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Research Article

Frequency of Major Aorto-Pulmonary Collaterals in Patients with Tetralogy of Fallot in Our Patients

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ABSTRACT

Introduction: Major Aortopulmonary Collateral Arteries (MAPCAs) are blood vessels that bring systemic blood flow to the pulmonary arteries, are a complex and heterogeneous condition. They develop in response to decreased pulmonary blood flow and cyanosis. Our institutional approach to this lesion emphasizes early management before complete repair.

Objectives: To determine the frequency of major aorto-pulmonary collaterals in patients with tetralogy of fallot in our patients.

Materials and methods: Cross sectional retrospective study planed in Children's Hospital and ICH Lahore, from January 2010 to December 2018. All children having Tetralogy of Fallot form 1 year above enrolled in the study underwent standard cine-angiograms after obtaining written consent following basic laboratory workup and ethical committee approval. Data was analysed using SPSS version19.The frequency, median with range was calculated for ordinal and scalar variables due to non-normally distributed data. Student T test was applied to check any significant difference between various variables with p less than 0.05 considered as significant.

Results: 425 patients of tetralogy of fallot were enrolled and 398 completed cardiac catheterization. The median age was ± 6 yrs with male predominance of 2.1:1. Confluent Branch pulmonary arteries were present in 395 (99%) children with 2 having disconnected LPA and 1 having PDA continuing as RPA. Pulmonary artery abnormalities seen in 72(18%) patients. 211(53%) patients had MAPCAs, while 88 (22.11%) had 2 or more MAPCAs. 195(92%) had hemodynamically significant MAPCAs i.e., supplying 3 or more lung segments.54 (28%) had small i.e., <1.33 mm at origin caliber. 105 (54%) had moderate size i.e., 1.33-1.67 mm and 36(18%) had large size i.e.,>1.67 mm at origin caliber. 166 (57%) MAPCAs were mainly arising from descending aorta, 54(19%) arising from right Subclavian artery, 45 (16%) from left Subclavian artery. 14(5%) from right internal mammary artery and 9(3%) left internal mammary artery.

Conclusion: The frequencies of major aortopulmonary collateral arteries were quite high in our population. Diagnostic cardiac catheterization is still a relevant invasive diagnostic as well as therapeutic procedure in children with tetralogy of fallot.

Keywords: Cardiac anomalies; Congenital heart defect; Angiography; Ventricular septal defect; Cardiac catheterization

INTRODUCTION

Tetralogy of Fallot (TOF) is among the most common cyanotic congenital heart disease in children contributing to 3.5-9 % of all congenital heart diseases [1,2]. Tetralogy of Fallot characterized by four peculiar features i.e. large Ventricular Septal Defect (VSD), Right Ventricular Outflow Tract (RVOT) obstruction, over riding of aorta and right ventricular hypertrophy [3]. The clinical features of TOF primarily depend on severity of Right Ventricular Outflow Tract (RVOT) obstruction and degree of aortic overriding. The more severe infundibular stenosis the more dramatic presentation of child with characteristic Tet spell. Optimal age for primary repair of TOF is 6-12 months of age [4,5].

Tetralogy of Fallot (TOF) with Major Aortopulmonary Collateral Arteries (MAPCAs) is a complex and heterogeneous disease with varying degrees of severity. While MAPCAs are present in 20% to 25% of patients with TOF and pulmonary atresia, the disease is rare and management varies considerably [6,7]. A number of investigators have described approaches to treatment for TOF with MAPCAs, but most series have been relatively small and

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results have been mixed [8].

Diagnostic right and left heart catheterization have been an integral part of diagnostic workup for TOF until late 1970's. With the advent for better echocardiographic assessment, the anatomic details can accurately established through this non-invasive test in children less than one year of age. Cardiac catheterization has thus become less popular due to its invasive nature and additional radiation exposure, especially in children with adequate pulmonary vasculature [9,10].

