IMAGES in PAEDIATRIC CARDIOLOGY

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Abstract
Crisscross heart (CCH) is a rare cardiac malformation characterized by crossing of the inflow streams of the two ventricles due to an apparent twisting of the heart about its long axis. The developmental mechanisms and causes of CCH are remaining unknown. Neonates mainly presents with cyanosis and a systolic murmur. We herein present a case of CCH with concordant atrioventriculo connections with double outlet right ventricle (DORV) which was diagnosed by echocardiography.

Keywords: *Congenital heart defect; Criss-Cross heart; Double outlet right ventricle; Echocardiography*

Introduction

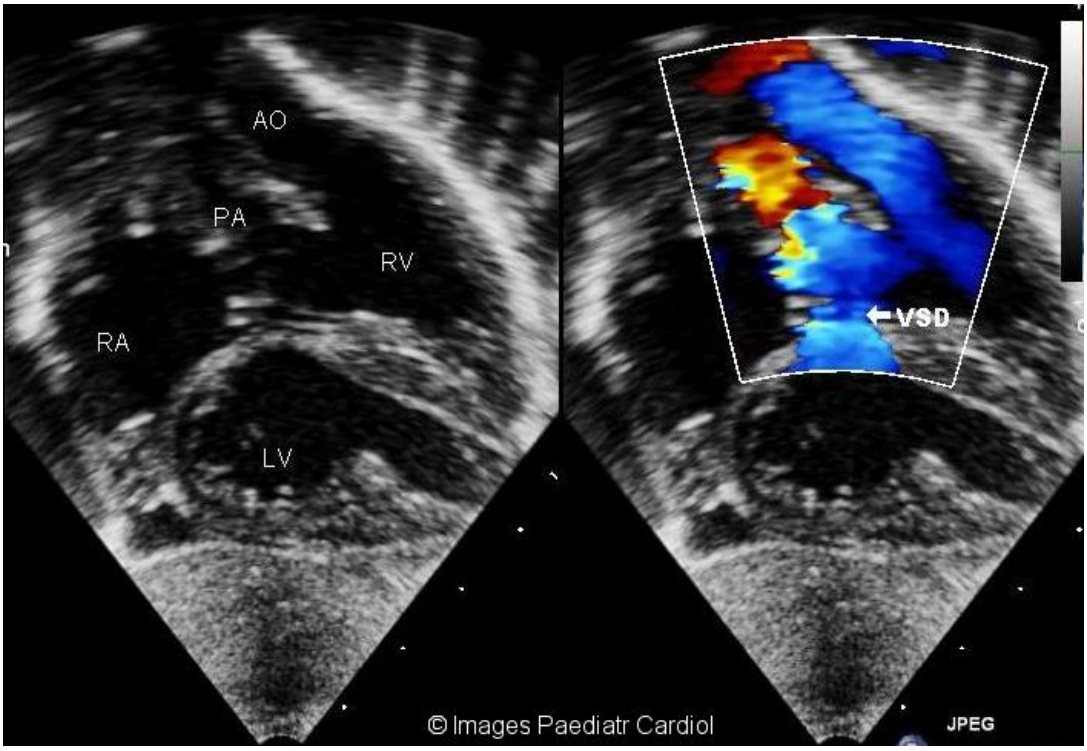
Criss-cross heart (CCH), or superoinferior ventricles, is a complex congenital rotational abnormality in which the systemic and pulmonary venous streams cross at the atrioventricular (AV) level without mixing. Its frequency is less than 8/1000000 and accounts for <0.1% of congenital heart defects (CHD). In the normal heart, AV structures are parallel to each other when viewed from the front, whereas in CCH the AV structures are not parallel but angulated by as much as 90 degrees.[1-4] The atrium connects with the contralateral ventricle and the ventricular chambers are arranged in a superoinferior fashion, with the right ventricle (RV) superiorly and the left ventricle (LV) inferiorly located, regardless of whether the AV connection is concordant or discordant. The diagnosis is made by using 2-dimensional and color Doppler echocardiography.[3-5] Here, we report a rare case of CCH with concordant atrioventricular connections with DORV which was diagnosed by echocardiography.

