

Single Coronary Artery from Single Sinus in Complete Transposition of Great Artery: An Exceptional Case

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Abstract

Complete Transposition of great arteries (D TGA) characterizes by Ventriculo-arterial discordance, along with AV concordance. In D-TGA, Aorta arises from morphologic right ventricle and Pulmonary artery arises from Left ventricle. In new born and infants this is a potentially lethal form of heart disease. The incidence of D-TGA is 1 in 2300 to 1 in approximately 5100 live birth. Ninety percent of d TGA are associated with dual sinus origin of coronaries, in this singular case, the baby had single sinus origin of coronary artery. In this exceptional study, a new born baby girl of 16 day age who could not maintain her oxygen saturation, found to have complete transposition of aorta, which was associated with single sinus origin of her coronary artery which indicates that both coronary arteries arise from one of the two but not both of the facing sinuses by a single ostium. Her left and right coronary and left circumflex artery arises from single coronary artery. She was taken for anatomic correction by arterial switch operation with re-implantation of coronary artery and she made excellent recovery. This case highlights that we have to be very careful to assess the coronaries thoroughly in all cases of d TGA as many cases of ischemia, infarct and LV failure have been documented in patients who were not treated appropriately with respect to coronary anatomy.

Keywords: Complete transposition of great arteries, Corrected transposition of great artery, D-TGA, Simple transposition

INTRODUCTION

Complete transposition or D-TGA is one the most fatal form of congenital cyanotic heart disease if remain untreated. Malformation consist of the origin of the Aorta from morphologic right ventricle and that of the pulmonary artery from the morphologic left ventricle.¹ Consequently, pulmonary and systemic circulation is connected in parallel rather than in series connection. In one circuit, the Systemic venous blood passes through the right atrium, right ventricle, and then to aorta. In the other, pulmonary venous blood passes through left atrium, left ventricle and to the pulmonary artery. This situation is incompatible with life unless mixing of the two circuits occurs.

The incidence of D-TGA is 1 in 2300 to 1 in approximately 5100 live birth.² The malformation represents 5-8 % of congenital cardiac malformations but accounts for 25 %

of death from congenital heart disease in the first year of life.³ Male outnumber females by a ratio of 4:1,⁴ and seldom occurs in first born infants, but a twofold increase in incidence occur in mother who have had three or more pregnancy.

Associated congenital anomalies which are seen with D TGA are Patent foramen ovale, septum secundum atrial septal defect, ductus arteriosus, pulmonary stenosis, aortic arch anomalies such as hypoplasia, coarctation, interruption of aorta and aortic arch, or tricuspid and mitral valve abnormalities.

CASE REPORT

A baby girl of 16 day old, born through normal uncomplicated delivery, got admitted on 29 September 2013 with history of respiratory distress, chest infection

and bluish discoloration of lips, fingers and tongue since birth. Baby is third sibling of her parent. Baby was consulted by her parents with pediatrician during the past 15 day and was given antibiotics for the same.

Subsequently baby got admitted as there was no improvement in her respiratory distress and cyanosis.

On examination her heart rate was 142 per minute, oxygen saturation was 40%, blood pressure was 90/60 mmHg respiratory rate was 44/minute, her birth weight was 3.6 kg there was presence of central cyanosis, on CVS examination she had central cyanosis, with full volume bounding pulse. Precordial palpation was normal, loud palpable second heart sound at the left base is felt. Mid systolic murmur heard over aortic area, mid diastolic flow murmurs of mitral and tricuspid origin⁵ were auscultated. Loud and single second heart sound of anterior place aorta was auscultated too.

Echocardiography reveals aorta arising from right ventricle, pulmonary artery arising from left ventricle with “sausage” appearance in short axis (Figures 1 and 2). The great arteries appear as double circles with aorta anterior and to the right of the main pulmonary artery. The anterior aortic root and posterior pulmonary trunk run parallel to each other and do not cross. There was small Patent ductus arteriosus which was about to close with left to right shunt and peak gradient of 13 mmHg.

There was single coronary artery arising from single sinus of left aortic sinus and gave rise to right coronary artery, left coronary artery and circumflex artery. Usually in more than 90% of DTGA, there is dual sinus origin of coronary artery (Figure 3).^{6,7}

There was no evidence of associated other congenital anomaly.

Chest X ray showed narrow aortic knuckle, increased pulmonary vascularity, and fairly normal size cardiac silhouette.

