International Journal of Medical Science in Clinical Research and Review
Online ISSN: 2581-8945
Available Online at https://ijmscrr.in/ Volume 7|Issue 06 (November-December) |2024 Page: 1209-1217

Case Report

Persistent Truncus Arteriosus & Lung Hypoplasia - A Case Report.

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Article Received: 29- September -2024, Revised: 19-October-2024, Accepted: 09-November-2024

ABSTRACT:
Persistent Truncus Arteriosus is a RARE 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 cono truncal anomaly It is a major problem postnatally and, if left untreated, approximately 80% of infants die within the first year. Early Diagnosis should be made by radiologists and cardiologists due to fatal illness. We found it valuable to present a case of truncus arteriosus with lung hypoplasia with Imaging & 2D ECHO findings because of its impact on patient's life and for it's rare occurrence.

Keywords: truncus arterious, Lung hypoplasia, Ventricular Septal defect.

INTRODUCTION:

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 [i ii iii]

TA typically overrides a large outlet Ventricular septal defect (VSD). 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 We observed a 16 year old female with TA Type II(Collett & Edwards) ,Type AII (Van praagh)with severely hypoplastic left lung , hypoplastic left pulmonary artery and Major RT pulmonary artery supplying the right lung. In view of the rarity of this condition it was considered worthy of case report.

CASE REPORT:

A 16 YR old female came to cardiology OPD from rural background with complaints of Chest pain, short ness of breath and fatigue on exertion since childhood. She is referred to a higher centre after visiting a primary centre.She is malnourished and weighs around 30 kg On auscultation Grade 3/6 systolic murmur is heard in Left parasternal area. Her Pulse rate is 134 / min.Her blood pressure is 118/58 mm hg. Her SPO2 levels are 75mm hg. She is on Tab sildenafil 20mg, Tab Bosentan 62.5mg & tab dytor.

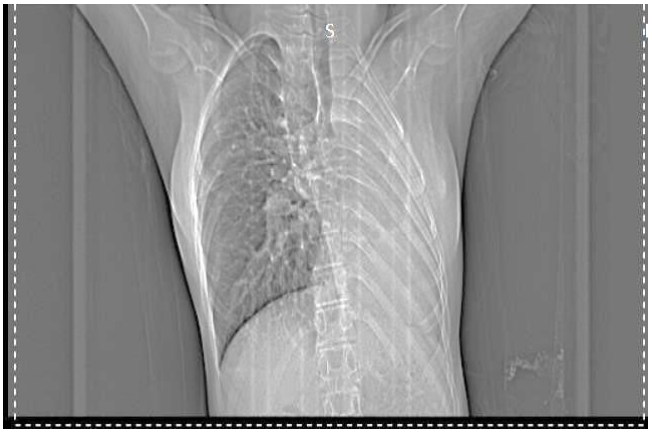
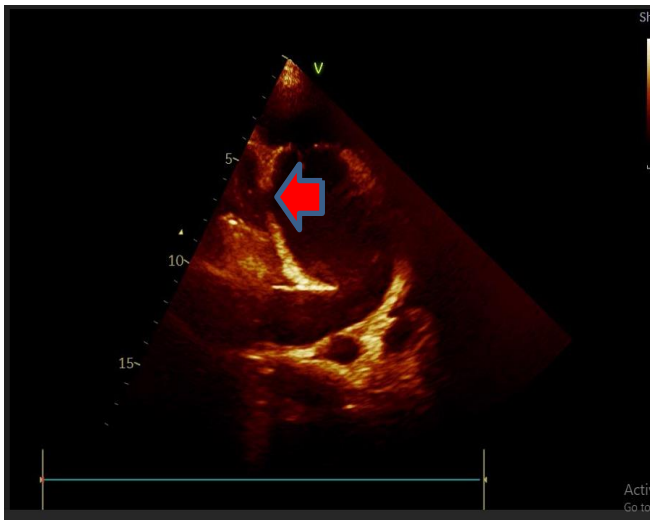
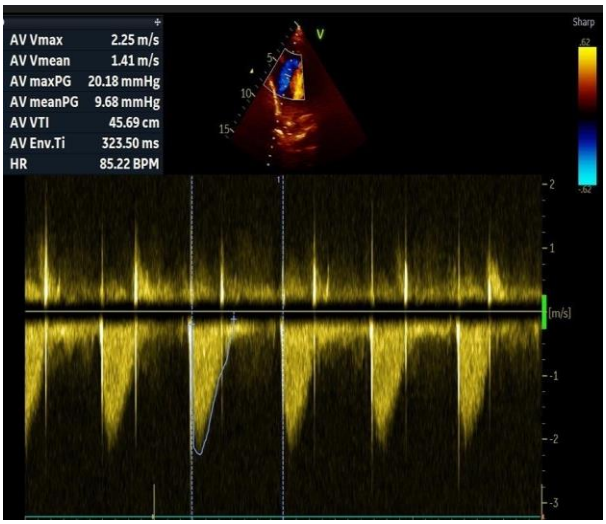


Figure 1. On chest x ray there is severe left lung hypoplasia, cardiomegaly, Right pulmonary plethora& mediastinal shift to LT side.

