Contents lists available at ScienceDirect

Heliyon



journal homepage: www.cell.com/heliyon

Anomalous venoatrial connections - CT and MRI assessment

Antonio Celona^{a,*}, Elio Caruso^b, Silvia Farruggio^b, Lilia Oreto^c, Maria Cristina Inserra^d, Maria Teresa Cannizzaro^d, Tommaso D'Angelo^{e, f}, Silvio Mazziotti^g, David Angel Ortiz^b, Davide Calvaruso^h, Christian Boozⁱ, Salvatore Agati^j, Corrado Di Mambro^j, Giambattista Privitera^k, Giuseppa Fiumanò^k, Placido Romeo^k

^a UOC Radiodiagnostica, San Vincenzo Hospital, Provincial Health Agency of Messina, Taormina, Italy

^b Centro Cardiologico Pediatrico del Mediterraneo (CCPM), San Vincenzo Hospital, Provincial Health Agency of Messina, Taormina, Italy

^c Dipartimento di Medicina Clinica e Sperimentale, Università di Messina, Italy

^d Radiologia 2, CAST, AOU Policlinico "G. Rodolico" San Marco, Catania, Italy

e Diagnostic and Interventional Radiology Unit, BIOMORF Department, University Hospital Policlinico "G. Martino", Messina, Italy

f Department of Radiology and Nuclear Medicine, Erasmus MC, Rotterdam, the Netherlands

g Diagnostic and Interventional Radiology Unit, BIOMORF Department, University Hospital "Policlinico G. Martino", Messina, Italy

h University Hospital Centre, Toulouse, France

¹ Division of Experimental Imaging, Department of Diagnostic and Interventional Radiology, University Hospital Frankfurt, Theodor-Stern-Kai 7,

60590, Frankfurt am Main, Germany

^j Bambino Gesù Pediatric Hospital, Rome, Italy

^k UOC Radiologia San Marco, AOU Policlinico "G. Rodolico" San Marco, Catania, Italy

ARTICLE INFO

CelPress

Keywords: Anomalous venous atrial connections Computed Tomography Magnetic resonance

ABSTRACT

Abnormal venous atrial (VA) connections present a congenital heart disease (CHD) challenge for pediatric cardiologists. Fully anatomical evaluation is very difficult in prenatal and perinatal follow-up, but it has a profound impact on surgical correction and outcome. The echocardiogram is first-line imaging and represents the gold standard tool for simple abnormal VA connection. CT and MRI are mandatory for more complex heart disease and "nightmare cases". 3D post-processing of volumetric CT and MRI acquisition helps to clarify anatomical relationships and allows for the creation of 3D printing models that can become crucial in customizing surgical strategy.

Our article describes a ten-year (2013–2022) tertiary referral CHD center of abnormal AV connections investigated with CT and MRI, illustrating most of these complex diseases with the help of volume rendering (VR) or multiplanar reconstructions (MPR). The nightmarish cases will also be addressed due to the complex cardiovascular arrangement that requires a challenging surgical solution for correction along with the post-surgical complications.

1. Introduction

Abnormal venous atrial (VA) connections pose a congenital heart disease (CHD) challenge to the pediatric cardiologists. Fully

* Corresponding author. *E-mail address:* antonio.celona@alice.it (A. Celona).

https://doi.org/10.1016/j.heliyon.2023.e18462

Received 4 January 2023; Received in revised form 16 July 2023; Accepted 18 July 2023

Available online 27 July 2023

^{2405-8440/© 2023} Published by Elsevier Ltd. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

anatomical evaluation is very difficult in prenatal and perinatal follow-up, but it has a deep impact on surgical correction and outcome. The echocardiogram is first-line imaging and represents the gold standard tool for simple abnormal VA connection. CT and MRI are mandatory for more complex heart disease and "nightmare cases". 3D post-processing of volumetric CT and MRI acquisition helps clarify anatomical relationships and allows for the fabrication of 3D printing models that can become crucial in customizing surgical strategy [1,2].

2. Materials and methods

Our article describes a ten-year (2013–2022) tertiary referral CHD center of abnormal VA connections investigated by means of CT and MRI, with the aim to comprehensively illustrate most of these complex diseases with volume rendering (VR) or multiplanar reconstructions (MPR).

Cases with complex cardiovascular anatomy will also be discussed showing some challenging surgical solutions for correction along with the most common post-surgical complications.

All patients involved in the present study gave their consensus for the publication of anonymized clinical data and radiological images.

Institutional Review Board approval was waived due to the retrospective nature of this observational study.

