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Case Report

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# Perioperative Identification and ligation of MAPCAs under TEE guidance: Case series of seven patients

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## Abstract

**Background:** Major aortopulmonary collateral arteries (MAPCAs) are systemic-to-pulmonary arteries of variable origin or size that carry blood to the lungs in complex congenital cyanotic heart defects (CCHD). Most notably CCHD involved in the MAPCAs genesis are Tetralogy of Fallot (TOF), transposition of great arteries (TGA), tricuspid atresia (TA), VSD, Pulmonary Atresia (PA) with Ventricular Septal Defect (VSD). MAPCAs are rare, and present in 4–10 per 10,000 live births and 2–3% of all CHDs. They are commonly associated with PA with VSD (50–70%), and 5–10% in patients of TOF, and most MAPCAs arise from the descending thoracic aorta (DTA). MAPCAs are essential for survival in most extremes of the lesions. These can be utilized for unifocalization (connecting to the native pulmonary system) during surgery. MAPCAs might serve as the primary sole source of blood supply to the lungs when the native pulmonary arteries are severely hypoplastic or absent. However, it needs to be ligated before or after CPB to prevent pulmonary over-perfusion, vessel narrowing, or lung damage, and low systemic perfusion pressure and tracheal bleeding during cardiac surgery. Management of MAPCAs is complex, often requires surgical ligation or embolization in Cath lab. Cardiac catheterization is a gold standard for diagnosis of the MAPCAs and their detailed anatomy, although CT angiography (CTA) is frequently used to visualize the anatomy before surgery. However, many a times either these facilities are unavailable or children are unsuitable due to poor hemodynamic in uncompensated heart failure or profound cyanosis requiring urgent surgery. It may be feasible to detect the MAPCAs on preoperative transthoracic echocardiography (TTE), although imaging them could be difficult at times due to their unpredictable site of origin and course in the mediastinum. In this study, we describe seven cases of MAPCAs diagnosis and ligation under the transesophageal echocardiography (TEE)-guidance in patients undergoing various cardiac surgical procedures (Table 1).

**Material and Method:** 2-D, TEE view of descending Thoracic aorta (DTA) facilitates the MAPCA diagnosis and ligation. Descending aortic short and long axis view (Desc Ao LAX) were obtained at a navigation sector angle of 0, and 90- 100 degrees respectively after rotating the probe leftward from the mid-esophageal (ME) 5-chamber view, to pass all the cardiac structures and using the transducer depth to 4–6 cm to optimize the view. The color Doppler and PWD are used to confirm the MAPCAs as long tortuous vessels with flow towards and away from the probe as red and blue respectively depending upon the direction of the blood flow on color Doppler and pulse wave Doppler.

**Patient selection:** A prospective observational study on potential cyanotic patients of more than 5 Kg, with cyanotic congenital heart defects diagnosed at GIPMER, Tertiary care institute and posted for repair of the

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defects were included. The included patients with TOF, PA-VSD-MAPCAs who were treated and followed up between 01/09/2022 and 31/08/2023 were used to identify patients eligible for inclusion. No exclusion criteria for patients were applied. This case series did not require approval by the Research Ethics Committee as perioperative TEE has been used routinely in cardiac OT for perioperative cardiac monitoring. An informed consent was obtained from the parents of the patients.

**Patient characteristics:** A total of 7 patients has been included as described in table 1. Tricuspid atresia with SA VSD, OP ASD for BD Glenn (1), TA with OS ASD with hypo plastic RV for BD Glenn(1),Infundibular PS with small VSD for VSD closure for RVOT augmentation and muscle bundle excision(1), TOF underwent BD Glenn (3) and total correction including direct VSD closure, RVOT muscle bundle excision and pericardial patch RVOT augmentation (1).

