

## Case Series

# Type A4 truncus arteriosus: series of 3 cases focused on dual source multidetector CT angiogram findings

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## ABSTRACT

Truncus arteriosus (TA) is a rare congenital cardiac anomaly caused by failure of normal conotruncal septation during the fetal development. This aberration leads to a common ventricular outflow artery over the malaligned large ventricular septal defect (VSD), supplying systemic, coronary and pulmonary circulation. People with such anomalous anatomy show variable presentation from early childhood to adult life depending on the severity of defects. We here present three cases of truncus arteriosus with aortic interruption / hypoplasia-coarctation (type A4 truncus arteriosus) with focus on relevant dual source MDCTA findings.

**Keywords:** Multidetector CT angiogram, Interrupted aorta, Truncus arteriosus, Ventricular septal defect, Patent Ductus arteriosus, Isthmic hypoplasia

## INTRODUCTION

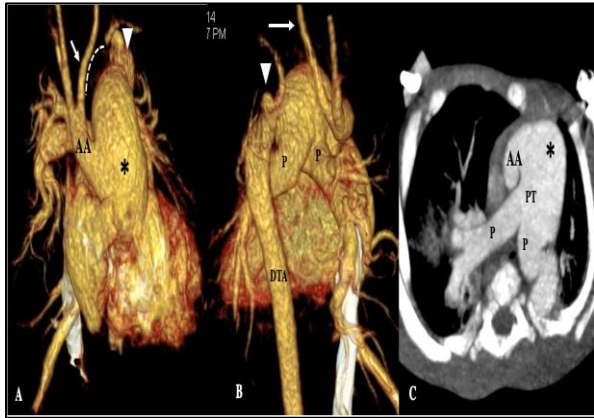
Truncus arteriosus is a rare congenital cardiac defect, categorized under conotruncal anomalies, affecting the ventricular outflow tract and great vessels.<sup>1</sup> It has an overall incidence of 95 per million live births.<sup>2</sup> The anomaly occurs due to the failure of conotruncal septation during intrauterine fetal development. This causes incomplete separation of aorta and pulmonary arteries with developmental aberration, leading to a large common ventricular outflow artery over the malaligned ventricular septal defect (VSD). Such anomaly leads to mixing of arterio-venous blood with a common arterial trunk supplying systemic, coronary and pulmonary circulation. These patients may present variably from early pediatric age to adult life depending on the severity of defects and associated other cardiac anomalies. Common pediatric symptoms include poor feeding, failure to thrive, cyanosis, recurrent lung infections and diaphoresis. In adulthood these symptoms may vary with palpitation and breathlessness. Proper knowledge and early detection with timely surgical correction are crucial to save the life of the patient. In this article, we will

present three cases of truncus arteriosus with interrupted aortic arch (IAA) or aortic hypoplasia-coarctation (type A4 truncus arteriosus). The article discusses relevant anatomical aberrations in truncus arteriosus, its types with focus on dual source MDCT angiogram findings.

### Case 1

A 13 years old female child presented to pediatric outdoor with progressive shortness of breath for one year. She was third child of parents born when age of mother and father were 35 years and 38 years respectively. First two children were normal with uncomplicated perinatal events. She has history of recurrent respiratory tract infection since early childhood. She also complained of palpitation and frequent sweating which have increased over past 4-5 months. The girl was otherwise alert and her developmental growth was normal for the age. On examination, she has pallor with grade-2 clubbing. A continuous murmur can be auscultated over her left upper parasternal area. Her lungs were clear with oxygen saturation (SpO<sub>2</sub>) of 96%. The 2-D echocardiogram showed a common ventricular outflow with single large artery. A large ventricular septal defect was present in

sub-arterial location. Left ventricle was mildly enlarged and ejection fraction was 46%. Considering the congenital heart anomaly, a cardiac angiogram was performed on dual source 256 slice CT scanner (Siemen's, Germany) with retrospective cardiac gating.



