Egyptian Journal of Radiology and Nuclear Medicine

RESEARCH Open Access

# The role of MDCT in the assessment of cardiac and extra-cardiac vascular defects among Egyptian children with tetralogy of Fallot and its surgical implementation

Ahmed Elshimy*, Rasha Tolba Khattab and Hend Galal Eldeen Mohamed Ali Hassan

## Abstract

**Background:** Tetralogy of Fallot (TOF) is considered the most common form of cyanotic congenital heart diseases (CHD), accounting for about 10% of cases. It includes four main cardiac defects, in addition to various extra-cardiac anomalies.

**Aim:** This study aimed to evaluate cardiac and extra-cardiac vascular defects associated with TOF among Egyptian children, regarding frequency and types with assessment of multi-slice or multi-detector computed tomography (MDCT) role in their diagnosis. Definitely, full detection of these vascular anomalies has utmost importance when evaluating such patients particularly before surgical intervention.

**Methods:** This study included 60 pediatric patients diagnosed as TOF, who underwent MDCT examination in our institute during period of 6 months from (March to September 2020), to confirm their trans-thoracic echocardiography (TTE) findings and detect other vascular abnormalities which cannot be precisely detected with TTE before their surgical interventions.

**Results:** The incidence of different extra-cardiac vascular defects diagnosed by MDCT among our patients was 85% which was significantly higher than that detected by TTE (55%). Moreover, MDCT was superior to TTE assessment as regards its diagnostic accuracy (96.6% vs. 80%), sensitivity (98% vs. 76.9%), and specificity (88.9% vs. 85.7%), in addition to both positive and negative predictive values. The most common anomalies detected were affecting the pulmonary artery (80%), followed by aorto-pulmonary vessels (45%), then aortic artery (40%), coronary arteries (20%), and lastly vena cava connection (6.7%). Patients' demographic characteristics and clinical presentations were also presented.

**Conclusion:** This study confirmed that many extra-cardiac vascular defects are commonly associating cardiac lesions in TOF and emphasizing the great value of MDCT in their diagnosis. Certainly, proper detection of these anomalies will help decision-making during preoperative evaluation, corrective interventions, and further management of these cases.

**Keywords:** Tetralogy of Fallot, Extra-cardiac vascular defects, MDCT

* Correspondence: ahmedchimi@hotmail.com; ahmedchimi@med.asu.edu.eg
Radiology Department, Faculty of Medicine, Ain Shams University, Cairo, Egypt

## Background

Although marked advances in both palliative and corrective cardiac surgery have been developed in recent years, congenital heart diseases (CHD) remain the leading death causes in children with different malformations [1]. These diseases include various presentations, as tetralogy of Fallot (TOF), which has prevalence rate of 3.5–9% [2]. As surgical repair considered the main curative management for these cases and it may be required as early as 3 to 6 months of age, so diagnosis of full anatomical structure of these conditions is mandatory before proceeding to operative management [3, 4].

In addition to the four diagnostic criteria of TOF that include ventricular septal defect, right ventricular hypertrophy with outflow tract obstruction, and overriding aorta, other extra-cardiac vascular anomalies are frequently coexist [5]. These may involve pulmonary, aortic, and coronary arteries with or without valve abnormalities, aorto-pulmonary vessels, and vena cava connections [6, 7].

Trans-thoracic echocardiography (TTE) is the initial classical procedure done for these patients, as it can demonstrate heart structure and estimate intra-cardiac pressures, gradients, and contractile functions as well as blood flow direction across any heart defect and emphasizing coronary vessels integrity. However, many extra-cardiac vascular defects associated with different heart diseases cannot be detected accurately by TTE; consequently, heart catheterization with its invasive complications was widely used for confirming these abnormalities before surgery [8, 9].

Recently, the evolution of multi-slice or multi-detector computed tomography (MDCT) with its multi-planar information, lower doses of radiation, short scanning time, and easy availability suggested it to be a non-invasive reliable diagnostic method for detection of different malformations in pediatric patients with complex CHD [10, 11].

The first surgical repair for TOF was started in 1954, and since that time, treatment strategies have continuously improved, resulting in excellent long-term survival rate (the range is 68.5 to 90.5% for 30-year survival) [12]. In fact, anatomical variations occurring in TOF, as position of cardiac chambers, either atria or ventricles with its relation to viscera and arrangement of great vessels, coronary arteries, and pulmonary and systemic venous connections can affect its management plan, so it must be determined quickly after birth, whether the case will need surgical repair, palliation, or catheter intervention [13, 14].