Case Report

A 2 month old female infant, weighing 3.7kg presented with history of breathlessness, feeding difficulty and cyanosis. She was a second product of non-consanguineous marriage and born by normal vaginal delivery. There was no family history of congenital heart disease. Physical examination was remarkable with evidence of pallor, cyanosis and dysmorphic facies. Her heart rate and respiratory rates were 154/min. & 60/min. respectively. Blood pressure was recorded at 75/35 mmHg and oxygen saturation 85% in room air. There was evidence of respiratory distress. All the peripheral pulses were well felt. Cardiac auscultation revealed normal first and second heart sound along with a grade 3/6 ejection systolic murmur audible best at the left upper parasternal area. The third heart sound was audible.

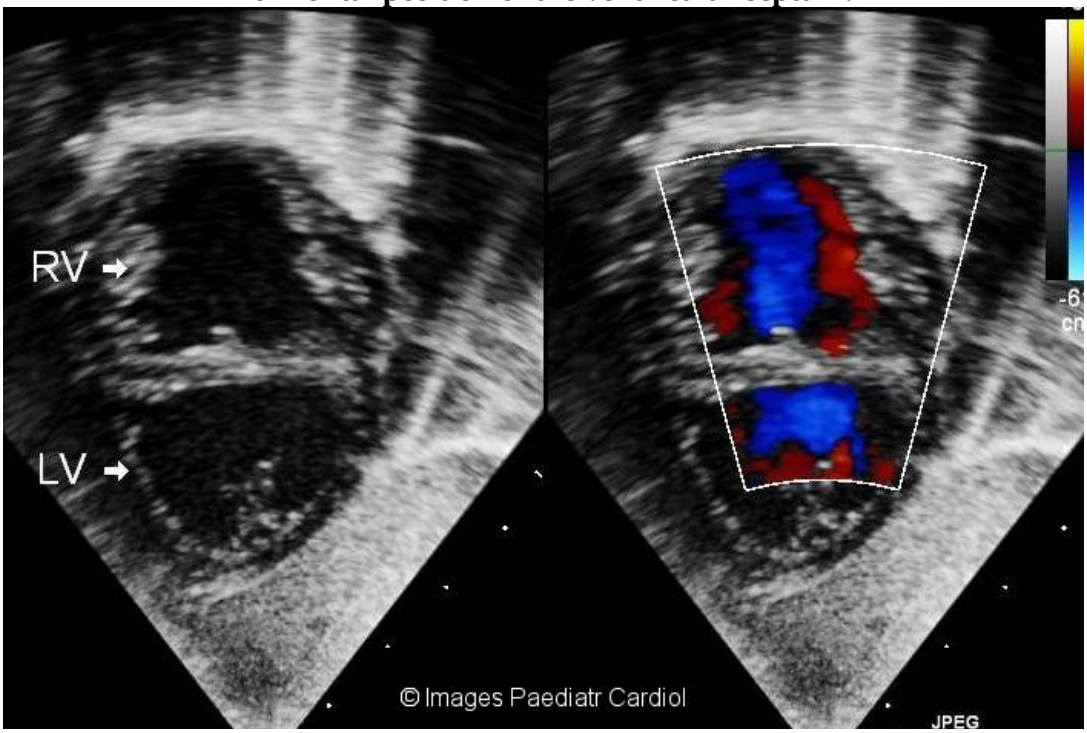
Chest roentgenogram revealed an enlarged heart (cardiothoracic ratio 0.6) with pulmonary plethora. The two-dimensional echocardiography, subcostal coronal views demonstrated the connection of the left-sided left atrium and the right-sided left ventricle through the mitral valve and the right-sided right atrium to be connected to the left-sided right ventricle through the tricuspid valve by anterior angulation of the transducer. It also shows the two great arteries arising from the right ventricle (figure 1).

