

# Anesthetic management in tetralogy of fallot with pulmonary atresia and major aortopulmonary collateral arteries in pediatric patients: One year experience

## ABSTRACT

**Background:** Unlike the typical tetralogy of fallot (TOF), the presence of pulmonary atresia and major aortopulmonary collaterals is recognized as a rare but severe variant of TOF. The objective was to describe the perioperative anesthetic management of pediatric patients who underwent unifocalization procedure and to describe their postoperative morbidity and mortality.

**Methods:** A retrospective observational study was conducted among pediatric patients who underwent unifocalization procedure at Prince Sultan Cardiac Center (PSCC) between October 2017 and October 2018. Unifocalization procedures were performed in a two-staged approach. Anesthetic management of both stages had similar concerns and challenges but with few peculiar issues. These included preoperative assessment, intraoperative management, vascular access, positioning, ventilation, optimizing hemodynamics, cardiopulmonary bypass monitoring, and postoperative management.

**Results:** A total of 19 unifocalization procedures were included. The average age was one year and 52.6% were females. Two patients (10.5%) arrived in theater already with intubated ventilation. Continuous mandatory ventilation was used in 11 (57.9%) patients while one-lung ventilation was used in the rest of patients. Approximately, 30% of patients encountered a stormy postoperative course, 52.6% underwent cardiopulmonary bypass with or without cross-clamping of the aorta, and 10.5% had reperfusion injury.

**Conclusions:** Despite the major challenges of unifocalization, significantly low rates of morbidity and mortality were observed in our patients. A thorough familiarity of different airway and ventilation issues, besides meticulous hemodynamic and anesthetic management, is of paramount importance. The maintenance of hemodynamic stability, hemostasis, and proper ventilation is critical for the success of the operation.

**Key words:** Anesthetic management; tetralogy of fallot; unifocalization procedures

## Introduction


Tetralogy of fallot (TOF) is a common congenital heart disease characterized by pulmonary stenosis, ventricular septal

defect (VSD), right ventricular hypertrophy, and overriding of the aorta.<sup>[1-3]</sup> Unlike the typical TOF, the presence of pulmonary atresia and major aortopulmonary collaterals

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## AHMAD ABUZAID, IBRAHIM ABD ELAAL<sup>1</sup>, AHMED ABDULAZIZ<sup>2</sup>, RAWAN ABUZAID<sup>3</sup>

Director of Cardiac Anesthesia and OR Department, <sup>1</sup>Consultant of Cardiac Anesthesia, <sup>2</sup>Senior Registrar of Cardiac Anesthesia, Prince Sultan Cardiac Center, <sup>3</sup>College of Pharmacy, King Saudi University, Riyadh, Saudi Arabia

**Address for correspondence:** Dr. Ahmad Abuzaid, Director of Cardiac Anesthesia and OR Department, Prince Sultan Cardiac Center, Riyadh, Saudi Arabia. E-mail: aabuzaid@yahoo.com

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in patients with TOF (TOF/PA/MAPCAs) is recognized as a rare but severe variant of TOF.<sup>[1-3]</sup> Unlike the surgical management of TOF/PA/MAPCAs which has been extensively described in the literature, the anesthetic management of such a complex procedure received little attention. The objective of the current study was to describe the perioperative anesthetic management of pediatric patients who underwent unifocalization procedure at Prince Sultan Cardiac Center (PSCC), as well to describe postoperative morbidity and mortality among these patients.

## Methods

### Setting and patient selection

The current study is a retrospective observational study conducted at PSCC between the beginning of October 2017 and mid-October 2018. It included all 19 pediatric patients who were admitted to the operative room (OR) at PSCC for either a unilateral or bilateral unifocalization procedure as a surgical correction of TOF/PA/MAPCAs.

