RESEARCH

Spectrum of thoracic systemic venous abnormalities using multidetector computed tomography

Samar Shehata, Ghada Abdulmonaem, Aya Gamal* and Mostafa Assy

Abstract

Background: Anomalies of the thoracic systemic venous return vary widely and range from those with completely normal physiology to severe right to left shunting thus requiring surgical correction. The aim of the study is to enhance the awareness of multidetector computed tomography (MDCT) role in evaluation of systemic venous abnormalities, and be familiar with the imaging characteristics of even the extremely rare abnormalities.

Results: Among 270 examined patients, 15.19% had systemic venous abnormalities. Inferior vena cava (IVC) congenital anomalies accounted for (24.4%) of the detected abnormalities (prevalence: 3.7% among the studied population) where IVC interruption with azygos continuation was the most common detected IVC abnormality accounting for 17.7% of the detected abnormalities (prevalence: 2.6%), while IVC thrombosis accounted for 21.9% (prevalence: 3.3%). Persistent left sided superior vena cava (SVC) accounted for 14.6% of the detected abnormalities (prevalence: 2.2%), while SVC syndrome represented 19.5% (prevalence: 2.9%) and SVC aneurysm represented 2.4% (prevalence: 0.37%). Retroaortic brachiocephalic vein (BCV) and BCV thrombosis accounted for 7.3% each (prevalence: 1.1). Finally, persistent levo-atrial cardinal vein represented 2.4% of the detected abnormalities (prevalence: 0.37%).

Conclusions: MDCT is a non-invasive modality that can provide detailed information about the systemic thoracic veins before surgical or interventional procedures, especially in patients with congenital anomalies.

Keywords: Multidetector CT angiography, Systemic venous abnormalities, Superior vena cava, Inferior vena cava

Introduction

CT scan is a non-invasive and rapid procedure used as an alternative to conventional angiography for assessing systemic vascular diseases [1].

It is critical to be aware of the presence of systemic venous abnormalities prior to surgical or interventional procedures to perform the procedure safely, especially in patients with congenital cardiac malformations [2].

Awareness of systemic venous abnormalities, particularly left SVC, is essential when considering a left superior venous approach to the heart in patients undergoing pacemaker placement or defibrillator, and when using retrograde cardioplegia for surgical approaches requiring cardiopulmonary bypass [3].

The aim of this study was to enhance the awareness of MDCT role in evaluation of systemic venous abnormalities, and be familiar with the imaging characteristics of even the extremely rare abnormalities.

Methods

Patients

This study was carried out at Radiology Department, Faculty of Medicine, from December 2019 to September 2020. Written informed consent was obtained from all participants and the study was approved by the research ethical committee of Faculty of Medicine. The work has been carried out in accordance with The Code

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of Ethics of the World Medical Association (Declaration of Helsinki) for studies involving humans.

This retrospective study included two hundred and seventy patients referred for cardiovascular imaging unit and underwent MDCT angiography. Patients had isolated systemic venous lesions or associated complex cardiac pathologies resulting in respiratory, cardiovascular, or feeding problems.

Patient inclusion criteria were: Patients of any age group and gender diagnosed with thoracic venous variations or abnormalities either accidentally discovered on MDCT or suspected by echocardiography.

Patient exclusion criteria were patients with: contraindications to contrast media, impaired renal functions [(Creatinine level ≥ 2 mg/dl) or (GFR < 30 ml/min/1.73 m^2) for adults and (GFR < 60 ml/min/1.73 m^2), calculated by Schwartz's formula [4], for children but not on dialysis], morbid obesity, pregnant/lactating females or inability to sustain breath hold for about 12 s. In neonates and children; we perform free breathing study as we concerned with gross congenital anomalies and the origin and proximal course only of the coronaries.