Figure 1: Subcostal coronal views shows the connection of the right-sided RA to be connected to the left-sided RV through the tricuspid valve, inlet VSD and the two great arteries arising from the RV.



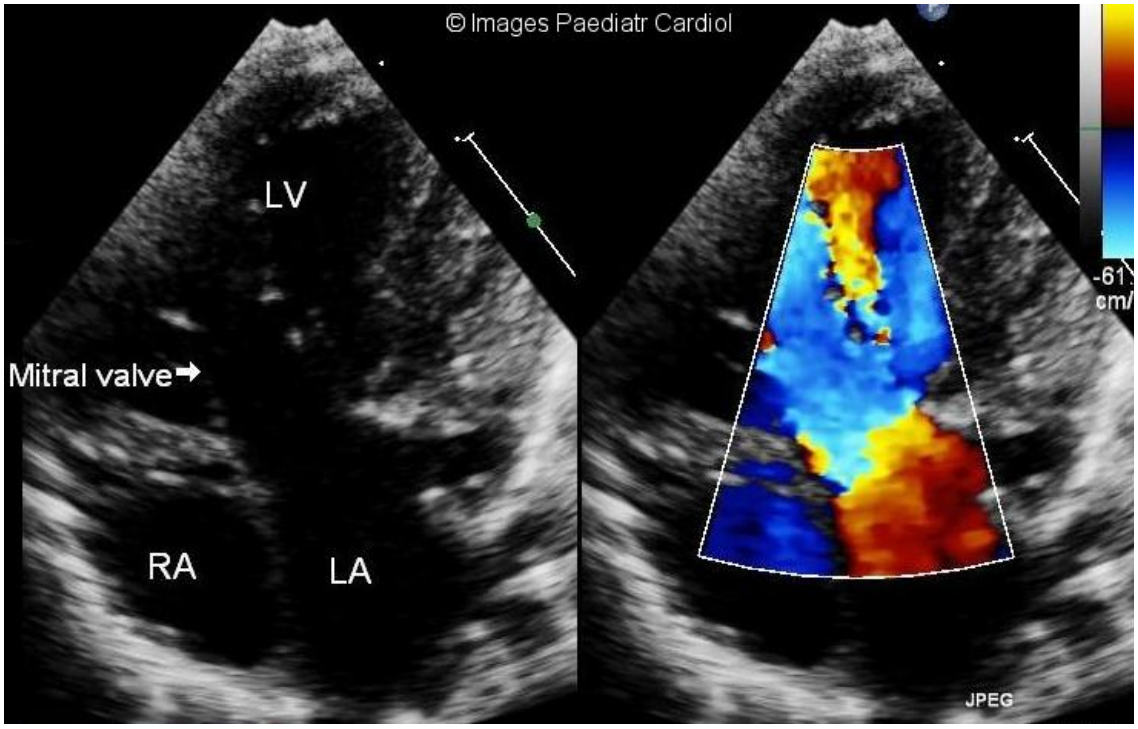
2-D and color Doppler imaging revealed the crossing of the atrioventricular connections. Short-axis views shows the right ventricle was superior and left ventricle was inferior with horizontal position of the ventricular septum (figure 2).

Figure 2: Short-axis views shows the superior (RV) and inferior (LV) ventricle with horizontal position of the ventricular septum.