MANAGEMENT

She was treated with oxygen supplementation 3 liter/hr, fluids and antibiotics, along with this baby was given Prostaglandin E1 to keep the duct patent so that baby could overcome the crises till she get her surgery done, subsequently she was taken for surgery. Although, Balloon atrial septostomy (BAS) was considered for the patient but in view of potential complications of BAS such as neurologic/embolic stroke^{8,9} and poor exercise performance^{10,11} after

BAS in comparison to Arterial switch, the baby was taken for Arterial switch operation. Although there are reports of intra operative and post operative coronary artery kinking, ischaemia and IV dysfunction^{12,13} but overall it gives the best possible result so the baby had undergone Arterial switch and made good recovery after the surgery and got discharged postoperatively (Figures 4-7).

DISCUSSION

Complete transposition of great arteries are one of the rare congenital heart disease of new born and in DTGA, chances of three coronary artery arising from single aortic sinus is less than 10%. To suspect complete transposition of great arteries in a new born, pediatric cardiologist or pediatrician should have vast knowledge to detect even minute details of the baby profile, gender, and clinical presentation. It should be aggressively confirmed with cardiac Doppler and Chest X-ray. The clinical recognition of D-TGA is based on following features (1) Male third or fourth child, (2) large birth weight (3) Cyanosis in the neonatal period (4) Radiological feature of increased blood flow in presence of cyanosis and egg-shaped cardiac silhouette with a narrow vascular pedicle and absent thymic shadow (5) Echocardiographic identification of Aortic alignment with morphologic Right ventricle and Pulmonary alignment with morphologic left ventricle (Ventricular arterial discordance) and right atrial alignment with right ventricle and left atrium alignment with left ventricle (Atrioventricular concordance).

Some communication between the two circulations must exist after birth to sustain life; otherwise unoxygenated systemic venous blood is directed inappropriately to systemic circulation and oxygenated pulmonary venous blood is directed to the pulmonary circulation. Almost all patients have an interatrial communication such as Patent ductus arteriosus, Ventricular septal defect or Atrial septa defect.

There could be various mode of coronary artery origin in DTGA, common amongst them are Dual sinus origin, in which left coronary artery arise from left aortic sinus which in turn give rise to left anterior descending artery and circumflex artery and right coronary from posterior aortic sinus. Dual sinus origin accounts for 90% of cases. Single sinus origin indicates that both coronary arteries arise from one of the two but not both of the facing sinuses by a single ostium or multiple ostia. In this exceptional study, there was single coronary artery from left aortic sinus which in turn gave rise to Left coronary, right coronary and circumflex arteries. The incidence of this among the DTGA is less than 10%.

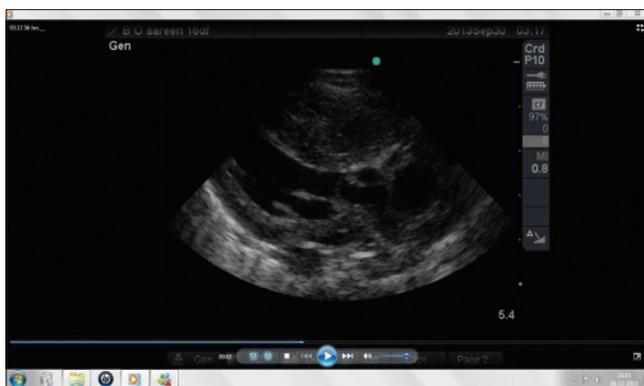


Figure 1: Echocardiography of transposition seen in long axis with great arteries side by side



Figure 2: Echocardiography of great arteries in short axis view in DTGA with sausage appearance

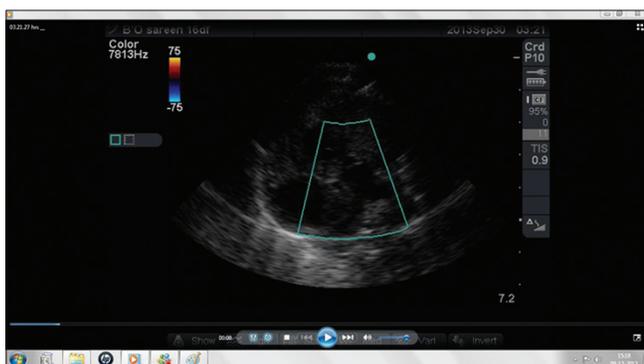


Figure 3: D-TGA in apical four chamber view with pulmonary artery arising from left ventricle and bifurcating into right and left pulmonary artery. Operative pictures of Arterial switch with single coronary artery and its branches {Left coronary artery-left main (LCA) , Right coronary artery (RCA) and left circumflex artery (Lcx)}