Accordingly, this study aimed to evaluate cardiac and extra-cardiac vascular defects among Egyptian children with TOF, regarding its frequency, types, and assessment of MDCT role in their diagnosis. Of course, proper detection of these defects will be of utmost importance in full evaluation of these children before operative intervention.

## Methods

### Patients

This pilot study was designed as an observational cohort study. A total of 60 patients diagnosed as Fallot tetralogy depending on their symptoms, signs, and echocardiographic assessment in the Pediatric and Cardio-surgery Departments of our institute from October 2019 till September 2020 were included in the study. They were then referred by their treating physicians to the Radiology department, for MDCT before surgical interventions during 6 months period from March to September 2020. Unstable or critically ill patients and those with history of renal impairment, allergy, or hypersensitivity to contrast material were excluded.

Parents were informed about study design, and as all patients included in this study were less than 16 years old, written informed consent for their participation was obtained by their parent or legal guardian. Research protocol was approved by Radiology Department Scientific Board as well as fulfilling the ethical guidelines of institute.

### Clinical and laboratory evaluation

Revision of history and clinical assessment data of all patients was done including their ages, sex, body weights, and heights, with calculation of body mass index (BMI). Also, their cardiologic symptoms and signs as heart rate, blood pressure, heart murmurs, cyanosis, hypoxic spells, and squatting history were documented. In addition, laboratory data revision, whatever available, especially hematologic data and renal functions was also done. Lastly, their previous echocardiography study findings were revised and documented.

### Radiologic interventions

All patients were examined by 128-slice MDCT scanner (GE Optima CT 660 SE 128, Germany), in the supine position after fasting for about 4–6 h before the study. Chloral hydrate sedation (by oral administration of a dose of 50–100 mg/kg; maximum dose, 2000 mg) for children less than 6 years was done before procedure. The scans were taken from thoracic inlet (the root of the neck including the proximal aspect of common carotid and subclavian arteries) to 2 cm below diaphragm (at the level of portal vein inferiorly) in a cranio-caudal direction.

Retrospective ECG gating utilizing low dose protocol was performed in all cases complying with ALARA (As Low As Reasonably Achievable) principle, in the form of reduction of Kvp and automatic adjustment of tube current according to body weight.

A contrast agent (nonionic) was injected into their ante-cubital veins (omnipaque 350 mg iodine/ml (Iohexol, GE health care Ireland, Cork, Ireland)), in a dose of 1.5–3 mL/kg followed by saline chase 15–20 ml, injection rate 1.5–3 ml/s injected by dual mechanical power injector.

The region of interest (ROI) was placed at descending aorta at main pulmonary artery level with trigger threshold set at 150 HU. Repetitive low dose monitoring examinations (100 kV, 50 mAs, 0.5-s scanning time) were performed 10 s after contrast medium injection began. When trigger threshold was reached, scan started immediately after breath holding command if possible, in older children. In younger children no breath hold was required. The patient is kept under observation for 15–30 min after the procedure till recovery of sedation.

The acquired axial images were reconstructed in sagittal and coronal planes. Furthermore, a variety of high-quality 2D reformatted and 3D reconstructed images were generated that aided in the understanding of complex cardiovascular anatomy. MDCT data were reviewed to obtain all clinically relevant information using a combination of three-dimensional maximum intensity projections (MIP), volume rendering (VR), multi planar reconstruction (MPR), and curved planer reformations (CPR); also, volumetric and functional analyses were done.

Image analysis was performed by two radiologists; one with 5 years of experience and other one with 7 years of MDCT imaging experience in congenital heart disease in an independent way and blinded to the clinical and previous TTE findings. If a non-clear finding or observer variability was present, a revision of this variable by the two radiologists (in a conjoint form) was done until an agreement on the finding between them was reached and no inter-observer variability will be present.

Analysis and confirmation for the 4 main TOF defects and detailed intra-cardiac structure were firstly done and recorded (situs, atrioventricular and ventriculo-atrial concordance, great vessel relationship, defects, size, etc.). Then, all extra-cardiac vascular defects associated with the condition (including aorta, pulmonary arteries, coronary arteries, pulmonary venous drainage, systemic venous drainage) were searched for and documented in a blinded approach to their TTE report. Lastly, after complete MDCT reporting, comparing these results to earlier TTE results and to the later postoperative findings was done.

### Statistical analysis

Study results were analyzed by Statistical program of social science (SPSS) version, 24. Quantitative variables were described in the form of range, mean, and standard deviation, while qualitative variables were described as number and percent and compared using chi-square ($\chi^2$) test. The diagnostic parameters, sensitivity, specificity, positive predictive value, negative predictive value, and diagnostic accuracy of MDCT and TTE for detection of the extracardiac anomalies, were also calculated and compared.

