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Criss Cross Heart with Normally Related Great Arteries

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Abstract **Case Report**

The detection of rare congenital cardiac anomalies gives us an opportunity to study such infrequent conditions in greater detail. In this report we describe such an entity that has been reported in the literature extremely rarely. A six weeks old baby boy presented with respiratory distress with features of cardiac failure. The echocardiogram revealed a Criss cross heart with atrioventricular segmental discordance, with a large ventricular septal defect, atrial septal defect and pulmonary arterial hypertension. The ventricles did not have the expected supero-inferior position. There was no ventriculoarterial discordance with no atrioventricular valve straddling. This condition is a rare congenital anomaly. Absence of supero-inferior position of the ventricles, ventriculoarterial discordance and straddling of atrioventricular valves make this case most unusual. Hence such variations need to be considered while entertaining a diagnosis of Criss cross heart in future reports.

Keywords: Congenital anomaly, Criss cross heart, Septal defect, NRGA.

INTRODUCTION

Congenital cardiac anomalies are seen in about 8 cases per 1000 new-borns at term. This percentage increases if premature and stillbirths are considered. Criss cross heart is an extremely rare cardiac defect, accounting for less than 0.1% of all congenital cardiac malformations, not exceeding 8 per 1,000,000 births [1]. In this report we highlight certain unique features in the heart of a baby boy, born with this rare abnormality.

CASE REPORT

A six weeks old baby boy presented to our Cardiology OPD with worsening respiratory distress for duration of one week. At presentation his heart rate was 154 beats per minute, respiratory rate was 62 beats per minute, blood pressure was 70/50 mm Hg and he had a room air oxygen saturation of 88%. Examination revealed a pan systolic murmur in the left sternal border. The twelve-lead electrocardiogram (ECG) revealed sinus tachycardia. The chest radiograph revealed cardiomegaly.

The ECHO revealed visceral situs solitus, atrial situs solitus and levocardia. The pulmonary and systemic venous drainages were normal. There was a 16mm non-restrictive ostium secondum atrial septal defect shunting left to right. The morphological right ventricle was found to be on the right of the morphological left ventricle – there was no ventricular inversion. Interestingly the right atrium was connected to the left ventricle through the mitral valve and the left atrium was connected to the right ventricle through the tricuspid valve – hence there was crossing over of the systemic and pulmonary circulations at the level of the atrioventricular junction with atrioventricular (AV) segmental discordance. There was a 14mm non-restrictive muscular ventricular septal defect shunting left to right. The ventricles we're not placed in a supero-inferior axis. There was ventriculoarterial (VA) concordance with normally related great vessels. The main pulmonary artery arising from the right ventricle was significantly dilated. There was no pulmonary stenosis. The aorta arising from the left ventricle had no subaortic stenosis or arch anomaly. There was no significant tricuspid regurgitation or pulmonary

regurgitation for accurate echocardiographic estimation of the pulmonary artery pressures. In view of the above, a diagnosis of criss cross heart was made, probably associated with pulmonary artery hypertension. The child required intubation and stabilisation with inotropic support. The patient's parents were explained about the need for pulmonary artery banding followed by corrective surgery at a later date. However, they were not keen on further management and hence the patient had to be discharged at request and could not be followed up further.

