

Criss-Cross Heart: A Case Report

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Abstract

Criss-cross heart was first described in 1974. It is a rare congenital heart malformation that occurs in 8 cases per 1,000,000 children, and represents only 0.1% of congenital malformations. The diagnostic methods of choice are transthoracic echocardiography, cardiac magnetic resonance (CMR), computed tomography angiography (CT) and, sometimes, cardiac catheterization. This report describes the case of a newborn with a criss-cross heart in addition to double-outlet right ventricle (RV), with poorly positioned vessels, in addition to atrial septal defect (ASD), interventricular septal defect, tricuspid valve dysplasia and persistent left superior vena cava. The exact etiology of this malformation is not known, but it seems to occur due to rotation of the ventricles in their longitudinal axis, not accompanied by rotation of the atrial and atrioventricular (AV) valves. This movement produces abnormal ventricular inlets, determining that the RV be positioned on a superior plane and the left ventricle on an inferior plane. Although the exact cause of this anomaly is still unknown, it is believed that a genetic abnormality may be leading to these cases: mutation of the Cx43 gene. Diagnosis of the case concerned was given by transthoracic echocardiography and computed CT of the aorta and pulmonary arteries, which showed, in addition to the criss-cross heart, other abnormalities, such as double-outlet RV, large ASD and ventricular septal defect (VSD).

Introduction

Criss-cross heart was first described in 1974, although it had been reported in 1961.^{1,2} It is a rare congenital heart malformation that occurs in 8 cases per 1,000,000 children, and represents only 0.1% of congenital malformations.^{3,4}

Criss-cross heart appears when, during the embryonic period, the heart rotates around its own axis, resulting in an anterosuperior RV and a posteroinferior left ventricle. Due to the complex structural alteration, diagnosis is complicated,

and the clinical picture depends on other congenital cardiac abnormalities, commonly present in cases of criss-cross heart.

The diagnostic methods of choice are transthoracic echocardiography, CMR imaging, computed CT and, occasionally, cardiac catheterization.⁵ Transthoracic echocardiography is usually the first test to be performed. It identifies the position and morphology of the four chambers and AV valves and the connections between vessels and chambers.⁶ Furthermore, in the performance of this method, there is, dynamically, the impression that the atrium empties into the contralateral ventricle due to the crossing of blood flows.^{7,8} CMR imaging and computed CT provide more detailed information and on other planes, such as coronal, axial and sagittal positions.⁹ Cardiac catheterization may be necessary to assess intracavitary or vessel pressures and oxygenation in different locations, in addition to ruling out septal defects not seen in other scans.⁵

The malformation regarding the rotation of the heart itself does not indicate a surgical approach, however, most cases are associated with other anatomic abnormalities, which need to be evaluated individually to determine the conduct. The most frequent associated malformations include: tricuspid valve and right ventricular hypoplasia, VSD, ventricular arterial discordance and pulmonary stenosis.⁷

In this report, we describe the case of a newborn with a criss-cross heart in addition to double-outlet RV, with poorly positioned vessels, in addition to ASD, interventricular septal defect, tricuspid valve dysplasia and persistent left superior vena cava.

Case report

Male child born on March 28, 2021, from home birth, was taken to a hospital in Colatina (ES) after birth, for evaluation. When examined by the attending physician, the heart test revealed an abnormality (saturation in the right upper limb = 92% and right lower limb = 92%). Transthoracic echocardiography was performed, suggesting a complex congenital heart disease with transposition of the great vessels with patent foramen ovale (PFO) ASD and large associated VSD.

Transferred to a reference pediatric cardiac surgery hospital in Vila Velha (ES) on March 30, 2021, for follow-up and treatment. Upon hospital admission to the pediatric intensive care unit (PICU), he had sucking issues, respiratory effort and drop in saturation during breastfeeding. On physical examination, he was afebrile, ruddy, acyanotic, diffuse toxic erythema, heart rate of 150 beats per minute, blood pressure of 60/35 mmHg in the right upper limb, 90/68 mmHg in the

Keywords

Congenital Heart Defects; Crisscross Heart; Heart Ventricles

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Manuscript received January 5, 2022; revised October 27, 2022; accepted January 11, 2023

Editor responsible for the review: Karen Saori Shiraishi Sawamura

DOI: <https://doi.org/10.36660/abcimg.2023282i>

right lower limb, 79/47 mmHg in the left upper limb and 79/55 mmHg in the left lower limb, systolic ejection murmur ++/6+ in the left sternal border, in addition to mild subcostal retraction.

