

Original Research Article

Endotracheal bleed after intra cardiac repair in tetralogy of fallot patients: a diagnostic challenge

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Received: 14 April 2022

Revised: 06 May 2022

Accepted: 19 May 2022

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ABSTRACT

Background: Tetralogy of Fallot (TOF) is the most common congenital cyanotic heart disease (CCHD) in children. With the development and advancement of paediatric cardiac surgery and intensive care units, a large number of TOF are presenting for cardiac surgery. Operated TOF may suffer from endotracheal (ET) bleeding in postoperative period which results in delayed extubation of patients. ET bleeding is multifactorial with MAPCAs, coagulopathy residual VSD, and high LA pressure being usual causes. The objective of this study was to evaluate those children of TOF who suffered from ET bleeding, their causes and management after intracardiac repair.

Methods: This was a single center retrospective study that included patients with a diagnosis of TOF. Total of 27 patients were operated at GB Pant Hospital, Department of CTVS from February 2021 to November 2021. After taking ethical approval we conducted a retrospective study to analyse the cause of endotracheal bleed. Statistical analyses were performed using STATA software version (16.0). Continuous variables were presented as mean±standard deviation (SD) or median (interquartile range [IQR]). Inclusion criteria was- all diagnosed case of tetralogy of fallot patient with age –1.5-8 years, both male and female. Exclusion criteria was- patients having co-morbidities, like juvenile diabetes, hypothyroidism, syndromic patients.

Results: Endotracheal bleed noted in majority of patient in those MAPCAs were not coiled, not able to be ligated intraoperatively, they bled in postoperative period which further delayed extubation of patients. Of the patients with Endotracheal bleed none had a residual cardiac defect. Post-operative mortality was 1/27.

Conclusions: We hereby conclude that cause of endotracheal bleed after intracardiac repair of tetralogy of fallot patients can be due to left uncoiled, non- ligated major aorto-pulmonary collateral arteries (MAPCAs).

Keywords: TOF, Intracardiac repair, Endotracheal bleed

INTRODUCTION

Tetralogy of Fallot (TOF) is the most common congenital cyanotic heart disease (CCHD) in children, and has a wide spectrum of pathology.^{5,6} Pulmonary blood flow is dependent on the presence of an aorto-pulmonary communication, which may exist in the form of a patent ductus arteriosus (PDA), other major nonductal

collaterals arising from the descending aorta that connect to the central native pulmonary arteries, or MAPCAs that directly enter the hilum and join the segmental pulmonary arteries. In utero, the pulmonary parenchyma is perfused by branches from the dorsal aorta as well as by the sixth aortic arch, which forms the basis for the central pulmonary arteries. The most severe form of TOF is with pulmonary atresia (TOF-PA).⁷ Approximately 85% of

MAPCAs are found in patients with TOF-PA.⁹ The objective of this study was to evaluate those children of TOF who suffered from ET bleeding, their causes and management after intracardiac repair.

METHODS

This was a single center retrospective study that included patients with a diagnosis of TOF, TOF-PA, double outlet right ventricle with TOF subtype (DORV-TOF), and DORV with pulmonary atresia subtype (DORV-PA) who had CT angiography at our tertiary care radiology department. Total of 27 patients were operated at GB Pant Hospital, Department of CTVS from February 2021 to November 2021. These patients were studied for echo findings, CT pulmonary angiography, cardiac catheterisation study Coagulation parameters, and presence of MAPCAs etc. These patients were operated by single surgeon. In every case, intracardiac repair was done. These patients were studied for demographic characteristics (Table 1), objective of study MAPCAs Coiling, CPB Time, Aortic cross clamp time, Left atrial pressure time, MUF after surgery, ACT time, endotracheal bleed after surgery, extubation time (Table 2).

After taking ethical approval we conducted a retrospective study to analyse the cause of endotracheal bleed. Statistical analyses were performed using STATA software version (16.0). Continuous variables were presented as mean $\pm$ standard deviation (SD) or median (interquartile range (IQR)).

Inclusion criteria

All diagnosed case of tetralogy of fallot patient with age $-1.5-8$ years. Both male and female.

