Introduction

Cyanotic congenital heart disease (CCHD) presents an increased tendency to bleed in view of subtle coagulation defects. Airway bleeding can be particularly difficult to manage while maintaining an adequate ventilation. An isolated lung bleed with the exclusion of possible traumatic, medical and surgical causes of bleeding, should alert the attending anesthesiologist to the possibility of the collateral-related bleeding. Preoperative coil embolization remains an important initial management step in a case of tetralogy of Fallot (TOF) with major aortopulmonary collaterals. Nevertheless, the coiling of the collaterals in certain specific case scenarios is not feasible, rendering the management of a lung bleed, all the more challenging. We, hereby discuss a case of a 7-year-old girl with a massive endotracheal bleed at the time of weaning from cardiopulmonary bypass after corrective surgery for TOF. The subsequent approach and management are discussed. The optimal management of tetralogy with collaterals mandates an effective communication among the cardiologist, radiologist, anesthesiologist, and the surgeon.

Case Report

A 7-year-old girl, weighing 16 kg, with the diagnosis of TOF, and a history of right-sided modified Blalock-Taussig (BT) shunt 4 years back, was scheduled for corrective surgery. On examination, she had pulse rate of 80 beats/min, sinus rhythm, arterial blood pressure 90/60 mm of Hg, and a Grade II ejection systolic murmur heard in the second left intercostal space. The peripheral oxygen saturation was 88% on room air. She was on Tablet propranolol 10 mg every 12 h. Preoperative hemoglobin was 18.2 g/dL with a platelet count of 269,000/µL. Chest X-ray postero-anterior view showed a boot-shaped heart with normal transverse diameter and clear lung fields. Electrocardiography showed sinus rhythm with right ventricular hypertrophy.

Preoperative echocardiography showed situs solitus, atrioventricular and ventriculoarterial concordance, large, nonrestrictive malaligned ventricular septal defect (VSD) of 20 mm size, with the right to left shunt and 50% aortic over-ride. There was severe infundibular and valvular pulmonary stenosis. The peak gradient across the right ventricular outflow tract was 90 mm of Hg. Pulmonary arteries were small and confluent with Mc Goon ratio of 0.7. The right ventricle was moderately hypertrophied. Ventricular function was good with an ejection fraction of 61%. The great vessels were normally related with a left aortic arch.

Cardiac catheterization confirmed the diagnosis. The coronaries were normal. Descending aortogram did not reveal any significant aortopulmonary collaterals [Figure 1]. However, injection into the right subclavian artery showed multiple small collaterals feeding the right upper lobe. Reformation of only a distal

**Abstract**

Cyanotic congenital heart disease presents an increased tendency to bleed in view of subtle coagulation defects. Airway bleeding can be particularly difficult to manage while maintaining an adequate ventilation. An isolated lung bleed with the exclusion of possible traumatic, medical and surgical causes of bleeding, should alert the attending anesthesiologist to the possibility of the collateral-related bleeding. Preoperative coil embolization remains an important initial management step in a case of tetralogy of Fallot (TOF) with major aortopulmonary collaterals. Nevertheless, the coiling of the collaterals in certain specific case scenarios is not feasible, rendering the management of a lung bleed, all the more challenging. We, hereby discuss a case of a 7-year-old girl with a massive endotracheal bleed at the time of weaning from cardiopulmonary bypass after corrective surgery for TOF. The subsequent approach and management are discussed. The optimal management of tetralogy with collaterals mandates an effective communication among the cardiologist, radiologist, anesthesiologist, and the surgeon.

**Keywords:** Airway bleed, aortopulmonary collaterals, coil embolization, lung bleed, tetralogy of Fallot

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**Address for correspondence:**

Prof. Neeti Makhija,
Department of Cardiac Anaesthesia,
Room No. 9, 7th Floor, Cardiothoracic Centre,
All India Institute of Medical Sciences,
New Delhi - 110 029, India.
E-mail: neetimakhija@hotmail.com

**Website:** www.annals.in

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right upper lobe branch of PA was seen [Figure 2]. Injection into right modified BT shunt showed filling of confluent pulmonary arteries. Mild narrowing of right pulmonary artery (RPA) at right modified BT shunt anastomosis site was appreciated and the upper lobe branch of RPA was cutoff [Figure 3].

In the operating room, induction of anesthesia was done with sevoflurane in 100% oxygen followed by administration of injection ketamine 2 mg/kg, fentanyl 3 µg/kg, and rocuronium bromide 1 mg/kg. A smooth direct laryngoscopy was performed and the trachea intubated with a 5.5 cuffed endotracheal tube (ETT). A pediatric transesophageal echocardiography (TEE) probe and temperature probe were gently placed followed by the acquisition of left radial artery and right internal jugular central venous access. Median sternotomy was done, and right modified BT shunt takedown was performed, followed by systemic heparinization. CPB was initiated after aortic and bicaval cannulation and an adequate activated coagulation time (ACT). The CPB lasted for 124 min with aortic cross-clamp duration of 66 min. The surgical field did repeatedly flood with blood during the CPB, which was managed with frequent venting.

