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CASE REPORT Open Access

Case report of coexisting transposition of the great arteries with supracardiac total anomalous pulmonary venous connection: focus on computed tomography features

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Abstract

Background The coexisting of transposition of the great arteries (TGA) with total anomalous pulmonary venous connection (TAPVC) is one of the rare anomalies. The incidence of coexisting TAPVC and TGA is unknown with very few cases ever reported.

Case presentation We reported a case of a 13-month-old female toddler with history of cyanosis. Echocardiography revealed atrioventricular ambiguity with pulmonary atresia, all PVs drain into the innominate vein via vertical vein (VV), ostium secundum atrial septal defect (ASD) and ventricular septal defect (VSD) were observed. The CT scan confirmed co-occurrence of TGA and TAPVC. All four confluence PVs behind the small left atrium (LA), drains into an ascending lateral large VV, coursing to innominate vein without any PV access into LA. The superior vena cava, right atrium and right ventricle (RV) were dilated. The RV is the origin of the aortic root. The aorta continues on the right side, with an arterial connection to the right pulmonary aberrant artery at the level of the aortic arch. Main pulmonary artery originates from LV and appears atretic with only connection to the left pulmonary artery. Large ASD and VSD were identified.

Conclusions TGA and TAPVC are a rare combination and should be suspected in mild-cyanotic cases with levocardia with situs solitus. CT angiography is one of the modalities of choice to characterize the vasculature anomalies.

Keywords Transposition of the great arteries, Total anomalous pulmonary venous connection, CT angiography, Case report

Background

An anatomical description of transposition of the great arteries (TGA), specifically dextro-TGA or D-TGA with total anomalous pulmonary venous connection (TAPVC), was first published in 1933 [1]. The coexistence of D-TGA and TAPVC is uncommonly described in the scientific literature. With the unknown incidence, only a few publications describing patients who underwent anatomically anomalous diagnostic workups or surgery. The case of coexisting D-TGA and TAPVC in a toddler with cyanotic cardiac disease was described.

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Case presentation

A 13-month-old female toddler was referred to the radiology department for a computed tomography (CT) angiography of the chest to evaluate the cardiac vasculature. The infant was diagnosed with cyanosis within her first week of life. Initially, two-dimensional echocardiography had been performed of which revealed atrioventricular ambiguity with pulmonary artery atresia, all pulmonary veins (PVs) drain into the innominate vein (IV) via vertical vein (VV), ostium secundum atrial septal defect (ASD) and ventricular septal defect (VSD) were observed. Left ventricle (LV) present with EF 83%. Right ventricle (RV) had good contractility with tricuspid annular plane systolic excursion of 1.3 cm.

A non-ECG-gated, free-breathing chest CT angiography was performed using a 128-multislice CT scanner (Revolution™ HD, GE HealthCare Technologies Inc., US), with 100 kVP and 150 mAs after 10.5 mL of non-ionic, water-soluble, iodine contrast material (Iohexol™ 350 mg/mL) was administered. Advantage Workstation™ VolumeShare 7 (GE HealthCare Technologies Inc, US) was used for image processing.

The CT scan detected levocardia with situs solitus. All four PVs are in confluence behind the small left atrium (LA), and their confluence converges into an ascending lateral large VV, coursing to IV without any PV access into LA. TAPVC was confirmed, with no observable PV access into LA. Large atrial septal defect (ASD) and ventricular septal defect (VSD) were identified (Fig. 1). The superior vena cava (SVC), right atrium (RA) and RV were dilated. D-TGA was suggested after it was discovered that the major arteries of both ventricles were reversed. The RV is the origin of the aortic root. The aorta continues on the right side, with an apparent connection to the aberrant right pulmonary artery at the level of the aortic arch via the arterial communication favors to major aortopulmonary collateral arteries (MAPCAs). Main pulmonary artery originates from LV and appears atretic with only connection to the left pulmonary artery. The liver is located in the midline, and no intraabdominal spleen was identified (Fig. 2). Other identified intraabdominal organs were unremarkable. Schematic cardiac findings are featured in Fig. 3.

