Role of Computed Tomography for the evaluation of Pulmonary artery sizes in Tetralogy of Fallot with Pulmonary Atresia

Faizah Hakim, Hasham Hafeez Hanjra, Zain ul Abidin, Syed Muhammad Yousaf Farooq, Sani Zahra, Quratulain Muhammad Akbar, Faryal Qazafi, Zareen Taj

Abstract— imaging requirement was to determine the extent and severness of reduction of pulmonary outflow, illustration of related abnormalities and presenting degree of pulmonary atresia or discontinuity in arteries. MDCT gave structural details in this regard. So, role of MDCT was well recognized in preoperative evaluation for surgical policy in atretic patients.

Index Terms— Tetralogy of Fallot, Multi detector Computed Tomography, pulmonary arteries, pulmonary atresia

1 Introduction

AMong cyanotic congenital cardiac disease tetralogy of fallot is most frequently occurring disease. In this condition the children have survived without given medical care beyond the neonatal age, with the need for surgical intervention in the beginning of their life. This considers congenital defects from 7 to 10 percent. In 3 to 5 of every 10,000 live births suffers from this disease [1]. In Pakistan, the true occurrence and incidence of congenital heart disease is unidentified due to limited access to large population studies for medical care and insufficient resources. The incidence of congenital heart disease in all population, where appropriate data is accessible, ranges betweeb 8-10 per 1000 live birth. Even through cyanotic congenital heart disease was recorded for less than 25% of heart defects [2].

The main role of heart is to pump blood throughout the body. When a person is born with some defects in heart. These defects are called congenital heart defects [3]. A significant complication of TOF is the reduction of the outflow of the right ventricle and the variable degree of contraction of the pulmonary artery and its branches [4]. Ventricle septal defect and aortic overriding are also included in it. Pulmonary atresia is a severe heart condition in which there is no proper formation of the pulmonary valve which allows blood from the heart to the lung of patient. Instead of opening and closing of the valve, a solid sheet of tissue develops. Therefore blood cannot flow by its normal path to take oxygen from the lung. Instead, inadequate blood travels to the lung through other natural routes within the heart and its arteries [5], [6]. Echocardiographic detection is a diagnostic modality of intracardiac malformation; however echocardiography is inadequate in the assessment of the peripheral pulmonary arteries due to improper acoustic windows and does not allow surgical operation with an accurate anatomic road map [7]. Imaging requirement is to determine the extent and severeness of reduction of pulmonary outflow and arteries, illustration of related abnormalities and presenting degree of pulmonary atresia or discontinuity. MDCT gave structural details in this regard. Pulmonary atresia is variation of TOF with VSD, due to complete atresia of pulmonary artery. Surgical policy is more complex in patients with large atretic segment. So, role of MDCT is well recognized in preoperative evaluation [8].

Cardiac MRI with breath-hold ECG- gated technique and MR angiography with contrast are also used for the assessment of complicated congenital cardiac defects. However, in closed MR environment prolonged sedation and administrating anesthesia in severely ill patient leads to substantial depression. Angiography by catheter is a procedure with many disadvantage as well as need for anesthesia, arterial puncture and vascular problems are also associated with this procedure [9]. Pre and post assessment of pulmonary atresia become easier with the development of the MDCT [10]. The application of multi slice spiral CT among patients with congenital heart disease has improved the clinical practice of cardiac CT imaging [11], [12]. It has the benefits of rapid scan rate, improved spatial resolution, achieving isotropic volume results and immediate assessment of airways and lung parenchyma, which is responsible for structural pathology in congenital heart disease patients and can non-invasively give detailed cardiopulmonary status in one review[13],[14].

For the surgical procedure of pulmonary atresia, it is therefore important to provide information on pulmonary artery measurement, although echocardiography is a non-invasive cardiac capacity assessment technique but in some cases the measurement of pulmonary arteries is limited.