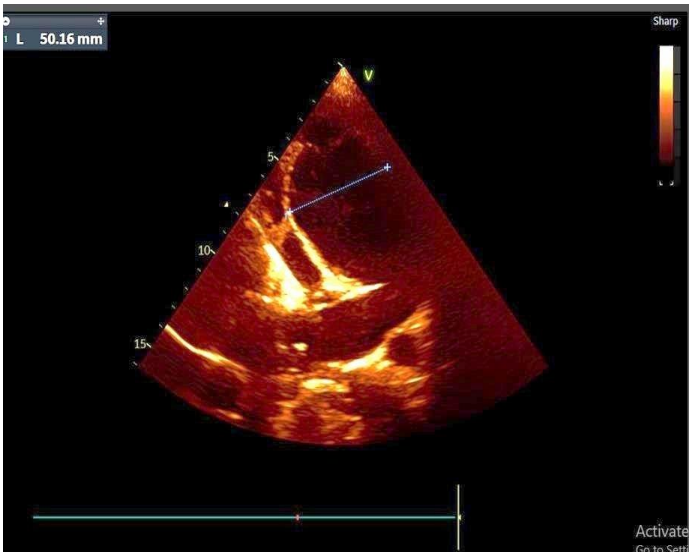
On Echocardiography [iv] in short axis view showed origin of common Truncal artery with a bicuspid valve .There is a large Sub arterial Ventricular septal defect(VSD) of 13mm with bidirectional shunt. There is moderate tricuspid regurgitation (TR) and Pulmonary arterial pressure of 52mm hg- Moderate PAH (Pulmonary arterial hypertension).Mitral and tricuspid valves are intact. Ventricular function is normal.

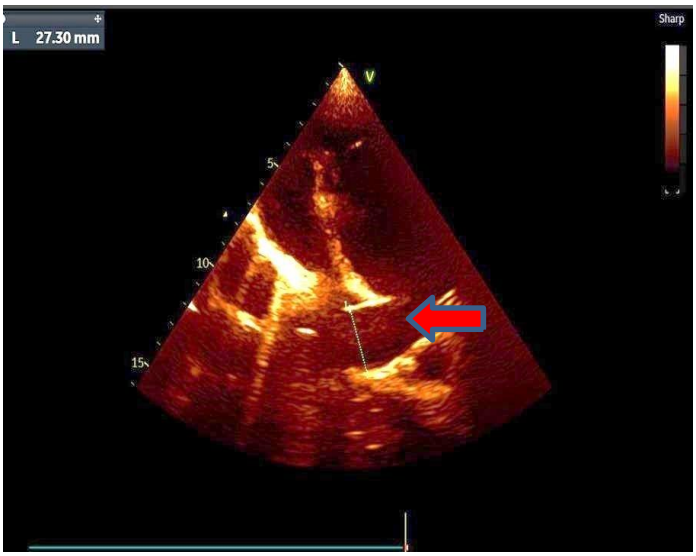


2A VSD on 2DECHO.



2BVSD show bidirectional flow





3A-2D ECHO- large common arterial Trunk
3B- Origin of RT pulmonary artery from Posterior part of arterial trunk (2.7cm)

On CT pulmonary angiography (CTPA), [v] [vi] Heart is positioned with in left hemi thorax with apex to left side. Rt pulmonary veins are prominent draining in to LT atrium with complete aplasia of LT pulmonary veins.A large sized single arterial common trunk (maximum Transverse and AP diameters of 7.3cms& 5.7cms respectively) is seen overriding the interventricular septum with concomitant membranous VSD.There are no signs of a common main pulmonary trunk. The Right Pulmonary artery is significantly hypertrophied(AP Diameter: 2.6cms)arising from the posterior aspect of common arterial trunk above the valve. The left pulmonary artery is under developed arising along the Posterior aspect of common arterial trunk close to Right branch. The aorta is in normal location with no anomalies. There is severe Left lung hypoplasia with parenchyma replaced with malformed multilocular cystic mass. The left main bronchus is atretic. There is compensatory hyperinflation of Right lung with prominent pulmonary arterial branches. No E/O Pulmonary thromboembolism.
All these findings are suggestive of Persistent Truncus arteriosus (Collett and Edwards type II & Van praagh type AII) with associated membranous VSD and severe LT lung hypoplasia.