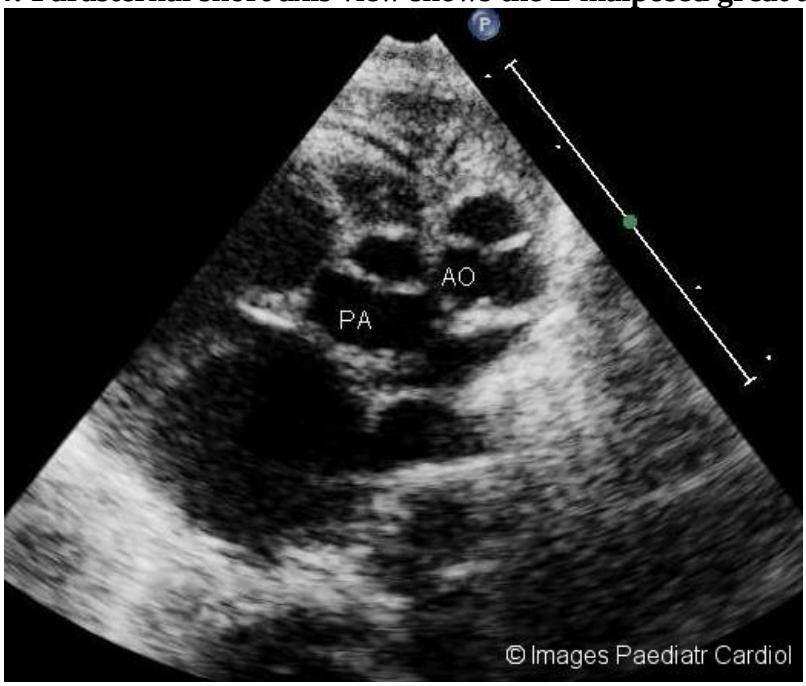
The standard 4-chamber view was not showing simultaneously all four chambers and both atrioventricular valves (figure 3).

Figure 3: Apical 4-chamber view was not showing simultaneously all four chambers and both atrioventricular valves.



Parasternal short axis view shows malposed great arteries (figure 4).

Figure 4: Parasternal short axis view shows the L malposed great arteries.



A large size inlet type of ventricular septal defect (bidirectional shunt) and a moderate size ostium secundum atrial septal defect (bidirectional shunt) were present. Biventricular function was normal.

Discussion

CCH is a complex congenital anomaly produced by the rotation of ventricular mass along its long axis. This positional anomaly can coexist with a horizontal displacement of the ventricular mass along the horizontal plane of long axis, which produces superior-inferior ventricles. In 1961, Lev and Rowlatt described unusual arrangement of the cardiac inlets that is ventricular chambers arranged in a superoinferior fashion, with the RV superiorly and the LV inferiorly.[2] In 1974, Anderson et al used the term CCH for cardiac anomaly producing the illusion of crossing of the systemic and pulmonary venous stream without mixing at the AV level.[3] In the later years, other cases was described with situs solitus or situs inversus or isomerism, mostly with AV concordance.[4-5] The physiology is determined by the concordant or discordant atrioventricular (AV) and ventriculoarterial (VA) connection and the associated cardiac defects.[6-7] CCH may be seen in three forms, that is complete transposition, corrected transposition, and normal hearts. In our case, there was AV and VA concordance with malposed great arteries. A review of the medical literature does not reveal any isolated presentation of CCH cases. Most patients with CCH has other anomalies such as VSD, transposition of great vessels, right ventricular hypoplasia, subpulmonary stenosis, straddling AV and others.[7-8]

The diagnosis should be suspected by echocardiography when the parallel arrangement of the AV valves and ventricular inlets cannot be achieved, and the two valves are not easily visualized simultaneously on apical 4 chamber view. Color flow mapping can help in assessing the AV connection, visualization of the direction of intracardiac blood flows and recognition of the crossover of the inflow streams.[9-10] Yang YL et al[11] reported that the failure to obtain a characteristic 4-chamber view was diagnostic for recognition of the CCH. The echocardiographic features suggested by yang YL et al[11] were present in our case. Surgical options vary according to the exact sequential segmental analysis and associated abnormalities. In conclusion, CCH is a rare cardiac anomaly than can be diagnosed by a transthoracic echocardiography by an alert echocardiographer to determine the relationships of the cardiac chambers and associated cardiac anomalies.

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