2 Material and method

This descriptive study was conducted in the Cardiology Department, The Children's Hospital and Institute of Child Health Lahore, Pakistan.
The data were obtained through convenient sampling. The study included children who had TOF with pulmonary atresia. The data were analyzed using Statistical Software for social sciences (SPSS version 25).

3 SCANNING TECHNIQUE

Toshiba, Aquilion ONE, 640 slice ECG gating 320 detector ar-

ray and 3rd generation CT scanner were used for scanning purposes. Ketamine has been used to sedate the patient. The dose of radiation is kept as minimum by sufficiently rising the kilvoltage and tube current. 80 KV and 80 mAs were used for patients weighing less than 10kg, and for 10-19 kg, 80 KV and 100 mAs, and for patients whose weight ranges from 20-30 kg 100 kV and 120 mAs respectively. Certain technical parameters were as follows: 0.24mm of detector collimation; 1.2 pitch scans; 0.5 mm of slice thickness; 0.5 mm of reconstruction interval; and 0.34 seconds of gantry rotation. The contrast medium for a 22-gauge cannula was injected at a rate of 1.5-2.0 mL/s and for a 20-gauge cannula contrast was injected at a rate of 3.0 mL/s with a dual-head power injector. By using a saline chaser the amount of contrast medium required for CT angiography was minimized. It is advised to use 10-20 mL of saline at a rate equal to contrast injection.

4 RESULTS

The size of MPA, LPA and RPA was measured. This study included 51 patients of TOF with pulmonary atresia having 37 males (72.5%) and 14 females (27.5%). Their ages range from 2 months (min) to 14 years (max). Out of total patients 32 (62.7%) have confluent branches while 19 (37.3%) were without confluent branches.

The size of pulmonary arteries was measured. Out of 51 patients 21(41.2%) were suffering from valvular atresia and their mean and standard deviation were 8.77±4.2 while 30 patients were without valvular atresia and their mean and standard deviation were 9.39±3.5. The minimum size of MPA was 3.7mm and maximum size was 25.3mm and their mean and standard deviation were 9.1±3.85.

Out of 51 patients, 14(27.5%) were suffering with RPA atresia. We took the size of RPA at proximal and distal sites. Mean & standard deviation of proximal RPA size was 7.39±2.39 while for distal RPA size was 4.56±3.46. The minimum size of proximal RPA atresia was 2.6mm and maximum size was 12.3mm. Their mean and standard deviation were 6.94±2.1. The minimum size of distal RPA atresia was 0.3mm and maximum size was 12mm. Their mean and standard deviation were 6.1±2.6. While 37(72.5%) patients were without RPA atresia, the mean & standard deviation of proximal and distal sizes were 6.78±2.06.

Similarly 15(29.4%) patients were suffering with LPA atresia. The size of left pulmonary artery at proximal and distal sites was measured. Mean and standard deviation of proximal LPA size was 5.5±2.6 while for distal LPA size was 4.4±3.3. The minimum size of proximal LPA atresia was 2mm and maximum size was 14mm. Their mean and standard deviation were 6.1±2.5. The minimum size of distal LPA atresia was 0.7mm and maximum size was 14mm. Their mean and standard deviation were 5.8±2.8. While 36(70.6%) patients were without LPA atresia, the mean and standard deviation of proximal and distal sizes were 6.39±2.44.

The results showed that multi detector computed tomography can accurately measure the size of MPA, RPA and LPA with atresia.

5 DISCUSSION

Tetralogy of fallot is the most common cyanotic heart condition along with pulmonary atresia, which aggravates the situation in such a way that a solid tissue sheet is developed instead of pulmonary valve, blood cannot pass through its natural path to the lung and different pathways form that carry oxygen-poor blood towards the lungs. Pulmonary artery and its branches may be very small or completely absent and children are always at high risk of death if the disease is not treated, even after treatment, children are vulnerable to multiple heart problems. To resolve this, a proper technique for determining cardiopulmonary status and their correct management is vital, so it is important to provide pulmonary arterial measurement information for the surgical procedure of pulmonary atresia.