After satisfactory correction, on resuming gentle ventilation, the aortic cross-clamp was released. Thereafter, lungs became progressively heavy, and ventilation was barely possible even at high airway pressures in spite of a patent ETT and circuit. The patient was nebulized with short acting β agonists. Injection deriphyllin, methylprednisolone, and hydrocortisone were added to the CPB reservoir. The difficulty in ventilation persisted. Soon, endotracheal bleed became apparent. The oral cavity was free from any bleeding. Gentle ETT suctioning was mandatory to allow any ventilation signifying a substantial bleed. About 100–150 ml of blood was suctioned from the ETT, and ventilation was then possible.

Once we were able to ventilate with an acceptable tidal volume, we came off CPB. On TEE, an adequate surgical repair was confirmed, demonstrating a good contractility with no right ventricular outflow tract obstruction or residual VSD. We could maintain hemodynamics on inotropic support of dopamine 5 µg/kg/min, dobutamine 5 µg/kg/min, and noradrenaline 0.1 µg/kg/min and peripheral oxygen saturation between 90% and 97% on 100% oxygen. Even, after adequate reversal of heparin with protamine, the ETT bleed continued. Adrenaline-saline (1:200,000) irrigation, gentle ETT suction of blood and intermittent ventilation was alternated. A post-CPB bolus dose of epsilon aminocaproic acid (EACA) 100 mg/kg was followed by an infusion of EACA. 20 ml/kg fresh frozen plasma, 0.1 U/kg platelet concentrates and packed red blood cell corpuscles were transfused to maintain a minimum hematocrit of 36%, in view of ongoing lung bleed. The sonoclot signature and the ACT were normal ruling out a medical cause of bleed. The above-mentioned measures were continued for an hour to achieve hemostasis. The amount of bleed had decreased sizeably but did not subside completely.

The immediate postoperative chest X-Ray demonstrated haziness in right upper and middle zone of the lung [Figure 4]. In the intensive care unit, we followed a comprehensive three-pronged management approach that constituted (i) lung protective ventilatory strategy with a low tidal volume, high frequency, minimizing the airway pressures, addition of positive end-expiratory pressure, recruitment manoeuvres with minimal and gentle suctioning, (ii) delayed sternal closure, and (iii) pharmacological control of bleeding and airway hyperreactivity with antifibrinolytics, nebulization, and steroids, respectively. The blood and blood products were transfused to maintain a minimum hematocrit of 36% and coagulation parameters within normal range. The patient was successfully weaned off and extubated on the third
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Discussion

The patients with TOF develop collaterals in response to decreased pulmonary blood flow and ongoing hypoxemia.\(^3\) The origin, number, size, course, and arborization of collaterals and concomitant variation of pulmonary blood supply is a valuable preoperative information. A segment of lung may be supplied solely from the true pulmonary arteries, solely from the aortopulmonary collaterals, or from both.\(^4,5\) Therefore, a meticulous computed tomographic angiography for detailed characterization of collaterals and pulmonary blood supply is indispensable.\(^6\) This helps to direct the appropriate management of major aortopulmonary collaterals (MAPCAs) as they contribute to the pulmonary blood flow.

Preoperative coil embolization of MAPCAs remains an important step in the management of patients coming for intracardiac repair (ICR). However, TOF with MAPCA as a sole blood supply to a segment of a lung precludes preoperative coil embolization. Airway bleeding due to the collaterals poses multitude challenges throughout ICR. In our patient, the multiple small tufts like collaterals were the sole supply to right upper lobe, and they were not easily amenable to coiling. Hence, they were left with the presumption that intraoperative unifocalization procedure would be done and these tufts like collaterals would be dealt in the postoperative period if required. Intraoperatively, it was discovered that a unifocalization procedure could not be done as the distal RPA was not well reformed and collaterals were small in size.

The patient was taken up with the background that significant MAPCAs were absent in the present case. At the same time, preoperative coil embolization of small tuft of collaterals that were present was not technically feasible. Despite the presence small collaterals, such a bleeding catastrophe was unexpected for the team. The team pondered into the various causes of airway bleed. The massive intraoperative lung bleed, was however, conservatively managed with the consideration of coil embolization of collaterals or lobectomy as a last resort, in the face of a failing conservative management strategy.

The index case highlights certain distinctive peculiarities of perioperative management of a patient of TOF with collaterals. The ongoing hypoxemia in TOF leads to ubiquitous development of collaterals of varying anatomy, morphology, and contribution to the pulmonary blood supply. The authors wish to elucidate the fact that an isolated lung bleed with the exclusion of possible traumatic, medical and surgical causes of bleeding, should alert the attending anesthesiologist to the possibility of the collateral-related bleeding, especially in the setting of a CCHD. Our case is unique in the sense that the tuft of small collaterals was the sole blood supply to the right upper lung lobe, rendering the perioperative coiling inappropriate. Thereby, the armamentarium for the management of a lung bleed in such a case scenario is limited to various pharmacological and nonpharmacological manoeuvres maintaining a much higher threshold for coil embolization of collaterals. The awareness on the part of anesthesiologist regarding the possibility of such a bleeding catastrophe due to the presence of small collaterals is of utmost importance.

Conclusion

The optimal management of tetralogy with collaterals mandates an effective communication among the cardiologist, radiologist, anesthesiologist, and the surgeon. The perioperative course of these patients can be complicated with lung bleed, the mitigation of the same requires a meticulous and well-informed attending anesthesiologist.
Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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