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Truncus Arteriosus Type 1 : A case Report

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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. Pulmonary arteries originate from the common arterial trunk distal to the coronary arteries and proximal to the first brachiocephalic branch of the aortic arch [1]. TA typically overrides a large outlet Ventricular septal defect (VSD). We observed a 16 year old boy with TA Type 1 with a hypoplastic left pulmonary artery, hypoplastic left lung and a Major aorto-pulmonary collateral artery (MAPCA) supplying the right lung. In view of the rarity of this condition it was considered worthy of report.

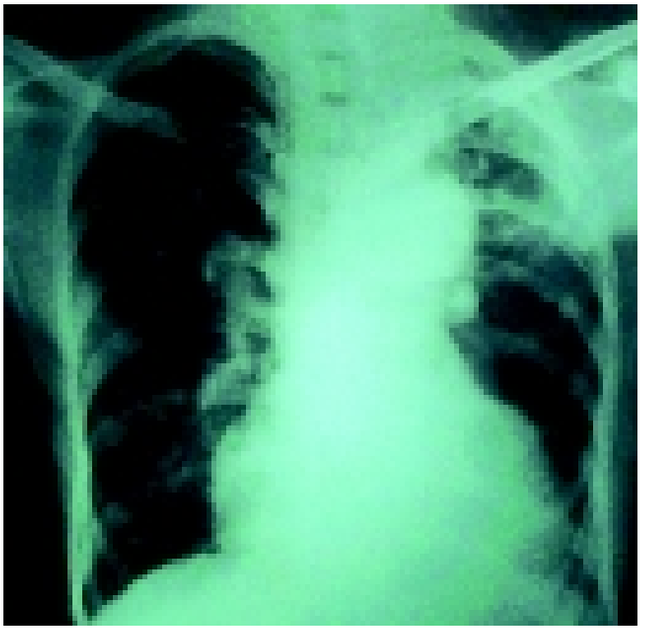

Fig 1

Case Report

A 16 year old boy was referred to us with complaints of shortness of breath and fatigue on exertion for 4-5 years and palpitations for 3 years. He gave a past history of lower respiratory tract infections since infancy. The patient was cyanosed and had grade 2 clubbing and a continuous murmur in the right parasternal area.

A chest radiograph of the patient showed hypoplasia of the left lung, an elevated right hilum with absence of hilar comma on the left side, notching of the left 5th -9th ribs posteriorly and cardiomegaly with a widening of the mediastinum due to a large aortic shadow. A Patch of consolidation was seen in the upper zone of left lung fields (Fig.1).

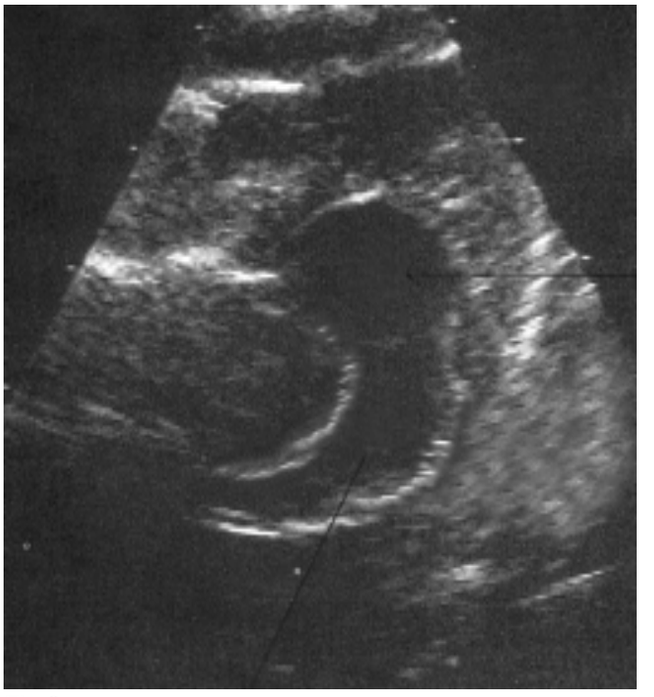

Fig 2

Echocardiography on high parasternal shortaxis view showed the origin of the right pulmonary artery from the posterior aspect of the Truncus. The left pulmonary artery was not visualized (Fig.2). The parasternal long axis view

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revealed a large subarterial VSD. The LV was dilated with normal LV systolic function. There was moderate truncal valve regurgitation. One MAPCA was visualized with continuous flow.