Patient preparation

Patients were prepared for examination after revising previous investigations (laboratory and cardiac) as follows; Fasting for 4 h before the examination; Proper amount of simple fluid intake for 3 h before the examination to maintain good hydration; Beta-blockers 1 day before the examination or Ivabradine 3 days before the examination to control heart rate; Respiratory training to hold breath for 12 s; Sedation by an anesthesiologist for patients below 8 years; Intravenous route in the right cubital vein in adults and the leg in infants.

Image acquisition

This retrospective study was done using 128 multi-detectors (Ingenuity Phillips health care, best, Netherlands) scanner as follows: Scanogram, calcium score calculation in adult, using bolus tracking technique; non-ionic contrast media (350/370 mg Iodine/ml) was injected (1.5 m /kg body weight for adults and 2 ml/kg body weight for children < 12 years) followed by saline chaser bolus via dual head Medrad stellant injector pump. 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 min after the procedure to check the vital signs (pulse and blood pressure).

Image reconstruction and interpretation

The cases were transferred to a dedicated Philips workstation (Version; V5.0.2.10010). Images were interpreted and revised by two radiologists with 7 and 15 years of experience in cardiac imaging, using axial images (as source images), reconstructed images (including MPR, MIP and volume rendering techniques).

Statistical analysis

Data were collected, coded, entered, and analyzed using Microsoft Excel software. Data were then transferred into Statistical Package for the Social Sciences (SPSS version 23.0) software for analysis.
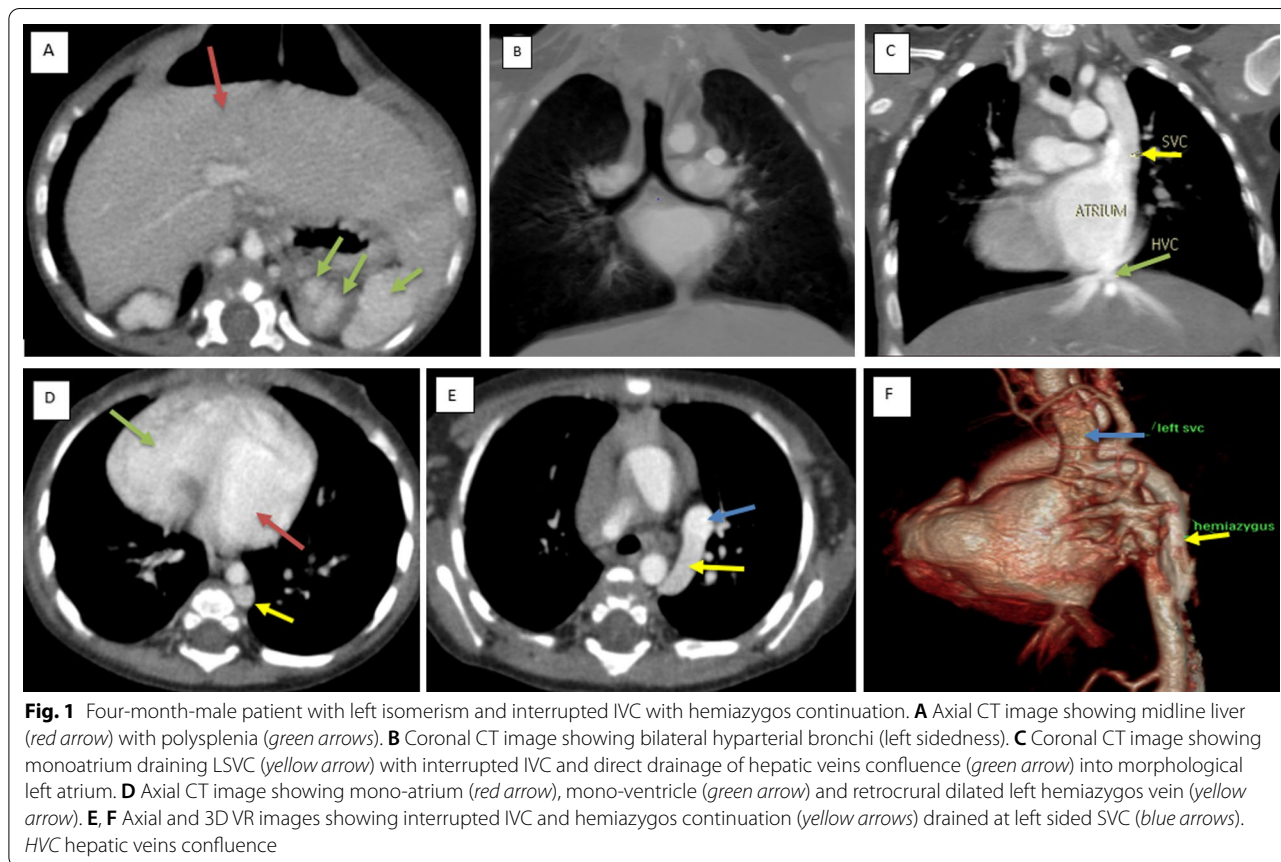
Results

