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Case Series

Dual-energy computed tomography imaging in comprehensive assessment of truncus arteriosus in neonates: A case series

Yogesh Shardul[1], Anushka Mittal[1], Samrin Haq[1], Priyanka Garg[1]

[1]Department of Radiodiagnosis, Lady Hardinge Medical College and Associated Hospitals, New Delhi, India.

***Corresponding author:**
Anushka Mittal,
Department of Radiodiagnosis,
Lady Hardinge Medical College
and Associated Hospitals,
New Delhi, India.

dranushkamittal23@gmail.com

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ABSTRACT

Truncus arteriosus (TA) is a rare congenital heart disease in which a single arterial trunk arises from the heart, providing systemic, coronary, or pulmonary circulation. This condition is frequently associated with ventricular septal defects and various other cardiac and pulmonary irregularities. In this case series, we described the radiological findings and the related cardiac and pulmonary complications in three neonates diagnosed with persistent TA (PTA). This series underscores the critical role of dual-energy computed tomography in the assessment of PTA and illustrates its utility in identifying associated anomalies, thereby enhancing comprehensive patient management.

Keywords: Interrupted aortic arch, Persistent truncus arteriosus, Ventricular septal defect

INTRODUCTION

In 1978, Wilson was the first to characterize a rare congenital defect known as persistent truncus arteriosus (PTA), which accounts for approximately 2% of all congenital cardiac anomalies with slight male predominance. This condition is characterized by the inadequate formation of distinct pulmonary trunk and aorta. Instead, a single large arterial vessel emerges from both heart ventricles, supplying blood to the major organs.[1] PTA is frequently associated with a non-restrictive anterior ventricular septal defect (VSD), which facilitates the flow of blood from the left ventricle to the right ventricle. This occurs because the left ventricle is typically hypoplastic and exhibits minimal to no functional capacity. Various structural abnormalities such as a right-sided aortic arch, double aortic arch, left superior vena cava, secundum atrial septal defect (ASD), interrupted aortic arch (IAA), and aberrant subclavian artery are also commonly seen with truncus arteriosus (TA).[2]

CASE SERIES

Case 1

A 8-day-old female presented with fever and respiratory distress. On examination, the patient had cyanosis, tachypnoea, and tachycardia. On auscultation, a grade 2 systolic murmur was present at the apex. On chest auscultation, decreased breath sounds toward the left side. Cardiac anomaly along with secondary pulmonary infection was suspected in this child. On computed tomography (CT) cardiac angiogram, both ventricles were seen to be opening into a single trunk. The ascending aorta and main pulmonary artery were seen to arise from a single trunk. A sub-truncal VSD and membranous ASD were seen in this patient. It was diagnosed as Van Praagh type A1 TA [Figure 1].

Case 2

A 14-day-old female presented with respiratory distress and difficulty in feeding. On examination, the patient had cyanosis and tachycardia. On pulse oximetry evaluation, the newborn had suboptimal oxygen saturation (SpO_2) and systolic murmur on auscultation. Echocardiography was suggestive of complex congenital heart disease (CHD). On CT cardiac angiogram, dextrocardia was seen along with the presence of a common ventricle. A single arterial trunk was seen arising from the common ventricle, suggesting TA. Both pulmonary arteries were seen to arise from the posterior aspect of the trunk. It was diagnosed as Van Praagh type A2 TA [Figure 2].

Case 3

A 7-day-old male was screened for CHD under pox (pulse oximetry) screening. On pulse oximetry evaluation, the newborn had suboptimal SpO_2 and was thus admitted to the neonatal intensive care unit for further workup. On clinical examination, the patient had pallor and a continuous murmur on auscultation. Echocardiography showed TA with VSD and IAA. On CT cardiac angiogram, a single large arterial trunk was seen arising from the ventricles along with the presence of a sub-truncal VSD. There was a confluent origin of the right and left pulmonary arteries from the posterior part of the main arterial trunk, crossing each other to supply the opposite side. The ascending aorta was seen to arise from the common arterial trunk with interruption of the arch of the aorta at its distal part. The descending thoracic aorta was supplied by the patent ductus arteriosus. It was diagnosed as Van Praagh type A4 TA due to the presence of an IAA [Figure 3].