Imaging purpose was to determine the extent and nature of pulmonary outflow reducstion, to demonstrate associated anomalies and to present degree of pulmonary atresia or discontinuity in the arteries. In this regard the MDCT gave structural descriptions. MDCT's role in preoperative assessment for surgical policy in atretic patients was thus well known.

The study was conducted in children hospital Lahore. The study included 51 patients of TOF with pulmonary Atresia. Their minimum age was 2 months and maximum age was 14 years having 37 males (72.5%) and 14 females (27.5%). 32(62.7%) patients were presented with confluent branches. 21 (41.2%) patients were suffering from valvular Atresia with minimum size of 3.7mm and maximum size of 25.3mm, and their mean and standard deviation was 9.1±3.8.

14 (27.5%) patients were with RPA Atresia .We also took their sizes at proximal and distal sides. The minimum size of proximal RPA Atresia was 2.6mm and maximum size was 12.3mm and their mean and standard deviation was 6.9±2.1. Similarly minimum size of distal RPA Atresia was 0.3mm and maximum size was 12mm and their mean and standard deviation were 6.1±2.6. Remaining 15 (29.4%) patients were presented with LPA Atresia. The minimum size of proximal LPA Atresia was 2mm and maximum size 14mm and their mean and standard deviation were 6.1±2.5. Similarly minimum size of distal LPA Atresia was 0.7mm and maximum size of distal LPA Atresia was 14mm and their mean and standard deviation were 5.8±2.8.

K Shi, et al conducted a study involving thirty five patients with CCHD who undertook DSCT and TTE to estimate the diameter of the MPA, RPA and LPA and concluded that"DSCT may provide more precise pulmonary artery measurement than TTE in cardiac surgery" [15]

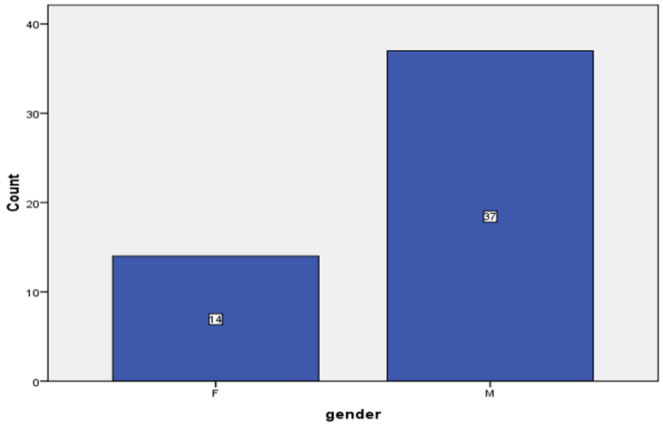
N Chaosuwannakit, et al also concluded that "sensitivity, specificity, positive and negative predictive value of MDCT for pulmonary artery confluence identification , lack of native pulmonary artery and pulmonary artery stenosis were all 100 %"[16]. Similarly a study conducted by Enaba MM, et al in 2017, sixty patients was included in their study. All images included axial, MPR, MIP and VRT and were presented in one

session, concluding that "MDCT has proved to be an important decision making modality in congenital cyanotic heart disease patients" [17].
Similarly, G Chandrasekhar, et al conducted a study in which 105 children were included. For the pulmonary artery evaluation, they go through 64-row MDCT, echocardiography and CCA, including visualization, presence of confluence, stenosis and collaterals. They concluded that "low dose64 row MDCT is strongly correlated with CCA and can provide reliable information on pulmonary arterial anatomy in children with congenital cyanotic heart disease and can eliminate invasive cardiac catheterization angiography with significantly lower radiation exposure to the patient"[18].
Similarly a study conducted by S Murai, et .al in 2004. Two cases of PA and VSD associated with MAPCAs pre and post operatively evaluated using angiography and MDCT using 3D –VR technique was recorded. They concluded that "MDCT had the ability to explain all MAPCAs discovered by invasive angiography with the 3D-VR technique. This method is useful with traditional preoperative angiography as a standard examination for the preparation of staged surgery" [19].
We get that cardiac computed tomography has the benefits of rapid scanning rate, high spatial resolution, achieving isotropic volume data and is responsible for the measurement of pulmonary artery sizes and other structural pathology in congenital heart disease patients.

