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ORIGINAL ARTICLE

Role of Multi-detector Computed Tomographic Angiography in Delineation of Pulmonary Venous Variants and Anomalies.

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ABSTRACT

Background: Diagnostic imaging is crucial in the evaluation of thoracic congenital anomalies. Although clinical assessment may provide insight into the possible diagnosis, imaging is usually necessary to confirm the diagnosis, treatment planning and postoperative evaluation. Aiming to assess the role of multidetector computed tomography (MDCT) in evaluating and calculating prevalence of pulmonary venous abnormalities, and be familiar with the imaging characteristics of these abnormalities.

Methods: Ninety patients were enrolled in this retrospective study (33.3% males and 66.7% females) their age ranged from 10 days to 60 years (mean age: 28.7±19.6 years), who underwent MDCT angiography using 128-slice MDCT scanner (ingenuity Philips health care, best, Netherlands) and have been diagnosed of pulmonary venous variants or anomalies.

Results: Pulmonary venous anomalies were detected in 12.2 % of the studied group while the pulmonary variants were 20%. The most common pulmonary venous congenital anomalies were partial anomalous pulmonary venous return (PAPVR) (45.5%) then pulmonary venous varix (27.3 %), followed by total anomalous pulmonary venous return (TAPVR) representing (18.2%) then the meandering pulmonary vein representing (9%) of detected anomalies. While the most common pulmonary venous variant is conjoined pulmonary vein representing 10 % followed by supernumerary right pulmonary vein representing 5.6 % then pulmonary vein open with common ostium representing 4.4% of the studied group.

Conclusion:(MDCT) is considered the modality of choice for accurate delineation of pulmonary venous normal and variant anatomy as well as anomalies and aids planning of interventional procedures especially radiofrequency ablation of arrhythmiogenic foci.

Keywords: MDCT; Total anomalous pulmonary venous return; Partial anomalous pulmonary venous return; Pulmonary venous anomalies.

INTRODUCTION

Vascular thoracic abnormalities represent a significant group of entities that can occur either in isolation or in conjugation with various forms of congenital heart diseases [1]. Diagnostic imaging is crucial when assessing pediatric patients with pulmonary venous anomalies. While clinical evaluation may provide insight into the possible diagnosis, imaging is usually required to confirm the diagnosis, treatment planning and postoperative evaluation [2,3].

The production of ectopic atrial beats is one of the mechanisms that may trigger the onset of atrial fibrillation. The pulmonary veins were found to be a site of this arrhythmogenic activity, which raises the need to detailed anatomic evaluation of pulmonary veins [4].

The advancement of multi-detector computed tomography (MDCT) has increased the clinical use of cardiac CT imaging in patients with pulmonary venous abnormalities and can be used as an alternative to conventional angiography for assessing the pulmonary vascular diseases. Multi-slice computed tomography has the advantages of rapid scan speed; high spatial resolution, simultaneous evaluation of airways and lung parenchyma,

thereby enhancing the ability to answer most clinical questions about structural abnormalities in patients with congenital heart diseases [5].

The aim of the study was to assess the role of MDCT in evaluating and calculating prevalence of pulmonary venous abnormalities, and be familiar with the imaging characteristics of these abnormalities.

METHODS

This retrospective study included 90 patients referred to the Radiology Department, Faculty of Medicine, Zagazig University Hospitals from the outpatient clinics and cardiology department for cardiac /coronary computed tomography angiography (CCTA) in the duration between June 2019 to February 2020. Our patients presented with chest pain or cardiac anomalies resulting in cardiovascular, respiratory or feeding problems.

Patient inclusion criteria were: Patients diagnosed with thoracic venous variants or abnormalities either accidentally discovered or suspected at echocardiography; of any age group and including both genders.Patient exclusion criteria were patients with**:** elevated renal functions (Creatinine level ≥ 2 mg/dl) not on dialysis, contraindications to contrast media, pregnant/lactating females, morbid obesity and inability to sustain breath hold for about 12 sec.