**Table 1:** Describes the details of the 7 patients as age, diagnosis, TTE findings, surgical procedures, and in addition the diagnosis and ligations of MAPCAS before and after the cardiac surgical procedure under the TEE guidance. TEE also delineates the various sites of the MAPCAs in different cardiac lesions.

| Age/ Sex | Wt. (kg) | TTE / CTPA/ Catheterization study findings | Diagnosis | Intra-op TEE findings | Surgery | Details of MAPCAs |
|---|---|---|---|---|---|---|
| 5yM | 15kg | TTE: Tricuspid atresia S/A restrictive VSD, large OP ASD (LàR), both AV valves at same level, incomplete AVSD | Tricuspid atresia, | TTE and CT findings confirmed. Additionally, MAPCAs noted. | BD Glenn | Three MAPCAs with tortuous course noted arising from medial wall of proximal descending aorta with flow towards and away from probe. All ligated intra-operatively. |
| | | No large collateral detected on cardiac catheterization study | SA VSD, | | | |
| | | | OP ASD | | | |
| 15yF | 30kg | TTE: Tricuspid atresia, 3cm OS ASD (RàL), Small restrictive VSD, mild PS | Tricuspid atresia, | TTE and CT findings confirmed. MAPCAs noted as described. | BD Glenn | Multiple MAPCAs with tortuous course noted arising from medial and posterior wall of proximal descending aorta with flow towards and away from probe. All ligated intra-operatively. |
| | | CT: Few thin MAPCAs from left subclavian artery, descending thoracic and arch of aorta, | OS ASD, hypoplastic RV with lumbar scoliosis | | | |
| | | PM VSD 6.2mm, Hypoplastic RV, OS ASD 24mm, Tricuspid atresia, hypoplastic RV, Scoliosis of lumbar vertebrae to left | | | | |
| 1.5yM | 8kg | TTE: Large S/A VSD(RV → *Ao* rta), <50% aortic override, infundibular and valvular PS, hypoplastic MPA | TOF | TTE and CT findings confirmed. MAPCAs noted as described. | BD Glenn | Multiple MAPCAs with tortuous course noted arising from medial wall of proximal descending aorta with flow towards and away from probe. All ligated intra-operatively. |
| | | No MAPCAs noted in cardiac catheterization study | | | | |
| 20yM | 40kg | S/A 2cm VSD (RV→aorta), <50% aortic override, infundibular PS, hypoplastic MPA | TOF | 1 MAPCA from descending aorta TTE and CT findings confirmed. MAPCAs noted as described. | BD Glenn | One large MAPCA noted arising from posterior wall of proximal descending aorta with flow towards the probe. The MAPCA could not be ligated due to its deep-seated location. |
| | | CT: Multiple aorto-pulmonary collaterals noted. S/A VSD 15mm, Infundibular PS, MPA: Aorta ratio<1, Right sided aortic arch | | | | |
| 13yM | 25kg | Large S/A VSD(R→L), <50% aortic override, infundibular PS, hypoplastic PA | TOF | TTE and CT findings confirmed. MAPCAs noted as described. | BD Glenn | Multiple large MAPCAs with tortuous course noted arising from lateral and posterior wall of proximal descending aorta with flow towards and away from probe. All ligated intra-operatively. |
| | | CT: Few MAPCAs from descending aorta >2mm to bilateral hila. S/A VSD 9.4mm, <50% aortic override, infundibular PS, McGoon ratio:1.19 | | | | |

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| | | | | | | |
|---|---|---|---|---|---|---|
| 9yF | 21kg | Severe Infundibular PS, Small S/A VSD (R→L)<br>No MAPCAs noted in cardiac catheterization study | Infundibular PS with small VSD | TTE and CT findings confirmed. Additionally, MAPCAs noted. | Direct VSD closure, RVOT muscle bundle excision, pericardial patch augmentation of RVOT | Multiple MAPCAs with tortuous course noted arising from lateral and posterior wall of descending aorta with flow towards and away from probe. All except one deep seated MAPCA arising from posterior wall ligated intra-operatively. |
| 18yM | 45kg | Large S/A VSD (R→L), 50% aortic override, infundibular and valvular PS<br>CT: S/A VSD 20.1mm, RA/RV dilated, dilated aortic root (34.7mm) and ascending aorta(35.9mm) with aortic over-ride. infundibular stenosis with Stenosis at LPA origin.<br>Multiple dilated MAPCAs from descending thoracic aorta talking tortuous course in mediastinum supplying both hilum/pulmonary circulation. | TOF | TTE and CT findings confirmed. MAPCAs noted as described. | Direct VSD closure, RVOT muscle bundle excision, pericardial patch augmentation of RVOT | Multiple MAPCAs with tortuous course noted arising from medial and posterior wall of proximal descending aorta with flow towards and away from probe. All ligated intra-operatively. |

**Aims and objectives:** We aimed (1). To investigate the presence of MAPCAs in various cyanotic congenital cardiac diseases (2). To detect the site of the MAPCAs in relation to various cardiac lesions in cardiac OT by TEE, (3). To guide and confirm their ligation by TEE (4). The effects of MAPCA on perioperative hemodynamic and morbidity and mortality.