All three neonates expired due to severity of congenital heart anomalies and their associated cardiovascular and respiratory complications.

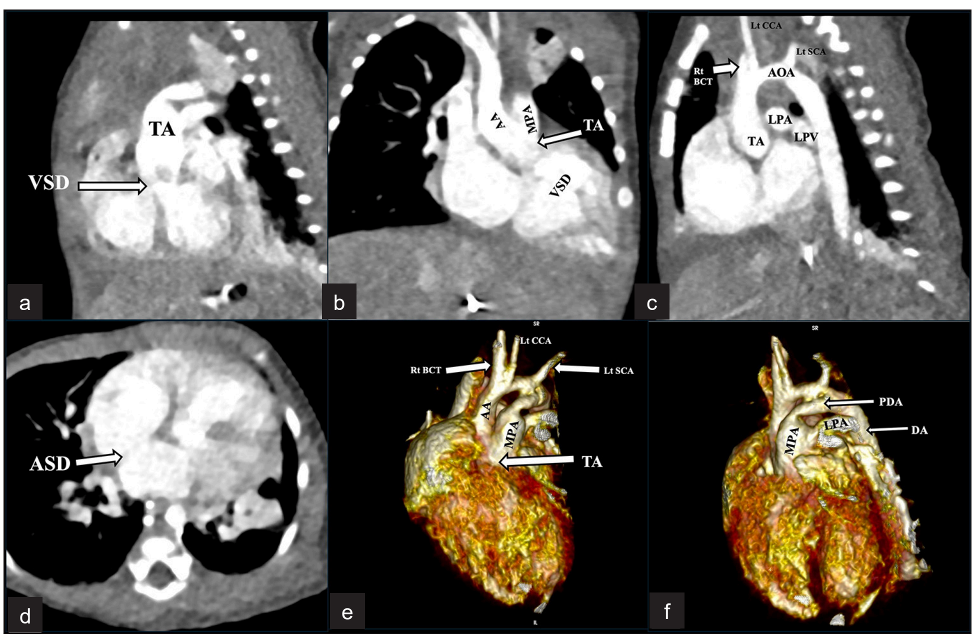

Figure 1: Cardiac CT angiogram images demonstrating truncus arteriosus and associated anomalies. (a-b) Sagittal and coronal MIP (maximum intensity projection) images show a single arterial trunk (truncus arteriosus) overriding both right and left ventricles, with a subtruncal ventricular septal defect (VSD). (c-d) Sagittal and axial MIP images demonstrate the arch of aorta and its branches arising from the truncus arteriosus, along with a membranous atrial septal defect (ASD). (e-f) Volume-rendered (VRT) images depict the truncus arteriosus giving rise to both the main pulmonary artery (MPA) and ascending aorta (AA) from a common origin. TA: Truncus arteriosus, VSD: Ventricular septal defect, ASD: Atrial septal defect, MPA: Main pulmonary artery, AA: Ascending aorta, LPA: Left pulmonary artery, LPV: Left pulmonary vein, DA: Descending aorta, PDA: Patent ductus arteriosus, BCT: Brachiocephalic trunk, CCA: Common carotid artery.