## Results

The demographic parameters and clinical data of TOF patients are presented in Table 1. Ages ranged from 1 month to 14 years (mean 2.84 ± 3.77 years) with 60% males and 40% females. Most of patients were underweight and their body mass index mean was 15.27 ± 1.99 (kg/m$^2$) which is also underweight. Their heart rates and blood pressures were within normal ranges. Regarding their clinical presentations, the most common one was cyanosis which was apparent in 73%, followed by clubbing in 53%, squatting position in 47%, and hypoxic spells in 33% of cases. Moreover, cardiac murmurs were heard in 87% of them.

Cardiac defects using MDCT of all TOF patients are shown in Table 2. It should be noted that all cardiac defects seen by TTE were detected by MDCT, except for 3 cases with right ventricular hypertrophy that showed normal right ventricle in MDCT.

In addition to the 4 main criteria of Fallot tetralogy, other associated extra-cardiac vascular defects are demonstrated in Table 3. It is apparent that by using MDCT,

**Table 1** Demographic parameters and clinical data of the patients

| Parameter | Mean ± SD (range)<br>*n* (%) |
|---|---|
| **Age** (years) | 2.84 ± 3.77 (1 month to 14 years) |
| **Sex** | |
| Male | 36 (60%) |
| Female | 24 (40%) |
| **Weight** (kg) | 10.78 ± 7.26 (3–32) |
| **Height**(cm) | 80.37 25.78 (50–146) |
| **BMI** (kg/m$^2$) | 15.27 ± 1.99 (12–22.2) |
| **HR** (beat/min) | 91.18 ± 7.69 (78–110) |
| **SBP** (mmHg) | 84.77 ± 16.63 (58–122) |
| **DBP** (mmHg) | 60.52 ± 11.26 (40–84) |
| **PP (**mmHg) | 50.05 ± 7.71 (35–65) |
| **Clinical symptoms** | |
| Cyanosis | 44 (73%) |
| Clubbing | 32 (53%) |
| Squatting | 28 (47%) |
| Hypoxic spells | 20 (33%) |
| Murmur | 52 (87%) |

*BMI* body mass index, *HR* heart rate, *SBP* systolic blood pressure, *DBP* diastolic blood pressure, *PP* pulse pressure

**Table 2** Cardiac defects diagnosed by MDCT in TOF patients

| Site of defect | Type of defect | | Number (%) |
|---|---|---|---|
| Right atrium | Dilated | | 24 (40%) |
| | Normal | | 36 (60%) |
| Right ventricle | Hypertrophy | | 56 (93.3%) |
| | Normal | | 4 (6.7%) |
| Left atrium | Dilated | | 0 (0.0%) |
| | Normal | | 60 (100%) |
| Left ventricle | Hypertrophy | | 2 (3.3%) |
| | Normal | | 58 (96.7%) |
| Pulmonary valve | Valvular stenosis | | 42 (70%) |
| | Supravalvular stenosis | | 4 (6.7%) |
| | Sub valvular stenosis | | 14 (23.3%) |
| Aortic valve | Bicuspid | | 2 (3.3%) |
| | Normal | | 58 (96.7%) |
| Atrial septal defect | Present | | 12 (20%) |
| | No | | 48 (80%) |
| Ventricular septal defect | Site | Subaortic | 30 (50%) |
| | | No | 30 (50%) |
| | Type | Membranous | 40 (66.7%) |
| | | Muscular | 20 (33.3%) |

pulmonary artery anomalies were the most common vascular defects found in TOF (80%), followed by aorto-pulmonary vessels defects (45%), then aortic artery anomalies (40%), coronary artery malformation (20%), and lastly venous connection defects (6.7%). Moreover, it should be mentioned that many cases had more than one extra-cardiac vascular defect at the same time.

The incidence of different extra-cardiac vascular defects diagnosed by MDCT among our 60 patients was 51 (85%) which was significantly higher than that detected by TTE 33 cases (55%); $p$ value < 0.01 (Table 4).

Moreover, when following up the postoperative findings of these patients, MDCT was found to be superior to TTE assessment for detection of these anomalies, as regards its sensitivity (98% vs. 76.9%), specificity (88.9% vs. 85.7%), positive predictive value (98% vs. 90.9%), negative predictive value (88.9% vs. 66.7%), and so its diagnostic accuracy (96.6% vs. 80%) (Table 5)

Only one case of pulmonary venous connection anomalies was not detected by MDCT. On the other hand, 9 cases with extra-cardiac vascular anomalies were not detected by TTE, including cases of venous connection defects, aortic arch anomalies, and coronary artery anomalies especially anomalous origin of right coronary artery from pulmonary artery (ARCA PA). Figures 1 and 2 demonstrate some of our TOF patients associated with extra-cardiac vascular anomalies diagnosed by MDCT examination.

