

## A criss-cross heart

### *Criss-cross kalp*

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### ABSTRACT

A criss-cross heart is an extremely rare congenital cardiac anomaly characterized by crossing of the inflow streams of the two ventricles and almost always associated with other severe cardiac diseases. In this article, a total of seven patients with criss-cross morphology were identified from the hospital records and clinical characteristics of these patients and associated congenital cardiac diseases were discussed in the light of the literature data. Age on admission ranged from one month to 13 years. Of the seven patients, three presented with cyanosis, three with murmur, and one with respiratory distress and heart failure. The viseroatrial situs was solitus in all patients including levocardia in five and dextrocardia in two. The atrioventricular connection was concordant in four patients and discordant in one patient, while double-inlet ventricle was present in two patients. The ventriculoarterial connection was concordant in four patients and discordant in one patient, while double-outlet ventricle was present in one patient and single outlet (pulmonary atresia) in another patient. Two patients underwent corrective biventricular repair, while one patient underwent bidirectional cavopulmonary shunting. Criss-cross heart is an extremely rare and complex anomaly which should be kept in mind to recognize with echocardiography.

**Keywords:** Abnormal twisting; criss-cross heart; straddling atrioventricular valve; superior-inferior ventricles.

A criss-cross heart is an extremely rare congenital cardiac anomaly characterized by crossing of the inflow streams of two ventricles due to abnormal rotation of the ventricular masses along its major axis.<sup>[1]</sup> The incidence of this anomaly is less than 8/1.000.000.<sup>[1]</sup>

### ÖZ

Criss-cross kalp iki ventrikülün giriş akımlarının birbirlerini çaprazlaması ile karakterize ve hemen hemen daima diğer ciddi kalp hastalıkları ile ilişkili olan, çok nadir görülen doğumsal bir kalp anomalisidir. Bu yazıda, hastane kayıtlarından criss-cross morfolojisi olan toplam yedi hasta belirlendi ve bu hastaların klinik özellikleri ve eşlik eden doğumsal kalp hastalıkları literatür verileri eşliğinde tartışıldı. Başvuru sırasındaki yaş, 1 ay ile 13 yıl arasında değişiyordu. Yedi hastanın üçü siyanoz, üçü üfürüm ve biri solunum sıkıntısı ve kalp yetmezliği ile başvurdu. Tüm hastalarda viseroatriyal situs solitus olup, beşinde levokardi ve ikisinde dekstroardi vardı. Atrioventriküler ilişki dört hastada konkordan ve bir hastada diskordan iken, iki hastada çift girişli ventrikül saptandı. Ventriküloarteriyel ilişki dört hastada konkordan ve bir hastada diskordan iken, bir hastada çift çıkışlı ventrikül ve bir diğer hastada tek arter çıkışı (pulmoner atrezi) vardı. İki hastaya düzeltici biventriküler tamir yapılırken, bir hastaya bidireksiyonel kavopulmoner şant yapıldı. Criss-cross kalp oldukça nadir görülen ve ekokardiyografi sırasında tanınması için akılda tutulması gereken kompleks bir anomalidir.

**Anahtar sözcükler:** Anormal burulma; criss-cross kalp; atrioventriküler kapak straddlingi; superior-inferior ventrikül.

Although the exact etiology of a criss-cross heart remains unclear, it is almost always associated with other severe cardiac anomalies and most of the patients, therefore, present early in infancy with cyanosis and murmurs.<sup>[1]</sup> The failure to obtain a characteristic four-chamber view in any plane with transthoracic



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echocardiography is diagnostic for recognition of the criss-crossed atrioventricular junctions.<sup>[1]</sup> The majority of the patients with a criss-cross heart can be staged toward a Fontan-type operation, as a two-ventricle repair cannot be always achieved.

In this study, we discuss seven patients with criss-cross morphology in the light of literature data.

## CASE REPORT

Between January 2006 and August 2014, seven patients with criss-cross morphology were identified from the hospital medical records and cardiac catheterization database. In addition to the demographic and clinical findings, echocardiography, cardiac catheterization, and angiography findings of all of the patients at the time of initial diagnosis were obtained from their medical records. The diagnosis of criss-cross morphology was established by echocardiography in case of inability to obtain a characteristic four-chamber view in any plane through detailed two-dimensional and Doppler echocardiographic examinations using the 3 or 7 MHz probe of a Vivid 7 ultrasound device (GE Vingmed Ultrasound AS, Horten, Norway). All patients had associated congenital heart defects. Cardiac catheterization and angiography were performed to obtain pressure or oxymetric data to identify associated defects.