### Surgical approach

The extreme variations in the number and course of collaterals in patients with TOF/PA/MAPCAs require individualization of the surgical approach. The surgeons at PSCC followed a two-staged approach in performing unifocalization procedure. The main advantage of such a staged approach is to allow small native central pulmonary arteries to grow. From the surgical point of view, the first stage involves a thoracotomy along with unilateral unifocalization of the MAPCAs into a single vascular graft that is connected to the systemic circulation by a central shunt. The second stage which is usually performed after at least 3 months from the first one involves a median sternotomy incision and the unifocalization of the other side of MAPCAs into a new graft. The earlier central shunt is then removed and the two vascular grafts (right and left) are connected together and then to the right ventricle (RV). The connection to the RV can be performed through the native pulmonary artery or via a synthetic conduit. Finally, the VSD is closed.

### Anesthetic management

The information herewith is focusing on crucial and specific anesthetic management information that should be in the mind of anesthetists who are planning to handle unifocalization procedures. They are presented separately for each stage.

#### **Anesthetic management of the first stage of correction**

##### Preoperative assessment

The preoperative assessment of the extent and severity of TOF/PA/MAPCAs was performed with the primary cardiology team. Furthermore, the anesthetist had at least one

preoperative meeting with the surgical team to evaluate other comorbidities and congenital anomalies. The implemented overnight preparation protocol ensured to minimize fasting hours. Intravenous (IV) maintenance fluid was then started using 5% dextrose in water and diluted normal saline which contains 10 mmol potassium chloride in 500 mL. Overnight sedation was not usually performed due to the risk of hypoxia. Upon arrival to OR reception, IV sedation was performed with ketamine 1 mg/kg plus glycopyrrolate 5 µg/kg.

##### Intraoperative management

After patient assumed supine position over bear hugger air warming blanket, monitoring was done through three leads ECG, pulse oximeter, noninvasive measurement of blood pressure (NIBP), and near-infrared spectroscopy (NIRS). The mainstay induction protocol included IV induction with ketamine 2 mg/kg, fentanyl 5 µg/kg and intubation facilitated with rocuronium 1 mg/kg. The fraction of inspired oxygen (FiO<sub>2</sub>) used during induction and maintenance was variable according to the degree of preoperative pulmonary over-circulation. NIBP was done frequently during induction to maintain a level of oxygenation and blood pressure that can appropriately balance the pulmonary to systemic blood flow ratio (Qp/Qs). Single lumen tracheal tube (TT) with bronchial blocker (Arndt Pediatric Endobronchial Blocker, Cook Medical, USA) was used for lung separation. Fiberoptic confirmation of final site and balloon inflation was performed after passing the blocker beside the TT. Finally, different lung areas were auscultated to ensure proper lung separation. Balanced anesthesia was then established using fentanyl infusion 4–10 µg/kg/h in combination with inhaled sevoflurane (1-2 minimum alveolar concentration) and repeated boluses of rocuronium 1.2 mg/kg.

##### Vascular access

Arterial catheter was inserted into the femoral artery and a central venous catheter was routinely inserted into the right internal jugular in all patients. While femoral venous cannulation is advisable, bilateral femoral venous was avoided in the majority of patients in order to spare these vessels for future cardiac catheterization procedures.

##### Positioning

Lateral positioning was conducted after TT placement and confirmation. It is of paramount importance for the anesthetic to experience extreme caution while placing a pediatric patient in position. Finally, reconfirmation of integrity of TT site and passage should be done after complete positioning.

##### Ventilation

One-lung ventilation in pediatric patients is generally challenging, due to the fact that the ventilated lung

blood flow is already compromised by the aberrant blood vessels. Single-dose of corticosteroids (1–2 mg/kg methylprednisolone) was given to minimize the reperfusion injury. More importantly, lung volume was restricted and hypervolemia was avoided while keeping hemoglobin (Hb) level above 15–16 g/dL. In addition, nitric oxide, frequent endotracheal suctioning, and aggressive pulmonary care with metered-dose inhaler (MDI) ventolin puffs were implemented to tackle this problem. Analysis of arterial blood gases (ABGs) was done every 30 min to early detect and aggressively manage any acid-base, oxygenation, and electrolytes abnormalities. Heparin boluses of 50–100 IU were repeated with the aim to keep activated clotting time (ACT) between 200 s and 250 s to avoid closure of anastomosis.