A written informed consent was obtained from all participants/ parents, the study was approved by research ethical committee of Faculty of Medicine, Zagazig University. The study was done according to the code of ethics of the world medical association (Declaration of Helsinki) for studies involving humans. After revising previous investigations (laboratory and cardiac investigations), patients were asked to fast 4 hours and ensure good hydration by adequate amount of simple fluid intake for 3 hours before examination. Heart rate was controlled by Beta – blockers administration one day before examination or Ivabradine three days before examination. Respiratory training was done to hold breath for 12 seconds. Sedation was applied to patients under 8 years old by anesthesiologist. Then intravenous route was applied in right cubital vein in adults and in leg in infants.

The study was performed using 128 multi-detectors (Ingenuity Phillips health care, best, Netherlands) scanner as follows: Scanogram (parameters: 120 KV for adults & 80-100 KV for children, 250-300 mA, a slice pitch of 1.17, slice thickness 1 mm and thin collimation), calcium score calculation in adult, then contrast media is administrated using bolus tracing technique, 1-2 ml/ kg of non-ionic contrast media (Ultravist 370/ Omnipaque 350) was intravenously injected through an 18 gauge catheter with a flow rate of 4-6 ml/sec followed by saline chaser bolus via dual head Medrad stellant injector pump. ROI is placed at descending aorta and acquisition starts when 150 Hounsfield unit (HU) is reached. Image acquisition started from carina till 1 cm below diaphragm for ECG gated coronary CTA and from root of the neck to 1 cm below diaphragm for cardiac CTA.

Patients are kept under observation for 15 minutes after the procedure to check the vital signs (pulse and blood pressure).

Images were transferred to a dedicated Philips work station, revised and interpreted by two independent radiologists with 7 and 12 years' experience in cardiac imaging, using axial images (as source images), reconstructed images (including multiplanar reformation (MPR) (curved and oblique), maximum intensity projection (MIP) and three dimentional (3D) volume rendering (VR) techniques). 3D volume rendering and thick thickness curved multiplanar reformation (C-MPR) were the best post processing view in delineation of the pulmonary veins.

Statistical analysis: Data were collected, coded, entered and analyzed using Microsoft Excel software. Data were then imported into Statistical Package for the Social Sciences (SPSS version 23.0) software for analysis. Qualitative data were represented as number and percentage; quantitative data were represented by mean ± SD. P value was set at <0.05 for significant results & <0.001 for high significant result.

RESULTS

90 patients were enrolled in this study (33.3% males and 66.7% females) their age ranged

from 10 days to 60 years (mean age: 28.7±19.6 years). The pattern of Pulmonary venous drainage was classified into three groups: normal four vessels pattern (67.8%, n: 61 patients), pulmonary venous drainage variants (20%, n: 18) and anomalous pulmonary venous drainage (12.2%, n: 11).

Pulmonary venous anomalies prevalence

Partial anomalous pulmonary venous return (PAPVR) was the commonest detected pulmonary venous anomaly (45.5% of detected anomalies, 5.6% prevalence) (figure 1 and 2) followed by pulmonary varix (27.3% of detected anomalies, 3.3% prevalence) then total anomalous pulmonary venous return (TAPVR) (18.2% of detected anomalies, 2.2% prevalence) (figure 3) and lastly a single pulmonary vein drained at persistent levo-atrio-cardinal vein (9% of detected anomalies, 1.1% prevalence) (Table 1).

Pulmonary venous variants prevalence

The commonest pulmonary venous variant detected was conjoined pulmonary veins which represents (50 % of detected variants, 10% prevalence) followed by supernumerary pulmonary vein (27.8 % of detected variants, 5.6 % prevalence) then pulmonary veins open with a common ostium (22.2 % of detected variants, 4.4 % prevalence) (Table 2).

Relation between ASD presence and pulmonary venous variants and anomalies

There was statistically significant association between different pulmonary veins abnormalities and presence of ASD (p: 0.04) where (100.0%, 83.3%, 66.7% & 33.3%) of TAPVR, PAPVR, conjoined pulmonary veins and Pulmonary varix had ASD respectively with absent ASD among other pulmonary veins variants (Table 3).