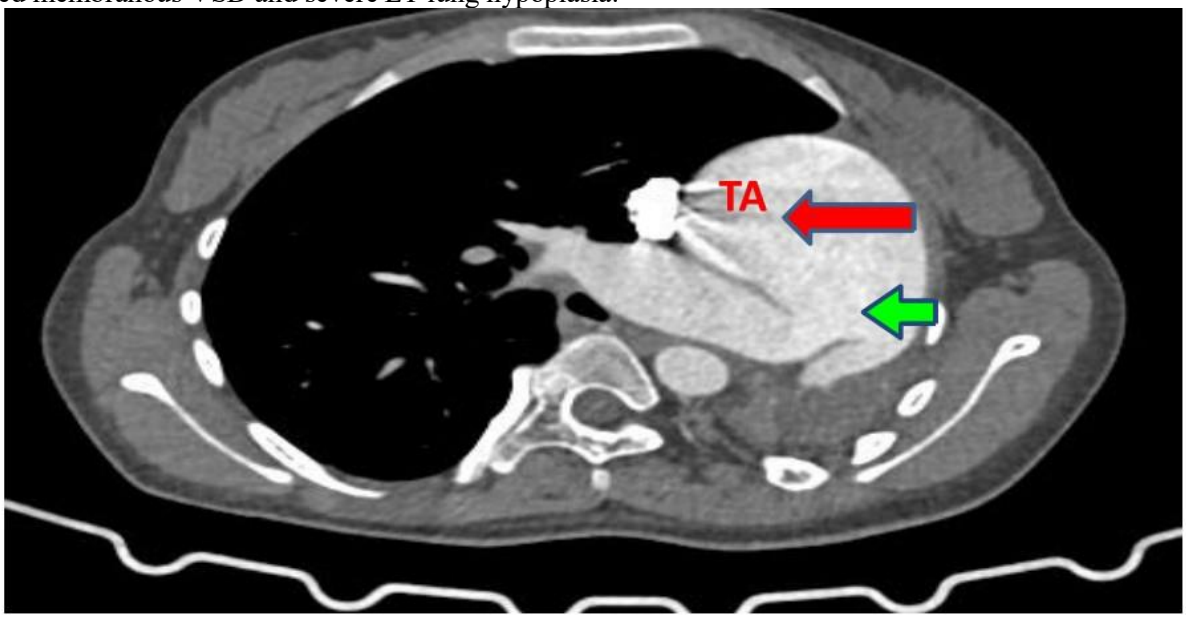


Fig 4A-CTPA showing origin of RT & LT pulmonary Arteries from posterior aspect of common trunk (Green Arrow).