#### Hemodynamics and postoperative management

Optimal fluid balance with acceptable Hb level was ensured. Furthermore, small doses of vasopressors and/or inotropic infusions were commenced in some patients. Acidosis and imbalance of electrolytes were vigorously corrected. The patients were transferred to a pediatric intensive care unit (PICU) after the first stage of surgical correction. As early extubation should not be done, the patients were kept on sedated ventilation to optimize the lung conditions.

#### **Anesthetic management of the second stage of correction**

##### Preoperative assessment

The preoperative assessment involved taking the history of the previous unifocalization procedure. A computed tomography (CT) of the chest to evaluate the space between the sternum and anterior border of the heart.

##### Intraoperative course

During the completion of unifocalization procedure, some patients required cardio-pulmonary bypass (CPB) with or without cross-clamping (CC) of the Aorta. Balanced anesthesia on bypass was ensured. This included IV fentanyl infusion 4 µg/kg/h during the period of CPB, along with inhaled sevoflurane. Finally, repeated boluses of rocuronium 1.2 mg/kg and small dose of midazolam 0.1–0.2 mg/kg were administered.

##### Ventilation

To avoid post-bypass reperfusion injury, frequent suctioning was done and aggressive pulmonary care was given to clear the airways of blood. In addition, recruitment maneuvers and increased positive end-expiratory pressure were required to recruit atelectatic lung segments and potentially minimize further hemorrhage. To improve the tolerability of RV for the new circulatory changes, gradual weaning or addition of inhaled nitric oxide was done. Open chest with or without

extracorporeal membrane oxygenation (ECMO) insertion could be additional last resort. Coagulopathy is common after bypass and frequently requires blood product administration.

#### Hemodynamics and postoperative management

Meticulous lines' connections and high flow rate were ensured after considering the patient's age, body weight, and body surface area. Continued assessments of end-organ functions were ensured through maintaining target MAP, in-line measurement of mixed venous saturation, pH, base excess, regular testing of blood for heparin and lactate levels, and other indices of end-organ perfusion including urine output, somatic, and cerebral oxygenation values (e.g., NIRS). All patients were transferred to PICU on sedated ventilation to optimize the lung conditions.

## Results

We had a total of 19 unifocalization procedures in PSCC during the one-year of the study. While the majority of patients underwent the previously mentioned two-staged correction, one patient underwent three-stage correction over a course of more than 2 years. The average age at operation was one year, ranging from 9 days to 2 years. Approximately three-fourth (77%) of the patients aged one year or less. Roughly, males and females were similarly represented (47.4% and 52.6%, respectively). The average duration for each operation was 9 h, ranging from 4 h to 15 h. The average ICU stay was 10 days, ranging from 3 days to almost a year.

Preoperative assessment showed some additional congenital anomalies in 4 (21.1%) patients, as shown in Table 1. Two patients arrived in theater already with intubated ventilation and on vasoactive drugs (epinephrine and dopamine), and alprostadil (prostaglandin E1) infusions [Table 2]. Continuous mandatory ventilation was used in 11 (57.9%) patients while one-lung ventilation was used in the rest of patients [Table 3].

The postoperative ICU course in the first stage of correction was variable, with 30% of patients encountered a stormy

**Table 1: Additional congenital anomalies in patients with TOF/PA/MAPCAs**

	Number	Percentage
DiGeorge syndrome, seizure disorder, and multiple emphysematous bullae	1	5.3%
Glaucoma	1	5.3%
Hydronephrosis with normal kidney function	1	5.3%
Lactose intolerance	1	5.3%
None	15	78.9%
Total	19	100.0%

postoperative course. The duration of mechanical ventilation ranged from 1 to 5 days and the patients were discharged from ICU within 3 to 10 days. For completion of unifocalization, 6 patients underwent CPB with CC. As shown in Table 4, the mean time for CPB was 230 min and the mean time for CC was 75 min. CPB without CC was conducted in two patients with a mean time of 275 min. Surgical management without CPB was performed in the remaining patients.