Until this report was made, the patient underwent routine follow-up for a surgical plan at a national cardiac surgery center elsewhere. She was scheduled for an initial Blalock–Thomas–Taussig shunt surgery to increase blood flow to the pulmonary circulation before undergoing other definitive surgery.

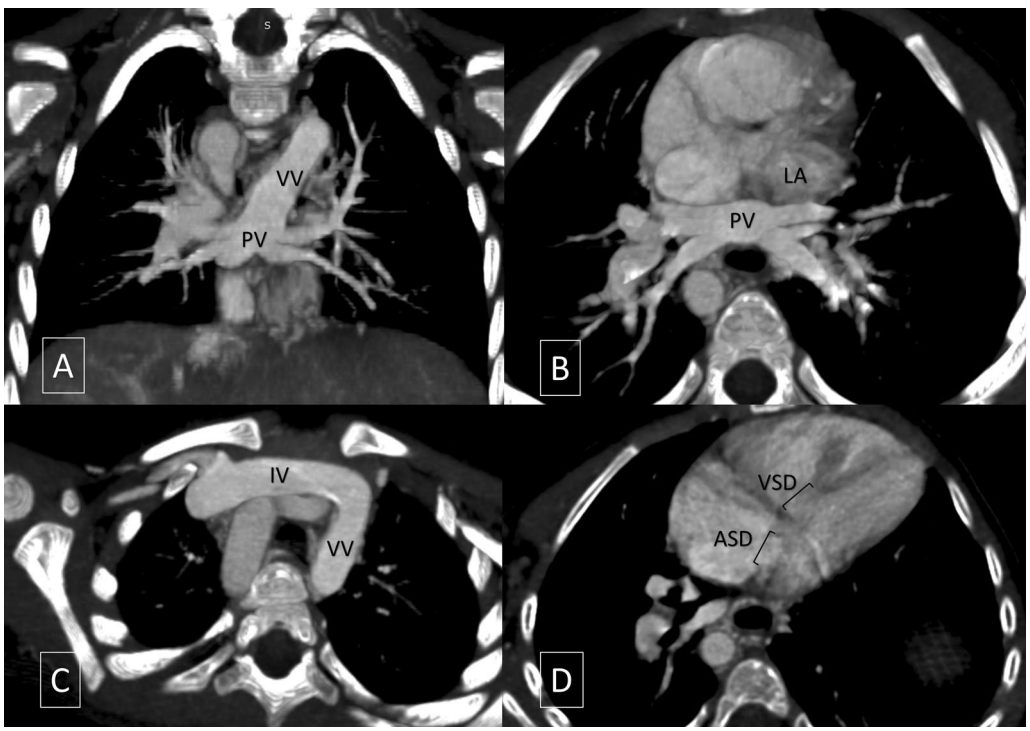

Fig. 1 Axial and coronal section of CT angiography of the cardiac vasculature. All four pulmonary veins (PVs) are in confluence behind the small left atrium (LA), and their confluence converges to an ascending lateral large vertical vein (VV) which drains into innominate vein (IV) (**A**–**C**). The entrance of PV into LA is absent. All the finding favors to supracardiac TAPVC. Large ostium secundum atrial septal defect (ASD) and ventricular septal defect (VSD) were identified (**D**)