**Case 1–** A 13-year-old male was referred to our clinic with murmur, palpitation, and easy fatigue. His physical examination revealed a 3/6 systolic murmur best heard at the upper left sternal area. He had no cyanosis. Echocardiography demonstrated criss-cross morphology as well as situs solitus levocardia, congenitally corrected transposition of great arteries (ccTGA) with right anterior aorta, straddling mitral valve, large inlet ventricular septal defect (VSD), atrial septal defect (ASD) and valvular and subvalvular pulmonary stenosis (PS) with a maximum pressure gradient of 70 mmHg. Cardiac catheterization and angiography were performed. Oxygen (O<sub>2</sub>) saturation in the descending aorta was 95%, while the mean pulmonary artery pressure was 19 mmHg. The patient is currently being followed without any surgical intervention.

**Case 2–** A 16-month-old girl was admitted to our center due to cyanosis. On physical examination, severe cyanosis and 3-4/6 systolic ejection murmur best heard at left upper sternal border were detected. Echocardiography revealed situs solitus levocardia, a criss-cross heart, double inlet left ventricle, right ventricular hypoplasia, restrictive small VSD, severe PS and pulmonary hypoplasia, and persistent left superior

vena cava. Cardiac catheterization and angiography confirmed these findings and additionally demonstrated an O<sub>2</sub> saturation of 65% in the descending aorta and the mean pulmonary artery pressure of 12 mmHg. Bidirectional cava pulmonary shunt was recommended to the patient; however, she was lost in follow-up.

**Case 3–** A five-year-old female patient was referred to our center with the diagnosis of complex congenital heart disease detected during the examination for murmur and dextrocardia. Physical examination demonstrated the apical heart beat on the right side of the chest and 3/6 systolic ejection murmur on the right side of the sternum without any additional finding. Echocardiography showed situs solitus dextrocardia, criss-cross heart with superior-inferior relation of the ventricles, juxtaposition of the atrial appendages, VSD, severe sub-aortic (discrete sub-aortic membrane), and mild pulmonary stenosis creating a pressure gradient of 106 and 30 mmHg, respectively. Corrective biventricular repair including the closure of VSD and relief of subaortic obstruction was performed. Transvenous pacemaker implantation was required during the postoperative period due to complete atrioventricular (AV) block.

**Case 4–** A one-month-old female baby was referred to our center with severe respiratory distress and heart failure. She was already followed in a local hospital with the diagnosis of pneumonia and was, then, referred for further evaluation. She required mechanical ventilation support on admission due to severe respiratory distress. Physical examination also revealed the apical heart beat on the right chest, 3/6 systolic murmur, hepatomegaly, and mild peripheral edema. Echocardiographic findings included situs solitus dextrocardia, juxtaposition of the atrial appendages, ASD, large VSD, aberrant origin of left subclavian artery and pulmonary hypertension with a tricuspid regurgitation velocity of 4.3 m/s as well as criss-cross morphology. Corrective surgery for VSD was carried out, when she was three months old.

**Case 5–** A three-month-old male patient was admitted to our clinic with murmur. Echocardiography confirmed the findings of a criss-cross heart, multiple VSD, straddling tricuspid valve, patent ductus arteriosus, and mitral regurgitation. Cardiac catheterization revealed an additional diagnosis of peripheral pulmonary hypoplasia and stenosis with a pressure gradient of 70 mmHg between the main pulmonary artery and its branches. Oxygen saturation in the descending aorta was 96%. The patient was a refugee and did not attend the scheduled follow-up visits.

