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Role of MDCT in Assessment of Pulmonary Artery Circulation in Tetralogy of Fallot Patients

Samar Mohammad Shehata[1], El Sayed H. Zidan[1], Besheer Abdallah Hassan[2], TarekMostafa Mohy[1]

[1]Radio-diagnosis Department, Faculty of Medicine, Zagazig University, Egypt.

[2]Pediatrics Department, Faculty of Medicine, Zagazig University, Egypt.

Corresponding author:**TarekMostafaMohy**, Email: Tareq_mohi2@yahoo.com

ABSTRACT

Background: Tetralogy of Fallot(TOF) is a congenital heart defect.Multi-detector computed tomography (MDCT has a pivotal role in the evaluation of complex anatomical findings in TOF patients.**Purpose**: To assess the role of MDCT angiography in preoperative evaluation of the pulmonary arterial circulation in TOFpatients.**Patients and methods**: thirty patients (17 males and 13 females) ranged in age from 3 to 74 months, diagnosed with TOF by clinical and echocardiographic examination underwent semi-prospective ECG-gated MDCT cardiac angiography using 128 detectors (Philips Healthcare, Ingenuity) scanner.**Results:** Pulmonary arterial anomalies were detected in 50 % of cases .MPA was atretic in 13.3%, RPA was hypoplastic in 13.3% of cases and LPA was hypoplastic in 10 % of cases. Aortopulmonary anastomosis was identified in 66 % of cases (PDA in 60% of cases, MAPCAs in 20%).The mean of McGoon's ratio was 2.44±1.23.**Conclusion:** MDCT angiography is a powerful and an essential tool in the preoperative examination of pulmonary circulation in TOF patients because it can demonstrate detailed anotomy of the pulmonary arteries and the aortopulmonary connections, which are important details to know before surgery.

Keywords: Tetralogy of Fallot, MDCT, Pulmonary Atresia.

INTRODUCTION

The congenital heart disease (CHD) incidence varies between studies, ranging from 4/1000 to 50/1000 live newborns. Tetralogy of Fallot is the most prevalent type of cyanotic CHD, accounting for about 10% of all CHD cases with an incidence of 421 cases per million live births **(1)**.Tetralogy of Fallot is a congenital heart defect that includes a ventricular septal defect, obstruction of the right ventricular outflow tract, aortic root override, and right ventricular hypertrophy. Several associated extracardiac vascular anomalies have been described especially those related to pulmonary arteries **(2)**.

The main aim of the surgical intervention is to maintain secure blood supply to the lungs either by palliative surgery by inserting a prosthetic tube between a systemic and pulmonary arteries or corrective surgery to relief the subpulmonic obstruction, so detailed characterization of the anatomy of the pulmonary circulation

ISSN: 0975-3583, 0976-2833 VOL 12, ISSUE 06, 2021

is essential for appropriate surgical intervention planning **(3,4)**. In cases of TOF with pulmonary atresia, blood flow to the pulmonary arteries comes from the consistently patent arterial duct or multiple aorto-pulmonary collateral arteries **(5,6)**.

Conventional cardiac angiography could assess both anatomical and hemodynamic data of pulmonary circulation in a single session with a high spatial resolution. Its main drawbacks are that it is an invasive technique and carries a 1% procedure-related mortality risk in neonates, a relatively large ionizing radiation doses and the requirement for anesthesia **(7)**.

The mainstay of TOF diagnosis is echocardiography. Its advantages include the lack of radiation, the ability to evaluate intracardiac structure and function, and the ability to perform hemodynamic assessments. However, it is an operator-dependent study, limited by a small acoustic window, and restricted in the examination of certain parts of the aorta, the distal pulmonary arteries, the pulmonary veins, and the small aortopulmonary collaterals **(8)**.

Magnetic resonance imaging (MRI) yields high-resolution images and is especially beneficial for determining the anatomy of the pulmonary arteries **(9)**. However, its long examination period necessitates a long term of anaesthesia, which is undesirable for children, Furthermore, when compared to CT, the spatial resolution is reduced, which means that minor anomalies may be missed. Pacemakers, mechanical prostheses, conduits, and coils are all contraindications for cardiac magnetic resonance (CMR)**(7)**.