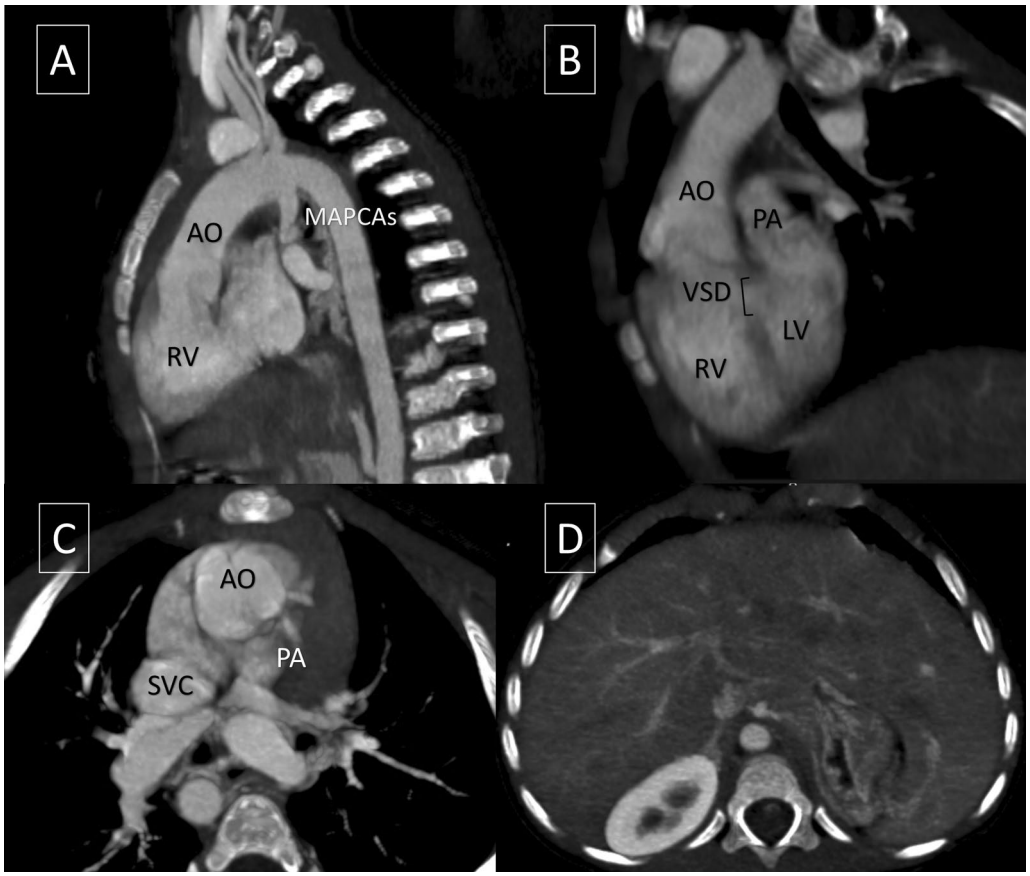

Fig. 2 Sagittal, coronal and axial section of cardiac CT angiography of the cardiac vasculature. Aortic root is originated from the right ventricle (RV). The aorta continues on the right side, with an apparent connection to the right pulmonary aberrant artery at the level of the aortic arch via the major aortopulmonary collateral arteries (MAPCAs) (**A**). Main pulmonary artery originates from left ventricle and appears atretic with only connection to the left pulmonary artery (PA) (**B**, **C**). Aortic valve is located on the anterior right to the pulmonary valve confirming the spatial relationship of semilunar cusps in D-TGA (**C**). The liver is transversal in position and located in the midline, and no spleen was identified, suggesting a possible heterotaxy syndrome, along with the cardiac anomaly (**D**)

Discussion

Epidemiology

Very few cases of D-TGA and TAPVC co-occurrence have ever been documented (Table 1). Adults with coexisting D-TGA and TAPVC are quite uncommon. The majority of these occurrences occurred in newborns, and younger children. The oldest patient documented in medical literature was 22 years old [2]. The incidence of TAPVC is increased in individuals with heterotaxy syndromes, especially asplenia [3], as is the case with our patient.

Presentation

Total anomalous pulmonary venous return (TAPVR), total anomalous pulmonary venous drainage (TAPVD), and total anomalous pulmonary venous connection (TAPVC) are terms that are arbitrarily used and generally regarded synonymous. Hines [22] defines TAPVC as the absence of confluence between the PV and the LA. In contrast, the junction of the pulmonary vein as an entrance to the LA is still maintained and the aberrant drainage is secondary to the presence of some level of left-sided structural atresia in TAPVR.

Consequently, in the isolated TAPVC, all PVs drain abnormally into the systemic venous circulation, rather than the LA. Thus, oxygenated pulmonary blood is rerouted to the right heart and pumped back to the lungs [23]. To gain survival, a portion of the RA's mixed blood is diverted to the left heart via a right-to-left shunt to supply the systemic circulation. This occurs commonly at the atrial level via an ASD, a patent foramen ovale, or less frequently a PDA [23, 24].

