Truncus arteriosus Type ii

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Background: Truncus arteriosus (TA) is an uncommon congenital cardiovascular anomaly that is characterized by a single arterial trunk arising from the normally formed ventricles by means of a single semilunar valve (ie, truncal valve).

Case presentation: A 3-month-old female infant product of spontaneous vaginal delivery complaining from dyspnea, tachypnea, mild cyanosis since birth, decrease in feeding and activity. She was admitted twice in different hospitals and diagnose as a case of VSD with pulmonary hypertension, given treatment but no response, the family picked her to Babylon for maternity and children hospital. ECHO and cardiac CT-angiography was done were revealed truncus arteriosus.

She was treated by anti failure and drug which lower pulmonary vascular resistance, her condition improve and become stable. after that they went to turkey where operation was done in specialized cardiac centre before age of six month.

Conclusion: In any infant with signs and symptoms of congestive heart failure and mild cyanosis truncus arteriosus shoud be suspected.

Corrective operation should be done in first six month of age

Truncus arteriosus (TA) is a rare cardiac disorder¹ accountant of about (0.7%-1.4%) of all congenital heart disease, in which aorta and pulmonary artery were not separated completely during fetal development and both originate jointly from left ventricle². It has an incidence of about 0.3/10,000 live birth, no striking sex difference although most series contain more male than female³⁴.

Etiology is multifactorial and it may be associated with microdeletion chromosome 22q11.2, maternal diabetes has been implicated as a risk factor for truncus arteriosus⁵. The number of truncal valve cusps varies from 2 to as many as 6 and the valve may stenotic, regurgitant, or both⁶. Its had been classified by Collet and Edwards in 4 different types in 1949.

Type I in which the pulmonary arteries can arise together from the posterior left side of the persistent truncus arteriosus and then divided into left and right pulmonary...
arteries, in type II and III, no main pulmonary artery is present, and the right and left pulmonary arteries arise from separated orifice on posterior (type II) or lateral (type III) aspect of truncus arteriosus. Type IV truncus is a term no longer used, this essentially a form of pulmonary atresia.

During past 20 years, diagnostic and therapeutic methods along with immediate intervention for surgery in infancy with complete reparation of VSD lead to evident improvement and less morbidity in newborns. Truncus arteriosus can be diagnosed in utero by fetal echocardiography.

Case presentation: A 3-month-old female infant, product of normal vaginal delivery, in a consanguineous marriage, she was admitted because of repeated chest infection, difficulty in breathing and feeding with mild cyanosis since birth, ECHO (echo cardio graphy) was done and was diagnosed as ventricular septal defect (VSD) and pulmonary hypertension, treatment was given but without benefit and patient still complaining, then they consult another cardiologist with the same result, so they came to our hospital (Babylon for maternity and children hospital).

On examination. The Patient had mildly cyanosis, dyspneic, tachypnic, Pulse rate=120 beat/min, bounding peripheral pulse.

Respiratory rate=62 breath/min, SPO2=90% at room air, no dysmorphic feature, Weight=3.8 Kg, length=53 cm,

Cardiovascular system: normal S1, loud S2 and single, on right side loud pansystolic murmur maximal at lower right border radiate to entire pericardium, grade 4/6 systolic thrill, apex beat on right side at 5th intercostals space, chest bilateral fine crepitation.

Abdomin: Soft with mild hepatomegaly

Work up: complete blood picture; all indices are normal, serum electrolytes were normal, Liver & renal function tests, plasma glucose all were normal.

CXR (Chest X ray): Dextrocardia, cardiomegaly. (plethoric lung) as shown in figure (1).

Figure (1): CXR, A-P View

ECG (Electrocardiography): sinus tachycardia, right axis deviation, biventricular hypertrophy. ECHO: situs solitus, dextrocardia, biventricular hypertrophy (RVH), (LVH), dilated right atrium (RA).

Large atrial septal defect (ASD) secondum, Large subtruncal ventricle septal defect (VSD) with left to right shunt. Single trunk with mild incompetence quadrivalent valve with both pulmonary arteries arise from
posterior aspect of trunk (truncus (A,B,C) ,Severe pulmonary hypertension arteriosus type II) as shown in figure 2 .