Transthoracic echocardiography was performed on March 31, 2021, which showed: situs solitus, levocardia; two-valve AV concordance with rotation of the AV connection and crossed ventricular inflow streams (Figures 1 and 2); double-outlet RV ventricular-arterial coupling (Figure 3), with poorly positioned vessels; wide fossa ovalis ASD, 5.4 mm in its largest measurement; no flow acceleration, mean gradient of 1.7 mmHg, left-right flow; interventricular septum with overriding greater than 50% and apparent double infundibulum; large inlet VSD, 10 mm, no significant gradient; moderate dilation of the right chambers and mild RV hypertrophy; preserved biventricular systolic function assessed by qualitative analysis; dysplastic tricuspid valve with straddling and moderate regurgitation of this valve allowing estimating right ventricular systolic pressure at 55 mmHg, 8.5 mm tricuspid annulus; trivalvular aortic valve anterior and to the right, no significant systolic gradient, mild regurgitation; trivalvular pulmonary valve with no significant systolic gradient at the time, mid-systolic notch and mild regurgitation; discrete stenosis in the left pulmonary artery; persistent left superior vena cava.

While in hospital, the child presented clinical and radiographic signs suggestive of pulmonary hyperflow. Adjustments were made to diuretic doses and computed CT of the thoracic aorta and pulmonary arteries was performed on April 8, 2021 for better anatomic evaluation and approach planning. The scan revealed: situs solitus; levocardia; concordant systemic and pulmonary venous couplings; luminal reduction of the ostium of the left internal pulmonary vein — 6.4 mm²; double RV outlet ventricular-arterial couplings (Figure 4); left ventricle inferior to the RV (criss-cross) (Figure 5); presence of a muscle band close to the aortic outflow tract; large inlet VSD; large ASD (Figure 6) and aortic arch on the left and abdominal aorta positioned on the left. Figure 7 reveals three-dimensional tomography reconstruction showing rotation of the ventricles around the largest axis and Figure 8 reveals normal AV couplings.

The patient underwent pulmonary artery banding cardiac surgery on April 20, 2021, uneventfully. In the immediate postoperative period, the patient developed supraventricular tachycardia, which improved after adjusting the temperature, required low-dose epinephrine, and presented oliguria requiring diuretic solution. The patient presented a positive outcome, allowing the diuretic solution and adrenaline to be suspended, and was extubated on April 22, 2021, uneventfully. Control transthoracic echocardiography on April 22, 2021 showed effective pulmonary banding.

Discussion

The exact etiology of this malformation is not known, but it seems to occur due to rotation of the ventricles around their longitudinal axis, not accompanied by atrial rotation and AV valve rotation. This movement produces abnormal ventricular inlets, determining that the RV be positioned on a superior plane and the left ventricle on an inferior plane.¹⁰ The other

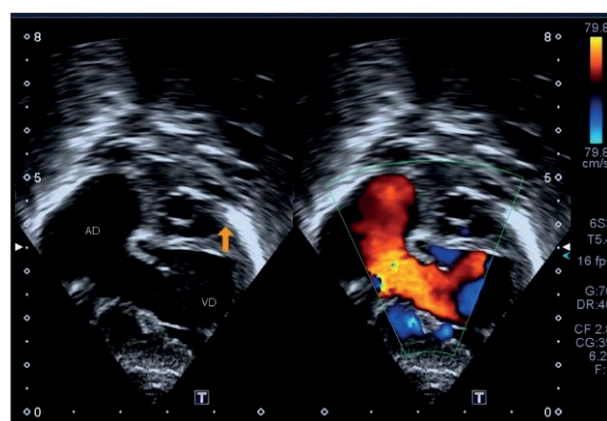


Figure 1 – Echocardiographic image showing AD-RV connection rotation. RV: right ventricle; RA: right atrium.

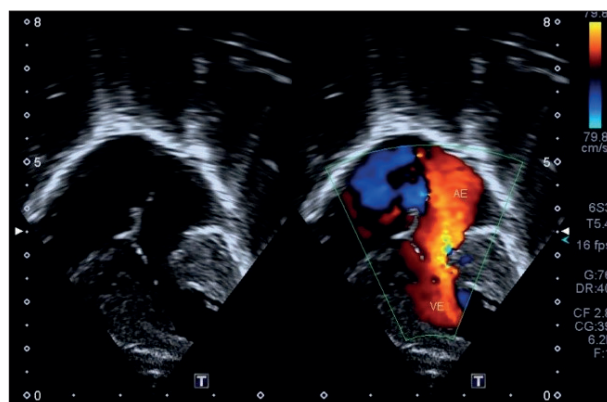


Figure 2 – Echocardiographic image showing LA-LV connection rotation. LA: left atrium.