Exclusion criteria

Patients having co-morbidities, like juvenile diabetes, Hypothyroidism. Syndromic patients.

RESULTS

Endotracheal bleed noted in 15 patients (55.5%). Preoperatively cath study and coiling of MAPCAs done in 19 patients (79%) (Table 2). On table ligation of MAPCAs were done in 4 patients (14.81%).

8 patients (29.62%) were more blue hence they had few MAPCAs, or they had history of recurrent episodes of spell, MAPCAs were not coiled. Hence, in those MAPCAs were not coiled, or not able to be ligated, they bleed in postoperative period which further delayed extubation of patients. Of the patients with ET Bleed none had a residual cardiac defect. The mean LA pressure evaluated in the OT was 14.07. Post-operative mortality was 1/27.

DISCUSSION

Patients with TOF-PA develop major aorto pulmonary collateral arteries (MAPCAs) in utero as a consequence of absence of antegrade pulmonary blood flow.⁷

Table 1: Study of demographic characteristics.

Demographic characteristics	Range	Mean $\pm$ SD	Median (IQR)
Age (years)	1.5-8	2.9 $\pm$ 1.59	2.3(2.3)
Gender	Male-17		
	Female-10		
Height (cm)	76-130	98.33 $\pm$ 15.72	100(32)
Weight (kg)	10.5-22	14.16 $\pm$ 2.79	14(4)
BSA (m ²)	0.45 - 0.9	0.54 $\pm$ 0.14	0.47(0.14)

Table 2: Study of objective of the study.

Studied parameters	Range	Mean $\pm$ SD
MAPCAs coiling		
Pre-operatively	19	
On table ligation of MAPCA'S	4	
CPB time (minutes)	110-122	116.81 $\pm$ 5.28
Aortic cross clamp time (minutes)	75 - 87	81.81 $\pm$ 5.24
Left atrial pressure	8-25	14.07 $\pm$ 3.92
Modified ultrafiltration (MUF) after surgery	15	
Activated clotting time (ACT)		
After protamine reversal	110-128	118.37 $\pm$ 7.13
After 2 hours of ICU arrival	138-155	144.33 $\pm$ 7.10
Endotracheal bleed		
Day 1	6	
Day 2-3	9	
Extubation		
After 17 hours	12	
After 41 hours	11	
After 65 hours	4	
Uneventful		
Post operative course	26	
Re-intubated	1	
Expired	1	

Embryology suggests that MAPCAs are congenital systemic arterial collaterals originating from the persistent intersegmental arteries which should normally regress once the connection between the central and intraparenchymal pulmonary arteries (PAs) has been established MAPCAs are also developed in TOF.^{7,8} Chronic hypoxia and high hemoglobin levels may

contribute to the development of MAPCAs in patients with TOF.¹⁰ Abnormal development of the central pulmonary arteries leads to persistence of the dorsal aortic branches, which ultimately develop into MAPCAs.

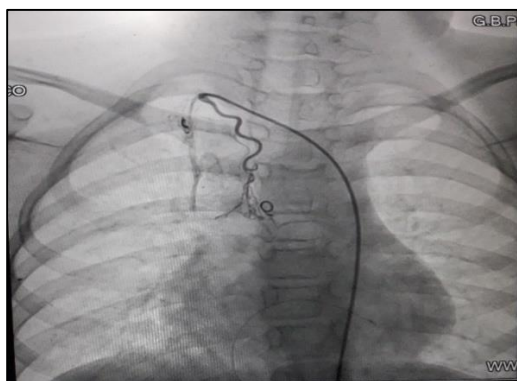


Figure 1: Preoperatively coiled MAPCAs.

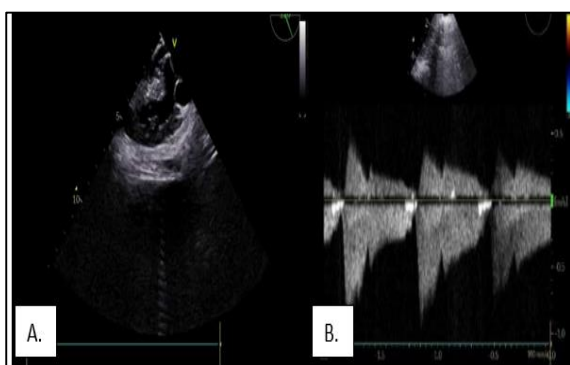


Figure 2: (A) TEE- Midoesophageal long axis view showing TOF anatomy; (B) there is large SA VSD >60% aortic override and RV hypertrophy.