In isolated D-TGA, the aorta originates from the RV and is located right and anterior to the PA, which, in the contrary, originates from the LV. Thus, the deoxygenated systemic venous blood goes to the right heart and then to

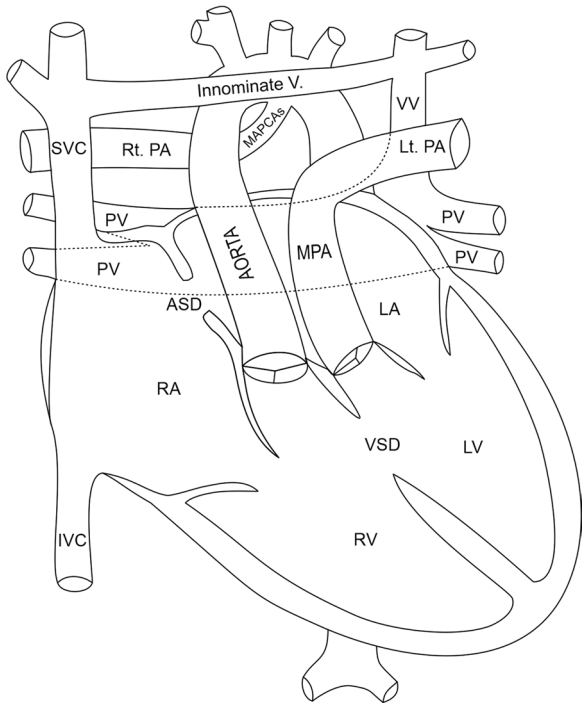

Fig. 3 Schematic of complex cardiac abnormalities of the reported case with the main feature of co-occurrence of D-TGA and supracardiac TAPVC. The right accessory pulmonary artery (Rt-PA) is supplied by arterial communication from the aorta, favoring major aortopulmonary collateral arteries (MAPCAs). Both ASD and VSD are present. *ASD* Atrial septal defect, *IVC* Inferior vena cava, *LA* Left atrium, *Lt-PA* Left pulmonary artery, *LV* Left ventricle, *PV* Pulmonary vein, *RA* Right atrium, *RV* Right ventricle, *SVC* Superior vena cava, *VSD* Ventricular septal defect

the systemic circulation via the aorta. Similar to TAPVC, a connectivity between the left and right sides, such as an ASD, patent foramen ovale, VSD, or PDA, is necessary for survival [25].

Cyanosis is usually present at birth in cases of isolated D-TGA or obstructed TAPVC. Both abnormalities have a high risk of inducing pulmonary arterial hypertension (PAH) at an early age, although through different mechanisms. In d-TGA, the process begins in utero with the preferential channeling of oxygen-rich inferior vena cava blood into the left heart through the VSD, causing dilatation of the pulmonary arteries and strain damage to the vascular intima, followed by secondary vasoconstriction. In obstructed TAPVC, however, the increase in pulmonary pressures is due to pulmonary venous hypertension, the severity of which is proportionate to the degree of obstruction. The coexistence of TGA and TAPVC reduces the hemodynamic impact of each individual anomaly by limiting severe desaturation and pulmonary overflow. Instead of the pulmonary circulation, the oxygen-rich blood that drains into the RA is delivered to the systemic circulation [21]. Possibly as a result of this reciprocal compensation, cyanosis is less noticeable and may go unrecognized at birth, culminating in an undeveloped and depleted LV as the individual ages [17, 18].

As oxygenated blood from the pulmonary veins is directed to the systemic circulation, the coexistence of TAPVC and D-TGA improves systemic saturations. However, it can cause a milder apparent cyanosis in a relatively asymptomatic infant, obscuring the discovery of a significant congenital heart disease [18]. Clinically, we should suspect this coexisting condition in children with a diagnosis of TGA and severe right ventricular enlargement, but who are significantly less cyanotic than the average child with uncomplicated TGA [9].