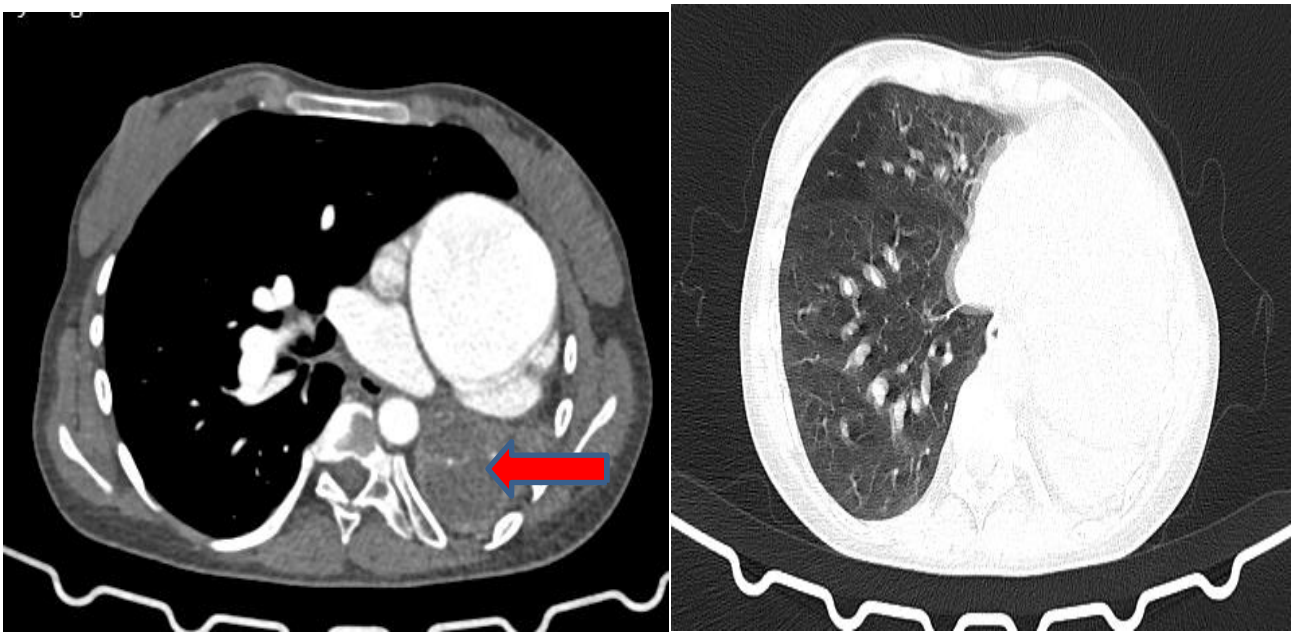


Fig 4B & 4C-Severe Left lung hypoplasia with malformed multiloculated mass.

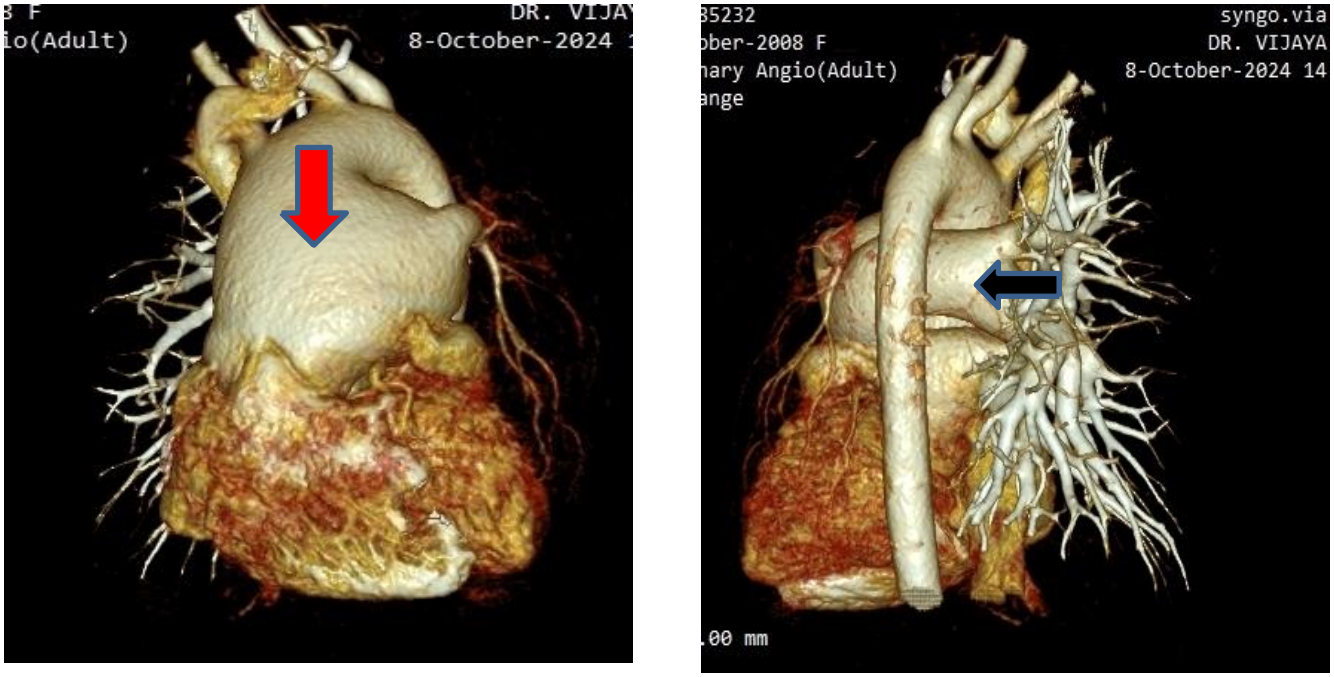


Fig 5A & 5B -Volume rendered 3D CT showing common trunk (Red Arrow) & origin of Right Pulmonary artery from posterior part of trunk(black arrow).