In developing country like Pakistan, numerous factors including lack of education, delayed referral, poor socioeconomic status, and lack of expertise in this arena frequently result in delayed diagnosis and treatment. Therefore, chronic long-standing hypoxia and polycythemia can contribute towards re-canalization of aorto-pulmonary collaterals, which cannot be evaluate through echocardiography alone. Such collaterals result in potential steal phenomenon on bypass during surgery and peri-operative pulmonary hemorrhage. Therefore, our institution establishes a policy to catheterize all patients with TOF preoperatively, not only to determine the underlying anatomical variations, also looked for significant additional Ventricular Septal Defects (VSDs) and hemodynamically significant Major Aortopulmonary Collateral Arteries (MAPCAs).

During the initial period of paediatric cardiac surgery at our centre, 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. The purpose of this study is to evaluate the frequency of MAPCAS, its hemodynamic significance, numbers and sites. Currently very little literature found regarding MAPCAS in tetralogy because most centres started doing corrective surgery before 6 months of life.

MATERIALS AND METHODS

Cross sectional retrospective study planed in Children's Hospital and ICH Lahore, from January 2010 to December 2018. All children having Tetralogy of Fallot or TOF with Blalock-Taussig (BT) form 1 year above enrolled in the study underwent standard cine-angiograms after obtaining written consent following basic laboratory workup and ethical committee approval.

Exclusion criteria

Patients having marked branch pulmonary hypoplasia in CT angiography, complete atrioventricular septal defect with TOF and Pulmonary atresia with VSD, complex cardiac lesion with pulmonary stenosis or isomeric children with pulmonary stenosis.

Anatomic characterization

Echocardiography was used primarily to determine intra-cardiac anatomy. The anatomy of the pulmonary blood supply, including the presence of central pulmonary artery and the nature of MAPCA supply, was determined primarily by angiography or computed tomography. Computed tomography can be used as the primary imaging modality in a subset of cases in which it clearly demonstrates all MAPCAs to be 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 pulmonary artery system. However, if a potential pulmonary artery to MAPCA connection cannot be defined adequately by computed tomography or the central pulmonary artery do not arborize normally; cardiac catheterization is performed for anatomic delineation and surgical planning. Traditional angiography remains our gold standard for anatomic delineation of MAPCAs and central pulmonary arteries, and cardiac catheterization is always perform in cases of unclear distribution.

Angiography procedure

Hemodynamically stable children with room air saturation more than 75% and not having any history of Tet spells were catheterize under local anaesthesia with aseptic technique. All patients kept sedated and pain free during the procedure, using midazolam and ketamine [11]. Children with history of Tet spells or room air saturation less than 75% and those who were hemodynamic unstable were catheterized under general anaesthesia. After femoral arterial access, an aortogram was done with pigtail in all the patients to visualize the MAPCAs with high pressure injection using non-ionic dye. Then the MAPCAs were engaged with Judkin Right (JR) catheter and selective angiogram was done for origin, size and distribution of MAPCAs. Right and left cardiac catheterizations carried out. Pressures and oximetry recorded and cine-angiograms were performed in the recommended positions [12]. Isolated MAPCAS angiography done at Anterio-Posterior (AP) view. For site detection selected angiography was performed at descending aorta, right and left Subclavian arteries. Pulmonary artery size measured by using z-scores. A vessel diameter of less than −3 z score was labelled as hypoplasia [13].

Statistical analysis

Data were analysed using SPSS version19. The frequencies, median and range calculated for ordinal and scalar variables due to non-normally distributed data. Student T test was applied to check any significant difference between various variables with p less than 0.05 considered as significant.

RESULTS

A total 425 patients underwent cardiac catheterization. Twenty-seven patients not compatible with inclusion criteria after cardiac catheterization, therefor they were excluded. The age range from 1 to 16 yrs with male predominance of M: F 2.1:1. Weight range from 6 kg-44 kg with median height of ± 102 cm and surface area of ± 0.63 m2. All patients had levocardia (heart lying in normal left hemi thorax) except 3 patients with dextrocardia, one of whom had sites inverses as well.

37 (9.3%) children with TOF had modified BT shunt done previously because of Tet spells or worsening hypoxemia. Regarding associated abnormalities, 17.8% have right aortic arch and 9% isolated LPA stenosis (Table 1).

Table 1: Patient characteristics.

