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# Type 3 Truncus Arteriosus with Type B Inturruption of Aortic Arch a Radiological Point of View

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## Abstract

Truncus Arteriosus (TA) is a rare congenital cardiac malformation in which a single common artery arises from the heart by means of a single semilunar truncal valve and supplies the systemic, pulmonary, and coronary circulations. Pulmonary arteries originate from the common arterial trunk distal to the coronary arteries and proximal to the first brachiocephalic branch of the aortic arch<sup>[1]</sup>. TA typically overrides a large outlet Ventricular septal defect (VSD).

# **1. Introduction**

The truncus arteriosus is a structure that is present during embryonic development. It is an arterial trunk that originates from both ventricles of the heart that later divides into the aorta and the pulmonary trunk. Truncus arteriosus is defined as a single trunc rising from the heart and supplying the coronary, pulmonary, and systemic circulations, with no remnants of an atretic aorta or pulmonary artery <sup>[11-13]</sup>. The number of semi lunar cusps may vary from 2 to 6. Typically a defect is present in the bulbar portion of the anterior ventricular septum, a defect lying just under the semi lunar cusps and entirely different from the usual ventricular septal defect involving or bordering on the membranous portion of the septum.

# 2. Structure

The truncus arteriosus and bulbus cordis are divided by the aorticopulmonary septum. The truncus arteriosus gives rise to the ascending aorta and the pulmonary trunk. The caudal end of the bulbus cordis gives rise to the smooth parts (outflow tract) of the left and right ventricles (aortic vestibule & conus arteriosus respectively).<sup>[8]</sup> The cranial end of the bulbus cordis (also known as the conus cordis) gives rise to the aorta and pulmonary trunk with the truncus arteriosus. This makes its appearance in three portions. Two distal ridge-like thickenings project into the lumen of the tube: the truncal and bulbar ridges.<sup>[9]</sup>

These increase in size, and ultimately meet and fuse to form a septum (aorticopulmonary septum), which takes a spiral course toward the proximal end of the truncus arteriosus. It divides the distal part of the truncus into two vessels, the aorta and pulmonary artery, which lie side by side above, but near the heart. Four endocardial cushions appear in the proximal part of the truncus arteriosus in the region of the future semilunar valves; the manner in which these are related to the aortic septum is described below. Two endocardial thickenings - anterior and posterior - develop in the bulbus cordis and unite to form a short septum, this joins above with the aortic septum and below with the ventricular septum. The septum grows down into the ventricle as an oblique partition, which ultimately blends with the ventricular septum in such a way as to bring the bulbus cordis into communication with the pulmonary artery, and through the latter with the sixth pair of aortic arches, while the left ventricle is brought into continuity with the aorta, which communicates with the remaining aortic arches.

## **3. Clinical Significance**

Failure of the truncus arteriosus to close results in the condition known as Persistent Truncus Arteriosus, or simply "Truncus arteriosus," one of many cyanotic heart defects or congenital heart defects.

Other pathologies of the truncus arteriosus include transposition of the great vessels (arteries in this case), and tetralogy of Fallot.<sup>[10]</sup>

## 4. Case Presentation

An 8 month old female baby came with the history of: Cyanosis Poor feeding Poor growth Since birth. RADIOGRAPH CHEST-Shows



- Cardiomegaly.
- Increased bronchopulmonary marking in bilateral lung fields.
  - Bilateral hyperinflation. PAEDIATRIC ECHOCARDIOGRAPHY



Short axis view showing chambers of heart



Ecocardiography, short axis view showing respective outflow tracts



Topogram shows cardiomegaly and bilateral hyperinfilation

Pictorial view of type 3 truncus arteriosus with type B inturruption of aortic arch Showing

- Right and left pulmonary arteries arising directly from the posterior aspect of the truncus.
- Common origin of innominate and left common carotid arteries from the truncus.
- Interruption of the aortic arch distal to the common origin of innominate and left common carotid arteries.
- Left subclavian artery arises from the distal arch.

artery

• Normal descending thoracic aorta. Indentation of left main bronchus by the left pulmonary



CECT Chest axial section at the level of spine of scapula showing arch of aorta on left side and shows proximal part of right and left pulmonary artries arising from posterior aspect of trances.



CECT chest and abdomen, saggital section shows common trances with left subclavian arteries arising from distal part of teancus



CECT Chest at the level of spine of scapula shows right and left pulmonary artries arising from the posterior aspect of trancus



CECT chest at the level of common trances shows Right pulmonary artery and immoninate arising from common trances with further branching (right subclavian, Right common caroted artery, left common carotid artery) arising from innominate artery.