Management of D TGA could be done by Mustard and Senning atrial switch procedures, but in view of increasing frequency of complications such as dysarrhythmias, sudden cardiac death and right ventricular dysfunction has led to go for anatomic correction with Arterial switch operation. Long term patency and growth of the coronary arteries are

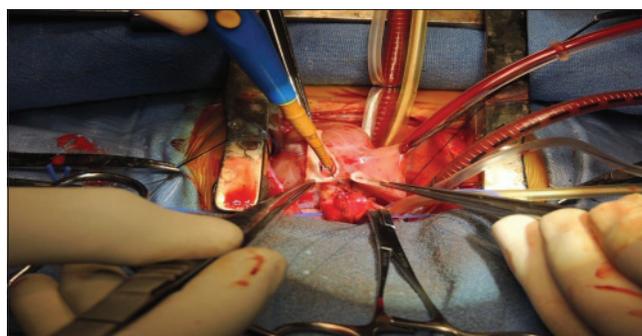


Figure 4: Transposition of great Artery (TGA)- great artery cut open

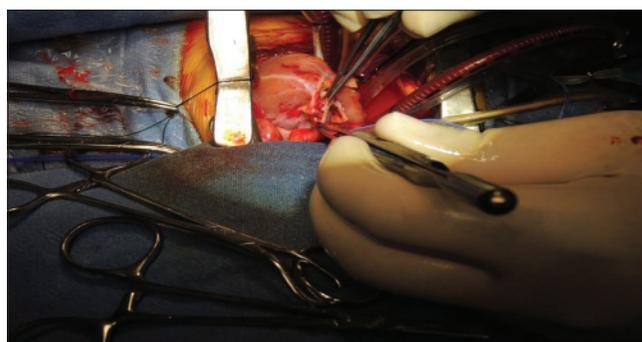


Figure 5: Single coronary artery arising from left aortic cusp with its branches- LCA, RCA, Lcx

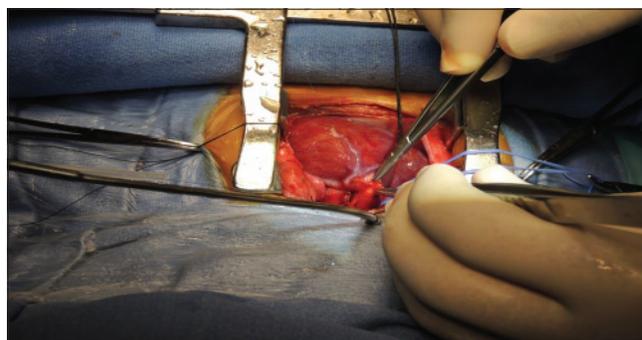


Figure 6: TGA with single coronary artery giving rise to LCA, RCA, Lcx

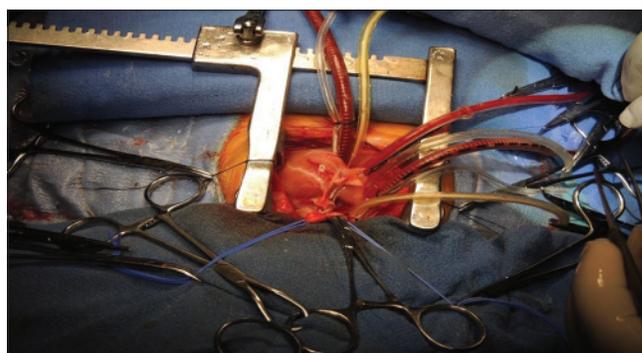


Figure 7: Post operative repair of TGA with coronary and its branches

crucial for the arterial switch operation to be considered the procedure of choice for the surgical management of TGA. Although intraoperative and post operative coronary artery kinking, occlusion can occur resulting in ischaemia and ventricular dysfunction.

CONCLUSION

Complete transposition of great arteries is one of the uncommon Cyanotic congenital heart diseases and single coronary sinus origin for all three coronary in DTGA is even exceptionally singular. Diagnosis of Complete transposition of great arteries should be suspected with the gender, para of pregnancy with its clinical presentation. It should be aggressively confirmed with echocardiography. Management has to be done aggressively with the idea to keep the shunt patent (Patent ductus arteriosus, Atrial septal defect or ventricular septal defect) with the help of Prostaglandin E1. Subsequently, whenever possible anatomical correction should be done with Arterial switch.

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