Clinical data of the studied group

Our patients were presented with either manifestations of systemic venous obstruction (7.4%) or manifestations of associated congenital cardiac anomalies (92.6%) (Table 1).

Inferior vena cava abnormalities prevalence

In our study, 7% of the studied population had IVC abnormalities representing 46.34% of the detected abnormalities. Congenital IVC anomalies were noted among 3.7% of the studied group and accounted for 24.4% of the detected abnormalities, where inferior vena cava interruption with azygos continuation was the most common with prevalence of 2.6% among the studied population followed by IVC interruption with hemiazygos/accessory hemiazygos continuation (Fig. 1) with a prevalence of 1.1% among the studied group. IVC thrombosis had a prevalence of 3.3% among the studied group and represented 21.95% of the detected anomalies (Table 2).

Superior vena cava abnormalities prevalence

In our study, 5.6% of the studied population had SVC abnormalities representing 36.58% of the detected abnormalities. The most common detected SVC abnormality was persistent left SVC (PLSVC) (Fig. 2) which represented 14.63% of the detected abnormalities with 2.2% prevalence among the studied group, followed by SVC syndrome (Fig. 3) which represented 19.5% of the detected abnormalities and 2.96% prevalence, then SVC aneurysm accounted for 2.4% of the detected abnormalities and 0.37% prevalence. Aberrant left brachiocephalic vein with retro-aortic anomalous course and BCV thrombosis represented 7.3% of the detected abnormalities each and prevalence of 1.1% each. Finally, persistent levoatriocardinal vein (PLACV) (Fig. 4) represented 2.4% of the detected abnormalities and had a prevalence of 0.37% (Table 3).

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Fig. 1 Four-month-male patient with left isomerism and interrupted IVC with hemiazygos continuation. **A** Axial CT image showing midline liver (*red arrow*) with polysplenia (*green arrows*). **B** Coronal CT image showing bilateral hyparterial bronchi (left sidedness). **C** Coronal CT image showing monoatrium draining LSVC (*yellow arrow*) with interrupted IVC and direct drainage of hepatic veins confluence (*green arrow*) into morphological left atrium. **D** Axial CT image showing mono-atrium (*red arrow*), mono-ventricle (*green arrow*) and retrocrural dilated left hemiazygos vein (*yellow arrow*). **E, F** Axial and 3D VR images showing interrupted IVC and hemiazygos continuation (*yellow arrows*) drained at left sided SVC (*blue arrows*). *HVC* hepatic veins confluence

Half of PLSVC patients had isolated persistent left SVC (IPLSVC) while the other half had dual SVC. PLSVC drains at right atrium through dilated coronary sinus in all cases apart from one case (0.37% of the studied population and 16.6% of PLSVC cases) where it continues as accessory hemiazygos» azygos» right SVC.

Most patients (83.3%) had associated cardiac anomalies in the form of mixed heterotaxy (16.6%), left isomerism (16.6%), partial anomalous pulmonary venous return (PAPVR) (33.3%), mitral valve atresia (16.6%), and monoventricle (16.6%).