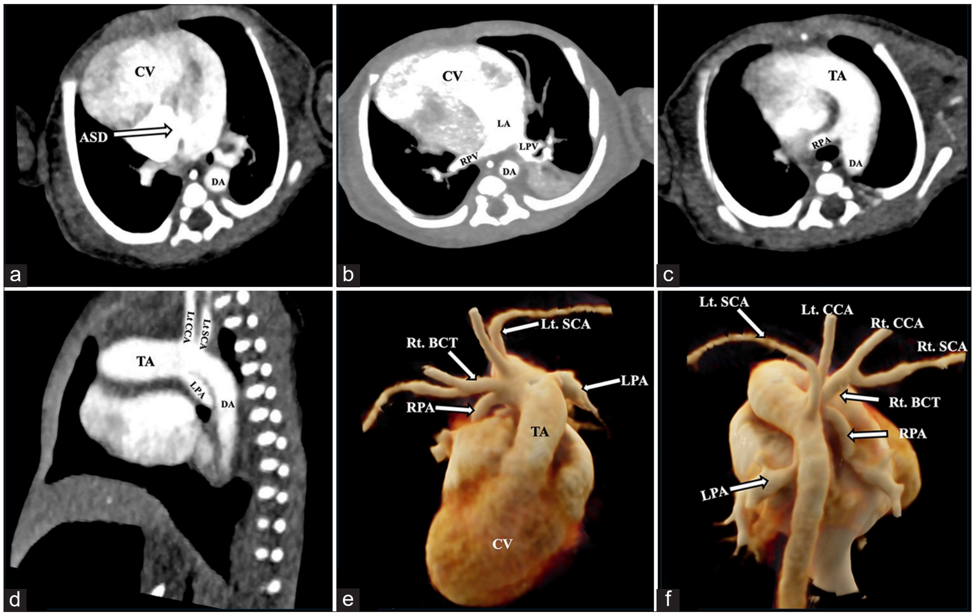

Figure 2: Cardiac CT angiogram images demonstrating truncus arteriosus with associated anomalies. (a-b) Axial MIP images show dextrocardia with a common ventricle receiving both atria, along with a large atrial septal defect (ASD). (c-d) Axial and sagittal MIP images demonstrate a single arterial trunk (truncus arteriosus) arising from the common ventricle, giving rise to both the pulmonary artery and aorta. (e-f) Volume-rendered (VRT) images depict the origin of truncus arteriosus from the common ventricle, with bifurcation into the pulmonary artery and aorta. CV: Common ventricle, LA: Left atrium, TA: Truncus arteriosus, LPV: Left pulmonary vein, RPV: Right pulmonary vein, ASD: Atrial septal defect, RPA: Right pulmonary artery, LPA: Left pulmonary artery, DA: Descending aorta, PDA: Patent ductus arteriosus, BCT: Brachiocephalic trunk, CCA: Common carotid artery, SCA: Subclavian artery.

DISCUSSION

The cyanotic heart defect known as PTA is characterized by a VSD, a single truncal valve, and a common ventricular outflow tract. At the site of the VSD, blood from systemic venous return mixes with pulmonary venous blood, resulting in the ejection of desaturated blood through the shared outflow tract. In PTA, pulmonary blood flow is largely regulated by pulmonary vascular resistance, which is generally elevated due to the direct attachment of the pulmonary arteries to the truncus. Truncal valve can be bicuspid, tricuspid (most common), or quadricuspid.[3]

The etiology of TA is not fully understood; however, it has been associated with abnormalities in neural crest cell development and migration. Microdeletions within chromosome 22q11.2 are identified in 30–40% of cases, frequently linked to defects such as branch pulmonary artery stenosis. Genes like TBX1 are critical for conotruncal development, though their precise role in the condition remains uncertain. Additional genetic contributors, including NKX2.6 and GATA6, as well as maternal diabetes, have been suggested as potential factors, but their overall impact is considered minor.[4]

IAA is a rare pharyngeal arch artery defect characterized by a discontinuity between the ascending and descending aorta, with the descending aorta typically supplied by a patent ductus arteriosus. The occurrence of IAA alongside TA is extremely uncommon, with Van Praagh type A4 TA representing 12% of all TA cases.[5,6] Classification and different modalities in evaluation of TA are shown in Figure 4 and Table 1, respectively.[1,7-9]

Dual-energy CT (DECT) plays a valuable role in the imaging of pediatric CHD by providing both anatomical and functional insights in a single acquisition. It enhances visualization of vascular structures, shunts, and myocardial and pulmonary perfusion, which is particularly useful in complex or cyanotic CHDs. DECT's material decomposition capabilities allow for automatic bone removal and iodine-specific imaging, improving vascular assessment without additional radiation. In addition, DECT offers a significant advantage in radiation dose optimization. By generating Virtual non-contrast (VNC) images from contrast-enhanced acquisitions, it obviates the need for separate non-contrast scans, thereby reducing cumulative radiation exposure an important consideration in the pediatric population. Importantly, DECT enables lung perfusion mapping and iodine overlay analysis, offering functional evaluation without the need for additional scans, making it a comprehensive, low-dose solution for pediatric cardiac

