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RESEARCH ARTICLE

TRUNCUS ARTERIOSUS TYPE 1 A RARE CONGENITAL HEART DISEASE

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ABSTRACT

The truncus arteriosus or common arterial trunk, is a form of congenital cyanotic heart disease where only one artery arises from the heart, being responsible for the systemic, pulmonary and coronary circulation. It occurs in 1.5% of cases of congenital heart defects in newborns, presenting variations in their presentation as to the origin of the pulmonary trunk. We present a case of 6 month old male boy who presented with complains of fast breathing and respiratory distress. On examination, there was no cyanosis and clubbing. A systolic murmur was heard at mitral area. Chest radiography showed B/L non homogenous opacity with cardiomegaly. Hence a clinical diagnosis of Acyanotic heart disease most probably VSD with congestive cardiac failure with pneumonia was made. But ECHO Cardiography showed Truncus arteriosus Type 1. Here we want to emphasize that truncus arteriosus, though classified as a cyanotic heart disease may present without cyanosis in late infancy.

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INTRODUCTION

Truncus Arteriosus (TA) is a rare*(<1.5%),* 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 (Mittal *et al*., 2006). TA typically overrides a large outlet Ventricular septal defect (VSD). Etiology is mostly genetic, 30-40% due to 22q11.2 microdeletion (Volpe *et al*., 2003) and rest due to environmental factors such as maternal 1st trimester exposure to alcohol, viral infection, dyes and dietary deficit in vitamins. About 34.8% of the patients are associated with cardiac and other extracardiac anomalies that is, right aortic arch 25–30% cases, interrupted aortic arch, aberrant right subclavian artery, abnormal coronary arteries, residual VSD, proper functioning of right ventricle to pulmonary artery conduit and global myocardial dysfunction (Freedom, 1992) Four types of are recognized. The most common variety is Type -1 which is characterized by short main pulmonary Artery that originate from truncus and give rise to Right and Left Pulmonary arteries. In Type – 2 Pulmonary artery arise from a separate ostia from the side of Truncus and in Type -3 Pulmonary arise from a separate ostia from back of Truncus. In Type -4 Pulmonary artery is absent or Atretic (Pathak and Singh, 2013). Truncal valve could be Quadricuspid (40-50%), bicuspid, Tricuspid or Hexa cuspid. Left Coronary artery arise from left posterior aspect of Truncus, right Coronary artery arise from right anterior aspect of Truncus. VSD is always Nonrestrictive.

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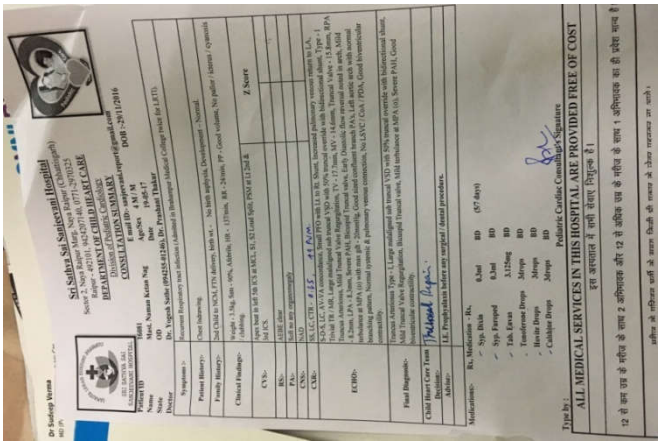
Case report

A 6 month old male baby was admitted to paediatric department of MKCG with complains of fever, fast breathing and respiratory distress for 7 days. There was past history of bluish discolouration during excess crying. The baby was born out of non consanguineous marriage by normal vaginal delivery. There was no history of birth asphyxia. The baby had not attained neck control. On examination there was no cyanosis and clubbing (Fig 1). There was tachypnea (64/min) and B/L crepitations. The Systolic murmur maximum at left lower 4th intercostals space with radiation upward to right. The chest radiography showed B\L non homogenous opacity with cardiomegaly. Echo cardiography showed Truncus arteriosus type 1, large VSD with 50% truncal overload with bidirection shunt, bicuspid truncal valve and severe Pulmonary hypertension (Fig 2) The parents were counseled about the prognosis and sent to higher center for corrective surgery after a course of intravenous antibiotics iv ceftriaxone for pneumonia.



DISCUSSION

Patients with truncus arteriosus have some degree of cyanosis during the first week of life. Congestive heart failure usually occurs by a few weeks of age. Excessive pulmonary blood flow at high pressure results in pulmonary vascular obstructive disease by 3 months. The diagnosis of truncus arteriosus is suspected in newborns with mild cyanosis, a cardiac murmur, and pulmonary overcirculation. Factors that limit pulmonary blood flow, such as pulmonary artery stenosis or persistently elevated pulmonary vascular resistance, may delay the appearance of symptoms (Mann *et al*., 2014) Systolic murmur across VSD becomes evident as pulmonary vascular resistance falls.There is decrescendo Systolic murmur maximum at left intercostals space with radiation upward to right. When pulmonary vascular resistance increases, the shunt across VSD is reduced and soft mid systolic murmur emerge across the truncal valve. Second heart sound is loud and single. ECG shows biatrial P wave and varying degree of ventricular hypertrophy. The diagnosis is established by echocardiography and cardiac catheterization (Van Praagh and Van Praagh, 1965). The only definitive treatment for this anomaly is surgical correction. 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 (Braunwald *et al*., 2001).

Conclusion

Making accurate diagnosis of congenital heart disease like truncus arteriosus can be quite challenging unlike the developed world. Also important is the issue of repairing the complex types because of the dearth of requisite personnel as well as the equipment. In management, surgery should be done at the earliest and should be followed up regularly for conduit patency and Pulmonary Hypertension.

Contributors

Dr.Nasreen Ali-conception,design and drafting
Dr.Sunil Kumar Agarwalla-revising it critically for important intellectual content.

Conflict of interest

There was no conflict of interest and no funds received.

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