Case Report

Interesting and Rare Crisscross Heart

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ABSTRACT

The crisscross heart is a rare congenital heart abnormality often associated with other cardiac anomalies. Echocardiography is the first-line diagnostic tool, whereas magnetic resonance imaging and catheterization are used for complementary assessments. In the absence of significant pulmonary valve stenosis or hypoplasia in the tricuspid valve or the right ventricle, total repair or, otherwise, the Fontan procedure is recommended. ***(Iranian Heart Journal 2023; 24(3): 85-88)***

KEYWORDS: Crisscross heart, Congenital heart abnormality, Congenitally corrected transposition of great arteries

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A 22-year-old man was referred to us with a history of congenital heart disease. He had no significant cardiac symptoms and appeared well-developed. A physical examination revealed decreased S2 intensity and systolic murmurs at the left sternal border.

INVESTIGATION

Electrocardiography showed normal sinus rhythm, and the chest X-ray and computed tomography findings were unremarkable for prominent pulmonary vascular markings. At age 9, the patient had undergone catheterization, revealing that the course of the venous catheter was from the femoral vein to the inferior vena cava, the right atrium, and, finally, the left ventricle. Left ventricular injection showed opacification in the main pulmonary artery. The course of the arterial catheter was from the femoral artery, the aorta, and, finally, the right ventricle. The aorta was right and anterior to the pulmonary artery. Superoinferior ventricles, a ventricular septal defect, and valvular pulmonary stenosis were detected, too. The size of the left and right pulmonary arteries was acceptable.

At the current visit, the patient underwent transthoracic and transesophageal echocardiographic examinations. The atrial situs was solitus, the right ventricle was anterior and right to the left ventricle (D-loop ventricles), atrioventricular discordance was detected, the left atrium was connected to a twisted right ventricle, and the right atrium was connected to the left ventricle (fig 1). Additionally, the ventriculoarterial connection was discordant, and the aorta and the pulmonary artery were parallel and had D-malposition. The systemic ventricle (the

morphologic right ventricle) was severely enlarged, and there was moderate systolic dysfunction. Further, the systemic atrioventricular valve (the tricuspid valve) had moderate regurgitation. The echocardiographic examinations also demonstrated a moderately sized ventricular septal defect (1 cm) and moderate subvalvular and valvular pulmonary stenosis. The pulmonary artery branches were well-developed. The echocardiographic findings were compatible with the congenitally corrected transposition of the great arteries (TGA) and a crisscross atrioventricular valve.

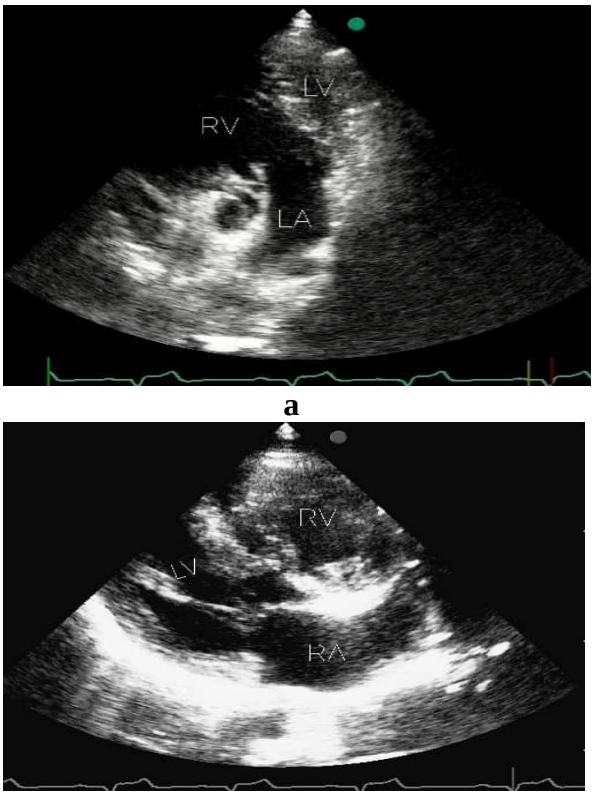

Figure 1: The images depict atrioventricular discordance. a) The LA is connected to a twisted RV. b) The RA is connected to the LV.

LA, Left atrium; RV, Right ventricle; RA, Right atrium; LV, Left ventricle

MANAGEMENT

We established a diagnosis of a corrected physiologic TGA and a crisscross atrioventricular valve. Given the absence of valvular and chamber hypoplasia and severe pulmonary valve stenosis, we recommended ventricular septal defect closure and pulmonary stenosis repair. The patient, however, refused surgery, obliging us to follow him up with regular periodic physical and echocardiographic examinations with no significant and specific cardiac symptom presentations.