Relation between VSD presence and pulmonary veins variants and anomalies

There was no statistically significant difference between the different pulmonary veins abnormalities regarding presence of VSD where (50.0%, 33.3% & 16.7%) of TAPVR, conjoined pulmonary veins and PAPVR respectively had VSD with absent VSD among other pulmonary veins abnormalities (Table 4).

Table (1): Frequency distribution of pulmonary venous anomalies.

Pulmonary venous anomalies	No. (90)	Incidence among anomalies %	Prevalence %
PAPVR	5	45.5	5.6
Meandering pulmonary vein	1	9	1.1
TAPVR	2	18.2	2.2
Pulmonary varix	3	27.3	3.3
Total	**11**	**100**	**12.2**

PAPVR: Partial anomalous pulmonary venous return
TAPVR: Total anomalous pulmonary venous return

Table (2): Frequency distribution of pulmonary venous variants.

Pulmonary venous variants	No. (90)	Incidence among variants %	Prevalence %
Conjoined Pulmonary Vein	9	50	10
Pulmonary veins open with common ostium	4	22.2	4.4

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Pulmonary venous variants	No. (90)	Incidence among variants %	Prevalence %
Supernumerary right Pulmonary Vein	5	27.8	5.6
total	18	100	20

Table (3): Relation between ASD presence and pulmonary venous variants and anomalies.

Pulmonary venous variants and anomalies (29)	ASD				χ^2	p-value
	Absent		Present			
	No (15)	%	No (14)	%		
PAPVR	1	16.7	5	83.3		
TAPVR	0	0.0	2	100.0		
Pulmonary varix	2	66.7	1	33.3		
Conjoined pulmonary Veins	3	33.3	6	66.7	5.9	**0.04****
Pulmonary veins open with common ostium	4	100.0	0	0.0		
Supernumerary right pulmonary vein	5	100.0	0	0.0		

ASD: Atrial septal defect
PAPVR: Partial anomalous pulmonary venous return
TAPVR: Total anomalous pulmonary venous return
There was a statistically significant difference among different pulmonary veins variants regarding presence of ASD

Table (4): Relation between VSD presence and pulmonary venous variants and anomalies.

Pulmonary venous variants and anomalies	VSD				χ^2	p-value
	Absent		Present			
	No (25)	%	No (4)	%		
PAPVR	5	83.3	1	16.7		
TAPVR	1	50.0	1	50.0		
Pulmonary varix	3	100.0	0	0.0		
Conjoined pulmonary Veins	7	66.7	2	33.3	1.6	**0.06***
Pulmonary veins open with common ostium	4	100.0	0	0.0		
Supernumerary right pulmonary vein	5	100.0	0	0.0		

VSD: Ventricular septal defect
PAPVR: Partial anomalous pulmonary venous return
TAPVR: Total anomalous pulmonary venous return
There was no statistically significant difference among different pulmonary veins variants regarding presence of VSD.

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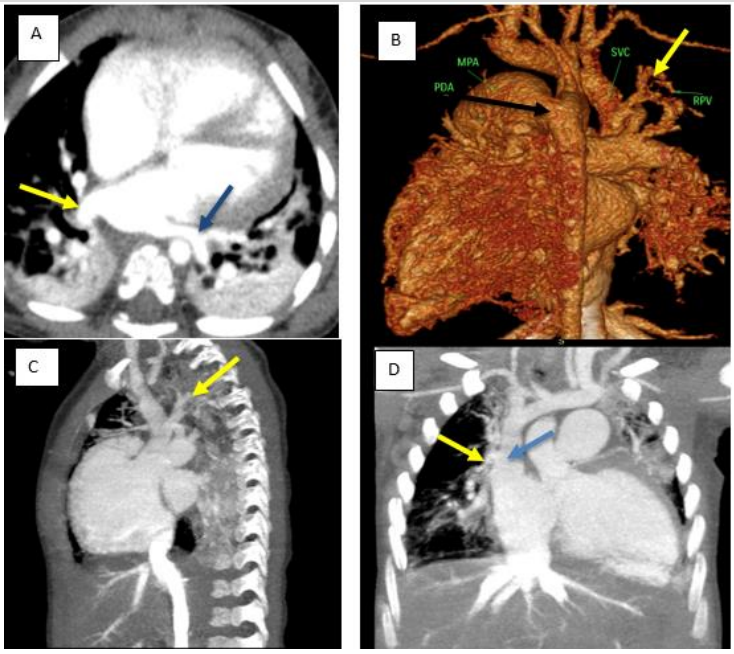