**Results:** The overall MAPCAs diagnosed and ligated in seven patients with congenital cardiac lesions who underwent various cardiac surgical procedures. The 7 patients in whom MAPCAs were diagnosed in OT by TEE belonged to age group of 1.3 yr to 20 yrs. The cardiac lesions included TOF (4). Tricuspid atresia with subaortic VSD and ostium primum ASD (1), TA with Ostium secundum ASD (1), ventricular septum defect with pulmonary stenosis PS (1). The MAPCAs were diagnosed in OR by preoperative TEE after induction of anesthesia and ligated successfully under the TEE guidance. The common sites of MAPCAs in TOF were posterior and anterior wall of DTA, in TA with VSD the site was anterior and medial wall of DTA (Video 1), and in one case they were also arising from the left subclavian artery in addition to DTA, in VSD with PS the origin was from posterior wall (Video 2). All the diagnosed MAPCAs were ligated before Glenn or total correction in 5 patients using same mid-sternotomy in 3 patients, and left thoracotomy in 2 patients. In 2 cases of TA with VSD MAPCAs were ligated after cardiac repair using left thoracotomy (Video 3). The ligation was confirmed by TEE on color Doppler examination and absence of flow velocity on pulse wave doppler. MAPCAs were ligated perioperatively and deleterious effects of their overflow were avoided like endotracheal bleed, desaturation and mortality.

**Video 2**: DTA SAX view of TEE showing 2-3 big MAPCAS originating from the posterior wall of the DTA. Color flow Doppler reveals distal proximal end of the aorta indicating the flow toward and away from the transducer

DTA-SAX- descending thoracic aorta - short axis, MAPCAs- major aorta-pulmonary collateral arteries.

**Conclusions:** Patients with various cyanotic congenital anomalies spontaneously develop MAPCAs to provide additional blood flow to the lungs to maintain arterial oxygenation. However, these can have several deleterious effects during and after CPB like low perfusion pressure, refractory endotracheal bleeding and even death. Many a times these are missing during the cardiac catheterization, or some patients are unsuitable for the cardiac catheterization study. The MAPCAs can be effectively diagnosed in OR by the TEE, and TEE not only diagnose the MAPCAs but also guides and confirms the complete surgical ligation before or after the CPB.

**Keywords:** Cyanotic congenital cardiac diseases; pulmonary circulation; TOF; PA-VSD; TGA; MAPCA; TEE; CPB; Cardiac surgery; MAPCAs ligation

## Introduction

Patent ductus arteriosus and MAPCAs are additional sources of pulmonary blood flow and influence the hypoxic symptoms of TOF and other congenital cyanotic lesions. TOF may be associated with MAPCAs in less than 5% of cases [1]. However, the incidence of MAPCAs increases to 20-25% in patients oof TOF with pulmonary atresia and presents one of the greatest challenges during total corrective surgery [2]. MAPCAS may play a big role in establishing the pulmonary blood flow and arterial oxygen saturation in severe cyanotic congenital cardiac anomalies, and largest MAPCA can be used for antilocalization surgery to reconstruct the pulmonary artery as a single source of pulmonary blood flow in TOF

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with pulmonary atresia [3]. Occasionally, the pulmonary blood flow through the MAPCAs can be excessive, producing high $So_2$, CHF, and possible PVR changes caused by over circulation and high-pressure flow into the PAs. In addition, the MAPCAS can undergo spontaneous rupture resulting in life threatening mediastinal or endotracheal bleeding [4]. The perioperative interruption of the large collateral vessels is often performed either by fluoroscopy-guided embolization in the preoperative or postoperative period [4,5]. Another option is to ligate the collaterals during the surgery [1,6]. Perioperative coil embolization under fluoroscopic guidance either in the preoperative or postoperative period remains an ideal initial management in the patients of TOF with MAPCAs. In addition, MAPCAs ligation during the surgery is another option. However, preoperative coiling of MAPCAs is not feasible in patients with severe desaturation and refractory recurrent Tet spells. Echocardiography, diagnostic cardiac catheterization, CT angiography, and cardiac MRI can all be used to delineate the TOF anatomy and MAPCAS [5-8]. In this study, we describe seven cases of MAPCAs diagnosis and ligation under the transesophageal echocardiography (TEE)-guidance in patients undergoing various cardiac surgical procedures.