Computed tomographic angiography (CTA) is an essential tool for the initial diagnosis of TOF, treatment planning, and post-interventional follow-up. The aorta-pulmonary connections, pulmonary arteries and veins, and the relationship of thoracic vessels to neighboring structures, especially the airways, can be clearly defined **(10)**.Therefore, this study aimed to assess the role of MDCT angiography in preoperative evaluation of the pulmonary arterial circulation in TOF patients.

PATIENTS AND METHODS

The study included patients who were diagnosed with TOF by transthoracic echocardiography (TTE) and referred to the Radiodiagnosis department for preoperative evaluation. During the period from April 2021 to October 2021 upon 30 patients (17 males (56.7%) and 13 females (43.3%). Their ages varied from three months to seventy-four months, with a mean of (21.75 ±30.04 months).The parents' signed an informed consent, and the approval of the medical ethics committee of our institution was obtained.

The exclusion criteria were: renal insufficiency, Poor general condition, Patients who refuse to complete the trial or who were missed at follow-up, and Iodine hypersensitivity.

Patient preparation:

-Beta blocker or calcium channel blocker to control heart rate one day before examination.4-6 hours fasting.Cannulation: Large bore 22G cannula was inserted at cubital fossa.Sedation : intravenous phenobarbital sodium was administered to

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patients under the age of four or who were uncooperativeat dose of (6 mg/kg; maximum dose, 200 mg). Children who respond well to verbal order were not sedated

All cases underwent:

- Perinatal, neonatal ,family and surgical history taking

-physical examination.

- Assessment of prior investigations as Echocardiogarphy , laboratory and operative reports .

- ECG gated MDCT cardiac angiography using a 128 -detectors scanner (Philips Healthcare Ingenuity, Philips Medical System, Best, The Netherlands) as follows:

- A scanogram : from thoracic inlet level to the L1–2 level
- Injection of mixture of non- ionic contrast media (omnipaque 350 mg Iodine/ml) and normal saline (2 ml/kg of contrast mixed with 1 ml/kg of saline) at flow rate of 1-3 ml /s.
- A large ROI was placed covering the four cardiac chambers with trigger threshold of 175 HU.

-Follow up for 15 minutes to be sure that the sedative effect has gone.

-CTA was reviewed by a professional radiologist on a dedicated workstation (Phillips workstation).

-Various image reformatting techniques was used, such as curved planar reformatting, Minimum intensity projection (miMIP), maximum intensity projection (MIP), and volume rendering (VR).

Images interprétation:

-Cardiac structure: Situs (viscer-atrio-bronchial), concordance (AV, VA), septal defect, chambers size, pericardial effusion, great vessel relationship.

- Extra-cardiac structure: Arteries (aorta, pulmonary arteries, coronary arteries, aorto-pulmonary anastomoses), Veins pulmonary veins, systemic veins), extracardiac structure (lung fields, pleura and upper abdomen)

Statistical analysis:

Data collected and analyzed using Microsoft Excel software. Data were then imported into Statistical Package for the Social Sciences (SPSS version 20.0) software for analysis. According to the type of data qualitative represent as number and percentage , quantitative continues group represent by mean ± SD. Differences between quantitative independent multiple by ANOVA or Kruskal Wallis,. P value was set at <0.05 for significant results &<0.001 for high significant result.

RESULTS

In this study upon 30 patients diagnosed with tetralogy of fallot (TOF), pulmonary arterial anomalies were found in 50% of cases. The main pulmonary

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artery (MPA) was normal in 60% of cases, with a mean diameter of 10.18±4.76 mm, while MPA atresia was the most common MPA anomaly in 13% of all cases. The RPA was normal in 73.3 percent of patients, with a mean diameter of 9.85 ±7.11mm. Hypoplasia was the most common RPA anomaly, occurring in 13.3 % of all cases. The LPA was normal in 76.6 percent of cases, with a mean diameter of 9.41±5.05mm, while hypoplasia and stenosis were the most common anomalies in LPA (13 % of all cases each). Total pulmonary arteries (MPA, RPA & LPA) atresia is present in only one case (3.3%) (table 1).(Fig.1-3)

In 66 % of cases, an aortopulmonary anastomosis was identified. PDA (Fig.2-3)was observed in 60% of the cases while major aortopulmonary collateral arteries (MAPCAs) (Fig.1)were detected in 20% of cases (with pulmonary artery atresia/hypoplasia accounting for 66% of MAPCA cases) (table2).