## Discussion

In this study, patients' age ranged from 1 month to 14 years with a mean of 2.84 ± 3.77 years and slightly higher male predominance. Their BMI mean was 15.27 ± 1.99 kg/m$^2$ (underweight), while heart rate and blood pressures were normal. Most of the cases presented with either cyanosis (73%) or cardiac murmur (87%), while other manifestations as clubbing, squatting, and hypoxic spells were noted in 53%, 47%, and 33% of cases.

In the study of Hu et al. [6], the mean age of TOF patients was 2.85 ± 2.30 years (range 5 months to 10 years) with 56% of them males. The mean body mass index was 14.05 ± 6.48 kg/m$^2$, heart rate 122.50 ± 16.73 bpm, systolic blood pressure 91.25 ± 24.84 mmHg, and diastolic pressure 54.15 ± 18.68 mmHg. The common symptoms noticed were heart murmurs (64.23%), followed by cyanosis (24.39%), post-exercise tachypnea (4.07%), and squatting (1.63%) that were comparable to the present study results.

In fact, clinical presentation varies based on severity of right ventricular outflow tract (RVOT) obstruction and amount of pulmonary blood flow. Often, cyanosis is not present at birth but appears later within a few months after birth, except in severe cases of (RVOT) obstruction. According to Loh et al., cyanosis appeared in 73% of cases from 3 months to 1 year of life, hypoxic spells in 45%, and squatting in 50% of cases [15]. The pulse and blood pressure are usually normal [16].

All the cases in the present study were diagnosed by their clinical manifestations and confirmed by echocardiography finding for major four TOF criteria. Searching for other associated vascular anomalies that are frequently present in these cases was done according to standard guidelines, before they were referred for MDCT assessment preoperatively [5, 8].

Although TTE combined with Doppler imaging is the first-line method for diagnosis of complex CHD, however, due to its small acoustic window, low spatial resolution, and operator-dependent nature, it is preferred mainly for diagnosing intra-cardiac anomalies with difficulty in detecting extra-cardiac ones [9, 11]. Also, the invasive nature, catheter problems and high radiation dose of cardiac catheterization deferred it as diagnostic method in many cases, although it is the gold standard for this purpose [17].

On the other hand, magnetic resonance imaging was considered as a promising imaging modality for delineating vascular defects in recent years; however, it has many limitations such as presence of pacemakers, lengthy sedation, and relatively low spatial resolution which limit its use for assessing smaller extra-cardiac vascular anomalies [18].

The development of MDCT, with its excellent image quality, rapid acquisition speed that decreases the need

**Table 3** Vascular defects detected by TTE and MDCT

| Types of vascular defects | Number (%) | |
|---|---|---|
| | TTE | MDCT |
| **Venous connection defects** | **0** | **4 (6.7%)** |
| Double superior vena cava | 0 | 2 (3.3%) |
| Pulmonary venous connection anomalies | 0 | 2 (3.3%) |
| **Pulmonary artery defects** | **36 (60%)** | **48 (80%)** |
| Pulmonary atresia | 18 (30%) | 18 (30%) |
| Hypoplastic main pulmonary artery | 6 (10%) | 8 (13.3%) |
| Abnormal atretic right pulmonary artery | 8 (13.3%) | 10 (16.7%) |
| Abnormal atretic left pulmonary artery | 8 (13.3%) | 12 (20%) |
| **Aortic artery anomalies** | **20 (33.3%)** | **24 (40%)** |
| Right-sided aortic arch | 16 (26.7%) | 16 (26.7%) |
| Double aortic arch | 2 (3.3%) | 2 (3.3%) |
| Hypoplastic aortic arch | 2 (3.3%) | 2 (3.3%) |
| Persistent 5th aortic arch | 0 | 2 (3.3%) |
| Aberrant left subclavian artery | 0 | 2 (3.3%) |
| **Aorto-pulmonary vessels defects** | **14 (23.3%)** | **27 (45%)** |
| Patent ductus arteriosus | 12 (20%) | 14 (23.3%) |
| Major aorto-pulmonary collateral vessels **(MAPCS)** | 2 (3.3%) | 14 (23.3%) |
| **Coronary arteries malformations** | **4 (6.7%)** | **12 (20%)** |
| Coronary artery venous fistula (AVF) | 1 (1.7%) | 2 (3.3%) |
| Anomalous origin right coronary artery from pulmonary artery **(ARCAPA)** | 0 | 2 (3.3%) |
| Anomalous origin right coronary artery from left main coronary artery | 1 (1.7%) | 2 (3.3%) |
| Left anterior descending artery dual coarse | 1 (1.7%) | 3 (5%) |
| Anomalous origin left coronary artery from pulmonary artery **(ALCAPA)** | 1 (1.7%) | 3 (5%) |