## Cases presentations

**Case 1**: 5 yrs- male, weighing 15 kg presented with cyanosis, chest pain and palpitations for one year of age. He was diagnosed with tricuspid atresia (TA) with multiple VSDs and large OP ASD. Cardiac catheterization revealed no major MAPCAs. He was scheduled for Glenn procedure (Video.3) After induction of anesthesia, a pediatric -TEE probe (Size- 11x8mm, Model- GE VIVID S60 N) was inserted and a comprehensive cardiac examination was performed to confirm the preoperative TTE findings. In addition, 2-3. MAPCAs were detected arising from descending thoracic aorta (DTA). At the upper -DTA level, 2-3 MAPCAs of 1-3 mm originating from the anterior wall were found on gentle counterclockwise rotation and zooming of the probe, and the same number were ligated after the Glenn procedure.

**Case 2:** A 15 yrs female, weighing 30 kg presented with cyanosis, palpitations and dyspnea since childhood. TTE revealed TA, hypoplastic RV,3cm OS ASD with right to left shunt, small PM VSD with a pressure gradient of 90mmHg and mild PS. CT revealed MAPCAs arising from left subclavian artery and distal aortic arch. She was scheduled for the bidirectional Glenn procedure. After anesthesia induction, an adult -TEE probe was inserted, and a comprehensive cardiac examination was performed using an ultrasound system (Model- GE VIVID S60 N) to confirm her preoperative TTE findings. In addition, on zooming the DTA view at 0 and 90 degrees, multiple MAPCAs were detected arising from medial aspect of the descending thoracic aorta (Video1). The color-Doppler confirmed their coursing towards and away from the probe. The MAPCAs were ligated via trans-sternotomy under the TEE guidance before the Glenn procedure.

**Video 1**: Mid- esophageal, DTA SAX view of TEE: Color Doppler examination reveals multiple tortuous vessels as MAPCAS arising from the medial wall of the DTA. The blue and red color confirms the flow away and towards the transducer and depicts distal and proximal aorta.

DTA- descending thoracic aorta, SAX- short axis, TEE- trans-esophageal echocardiography, MAPCAs- major aorta-pulmonary collateral arteries.

**Case 3**: A 1.5-year-old male, weighing 8 kg, child presented with cyanosis since birth. He was diagnosed as TOF. TTE revealed a large subaortic VSD, infundibular and valvular PS, severe aortic override >90%. Cardiac catheterization was not performed. After anesthesia induction, a pediatric TEE probe (Size- 11x8mm, Model- GE VIVID S60 N) was inserted and a comprehensive cardiac examination was a performed, which also revealed a 1.9cm, VSD with bidirectional shunt, a PFO with bidirectional, large PDA, LVIDD 1.6cm, posterior wall thickness of 0.4cm, LV mass of 14.81 Gm, McGoon's ratio 0.6, and EF of 65%. In addition, multiple MAPCAs were detected arising from the descending aorta. At the proximal -DTA level, several MAPCAs of different sizes, originating from the medial wall of the DA and traversing in multiple directions, and showing blood flow towards and away from the probe were detected on zooming and gentle counterclockwise rotation of the probe. All the MAPCAs were confirmed and ligated before establishing CPB by the operating surgeon via same mid-sternotomy incision used for Glenn procedure, to ameliorate the deleterious effects of MAPCAs during CPB. Color- flow Doppler TEE examination also confirmed the absence of the flow, and the optimum ligation of the multiple MAPCAs. (Video 3).

**Video 3:** Upper -Esophageal, Ascending Aorta short Axis view of TEE revealed a laminar flow (blue color) in the RPA as an unobstructed and functional Glenn, depicted above the ascending aorta. RPA-right pulmonary artery.