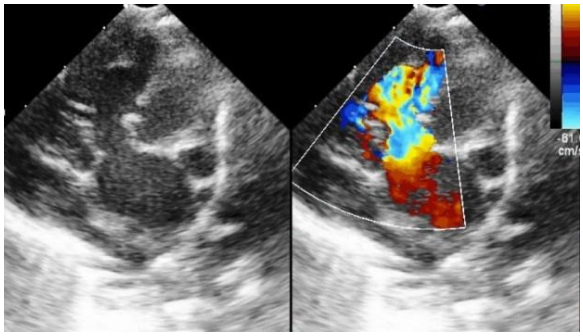

Fig-1: LA connecting to the RV

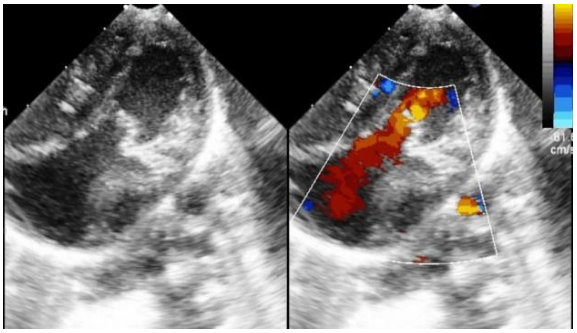

Fig-2: RA connecting to the LV

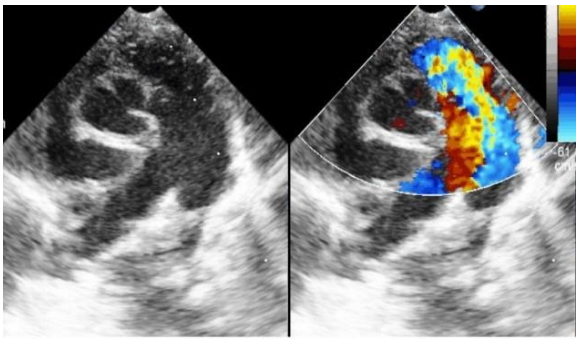

Fig-3: Normally related great arteries

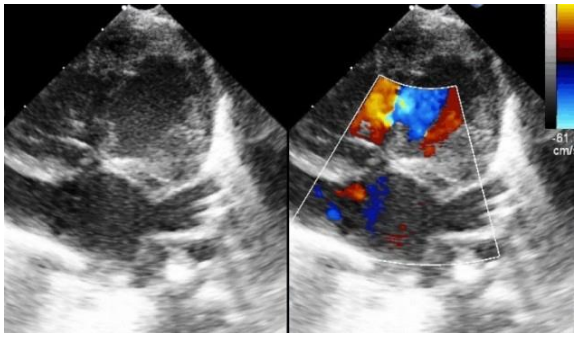

Fig-4: Ventricular septal defect

DISCUSSION

M Lev and UF Rowlatt were the first people to describe criss cross heart in 1961 [2]. In their report they called this rare entity "mixed levocardia". They reviewed thirteen cases that they had come across with atrial or ventricular inversion with or without corrected transposition. The terminology "Criss cross heart" was introduced by Robert H Anderson and his colleagues in 1974. [3].

The diagnosis of criss cross heart is based on the demonstration of crossing of the long axes of the atrioventricular valves. The condition is characterised by a basic defect produced by the rotation of the ventricular mass along its long axis. This positional anomaly may coexist with a horizontal displacement of the ventricular mass along the horizontal plane of long axis. This produces supero-inferior ventricles, with the right ventricle placed superiorly and the left ventricle placed inferiorly.

In most patients with criss cross heart, with atrial situs solitus, and d-looped or right-handed ventricles (concordant atrioventricular segmental situs), the right atrium is aligned with and opens into the right ventricle and the left atrium aligns with and opens into the left ventricle (concordant atrioventricular alignments). Consequently, very rarely, patients with criss cross heart with atrial situs solitus, will have the right atrium aligned with the right sided right ventricle, but opening into the left sided left ventricle and vice versa for the left atrium – a condition characterised by a discordant atrioventricular segmental situs with concordant atrioventricular alignment. Our patient fell into this extremely rare second category. In about 80% of patients with criss cross hearts, there is ventriculo-arterial discordance – this was not the case in our patient. Other associated defects that may be seen with this condition are a straddling mitral valve, a straddling tricuspid valve, sub-aortic stenosis, aortic arch obstruction and mitral stenosis. Very rarely it can be associated with transposition of the great arteries with an intact ventricular septum. Criss cross hearts may be seen in three forms - complete transposition, corrected transposition, and normal hearts.

CONCLUSION

In our infant with criss cross heart, there was AV segmental discordance and VA concordance with normally related great arteries. Interestingly, the two ventricles were aligned side by side rather than supero-inferiorly, with no atrioventricular valve straddling – both of which are very unusual of this condition. Thus, in our patient, even in the presence of a criss cross heart, the great vessels were normally related and there was no supero – inferior relationship of the ventricles. These two entities are rare in an already absolutely rare condition making this patient worth reporting.

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