6 Conclusion

Cardiac computed tomography can accurately measure the diameter of main pulmonary artery, right and left pulmonary arteries including visualization of confluent branches, stenosis and collaterals and is used as standard examination for preoperative planning of pulmonary atresia.

7 FIGURES



(1)

Figure 1: Graphical Presentation of Gender

Figure 1 showed gender description in which 51 patients were included, 14 were females and 37 were males.

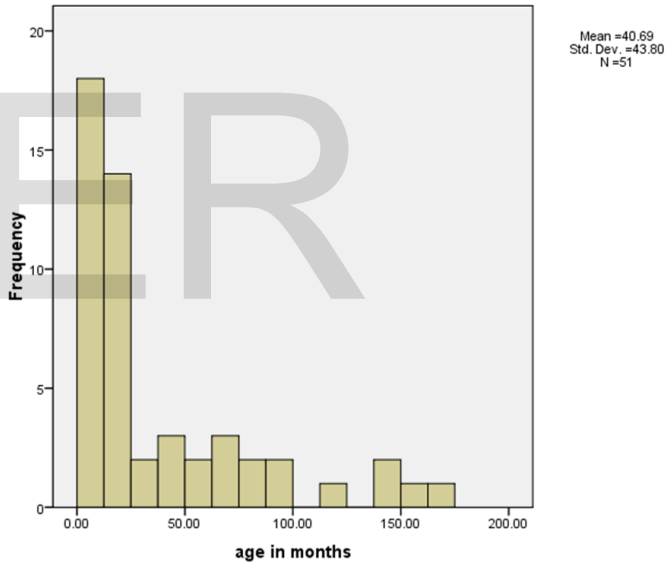


Figure 2: Graphical representation of age group

Figure 2 showed minimum age of 2 months and maximum age of 14 years (168 months) and their mean and standard deviation were 40.68±43.8

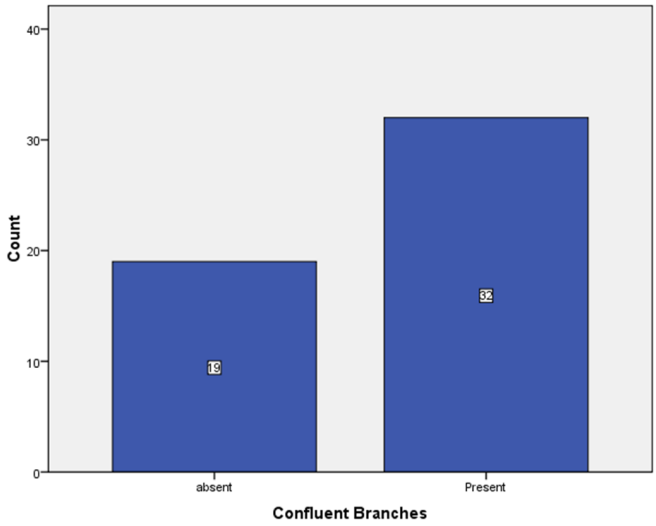


Figure 3: Graphical representation of Confluent Branch

Figure 3 showed that out of 51 patients 32 were presented with confluent branches while 19 were without confluent branches.