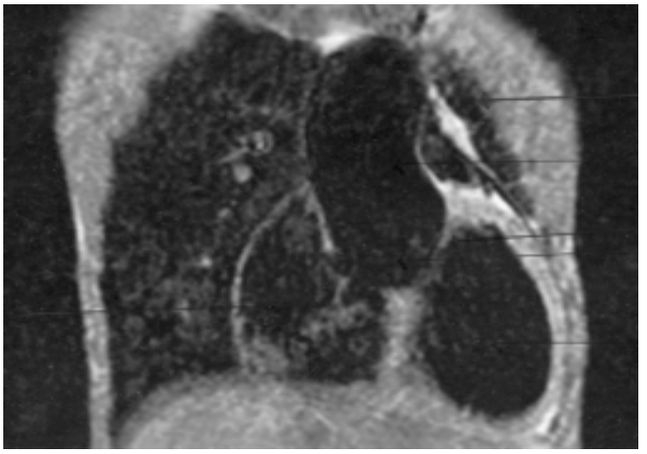

Fig 3

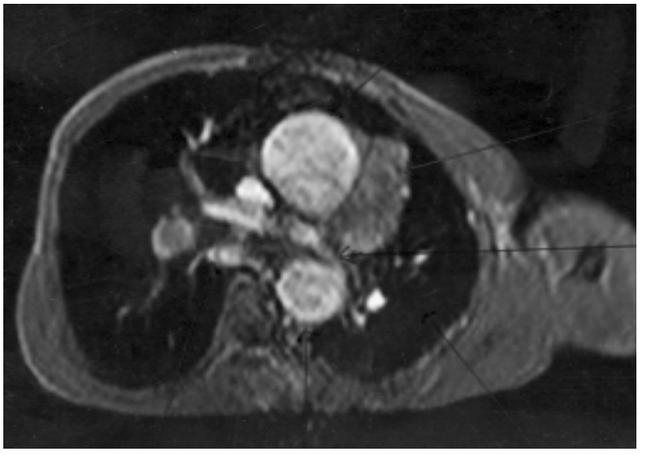

Fig 4

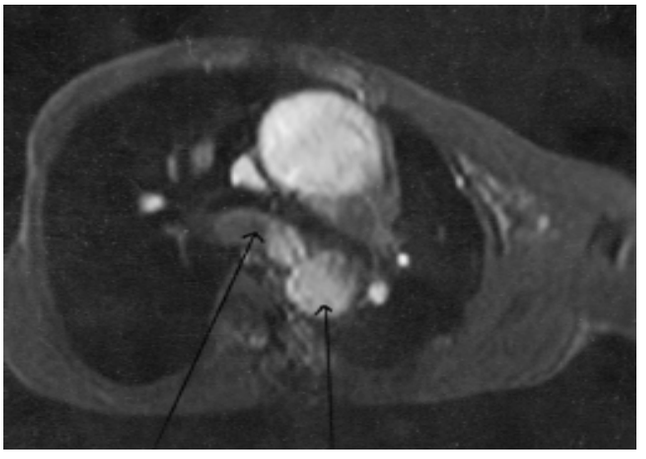

Fig 5

Coronal MRI T1W SE (ECG gated) sequences showed a common trunk overriding a membranous VSD (Fig.3). Axial MRI T1W SE sequences (ECG gated) showed the origin of the pulmonary trunk from the left posterior aspect of the truncus. The right pulmonary artery was seen to arise from the pulmonary trunk normally. The left pulmonary artery and left lung were hypoplastic (Fig.4). One MAPCA was seen arising from the descending aorta and supplying the right lung (Fig.5). Cardiac catheterization and angiography could not be done as the general condition of the patient did not permit. Karyotyping was negative.

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) [1]. There is no striking sex difference in its incidence although most series contain more males than females [1]. 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 [1].

TA is frequently associated with other cardiac and great vessel anomalies which are present in 34.8% cases [2], such as Right aortic arch (25-30 % cases [4]), 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 [2]. Majority of extra cardiac anomalies are associated with CATCH22 syndrome which is present in 30-35% patients with TA [1]. 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 [1,2,3].

Two classifications have been proposed for TA. Collett and Edward's (1948) classified TA into 4 types [2]. 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 [4], 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 [2].

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 [3]. 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 aorticopulmonary 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 [1]. The truncus consists largely of the main pulmonary artery component with a large PDA supplying the descending aorta [3]. In these cases a well documented association with Di George syndrome is observed [1]. TA type 1 and type 2 are the most common forms. Type 3 is least common [5].

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

ECG is non specific [1]. Chest radiograph findings depend on the hemodynamic circumstances [4]. 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 [4, 5]. In cases with an absent pulmonary artery, the pulmonary vascular pattern is diminished on that side [3]. A Right aortic arch is common.

On Echocardiography the origin of the pulmonary artery orifices are best observed in parasternal short axis view [6]. 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 [1], but may have up to six leaflets [6] and is frequently incompetent and/or stenotic. The posterior truncal wall is seen in fibrous continuity with the anterior mitral leaflet [6]. 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 [7]. Prenatal diagnosis of truncus has been reported [2] and when it is suspected by fetal echocardiography, karyotyping for Band 22q 11 deletion should be done.

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 [4].

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