**Figure 1: VR images in variable orientation and a Thin MIP axial oblique image. The images show a common ventricular outlet with single large artery (\*).**

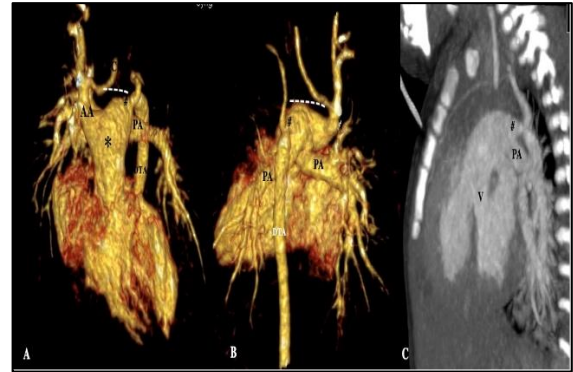
There is confluent origin of the right and left pulmonary arteries (P) from the main artery through a pulmonary trunk (PT). The ascending aorta (AA) is arising from the same arterial trunk, the interrupted (curved dotted line) between left common carotid (arrow) and left subclavian (arrow head) arteries (Type B interruption). This anatomy represents Type-A4 Truncus arteriosus.

Reconstructed CTA images (Figure 1A, B, C) show left ventricular enlargement with membranous ventricular septal defect. A common ventricular outflow tract was seen with single large artery. There was confluent origin of the right and left pulmonary arteries from posterior part of the main arterial trunk. The ascending aorta was small, originated from the proximal trunk. There was interruption of the aortic arch segment between the left common carotid and subclavian artery (Type B interruption). The descending thoracic aorta (DTA) distally was supplied by patent ductus arteriosus (PDA). This conotruncal anomaly with truncus arteriosus and interrupted aortic arch represents Type-A4 Truncus arteriosus.

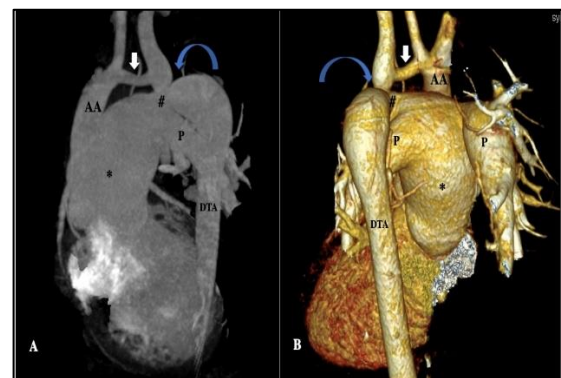
## Case 2

An 8 year old male child presented with history of recurrent respiratory tract infection. He was treated repeatedly in a peripheral private facility with clinical improvement on antibiotics without need of hospitalization. However, the issue of recurrence remained unresolved. The patient also complained of breathless during playing which hindered his activity. He had occasional palpitation and grade II clubbing.

His physical growth corresponded to 6years, though reflexes and mentation were normal. The patient had a left parasternal diastolic murmur and his SpO<sub>2</sub> was 98%. Single ventricular outflow artery was seen on 2-D echocardiogram with ventricular septal defect and dilated left ventricle. Ejection fraction was normal.



**Figure 2: VR images in anterior and posterior orientation and a sagittal oblique thin MIP image: show a single outlet arterial trunk (\*). Both the pulmonary arteries (P) arising from arising from this main trunk very close to each other. The ascending aorta (AA), arising from the arterial trunk is interrupted (curved dotted line) distal to left common carotid (white arrow) artery (Type B interruption). A short PDA (#) with stenosis at confluence is supplying the descending thoracic aorta (DTA). There is also a good size VSD (V). Type-A4 Truncus arteriosus.**



**Figure 3: Thick MIP image oblique and VR images in view: show a single large arterial trunk (\*) arising from ventricular outlet. Both the pulmonary arteries (P) arising from this main trunk with origin far from each other. The ascending aorta (AA) is also arising from the same arterial trunk with segmental hypoplasia of isthmus (arrow) with focal coarctation distally (arrowhead). A short PDA (#) is present with mild dilatation of the proximal descending thoracic aorta. This is Type A4 Truncus arteriosus.**

MDCTA done on the same scanner showed a single outflow arterial trunk, positioned over the ventricular septum (overriding) (Figure 2A, B, C). Right and left

pulmonary arteries were seen arising from this common artery on posterior aspect close to each other. The ascending aorta was also arising from the same arterial trunk with Type B aortic interruption. A short PDA was supplying the descending thoracic aorta with stenosis at its aortic confluence.