Inter-observer agreement

There was a highly significant excellent agreement between both observers regarding the role of multidetector computed tomographic angiography in delineating thoracic venous variants and abnormalities; both observers detected all positive cases (Table 4).

Discussion

Awareness of systemic venous abnormalities, especially left superior vena cava, is critical when considering a left SVC approach to the heart in patients undergoing defibrillator

Table 1 Clinical picture of the studied patients

Clinical data	%
Arrhythmia	20
Dyspnea	13.33
Cyanosis	17
Tachycardia	22.2
Chest pain	20
Leg heaviness and swelling	1.1
Upper limb edema	1.1
Chest wall collaterals	1.48
Follow-up malignancy	3.7

or pacemaker placement, and when using retrograde cardioplegia for surgical procedures requiring cardiopulmonary bypass [3].

In our study, our patients were presented with either manifestations of systemic venous obstruction or manifestations of associated congenital cardiac anomalies, our results agree with Berko et al. [5], who reported no significant difference in symptoms in patients with systemic vascular variants and anomalies.

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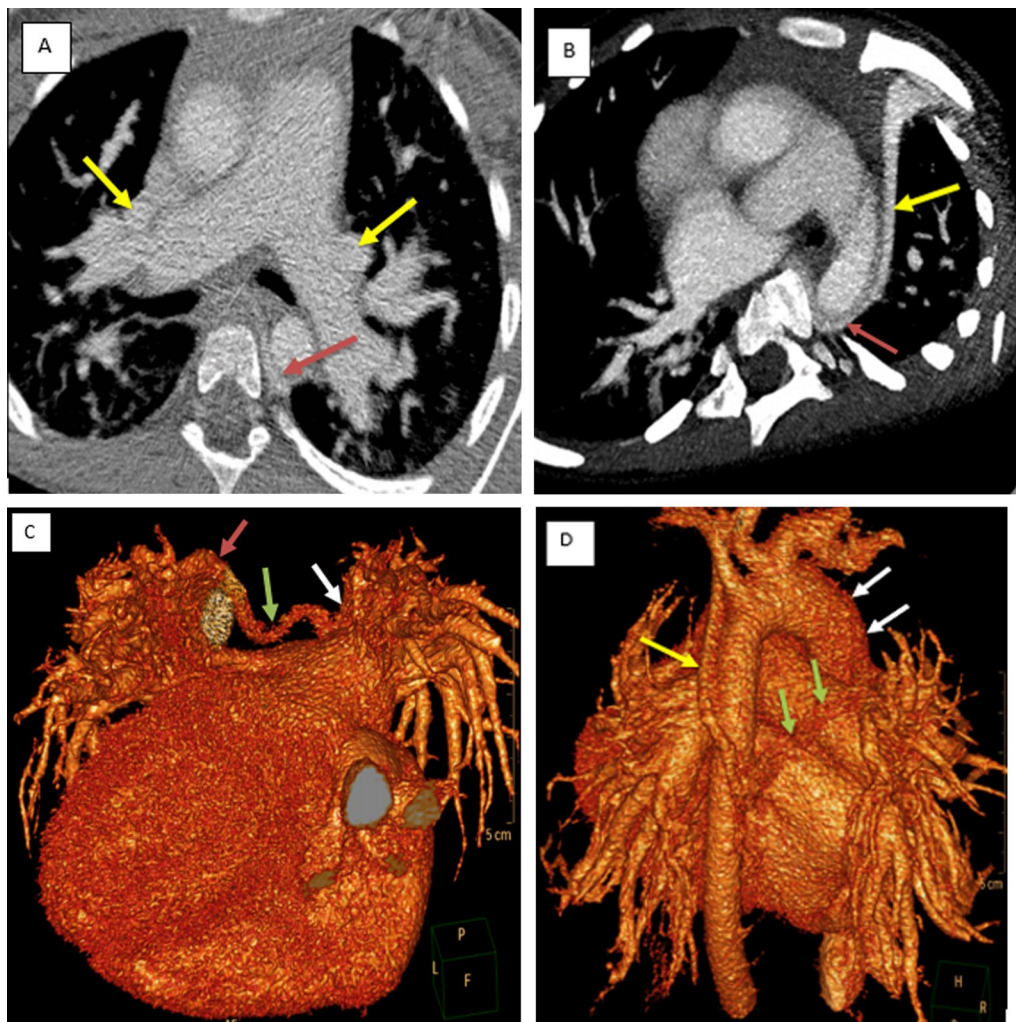
Fig. 2 One-year-old male patient with dual superior vena cava (DSVC) with PLSVC draining into the RSVC through azygos system. **A** Axial CT showing dual SVC (*yellow arrows*) with dilated accessory hemiazygos vein (*red arrow*). **B** Axial CT MIP and **C**, **D** 3D VR images showing PLSVC (*yellow arrow*), which continues as accessory hemiazygos (*red arrow*) connected through midline anastomotic collaterals (*green arrows*) to the azygos veins (*white arrows*) which drained into RSVC