**Case 4:** A 20 Years - 40 kg male presented with dyspnea on exertion, and chest pain. He was diagnosed as TOF. TTE revealed 2cm subaortic VSD with R-L shunt, dilated RA, severe infundibular PS, < 50% aortic override. CT also confirmed the diagnosis of the TOF, and in addition also revealed multiple MAPCAs. He was scheduled to undergo total correction for TOF. After induction of anesthesia, an adult TEE probe was inserted for comprehensive cardiac examination. TEE evaluation also confirmed the findings suggestive of TOF. In addition, TEE examination on 2-D and color doppler examination of DTA view (0 and 90 degrees) also revealed multiple MAPCAs originating from the anterior and posterior wall of the descending aorta, and several of these were ligated via sternotomy under the TEE guidance before the total correction of the TOF.

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**Case 5:** A 13-year male weighing 37 kg presented with DOE, cyanotic spells. TTE revealed subaortic VSD with a gradient of 76 mmHg, infundibular and valvular PS, hypoplastic PA, LV -15 Gm, < 50% aortic override suggestive a diagnosis of TOF. CT revealed MAPCAs arising from the descending aorta. The patient was operated on for the Glenn procedure. TEE examination also confirmed poor LV with 23 Gm mass and in addition multiple MAPCAs originating from the posterior wall of the descending aorta and distal aortic arch and traversing towards and away from the probe were also revealed and ligated under the TEE guidance before the Glenn procedure via the left thoracotomy (Video 4).

**Video 4**: The Aortic arch LAX view of the aortic arch of TEE: the color doppler depicts multiple MAPCAS from the anterior and posterior wall of the distal aortic arch.

**Case 6:** 1.5 yrs, 10 kg male child presented with dyspnea and repeated chest infections for 3 months of age. TTE revealed a large peri membranous VSD (1.4 cm) with a pressure gradient of 33 mmHg and left to right shunt and muscle bundles in the RVOT, and normal biventricular functions. The patient was posted for excision of RVOT muscle bundles and the VSD patch repair. After induction of anesthesia the pediatric TEE probe was inserted. The TEE examination also confirmed the large peri membranous VSD and muscle bundles in the RVOT. In addition, it also revealed two big MAPCAs arising from the posterior wall of the descending aorta, and color Doppler examination showed the blood flow towards the probe. Both MAPCAs were ligated via the trans -sternotomy route under the TEE guidance after the intracardiac repair.

**Case 7:** 18 yrs, 40 Kg male presented with cyanosis and dyspnea on exertion's revealed large S/A VSD with R-L shunt, more than 50% aortic over ride, infundibular and valvular PS, CT also confirmed S/A VSD20.1 mm, RA and RV dilated, dilated aortic root(34.7 mm) and ascending aorta(35.9mm) with aortic override, infundibular stenosis and stenosis at LPA origin. In addition, multiple dilated MAPCAs from descending thoracic aorta traveling tortuous course in the mediastinum supplying hilum pulmonary circulation was confirmed. Direct VSD closure, RVOT muscle bundle excision, pericardial patch augmentation of RVOT were performed under standard moderate hypothermic CPB. Multiple MAPCAs with tortuous course noted arising from lateral and posterior wall of descending aorta with flow towards and away from probe were confirmed by TEE. All except one deep seated MAPCA arising from posterior wall ligated intra-operatively.