Figure (1): A 2-years-old female patient with partial anomalous pulmonary venous return (PAPVR), signs of pulmonary artery hypertension and patent ductus arteriosus (PDA). (A) Axial CT image showing cardiomegaly with dilated right atrium (RA) and right ventricle (RV) with one right pulmonary vein (RPV) (yellow arrow) and conjoined left pulmonary veins (blue arrow) drain at left atrium (LA) with bilateral basal consolidation. (B) & (C) 3D VR and sagittal MIP images showing patent ductus arteriosus (PDA) (black arrow) connecting left sided aortic arch to dilated main pulmonary artery (MPA) & an anomalous right pulmonary vein (RPV) (yellow arrows) drains at the superior vena cava (SVC). (D) Coronal MIP showing another anomalous right pulmonary vein (yellow arrow) drains at the superior vena cava (blue arrow).

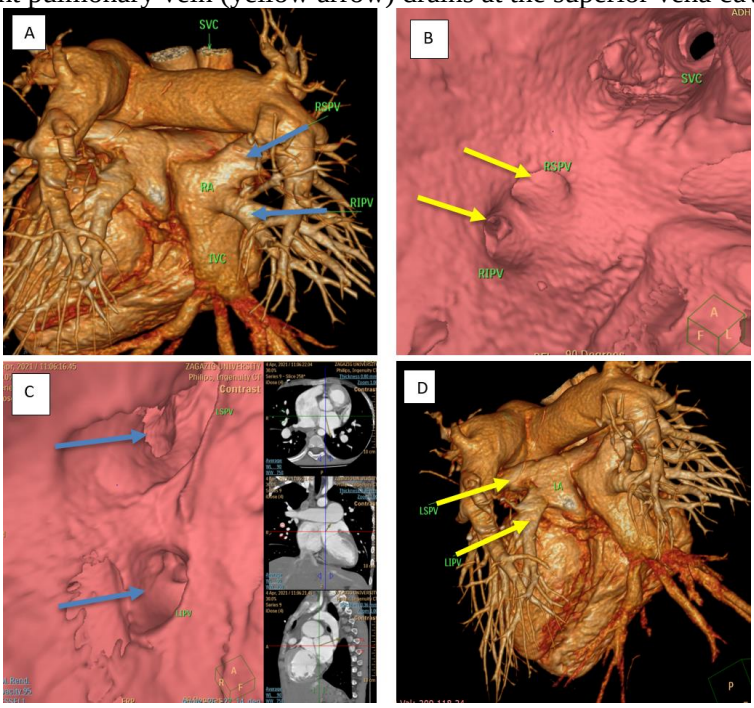

Figure (2): An 8-years-old male patient with cardiac PAPVR and veno-atrial discordance. (A) 3D VR image shows right superior (RSPV) and right inferior (RIPV) pulmonary veins (blue arrows) drain into right atrium (RA). (B) Endocardial VR image (endocardial view) from multidetector CT shows right pulmonary veins ostia (RSPV and RIPV) (yellow arrows) as well as superior vena cava (SVC) draining into right atrium. (C) Endocardial VR image (endocardial view) from multidetector CT shows left superior (LSPV) and left inferior (LIPV) pulmonary veins ostia (blue arrows) draining into left atrium. (D) 3D VR image shows left pulmonary veins (yellow arrows) drain into left atrium (LA).