*TTE* trans-thoracic echocardiography, *MDCT* multi-detector computed tomography

for sedation or anesthesia, lower radiation doses, high-resolution, and so excellent visualization of anatomic structures, made it a rapid reliable non-invasive tool for detecting various anomalies in children with complex CHD, even critically ill patients [9, 19]. It may be superior to echocardiography in detecting extra-cardiac anomalies as those of coronary artery, pulmonary artery branches, and multiple aorto-pulmonary collaterals (MAPCVs) [5, 13, 20], which is going with these study findings.

It was interesting to find that many vascular anomalies were newly detected by using MDCT other than those detected before during echocardiography examination.

**Table 4** Comparison of number of TOF patients with extra-cardiac vascular anomalies detected by TTE and MDCT

| Vascular anomalies | Method of detection | | $\chi^2$ test | *P* Significance |
|---|---|---|---|---|
| | TTE | MDCT | | |
| **Present** | 33 (55%) | 51 (85%) | 12.857 | < 0.01* Highly significant |
| **Absent** | 27 (45%) | 9 (15%) | | |

*TTE* trans-thoracic echocardiography, *MDCT* multi-detector computed tomography

By using MDCT, 85% of our cases showed associated extra-cardiac anomalies compared to only 55% detected by TTE and this difference was statistically highly significant. Moreover, when following up the postoperative findings of these patients, MDCT was found to be superior to TTE assessment for detection of these anomalies, as regards its sensitivity (98% vs. 76.9%), specificity (88.9% vs. 85.7%), positive predictive value (98% vs. 90.9%), negative predictive value (88.9% vs. 66.7%), and so its diagnostic accuracy (96.6% vs. 80%).

These are going with the findings of Hu et al.'s [6] study who compared MDCT to TTE and found that MDCT had greater value in detecting associated extra-cardiac vascular anomalies in TOF patients {diagnostic accuracy 99.13% vs. 97.39%; sensitivity 92.45% vs. 77.07%; specificity 99.81% vs. 99.42%; positive predictive value 98.00% vs. 93.08%; and negative predictive value 99.24% vs. 97.74%}. Also, Bu et al. [21], after finding similar results, mentioned that MDCT not only had overall higher sensitivity than TTE (97.2% vs. 90.6%; $P < 0.05$), but also was much more sensitive for the diagnosis of extra-cardiac vascular abnormalities (92.0% vs. 68.0%; $P < 0.05$).

**Table 5** Comparison of diagnostic parameters of TTE and MDCT in detection of extra-cardiac vascular anomalies in TOF patients

| Diagnostic parameter | Method of detection | | *Z* value | *P* Significance |
|---|---|---|---|---|
| | TTE | MDCT | | |
| **Sensitivity** | 76.9% | 98% | 3.489 | < 0.01* (HS) |
| **Specificity** | 85.7% | 88.9% | 0.526 | > 0.05 (NS) |
| **Positive predictive value** | 90.9% | 98% | 1.700 | > 0.05 (NS) |
| **Negative predictive value** | 66.7% | 88.9% | 2.926 | < 0.01* (HS) |
| **Diagnostic accuracy** | 80% | 96.6% | 2.896 | < 0.01* (HS) |

*TTE* trans-thoracic echocardiography, *MDCT* multi-detector computed tomography, *HS* highly significant, *NS* non-significant

