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Perioperative major aortopulmonary collateral arteries (MAPCAs) coiling in tetralogy of fallot patients

Abstract

Background: Tetralogy of Fallot (TOF) is characterized by ventricular septal defect, an overriding aorta, varying degrees of pulmonic stenosis, and right ventricular hypertrophy. It may be associated with a number of lesions including development of major aortopulmonary collateral arteries (MAPCAs) in less than 5% of cases. In this study we just focused on the management of MAPCAs.

Methodology: TOF patients with significant MAPCAs were selected after cardiac catheterization and planned for transcatheter embolization just before surgery.

Results: A total of 45 patients underwent transcatheter closure of MAPCAs just before surgical correction with 100% success rate. The age varied from 3-30years and 55% (n=25) were female. In majority 60% (n=27) the procedure was carried out under deep sedation with local anaesthesia while 18 (40%) patients had general anaesthesia. There was insignificant residual leak in 2 (4%) patients, transient bradycardia (n=1) and hypercyanotic spell in 4% (n=2). No serious complications or death occurred during this study period.

Conclusion: Transcatheter coiling of MAPCAs is safe, easy, and effective procedure and protects the patient from its long term devastating complications.

Keywords: tetralogy of fallot, transcatherter closure, major aortopulmonary collateral arteries

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Abbreviations: TOF, tetralogy of fallot; MAPCAs, major aortopulmonary collateral arteries; CHD, congenital heart disease; APVS, absent pulmonary valve syndrome; CAVSD, complete atrioventricular septal defect

Introduction

Major aortopulmonary collateral arteries (MAPCAs) are blood vessels that bring systemic blood flow to the pulmonary arteries. They develop in response to decreased pulmonary blood flow and cyanosis. MAPCAs may be an additional source of blood supply to the lungs along with the native pulmonary arterial supply in less than 5% patients with TOF. Sometimes these MAPCAs may provide enough blood to the lungs so that the patients appear pink and pose difficulty and late in diagnosis. On the other hand MAPCAs may the sole supply to the lungs and the life will be dependent on these collaterals as in patients with TOF with Pulmonary atresia. In this study we have focused only in patients with TOF with pulmonary stenosis who developed MAPCAs in due course of time.

TOF is one of the most common cyanotic congenital heart disease (CHD) and accounts for 3.5-9% of CHD.[1,2] The anatomic spectrum is wide and may include TOF with pulmonary stenosis, TOF with pulmonary atresia, TOF with absent pulmonary valve syndrome (APVS) and TOF with complete atrioventricular septal defect (CAVSD). TOF with pulmonary stenosis comprises 79.7% of all cases and have variable clinical spectrum.[3–5] Total correction is an ideal treatment in these patients and should be offered as early as possible in order to prevent the long term complications.[6] In industrialized countries total correction is done usually before 6 months of age or in the neonatal period.[7–9] In developing countries like Pakistan, multiple factors including illiteracy, late referral, poor socioeconomic status and shortage of professional hands and tertiary care institutions in this field play important role in the delay of treatment. The chronic hypoxia and high hemoglobin levels may contribute for the development of MAPCAs in these patients [a].

Our institution is one of few centers that is providing a wide range of transcatheter interventional and surgical management in patients with CHDs. During the initial period of paediatric cardiac surgery at our center, few patients after total correction for TOF developed pulmonary edema underwent cardiac catheterization that revealed significant MAPCAs, which were coiled and resulted in smooth postoperative recovery. In this study we analyzed the success and effectiveness of perioperative transcatheter MAPCAs coiling in patients undergoing total correction for TOF with pulmonary stenosis.

Materials and methods

A total of 45 patients with TOF with pulmonary stenosis underwent MAPCAs coiling during Jan 2011 to Dec 2014. These patients had previous cardiac catheterization and planned for transcatheter MAPCAs coiling one day before or on the day of surgery. All patient who had significant MAPCAs (>2mm) were included in the study. The patients were admitted in the hospital and a written consent was taken before the procedure. A complete blood picture, c-reactive protein, renal functions and saturations were taken before the procedure. We continuously monitor the saturations throughout the procedure and after the MAPCAs coiling till the patient undergoes for total correction. After femoral arterial access, an aortogram was done with pigtail in all the patients to visualize the MAPCAs. Then the MAPCAs were engaged with Judkin Right (JR) catheter and selective angiogram was done for origin, size and distribution of MAPCAs. The JR catheter advanced as far as possible from the main vessel into the collateral and free coil taken and load into the catheter and advanced

with the hard end of .038 guide wire. The cook free coils delivered in the collaterals and then contrast injection given to check for any residual leak or other collateral (Figure 1&2). After completion of the procedure the patients were shifted to ward for monitoring and then preparation for total correction.