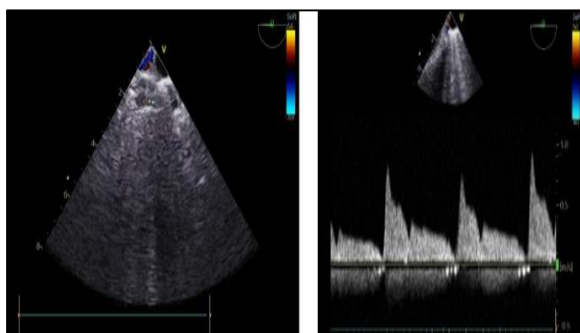


Figure 3 (A and B): Colour doppler assessment of MAPCAs in descending thoracic aorta short axis view showing pulsatile flow (towards and away from transducer).

The lungs may be supplied by the native pulmonary arteries, by MAPCAs, or by both. If confluent central pulmonary arteries are present, the blood supply to the lung may arise from a PDA, a central “ductlike” collateral, or a MAPCA. The behaviors of these three sources of pulmonary blood flow differ in their

predisposition to constrict over time, which has implications for the stability of pulmonary blood flow. Whereas MAPCAs or ductlike collaterals might remain patent beyond the neonatal period, the PDA is likely to constrict postnatally because of the presence of prostaglandin-responsive ductal tissue.

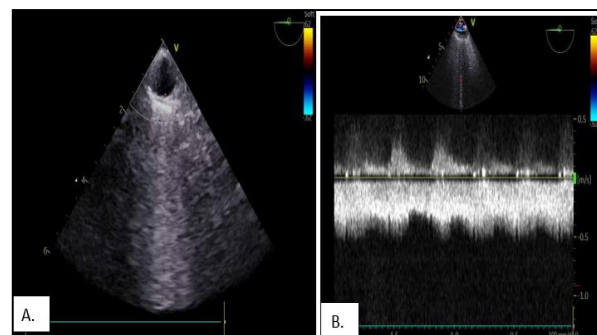


Figure 4 (A and B): Colour doppler assessment of MAPCAs in descending thoracic aorta short axis view confirm adequate coil embolization as there is no pulsatile flow.

Although the distinction may be difficult to establish by preoperative imaging studies, the location on the aorta from which the collateral originates is suggestive. Computed tomography (CTA) has been considered an accurate diagnostic modality for the assessment of MAPCAs.^{4,12,13} A solitary collateral that arises just distal to the left subclavian artery (if left aortic arch) and travels to a central pulmonary confluence is likely to be a PDA, whereas a collateral that arises from any other location off the aorta and supplies a central confluence is termed a ductlike collateral. A vessel that exits the descending aorta and travels directly to the pulmonary parenchyma is likely a MAPCA. There may be heterogeneity in the sources of pulmonary blood flow within any given patient, such that one segment of lung may be supplied by the native pulmonary artery whereas another segment may be supplied by a MAPCA. Some segments may have a dual supply. Pulmonary circulation in patients with TOF and TOF-PA is derived from PAs and/ or MAPCAs. The native PAs can vary from mild hypoplasia to absence of a branch or all PAs, and from confluent to non-confluent PAs.^{9,11} Patients with confluent central pulmonary arteries that are supplied by a PDA are less likely to have significant MAPCAs, and those with nonconfluent central pulmonary arteries depend on MAPCAs for pulmonary blood flow. MAPCA are not frequent in the setting of tetralogy of Fallot with pulmonary stenosis.¹ Patients with dual supply MAPCAs represent a separate anatomic category that is important to identify in the neonatal time frame. By definition, the phrase “dual supply” indicates that portions of the lung receive blood supply from both the pulmonary arterial tree and from MAPCAs. There are some patients who have completely normal arborization of the pulmonary tree, with all 18 segments of the lung in continuity with the pulmonary artery.

