

Criss-cross Heart: A Complex Congenital Heart Disease

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Abstract

A criss-cross heart is a rare congenital heart disease that is commonly associated with complex cardiac defect. Criss-cross heart is characterized by crossing of the inflow streams of the two ventricles, due to twisting of the heart during development. As a result the axis of the openings of the atrioventricular (AV) valves remain non parallel. We report a case of 2.5 months old baby with criss-cross spatial relationship of the atrioventricular valve with double outlet right ventricle with Pulmonary Arteria and atrial and ventricular septal defect. The diagnosis was confirmed by 2D and color Doppler Echocardiography in subxiphoid long-axis and coronal plane sweeps. Due to the complex structural changes and rarity of the anomaly, the diagnosis is often missed. In this case report we emphasized on diagnosis feature of presentation and surgical techniques required to correct or palliate the defects.

Introduction

Criss-cross heart is a rare congenital heart defects accounting for more than 0.1% of all congenital heart diseases (CHD). The morphological essence of the criss-cross heart is a rotation of ventricular mass along with major axis resulting in different relationships and connections between the atria, ventricle and great vessels. Atrioventricular concordance and ventriculoarterial discordance are seen in 80% of cases. A ventricular Septal defect is seen in almost all the cases. Other anomalies such as pulmonary stenosis, straddling of atrioventricular valves, tricuspid atresia, double outlet right ventricle and congenitally corrected transpositions are seen.

Case Report

Baby-X, a 2.5 month's old female infant, weighting 4.3 kg presented with history of respiratory distress, feeding difficulty and cyanosis since one month of age. Baby was 1st issue of consanguineous marriage and born by caesarean delivery. There was no family history of congenital heart disease. Physical examination revealed evidence of cyanosis, pallor, tachycardia with heart rate 120/min, tachypnea with respiratory rate 50/min, blood pressure 75/40 mm of Hg, peripheral pulses were present and symmetrical with surface oxygen saturation of 40-50% in room air. Cardiac auscultation revealed normal first and second heart sound along with a grade 3/6 ejection systolic murmur best audible at right upper parasternal area. Provisional diagnosis was complex congenital heart disease. Chest X-ray showed oligemic lung field with Mesocardia,

ECG showed right ventricular hypertrophy and thorough echo study showed Mesocardia, L- loop ventricle. Left sided left atrium and right sided left ventricle connected through the mitral valve, right sided right atrium (RA) was connected to left sided right ventricle through tricuspid valve. So ventricles were L-looped with concordant atrioventricular and discordant ventriculoarterial connection. Aorta was arising from RV and PA was also committed to right side. Main pulmonary artery was not seen (Pulmonary atresia) and small Patent ductus arteriosus (PDA) was seen supplying small pulmonary arteries. Large atrial septal defect (ASD secundum type) and large inlet ventricular septal defect (VSD) was noticed. So final diagnosis was Mesocardia, L-loop, Criss cross heart with double outlet right ventricle (DORV) with pulmonary arteria (PA) with PDA dependent pulmonary circulation, large ASD secundum and large inlet VSD.

Initially patient was managed by giving treatment of adequate nutrition and hydration with management of cyanotic spell-by: O₂ inhalation, normal saline infusion and injection Esmolol. Antibiotics: injection Meropenem and Prophylaxis against spell by tab Propranolol and Phenobarbitone. As patients pulmonary blood flow was inadequate, immediate palliation to maintain pulmonary blood flow was planned and referred to advanced cardiac center of India as native surgeons refused the case. PDA anatomy was not suitable for stenting. Plan of surgery from pediatric cardiologists were; Blalock Taussig shunt (BT shunt) in first stage then Single ventricle/ biventricular repair later.



Figure-I: Baby-X

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Figure-II: Color Doppler imaging revealed the crossing of the atrioventricular connections. There were concordant atrioventricular connections, morphological left ventricle on the right side.



Figure-III: The standard 4 chamber view failed to show simultaneously all the four chambers and both atrioventricular valves.



Figure-IV: Short Axis view shows aorta is anterior and Malposed and small pulmonary Arteries are getting supply from PDA.



Figure-V: Echocardiography Showing DORV with PA and Malposed Aorta.



Figure-VI: ECG Showing Right Ventricular Hypertrophy



Figure-VII: Chest X-ray Showing Mesocardia Normal sized heart and oligemic lung Fields

Discussion

The criss-cross heart is an extremely rare malformation of heart. This congenital defect was first described by Lev. Rowlett¹ in 1961, but it was only in 1974 that Anderson et al² first used the term

criss-cross heart. The diagnosis of criss-cross heart is based on crossing of the long axes of the atrioventricular valves. This gives the appearance of each atrium emptying into the contralateral ventricle. Criss-cross heart is almost always associated with other severe cardiac anomalies. The majority of patients with criss-cross heart have hypoplasia of the tricuspid valve and right ventricle, a VSD, abnormal ventricular-arterial alignments (either transposition of the great arteries or double outlet right ventricle) and pulmonary stenosis or atresia. Consequently, neonates present with cyanosis and a systolic murmur. Some patients with criss-cross heart have only a VSD with normally related great arteries and present with symptom of heart failure^{3,4}. Some patients with criss-cross heart have juxtaposed atrial appendages^{5,6,9}, rarely patient present with atrial situs solitus but L-Looped or left handed ventricles (discordant AV segment) but have AV concordance. Despite segmental situs discordance (atrial situs solitus but L-Loop ventricle) the right atrium is aligned with the right ventricles and the left atrium with the left ventricle. Similarly rare patients with atrial situs solitus and D-Looped or right handed ventricles (concordant AV segmental situs), have AV alignment disorder. That is the right atrium is aligned with the left ventricle. Criss-cross heart illustrates the important concepts that AV alignments in L-loop and D-loop heart are important and both must be elucidated and described independently¹⁰. Associated defects with criss-cross heart include: straddling mitral valve¹¹⁻¹⁵ and tricuspid valve, sub aortic stenosis and aortic arch obstruction, mitral stenosis. Initial management is usually determined by the severity of pulmonary stenosis. If pulmonary blood flow is inadequate, short term palliation with prostaglandin E1 is indicated to maintain patency of the Ductus arteriosus. Then a systemic to pulmonary shunt is created to provide adequate pulmonary blood flow until more definitive surgical management can be undertaken. PDA stenting may also be done.

Echocardiography is the primary diagnostic tool. The diagnosis is made easily in a subxiphoid long axis scan of the heart by showing the crossing axes of the two atrioventricular valves^{11,13}. Similarly a scan from posterior to anterior swiping the apical four chamber view shows the crossing axis of the atrioventricular valves. The great arterial connections are better visualized in the parasternal window. Doppler color flow mapping has been reported to facilitate detection of crossing of the inflow Streams¹⁴. Angiography can also use for diagnosis. More recently magnetic resonance imaging has been shown capable of detecting the atrioventricular valves and associated defect¹⁵⁻¹⁷. Prenatal diagnosis can be carried out with fetal echocardiography. The optimal time for imaging the fetal heart is seventeen weeks of gestation.

Conclusion

The study of this case reflects the structural abnormalities associated with the criss-cross heart, way of presentation, and plan of management. The initial management was planned by creating a BT shunt to maintain pulmonary flow and growth of pulmonary artery. The plan of surgery is determined by the function of both ventricles. In single ventricle physiology,

Bidirectional Glenn operation can be done followed by completion of a Fortran type operation. Only small minority of these patients are suitable for two ventricle repair because of hypoplasia of the tricuspid valve and right ventricle. For this case surgical repair may be possible in future by placing a conduit from right ventricle to pulmonary artery.

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