Associated cardiac lesion	Frequency (percentage)
Bilateral SVC	30 (7.5)
Additional muscular VSDs	14 (3.5)
Isolated LPA stenosis	36 (9)
Coronary artery abnormalities	11 (2.8)
PDA	9 (2.4)
Right aortic arch	71 (17.8)
MAPCAs	283 (53)

Confluent Branch pulmonary arteries were present in 395 (99%) children with 2 having disconnected LPA and 1 having PDA continuing as RPA. Median MPA z score was -0.17 (median range -1.8-4.5), RPA median z-score was -0.18 (median range -0.9-4.3) and LPA median z-score was -0.2 (median range -1.9-4.6). Pulmonary artery abnormalities identified in 72 (18%) patients. The most common anatomical abnormality not previously documented on echo was isolated LPA origin stenosis 36 (9%), followed by bilateral proximal branch pulmonary artery hypoplasia 18 (4.5%) and isolated RPA stenosis 14 (3.5%) (Table 2).

211 (53%) patients of tetralogy had MAPCAs, while 88 (44.7%) children having two or more MAPCAs. Regarding origins of these MAPCAs, 166 (57%) MAPCAs were mainly arising from descending aorta, 54 (19%) arising from right Subclavian artery, 45 (16%) left Subclavian artery, 14 (5%) form right internal mammary artery and 9 (3%) left internal mammary artery (Figure 1).

195 (92%) had hemodynamically significant MAPCAs i.e., supplying three or more lung segments. Among the children with hemodynamically significant MAPCAs, 54 (28%) had small in size (<1.33 mm at origin), 105 (54%) had moderate in size (1.33-1.67 mm at origin) and 36(18%) had large in size (>1.67 mm at origin) (Figure 2).

There was no significant difference in frequency of MAPCAs with gender, age, weight, height or surface area (Table 3).

Table 2: Branch pulmonary arteries and Z-score of patients.

	N	Range	Minimum	Maximum	Mean		Std. Deviation	Variance
	Statistic	Statistic	Statistic	Statistic	Statistic	Std. error	Statistic	Statistic
MPA	400	28.37	4.25	32.62	12.247	0.2193	4.387	19.249
RPA	400	32.35	3.46	35.81	11.04	0.2181	4.362	19.027
LPA	400	29.93	2.8	32.73	11.064	0.2152	4.304	18.531
Z- score (MPA)	399	6.29	-1.787	4.51	-0.0068	0.0496	0.991	0.984
Z –score (RPA)	400	13.577	-0.9148	12.663	0E-7	0.05	1	1
Z –score (LPA)	400	6.886	-1.91	4.969	0E-7	0.05	1	1
Valid N (list wise)	399							

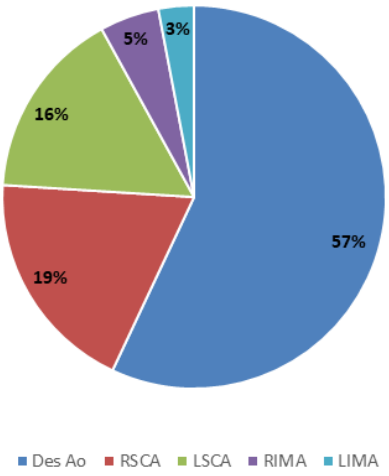

Figure 1: Distribution of MAPCAs.

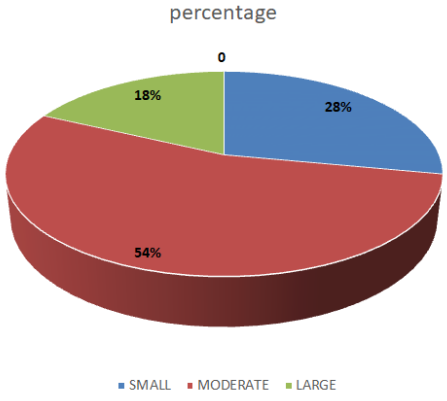

Figure 2: Hemodynamically significant MAPCAs size distribution.

Table 3: MAPCAs relation with demographic data of patients.

	Test Value = 0					
	t	df	Sig. (2-tailed)	Mean Difference	95% Confidence Interval of the Difference	
					Lower	Upper
Sex	56.515	399	0	1.3225	1.2765	1.3685
Age	35.001	399	0	6.559	6.1906	6.9274
Weight	45.641	399	0	16.05075	15.3594	16.7421
Height	113.01	399	0	104.315	102.5004	106.1296
Surface area	68.435	399	0	0.67407	0.6547	0.6934

DISCUSSION

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 be 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 [14].