3. Tecnique

Contrast-enhanced chest CT scans (25 patients over a 10 years period) were performed with a 64-detector row scanner (Philips Ingenuity 64; Philips Medical System; The Netherlands); where required, in consideration of the age or conditions of the young patients, sometimes suffering from even serious cognitive deficits, the tests were performed with the help of anesthetist-resuscitator or pediatric intensivist staff who provided for a mild pharmacological sedation of the patients (intravenous administration of propofol or midazolam).

Only contrast enhanced images were acquired instantly after bolus administration of iodinated contrast medium using a dual-head automatic injector; contrast medium with a high iodine concentration (370–400 mgI/mL) was used, administered at a dose of 1.5 mL/kg of body weight, at a flow rate of 1.8 mL/s, followed by a bolus of 12 mL of saline; the contrast medium was administered through a peripheral venous access (24 Gauge catheter for newborns, 22 Gauge for older children) placed in the antecubital area or forearm, rather on the right, with the aim of reducing streak artifacts related to the passage of high-density blood contrast medium in the left brachiocephalic venous trunk which compromises the visualization of the aortic arch; in infants and very young children, the venous access was placed in the lower limbs to avoid streak artifacts related to the "stagnation" of the contrast medium in the superior vena cava which could affect the quality of the image in suspected anomalous venous return of the right lung.

The images were acquired after a "fixed" delay time; two series of images were acquired, the first of which after a delay of about 13–15 s for adequate opacification of the arterial side of the circulation and the second after a delay of about 35 s for optimal opacification of the venous side.

Images were acquired as follows: slice thickness 0.625 mm, reconstruction slice thickness 1 mm, increment 0.50, pitch 1.2, tube voltage 80 kV, tube current 150–200 mAs.

Images were reconstructed using MPR (slice thickness 1 mm; images reformatted in para-axial, sagittal and coronal oblique plans), MIP (slice thickness 5 mm; images reformatted in para-axial, sagittal and coronal oblique plans) and VRT algorithms on an accessory console [3,4].

A small number of patients (only 5) underwent MRI scanning performed on a 1.5T scanner (Aera; Siemens Healthcare, Forcheim, Germany); despite a better safety profile due to the lack of use of ionizing radiation and iodinated contrast medium, the prolonged time necessary to perform an MRI makes the use of this instrument difficult in the pediatric population, in consideration of the absolute need for deep sedation and assistance anesthesiology of the young patient (the anesthetist/resuscitator present in the magnet room, on the indication of the radiology technician or the automatic voice of the equipment, induces apnea by momentarily interrupting the patient's ventilation).

There are undoubtedly many advantages offered by MRI, which can provide very detailed information on wall kinetics and ventricular volumes as well as on vascular flows through the use of the Steady State Free Precession (b-ssfp) and Phase Contrast (PC) techniques respectively.

MR angiography can be performed using the "white blood" and "black blood" techniques, without the intravenous administration of contrast medium; in the first case, a free-breathing 3D volumetric stack (3D Whole Heart Imaging) is acquired, triggered by the patient's cardiac and respiratory activity; the second group includes the 3DT1 acquisitions, also synchronized with the ECG tracing and the respiratory activity of the young patient.

Contrast-enhanced magnetic resonance angiography (CE-MRA) is performed using 3D Gradient (3D GRE) techniques that allows for heavily T1-weighted acquisitions of an entire volume in a single apnea following intravenous administration of a bolus of contrast medium followed by a bolus of saline; the dose of contrast medium administered is 0.1 mL/kg of body weight at a flow rate of 1.5 mL/s. MR fluoroscopy is used to synchronize the acquisition of images with the transit of the contrast medium: after the intravenous administration of the contrast medium, rapid images are acquired with the 2D GRE technique relating to a specific anatomical region of reference (generally a coronal including superior cava) and the start of the acquisition is activated manually by the operator upon visualization of the transit of the contrast medium [5,6].

The images obtained are subsequently reprocessed using different reconstruction algorithms: MIP (slice thickness 5 mm; images

Table 1

Types of systemic venous connection anomalies.

