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Original Article

# Criss-cross heart: Transthoracic echocardiographic features

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ABSTRACT

*Objective:* To study the echocardiographic features of criss-cross heart (CCH), a congenital cardiac anomaly characterized by crossed ventricular inflow streams, in Indian patients.
*Methods:* In this retrospective observational study, all pediatric echocardiograms performed in a single tertiary care institution in South India over a three-year period were scrutinized for a diagnosis of CCH. Demographic, clinical and echocardiographic data were collected from patients' medical records and echocardiographic database. Crossed ventricular inflow streams was identified when there was inability to visualize both atrio-ventricular valves in a single imaging plane in cardiac four chamber view.
*Results:* CCH was diagnosed in five patients from 10,500 pediatric echocardiographic studies. The age at diagnosis ranged from one month to 8 years. Cyanosis was present in all but one of the five cases. Crossed ventricular inflow streams was present by definition in all cases, whereas superior-inferior ventricular relationship was present in only three cases. All cases were associated with ventricular septal defects. Atrio-ventricular discordance was seen in three cases and concordance in two. Ventriculo-arterial discordance was seen in three cases, concordance in one and double outlet right ventricle in one. Three cases had pulmonary stenosis and the other two had pulmonary arterial hypertension. Straddling of AV valve was observed in four cases and hypoplastic aortic arch in one case.
*Conclusion:* CCH is an extremely rare congenital cardiac anomaly. Superior-inferior ventricular relationship often co-exists with CCH, but is not necessarily present in all cases. CCH requires early diagnosis because of its common association with diverse cardiac anomalies.


## 1. Introduction

Criss-cross heart (CCH) is a rare congenital anomaly of cardiac rotation resulting in crossing of ventricular inlets and drainage of the atria into contra-laterally located ventricles.[1] The atrio-ventricular (AV) and ventriculo-arterial (VA) connections can be concordant or discordant.[2] There can be either side-by-side or superior-inferior ventricular arrangement (superior-inferior ventricles, SIV); Lev and Rowlatt, in 1961, were the first to describe an unusual arrangement of ventricular inlets with the right ventricle abnormally positioned superior the left ventricle.[3] In 1974 Anderson et al. used the term "criss-cross heart" for the first time.[1] The complex and distorted cardiac anatomy seen in CCH makes accurate diagnosis difficult. Because of its common association with diverse cardiac anomalies, CCH requires early diagnosis to provide timely operative management and achieve a good functional outcome. Transthoracic echocardiography is the preferred diagnostic modality, though cardiac magnetic resonance imaging can provide valuable additional information, especially in challenging cases. Cardiac catheterisation and angiography have little value in the management, except in patients with features of pulmonary hypertension and in indeterminate cases.[4] The paucity of data on CCH from India provided the impetus to study its echocardiographic features in Indian patients.

## 2. Methods

This was a retrospective observational study. All pediatric echocardiograms performed over the preceding three years in a single tertiary care institution were scrutinized and patients with a diagnosis of CCH identified. Demographic, clinical and echocardiographic data were collected from patient's medical records and echocardiographic database. Echocardiograms were obtained using the Philips IE33 system by a single operator. Chloral hydrate was administered as necessary during scanning. Images were stored digitally for subsequent offline analysis by the same

*Abbreviations:* AV, atrio-ventricular; PA, pulmonary artery; VA, ventriculo-arterial; CCF, congestive cardiac failure; SV, single ventricle; PS, pulmonary stenosis; DORV, double outlet right ventricle; VSD, ventricular septal defect; ccTGA, congenitally corrected transposition of great arteries; PAH, pulmonary arterial hypertension.