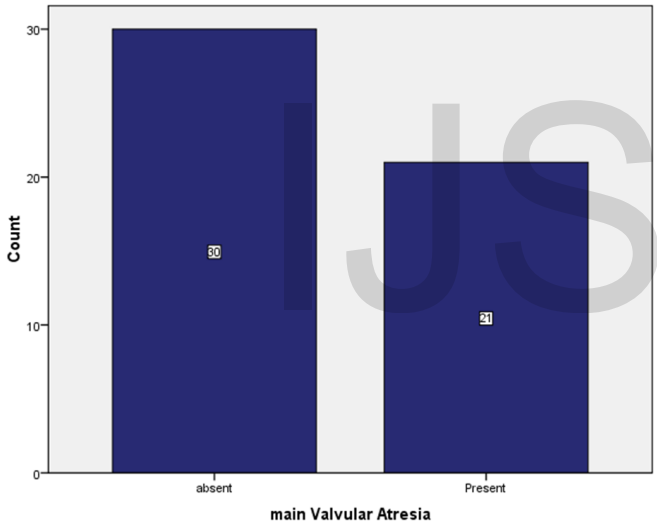


Figure 4: Graphical representation of MPA Atresia.

Figure 4 showed that out of 51 patients 21 were presented with MPA valvular atresia while 30 patients were not.

[1]Main pulmonary artery

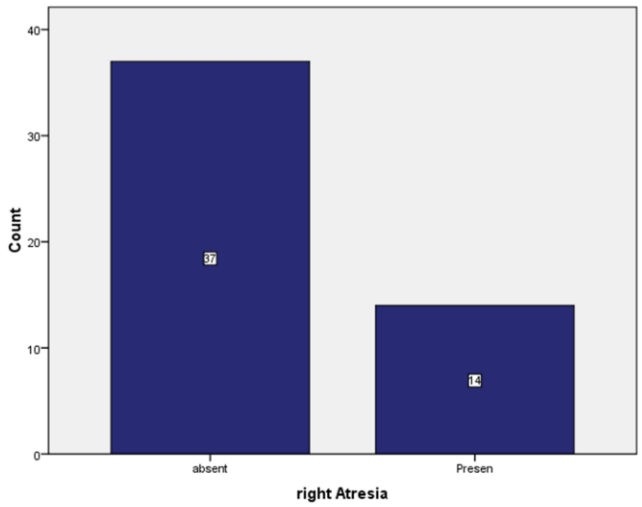


Figure 5: Graphical representation of RPA Atresia

Figure 5 showed that out of 51 patients 14 were presented with RPA atresia while 37 were without RPA atresia.

[2]Right pulmonary artery

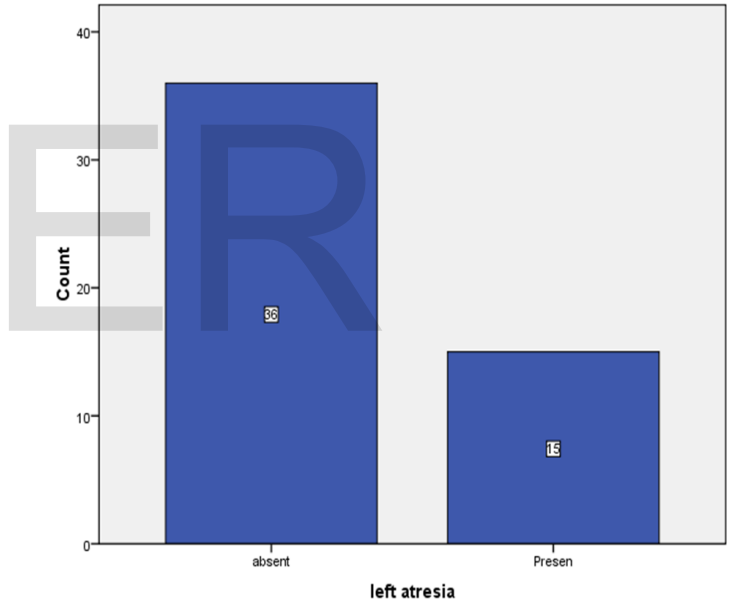


Figure 6: Graphical representation of LPA Atresia

Figure 6 showed out of 51 patients 15 were suffering from LPA atresia while 36 were without LPA atresia.