Another classification scheme divides patients further according to specific patterns of pulmonary artery anatomy that are most commonly encountered.

Valvar pulmonary atresia with a decent-sized main pulmonary artery and confluent central pulmonary arteries. This variant is associated with a PDA and therefore not associated with MAPCAs.

Absent main pulmonary artery with confluent, decent-sized or mildly hypoplastic central pulmonary arteries supplied by a PDA, without MAPCAs.

Confluent central pulmonary arteries with MAPCAs but no PDA.

Central pulmonary artery z-score > -2.5 .

Central pulmonary artery z-score < -2.5 .

Nonconfluent or absent central pulmonary arteries with MAPCAs.

Nonconfluent central pulmonary artery with PDA and MAPCAs.

The majority of MAPCAs originate from the anterior surface of the descending aorta. These MAPCAs can be found in the window between the ascending aorta and superior vena cava and beneath the bifurcation of the trachea (Figure 1). These MAPCAs initially course anterior for the first centimeter before turning laterally to reach either the right or left lung. The majority of MAPCAs will be anterior to the bronchi, although some branches may also go behind the bronchus. MAPCAs originating from the descending aorta may take an alternative course and traverse behind the esophagus. These retroesophageal MAPCAs originate from the lateral surface of the aorta and then course posteriorly to go behind the esophagus. As a consequence of the intrathoracic anatomy, retroesophageal MAPCAs are only found on the side contralateral to the aortic arch. Some retroesophageal MAPCAs will traverse through some of the esophageal muscle fibers, and thus are intraesophageal. The prevalence of intraesophageal MAPCAs was also found to be different based on the side of the arch (72% for left arch versus 32% for right arch). Many retroesophageal MAPCAs will have stenoses at the point where they cross behind the esophagus. The etiology of these stenosis remains unclear, although presumably it is on an embryologic basis. Because the majority of intraesophageal MAPCAs have significant midsegment stenoses, they should not be used as unaltered conduits but should either be trimmed distal to the stenosis or augmented with a patch. The significant number of patients with retroesophageal MAPCAs indicates that they should routinely be sought on the side contralateral to the aortic arch. MAPCAs may originate from a variety of other sources, including the subclavian arteries, innominate artery, thyrocervical trunk, and

coronary arteries. These MAPCAs are typically smaller in size and supply a lesser number of segments than those originating from the descending aorta. MAPCAs originating from the brachiocephalic vessels will usually course along the airway before entering the lung. In addition to the anatomic features outlined above, there are a small number of patients with PA/VSD/MAPCAs who present with signs and symptoms of congestive heart failure early on in life. These patients invariably have large MAPCAs with excessive pulmonary blood flow. As a result of the anatomical heterogeneity of pulmonary circulation and essentially uniqueness of each patient, Barbero-Marcial's classification based on the anatomy of PAs and MAPCAs, has been proposed to facilitate the risk stratification for patients with pulmonary atresia.⁸ Some of these patients will be ventilator dependent due to their heart failure condition. There is no effective medical treatment that will restore these patients to a favorable course, and thus early complete repair is indicated.^{2,3} We aimed to find out intraoperatively uncoiled MAPCAs by TEE (Figure 2, 3, 4) to ligate if possible.

Limitation of this study was some MAPCAs couldn't be found intraoperatively, and hence were left unligated. Hence, ET bleed was the cause for prolonged extubation time, and prolonged ICU stay.

CONCLUSION

We hereby conclude that cause of endotracheal bleed after intracardiac repair of tetralogy of fallot patients can be due to left uncoiled, non-ligated major aortopulmonary collateral arteries (MAPCAs), which bleed postoperatively, which could be surgeons nightmare. Hence, all possible effort should be made to coil/ligate these pre-operatively.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: The study was approved by the Institutional Ethics Committee

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Cite this article as: Sinha L, Solanki HJ, Ali M, Agarwal S, Naqvi SHE, Geelani MA et al. Endotracheal bleed after intra cardiac repair in tetralogy of fallot patients: a diagnostic challenge. *Int Surg J* 2022;9:1226-30.