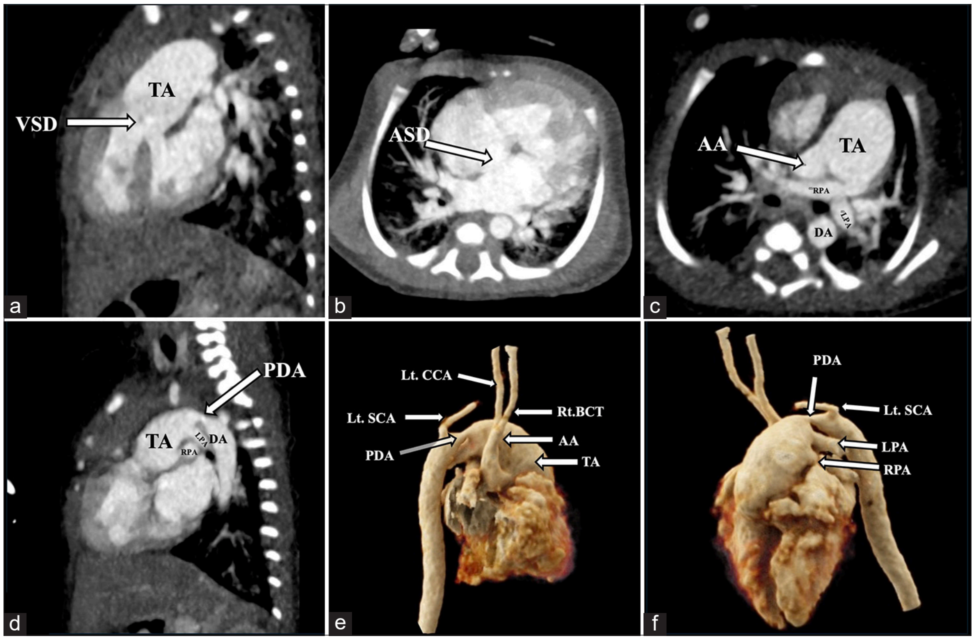

Figure 3: Cardiac CT angiogram images demonstrating truncus arteriosus with associated anomalies. (a-b) Sagittal and axial MIP images show a single arterial trunk (truncus arteriosus) overriding both right and left ventricles, with a subtruncal ventricular septal defect (VSD) and a membranous atrial septal defect (ASD). (c-d) Axial and sagittal MIP images demonstrate the pulmonary arteries and ascending aorta arising from the truncus arteriosus. The pulmonary arteries cross each other and supply the contralateral lungs. (e-f) Volume-rendered (VRT) images reveal truncus arteriosus with interrupted aortic arch and an aberrant left subclavian artery originating from the descending aorta. TA: Truncus arteriosus, VSD: Ventricular septal defect, ASD: Atrial septal defect, RPA: Right pulmonary artery, LPA: Left pulmonary artery, DA: Descending aorta, PDA: Patent ductus arteriosus, BCT: Brachiocephalic trunk, CCA: Common carotid artery, SCA: Subclavian artery.

Table 1: Role various modalities in imaging of truncus arteriosus.

Modality	Key findings/advantages	Disadvantages
Chest radiography	• Increased pulmonary vascular markings- simple, quick, widely available, inexpensive	• Low specificity, limited anatomical detail
Echocardiography	• Determines hemodynamic, visualizes congenital anomalies (e.g., VSD, pulmonary artery origin) – Non-invasive, portable, non-radiating	• Operator-dependent, limited visualization of vessel anatomy and extra-cardiac structures
Cardiac catheterization and angiography	• Direct visualization of anatomy, assesses hemodynamic and associated anomalies – Useful for interventional procedures (e.g., stenting)	• Invasive, requires sedation, high radiation, risk of complications
Cardiovascular MRI	• Provides detailed structural/functional data, non-invasive, excellent soft-tissue contrast – Ideal for diagnosis and functional evaluation	• Expensive, long scan time, limited accessibility, artifacts with metallic implants
CT	• Detailed evaluation of cardiac and extra-cardiac anomalies (e.g., arterial trunk, aortic arch, and airways) – Quick acquisition, practical for surgical planning, minimal sedation needed for younger patients	• Uses ionizing radiation and contrast media, sensitivity concerns in pediatric patients
DECT (Dual-energy CT)	• Advanced MDCT with weight-based low-dose protocol, high resolution for high heart rates, and minimal breath-holding	• Limited availability