[3]left pulmonary artery

8 TABLES

Table 1:

Mean &Std. Deviation of MPA Atresia

Valvular Atresia	Mean	N	Std. Deviation
Absent	9.3967	30	3.59036
Present	8.7762	21	4.26789
Total	9.1412	51	3.85456

According to table mean and standard deviation in patients with valvular atresia was 8.7±4.2 and mean and standard deviation in patients without valvular atresia was 9.3± 3.5.

Table 2:

Mean & Std. Deviation of RPA Atresia and its size on proximal and distal sides.

Mean &Std. Deviation of RPA Size

RPA Atresia		RPA Distal Size	RPA Proximal Size
Absent		6.7811	6.7811
		37	37
		2.06866	2.06866
Present		4.5643	7.3929
		14	14
		3.46157	2.39694

Table 3:

Mean & Std. Deviation of LPA Atresia with its Size on its proximal and distal sides:

Mean &Std. Deviation of LPA Size

LPA Atresia	LPA Proximal Size	LPA Distal Size
absent	6.3972	6.3972
	36	36
	2.44137	2.44137
Present	5.5000	4.4467
	15	15
	2.63574	3.30495

Table 4:

Descriptive Statistics of Mean & Std.Deviation of Pulmonary Arteries Sizes

Descriptive Statistics

	N	Minimum	Maximum	Mean	Std. Deviation
right proximal	51	2.60	12.30	6.9490	2.15661
Main size (mm)	51	3.70	25.30	9.1412	3.85456
right distal	51	.30	12.00	6.1725	2.68232
left distal	51	.70	14.00	5.8235	2.83483
left proximal	51	2.00	14.00	6.1333	2.50756

According to table the minimum size of MPA was 3.7mm and maximum size was 25.3mm and their mean and standard deviation was 9.1±3.8.The minimum size of proximal RPA was 2.6mm and maximum size was 12.3mm and their mean and standard deviation was 6.9±2.1.The minimum size of distal RPA was .30mm and maximum size was 12mm and their mean and standard deviation was 6.1±2.6.The minimum size of proximal LPA was 2mm and maximum was 14mm and their mean and standard deviation was 6.1±2.5.while the minimum size of distal LPA was .70mm and maximum was 14mm and their mean and standard deviation was 5.8

Acknowledgement

Alhamdulillah, in the name of Allah, the Most Gracious and the Most Merciful, praises Allah for His blessing and the power to complete this article.

We would like to pay special Appreciation to our dear parents for their continuous encouragement and support.

We would like to thanks Hasham Hafeez Hanjra and Zain ul abidin for their supervision and constant support. Their unceasing help and suggestion throughout the completion of this manuscript made this research a success.

We take this opportunity to thank all faculty members of the department for their help and support.

Last but not least we also thank to our colleagues for their support and help in completing this article.

Funding sources:

This research did not receive any special grant from any public, commercial or non-profit funding agency.

REFERENCES

[1] Khan SM, Drury NE, Stickley J, Barron DJ, Brawn WJ, Jones TJ, et al. Tetralogy of Fallot: morphological variations and implications for surgical repair. Eur J Cardiothorac Surg. 2019;56(1):101-9.

[2] Humayun KN, Atiq M. Clinical profile and outcome of cyanotic congenital heart disease in neonates. Journal of the College of Physicians and Surgeons Pakistan. 2008;18(5):290.

[3] Hsu DT, Pearson GD. Heart failure in children: part I: history, etiology, and pathophysiology. Circ Heart Fail. 2009;2(1):63-70.