Table 2 Frequency distribution of inferior vena cava abnormalities among the studied group

Inferior vena cava	No. (270)	Incidence among anomalies % No. (41)	Prevalence %
IVC interruption with azygos continuation	7	17.1	2.6
IVC interruption with hemiazygos continuation	3	7.3	1.1
IVC thrombosis	9	21.95	3.3
Total	19	46.34	7

Table 3 Frequency distribution of systemic venous abnormalities among the studied group

Systemic veins variants	No. (270)	Incidence among anomalies % No. (41)	Prevalence %
Persistent left SVC	6	14.6	2.2
SVC syndrome	8	19.5	2.96
SVC aneurysm	1	2.4	0.37
Persistent LACV	1	2.4	0.37
BCV thrombosis	3	7.3	1.1
Retro-aortic BCV	3	7.3	1.1
Total	22	53.66	8.1

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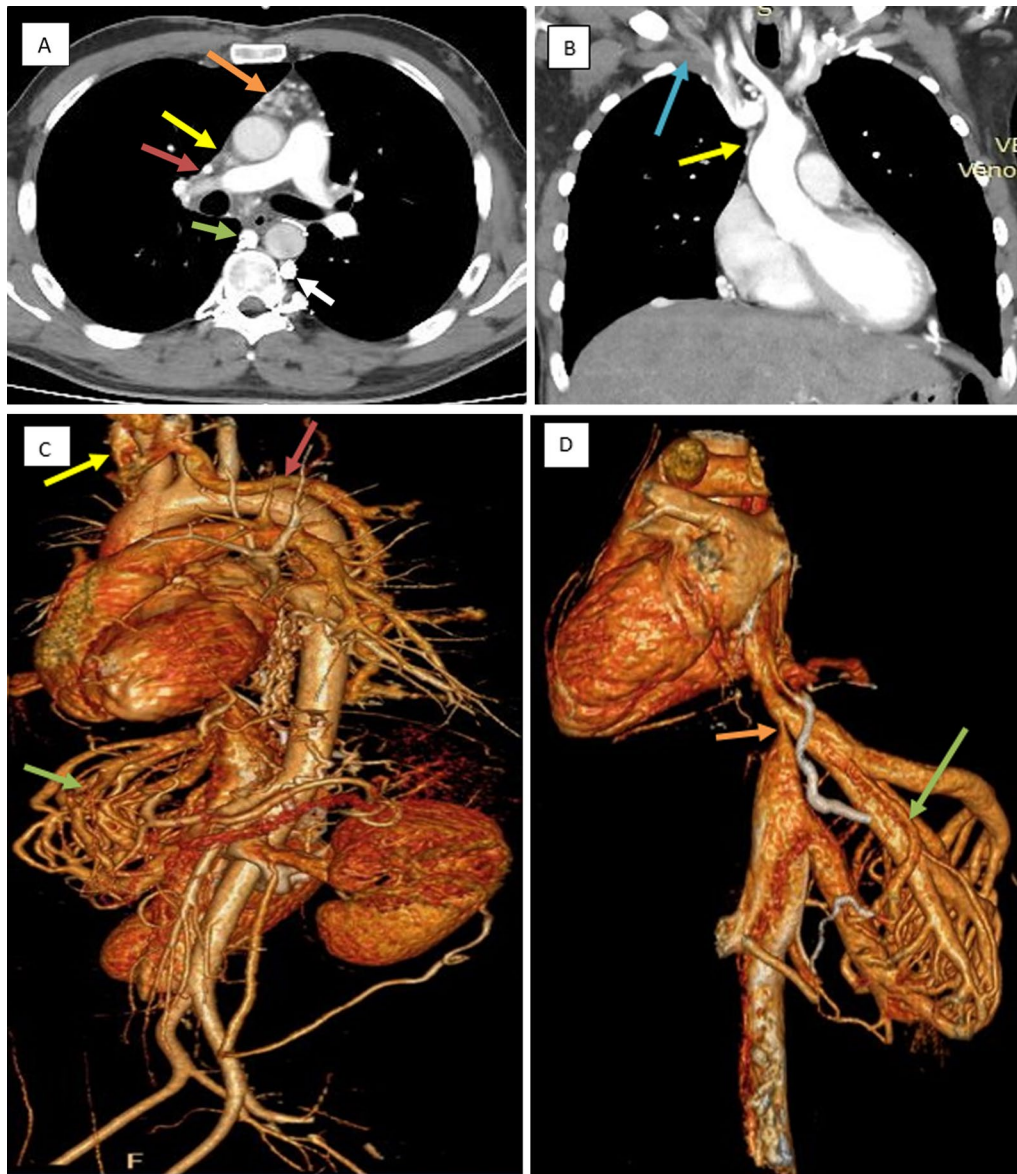
Fig. 3 28-year-old male patient with chronic vena cava thrombosis. **A**, **B** Axial and coronal CT chest showing attenuated thrombosed SVC (*yellow arrow*), thrombosed right subclavian vein (*blue arrow*) and azygos arch (*red arrow*) with dilated azygos (*green arrow*) and hemiazygos veins (*white arrow*) and multiple collaterals (*orange arrow*). **C**, **D** 3D VR images showing stenosed suprahepatic portion of IVC (4 mm) (*orange arrow*) with multiple intrahepatic venous collaterals (*green arrow*), and dilated hemiazygos and accessory hemiazygos vein (*red arrow*) draining into LBCV (*yellow arrow*)

Table 4 Concordance (inter-observer agreement) between observer A and B among the studied group regarding the role of multidetector computed tomographic angiography in delineation of systemic venous abnormalities

Systemic venous abnormalities	Observer A				Kappa agreement	*p* value
	Yes		No			
	No. (39)	%	No. (231)	%		
Observer B						
Yes (40)	38	97.4	2	0.9	0.98	0.00*
No (230)	1	2.6	229	99.1		

*Statistically highly significant difference (*P* ≤ 0.001)