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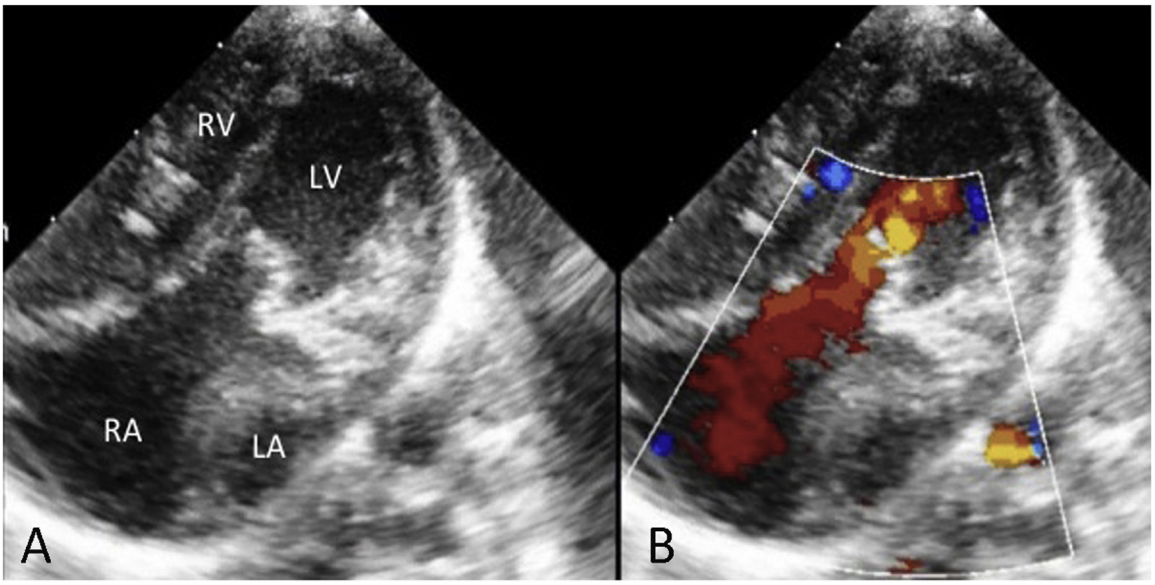


**Fig 1.** A. Two-dimensional and B. color Doppler transthoracic echocardiographic images obtained in apical four-chamber view showing twisted and discordant atrio-ventricular connections with the right atrium (RA) draining into the contra-laterally located morphological left ventricle (LV).

operator. Apex position, situs of the atrium, AV and VA relationships were determined by segmental analysis as previously reported.[5] The relationship of the inflows of the two ventricles was assessed in apical or subcostal four-chamber views with two-dimensional and color Doppler echocardiography. Crossed ventricular inflow streams was defined as: (1) inability to visualize both AV valves in a single imaging plane in cardiac four chamber view and (2) the presence of crossed AV inflow blood streams with each atrium draining into the ventricle located contra-lateral to it (Figs. 1 and 2). Presence of SIV was defined as horizontal alignment of the inter-ventricular septum in mid-ventricular parasternal and/or subcostal short-axis views (Fig. 3). Vertical inter-ventricular septal alignment was deemed present when both ventricles lay side-by-side (Fig. 4). Presence of associated cardiac defects was recorded in detail.

## 3. Results

Among the 10,500 pediatric echocardiographic studies done during the period January 2013 to May 2016, congenital anomalies were detected in 3500 children. A diagnosis of CCH was made in 5 cases (3 girls, 2 boys) which comprised 0.14% of all congenital anomalies. The age at diagnosis ranged from one month to 8 years. Cyanosis was the presenting feature in all but one of the five cases. Features of cardiac failure were seen in two children (Table 1).