In the cases of TOF with atretic MPA(Fig.3), the pulmonary circulation was PDA dependent in 50% of cases, MAPCAs dependent in 25% and it depended on both PDA and MAPCAs in 25 %of cases (table 3).McGoon ratio was calculated in all cases and its mean value was 2.44±1.23. 3.3 % of cases had a ratio less than 0.8 which is considered insufficient for complete surgical repair.

Table (1): pulmonary arteries anomalies among the studied patients:

pulmonary arterial anomalies	No.	%
Total pulmonary arterial anomalies	15	50
Total pulmonary atresia	1	3.3
Isolated MPA Atrsia with confluent RPA and LPA	1	3.3
Isolated MPA stenosis	2	3.3
MPA,RPA and LPA hypoplasia	2	3.3
MPA atresia with stenotic RPA	1	3.3
MPA hypoplasia with stenotic LPA	1	3.3
MPA hypoplasia with stenotic LPA and RPA	1	3.3
Isolated MPA hypoplasia	1	3.3
MPA and RPA hypoplasia	1	3.3
Isolated RPA hypoplasia	1	3.3
Atretic MPA and RPA with hypoplastic LPA	1	3.3
Isolated LPA stenosis	1	3.3
MPA		
Atretic	4	13.3
Hypoplastic	6	20
Stenotic	2	6.6
Normal	18	60
Mean diameter	10.18±4.76 mm	
RPA		
Atretic	2	6.6
Hypoplastic	4	13.3
Stenotic	2	6.6
Normal	24	73.3
Mean diameter	9.85± 7.11 mm	
LPA		
Atretic	1	3.3
Hypoplastic	3	10
Stenotic	3	10
Normal	24	76.6
Mean diameter	9.41±5.05mm	

ISSN: 0975-3583, 0976-2833 VOL 12, ISSUE 06, 2021

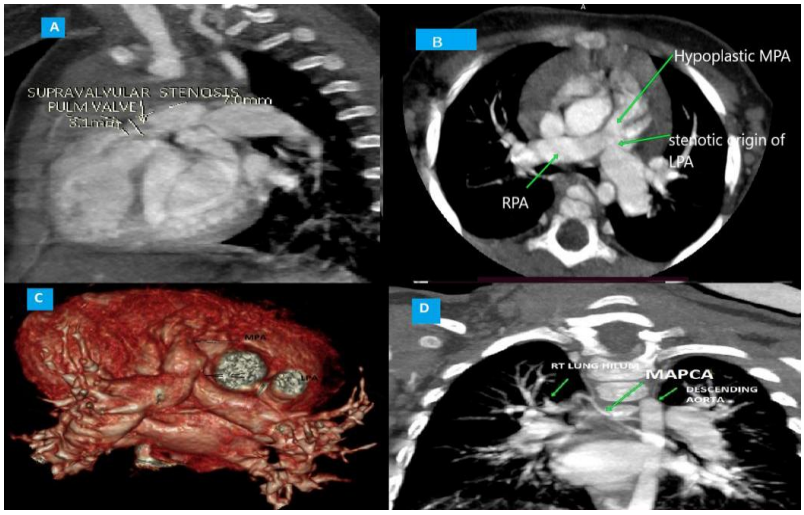

Figure (1):9-month-old boy diagnosed as TOF with hypoplastic MPA and MAPCA. (a)Oblique MIP of RVOT reveals valvular and supravalvular pulmonary stenosis. (b,C) axial MIP and VR image of pulmonary artery shows stenotic origin of LPA with post stenotic dilation and hypoplastic MPA.(d) oblique MIP image reveals MAPCA from descending aorta to RT lung hilum.

