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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.

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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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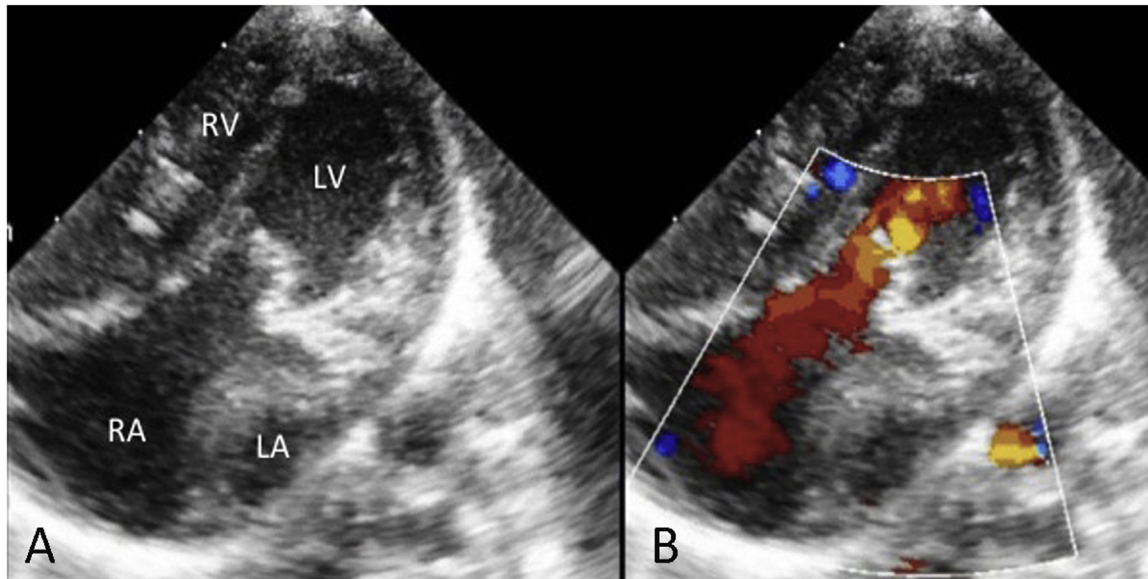


Fig 1. A. Two-dimensional and B. color Doppler transthoracic echocardiographic images obtained in apical four-chamber view showing twisted and discordant atrio-ventricular connections with the right atrium (RA) draining into the contra-laterally located morphological left ventricle (LV).

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

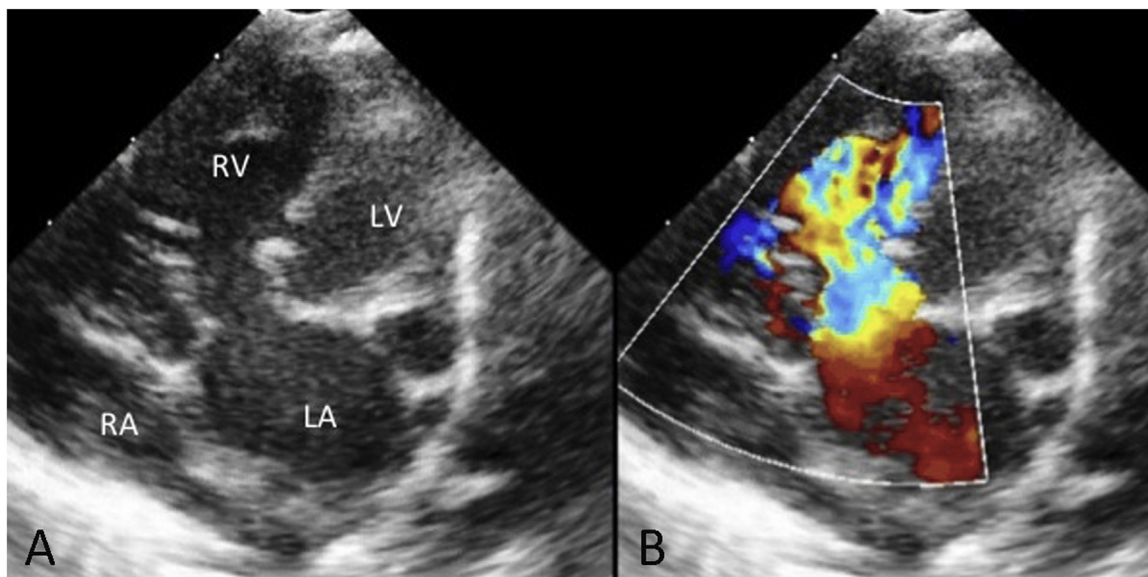


Fig. 2. A. Two-dimensional and B. color Doppler transthoracic echocardiographic images obtained in apical four-chamber view showing twisted and discordant atrio-ventricular connections with the left atrium (LA) draining into the contra-laterally located morphological right ventricle (RV).

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