Systemic Veins	Anomaly
SVC	Persistent left superior vena cava (LSVC)
	Drainage of SVC in left atrium with a sinus venosus defect
	Atresia of right SVC
	Absent right SVC with systemic vein drainage through left persistent SVC in RA via enlarged CS or directly
IVC	Interruption of IVC with azygos continuation (AC). This variant is usually associated with left isomerism. Rare with usual
	arrangement. AC drains most of the time in SVC, while direct hepatic vein drainage in RA may be associated
	Direct connection to the left atrium is a rare but possible variant. When described they are associated with a venous sinus
	defect
	Bilateral IVC
	Absent right IVC, left IVC to RA
	Retro-aortic innominate vein
CS	Fenestration
	Unroofing
	Atresia with a persistent left superior vena cava (LSVC) that remains patent to drain blood from the CS to the right SVC
Totally anomalous systemic venous	Includes IVC, SVC and CS and should be differentiated from left isomerism
drainage	
Persistent superior right caval vein into	Without a drainage into CS
left atrium	



Fig. 1. Contrast-enhanced computed tomography para-axial (a) and para-coronal (b) MPR reconstruction of a duplicated SVC in a patient with dextrocardia in situs solitus. RSVC = Right superior vena cava; LSVC = Left superior vena cava; RA = Right atrium; CS = Coronary sinus.

reformatted in para-axial, sagittal and coronal oblique plans), MPR (slice thickness 1 mm; images reformatted in para-axial, sagittal and coronal oblique plans) or Volume Rendering.

4. Embriology

The cardiovascular anatomy of the adult is strongly dependent on the development of the embryonic cardiovascular system. The primitive heart develops from cardiogenic mesoderm while the vascular supply originates from both mesoderm, yolk sac and placenta. The rapidly growing heart disc forms a bulge initially consisting of two paired tubes within the pericardial cavity until it reaches its final position within the thoracic cavity [7]. After union along the midline, a single heart tube originated from, and after "ballooning" and "looping" have occurred, the four-chambered division of the heart takes place.

Fetal development of systemic venous returns (SVR) consists of a dorsal systemic network and a double nutritional network, the vitelline veins, which carry extraembryonic blood from the yolk sac, and the umbilical-allantoic system, which carries oxygenated blood from the placenta. Initially these are symmetrically paired but, after cross and transverse anastomoses, they transform into single major trunks on the right side, while the left side vessels regress to obliteration [8].

The pulmonary veins initially develop from the dorsal mesocardium as outpouching of the left atrium and subsequently channelling into the mediastinum reach the primitive pulmonary venous system from the developing lung. After its initial appearance as a single midline venous channel, it remodels to give separate right and left orifices in the posterior wall of the left atrium. Subsequently, reaching their final position in the roof of the atrial wall, both the left and right orifices separate into the orifices of the superior and



Fig. 2. Contrast-enhanced computed tomography para-coronal MPR (a) and VR reconstruction (b) of right SVC interruption and drainage of blood via a persistent left persistent SVC in a patient with left isomerism, destrocardia and situs viscerum inversus. RSVC = Right superior vena cava; LSCV = Left superior vena cava; BCT = Brachio-cefalic trunk, RA = Right atrium; RV= Right ventriculum.

Table 2		
Types of pulmonary v	venous connection	anomalies.

Pulmonary veins	Anomaly	
Totally Anomalous Pulmonary	I Supracardiac	The pulmonary veins drain into the innominate vein or azygos vein or right or left superior vena cava
venous connections	II	PVs drain directly into the coronary sinus or right atrium directly
	Cardiac III Infracardiac	The PVs form the vertical vein that drains caudally into the portal vein or ductus venosus, hepatic vein, or
	INFACATALAC IV	A combination of connections at different levels
Partially Anomalous Pulmonary Venous Connections Levo-atrial cardinal vein	Mixea	One or more veins, but not all, do not drain into the left atrium. It could be unilateral if only one lung has bad connections Better defined as an anomalous to systemic venous collateral channel providing overflow from LA to systemic return in case of mitral atresia/stenosis with occluded oval fossa

Table 3

Prevalence of the most common types of systemic venous connection anomalies.

Systemic Veins	Anomaly (prevalence)
SVC IVC	Persistent left superior vena cava (LSVC) (<0,5%) Interruption of IVC with azygos continuation (AC). This variant is usually associated with left isomerism. Rare with usual arrangement. AC drains most of the time in SVC, while direct hepatic vein drainage in RA may be associated (0,6%) Bilateral IVC (0,2–0,5%) Retro-aortic innominate vein (0,55%)

inferior pulmonary veins [9].

In normal anatomy, systemic venous returns, from the lower body via the inferior vena cava (IVC), from the upper body via the superior vena cava (SVC), and from the coronary venous system via the coronary sinus (CS), are directed to the right atrium (RA). Conversely, the right and left pulmonary veins flow into the left atrium (LA).

Abnormalities in embryonic development can lead to alterations of normal anatomical connections. Abnormal pulmonary atrial connections could represent an emergency for the pediatric population while anomalous systemic VA returns are instead in most cases simple CHD, but sometimes they can also lead to blood desaturations requiring interventional or surgical treatment.