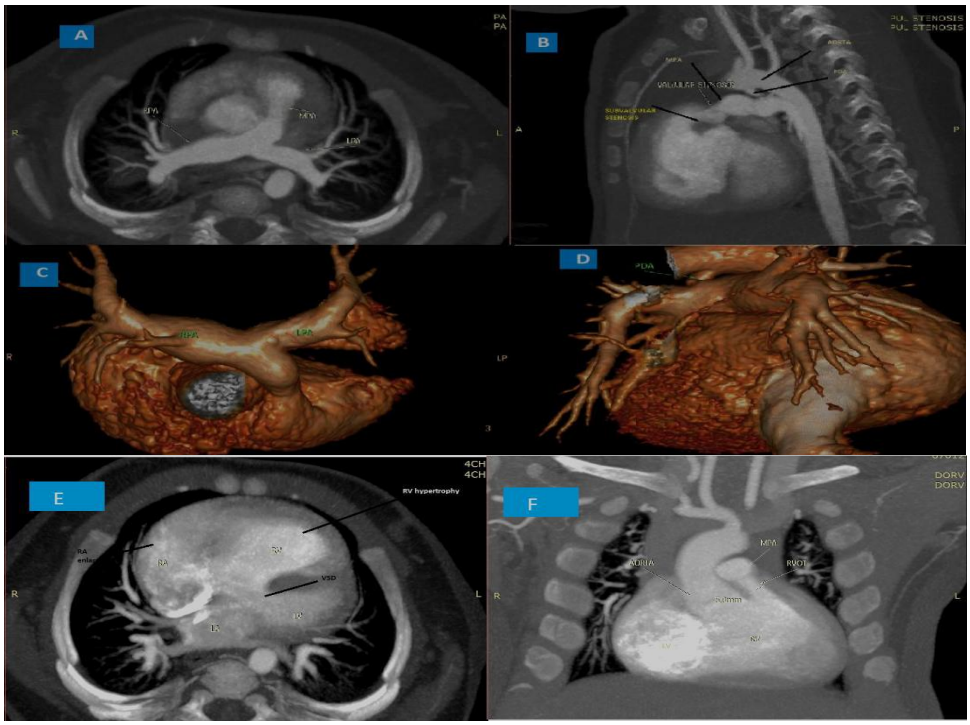

Figure (2): 4-Month-old boy with DORV type TOF and PDA.(a) axial MIP image reveals average caliber of pulmonary arteries. (b) Oblique MIP image shows valvular and subvalvular level RVOT stenosis and PDA from undersurface of descending aorta to LPA. (c) VR image reveals normal caliber of pulmonary arteries with valvular and subvalvularstenosis .(d) VR image of PDA between aorta and LPA. (e) Axial MIP image reveals hypertrophied RV myocardium ,dilated RA and sub

ISSN: 0975-3583, 0976-2833 VOL 12, ISSUE 06, 2021

valvular VSD. (f) Coronal MIP image reveals overrindingaorta(75 % from RT ventricle & 25 % from LT ventricle).

