Case Report

Survival of truncus arteriosus in 5th decade - a case report

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Abstract Persistent Truncus arteriosus is congenital cyanotic heart disease characterised by a single great artery that leaves the base of heart and gives rise to coronary, pulmonary and systemic arteries. Majority of patients die of congestive heart failure before their first birthday .Only occasional patients survive in adulthood. We are presenting such a case
Keywards: Truncus arteriosus , Pulmonary vascular Resistance.

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CASE REPORT

A 43 year old female was referred to our institute for exertional dyspnoea and cyanosis. She gave history of having been diagnosed with some cardiac anomaly at the age of six months. There was history of poor weight gain and recurrent episodes of pneumonia till the age of three years. She was subsequently well in adolescence and in early adulthood and was even able to participate in sports without significant discomfort. This continued till the age of fourty three, when she developed exertional dyspnoea with deepening of fixed bluish discolouration of tongue and extremities. Clinical examination revealed an averagely built, poorly nourished female with central cyanosis and clubbing. Her pulse was irregular, blood pressure was 110/ 40 mm Hg in upper limbs JVP was raised with absent a waves. There was evidence of right heart failure in the form of pedal edema, icterus and right hypochondriac tenderness. Cardiovascular examination revealed an apex localised to left 6th intercostal space in the anterior axillary line, hyperdynamic in nature with a diastolic thrill over the apical area. Auscultation revealed a single loud second heart sound with a long diastolic murmur in the apical region. ECG showed atrial fibrillation with right ventricular hypertrophy. 2 D echo showed evidence of truncus arteriosus with subaortic 21 mm VSD with bidirectional shunt with moderate truncal regurgitation with moderate biventricular function. There was a mass in RA which was confirmed to be a thrombus on cardiac CT. Hence we reached the diagnosis of Type I truncus arteriosus with moderate truncal regurgitation with moderate biventricular function. The patient was started on anticaoagulation and was advised to undergo truncal valve replacement. The patient was investigated in the form of routine blood investigations, ECG, 2D ECHO, Chest X Ray, Trans esophageal echo (TEE),

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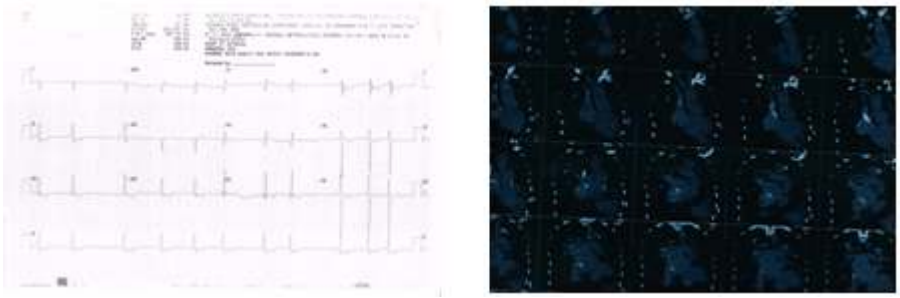

Figure 1: Cardiac CT

Figure 2: Cardiac CT

The patient was put on Digoxin, Diuretics, Vasodilators for cardiac failure and anticoagulation with Warfarin in view of right atrial thrombus. We would like to point out that this is an exceedingly rare example of survival of isolated truncus arteriosus into the fifth decade.

DISCUSSION AND RESULTS

Persistent truncus arteriosus is defined as a single trunk arising from the heart and supplying the coronary, pulmonary, and systemic circulations, with no remnants of an atretic aorta or pulmonary artery. The number of semilunar cusps usually varies from 2 to 6, commonest being three. Typically a defect is present in the bulbar portion of the anterior ventricular septum, a defect lying just under the semilunar cusps. This must be differentiated from the usual ventricular septal defect involving or bordering on the membranous portion of the septum. The reported incidence of truncus arteriosus (TA) ranges from 6 to 10 per 100,000 live births. Although only 0.7 percent of all congenital heart lesions are due to TA, it accounts for 4 percent of all critical congenital heart disease cases. The classifications most commonly used are:

1. Collett/Edwards and
2. Van Praagh schema.

Collet and Edwards classification — The Collet and Edwards. classification, proposed in 1949 and considered to be the most simple system, consists of:

