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## Case Report

# Contrast enhanced computed tomography findings of persistent truncus arteriosus; A rare congenital heart disease <sup>☆</sup>

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## ABSTRACT

Persistent Truncus Arteriosus is a cyanotic congenital heart anomaly in which a single trunk supplies both the pulmonary and systemic circulation, instead of a separate aorta and a pulmonary trunk. It is usually classified as a conotruncal anomaly. Due to parallel fetal circulation, truncus arteriosus does not cause any haemodynamic problem in utero. However it is a major problem postnatally and, if left untreated, approximately 80% of infants die within the first year. Diagnosis should be made early by radiologists and cardiologists due to fatal illness. We found it valuable to present a case of truncus arteriosus with computerized tomography findings because of its rare occurrence.

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## Introduction

Persistent Truncus Arteriosus (PTA) is a cyanotic congenital heart anomaly in which a single trunk supplies both the pulmonary and systemic circulation, instead of a separate aorta and a pulmonary trunk. It is usually classified as a conotruncal anomaly.

It accounts for up to 2% of congenital cardiac anomalies and is almost always associated with a ventricular septal defect (VSD) to allow circulatory flow circuit completion [1]. We found it valuable to present a case of truncus arteriosus with computerized tomography findings because of its rare occurrence.

## Case report

A 6-day-old girl was admitted to the pediatric intensive care unit due to respiratory distress and cyanosis, and a 2/6-degree systolic murmur was detected in the left sternal area. Pulmonary hypoplasia and cardiomegaly were observed due to enlargement of the mediastinum due to large aortic shadow on the chest X-ray of the patient.

On the thorax Computed Tomography; It was observed that the cardiomegaly, and aorta and the pulmonary trunk were in the middle trunk (Fig. 1 A-B-C). The trunk emerged from the right ventricle and a VSD of 5 mm diameter was observed (Fig. 2). Pulmonary arteries were more proximal and

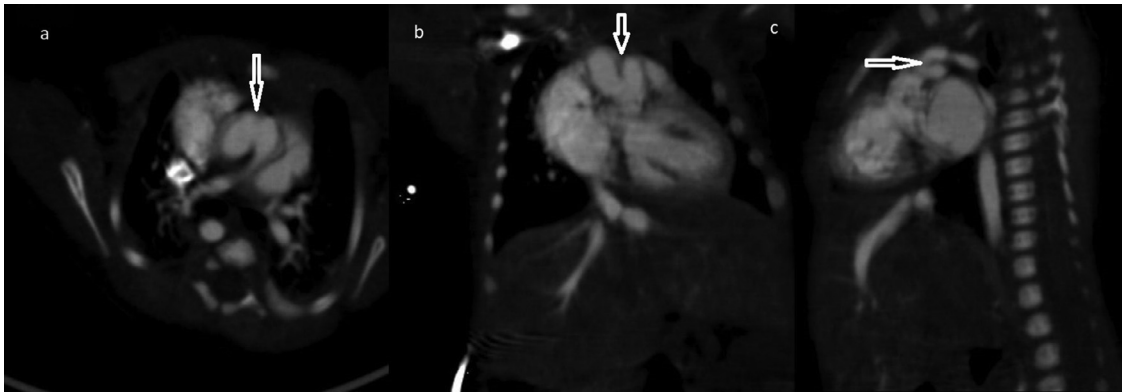
<sup>☆</sup> Competing Interests: The authors declare that they have no conflict of interest.