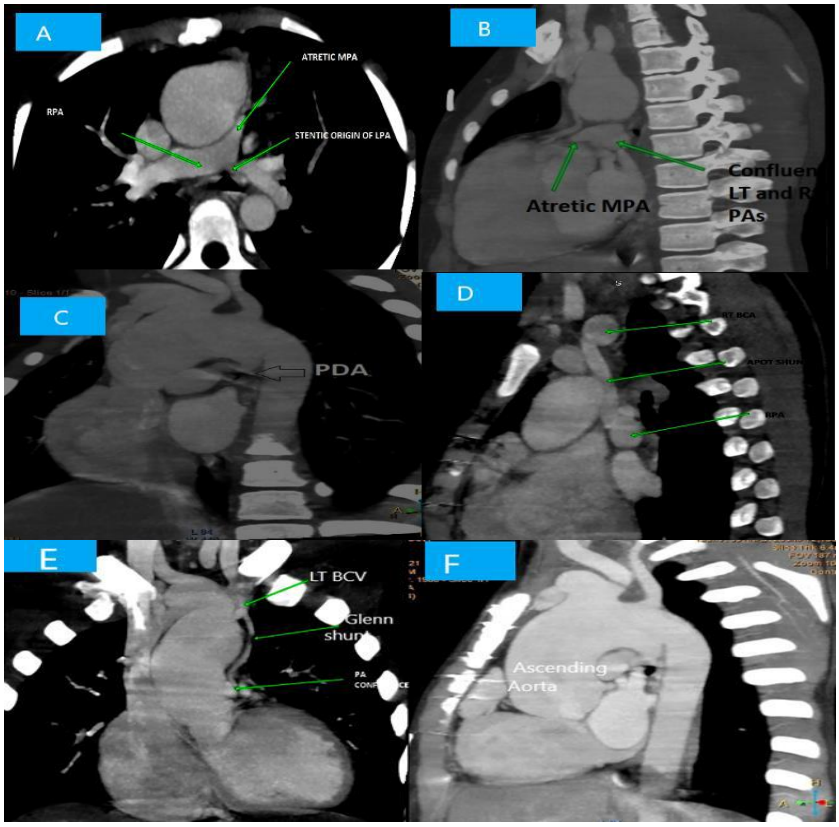

Figure (3): 6 -month -old girl diagnosed as TOF with atretic MPA, PDA, aortic root dilatation , modified Blalock Taussig shunt and Glenn shunt **.** (a) axial, (b) Oblique MIP images reveal atretic MPA with confluent RPA and LPA and stenotic origin of LPA. (c) Oblique MIP image shows S-shaped PDA from the aortic arch to the pulmonary arteries confluence. (d,e) Oblique MIP image reveals modified Blalock Taussig shunt (MBTS) between BCA and RPA and GLENN shunt from the LT. BCV to the confluence of the PA (the patient underwent palliative surgery to secure pulmonary blood flow) .(F) Sagittal MIP image reveals ascending aortic dilatation.

Table (2): Aortopulmonaryconnectionsamong the studied patients:

Aortopulmonary connections	No.	%
PDA	**18**	**60**
MAPCA	**6**	**20**
MAPCA with PDA	4	13.3
MAPCA alone	2	6.6
MAPCA with pulmonary atresia/hypoplasia	4	13.3
MAPCA without pulmonary atresia/hypoplasia	2	6.6

ISSN: 0975-3583, 0976-2833 VOL 12, ISSUE 06, 2021

Table (3): Pulmonary circulation supply in cases of atretic MPA

Pulmonary circulation of atretic MPA	No=4	%
PDA dependent	2	50
MAPCA dependent	1	25
PDA and MAPCA dependent	1	25

DISCUSSION

The overall survival of newborns with TOF has improved over time as diagnostic procedures and patient care for CHDs have improved. According to the most recent statistics, surgical correction of TOF is successful in 98% of infants when done during the first year of life. Prior to TOF repair surgery, diagnostic imaging findings provide critical information such as anatomical data and hemodynamic indicators. Multidetector CT has enhanced its spatial and temporal resolution, as well as its ability to create static and 3D reconstructed images of the heart and main vessels, and it is currently one of the most common imaging modalities used to assess the anatomy of patients with TOF **(11)**.

Tetralogy of Fallot is characterized by abnormalities in pulmonary artery architecture and pulmonary blood flow. As each patient's pulmonary blood supply varies, surgical strategy must be adjusted to their specific needs. The complexity of pulmonary blood flow contributes to the difficulty of managing TOF (extent of MPA atresia, patent ductusarteriosus, native pulmonary arteries, aortopulmonary collaterals and distal pulmonary vascular arborization). Non-confluent central pulmonary arteries result from the extension of MPA atresia to its bifurcation. The presence or absence of confluent PAs has a substantial impact on surgical results. When confluent pulmonary arteries are present at birth, PDA becomes an important source of pulmonary blood flow**(12)**.

Aortopulmonary collaterals (APCs) are found in 30 to 65 percent of patients with pulmonary atresia and typically 2 to 6 in number. The descending thoracic aorta at the level of the carina, subclavian arteries, abdominal aorta, and coronary arteries are all known sources of APCs **(13)**. In newborns with adequate pulmonary blood flow who are eligible for a relatively late definitive repair, the distinction between PDA and APCs is critical. Despite the fact that APCs are susceptible to stenosis over a period of weeks to months, they remain patent longer than PDA until surgical correction is undertaken at a few months of age **(14)**.

In this study, the pulmonary arterial anomalies were found in 50% of TOF cases. The main pulmonary artery was normal in 60 % (mean diameter; 10.18±4.76 mm) and was atretic in 13.3% of cases, hypoplastic in 20% , and stenotic in 6.6% of cases. A much higher incidence was reported by **El shimy et al. (15)**and**Zakaria et al. (16)** found Pulmonary artery defects among (80% & 100% of cases), while the main pulmonary artery was atretic in (30% & 22%) and hypoplastic in (13.3% &43 %).

ISSN: 0975-3583, 0976-2833 VOL 12, ISSUE 06, 2021

We found that the right pulmonary artery was normal in 73.3% (mean diameter: 9.85± 7.11 mm), atretic in 6.6%, hypoplastic in 13.3%, and stenotic in 6.6%.**El shimy et al. (15)**and **Zakaria et al. (16)** reported an atretic right pulmonary artery in (16.7% & 9 %) and hypoplastic RPA in (0% & 38%) of cases.