CT: Computed tomography, MRI: Magnetic resonance imaging, DECT: Dual-energy computed tomography, MDCT: Multidetector computed tomography, VSD: Ventricular septal defect

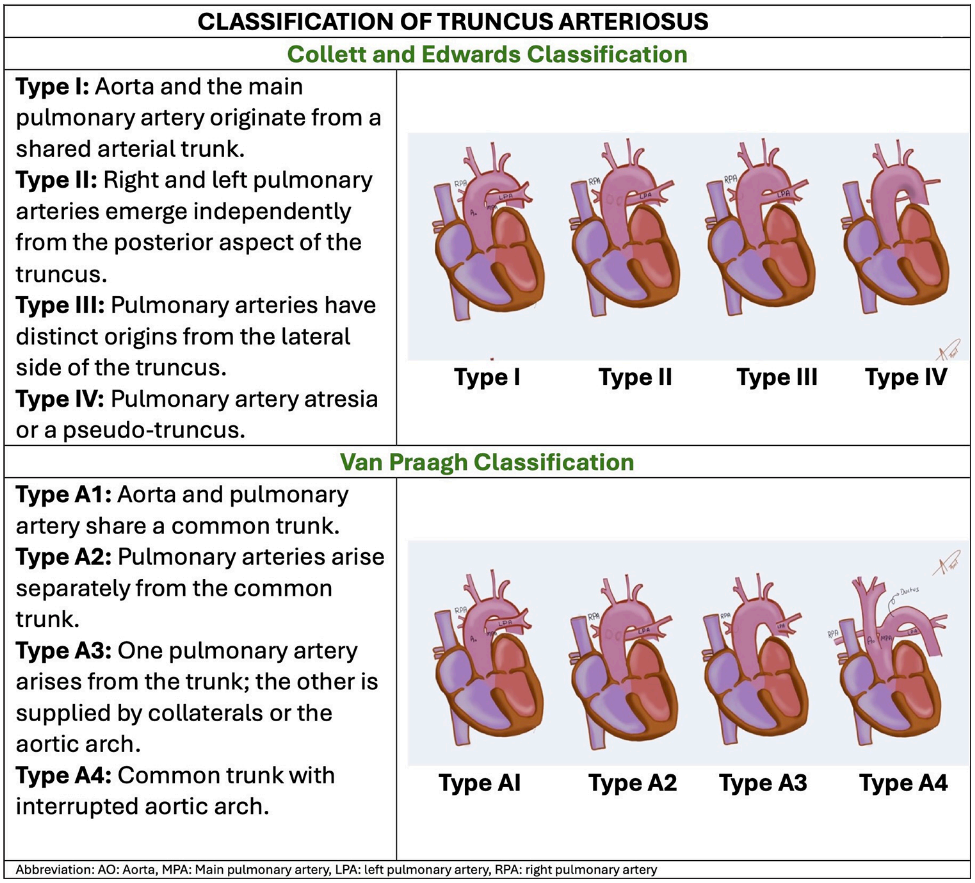

Figure 4: Classification of truncus arteriosus.