### *3.1. Segmental analysis*

All five CCH cases in our study had visceral and atrial situs solitus (Table 1). Four cases had levocardia and one had dextrocardia. AV discordance was observed in three cases and

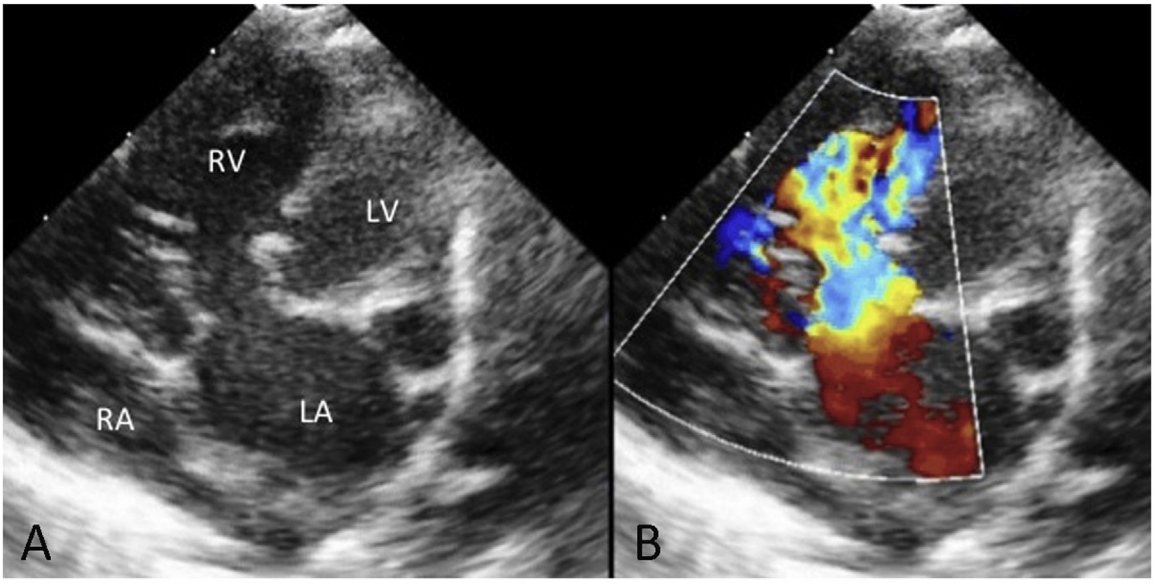


**Fig. 2.** A. Two-dimensional and B. color Doppler transthoracic echocardiographic images obtained in apical four-chamber view showing twisted and discordant atrio-ventricular connections with the left atrium (LA) draining into the contra-laterally located morphological right ventricle (RV).

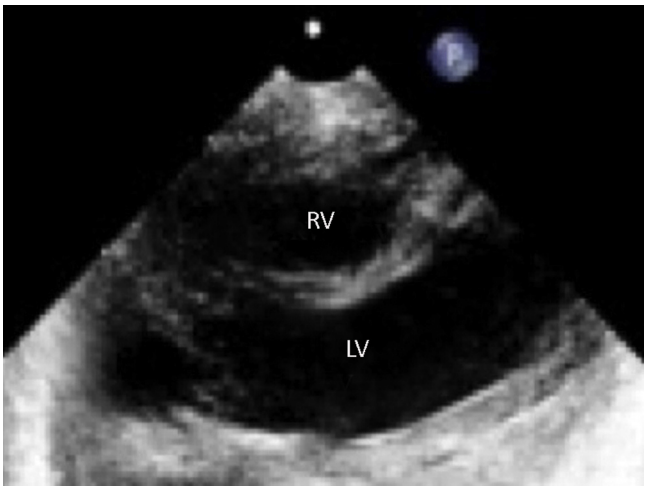


**Fig. 3.** Two-dimensional transthoracic echocardiographic image obtained in parasternal short-axis view showing horizontal orientation of the inter-ventricular septum and superior-inferior ventricular relationship.

concordance in the other two. VA discordance was seen in three cases, concordance in one and double outlet right ventricle in one. Crossed ventricular inflow streams were present by definition in all cases. However, SIV relationship was present in only three cases, while two had vertical inter-ventricular septal alignment with side-by-side arrangement of the ventricles.