Tetralogy of Fallot is one of the most common cyanotic congenital heart defect beyond neonatal period of life with an incidence of 0.28/1000 live births requiring surgical repair [15]. Cine-angiogram has been the gold standard for pulmonary vasculature evaluation prior to surgery [16]. Magnetic resonance angiography has been used but frequently requires anaesthesia in small children and doesn't allow intervention [17,18]. In developed world, the diagnosis of TOF is frequently making prenatally [19]. However, in developing countries like Pakistan there are many social factors in addition to lack of expertise.

In our study, majority of patients were relatively older between 5 and 15 years of age due to less efficient primary and secondary healthcare structure. TOF has equal gender distribution [20], though in our study, there was a male predominance. This finding was more of a social norm rather than a true statistical difference as previously documented in various local studies [21].The overall incidence of pulmonary artery abnormalities in our population was found to be 18%, which was comparable to the data reported by Sharma et al [21]. Bacha et al. also observed pulmonary artery abnormalities in 20% cases [22]. The incidence recorded in various other studies have been as high as up to 39%, though unlike our study, they had included all forms of TOF and hence cannot be standardized [23]. Isolated LPA stenosis was identify in 9% of our study cases. This finding was equal to the reported occurrence of 3% and 10% from Asia and Europe respectively [24,25]. RPA stenosis (3.5%) was also comparable with data by Farsani, where it was found to be 2.2% [26,27]. A local study conducted at National Institute of Cardiovascular Diseases (NICVD) Karachi, showed 5 out of 31 patients (16%) with TOF had markedly hypoplastic pulmonary arteries [9].

Additional muscular VSD's were present in 3.5% of our patients. This is again comparable to other workers study [12]. There is no functional importance of right sided aortic arch but its presence signifies to paediatric cardiologists to further investigations in Tetralogy of Fallot patients. Right aortic arch was present in 17.8% patients similar to previously reported incidence of 20%-25% cases [22]. It was also comparable to study conducted locally in Pakistan [20].

Major Aortopulmonary Collateral Arteries (MAPCA) were identifying in 211(53%) patients in our study. The incidence was higher than previous reports showing less than 1%-5% cases [23]. MAPCAs are congenital vascular malformations that bring systemic blood flow to the pulmonary arteries so they represent persistence of splanchnic circulation. They can re-canalize in response to diminish pulmonary blood flow and hypoxemia. Hemodynamically significant MAPCAs may be an additional source of blood supply to the lungs in less than 5% patients with TOF [28]. The incidence and distribution of MAPCAs were independent of disease severity or morphological variations. The incidence of high percentage in our study was probably due to late presentation and chronic long standing hypoxemia. MAPCAs have significant clinical implications pre and post-operatively and can lead to development of a various complications like massive hemoptysis because of erosion of bronchial vessels due to gross enlargement development of very fatal pulmonary edema postoperatively if not embolized [29]. Similarly, if left untreated it will lead to 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 sequale. If remain undetected can lead to pulmonary edema after operation and difficulty in weaning off the patient thus prolonging the ICU stay [30].

Therefore, we still believed to do imaging like CT angiography or cine-angiography in our patients with tetralogy of fallot prior to corrective surgery to save the post operation complication and mortality.

CONCLUSION

The frequency of MAPCAs in our population even with adequate size branch pulmonary arteries was still high. These frequency size and distribution of MAPCAS were independent of age, surface area, and degree of cyanosis or underlying pulmonary vasculature anatomy. Therefore, diagnostic cardiac catheterization is still a relevant invasive procedure in children with TOF.

LIMITATIONS

We did not study the clinical peri-operative outcome of these children. In view of the high frequency of MAPCAs in even children with "optimal" morphology, a comprehensive prospective

study is required to pinpoint and further elaborate the significance of such collaterals.

ETHICS APPROVAL AND CONSENT TO PARTICIPATE

This complies with national guidelines. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standard. Ethical approval was obtained from the Ethics and Research committee of the University of Nigeria Teaching hospital Enugu (IRB number of 00002323).

CONFLICT OF INTEREST

None.

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