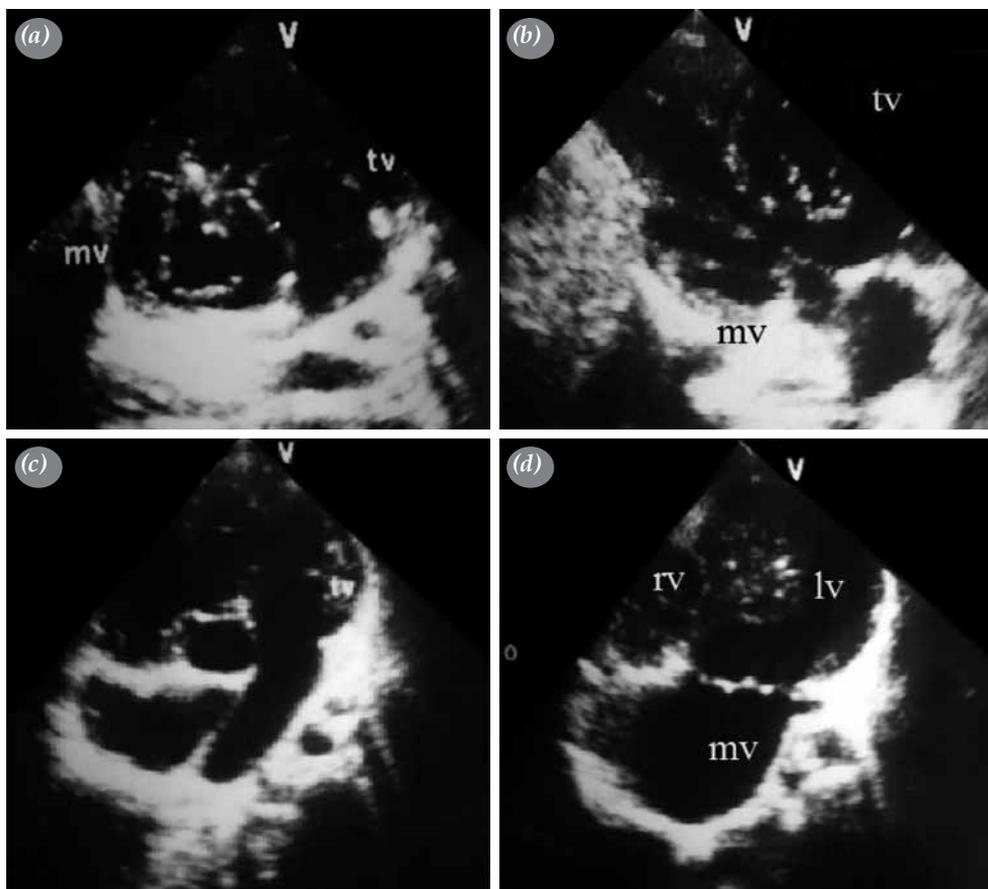
**Case 6**– A two and a half year old male patient with a previous history of modified left Blalock-Taussig shunt operation was referred to our center for further evaluation after cardiac catheterization and angiography which showed the narrowing of the shunt flow. His physical examination revealed cyanosis (O<sub>2</sub> saturation of 68% with pulse oxymetry), 2/6 systolic murmur on left sternal border, and a weak continuous murmur on the left upper chest. Echocardiography demonstrated criss-cross morphology, right ventricular hypoplasia, pulmonary atresia, restrictive VSD, ASD and straddling of the tricuspid valve, and diminished shunt flow. Bidirectional cava pulmonary shunt was recommended to the patient; however, he was lost in follow-up.

**Case 7**– A seven-year-old male patient was referred to our center for the indication of surgical treatment due to cyanotic congenital heart disease. On admission, cyanosis with an O<sub>2</sub> saturation of 62% and 4/6 systolic

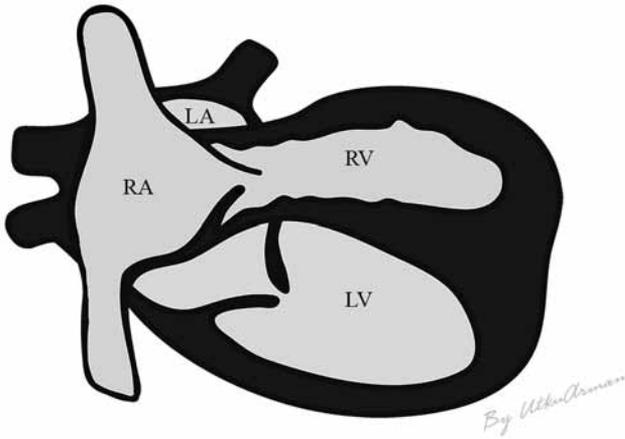
ejection murmur were present. Echocardiography and cardiac catheterization and angiography revealed a criss-cross heart, double-inlet left ventricle, double-outlet right ventricle, VSD, ASD, and severe PS (Figure 1). Bidirectional cavopulmonary shunt was carried out. He is currently being followed with an O<sub>2</sub> saturation of 88%.

## DISCUSSION

A criss-cross heart accounting for less than 1% of congenital heart defects was first described in 1961 by Lev and Rowlatt.<sup>[2]</sup> In the etiology of the disease, the ventricles twist around their long axis without concomitant movement of the atria, thus producing crossing of systemic and pulmonary venous blood streams each other at the atrioventricular level without mixing, giving the appearance of each atrium emptying into the contralateral ventricle (Figure 2).



**Figure 1.** Echocardiographic images of Case 7 demonstrating the (a) absence of parallelism of the atrioventricular valves [mitral valve was cut through the short axis, whereas tricuspid valve through the long axis] and (b) vice versa and the (c, d) absence of characteristic four-chamber view demonstrating only one of the atrioventricular valve seen.