[4] Neill CA, Clark EB. Tetralogy of Fallot. The first 300 years. Tex Heart Inst J. 1994;21(4):272.

[5] Hoffman JI, Kaplan S. The incidence of congenital heart disease. J Am Coll Cardiol. 2002;39(12):1890-900.

[6] Bergersen L, Foerster S, Marshall AC, Meadows J. Congenital heart disease: the catheterization manual: Springer Science & Business Media; 2008.

[7] Saric M, Armour AC, Arnaout MS, Chaudhry FA, Grimm RA, Kronzon I, et al. Guidelines for the use of echocardiography in the evaluation of a cardiac source of embolism. J Am Soc Echocardiogr. 2016;29(1):1-42.

[8] Bhat V, BeLaVaL V, Gadabanahalli K, Raj V, Shah S. Illustrated Imaging essay on congenital heart diseases: multimodality approach part I: clinical perspective, anatomy and imaging techniques. Journal of clinical and diagnostic research: JCDR. 2016;10(5):TE01.

[9] Han BK, Rigsby CK, Hlavacek A, Leipsic J, Nicol ED, Siegel MJ, et al. Computed tomography imaging in patients with congenital heart disease part I: rationale and utility. An expert consensus document of the Society of Cardiovascular Computed Tomography (SCCT): endorsed by the Society of Pediatric Radiology (SPR) and the North American Society of Cardiac Imaging (NASCI). J Cardiovasc Comput Tomogr. 2015;9(6):475-92.

[10] Yang S-h, Luo P-h, Tian X-x, Li X-y, Li X-q, Yang Z-j, et al. Prenatal diagnosis of pulmonary atresia with ventricular septal defect. Journal of Medical Ultrasonics. 2018;45(2):341-4.

[11] Hedgire S, Baliyan V, Ghoshhajra B. Advanced Imaging in Adult Congenital Heart Disease. Adult Congenital Heart Disease in Clinical Practice: Springer; 2018. p. 477-510.

[12] Burrill J, Dabbagh Z, Gollub F, Hamady M. Multidetector computed tomographic angiography of the cardiovascular system. Postgrad Med J. 2007;83(985):698-704.

[13] Chen S-J, Huang J-H, Lee W-J, Lin M-T, Chen Y-S, Wang J-K. Diagnosis of Pulmonary Arterial Hypertension in Children by Using Cardiac Computed Tomography. Korean journal of radiology. 2019;20(6):976-84.

[14] Gupta SK, Saxena A, Gulati GS. Evaluation of pulmonary hypertension in a child: role of computed tomography. The Indian Journal of Pediatrics. 2011;78(11):1417-9.

[15] Shi K, Yang Z-g, Xu H-y, Zhao S-x, Liu X, Guo Y-k. Dual-source computed tomography for evaluating pulmonary artery in pediatric patients with cyanotic congenital heart disease: comparison with transthoracic echocardiography. Eur J Radiol. 2016;85(1):187-92.

[16] Razek AAKA, Al-Marsafawy H, Elmansy M. Imaging of Pulmonary Atresia With Ventricular Septal Defect. J Comput Assist Tomogr. 2019;43(6):906-11.

[17] Richardson RR. Coronary Artery Anomalous Origins in Patients with Congenital Heart Disease. Atlas of Pediatric CTA of Coronary Artery Anomalies: Springer; 2020. p. 43-5.

[18] Sarawagi AK, Sodani V, Sodani RK, Verma M. Stratification of Tetralo-

gy of Fallot and Status of Pulmonary Artery by Cardiac CT (Pulmonary Angiography). Journal of Evolution of Medical and Dental Sciences. 2019;8(36):2787-93.

[19] Murai S, Hamada S, Yamamoto S, Khankan AA, Sumikawa H, Inoue A, et al. Evaluation of major aortopulmonary collateral arteries (MAPCAs) using three-dimensional CT angiography: two case reports. Radiat Med. 2004;22(3):186-9.