Abnormalities of the pulmonary and systemic atrial connection can be found in both the "situs solitus" or "mirror-image" phenotypes and in the heterotaxic syndrome [8,10]. It must be said that semantically it is possible to define regularly connected systemic veins only if the recipient cavity morphologically presents a left atrial appendage. Conversely, the pulmonary veins may be addressed as abnormally connected if a morphological right atrial appendage is present. In the event that the pulmonary veins drain elsewhere



Fig. 3. Contrast-enhanced computed tomography para-sagittal MPR (a) and VR reconstruction (b) of azygos continuation helping drainage of inferior body venous circle in a patient with complex congenital heart disease and absence of IVC. SVC = Superior vena cava; HV = Hepatic vein; PV = Portal vein; AAz = Azygos arch, AC = Azygos continuation.

Fig. 4. - Contrast-enhanced computed tomography para-sagittal MPR (a) and VR reconstruction (b) of anomalous left superior pulmonary veins drainage in left brachiocephalic trunk. LV = Left ventricle; RV = Right ventricle; PA = Pulmonary artery; LSPV = Left superior pulmonary vein; AO = Aorta; LBCT = Left brachio-cephalic trunk; RBCT = Right brachio-cephalic trunk.

than the atrial cavity it is better to define it as abnormal drainage. Completely abnormal systemic venous connections are possible only in the presence of a usual or mirror arrangement. Left isomerism with systemic veins draining into a left atrium is best addressed as abnormal systemic venous drainage in the context of left isomerism.

5. Results - SVR anomalies

SVR abnormalities (Table 1), usually asymptomatic, are often incidental diagnoses. Typically, venous blood from the upper half of the body is carried by the right superior vena cava (SVC) resulting from the confluence of the brachiocephalic veins. Venous blood from

Fig. 5. - Contrast-enhanced computed tomography para-sagittal MPR (a e c) and VR reconstruction (b) of anomalous right pulmonary veins drainage in SVC in a case of Unilateral Anomalous Pulmonary Venous Connection. SVC = Superior vena cava; RA = Right atrium; RV = Right ventricle; PA = Pulmonary artery; RSPV = Right superior pulmonary vein; RIPV = Right inferior pulmonary vein; LBCT = Left brachio-cephalic trunk; RBCT = Right brachio-cephalic trunk.

the lower half commonly drains through the right inferior vena cava (IVC), formed by the confluence of the common iliac veins. The azygos-emiazygos (Azy-HAzy) venous system, which travels on either side of the spine, can connect these two systems and provide an alternate pathway to the right atrium when SVC or IVC is disrupted. The persistence or regression of segments of the primitive embryological venous network can lead to variations in the joint anatomy.

The most common congenital anomaly encountered is persistent left SVC, in most cases connected to CS, with double drainage of the upper body in RA (Fig. 1a and b) [10]; less frequent duplication of IVC, abnormal drainage of brachiocephalic or SVC, absence or interruption of SVC (Fig. 2a and b), azygos continuation (AC) of IVC, and left IVC. Persistent left SVC is an incidental finding, representing <0.5% of the general population (Table 3), and occurs more frequently in more complex CHDs, where the incidence can be as high as 4%. The prevalence of duplicated ICV is about 0.2–3%, while left-sided IVC reaches a prevalence of about 0.2–0.5%. The

Fig. 6. - Contrast-enhanced computed tomography para-coronal (a), para-sagittal (c) MPR and VR (b) reconstruction of type I TAPVC draining in SVC via a vertical vein in a complex CHD with dextrocardia in situs inversus. SVC = Superior vena cava; AO = Aorta; VV = Vertical vein; RSPV = Right superior pulmonary vein; RIPV = Right inferior pulmonary vein; LSPV = Left superior pulmonary vein; LIPV = Left inferior pulmonary vein.

prevalence of infrarenal ICV interruption is about 0.6%, but can reach 1.3% when there is concomitant complex CHD or heterotaxy syndrome (Fig. 3a and b) [8].

No statistically significant difference was found in the gender distribution of these anomalies.

Direct connection of the IVC to the left atrium is also a possibility, but usually associated with a venous sinus defect. Absent right IVC with a left IVC directed to the RA is described but usually associated with heterotaxis. Retroaortic innominate vein associated with other congenital heart defects, most commonly tetralogy of Fallot and right aortic arch, has been found to have a prevalence of approximately 0.55% [11]. Some CS malformations, such as "unroofing" and "fenestration", are not readily evaluable with CT and MRI and are primarily up to echocardiography to demonstrate them. Coronary sinus atresia with persistent left SVC to drain coronary blood into the right SVC is also described. This represents a benign anomaly without physiological consequences, but its recognition is of vital importance for the cardiac surgeon because it can potentially lead to myocardial ischemia and necrosis [12]. Totally anomalous SVR is described as a syndrome in its own right or associated with left isomerism.