CECT chest showing left subclavion arising from aortic arch



Volume rendered images shows as labeled below -RPA – Right pulmonary artery -RTSCLA- right sub clavion artery -RTCCA- Right common carotid artery -RTCCA- Left common carotid artery -LCCA- Left common carotid artery -LTVA- Left vertohral artery -LSCA-Left sub clavion artery -LPA-left pulmonary artery



Volume rendered image (porteroantorior view)shows digrametic representation of trances and branching pattern



Volume rendered image (anterioposterior view)shows diagrammatic representation of trances and its branching pattern



Volume rendered showing trachea with right and left main branchus



Volume rendered image shows left subclavian artery and left vertebral artery aroming from arch of aorta

post operative radiograph chest



Post operative diagram of trancus-done already

#### 5. Truncus Arteriosus

Truncus arteriosus, also known as common arterial trunk, is characterized by a single great artery that arises from the base of the heart and supplies the systemic, coronary, and pulmonary blood flow Truncus arteriosus accounts for 1% of cardiac lesions detected in fetal life. It is associated with a right aortic arch in 35% of cases and is commonly related to genetic anomalies. MR angiography and CT angiography clearly show this condition and its associated anomalies and are also used as follow-up imaging methods

The prognosis is poor in untreated patients (>90% mortality before 1 year of age) Surgical correction (i.e., closure of the VSD, separation of pulmonary arteries from the truncus, and implantation of a right ventricular–to–pulmonary artery conduit [the Rastelli procedure]



Classification of truncus arteriosus according to Collett and Edwards.

Type I, the pulmonary artery trunk arises from the proximal portion of the truncus arteriosus.

Types II and III, there is no pulmonary artery trunk and the pulmonary branches arise from the posterior and lateral mid segments of the truncus, respectively.

Type IV, the pulmonary circulation is dependent on major aortopulmonary collateral arteries.

### 6. Interrupted Aortic Arch

Interrupted aortic arch (IAA) is defined as a complete luminal and anatomic discontinuity between the ascending and descending aorta (IAA) is rare, accounting for only 1% of congenital heart diseases. It may occur as a simple or complex anomaly. In simple IAA, only VSD and PDA are seen. The complex form is associated with truncus arteriosus, transposition of the great arteries, double-outlet right ventricle, aortopulmonary window, and functional single ventricle. Patients with IAA usually undergo cardiac surgery during the first year of life during the preoperative evaluation, echocardiography might not allow differentiation of IAA from severe aortic coarctation with a hypoplastic arch in these cases, the assessment is complemented by CT or MR imaging.



Celoria and Patton classification of IAA.

*Type A*, is defined as an interruption distal to the left subclavian artery (*LSA*).

*Type B, the* absent segment is between the left common carotid artery (*LCC*) and left subclavian artery.

*Type C*, is defined as an interruption distal to the innominate artery *(IA)*.

## 7. Discussion

Truncus Arteriosus (TA), synonymous with common arterial trunk and Common aortico-pulmonary trunk accounts for 0.7-1.4% of all congenital heart diseases in live born infants (incidence of 0.03-0.056/1000 live births)<sup>[1]</sup>. There is no striking sex difference in its incidence although most series contain more males than females<sup>[1]</sup>. 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 bisdiamine have been blamed<sup>[1]</sup>.

TA is frequently associated with other cardiac and great vessel anomalies which are present in 34.8% cases<sup>[2]</sup>, such as Right aortic arch (25-30 % cases<sup>[4]</sup>), interrupted aortic arch, aberrant right subclavian artery, abnormal coronary arteries, atrial septal defect, tricuspid atresia, double aortic arch. Occurrence of aortic arch anomalies with TA has a strong association with 22q11.2 microdeletion<sup>[2]</sup>. Majority of extra cardiac anomalies are associated with CATCH22 syndrome which is present in 30-35% patients with TA<sup>[1]</sup>. CATCH22 syndrome (caused by a microdeletion in chromosome 22q11.2 -thought to affect migration or development of cardiac neural crest cells) is a combination of DiGeorge, Velocardiofacial

and Conotruncal anomaly face syndromes manifestations of which include cleft lip and palate, thymus and parathyroid dysfunction. Other extra cardiac manifestations reported include unilateral renal agenesis, dysplastic kidneys, holoprosencephaly, esophageal and duodenal atresia, imperforate anus, asplenia<sup>[1],[2],[3]</sup>.

Two classifications have been proposed for TA. Collett and Edward's (1948) classified TA into 4 types<sup>[2]</sup>. In Type 1, a single pulmonary trunk arises from the TA just distal to the truncal valve. In Type 2 and 3, the pulmonary trunk is absent and the two pulmonary branches arise from the dorsal wall of the truncus(Type 2) or from the side of the truncus(Type 3). In Type 4 also called Pseudo truncus<sup>[4]</sup>, pulmonary arteries are absent and the pulmonary circulation is supplied by MAPCA's arising from the descending aorta. Type 4 now corresponds to pulmonary atresia with VSD<sup>[2]</sup>.

