE)	Good (TEE)	Good (TEE)		Good (TTE)
LVEDA	NR		NR	R, kissing papillary	R	NR		
Intervention(s)	• Art line • Fluid	• CVC • PE • Fluid	• PRBC • PE • Vitamin K • Tranexamic acid	• IAP< 12 • VP 1.2 • FFP • Fluid	• IAP< 12 • VP 2.0 • NaHCO ₃ • PRBC • FFP	• VP 1.2 • Fluids • Plasmalyte	• VP 0.5 • Fluids • Plasmalyte	• Spirometry • O ₂ 5 L min ⁻¹ • Fluids • PCM

HR: heart rate; MAP: mean arterial pressure; EtCO₂: end tidal carbon dioxide; CVP: central venous pressure; Hb: haemoglobin; INR: international normalized ratio; PaO₂: partial pressure of oxygen; PaCO₂: partial pressure of carbon dioxide; pH: power of hydrogen; PT: prothrombin time; PTT: partial thromboplastin time; ScvO₂: central venous oxygen saturation; S lactate: serum lactate; PAW: peak airway pressure; TV: tidal volume; RR: respiratory rate; spont: spontaneous; PEEP: positive end expiratory pressure; TTE: trans thoracic echocardiography; TEE: trans oesophageal echocardiography; LVEDA: left ventricular end-diastolic area; NR: not reduced; R: reduced; Art line: arterial line; CVC: central venous catheterisation; PE: phenylephrine; PRBC: packed red blood cells; IAP: intra-abdominal pressure; FFP: fresh frozen plasma; VP: vasopressin (units/hr); PCM: paracetamol; SpO₂: oxygen saturation by pulse oximetry; SaO₂: oxygen saturation

in well perfused lungs with adequate Qp) (3, 6). A laparoscopy-induced raised IAP resulting a decreased venous return could have been detrimental in this patient (affecting preload, CO and Qs). A subsequent rise in intrathoracic pressures would have further compromised the Qp by increasing the PVR. Higher A-a gradients (V/Q mismatch) were minimised by avoiding hypoxemia, hypercarbia, acidosis and low Qp. IAP was kept below conventionally recommended 12 mmHg

by gradual gas insufflation, and by constantly monitoring IAP and abdominal compliance (10). Fall in preload was partially offset by the Trendelenburg position. CO augmentation was further managed with TEE-guided fluid administration and monitoring LV contractility. TEE was also used to monitor gas embolism. In this particular patient with good biventricular function, we could maintain adequate CO perioperatively by ensuring appropriate preload without much requirement

of inotropes (perioperative CO was monitored continuously using TEE). An optimal MAP, essential to maintain the end-organ perfusion, was maintained using minimal dose of vasopressin (not exceeding 2.4 units hr⁻¹). Vasopressin was the preferred vasoconstrictor in this patient, considering its favourable profile on pulmonary vasculature compared to systemic vasculature. Fortunately, there were no episodes of air embolism as monitored by TEE, probably because of the precautionary measures to avoid air embolism (air filters, fluid administration sets with air lock, deairing of crystalloid bags and vigilant administration of medications following strict prior aspiration protocol). The recommended dynamic variables of fluid responsiveness (stroke volume variation, pulse pressure variation, echocardiography-based V_{\max} and velocity time integral (VTI) variability) were not much reliable in this particular patient with large VSD (which alters the expected cardiorespiratory interactions to hypovolemia). Therefore, qualitative assessment of left ventricular end-diastolic area (LVEDA) was the only acceptable surrogate indicator of preload status (11). A hyperkinetic LV with small LVEDA (corresponding to approximation of papillary muscles in TG mid pap SAX view) served as a static indicator of low preload in this patient (12). The 'A' wave amplitude and duration obtained from the pulmonary venous flow served as a good indicator of left atrial pressures, safeguarding against excessive fluid administration. Therefore, TEE-guided hemodynamic management to optimise Q_s and Q_p enabled this patient with single ventricle physiology to function on the flatter part of the Frank-Starling curve. Thus, an adequate tissue oxygenation was achieved over a wide range of $Q_p:Q_s$ (closer to 1) (5), facilitating an early extubation and a favourable perioperative course (6).

Conclusion

ACHD can be best managed in a tertiary care hospital by formulating an effective perioperative plan involving a multidisciplinary team aiming for realistic therapeutic goals (13). The importance of TEE in the management of hemodynamics in this critically balanced circulation cannot be overemphasised.

Informed Consent: Written informed consent was obtained from the patient.

Peer-review: Externally peer-reviewed.

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Conflict of Interest: The authors have no conflicts of interest to declare.

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Supplementary digital content video 1. Transgastric short axis mid-papillary view shows a well contracting non-dilated left ventricle

References

- Hoffman JI, Kaplan S. The incidence of congenital heart disease. *J Am Coll Cardiol* 2002; 39: 1890-900. [\[Crossref\]](#)
- Sinto R, Nasution SA. Pregnancy in a woman with uncorrected tetralogy of fallot. *Acta Med Indones* 2009; 41: 81-6.
- Zach KJ, Ramakrishna H, Chandrasekaran K, Weis RA. Laparoscopic colectomy in an adult with single ventricle physiology: anesthetic implications and management. *Ann Card Anaesth* 2015; 18: 252-6. [\[Crossref\]](#)
- Fairley SL, Sands AJ, Wilson CM. Uncorrected tetralogy of Fallot: adult presentation in the 61st year of life. *Int J Cardiol* 2008; 128: e9-e11. [\[Crossref\]](#)
- DiNardo JA, Zvara DA. Anesthesia for cardiac surgery. Blackwell publishing 2008; chapter 6, pp 167-251. [\[Crossref\]](#)
- Lovell AT. Anaesthetic implications of grown-up congenital heart disease. *Br J Anaesth* 2004; 93: 129-39. [\[Crossref\]](#)
- Ishizaka T, Ichikawa H, Sawa Y, Fukushima N, Kagisaki K, Kondo H, Kogaki S, Matsuda H. Prevalence and optimal management strategy for aortic regurgitation in tetralogy of Fallot. *Eur J Cardiothorac Surg* 2004; 26: 1080-6. [\[Crossref\]](#)
- Gatzoulis MA, Till JA, Somerville J, Redington AN. Mechano-electrical interaction in Tetralogy of Fallot. QRS prolongation relates to right ventricular size and predicts malignant ventricular arrhythmias and sudden death. *Circulation* 1995; 92: 231-7. [\[Crossref\]](#)
- Alonso A, Downey BC, Kuvlin JT. Uncorrected tetralogy of Fallot in an 86-year-old patient. *Am J Geriatr Cardiol* 2007; 16: 38-41. [\[Crossref\]](#)
- Ott DE. Abdominal Compliance and Laparoscopy: A Review. *JLS* 2019; 23: e2018.00080. [\[Crossref\]](#)
- Levitov AI, Marik PE. Echocardiographic assessment of preload responsiveness in critically ill patients. *Cardiol Res Pract* 2012; 2012: 819696. [\[Crossref\]](#)
- Feissel M, Michard F, Mangin I, Ruyet O, Faller JP, Teboul JL. Respiratory changes in aortic blood velocity as an indicator of fluid responsiveness in ventilated patients with septic shock. *Chest* 2001; 119: 867-73. [\[Crossref\]](#)
- Alatassi A, Mulero SF, Massoud N, Alzayer Z, Mahmoud AH. A case series of different anesthesia approaches for single ventricular physiology patients in various stages of palliation underwent noncardiac procedures. *Saudi J Anaesth* 2018; 12: 629-33.