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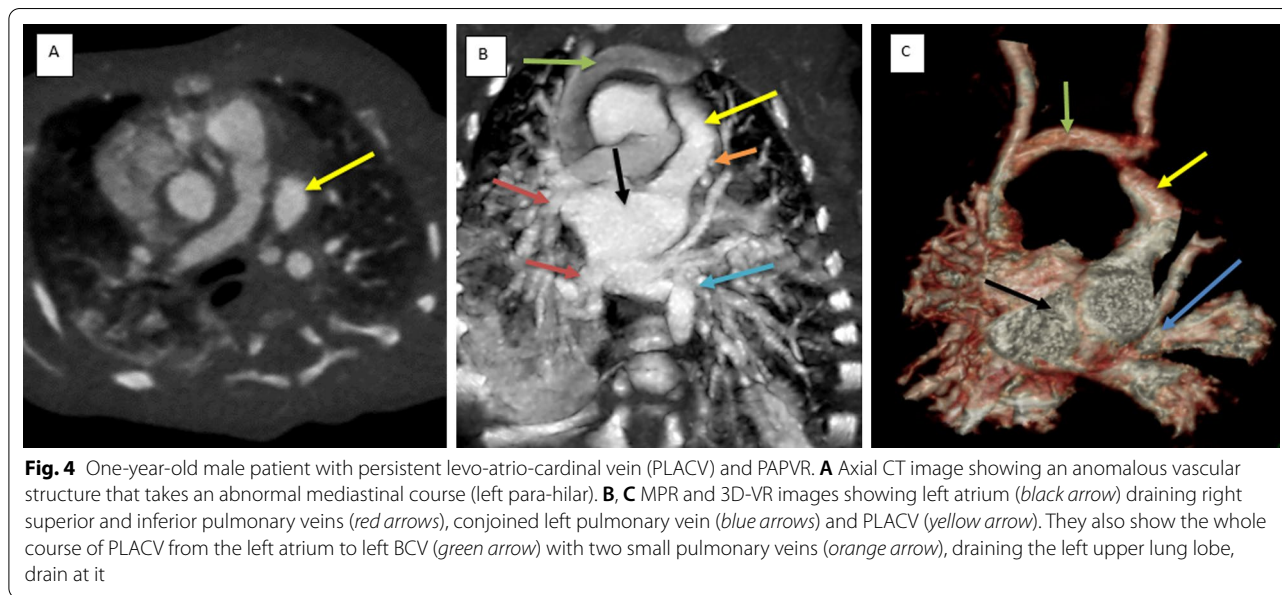
Fig. 4 One-year-old male patient with persistent levo-atrio-cardinal vein (PLACV) and PAPVR. **A** Axial CT image showing an anomalous vascular structure that takes an abnormal mediastinal course (left para-hilar). **B**, **C** MPR and 3D-VR images showing left atrium (*black arrow*) draining right superior and inferior pulmonary veins (*red arrows*), conjoined left pulmonary vein (*blue arrows*) and PLACV (*yellow arrow*). They also show the whole course of PLACV from the left atrium to left BCV (*green arrow*) with two small pulmonary veins (*orange arrow*), draining the left upper lung lobe, drain at it

IVC abnormalities represented 7% of the studied group, 3.7% of which were congenitally interrupted IVC with either azygos (2.6%) or hemiazygos (1.1%) continuation. Two-thirds of those patients had left isomerism, while the rest had an isolated IVC interruption. This finding is consistent with Obernosterer et al. [6], who reported that the IVC anomalies have a prevalence of 0.07–8.7% in general population.

We relatively agree with Mandato et al. [7], who stated that the interruption of the inferior vena cava (IVC) with azygos continuation is a rare congenital anomaly. In contrast, Aborashed et al. [8], reported that the azygos/hemiazygos continuation percentage reaches 23.5% of the studied population. This difference may be because they had a larger sample size.

In agreement with Hollingsworth et al. [9], who reported that IVC thrombosis is considered rare, IVC thrombosis in the current study represented only 3.3% of the studied group and was common among drug addicts.

The diagnosis of PLSVC is crucial prior to some cardiac surgeries, such as venous rerouting procedures, cavopulmonary anastomosis, and heart transplantation [10].

In our study, superior vena cava abnormalities accounted for 36.58% of the detected abnormalities among 5.6% of the studied population. The prevalence of persistent left SVC (PLSVC) was 2.2%, while the prevalence of acquired lesions was 3.3% in the form of SVC syndrome or SVC aneurysm; 2.96% and 0.37%, respectively.

Our results agreed with Aborashed et al. [8], who found that persistent left SVC is considered the most frequent anomaly of thoracic venous anomalies along with partial anomalous pulmonary venous return.