### 3.2. Associated anomalies

All cases were associated with ventricular septal defect, which was of inlet type in two cases, muscular type in two cases and perimembranous type in one case respectively (Table 2). Ostium secundum atrial septal defect was present in three cases and fenestrated inter-atrial septum was observed in one case. Three of the cases had pulmonary stenosis – infundibular and valvular stenosis in two and isolated infundibular stenosis in one. The two cases which were not associated with pulmonary stenosis were found to have pulmonary arterial hypertension. Straddling of AV valves was observed in four cases – tricuspid valve in two and mitral valve in the other two. Hypoplastic aortic arch was observed in one case. Due to significant tricuspid valve straddling, the right ventricle was hypoplastic with single ventricle physiology in two

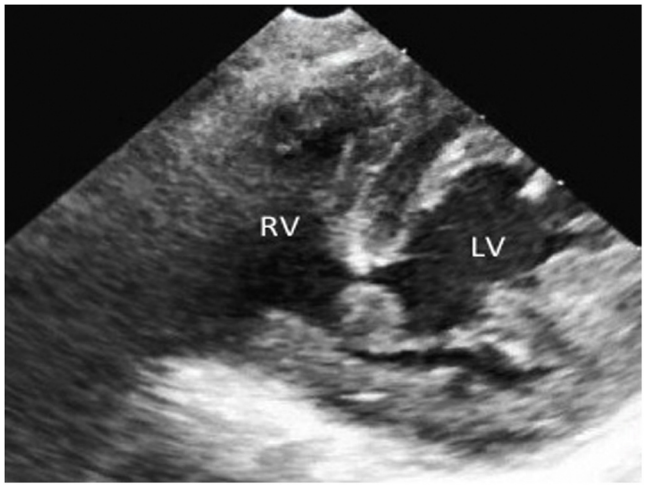


**Fig. 4.** Two-dimensional transthoracic echocardiographic image obtained in parasternal short axis view showing vertical orientation of the inter-ventricular septum and side-by-side ventricular relationship.

cases. Congenitally corrected transposition of great arteries and double outlet right ventricle was seen in one case each. None of the patients underwent surgery as the anatomy was deemed too complex for correction.

## 4. Discussion

CCH has been attributed to a rotational abnormality of the ventricular mass, as a result of which the relationships of the ventricular chambers are not as expected for a particular AV connection; the abnormal rotation of the ventricular mass occurs around its long axis and takes place after ventricular septation.[1,6] Freedom showed that the degree of rotation is variable and can result in either complete or partial criss-crossing of ventricular inlets.[7] CCH can be found with any biventricular AV connection, any VA connection and any atrial situs.[6] It is important to distinguish between connections of cardiac segments and the relationship of the chambers within the segments; these are separate and independent features and should be described using mutually exclusive terms.[6] In CCH, the abnormal rotation of ventricles along its long axis during the embryonic period causes criss-crossing of ventricular inlets producing unexpected ventricular relationships, but this can occur with both concordant and discordant AV connections.[6] Inability to visualize both the AV valves in a single imaging plane in cardiac four chamber view indicates the presence of crossed ventricular inflow streams during echocardiography, and its demonstration is a *sine qua non* for the diagnosis of CCH. This finding was present by definition in all five cases in our series as in other reported echocardiographic studies of CCH.[8,9]

CCH and SIV (upstairs-downstairs hearts) frequently co-exist, but the two are not synonymous and either can be found in isolation.[6] SIV arises due to displacement of the ventricular mass along the horizontal plane, a phenomenon independent of the rotational abnormality of the ventricular mass that results in CCH.[6] SIV was not found in two cases (40%) in our series. Although vertical alignment of interventricular septum has been reported in CCH, horizontal alignment is much more common. The 60% prevalence of horizontal alignment of inter-ventricular septum and SIV seen in our series was less than that observed in other series. Fang reported that nine of 10 patients with CCH in their study showed SIV and concluded that SIV was an important feature of patients with CCH.[8] In another study Yang reported SIV in all of their 4 cases.[9] SIV, however, is not specific for CCH and may also be present without CCH.[10]