Figure 2 A

Cardiac CT-angiography: Reveal dextrocardia, large ASD secondum, large subtruncal VSD, single trunk arise from both ventricles, absence of pulmonary artery from right ventricle, both pulmonary arteries arise from posterior aspect of the trunk , as shown in figure (3) .
Figure (3): cardiac CT-angiography

So the diagnosis was truncus arteriosus type II and patient received antifailure treatment. Total surgical correction was done at age of six month in turkey.

**Literature review**: Persistent truncus arteriosus is an uncommon congenital cardiovascular malformation. Truncus arteriosus usually occurs as an isolated cardiovascular malformation, although on occasion it has been reported in association with anomalies of other systems, particularly the DiGeorge or velocardiofacial syndrome (microdeletion chromosome 22q11.2) (8,9). During the last 25 years, surgical correction of truncus arteriosus during infancy has become routine (10). Among 400 cases of truncus arteriosus from four publications reviewed by Fuglestad et al. (11), the truncal valve was tricuspid in 277 (69%), quadricuspid in 86 (22%), bicuspid in 35 (9%), pentacuspid in 1 (0.3%), and unicommisural in 1 (0.3%). The semilunar valve is in fibrous continuity with the mitral valve in all patients but is continuous with the tricuspid valve in only a minority.

Of the Mayo Clinic's previously published series of patients with truncus arteriosus, 16% (11 of 70) had only a single pulmonary artery (13). In 9 of the 11 patients, the pulmonary artery was absent on the side of the aortic arch. Thus, in truncus arteriosus, the pulmonary artery is most frequently absent on the side of the aortic arch, in contrast to tetralogy of Fallot, in which the pulmonary artery is more frequently absent on the side opposite the aortic arch. Patients with truncus arteriosus are at risk of having pulmonary vascular obstructive disease develop at an early age, and this has provided the major impetus for early surgical correction (14), however, pulmonary vascular disease tends to progress postoperatively more often in patients with single pulmonary artery than two artery (15).

This hazard of ventricular dysfunction may explain in part the observation that repair of truncus at 6 to 12 months of age is associated with a mortality twice that for repair between 6 weeks and 6 months of age (16). The natural history of this condition is generally dismal. one series reported a survival of only 15% beyond the age of 1 year. Death is caused by heart failure, the complications of hypertensive pulmonary vascular disease and infective endocarditis. so early surgical intervention is advocated for these patients (17).

**Discussion**: Truncus arteriosus is a rare anomaly, defined as a single great artery that originates from the base of the heart and gives rise to the
pulmonary, systemic, and coronary circulation. It is frequently associated with other cardiac anomalies including: VSD, aortic arch abnormalities, single ventricle, atrioventricular valve abnormalities, absent ductus arteriosus, and abnormal pulmonary venous return. Extracardiac anomalies occur in up 20% to 50% of the cases and include: situs inversus, asplenia, bony defects, cleft lip/palate, urinary tract abnormalities, absent gallbladder, hypoplastic lung, and neural tube defects. Patient in early infancy developed signs of heart failure, tachypnea, tachycardia, and failure to thrive. In this case with truncus arteriosus and increased pulmonary blood flow, may have to differentiated the condition from the other congenital cardiac conditions that cause early heart failure and are associated with either mild or no cyanosis. Such malformations include ventricular septal defect (Which is common cardiac lesions so the diagnosis didn't settled early), patent ductus arteriosus, aorticopulmonary window, pulmonary atresia with ventricular septal defect and patent ductus arteriosus, or large collateral arteries, double-outlet right ventricle, univentricular heart, and total anomalous pulmonary venous connection.

- In any infant with mild cyanosis and signs and symptoms of congestive heart failure Truncus arteriosus should be suspected even it is a rare congenital cardiac malformation
- Operation should be done in first six month of age to avoid changes of pulmonary vascularity.

**Recommendation:**

1- It is crucial to diagnosis truncus arteriosus early in life to refer suspected case to tertiary cardiological centre.

2- Surgical operation should be done as early as possible because delayed after six month of age increase mortality and morbidity.

3- Good preparation before surgery by using antifailure treatment and drug that lower pulmonary vascular resistance.

4- Follow up the patient after surgery by pediatric cardiologist looking for development of any complications.

**References**