- Type I: a main pulmonary artery is present, and arises from the left side of the truncal root.
- Type II: the right and left branch pulmonary arteries have two closely-spaced but separate origins from the posterior aspect of the truncal root.
- Type III: the branch pulmonary artery origins are widely separated from the truncal root.
- In the original type IV, the pulmonary arteries originate from the descending aorta. Patients in this category, however, are now considered to have a different form of congenital heart disease called pulmonary atresia.
- Van Praagh classification — The Van Praagh (VP) classification is similar to the Collet and Edwards (CE) schema; however, it also includes aortic abnormalities and unilateral pulmonary atresia, which are of particular importance for surgical repair.
- Type A1 is the same as CE type 1, in which a main pulmonary artery is present, and arises from the left side of the truncal root.
- Type A2 includes both CE types II and III, as the two CE types do not differ embryologically and the surgical approach is the same. Type A2 consists of right and left branch pulmonary arteries with separate origins (regardless of the distance separating the two pulmonary arteries) from the truncal root.
- In type A3, there is unilateral pulmonary atresia with collateral supply to the affected lung.
- In type A4, the truncus is associated with an interrupted aortic arch.
- STS modification — In 2000, the following modification of the Van Praagh schema into three categories was proposed by members of the STS Congenital Heart Surgery Database

committee and European Association for Cardiothoracic Surgery. This classification is primarily used by pediatric cardiovascular surgeons.

- TA with confluent or near confluent pulmonary arteries (large aorta type, VP types A1 and A2)
- TA with absence of one pulmonary artery (VP type A3)
- TA with interrupted aortic arch or coarctation (large pulmonary type, VP type A4)

In patients with TA, mixing of systemic and pulmonary system occurs due to the presence of a single large vessel draining both ventricles. The pathophysiology and therefore the clinical manifestations and outcome of TA depend on the volume of pulmonary blood flow determined by the pulmonary vascular resistance, the degree of truncal valve insufficiency (regurgitation), and whether there are significant aortic anomalies.) At birth, there is little cardiovascular instability in patients with TA, due to the persistence of high fetal pulmonary vascular resistance (PVR) which restricts pulmonary blood flow. These patients may present with cyanosis, which may be detected by pulse oximetry screening. With the normal fall of PVR in the first weeks of life, flooding of the pulmonary system leads to pulmonary oedema and heart failure. In these infants, clinical features include a history of poor feeding and lethargy, signs of respiratory distress (tachypnea, costal-sternal retractions, grunting, and nasal flaring), tachycardia, hyperdynamic precordium, and hepatomegaly. Cardiovascular findings of TA consist of those due to a single valve draining the ventricles (i.e. a single loud second heart sound), and due to truncal regurgitation (leading to systolic ejection murmur at the left sternal border, and bounding peripheral pulses). One of every four to five cases of TA appears to be associated with 22q11.2 deletions, which are seen in patients with DiGeorge syndrome. These patients may present with clinical features of DiGeorge syndrome including hypocalcemia due to parathyroid hypoplasia, hypoplastic thymus, and palatal abnormalities (eg, cleft palate). Uncorrected patients with TA have a poor outcome, with a survival rate of only 15 percent at one year of age. Subsequently, survivors beyond one year of age develop severe pulmonary vascular obstructive disease (i.e., Eisenmenger syndrome) with profound cyanosis and functional impairment. Initial laboratory testing includes an unspecific electrocardiography and chest radiography with typical findings of cardiac enlargement and increased pulmonary vascular markings. Other chest radiographic findings may include a right aortic arch in one-third of patients, or an absent thymus (which is associated with DiGeorge syndrome). TA is diagnosed by echocardiography and by cardiac CT by identifying a single overriding great vessel arising from the heart. Although the diagnosis can be made antenatally, it is often difficult to distinguish TA from other conotruncal cardiac lesions. The differential diagnosis of TA includes other congenital cardiac conditions that cause early heart failure such as large ventricular septal defect (VSD), pulmonary atresia with VSD, and univentricular heart. TA is distinguished from these other cardiac diseases by echocardiography. Primary surgical repair in the neonatal period has improved overall survival despite a perioperative mortality of 10 percent. As a result, we recommend that all patients with TA undergo primary surgical repair during the neonatal period (Grade 1B). Reintervention is common and includes replacement of right ventricle to pulmonary artery, and repair or replacement of truncal valve. Prior to surgical repair, initial medical management includes stabilization of cardiopulmonary function. Patients with heart failure generally require care in the intensive care unit that includes diuretic therapy, ionotropic support, angiotensin blockade, and positive pressure ventilation. In addition, for patients with TA and critical coarctation or interrupted aorta, we recommend continuous infusion of prostaglandin E1 (also known as alprostadil) to maintain patency of the ductus arteriosus and avoid life-threatening heart failure prior to surgical repair (Grade 1B). Regular follow up is necessary to detect right heart failure early, and initiate treatment. Many patients will eventually require surgical correction of the tricuspid valve.

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