**Table 1: Classification systems for truncus arteriosus.**

Collett and Edward		Van Praagh system	
<b>Type-I</b>	Both PA arising from a short pulmonary trunk	Type A1	Same as type I of Collet and Edward system
<b>Type-II</b>	separate origin of the PA from the posterior aspect of the truncus	Type A2	Same as type II of Collet and Edward system
<b>Type-III</b>	separate origin of the PA from the lateral aspect of the truncus	Type A3	Left or right PA atresia, with collateral flow to the ipsilateral lung
<b>Type-IV</b>	a pseudo truncus (now considered a form of PA atresia with VSD)	Type A4	Truncus with associated aortic arch interruption, hypoplasia or coarctation

### Case 3

A 30 years old female was referred by an obstetrician to cardiology for consultation regarding her second pregnancy. She was diagnosed to have congenital cardiac anomaly after her first child birth, though her first pregnancy was uneventful. Her first child was also normal clinically with unremarkable echocardiography. She has history of mild dyspnea on climbing stairs. Her palpitation has become more frequent since her last pregnancy. She also had history of recurrent respiratory tract infection in childhood. Her examination revealed grade III clubbing and a continuous murmur in parasternal and diastolic murmur in mitral region. 2-D echocardiogram showed, enlarged left ventricle with mitral regurgitation. Single large ventricular outflow artery was seen with membranous ventricular septal defect. Ejection fraction was normal.

Reconstructed MDCTA images (Figure 3A, B) showed a single ventricular outflow tract with a common large arterial trunk. Both the pulmonary arteries arising from the posterior part of the main outflow trunk with origin far from each other. The ascending aorta is also arising from the proximal part of same arterial trunk. The aortic arch has segmental hypoplasia of isthmus. Arch also had a focal coarctation distal to the left subclavian artery. A short patent ductus arteriosus (PDA) was present between common trunk and aorta distally.

## DISCUSSION

Truncus arteriosus is characterized by single arterial outlet from the heart overriding the ventricular septum and supplying coronary, pulmonary and systemic circulations from the ascending aorta's proximal part.<sup>3</sup> The exiting arterial trunk is usually larger compared to normal aorta and positioned above the ventricular septum. Most of the time, it is associated with valvular abnormalities. A tricuspid valve is most common followed by quadricuspid or bicommissural valves, leading to stenosis or incompetence. The outflow may dominate over either of the ventricle or shared both ventricles equally. A subarterial non-restrictive ventricular septal defect (VSD) is present in nearly all patients with absent conal septum.

It is a rare anomaly with overall incidence of 95 per million live birth.<sup>2</sup> DiGeorge syndrome and chromosome 22q11 deletion have a well-established relation with the anomaly.<sup>4</sup> Amongst other associated anomalies, an interrupted aortic arch is commonest (11%–14% of cases), however; mitral valve, coronary arteries, and pulmonary venous connections abnormalities are also not uncommon.<sup>5</sup> Two of our cases have type-B aortic arch interruption while third case has a hypoplastic aortic isthmus with focal post ductal coarctation. All three cases have subarterial VSD and a patent ductus arteriosus.

On the basis of pulmonary artery origin, truncus arteriosus is divided into different types. The original classification system by Collett and Edward, divided truncus arteriosus into 4 types from Type I to IV.<sup>6</sup> This classification was further modified by Van Praagh.<sup>7</sup> Both the classification systems are briefed in Table 1.

As the presentation is variable at different ages from childhood to adult, early diagnosis and surgical repair is crucially important for survival. In this series two of our cases were diagnosed in early adolescent age, while third lady referred for planning of pregnancy was adult. Sundararajan et al in 1972 reported a case of TA with IIA in a three-year-old child.<sup>8</sup> Similar to our third case, Verhaert et al, reported an adult case of truncus with interrupted aortic arch who presented for counselling regarding her pregnancy.<sup>9</sup> In 2002, Lim et al reported a case of type B interruption with type II truncus arteriosus and coarctation of persistent 5th aortic arch.<sup>10</sup>

The correction involves patch repair of the VSD with alignment of the truncal valve which forms a neo-aortic valve from left ventricle. After detachment of the pulmonary arteries from the parent arterial trunk, these are connected to the right ventricle using a valved homograft. Associated critical anomalies like interrupted aortic arch or obstructive lesions are repaired in same setting. Conduit stenosis or regurgitation, branch pulmonary artery stenosis, neo-aortic truncal valve insufficiency or stenosis, VSD patch leak, and aortic arch obstruction are amongst repair related complications. Due

to increasing obstruction, most of these patients require homograft replacement over time.

## CONCLUSION

Truncus arteriosus (TA) is a rare congenital cardiac anomaly affecting ventricular outflow and septum with single artery supplying systemic, coronary and pulmonary circulation. Proper knowledge of key diagnostic features on MDCTA and structure reporting can help in early diagnosis and planning of definitive management.

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