6. Results - PVR anomalies

Anomalous pulmonary venous connections (APVC) are abnormalities due to failure of channeling of the pulmonary venous channel into the mediastinum.

APVC are divided into total anomalous pulmonary venous connection (TAPVC) if all four veins drain into the systemic venous system and partial anomalous pulmonary venous connection (PAPVC) if only one or more veins, but not all, do not drain into the left

Fig. 7. Contrast-enhanced computed tomography 3D VR (a), para-axial (b) and para-sagittal (c) MPR reconstruction of type II TAPVC draining in RA via an enlarged CS in a complex CHD. CS = Coronary sinus; LV = Left ventricle; RV = Right ventricle; RSPV = Right superior pulmonary vein; RIPV = Right inferior pulmonary vein; LSPV = Left superior pulmonary vein; LIPV = Left inferior pulmonary vein; RPVV = Right pulmonary vein; LPVV = Left pulmonary veins.

atrium (Fig. 4a and b). Unilateral anomalous pulmonary venous connection (UAPVC) is a condition of PAPVC in which only one lung has bad connections (Fig. 5a, b, c).

TAPVC characterized by an asymmetric ("3 + 1") connection in which 3 of the pulmonary veins drain to a common site through a confluence and the remaining pulmonary vein drains to a separate site are termed mixed TAPVC.

Bilateral symmetrical connections with separate drainage of veins from the right and left lung ("2 + 2") are also frequently described [13].

The Darling classification divides TAPVC into four categories [14] according to the sites where the anomalous connection occurs (Table 2). Type I, supracardiac (Fig. 6a, b, c), represents the most common phenotype, 45–55% of TAPVC. Type II, cardiac (Fig. 7a, b, c), accounts for about 20–30% of TAPVC. Type III, infracardiac (Fig. 8a and b) achieves about 13–25% of TAPVC. Type IV, described as mixed type, has an incidence of approximately 10% of TAPVC (Table 4) [15].

The incidence of TAPVC ranges from 0.6 to 1.2 per 10,000 live births. Among newborns with congenital heart disease, the incidence of TAPVC ranges between 0.7 and 1.5% [16,17].

The overall incidence of PAPVC is estimated to be 0.7% of the population [18]. However, as this rate is based upon autopsy data, the true prevalence of PAPVC may actually be higher [19–22]. Multiple case series report PAPVC as an incidental finding without associated symptoms. Although PAPVR can present as an isolated structural abnormality, it commonly occurs with other cardiac abnormalities, most often an atrial septal defect (ASD). In addition, patients with Turner syndrome are at increased risk for PAPVC [23, 24].

They have a prevalence of approximately 2.5% of all CHDs at birth (seven tenths are PAPVR) - (5) and the lowest prenatal diagnosis rate, approximately 9% of all fetal CHD diagnoses [25].

No statistically significant difference was found in the gender distribution of these anomalies.

From a diagnostic point of view, in the case of APVC, the consideration of the presence of pulmonary venous obstruction (PVO) is mandatory due to the conditioning of the surgical timing.

A particular case of left to right shunt is the persistence of the so-called "Levo-atrial cardinal vein" which should be better defined as

Fig. 8. Contrast-enhanced computed tomography para-sagittal MPR (a) and VR reconstruction (b) of type III TAPVC draining in portal vein via an duct crossing diaphragm in a complex CHD. CS = Coronary sinus; HV= Hepatic vein; RA = Right atrium; RMHV = Right medium hepatic vein; TAPVC = Total anomalous pulmonary venous connection duct; LSPV = Left superior pulmonary vein; LIPV = Left inferior pulmonary vein; RPPVV = Right pulmonary veins; PV = Portal vein.

Table 4	ŀ
---------	---

Frequency of the four types of TAPVC.

Pulmonary veins	Anomaly (frequency)		
Totally Anomalous Pulmonary Venous Connections	I Supracardiac II Cardiac III Infracardiac IV	The pulmonary veins drain into the innominate vein or azygos vein or right or left superior vena cava (SVC) (45–55%) PVs drain directly into the coronary sinus or right atrium directly (20–30%) The PVs form the vertical vein that drains caudally into the portal vein or ductus venosus, hepatic vein, or inferior vena cava (IVC) (13–25%) A combination of connections at different levels (10%)	
	Mixed		

pulmonary-to-systemic venous collateral channel that provides overflow from the LA to the systemic return in case of mitral atresia/ stenosis with oval fossa occluded (Fig. 9a, b).