In our series, 40% of cases had concordant AV connections, which was in contrast to the findings of Valdes-Crus who reported 81% AV concordance.[11] Criss-cross AV connection can occur even in double inlet ventricles when the axes of the two AV valves cross each other.[6] Ngeh reported three cases of CCH diagnosed using prenatal ultrasound examination at 19–23 weeks of gestation; double-inlet ventricle was present in these three cases.[12] Prenatal screening by an experienced operator may thus have a role in early detection of this complex congenital anomaly. Of the four cases that had AV valve straddling in our study, two had hypoplastic right ventricle, precluding biventricular repair in these patients. The presence of ventricular septal defect in all of our cases was similar with the pattern described in other case series.

Sixty percent of cases had discordant VA connections in our study, which was similar to the 54% prevalence reported in the study by Valdes-Crus.[11] One patient in our study had concordant VA connection, which is in contrast to the uniform absence of normal VA connection in all cases reported in recently published studies.[12,13] Double outlet right ventricle was seen in only one patient (20%) in our study, whereas a higher prevalence was reported by Fang (50%), Yang (50%), Valdes-Crus (30%), and

**Table 1**
Echocardiographic characteristics and clinical presentation of criss-cross heart.

| Case | Age at diagnosis, Sex | Atrial situs, Cardiac position | Atrio-ventricular connection | Great artery relationship | Ventriculo-arterial connection | Clinical presentation, Physiology |
|---|---|---|---|---|---|---|
| 1. | 1 month, Male | Situs solitus, levocardia | AV discordance | Aorta right and posterior to PA | VA concordance | Cyanosis, CCF, Large VSD/ PAH |
| 2. | 8 years, Female | Situs solitus, levocardia | AV concordance | Aorta anterior to PA | VA discordance | Cyanosis, SV/PS |
| 3. | 4 years, Female | Situs solitus, dextrocardia | AV concordance | Aorta left and anterior to PA | DORV | Cyanosis, DORV/VSD/PS |
| 4. | 3 years, Female | Situs solitus, levocardia | AV discordance | Aorta left and anterior to PA | VA discordance | Cyanosis, ccTGA/VSD/PS |
| 5. | 6 months, Male | Situs solitus, levocardia | AV discordance | Aorta anterior to PA | VA discordance | CCF, SV/PAH |

**Table 2**
Associated anomalies of criss-cross heart.

| Case | Type of VSD | PS/PAH | Straddling AV valves | Other anomalies |
|---|---|---|---|---|
| 1. | Inlet | PAH | Absent | ASD |
| 2. | Muscular | PS | Present | Hypoplastic RV |
| 3. | Inlet | PS | Present | ASD |
| 4. | Peri-membranous | PS | Present | Fenestrated inter-atrial septum |
| 5. | Muscular | PAH | Present | Hypoplastic aortic arch, hypoplastic RV |

Abbreviations: VSD, ventricular septal defect; PS, pulmonary stenosis; PAH, pulmonary arterial hypertension; AV, atrio-ventricular; ASD, atrial septal defect; RV, right ventricle.

Hoffmann (57%)[8,9,11,13] Sixty percent prevalence of pulmonary outflow tract obstruction at the time of diagnosis in our series was close to the 55% prevalence reported by Valdes-Crus, whereas all cases reported by Hoffmann had pulmonary outflow tract obstruction.[11,13]

## 5. Conclusion

CCH is an extremely rare congenital cardiac anomaly. Systematic echocardiography using segmental analysis approach is essential for diagnosis. Inability to visualize both AV valves in a single imaging plane in cardiac four chamber view identifies crossed ventricular inflow streams and is a prerequisite for the diagnosis of CCH. SIV very often co-exists with CCH, but is not necessarily present in all cases.

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