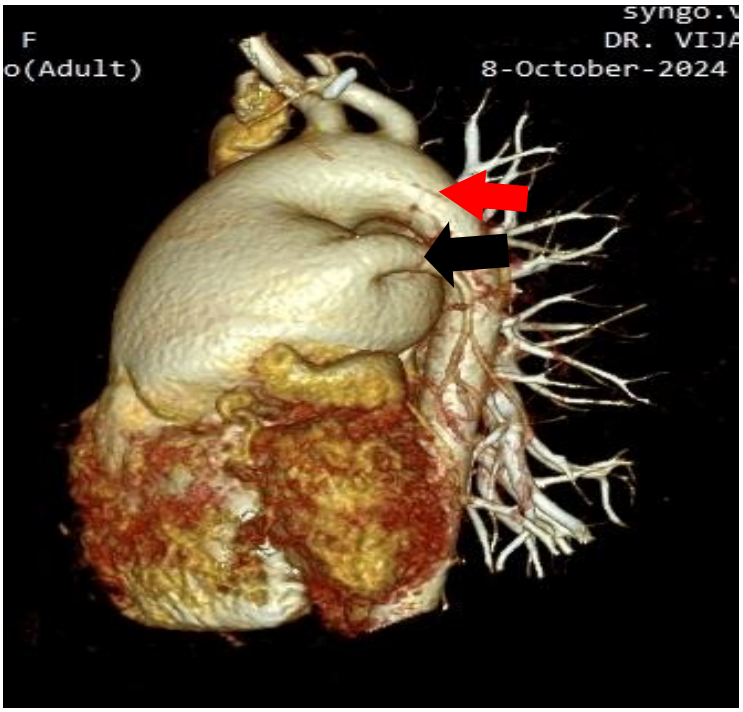
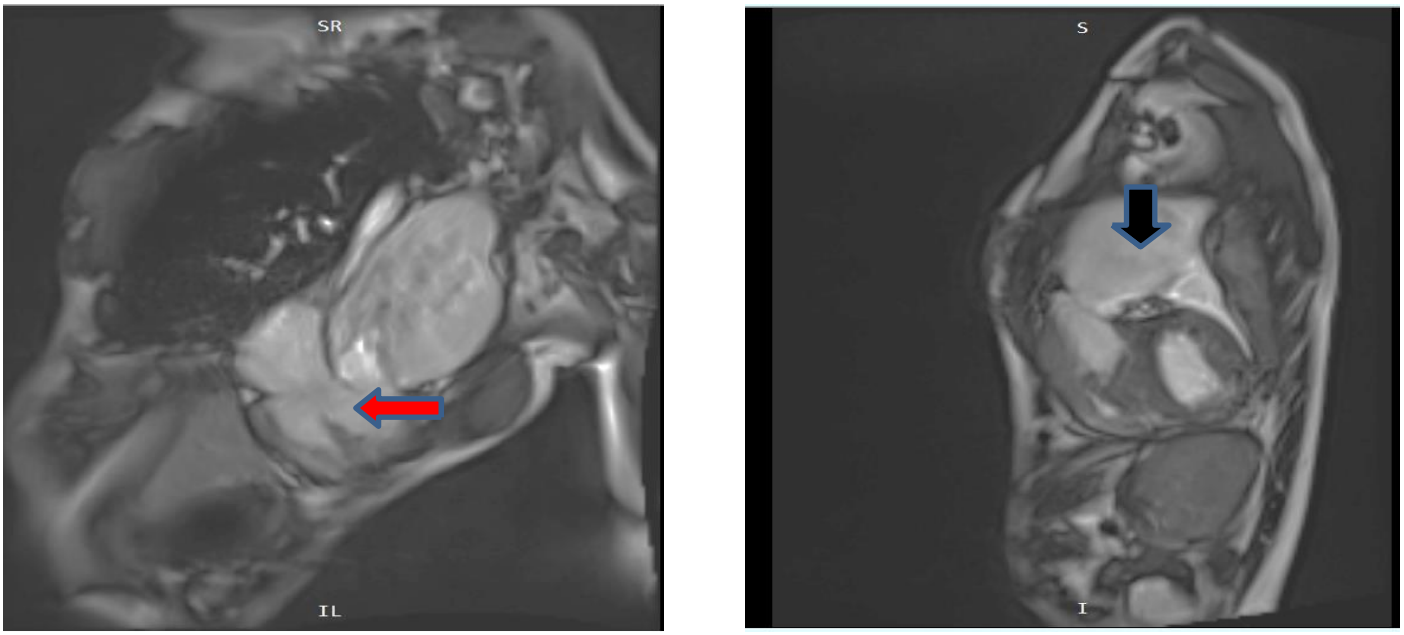


Fig 5C-VR CT showing normal Aortic arch(Red Arrow) and Origin of Left Pulmonary artery(Black arrow).