7. Treatment

Surgical correction of the anomalous VA connection is often demanding and is not free from post-surgical complications such as stenosis (Fig. 10) or dilatation of the anastomoses (Fig. 11) which require reoperation. Interventional radiological procedures are sometimes problem-solving and metallic stents may be implanted to salvage the anastomosis (Fig. 12).

Furthermore, the correction of very complex CHDs such as in isomeric patients could be a nightmare for surgeons who are faced with devastated anatomy and have to plan the surgery step by step. As an example, we report a case of type I TAPVC draining into a left SVC, associated with a single atrium, a double outlet right ventricle, transposition of the great arteries (anterior aorta and posterior allocation of the pulmonary artery and patent ductus arteriosus in a right isomeric patient with dextrocardia and situs viscerum inversus) (Figs. 6–12). In reality, the definitive treatment of TAPVC from a surgical point of view must be undertaken urgently only in the event that they are obstructed. In unobstructed TAPVC there is no immediate urgency and elective surgery can be planned before six months of age. Patients with minor degrees of obstruction usually have a degree of right heart failure and should be repaired as a

Fig. 9. Cardiac Magnetic Resonance para-coronal MIP (a) and VR reconstruction (b) of a patient with a levo-cardinal vein connecting left atrium to right superior vena cava with a left to right shunt. RV = Right ventricle; RA = Right atrium; SVC = Superior vena cava; LCV = Levo-cardinal vein; RBCT = Right brachio-cephalic trunk LBCT = Left brachio-cephalic trunk; LIPV = Left inferior pulmonary vein; RIPV = Right inferior pulmonary vein; AO = Aorta; PA = Pulmonary artery.

Fig. 10. - Contrast-enhanced computed tomography VR reconstruction after surgical repair of a Type I TAPVC. A stenosis at anastomotic site of right pulmonary veins is seen. RPPVV = Right pulmonary veins; LPPVV = Left pulmonary Veins; PA = Pulmonary artery; Black Arrow = Anastomotic stenosis.

priority.

Surgical repair involves anastomosing the pulmonary venous confluence into the left atrium to achieve proper drainage. In infraand supracardiac TAPVC redirection can be achieved by opening the confluence sitting behind the pericardium and making a corresponding incision in the posterior wall of the left atrial appendage, anastomosing the two together. The draining vein may be ligated or left open and the atrial septal defect or patent foramen ovale closed. In cardiac TAPVC the atrial septum may be partially resected, the coronary sinus exposed in the left atrium, and a patch used to close the atrial septum, engaging the return of the coronary sinus into the left atrium.

The associated complex cardiac abnormalities often need to be addressed in stages and require careful surgical planning and timing that is highly dependent on the native cardiovascular anatomy.

Fig. 11. Contrast-enhanced computed tomography para-axial MPR reconstruction after surgical repair of PAPVC with superior right pulmonary vein draining into SVC. Anastomotic site look to be enlarged. RSPV = Right superior pulmonary vein; DAT = Dilated anastomotic tract; RPA = Right pulmonary artery; SVC = Superior vena cava; LA = Left atrium; LV = Left ventriculum; RV = Right ventriculum.

Fig. 12. Contrast-enhanced computed tomography paracoronal MPR reconstruction after surgical redirection of superior vena cava into right atrium. Stenosis of surgical anastomoses treated with a metallic stent; SVC = Superior vena cava; RA = Right atrium RV= Right ventriculum.

8. Conclusions

Abnormal systemic venous or pulmonary venous returns are not uncommon congenital anomalies that must be fully clarified before treatment. 3D CT and MRI can add value to the echocardiogram particularly in complex cases. We hope that this document will help the radiologist to better understand these pathological findings by avoiding pitfalls and properly evaluating the findings and their implications for invasive diagnostic or therapeutic procedures during reporting [26].

Funding statement

This research did not receive any specific grant from funding agencies in the public, commercial or not-for-profit sector.

Declarations

Patients whose data or images are included in the publication have consented for all images and clinical data and other data

included in the manuscript.

APC waived - waiver code: Heliyon - Contractual payment discounts.

Author contribution statement

Antonio Celona: Conceived and designed the experiments; Performed the experiments; Wrote the paper.

Elio Caruso, Silvia Farruggio: Conceived and designed the experiments; Wrote the paper.

Lilia Oreto, Maria Teresa Cannizzaro and Tommaso D'Angelo: Analyzed and interpreted the data; Contributed reagents, materials, analysis tools or data.

Maria Cristina Inserra: Performed the experiments; Analyzed and interpreted the data; Contributed reagents, materials, analysis tools or data.