Diagnostic workup

Prompt diagnosis and surgical strategy are essential to reducing morbidity and mortality in severe cases. Some of the most intricate aspects of TAPVC may not be depicted by echocardiography, the first and safest imaging technique for cardiovascular anomalies [11, 16]. For mapping the pulmonary veins and other vascular and structural anomalous anatomy without the necessity for invasive cardiac catheterization, CT angiography provides a noninvasive and sensitive alternative. Contrast-enhanced magnetic resonance (MR) angiography is a radiation-free alternative to CT angiography, although younger children may require general anesthesia. For morphological identification in TAPVC, electrocardiography-gated CT and phase-contrast cine MR imaging (PC-MRI) are preferred above non-gated CT [26].

Treatment

The treatment strategy for a patient with coexisting D-TGA and TAPVC must be supported by clinical and radiological data indicating preservation of LV mass and the absence of irreversible pulmonary vascular disease despite the presence of PAH. [18]. A few authors reported surgery as the mainstay of treatment for coexisting TGA and TAPVC. Successful results were obtained using modified atrial switching techniques based on the principles of either Senning [2, 6, 10, 18] or Mustard [5, 7–9, 17]. However, in recent years, arterial switch operation (ASO) has become the preferred approach for the TGA.

Few cases have been reported of successful anatomic repair through ASO. The first successful ASO was performed by Lopes et al. on a 22-h-old infant with D-TGA and subdiaphragmatic TAPVC [11], followed by Seliem et al. on a 4-year-old toddler who had d-TGA with supracardiac TAPVC [13], and by Mykychak et al. on TGA and subdiaphragmatic TAPVC [14]. A few number procedures have been performed elsewhere [15, 16, 20]. The

Table 1 Cases of coexisting D-TGA and TAPVC reported in the literature

Author(s)	Year	Age/sex	Country	Imaging modalities	Type of TAPVC	Associated congenital defect	Surgery
Whitaker et al. [4]	1964	9-year-old/Female	UK	Cardiac catheterization and angiography	Supracardiac	ASD	None
Sapsford et al. [5]	1972	16-year-old/Female	UK	Cardiac catheterization and angiography	Supracardiac	None	Atrial (Mustard)
Barbero-Marcial et al. [6]	1984	8-month-old/Male	Brazil	Cardiac catheterization and angiography	Supracardiac	None	Atrial (Senning)
Amodeo et al. [7]	1990	Neonate/Female	Italy	Echocardiography; cardiac catheterization and angiography	Cardiac (right atrium)	ASD, VSD, PDA	Atrial (Mustard)
Thies et al. [8]	1990	3-day-old/Male	Germany	Echocardiography; cardiac catheterization and angiography	Subdiaphragmatic	ASD, VSD, cor triatum	Atrial (Mustard)
Gontijo et al. [9]	1994	11-month-old/Female	Brazil	Echocardiography	Cardiac (coronary sinus)	None	Atrial (Mustard)
Ueda et al. [10]	1994	1-year-old/Male	Japan	Cardiac catheterization and angiography	Subdiaphragmatic	None	Atrial (Senning)
Lopes et al. [11]	2001	22-hour-old/Male and 5-month-old/Male	Brazil	Echocardiography; cardiac catheterization and angiography	Partial subdiaphragmatic and supracardiac	VSD, ASD, single coronary ostium	ASO
Raff et al. [12]	2002	13-year-old/Female	US	Echocardiography	Supracardiac	None	Atrial (Senning)
Seliem et al. [13]	2003	4-year-old/Male	Saudi Arabia	Echocardiography; cardiac CT; cardiac catheterization and angiography	Supracardiac	ASD	ASO
Mykychak et al. [14]	2016	5-hour-old/Male	Ukraine	Echocardiography; cardiac MRI; cardiac catheterization and angiography	Subdiaphragmatic	Single coronary ostium	ASO
Salve et al. [15]	2017	6-day-old/Male	India	Echocardiography	Cardiac (coronary sinus and right atrium)	ASD	ASO
Meliota et al. [16]	2019	6-day-old/Female	Italy	Echocardiography	Subdiaphragmatic	None	ASO
Samaddar et al. [17]	2019	7-month-old/Not available	India	Echocardiography; cardiac CT	Cardiac (coronary sinus)	None	Atrial (Mustard)
Aggarwal et al. [18]	2019	24-day-old/Male	India	Echocardiography	Supracardiac	None	Atrial (Senning)
Scrascia et al. [19]	2020	21-day-old/Female	Italy	Cardiac catheterization and angiography	Subdiaphragmatic	ASD	ASO
Mishra et al. [2]	2020	Four patients (7-month-old, 3-year-old, 7-year-old, 22-year-old)/Not available	India	Echocardiography; cardiac CT	1 patient of cardiac (coronary sinus); 3 patients of supracardiac	VSD	Atrial (modified Senning)
Abah et al. [20]	2021	1.5-year-old/Female	Nigeria	Echocardiography	Supracardiac	ASD	ASO
Chatterjee et al. [21]	2021	18-year-old/Not available	India	Echocardiography; cardiac MRI	Cardiac (right atrium)	ASD	None