Cardiac MRI images 6 A & 6 B showing Interventricular Septal defect (Red Arrow)& common arterial trunk over riding IV septum (black arrow).

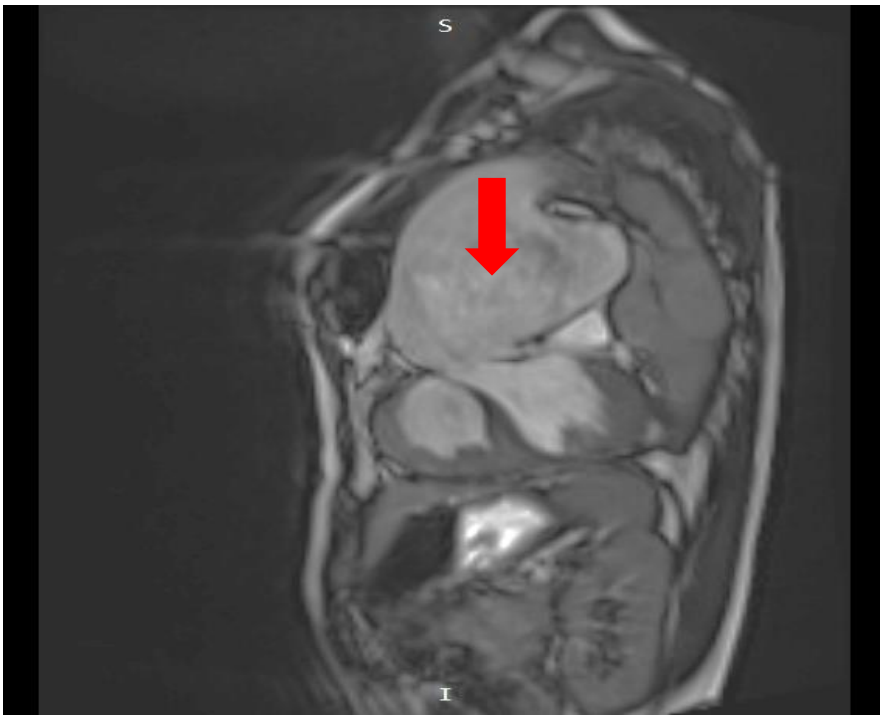


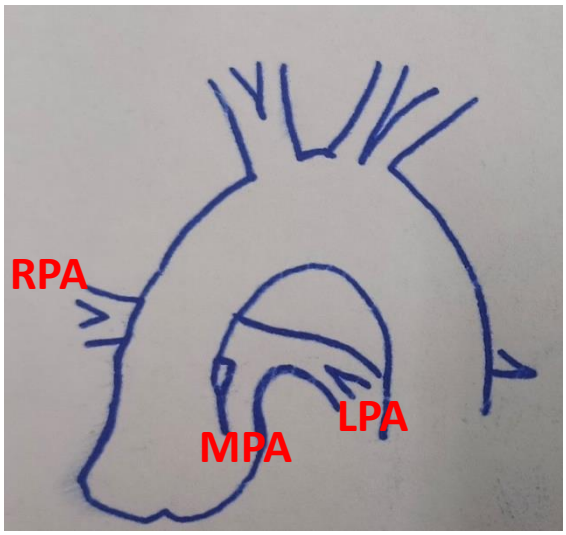
Fig 6C shows Common arterial trunk

DISCUSSION:

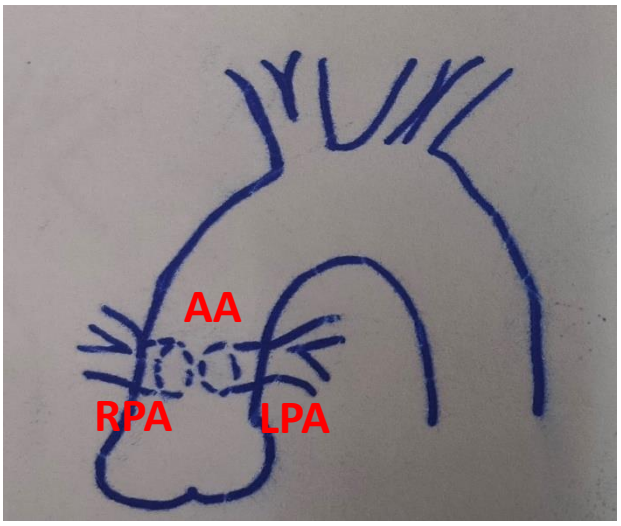
Truncus Arteriosus (TA), synonymous with common arterial trunk and Common aortico-pulmonary trunk accounts for 0.7-1.4% of all congenital heart diseases in live born infants (incidence of 0.03-0.056/1000 live births). In the embryological development process, at the 4th week, the truncus arteriosus is divided into aorta and pulmonary truncus. 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(Di George syndrome), maternal diabetes mellitus in pregnancy and teratogens such as retinoic acid and bisdiamine have been implicated.

TA is frequently associated with other cardiac and great vessel anomalies which are present in 34.8% cases, such as Right aortic arch (25-30 % cases), 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. Majority of extra cardiac anomalies are associated with CATCH22 syndrome which is present in 30-35% patients with TA . 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 and asplenia.

Classification:
Collett and Edward's (1948) classified TA into 4 types [vii]



TYPE 1



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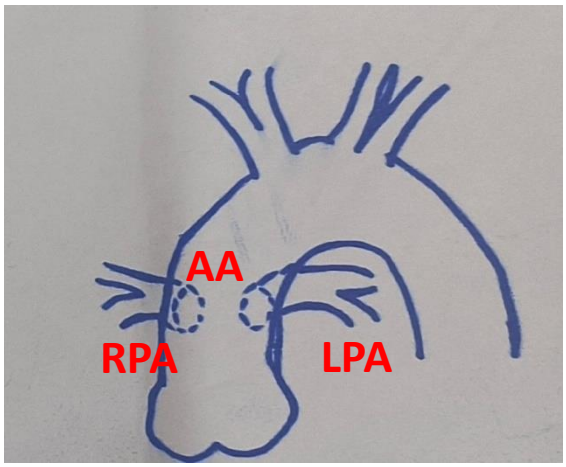

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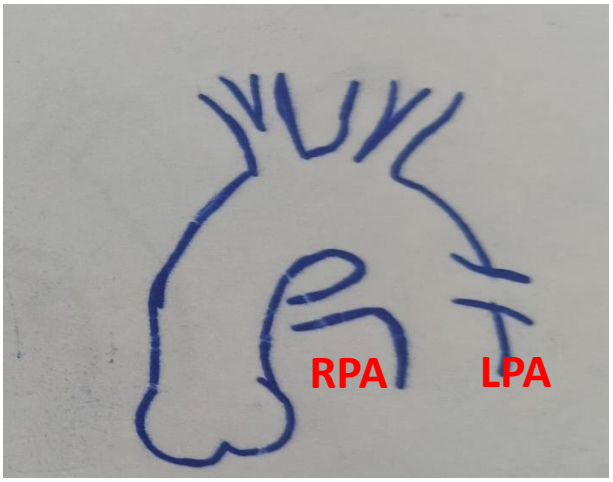