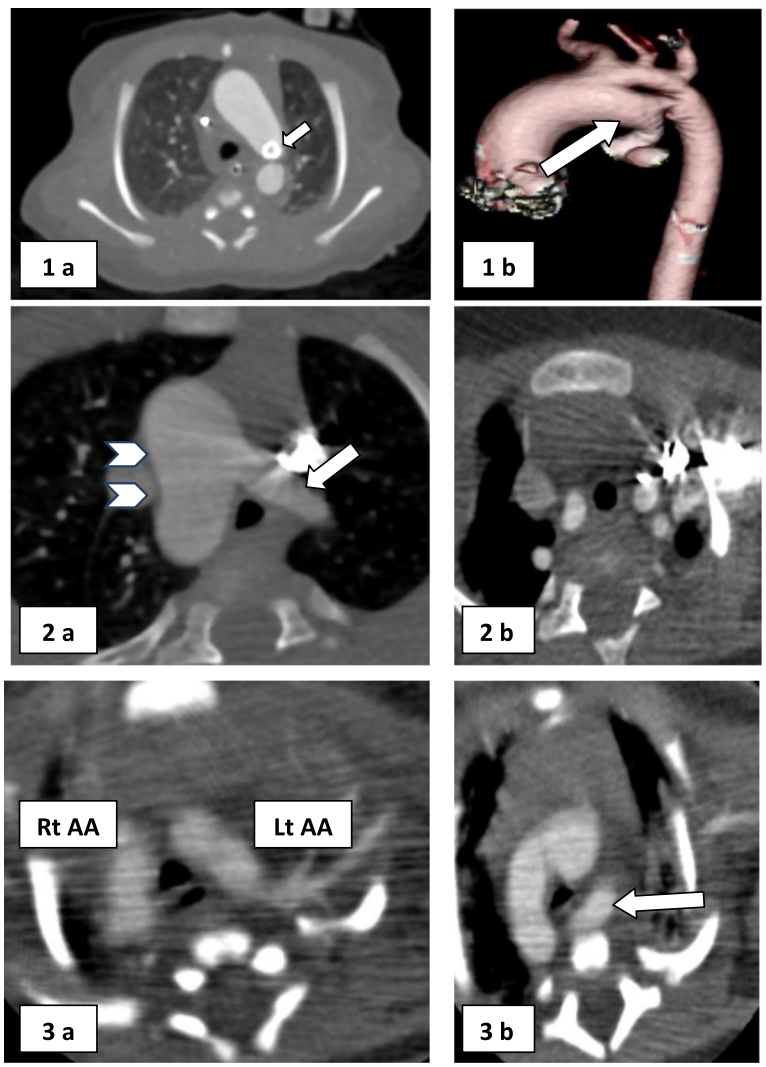


**Fig. 1** MDCT chest with contrast with thin axial cuts of 3 different cases of AA abnormalities (**1a**, **1b** 3D reformate). A 1-month old male neonate with patent PDA stent (short arrow) shows split AA with (persistent embryological 5th arch (long arrow). **2a**, **2b** A 2-year-old female child with double AA with dominant Rt arch (double arrowhead) and hypoplastic Lt arch (short arrow) with no vascular ring. **3a**, **3b** A 3-month-old male infant with double AA (co-dominant) with complete vascular ring (long arrow)