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<sup>[3]</sup>. The 2 types are sub classified into 4 subtypes. In type A1, the aorticopulmonary septum is partly formed, resulting in a partially separate main pulmonary artery that arises from the common trunk. The aortic pulmonary septum is completely absent in type A2, and both pulmonary arteries arise directly from the common trunk. Type A2 includes types 2 and 3 of Collett and Edward's. In Type A3 one pulmonary artery is absent and that lung is supplied by collateral vessels (e.g. bronchial arteries) or a pulmonary artery from a patent ductus arteriosus (PDA) or MAPCA's from the descending aorta. Type A4 is defined not by the pattern of origin of the pulmonary arteries but by the coexistence of hypoplasia, coarctation, atresia, or absence of the aortic arch<sup>[1]</sup>. The truncus consists largely of the main pulmonary artery component with a large PDA supplying the descending aorta<sup>[3]</sup>. In these cases a well documented association with Di George syndrome is observed<sup>[1]</sup>. TA type 1 and type 2 are the most common forms. Type 3 is least common<sup>[5]</sup>.

### 8. Clinical Presentation

Patients usually present in infancy with signs of congestive cardiac failure, tachypnea, tachycardia, failure to thrive<sup>[4]</sup>. 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<sup>[1]</sup>.

## 9. Radiological Findings

ECG is non specific<sup>[1]</sup>. Chest radiograph findings depend on the hemodynamic circumstances<sup>[4]</sup>. Cardiomegaly with a small or absent main pulmonary segment (does not develop in its usual position) with pulmonary vascular engorgement (pulmonary arteries receive blood at systemic pressures) are the usual features<sup>[4],[5]</sup>. In cases with an absent pulmonary artery, the pulmonary vascular pattern is diminished on that side<sup>[3]</sup>. A Right aortic arch is common.

On Echocardiography the origin of the pulmonary artery

orifices are best observed in parasternal short axis view<sup>[6]</sup>. The long axis-parasternal view shows the size of the truncus with the truncal valve and the VSD as well as the degree of overriding. The single truncal valve is usually tricuspid (61% cases) and is quadricuspid in most of the rest<sup>[1]</sup>, but may have up to six leaflets<sup>[6]</sup> and is frequently incompetent and/or stenotic. The posterior truncal wall is seen in fibrous continuity with the anterior mitral leaflet<sup>[6]</sup>. Color Doppler study will demonstrate the regurgitation or stenosis of the truncal valve.

Cardiac catheterization with angiography is indicated when pulmonary vascular disease is suspected and to define great vessels and coronary artery anatomy.

Sagittal and Transverse MR Images at the base of the heart can demonstrate a large arterial trunk with truncal valve over riding the inter ventricular septum and the origin of the pulmonary arteries. Coronal or oblique images are useful for determining the size and location of the VSD. Potential pulmonary artery stenosis can also be sought after<sup>[7]</sup>. Prenatal diagnosis of truncus has been reported<sup>[2]</sup> and when it is suspected by fetal echocardiography, karyotyping for Band 22q 11 deletion should be done.

#### 10. Prognosis

Prognosis is poor without treatment. Corrective operation (Closure of VSD, Separation of pulmonary arteries from primitive truncus and right ventricular to pulmonary artery conduit -Rastelli's procedure) is indicated before 3 months of age to avoid development of severe pulmonary vascular obstructive disease<sup>[4]</sup>.

#### References

- [1] Cifarelli A, Ballerini L. Truncus arteriosus. Pediatr Cardiol 2003; 24:569-73.
- [2] Volpe P, Paladini D, Marasini M, et al. Common arterial trunk in the fetus: characteristics, associations, and outcome in a multicentre series of 23 cases. Heart 2003; 89:1437-1441.
- [3] Ferdman B, Singh G. Persistant truncus Arteriosus. Current Treatment Options in Cardiovascular Medicine 2003, 5:429-438
- [4] Braunwald E, Zipes PD, Libby P. Heart Disease, 6thed. WB Saunders, 2001:1536-1537.
- [5] Sutton D. Text book of radiology and imaging, 7thed.Churchill Livingstone, 2003:397-398.
- [6] Feigenbaum H. Echocardiography, 5thed. Lea & Febiger, 1994: 418-421.
- [7] Stark DD, Bradley GW Jr. Magnetic Resonanance Imaging, 3rded. St. Louis, Missouri: Mosby, 1999:399-400.
- [8] Le, T., Bhushan, V., et al. 'First Aid for the USMLE STEP1'. 2009.
- [9] Le, Tao; Bhushan, Vikas; Vasan, Neil (2010). First Aid for the USMLE Step 1: 2010 20th Anniversary Edition. USA: The McGraw-Hill Companies, Inc. p. 123. ISBN 978-0-07-163340-6.

- [10] Le, Tao; Bhushan, Vikas; Vasan, Neil (2010). First Aid for the USMLE Step 1: 2010 20th Anniversary Edition. USA: The McGraw-Hill Companies, Inc. p. 123. ISBN 978-0-07-163340-6
- [11] ABBOTT, 2M. E.: Atlas of Congenital Cardiac Disease. New York, the American Heart Association, 1936, pp. 52, 60.
- [12] LEV, M., AND SAPHIR, 0.: Truncus arteriosus communispersistens. J. Pediat. 20: 74, 1942.
- [13] COLLETT, R. Wr., AND EDWARDS, J. E.: Persistenttruncus arteriosus: A classification according toanatomic types. Surg. Clin. North America 29:1245, 1949.