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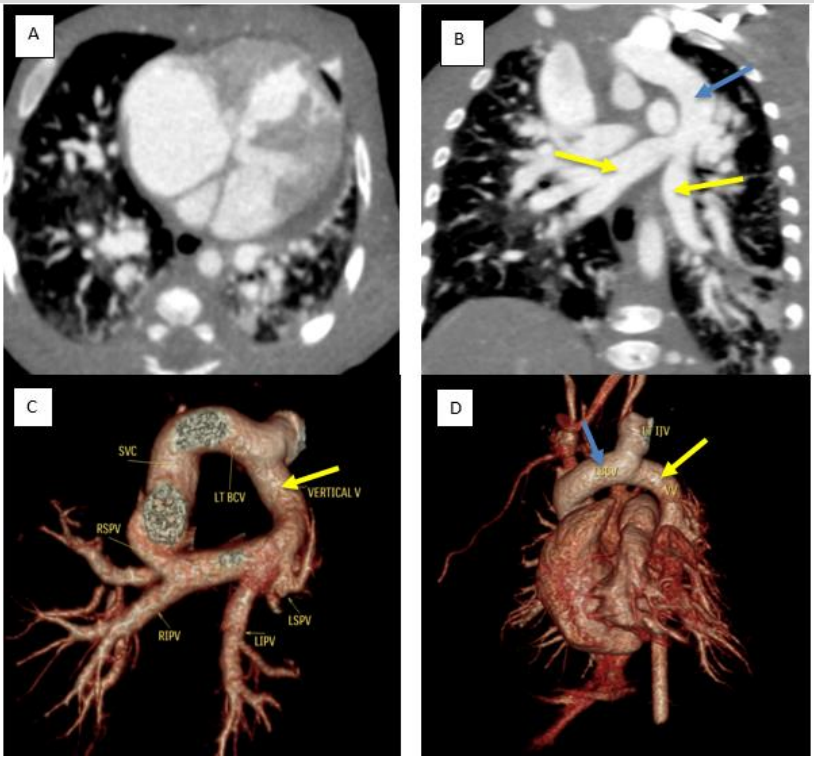

Figure (3): A 30-day-old male patient with non-obstructed supra-cardiac TAPVR and cortriatriatum dextrum. (A) Axial CT image showing enlarged right atrium subdivided by thin membrane (cortriatriatum dextrum) with hypoplastic left atrium and left ventricle. (B) Curved planar reformatted CT image showing right and left pulmonary veins (yellow arrows) drain into a left vertical vein (blue arrow). (C &D) 3D VR images showing right superior (RSPV), right inferior (RIPV), left superior (LSPV) and left inferior (LIPV) pulmonary veins drain into a common chamber then to left vertical vein (yellow arrow) which drains into left brachiocephalic vein (BCV) (blue arrow). IJV: internal jugular vein.

DISCUSSION

A wide variety of pulmonary veins arrangement and anomalies exists. Detailed examination of the pulmonary veins pattern should be done when evaluating cross-sectional images [6].

Pathology of the pulmonary veins involves both congenital and acquired conditions. Some of these disorders are benign, disruption of the pulmonary vasculature is often life-threatening, making these conditions essential to identify on imaging [7].

The pulmonary veins were found to be a significant site of arrhythmogenic activity, resulting in up to 96% of foci triggering paroxysmal episodes of AF. Ectopic foci most commonly derive from the left superior pulmonary vein due to its longer myocardial sleeve and thicker atrio-pulmonary venous junction. Radiofrequency ablation (RFA) has been used to disconnect the electrical connection between the pulmonary veins and the left atrium, and thus treat atrial fibrillation [4].

Types of anomalous pulmonary venous drainage are; supracardiac, cardiac, infracardiac and mixed types; the most common mixed type is the combination between cardiac and supracardiac types [8].

Partial anomalous pulmonary venous return (PAPVR) is the draining of blood from one or more anomalous pulmonary vein to the systemic circulation forming a right to left shunt. The overall occurrence of partial anomalous pulmonary venous return (PAPVR) is 0.5–0.7% approximately [8].

In our study, we found that PAPVR was the most common pulmonary venous congenital anomaly with 5.6% prevalence, 60% of which were supracardiac type while 40% were cardiac type. The prevalence of PAPVR raises

to 6.7% when adding the case of anomalous drainage of left superior pulmonary vein to persistent levo-atrio-cardinal vein. (Levo atrio cardinal vein is anomalous pulmonary vein connecting the left atrium with the systemic venous system and is commonly associated with hypogenetic lung syndrome [9]. This comes in agreement with Aborashed et al. [10] who stated that the most common thoracic venous anomalies were PAPVR by a prevalence of 8.75% of all studied thoracic vascular anomalies. However, our study disagreed with Berko et al. [11] and Dillman et al. [12] who reported much lower prevalence of PAPVR (0.3 % and 0.4% - 0.7%) respectively, however, their study included adult population only.