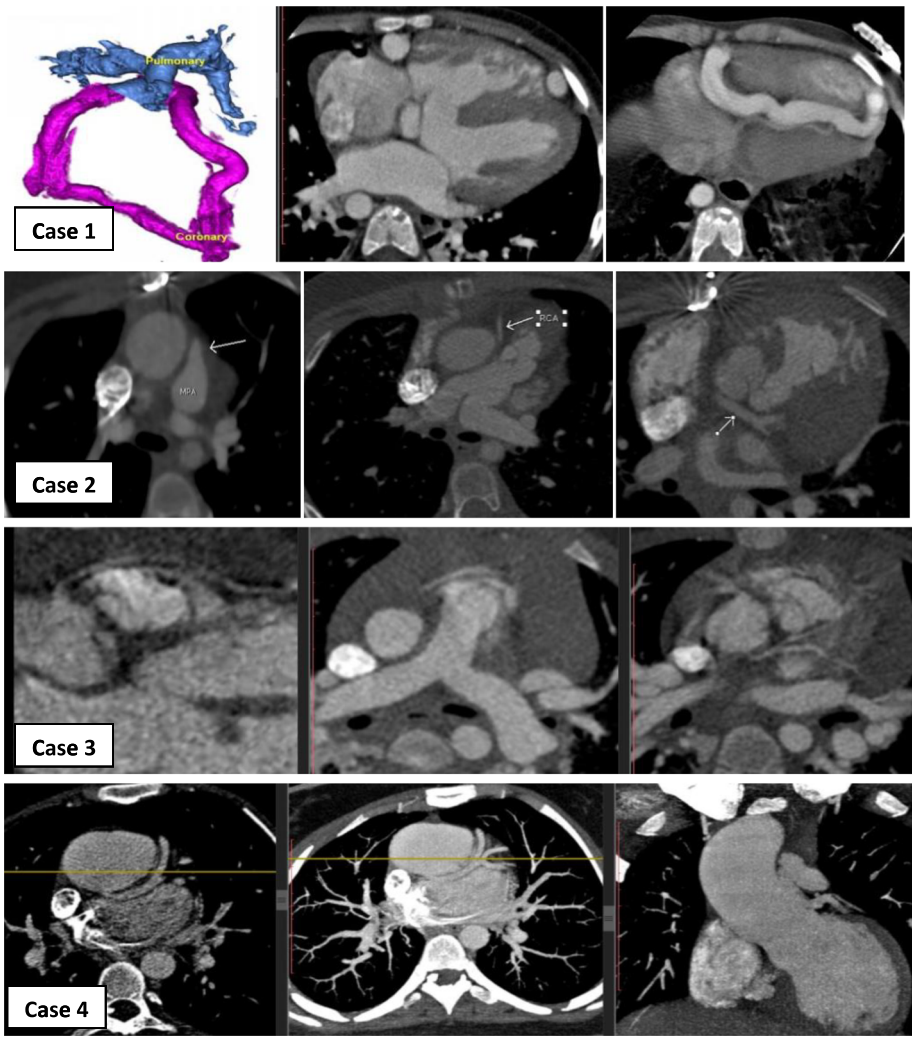


**Fig. 2** MDCT chest with contrast thin cuts of 4 cases of abnormal coronary arteries. (Case 1) A 5-year-old female child with dilated tortuous coronaries and fistulas connection between distal LAD and RCA which showed anomalous origin from pulmonary artery as well. (Case 2) A 8-year-old female child with anomalous origin of RCA from MPA (ARCAPA) with anomalous LM origin from non-coronary cusp. (Case 3) A 1.5-year-old male child with dual LAD origin and pre-pulmonic course of abnormal origin from RCA. (Case 4) A 6-month-old female infant with abnormal origin of RCA from 6 O'clock long left main artery with proximal pre-aortic course

Due to the small field of view during echocardiography examination from the supra-sternal direction in addition to the short neck of pediatric patients, the overlying bone, and aerated lung, all these factors may influence the diagnostic value of TTE compared to MDCT in identifying accurately the great vessels and extra-cardiac anomalies [6, 9].

Moreover, Nie et al. [9], in addition to finding significant differences in the diagnostic accuracy and sensitivity of high pitch MDCT angiography and TTE for detecting congenital extra-cardiac vascular anomalies ($P < 0.05$), mentioned that MDCT offers more information about the relations of these thoracic vascular anomalies to the airways and lung parenchyma, resulting in greater anatomic details, that help the surgeon during correction procedures.

Also, Abd El-Rahman et al. [22] found that the overall sensitivity of MSCT angiography in diagnosis of extra-cardiac vascular anomalies was 98.1% which was higher than that of TTE (80%) and they concluded that it provides important complementary information to TTE with regard to extra-cardiac vascular structures and coronary artery anatomy.

More recently, Alsalihi et al. [23] confirmed these findings and they concluded that MDCT can provide confident detection and exclusion of extra-cardiac vascular abnormalities with superb anatomical description (sensitivity 98.41%, specificity 99.76%, PPV 96.88%, and NPV 99.88%).

It is worthy to mention that the most common anomalies detected in our patients by MDCT were those of

pulmonary artery (80%), followed by aorto-pulmonary vessels (45%), then aortic artery (40%), coronary arteries (20%), and lastly vena cava connection anomalies (6.7%).

Patients with pulmonary artery (PA) defects included 18 patients with main PA atresia, 8 with hypoplastic main PA, 10 with abnormal atretic right PA, and 12 with abnormal atretic left PA. Patients with aorto-pulmonary vessels defects included 14 patients with patent ductus arteriosus and another 14 with major aorto-pulmonary collateral vessels (MAPCS). On the other hand, those with aortic artery anomalies included 16 patients with right-sided aortic arch (AA), and 2 patients with each of double AA, hypoplastic AA, persistent 5th AA and aberrant left subclavian artery. As regards coronary arteries (CA) malformations, there were 2 patients with each of coronary AVF, anomalous origin of right CA from PA (ARCAPA), and anomalous origin of right CA from left main CA, in addition to 3 patients with each of left anterior descending artery dual coarse and anomalous origin of left CA from PA (ALCAPA). Lastly venous connection anomalies included 2 patients with double superior vena cava and another 2 with pulmonary venous connection anomalies.