DISCUSSION

The crisscross heart is a rare congenital heart abnormality defined by a spatial atrioventricular relation resulting in crossing ventricular inflow streams. [1] The incidence is about 0.1% of all congenital heart diseases and 8 per 1 000 000 live births. [1, 2] The crisscross heart is often accompanied by other cardiac anomalies, including ventricular septal defects, double-outlet right ventricles, TGA, tricuspid valve hypoplasia, right ventricular hypoplasia, pulmonary stenosis, and less commonly mitral or tricuspid straddling, mitral stenosis, subaortic stenosis, and aortic arch anomalies. [1, 3]

In the normal heart, the inflow streams through the atrioventricular valves are parallel. The tricuspid and mitral valves open during ventricular diastole; then, the atria drain into their respective ventricles. [1, 4]

In the crisscross heart (or superoinferior ventricles), the base of the heart is unchanged, whereas the ventricles appear to have rotated along their longitudinal axis. This condition positions each atrium into the contralateral ventricle, and the pulmonary and systemic venous streams cross each other at the atrioventricular level without any mixing. [3, 5]

The classification of the crisscross heart is based on atrioventricular and ventriculoarterial connections (fig 2 & 3) and is associated with other cardiac defects. There is often harmony between the segmental situs and atrioventricular alignment and also discordance in the

ventriculoarterial segment situs. [1,6] The most common type is characterized as atrial situs solitus, L-loop ventricles, and ventriculoarterial discordance. [7]

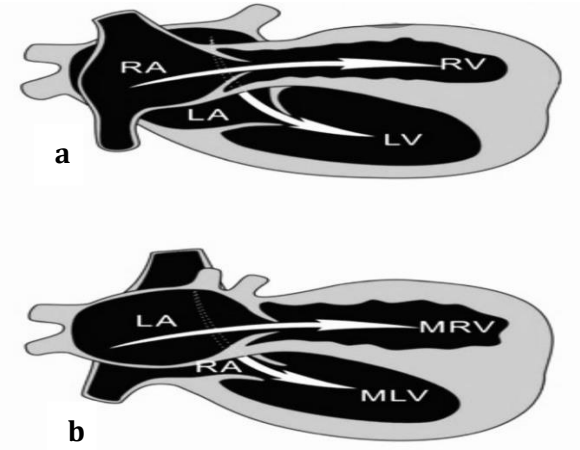

Figure 2: The images illustrate a) atrioventricular concordance with crisscross morphology and b) atrioventricular discordance with crisscross morphology.

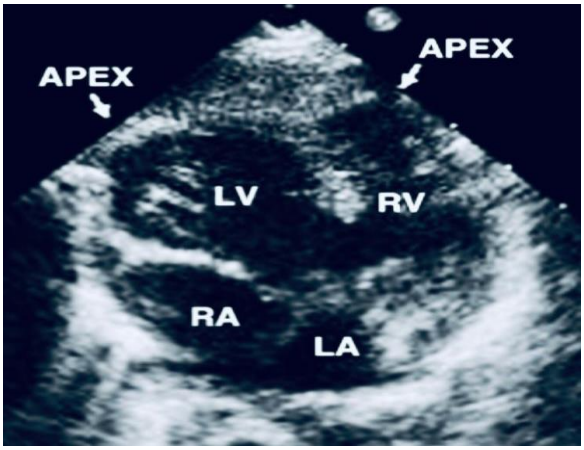

Figure 3: echocardiography view of atrioventricular discordance with crisscross morphology

Echocardiography is a first-line appropriate diagnostic tool for the crisscross heart when the parallel arrangement of the atrioventricular valve and ventricular inflow regions cannot be obtained in the 4-chamber view. Flow color can also assess intracardiac blood flow and recognize the crossed inflow streams in echocardiography. Magnetic resonance imaging is a complementary diagnostic imaging modality for detecting crossed atrioventricular valves and associated congenital defects, especially in challenging cases. Moreover, catheterization can show this crossing of the atrioventricular inflow streams. [1, 6]

Decision-making vis-à-vis crisscross heart management and the surgical approach is based on the severity of pulmonary stenosis, the adequacy of pulmonary blood flow, and the possibility of complete ventricles repair because of concomitant pulmonary stenosis and tricuspid valve or right ventricular hypoplasia. If the surgical method is possible (in a minority of patients), it is recommended to close the ventricular septal defect so as to achieve an alignment of the ventricles with the great arteries (usually the left ventricle with the pulmonary artery and the right ventricle with the aorta) and then perform the arterial switch on the cardiopulmonary bypass pump. In pediatric open-heart surgery on the cardiopulmonary bypass pump, a rise in the volume of body fluids occurs, especially in children with low weight and age, because of the stimulation of the inflammatory system and increased vascular permeability. Some studies have shown that the use of combined conventional ultrafiltration and modified ultrafiltration is related to improved hemodynamic status in patients and significantly decreases the duration of mechanical ventilation and inotropes requirement within 48 hours after surgery. A staged Fontan procedure is drawn upon if the ventricles are not suitable for repair. [1, 3, 8]

CONCLUSIONS

The crisscross heart is a rare congenital heart disease whose early diagnosis can be useful because of concomitant cardiac anomalies. Treatment decision is based not only on the disease itself but also on associated congenital cardiac abnormalities. Regular

life-long follow-ups are required regardless of the treatment methods selected.

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