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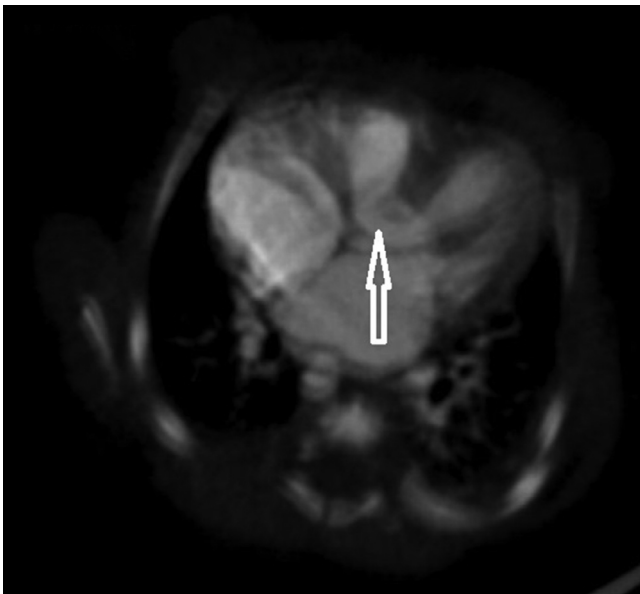
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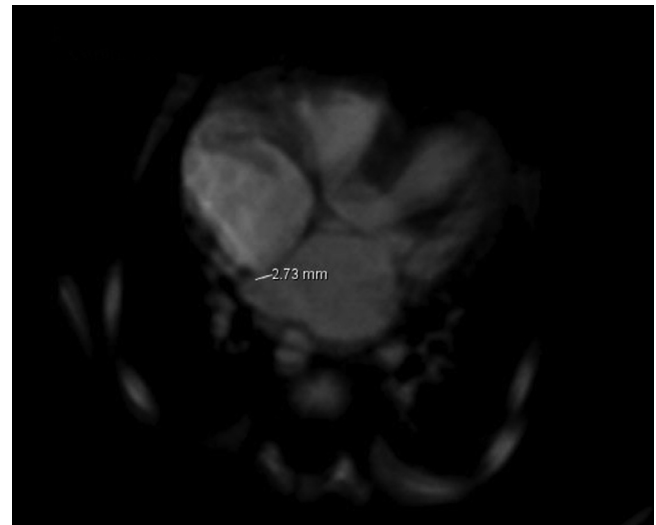
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**Fig. 1 – It was observed that the cardiomegaly, and aorta and the pulmonary trunk were in the middle trunk.**



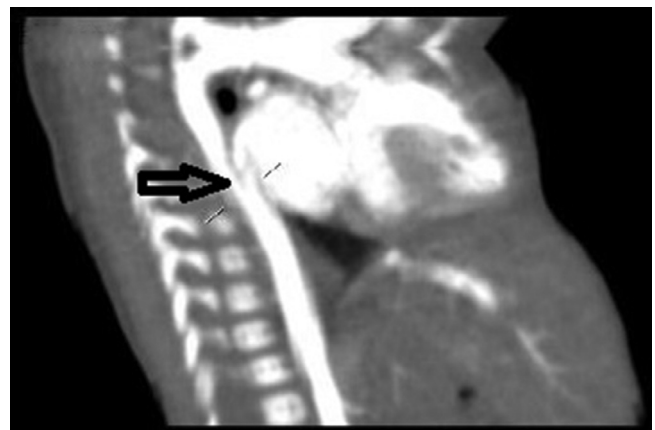
**Fig. 2 – The trunk emerged from the right ventricle and a VSD of 5 mm diameter was observed.**



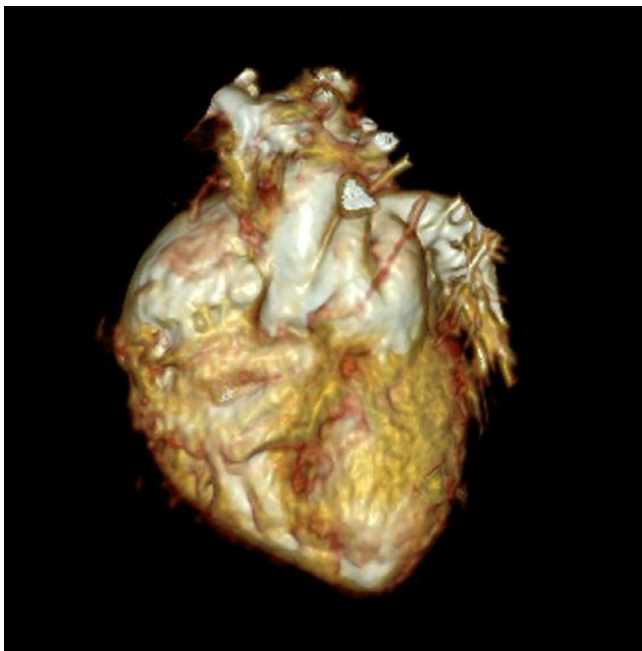
**Fig. 3 – Secundum type ASD was observed in the interatrial septum.**

slightly narrower. The left subclavian artery was found to have originated from the left main pulmonary artery. The vascular structure, which showed the right atrium of the aorta and extending from the inferior aorta to the left atrium, attracted attention. Secundum type ASD was observed in the interatrial septum (Fig. 3). Patent ductus arteriosus is observed in the sagittal reformat images (Fig. 4). In addition, PTA is observed in 3D Volume Rendering images (Fig. 5).