## Discussion

The arteries originating from the aorta or its branches supplying the pulmonary blood flow for a segment or a lobe of the lung without any connection to the central pulmonary arteries are defined as MAPCAs and develop spontaneously providing additional blood flow to the lungs as the major contributors. Indeed, MAPCAs are frequently encountered in cyanotic cardiac anomalies such as severe TOF with PA, tricuspid atresia (TA), single ventricle pathology, d- TGA, DORV, truncus arteriosus etc. [1,9] TOF patients with significant RVOT obstruction or pulmonary atresia may require some additional sources of pulmonary blood flow from PDA or MAPCAs for maintaining the arterial oxygen saturation, and frequently MAPCAs serve as the main source of pulmonary flow.[10] At times, MAPCAs serve as the primary, often sole, source of blood supply to the lungs when the native pulmonary arteries are severely hypoplastic or absent. MAPCAs are rare, and present in 4–10 per 10,000 live births and 2–3% of all CHDs. They are commonly associated with PA/VSD (50–70%), and 5–10% of TOF. Incidence of MAPCAs increased to 20% to 25% of patients with TOF with pulmonary atresia.[11] In the presented cases series, the MAPCAs have been reported in patients of TOF with PS and PA (4cases), and tricuspid atresia (2 cases) and VSD with PS (1 case). Although the precise mechanisms that lead to the development of the MAPCAs are incompletely understood. However, chronic hypoxia, high hemoglobin levels, diminished global or regional pulmonary blood flow and non-pulsatile flow in the pulmonary arteries may contribute for the development of MAPCAs in these patients [1]. The MAPCAs most frequently arise from DTA and aortic arch, but can also originate from the ascending aorta, head and neck vessels (most commonly the subclavian artery), abdominal aorta and even from the coronary arteries or from the infra-diaphragmatic arteries.[12,13] In the presented case series, The common sites of MAPCAs in TOF were posterior and anterior wall of DTA, in TA with VSD the site was anterior and medial wall of DTA, and in one case they were also arising from the left subclavian artery in addition to DTA, in VSD with PS the origin was from posterior wall. All the diagnosed MAPCAs were ligated before Glenn or total correction in 5 patients using same mid-sternotomy in 3 patients, and left thoracotomy in 2 patients. In 2 cases of TA with VSD, MAPCAs were ligated after cardiac repair using left thoracotomy On radiological assessment, These have been classified as [1] single-supply (MAPCA serves as the only source of blood flow to the lung segments), with no connection to a central PA, (2) isolated supply to central PAs (the only MAPCA connecting to the central PAs; (3) dual-supply (1 of ≥2 MAPCAs connecting to the central PAs); or (4) mixed-supply (≥1 single-supply and ≥1 dual-supply branches) [12]. MAPCAs may also act as a dual supply, meaning that the central pulmonary arteries arborize normally, with no lung segments receiving blood flow solely from a MAPCA without connection to the central PA system. The pulmonary blood flow through the MAPCAs can be excessive, causing variable clinical presentation, producing high SaO2, CHF, and possible PVR changes caused by

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over circulation and high-pressure flow into the pulmonary arteries.[14]

The evaluation of TOF with PA and MAPCAs should include chest radiography, magnetic resonance imaging (MRI), and multidetector computed tomography (MDCT) scanning. 2-D echocardiography with color flow and pulse wave Doppler is the most important tool in the diagnosis of tetralogy of Fallot with pulmonary atresia. Color-flow imaging may help to identify sources of pulmonary artery blood flow, including the PDA and MAPCAs. A systematic approach is recommended to ensure visualization of all the zones of the descending thoracic aorta (DTA). A common method is to begin the examination after obtaining the ME 4-chamber view and turning the probe to the left until the descending aorta is seen in SAX at 0 to 15 degrees. From here, the transducer depth is decreased to 6 to 8 cm, and the probe is withdrawn or advanced, following the course of aorta. If collaterals are suspected, echocardiography alone is inadequate for complete delineation of pulmonary blood flow, and further imaging by MRI or angiography is recommended, as viewing them could be difficult at times due to their unpredictable site of origin and course in the mediastinum [5-8]. MRI and MDCT may be used as a non-invasive method and provide excellent delineation of the pulmonary arterial circulation and their MAPCAs. Cardiac catheterization with angiography is recommended not only for delineation of TOF pathology to facilitate surgical planning but also the presence MAPCAs that may need to be either ligated or incorporated into the repair. Traditional angiography remains gold standard for anatomic delineation of MAPCAs and central PAs, and cardiac catheterization is always performed in cases of unclear distribution. But this is not available to all the TOF patients. Preoperative cardiac catheterization evaluation and MAPCAs ligation is limited in patients with very low arterial oxygen saturation and refractory recurrent Tet spells. This is essential for mapping the anatomy, identifying connections between the MAPCAs and the pulmonary arteries, assessing the presence of narrowing (stenosis), and measuring blood pressure within the collaterals before the cardiac surgery. However, many a times either these facilities are unavailable or children are unsuitable due to poor hemodynamic such as uncompensated heart failure or profound cyanosis requiring immediate surgery. In such scenarios, it may be feasible to detect the MAPCAs on preoperative transthoracic echocardiography (TTE), although imaging them could be difficult at times due to their unpredictable site of origin and course in the mediastinum. In this study, we describe seven cases of MAPCAs diagnosis and ligation under the transesophageal echocardiography (TEE)-guidance in patients undergoing various cardiac surgeries. Differential diagnosis of MAPCA on echocardiography includes PDA and posterior intercostal arteries (PICA), which may arise from the distal aortic arch or DTA. The MAPCA should be differentiated from a PDA based on number, size, origin, and course. multiple vessels having similar Doppler flow characteristics are invariably aortopulmonary collateral arteries, which must be differentiated from the PDA. The color Doppler and spectral Doppler examination of a PDA reveals turbulent PDA jet with marked spectral dispersion in systole and diastole (throughout the cardiac cycle) along the anterolateral wall of the main PA probably because of the anatomic arrangement [15,16].