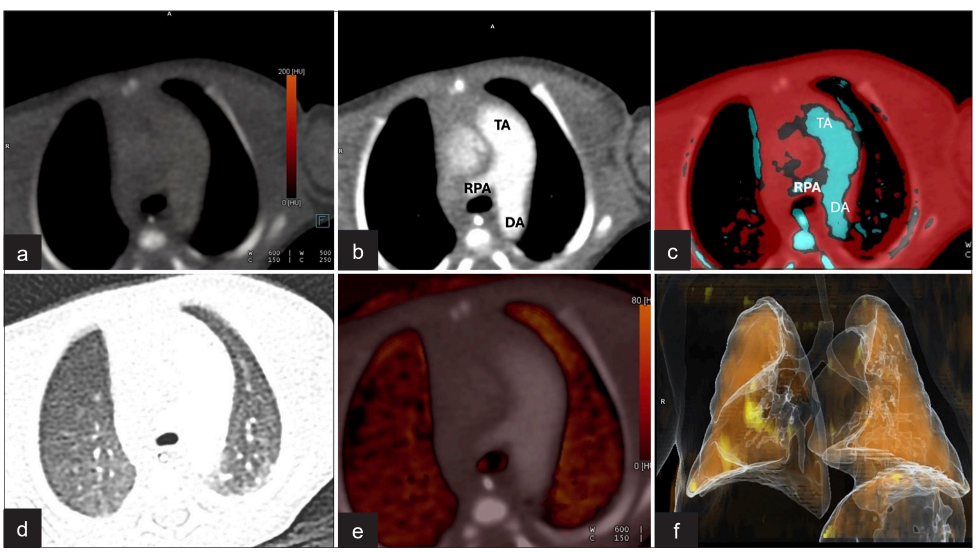

Figure 5: Representative material-specific images from DECT examination performed in case no 2. (a) Axial virtual non-contrast image. (b) Axial contrast enhanced CT image of truncus arteriosus and its branches. (c) Axial pulmonary vessel image showing intravascular contrast in TA, RPA and DA, and color coded blue. (d) Axial lung window image shows no significant abnormality. (e) Axial perfused blood volume image shows good perfusion, color-coded reddish orange. (f) Volume rendering reconstruction image shows normal lung perfusion. DECT: Dual-energy computed tomography, CT: Computed tomography, TA: Truncus arteriosus, RPA: Right pulmonary artery, DA: Descending aorta.

imaging.[10,11] Representative material-specific images from the DECT examination performed in Case no. 2 are shown in Figure 5.

Surgical repair of TA is typically performed within the first few weeks of life. The surgery involves closing the VSD with a patch, connecting the pulmonary arteries to the right ventricle using a homograft, and repositioning the truncal valve to serve as the neo-aortic valve aligned with the left ventricle. CT angiography is crucial before surgery for identifying truncus type, visualizing pulmonary arteries, and detecting issues such as aortic arch interruption or coarctation. Double outlet right ventricle and pulmonary atresia are close differential of truncus arteriosus. Imaging modality like cardiac CT angiography helps to differentiate these entities. Postoperatively, it helps to evaluate the pulmonary homograft and any residual or recurrent stenosis in the aortic arch or pulmonary arteries.[12]

DIFFERENTIAL DIAGNOSIS

Double outlet right ventricle and pulmonary atresia are close differential of truncus arteriosus. Imaging modality like cardiac CT angiography helps to differentiate these entities.

CONCLUSION

CT imaging plays a critical role in diagnosing and conducting pre-operative evaluations for TA, as it provides comprehensive insights into its anatomical characteristics and related anomalies. Dual-energy CT serves as a valuable adjunct to echocardiography and is essential for effective surgical planning due to its high-resolution and three-dimensional representations of cardiovascular structures.

TEACHING POINTS

1. Early clinical suspicion of TA should arise in neonates presenting with cyanosis, respiratory distress, or a systolic murmur, especially if SpO_2 remains low despite supportive care.
2. Associated anomalies such as VSD, IAA, aberrant subclavian artery, and ASD are common and must be actively sought on imaging, as they significantly impact surgical approach and prognosis.
3. DECT offers a significant advantage in radiation dose optimization. By generating VNC images from contrast-enhanced acquisitions, it obviates the need for separate non-contrast scans, thereby reducing cumulative radiation exposure an important consideration in the pediatric population.

MCQs

1. Which Van Praagh classification type of TA is associated with an IAA?
   a. Type A1
   b. Type A2
   c. Type A3
   d. Type A4

Answer Key: d

2. What congenital anomaly is most commonly associated with PTA?
   a. ASD
   b. VSD
   c. Coarctation of the aorta
   d. Tetralogy of Fallot

Answer Key: b

3. What is the primary reason for early surgical intervention in neonates with TA?
   a. To improve SpO_2 levels
   b. To prevent arrhythmias
   c. To separate systemic and pulmonary circulations and correct associated defects
   d. To close the patent ductus arteriosus

Answer Key: c

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