While the main post-bypass intraoperative pharmacological support was milrinone 0.5 to 0.75 µg/kg/min, four patients did not receive any inotropic drugs. Inhaled nitric oxide 20 ppm was used in 4 patients. One patient shifted to ICU with open chest while no patients needed ECMO insertion. With the exception of one patient, the majority of the patients had a smooth postoperative ICU stay. One patient experienced a remarkable volatile ICU course for almost a year. Another patient underwent catheter-based coil embolization of MAPCs and three patients underwent surgical diaphragmatic plication. One patient suffered sepsis (pneumonia and urinary tract infection). Finally, two patients had reperfusion injury with lung congestion and failed extubation, hypertension with oral antihypertensive drugs, and gastrointestinal stress ulcers.

## Discussion

While unifocalization is considered a variant of TOF, the management of anesthetic problems differs considerably

**Table 2: Induction in patients undergoing unifocalization procedure**

	Number	Percentage
Arrival to OR with mechanical ventilation and ETT <i>in situ</i>	2	10.5%
Inhalational	2	10.5%
Intravenous	15	78.9%
Total	19	100.0%

**Table 3: Bronchial blocker and ventilation during unifocalization procedure**

	Number	Percentage
Bronchial blocker		
No	12	63.2%
Yes	7	36.8%
Type of ventilation		
Continuous mandatory ventilation	11	57.9%
One-lung ventilation	8	42.1%

**Table 4: Average time in minutes of CPB with CC**

	Number	Mean	Minimum	Maximum
Cardiopulmonary bypass	6	230	159	316
Cross clamping of the aorta	4	75	40	152

from typical correction of TOF. Despite the major challenges of unifocalization, significantly low rates of morbidity and mortality were observed in our patients. Despite the fact that there are various approaches for the management of TOF/PA/MAPCAs,<sup>[4-9]</sup> the extreme variation in the anatomy should be stressed, which requires individualization of the approach used for individual patients.

The surgical approach conducted in our center was two-staged correction. This allows small native central pulmonary arteries to grow gradually. The later can result in augmentation of blood flow to the central pulmonary arteries, which can enhance the rehabilitation outcomes. Because of its complex anatomy, each surgical stage has its own risks and challenges in addition to the usual problems anticipated in both stages. Preoperative meticulous assessment is a must in both stages. A common associated anomaly is DiGeorge syndrome, which was observed in one patient in our study. It is usually associated with absent thymus, hypocalcemia, and a difficult airway.<sup>[10]</sup>

In the first stage of correction, the main challenge was to keep acceptable blood gases throughout the procedure. Arendt blocker passed beside the TT for lung isolation was our choice. The adjustment of the blocker site to secure the airway and maintain ventilation especially after repositioning laterally is very tricky and demands expertise. Repeated ABG sampling and correction of oxygenation, ventilation, and acidosis was the mainstay element in our anesthetic management. In our center, the management policy stipulates to repeat ABG sampling regularly every 30 min.

In the second stage of correction, CPB was required in almost a third of patients, specifically when saturation remains low (around 70%) despite all correction measured applied. Lung reperfusion injury in the setting of unifocalization surgery has been estimated in the previous studies at 65%.<sup>[11]</sup> Although only two patients (10.5%) had lung reperfusion injury in our study, more research should be conducted to underscore the significance of certain practices to tackle such a problem during unifocalization procedures. Proton pump inhibitor in a single dose with corticosteroid was used to decrease the incidence of reperfusion injury in both stages. RV dysfunction occurrence is usually secondary to increased afterload because of an inadequate pulmonary vascular bed. The key management strategies were optimization of preload, starting inotropic support, and proper adjustment of ventilation with the aim of decreasing pulmonary vascular resistance. Inhaled nitric oxide may provide assistance as a potent short-acting pulmonary vasodilator in this setting.

In conclusion, the anesthesiologist is a key factor in the management of this uncommon group of patients. A thorough familiarity of different airway and ventilation issues, besides meticulous hemodynamic and anesthetic management, is of paramount importance. The maintenance of hemodynamic stability, hemostasis, and proper ventilation in a precise manner is critical for the success of the operation. Despite the fact that our approach in anesthetic management has been established based on the surgical practices at PSCC, we believe that our experience may be useful for other anesthesiologists caring for similar patients.

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#### Conflicts of interest

There are no conflicts of interest.

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