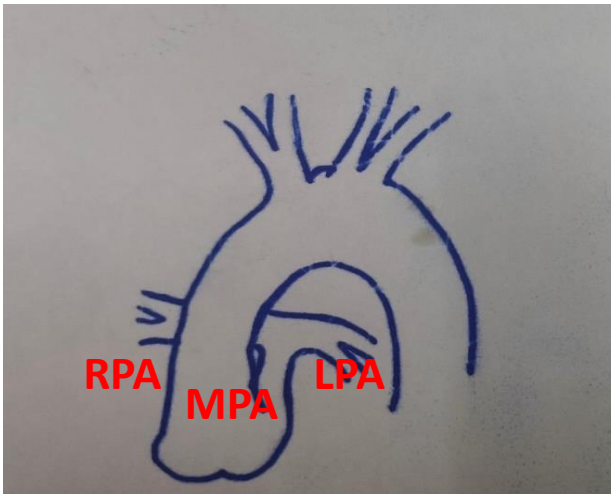
TYPE IV

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

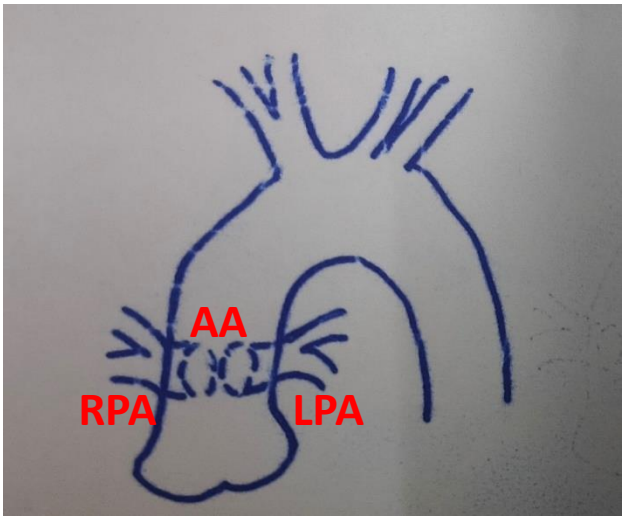
Type II: Pulmonary arteries arise separately from the dorsal wall of trunk, close to each other just above the truncal valve

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

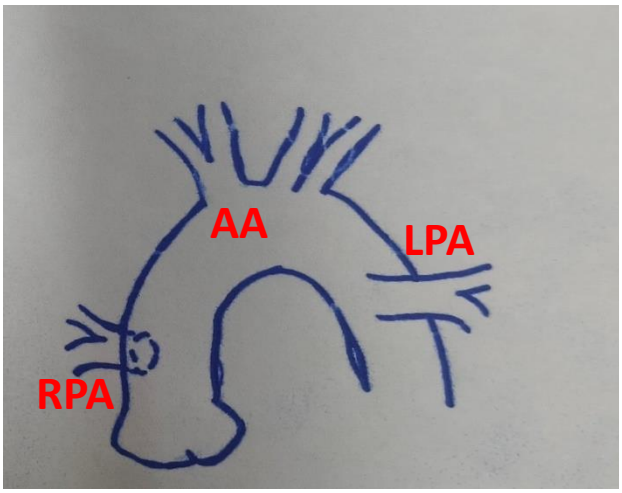
Type IV: also called Pseudo truncus, pulmonary arteries are absent and the pulmonary circulation is supplied by MAPCA's arising from the descending aorta. currently considered a form of pulmonary atresia with a VSD.



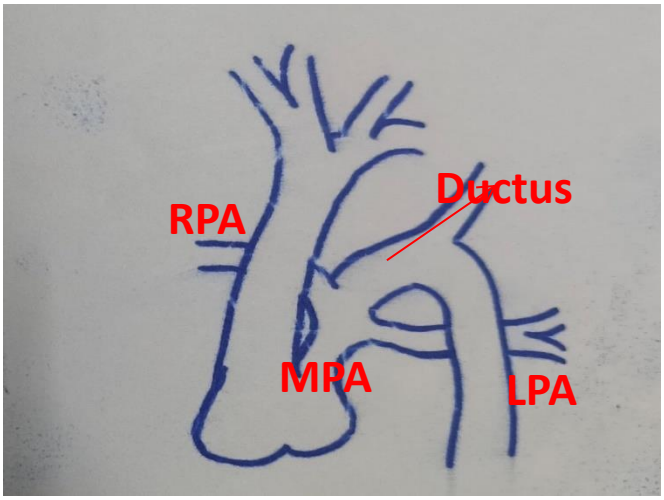
Type A1



Type A2



Type A3



Type A4

Van Praagh (1965) classification: [viii]

Type A1: Aorta and main pulmonary artery share common arterial trunk. - 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 trunkType A2 includes types 2 and 3 of Collett and Edward's.

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 . Condition should be differentiated in the neonatal period from other congenital heart diseases causing early heart failure with absent or mild cyanosis and neonatal sepsis. Chest radiograph findings [ix] include 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. In cases with an absent pulmonary artery, the pulmonary

vascular pattern is diminished on that side. Pulmonary plethora is seen. On Echocardiography the origin of the pulmonary artery from common trunk is best observed in parasternal short axis view. 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. Cardiac catheterization with angiography is indicated when pulmonary vascular disease is suspected and to define great vessels and coronary artery anatomy.
Sagittal and Transverse MRI Images at the base of the heart can demonstrate a large common arterial trunk over riding the interventricular septum and the origin of the pulmonary arteries. Coronal or oblique images are useful for determining the size and location of the VSD. Prenatal diagnosis of truncus has been reported by fetal echocardiography, karyotyping for Band 22q 11 deletion should be done.
Prognosis is poor without treatment. Corrective operations [x] (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.

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 as the disease carries poor prognosis if untreated.

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