Also, Zakaria et al., by using multi-detector CT for diagnosis of TOF associations, found pulmonary artery abnormalities in 100% of their cases, either atresia or stenosis of the main artery or its right or left branches, infundibular pulmonary artery stenosis in 43%, deformities of the MAPCVs in 57%, and right aortic arch in 30%. In addition to that, 4% of their cases had abnormal coronary arteries and 44% had patent ductus arteriosus [7].

Recently, Chelliah et al. [13] and others, when comparing MDCT to TTE, found that it was of greater value in the visualization of anomalies of the pulmonary artery [11, 21] and valve, as well as deformities of the MAPCVs [12], abnormal vena cava connections, and aortic artery and valve disorders [24]. They mentioned that TTE can only identify relatively large ones, while MDCT can visualize the number, origin, and supplied lung lobes of MAPCVs, regardless of the size of the vessels [25]. Moreover, MDCT was found to have both 100% sensitivity and specificity for the detection of coronary artery anomalies in these patients [10].

It should be mentioned that MDCT highlights the importance of specific critical extra-cardiac vascular anomalies with its implication to management plane as in the following categories:

1 Finding needs immediate intervention as anomalous right coronary artery origin from pulmonary artery (ARCAPA) especially if associated with severe degree of MPA stenosis, coronary arteriovenous fistula (AVF).
2 Findings important to be clarified before the time of operation such as size and numbers of major aorto-pulmonary collateral vessels (MAPCVs), persistent left sided SVC, right sided AA, pre-pulmonic course of dual LAD, and total anomalous pulmonary venous return (TAPVR).
3 Findings of implication in surgical access or dating like double AA, and partial anomalous pulmonary venous return (PAPVR).

Lastly, although MDCT provide accurate, non-invasive, rapid, diagnostic imaging of the extra-cardiac vascular structures, however, many limitations for its use are present as the need for sedation of children especially those below 6 years of age in addition to exposing these young developing individuals to higher doses of ionizing radiations and to iodinated contrast material. In this study, we use chloral hydrate for sedating these children and non-iodinated contrast materials to avoid side effects of these drugs as much as it could be however MDCT is indispensable prior to operative correction as the most accurate imaging technique for this purpose.

The study used a relatively small sample size; for better analysis and results, it would have been presented with a larger sample size. The imaging technique used was a retrospective ECG-gated one which still provides more irradiation than its prospective counterpart, with however using low dose according to the ALARA dose lowering protocol. In future studies, it might be warranted using prospective ECG-gated technique with low dose radiation adaptive statistical iterative reconstruction (ASiR) technique to benefit from high image quality with dose reduction.

## Conclusion

In conclusion, we emphasize that vascular defects especially extra-cardiac one are commonly associated with cardiac lesions in TOF patients and MDCT has proved a key role in diagnosis of these defects complementing the echocardiography assessment. Certainly, proper detection of these anomalies will help decision-making during preoperative evaluation, surgical intervention, and further management of these patients.

### Abbreviations

AA: Aortic arch; ALCAPA: Anomalous origin left coronary artery from pulmonary artery; ARCAPA: Anomalous origin right coronary artery from pulmonary artery; AVF: Artery venous fistula; BMI: Body mass index; CA: Coronary arteries; CHD: Congenital heart diseases; HS: Highly significant; MAPCS: Major aorto-pulmonary collateral vessels; MDCT: Multi-detector computed tomography; NS: Non-significant; PA: Pulmonary artery; TOF: Tetralogy of Fallot; TTE: Trans-thoracic echocardiography

### Acknowledgements

The authors thank all the study participants for their patience and support.

### Authors' contributions

A.S (corresponding author). Research study conception. Participation in practical part. Paper editing and revision. Interpretation of data and statistical preparation. References collection. Approval of the submitted version. Agreed to be personally accountable for the author's own contributions. Read and approved the manuscript. R.K: Revision of the work and paper editing. Read and approved the manuscript. H.G.H: Research study work design. Main researcher in practical part. Interpretation of data. Revision of the work and paper editing. References collection. Approval of the submitted version. Agreed to be personally accountable for the author's own contributions. Read and approved the manuscript.

### Funding

This study had no funding from any resource.

### Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

### Ethics approval and consent to participate

This study was approved by the Research Ethics Committee of the Faculty of Medicine, Ain Shams University, in Egypt on 16/03/2020; Reference Number of approval: FWA000017585. FMASU R18/2020.
A written informed consent was obtained by the parent or legal guardian of each patient.

### Consent for publication

All patients included in this study are less than 16 years old so; written informed consent for their participation was obtained by their parent or legal guardian.

### Competing interests

We have no competing interests to declare.

Received: 1 October 2020 Accepted: 25 January 2021
Published online: 08 February 2021

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