The left pulmonary artery was atretic in 3.3% of cases, hypoplastic in 10% of cases, stenotic in 10% of cases and normal in 76.6% of our cases (mean diameter; 9.41±5.05mm). In **El shimy et al. (15)**, atretic left pulmonary artery was noticed in (20%) of cases. In **Zakaria et al.(16)**. The LPA was atretic in 9 % of cases and hypoplastic in 58 % of cases.

A lower incidence of pulmonary artery anomalies was reported by multiple previous studies.In**Sheikh et al.(17)** study, Pulmonary artery abnormalities were reported in (18.92%) of cases, MPA was stenotic in 2.8 % of cases and hypoplastic in 1.4 % of cases. The RPA was stenotic in 2.8 %, hypoplastic in 0.4 % and absent in 0.2 % of cases. The LPA was stenotic in 12.3 %, hypoplastic in 1.2 % and absent in 1.4% of cases. **Moustafa et al. (18)** reported complete atresia in 2.6 % of cases .MPA was absent in 5.1%, hypoplastic in 29.8 % and stenotic in 5.1 % .RPA was absent in 3.9% ,hypoplastic in 23.4% and stenotic in 3.9 % of cases .LPA was absent in 2.6 %,hypoplastic in 25.9 % and stenotic in 10.4 % of cases.

Tiwari et al. (19) found pulmonary artery abnormalities among 35 % of their cases. This may be attributed to the different inclusion criteria between studies as **Sheikh et al.(17)**excluded cases withComplete AtrioventricularSeptal Defect with TOF, pulmonary atresia with VSD and operated cases of BlalockTaussig shunt for TOF.

In our study, in cases of main pulmonary artery atresia, the pulmonary circulation was PDA dependent in 50% of cases, MAPCAs dependent in 25% and it depended on both PDA and MAPCAs in 25% of cases. We found aorto-pulmonary connection in 66.6 % of cases in the form of patent ductusarteriosus (PDA) in 60 % of cases, major aortopulmonary collaterals (MAPCAs) in 20 % of case where 66% of MAPCA cases was associated with pulmonary artery atresia/Hypoplasia. **El shimy et al.(15)** found aorto-pulmonary anastmosis among 45% of cases in the form of PDA (23.3%) or MAPCS (23.3%) of patients.**Zakaria et al.(16)**reported aortopulmonary collateral Circulation among 57% of cases. A**Tiwari et al. (19)**foundaortopulmonary collaterals in (37%) and Patent ductusarteriosus in (29%) of cases.

In **Moustafa et al.(18)** study aortopulmonary collaterals was detected in (74%) in which MAPCAs were seen at 28.5% and delicate collaterals were seen at 45.5% and PDA in 35 % of cases.**Hu et al. (20**) detected PDA in 22.64% of cases and aortopulmonary collaterals in 13.8% of cases.

To determine the adequacy of pulmonary artery blood flow before surgery, many pulmonary artery indices were developed, including McGoon ratio.The sum of the diameters of the RPA (at the level of crossing the lateral margin of the vertebral column on angiography) and LPA (just proximal to its upper lobe branch on angiogram) is divided by the diameter of the aorta at the level above the diaphragm

ISSN: 0975-3583, 0976-2833 VOL 12, ISSUE 06, 2021

to get McGoon ratio.In normal people, a range of 2-2.5 was found. In Tetralogy of Fallot, a ratio greater than 1.2 is associated with appropriate postoperative RV systolic pressure. When a ratio is less than 0.8, VSD closure is postponed at the time of repair or they had an aortopulmonary shunt operation as a first step **(21,22)**.

In our study, the McGoon ratio was below the cutoff value of 0.8 in one case (3.3%), while the rest of the cases yielded normal values with mean value 2.44±1.23**.** In the study of **Moustafa et al. (14)**McGoon ratio (mean ± SD) was 1.8 ± 0.81.

CONCLUSION

Using MDCT cardiac angiography is an essential preoperative investigation tool that can clarify accurately the anatomy of the pulmonary arterial circulation and associated anomalies which is crucial for surgical planning of TOF patients.

No Conflict of interest.

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