In our study the prevalence of SVC syndrome, including both SVC indentation and thrombosis, was nearly 3%. It accounted for 19.5% of the detected abnormalities. This finding was attributed to mediastinal malignancy (37.5%) or the presence of central vein catheters (62.5%). In contrast, Birch et al. [11], stated that SVC syndrome resulting from SVC and internal jugular vein (IJV) thrombosis is rare. Rice et al. [12], in their study reported that, malignancy is the most common cause of the superior vena cava syndrome (60%); however, they reported an increasing incidence of benign etiologies of SVC syndrome due to the increasing use of intravascular devices [12].

Superior vena cava aneurysm is a very rare mediastinal vascular lesion accounting for 0.37% of the studied population. To our knowledge, the first case was reported by Abbot [13] in 1950, and approximately 37 cases were only reported till 2017. These results agree with Janczac et al. [14], and Sharma et al. [15].

Anomalous left brachiocephalic vein (ALBCV) is a rare systemic venous anomaly (1%). This vein takes an abnormal retro-aortic course beneath the aortic arch to create the superior vena cava. This condition is frequently seen in association with other congenital anomalies, mostly tetralogy of fallot (TOF) and right sided aortic arch [10, 16]. ALBCV accounted for 1.1% (3/270) of our studied population and 7.3% of the detected anomalies where two cases were associated with TOF and one with right isomerism.

The levoatriocardinal vein (LACV) is an extremely rare form of pulmonary systemic connection. Approximately 50 cases were reported in the literature till 2015. It originates from the left atrium (68%) or pulmonary vein (32%) and drains into one of the systemic venous structures,

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mostly into the left brachiocephalic vein (LBCV) (48%) [10, 17]. In our study, LACV accounted for 2.4% of the detected anomalies with a prevalence of 0.37%. While the whole left lung is drained by a conjoined left pulmonary vein, two small pulmonary veins drain left upper lung lobe into LACV. It was associated with patent ductus arteriosus (PDA) and atrial septal defect (ASD).

In our study, there is a highly significant excellent agreement between both observers regarding the role of multidetector computed tomographic angiography (MDCTA) in delineating thoracic venous abnormalities. We recommend using MDCTA as a first line diagnostic tool that can accurately delineate systemic venous abnormalities before interventional procedures.

Limitations to this study were: First, the lack of reference standard which was overcome using inter-observer agreement method. Second, respiratory motion and high heart rate in children that we partially overcame by using intravenous Fentanyl by anesthesiologists. Third, we included a wide range of ages in the study. Finally, and most important, it is a retrospective analysis involving a selected group of patients, this makes the proportion of normal subjects to those with congenital heart disease (CHD) very different from the true proportion in the population (selection bias).

Conclusions

MDCT is a non-invasive modality which can provide detailed information about the thoracic systemic veins prior to surgical or interventional procedures, especially in patients with congenital anomalies.

Abbreviations

ALBCV: Anomalous left brachiocephalic vein; ASD: Atrial septal defect; BCV: Brachiocephalic vein; CHD: Congenital heart disease; CTA: Computed tomographic angiography; IJV: Internal jugular vein; IVC: Inferior vena cava; LBCV: Left brachiocephalic vein; MDCT: Multidetector computed tomography; MDCTA: Multidetector computed tomographic angiography; MIP: Maximum intensity projection; MPR: Multiplanar reformatting; PDA: Patent ductus arteriosus; PLACV: Persistent levoatriocardinal vein; PLSVC: Persistent left superior vena cava; SVC: Superior vena cava.

Availability of data and materials

The data supporting the results of this article are included within the article.

Declarations

Ethics approval and consent to participate

The study was approved by the institutional ethics committee of Faculty of Medicine, Zagazig University. IRB statement code is 5340 and date of approval was 26/3/2019. A written consent was taken from all of the participants after explaining the details, benefits, and risks to them.

Consent for publication

Consent for publication has been obtained from the participants involved in the study to report their individual patient data.

Competing interests

The authors declare that they have no competing interests.

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