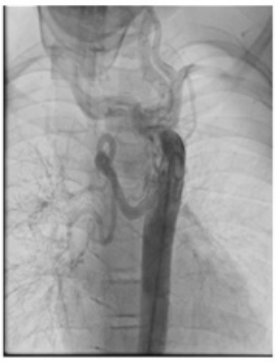

Figure 1

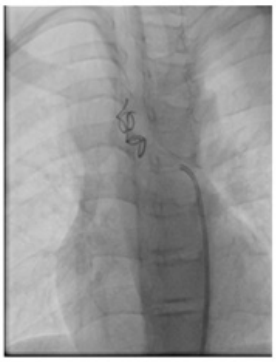

Figure 2

Figure 1&2 Showing large MAPCA and in situ cook free coils resulting complete occlusion.

Results

100% patients (n=45) had successful closure. The age varied from 3.2-30years and 45% (n=20) were male while 25 were female patients. The procedure was carried out with deep sedation with local anaesthesia in 27 (60%) while 40% had general anaesthesia. The mean procedure time was 55±31minutes (25-130min) and mean fluoroscopic time was 20±19minutes (4-91min). The minor complication rate remained 11% (n=5) including slight residual leak in 2, transient bradyarrhythmia in 1 patient. The two patients desaturated significantly immediately after embolization of two MAPACAs and shifted to operation theater for total correction with good postoperation result. There was no vessel wall perforation, bleeding, hematoma formation, coil embolization or death during this period. Data was entered in SPSS-17 and descriptive analysis including frequencies with percentages, mean and standard deviations were calculated as shown in Table 1.

Table 1 Showing demographic variables

	Age (years)	Height (cm)	Weight (kg)	Procedural Time(min)	Fluoroscopic Time(min)
Mean	15.738	142.1538	34.8077	55.12	20.7885
Std. Deviation	7.446	24.87198	14.18737	31.033	19.17194
Minimum	3.2	90	11	25	4
Maximum	30	168	57	130	91

Discussion

All patients after percutaneous MAPCAs coiling had smooth surgical and postoperative recovery in our study. Although two patients desaturated significantly after coiling and became unstable so surgery was performed immediately with uneventful postoperative course. MAPCAs are basically persistence of splanchnic circulation and the word 'Major' because they are not acquired one.[10,11] MAPCAs can develop in other conditions like TOF with pulmonary atresia and in this condition pulmonary circulation is dependent on these collateral.[12] MAPCAs can be an additional source of blood flow to lungs along with native pulmonary arteries like TOF with pulmonary stenosis.[13,14] A number of factors influence the development of MAPCAs including hypoxia and cyanosis to augment pulmonary blood flow.[15] Tetralogy of Fallot with MAPCAs is reported in about 2% of the cases in pediatric population, while in unrepaired adults, it is much more common ranging between 13% and 25%.[b]

The development of MAPCAs is much more common in our patients due to chronic hypoxia and dense cyanosis due to delay of treatment as these are contributing factors.[16–19] On contrary in developed countries patients with TOF with pulmonary stenosis have usually total correction before 6 months of age[7–9] so the development of MAPCAs is very unlikely in these patients.

MAPCAs can result in a number of complications including gross enlargement with erosion of bronchi resulting massive hemoptysis.[20] Occlusion of the MAPCAs before open heart surgery is important because otherwise there is excessive return to the left heart when the aorta is cross clamped on cardiopulmonary bypass, flooding the operative field thus interfering the surgery. MAPCAs may contribute low output throughout surgery which can lead to cerebral anoxia and renal hypoperfusion and devastating postoperative sequalea.[21] If remain undetected can lead to pulmonary edema after operation and difficulty in weaning off the patient thus prolonging the ITC stay.[22] In the long term postoperatively patients may develop CCF refractory to medical treatment.[23] Considering all these necessitates that all MAPCAs in patient with TOF with pulmonary stenosis should be closed before operation.[24]

The closure can be done by two ways either surgical ligation or transcatheter embolization.[22,23] The surgical ligation requires extensive surgery and tissue damage, difficult to locate, excessive blood loss and prolongs the procedure time. However large MAPCAs may be easily located by the expert surgeon and ligated. In the recent years the emerging technique of percutaneous approach is easy and effective as in our study all patients (n=45) had successful closure (Figure 1&2). The mean procedure time remained 55min. sometimes one may have difficulty via transcatheter coiling as in one of our patient MAPCAs were arising from left subclavian artery and had difficulty in engaging the collaterals so the procedure time was prolonged, however MAPCAs from descending aorta were straight forward with much less procedure and fluoroscopic time.

Conclusion

Transcatheter MAPCAs embolization is safe, easy and effective and prevents the devastating long-term morbidity and mortality in patients with TOF with pulmonary stenosis. All patients with significant MAPCAs should undergo coil embolization prior to total correction.

Ethical issue

An approval from ethical committee of the institute taken before the study.

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Conflicts of interest

Author declares there are no conflicts of interest.

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