David Angel Ortiz, Davide Calvaruso, Christian Booz, Salvatore Agati, Corrado Di Mambro, Giambattista Privitera, Silvio Mazziotti and Giuseppa Fiumanò: Analyzed and interpreted the data.

Placido Romeo: Conceived and designed the experiments; Performed the experiments; Wrote the paper.

Data availability statement

Data will be made available on request.

Additional information

No additional information is available for this paper.

Declaration of competing interest

The authors declare the following financial interests/personal relationships which may be considered as potential competing interests:One of the authors (Tommaso D'Angelo) is editor of the journal.

References

- [1] K. Hadeed, P. Acar, Y. Dulac, F. Cuttone, X. Alacoque, C. Karsenty, Cardiac 3D printing for better understanding of congenital heart disease, Arch. Cardiovasc. Dis. 111 (1) (2018) 1-4, https://doi.org/10.1016/j.acvd.2017.10.001.
- E. Caruso, S. Farruggio, A. Secinaro, S. Agati, D. Calvaruso, C. Di Mambro, L. Borro, A. Celona, D. Petruccelli, P. Romeo, Impact of computerized tomography scan and three-dimensional printing model for surgical procedure in complex congenital heart disease, J. Radiol. Rev. 8 (1) (2021) 1-3, https://doi.org/ 10.23736/S2723-9284.20.00093-7
- [3] L.S. Alizadeh, T.J. Vogl, S.S. Waldeck, D. Overhoff, T. D'Angelo, S.S. Martin, I. Yel, L.D. Gruenewald, V. Koch, F. Fulisch, C. Booz, Dual-Energy CT in cardiothoracic imaging: current developments, Diagnostics 13 (12) (2023) 2116, https://doi.org/10.3390/diagnostics13122116. PMID: 37371011; PMCID: PMC10297493.
- L. Lenga, M.H. Albrecht, A.E. Othman, S.S. Martin, D. Leithner, T. D'Angelo, C. Arendt, J.E. Scholtz, C.N. De Cecco, U.J. Schoepf, T.J. Vogl, J.L. Wichmann, [4] Monoenergetic dual-energy computed tomographic imaging: cardiothoracic applications, J. Thorac. Imag. 32 (3) (2017) 151–158, https://doi.org/10.1097/ RTI.00000000000259. PMID: 28198752.
- [5] L.R.M. Lanzafame, G.M. Bucolo, G. Muscogiuri, S. Sironi, M. Gaeta, G. Ascenti, C. Booz, T.J. Vogl, A. Blandino, S. Mazziotti, T. D'Angelo, Artificial intelligence in cardiovascular CT and MR imaging, Life 13 (2) (2023) 507, https://doi.org/10.3390/life13020507. PMID: 36836864; PMCID: PMC9968221.
- I. Mia, M. Le, C. Arendt, D. Brand, S. Bremekamp, T. D'Angelo, V.O. Puntmann, E. Nagel, Quantitative perfusion-CMR is significantly influenced by the [6] placement of the arterial input function, Int. J. Cardiovasc. Imag. 37 (3) (2021) 1023-1031, https://doi.org/10.1007/s10554-020-02049-3. Epub 2020 Oct 12. PMID: 33047177: PMCID: PMC7969553.
- [7] P. Stein, Total anomalous pulmonary venous connection, AORN J. 85 (3) (2007) 509-520, https://doi.org/10.1016/S0001-2092(07)60123-9.
- J.D. Oliveira, I. Martins, Congenital systemic venous return anomalies to the right atrium review, Insights Imaging 10 (1) (2019) 115, https://doi.org/10.1186/ [8] s13244-019-0802-v
- [9] R.H. Anderson, K. Krishna, K.A. Mussato, A. Redington, J.S. Tweddell, J. Tretter, et al., Anderson's pediatric cardiology, 4th edition [internet]. Elsevier [citato 6 febbraio 2021]. Disponibile su: https://www.us.elsevierhealth.com/andersons-pediatric-cardiology-9780702076084.html, 2019.
- [10] J.P. Jacobs, R.H. Anderson, P.M. Weinberg, H.L. Walters 3rd, C.I. Tchervenkov, D. Del Duca, R.C.G. Franklin, V. D'Aiello, M.J. Beland, S.D. Colan, J.W. Gaynor, O.N. Krogmann, H. Kurosawa, B. Maruszewski, G. Stellin, M. Elliot, The nomenclature, definition and classification of cardiac structures in the setting of heterotaxy, Cardiol. Young 17 (Suppl 2) (2007) 1-28, https://doi.org/10.1017/S1047951107001138.
- [11] K.W. Tyrak, M.K. Holda, M. Koziej, K. Piątek, W. Klimek-Piotrowska, Persistent left superior vena cava, Cardiovasc. J. Afr. 28 (3) (2017) e1-e4, https://doi.org/ 0.5830/CVJA-2016-084
- [12] S. Kulkarni, S. Jain, P. Kasar, S. Garekar, S. Joshi, Retroaortic left innominate vein incidence, association with congenital heart defects, embryology, and clinical significance, Ann. Pediatr. Cardiol. 1 (2) (2008) 139-141, https://doi.org/10.4103/0974-2069.43881.
- R. Santoscoy, H.L. Walters 3rd, R.D. Ross, J.M. Lyons, M. Hakimi, Coronary sinus ostial atresia with persistent left superior vena cava, Ann. Thorac. Surg. 61 (3) (1996) 879-882, https://doi.org/10.1016/0003-4975(95)01137-4.
- [14] F.J. Ross, D. Joffe, G.J. Latham, Perioperative and anesthetic considerations in total anomalous pulmonary venous connection, Semin. CardioThorac. Vasc. Anesth. 21 (2) (2017) 138–144, https://doi.org/10.1177/1089253216672012.
- [15] J.M. Craig, R.C. Darling, W.B. Rothney, Total pulmonary venous drainage into the right side of the heart; report of 17 autopsied cases not associated with other major cardiovascular anomalies, Lab. Invest. 6 (1) (1957) 44–64.
- [16] M.D. Reller, M.J. Strickland, T. Riehle-Colarusso, et al., Prevalence of congenital heart defects in metropolitan Atlanta, 1998-2005, J. Pediatr. 153 (2008) 807, https://doi.org/10.1016/j.jpeds.2008.05.059.
- [17] J.I. Hoffman, S. Kaplan, The incidence of congenital heart disease, J. Am. Coll. Cardiol. 39 (2002) 1890, https://doi.org/10.1016/s0735-1097(02)01886-7. [18] J.E. Healey Jr., An anatomic survey of anomalous pulmonary veins: their clinical significance, J. Thorac. Surg. 23 (1952) 433.