Pulmonary varix is a rare condition characterized by focal enlargement of segmental pulmonary veins and may be mistaken as lung nodule at radiographs [13].

Sabri et al. [14] in their study stated that the most common pulmonary anomaly is pulmonary varix which represents about (20.3%), this almost agrees with our results that pulmonary varix was the 2nd most common pulmonary venous anomaly (27.3% of anomalies with 3.3% prevalence). In contrast to our results, Couvreur and Ghaye [15] reported that the pulmonary venous varix is rare (as only 71 cases have been reported up to 1988).

Total anomalous pulmonary venous return (TAPVR) is the abnormal drainage of the entire pulmonary venous system into a systemic vein and always associated with ventricular septal defect (VSD) [13]. Total anomalous pulmonary venous return (TAPVR) is most commonly a cyanotic condition, occurring in 1%–3% of children with congenital heart defects [8].

In our study, supra-cardiac TAPVR accounted for 18.2% of detected anomalies with 2.2% prevalence. In Partial agreement with our results, EL Fiky et al. [1] and Aborashed et al. [10] stated that 5.8 % of the studied population had TAPVR. However, Oh et al. [16] and Osama et al. [17] reported much higher prevalence of such anomaly (50% and 31%). This difference can be attributed to different study populations. Our study stated that meandering vein is rare with 1.1 % prevalence. We agreed with Porres et al. [9] who stated that anomalous unilateral single pulmonary vein (meandering vein) is extremely unusual.

In our study pulmonary venous anomalies were commonly associated with other congenital cardiac anomalies. TAPVR was associated with atrial septal defect (ASD) (100%) and ventricular septal defect (VSD) (50%) while PAPVR was associated with various congenital cardiac anomalies the most common was ASD (83.3%), PDA (50%) and VSD (16.7%), which agrees with Hellinger et al. [18] who stated that PAPVR may be associated with a sinus venosus defect. Also Broy et al. [19] stated that PAPVR is found most commonly in association with congenital cardiac anomalies with the closest association with ASD of the sinus venosus type. Our results showed a statistically significant association between pulmonary venous anomalies and presence of ASD (p= 0.04).

There were typically four pulmonary veins, two draining each lung into the left atrium in 61% of studied population, while anatomic variants were detected in 20% of our patients; the commonest was conjoined pulmonary veins draining the left lung by a common trunk (50% of variants, 10% prevalence) followed by right supernumerary vein /veins (27.8% of variants, 5.6% prevalence) then left pulmonary veins drained by a common ostium into left atrium (22.2% of variants, 4.4% prevalence). Our results agreed with Prasanna et al. [20] who reported that the most common drainage pattern was two pulmonary veins with two separate ostia on each side, the next common drainage pattern on the right side was three pulmonary veins with three ostia (24%).

Inter-observer agreement was done between two radiologists. They agreed in diagnosis of all cases except only one case where one radiologist diagnosed it as meandering vein while the other diagnosed it as PAPVR in which left superior pulmonary vein drains at persistent levo-atrio-cardinal vein. There were some limitations to this study. First, there is lack of reference standard which we overcome by inter-observer agreement method. Second, the wide range of age which was included in the study. Finally, and most important,

selection bias thus makes the proportion of variants and anomalies among patients with congenital heart diseases very different from the true proportion in general population.

In conclusion, MDCT angiography is considered the modality of choice for accurate delineation of pulmonary venous normal and variant anatomy as well as anomalies and aids planning of interventional procedures especially radiofrequency ablation of arrhythmiogenic foci.

List of abbreviations: ASD: Atrial septal defect; MIP: Maximum intensity projection; MPR: Multiplanar reformation; PAPVR: Partial anomalous pulmonary venous return; TAPVR: Total anomalous pulmonary venous return; VSD: Ventricular septal defect.

Conflict of Interest: None

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