Free flowing MAPCAs have several detrimental effects during CPB, such as a significant systemic to pulmonary shunt causing flooding of the surgical field and interfering the surgery, increased blood return in the CPB circuit, stealing from the cerebral circulation, and persistent low perfusion pressure, reduces effective systemic blood flow or even life threatening mediastinal or endotracheal bleeding due to spontaneous rupture [4,17,18]. The presence of MAPCAs is a significant factor associated with late mortality following cardiac corrective surgery [19]. Thus, MAPCAs have a great impact on CPB management; a bypass flow index of 2.4 L/min/m (2) may not be sufficient, and the maximum requirement of bypass flow index may be 3.2 L/min/m (2) or more in this patient population to maintain the tissue perfusion and normal lactate levels. MAPCAs may contribute low output throughout surgery which can lead to cerebral anoxia and renal hypoperfusion and devastating postoperative sequalae [17]. MAPCAs should be occult included in the cardiac catheterization lab. or should be ligated via thoracotomy before initiation of CPB to avoid the generalized tissue hypoperfusion particularly of the brain. This is the most effective method for preventing a dangerous steal from systemic circulation during CPB. It will also accelerate postoperative recovery [20,21]. Contrary, the MAPCAS may play a big role in establishing the pulmonary blood flow and arterial oxygen saturation, by utilizing largest MAPCA for antilocalization surgery to reconstruct the pulmonary artery in TOF with pulmonary atresia [3]. Lung reperfusion injury in the setting of antilocalization surgery has been estimated in the previous studies at 65% [22]. It is not always possible to perform cardiac catheterization and coil embolization of MAPCAs in a TOF with severe pulmonary artery atresia, risking precipitation of very low arterial oxygen saturation and recurrent refractory Tet spell after coil embolization [23]. In this case series, the MAPCA ligation could not be performed in the preoperative period due to persistent low oxygen saturation and patients were exhibiting repeated Tet spells. However, perioperative TEE evaluation diagnosed MAPCAs commonly originating from the proximal descending aorta or aortic arch and successfully ligated via mid-sternotomy before establishing the CPB or post repair. In addition to the TTE values, the TEE findings also confirmed inoperability for total TOF correction in five patients, and the Glenn procedures were performed as

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LV and RV were borderline, LV mass was inadequate and inadequate Mc Goon ratio i.e. 0.5-0.8, (normal average 2.1), a ratio below 0.8 is deemed inadequate for complete repair of pulmonary atresia cases with TOF anatomy (TOF/PA).[24]

## Conclusions

All patients with significant MAPCAs should undergo coil embolization prior to total correction. However, in patients with TOF/ PA/ MAPCAs and severe hypoxemia due to refractory recurrent Tet spells, preoperative cardiac catheterization and coil embolization is unsafe and risking desaturation and morbidity and mortality. In such a high-risk patient, the MAPCAs can be successfully identified and confirmed or first time detected by intraoperative TEE. These can be surgically ligated before initiation of CPB, or after coming off bypass via trans-sternotomy or thoracotomy route under the TEE guidance. Surgical ligation of MAPCAs under the TEE guidance is safe, easy, and effective, and prevents the devastating complications of MAPCAs on CPB as well as postoperatively.

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## Declaration:

All authors were involved in the patient's management and preparation of the manuscript.

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