- [19] C. Garduno, S. Chew, J. Forbess, et al., Persistent left superior vena cava and partial anomalous pulmonary venous connection: incidental diagnosis by transesophageal echocardiography during coronary artery bypass surgery, J. Am. Soc. Echocardiogr. 12 (1999) 682, https://doi.org/10.1053/je.1999.v12. a98795.
- [20] K. Miwa, S. Takamori, A. Hayashi, et al., Incidental partial anomalous pulmonary venous connection in left lung cancer, Jpn. J. Thorac. Cardiovasc. Surg. 52 (2004) 189, https://doi.org/10.1007/s11748-004-0105-0.
- [21] H. Sasaki, N. Naka, N. Kitahara, et al., Right partial anomalous pulmonary venous connection found during lobectomy for coexisting lung cancer and tuberculosis: report of a case, Clin. Lung Cancer 7 (2006) 350, https://doi.org/10.3816/clc.2006.n.018.
- [22] G. Sobrinho, J. Salcher, Partial anomalous pulmonary vein drainage of the left lower lobe: incidental diagnostic after central venous cannulation, Crit. Care Med. 31 (2003) 1271, https://doi.org/10.1097/01.CCM.0000059648.07978 (EC).
- [23] S.M. Bechtold, R. Dalla Pozza, A. Becker, et al., Partial anomalous pulmonary vein connection: an underestimated cardiovascular defect in Ullrich-Turner syndrome, Eur. J. Pediatr. 163 (2004) 158, https://doi.org/10.1007/s00431-003-1384-7.
- [24] V.B. Ho, V.K. Bakalov, M. Cooley, et al., Major vascular anomalies in Turner syndrome: prevalence and magnetic resonance angiographic features, Circulation 110 (2004) 1694, https://doi.org/10.1161/01.CIR.0000142290.35842. B0.
- [25] C.-C. Kao, C.-C. Hsieh, P.-J. Cheng, C.-H. Chiang, S.-Y. Huang, Total anomalous pulmonary venous connection: from embryology to a prenatal ultrasound diagnostic update, J. Med. Ultrasound 25 (3) (2017) 130–137, https://doi.org/10.1016/j.jmu.2017.08.002.
- [26] V. De Matteo, M.D. Quartermain, M. Di Lorenzo, M. Liu, C. Tinker, M.S. Cohen, Total anomalous pulmonary venous return with no connection, CASE 2 (3) (2018) 85–88, https://doi.org/10.1016/j.case.2017.11.001.