On the echocardiographic evaluation; From left to right, shunt, secundum type ASD and 5.5 mm VSD were detected. Single cardiac output and truncal bicuspid were observed in the heart. Trunks were observed with 50% dextroposes. Our findings were compatible with Type A1 according to Collett and Edwards and Type A1 according to Van Praagh.



**Fig. 4 – Patent ductus arteriosus is observed in the sagittal reformat images.**



**Fig. 5 – PTA is observed in 3D Volume Rendering images.**

## Discussion

The estimated incidence of PTA is 1 in 10,000 births. There is a lack of normal separation of the embryological truncus arteriosus into a separate aorta and pulmonary trunk. This results in a single arterial vessel that originates from the heart that supplies the systemic, pulmonary and coronary circulations. It may also result in a common truncal valve which can contain 2 to 4 cusps. In the embryological development process, at the 4th week, the truncus arteriosus is divided into aorta and pulmonary trunk. At the end of this period, PTA occurs if there is a lack of development steps [2].

Chest radiographs often show moderate cardiomegaly with pulmonary plethora and widened mediastinum.

However, the main pulmonary artery which is arising from common trunk, may be small/unusual in position which may result in a narrow mediastinum. This along with moderate cardiomegaly and pulmonary plethora gives an appearance that is similar to D-loop transposition of great arteries [3].

TA is caused by the failure of the aortico-pulmonary septum to develop and separate the embryonic truncus into the aorta and main pulmonary artery. Etiology is multifactorial and 22q11.2 deletion, maternal diabetes mellitus in pregnancy and teratogens such as retinoic acid and bisdiazine have been blamed [4].

Collett and Edward's (1948) classified PTA into 4 types [5](Table 1). Van Praagh and Van Praagh (1965) classified TA into 2 types based on the presence (type A) or absence (type B) of a VSD with the latter type being rare [6]. The 2 types are sub classified into 4 subtypes.(Table 2).

## Collett and Edwards classification(Table 1)

Type I: (Most common) both aorta and main pulmonary artery arise from a common trunk.

Type II: Pulmonary arteries arise separately from the posterior aspect of trunk, close to each other just above the truncal valve (negligible main pulmonary artery segment).

Type III: (Least common) pulmonary arteries arise independently from either side of the trunk.

Type IV: Neither pulmonary arterial branch arising from the common trunk (pseudotruncus), currently considered a form of pulmonary atresia with a VSD.

## Van Praagh modified the classification system(Table 2)

Type A1: Identical to the Type I of Collett and Edwards.

Type A2: Separate origins of the branch pulmonary arteries from the left and right lateral aspects of the common trunk.

Type A3: Origin of one branch pulmonary artery (usually the right) from the common trunk, with other lung supplied either by collaterals or a pulmonary artery arising from the aortic arch.

Type A4: Coexistence of an interrupted aortic arch.

Patients usually present in infancy with signs of congestive cardiac failure, tachypnea, tachycardia, failure to thrive [7]. Clinically the condition may have to be differentiated in the neonatal period from other congenital heart diseases causing early heart failure with absent or mild cyanosis and neonatal sepsis [3]. Chest radiograph findings depend on the hemodynamic circumstances [7]. Cardiomegaly with a small or absent main pulmonary segment with pulmonary vascular engorgement are the usual features [7–8].

In cases with an absent pulmonary artery, the pulmonary vascular pattern is diminished on that side [5]. A Right aortic arch is common. Cardiac catheterization with angiography is indicated when pulmonary vascular disease is suspected and to define great vessel.

## Conclusion

Due to parallel fetal circulation, truncus arteriosus does not cause any haemodynamic problem in utero. However it is a major problem postnatally and, if left untreated, approximately 80% of infants die within the first year. Diagnosis should be made early by radiologists and cardiologists due to fatal illness.

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