**Figure 2.** Atrioventricular concordance with criss-cross morphology: right-sided morphologic right atrium connected to a left-sided morphologic right ventricle and left-sided morphologic left atrium connected to a right-sided morphologic left ventricle. RA: Right atrium, RV: Right ventricle, LA: Left atrium, LV: left ventricle

Although the exact etiology of a criss-cross heart has been not clearly understood yet, it seems to be due to abnormal twisting of the apex of the heart, while the base remains relatively fixed.<sup>[1,3,4]</sup> In several studies, connexin 43 (Cx43) gene mutations were associated with the pathogenesis of the disease.<sup>[3,4]</sup> Reaume et al.<sup>[3]</sup> reported malformation of the subpulmonary outflow tract in mice lacking gap junctions of Cx43 type. In another study conducted by Ya,<sup>[4]</sup> Cx43 deficient mice developed a criss-cross-like morphology, in which the craniomedial position was maintained by the right ventricle, resulting in a 90° rotation of the atrioventricular mass. On the other hand, in a study by De Luca et al.,<sup>[5]</sup> no pathogenic mutation in the Cx43 gene was detected in patients with a criss-cross heart.

The diagnosis of a criss-cross heart and associated cardiac defects can be successfully established with transthoracic echocardiography in most cases.<sup>[6,7]</sup> However, cardiac catheterization may be necessary in some to obtain pressure and oxymeter data or view additional defects. Studies with cardiac magnetic resonance imaging demonstrated clear visualization of the heart with criss-cross morphology and provided reconstructed three-dimensional images of the heart.<sup>[8]</sup>

Echocardiographic findings include inability to obtain a characteristic four-chamber view in any plane, displaying the crossing of long axes of atrioventricular valves as seen in the subcostal long axis or coronal plane sweep.<sup>[6,7]</sup> This anomaly can be also diagnosed prenatally with fetal echocardiography considering these findings.<sup>[9]</sup>

To the best of our knowledge, isolated criss-cross morphology has not been reported in the literature, that it is almost always associated with other severe cardiac anomalies. The majority of patients with a criss-cross heart have a hypoplastic tricuspid valve and right ventricle, a VSD, abnormal ventriculoarterial alignments (complete transposition of great arteries, double-outlet right ventricle), and pulmonary stenosis. Additional abnormalities include dextrocardia, straddling atrioventricular valve, subaortic stenosis, aortic arch obstruction, mitral stenosis, superior-inferior ventricle, and juxtaposition of atrial appendages.<sup>[6-13]</sup>

In our study, ventriculoarterial connection was concordant in four, discordant in one, double outlet in one and single outlet in one patient. In another study carried out by Fang et al.,<sup>[7]</sup> of 10 patients, all had abnormal ventriculoarterial connections including transposed in five and double-outlet right ventricle in the remaining patients.

Congenitally corrected transposition of the great arteries in criss-cross morphology was reported previously.<sup>[10-14]</sup> Our patient had a d-transposed aorta (right anterior aorta), described as “S,L,D” according to the Van Praagh notation system, which is a rare finding in ccTGA. In a study by Allwork et al.,<sup>[15]</sup> of the 32 patients with ccTGA, two had a right-sided aorta. Symons et al.<sup>[10]</sup> reported a case with a criss-cross heart presenting with ccTGA and a d-transposed aorta.

Of the seven patients, all had ventricular septal defect which was restrictive in two patients with right ventricular hypoplasia. Right ventricular hypoplasia and pulmonary stenosis are relatively common findings in criss-cross morphology. Fang et al.<sup>[7]</sup> reported the incidence of hypoplastic right ventricular and pulmonary stenosis in 30% and 60% of the patients, respectively. However, it is unclear that whether these are secondary or primary in the pathogenesis of a criss-cross heart.

Straddling atrioventricular valve and double-inlet ventricle were present in five of our patients, which were also reported in the literature with a criss-cross heart.<sup>[7]</sup>

Furthermore, juxtaposition of the atrial appendages is a rare malformation with the appendages both located on the left or right side of the great arteries reported with criss-cross morphology.<sup>[12]</sup> Juxtaposition of the atrial appendages were present in two of our patients and both had situs solitus dextrocardia.

Since it is almost always with other complex cardiac defects, patients usually present early in life

with cyanosis, murmur or with heart failure symptoms, as seen in our patients. The clinical outcome of a criss-cross heart predominantly depends on the underlying hemodynamic abnormalities. Surgical treatment varies from palliative correction to definitive anatomic correction. In the majority of the patients with a criss-cross heart, a two-ventricle repair may not be possible and these patients are staged toward a Fontan-type operation.

In conclusion, a criss-cross heart is a rare and complex anomaly which should be kept in mind to recognize during echocardiography. Inability to obtain a characteristic four-chamber view in any echocardiographic plane is diagnostic for this anomaly in most cases.

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