similar observations of the late presentation and early regression of the LV serve as limiting factors for ASO.

In D-TGA, the regression of LV mass often begins during the first 2–3 weeks. Beyond this period, the steady decline in PVR deconditions the performance of the LV with a progressive decrease in its muscle mass [18]. In the coexisting D-TGA and TAPVC, only a few have documented variability in the development of LV mass regression, which can occur early [18] or late [20, 21]. Nevertheless, ASO is encouraged within the first 2 to 3 weeks of life. Beyond that period, it may pose the risk of incapability of LV to cope with the acutely increased work of systemic circulation following surgery [9].

LV mass is preserved in TGA patients with a large VSD or PDA, obstruction of LV outflow, unregressed pulmonary vascular resistance (PVR), or a significant aortopulmonary collateral due to the presence of a greater afterload. In cases of extensive interatrial communication alone, the LV mass is often not adequately preserved following a decrease in PVR since there is no afterload and a substantial decrease in LV inflow [18].

A case report by Abah et al. [20] described a satisfactory outcome of a late-treated coexisting TAPVC and TGA, with the advantage of PAH in preserving the LV mass being highlighted. PAH and volume load from the significant right-to-left shunt at the atrial level due to the large ostium secundum ASD provide a pressure stress on the LV without resulting permanent pulmonary vascular disease. The existence of obstructed TAPVC will result in increased PA pressures, which may be beneficial for LV readiness.

Conclusions

Even in the presence of normal cardiac and abdominal situs, TGA can be associated with anomalous pulmonary venous connection. In order to aid clinical decisions and the planning of surgical and catheter-based procedures, it is essential to map the relevant anatomy. For these reasons, understanding of anomalous pulmonary venous drainage and variant anatomy is essential for optimizing the prognosis, particularly in the diagnostically challenging patient.

Abbreviations

ASD	Atrial septal defect
ASO	Arterial switch operation
CT	Computed tomography
D-TGA	Dextro-transposition of the great arteries
LA	Left atrium
LV	Left ventricle
MAPCAs	Major aortopulmonary collateral arteries
MRI	Magnetic resonance imaging
PAH	Pulmonary artery hypertension
PV	Pulmonary vein
RA	Right atrium
RV	Right ventricle
SVC	Superior vena cava
TAPVC	Total anomalous pulmonary venous connection
TAPVD	Total anomalous pulmonary venous drainage
TAPVR	Total anomalous pulmonary venous return
VSD	Ventricular septal defect
VV	Vertical vein

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Author contributions

YSN contributed to study design, data collection, manuscript preparation, literature search, and data interpretation; IPS contributed to data collection, manuscript preparation, and literature search; IPM contributed to study design, manuscript preparation, and data interpretation; RFR contributed to literature search and manuscript preparation; SLW contributed to data collection; data interpretation; and manuscript preparation. All authors read and approved the final manuscript.

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Availability of data and materials

Not applicable.

Declarations

Ethics approval and consent to participate

Non-identifiable images of the patient have been only used. Careful attention has been provided to make sure that no patient identifiable information is in the images provided.

Consent for publication

Informed consent from the patient has been obtained, and signed consent was provided by the patient.

Competing interests

The authors declare that they have no competing interests.

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