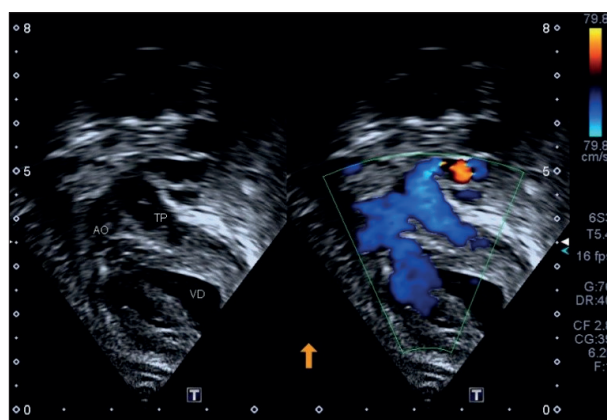


Figure 3 – Echocardiographic image showing double outflow of the RV. RV: right ventricle; AO: aorta; PT: pulmonary trunk.

anomalies normally found are hypoplasia of the right tricuspid valve, pulmonary stenosis, inlet VSD and abnormal ventricular-arterial coupling. Discordant coupling is more frequent, and double-outlet RV is rare.^{10,11}

Case Report

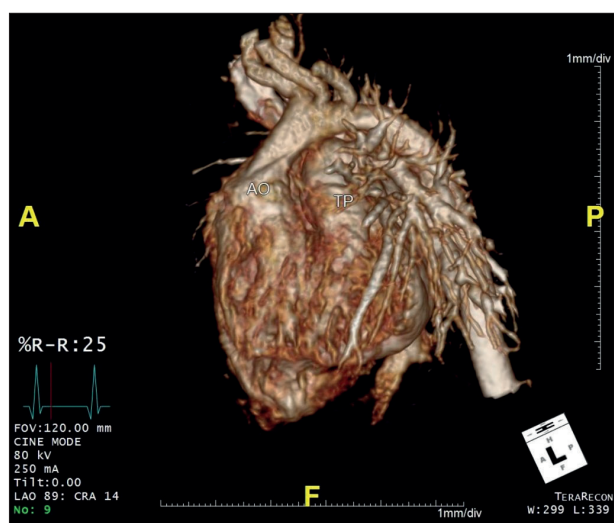


Figure 4 – Double Outlet RV. AO: aorta; PT: pulmonary trunk.



Figure 5 – Left Ventricle Inferior to RV. RV: right ventricle; LV: left ventricle.

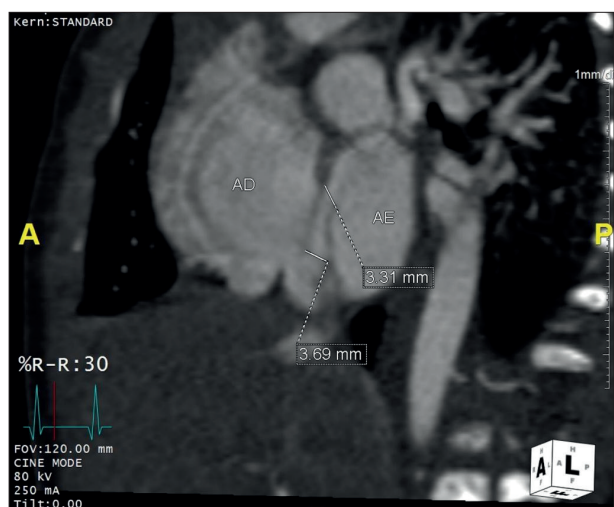


Figure 6 – Wide Interatrial Communication. RA: right atrium; LA: left atrium.

Although the exact cause of this anomaly is not yet known, it is believed that a genetic abnormality may be leading to these cases — mutation of the Cx43 gene — and the exclusion of this gene would lead to a delay in the dextroposition of the heart, thus causing right ventricular defect, and not taking it into the correct position.¹²

The diagnosis of the case in question was given by through transthoracic echocardiography and computed CT of the aorta and pulmonary arteries, which showed, in addition to criss-cross heart, other abnormalities, such as double-outlet RV, and large ASD and VSD. Due to the presence of double-outlet RV, in view of the high systemic resistance leading to the flow preferentially through the pulmonary trunk, with exacerbated pulmonary hyperflow, it was decided to perform the banding of the pulmonary arteries in order to increase or, at least, equalize the pulmonary resistance and, thus, cause the blood to be ejected preferentially to the systemic arterial bed instead of the pulmonary venous bed, thus protecting the pulmonary arterial vasculature.¹³ The procedure was uneventful, but, due to other neonatal problems, the child had to remain hospitalized after hospital discharge from a cardiovascular point of view, but remained hemodynamically stable and in room air, which demonstrates the effectiveness of the procedure performed, in addition to the echocardiography postoperatively. An outpatient follow-up schedule was created so that, in the future, the best therapeutic strategy could be defined.

Author Contributions

Conception and design of the research: Potratz MO, Garbo LZ, Pessimilio KP, Loss AS, Ambrozim CB, Lima ALTA, Rocha DL; acquisition of data and critical revision of the manuscript for intellectual content: Potratz MO, Garbo LZ, Rocha DL; writing of the manuscript: Lima ALTA.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

Sources of Funding

There were no external funding sources for this study.

Study Association

This study is not associated with any thesis or dissertation work.

Ethics Approval and Consent to Participate

This article does not contain any studies with human participants or animals performed by any of the authors.

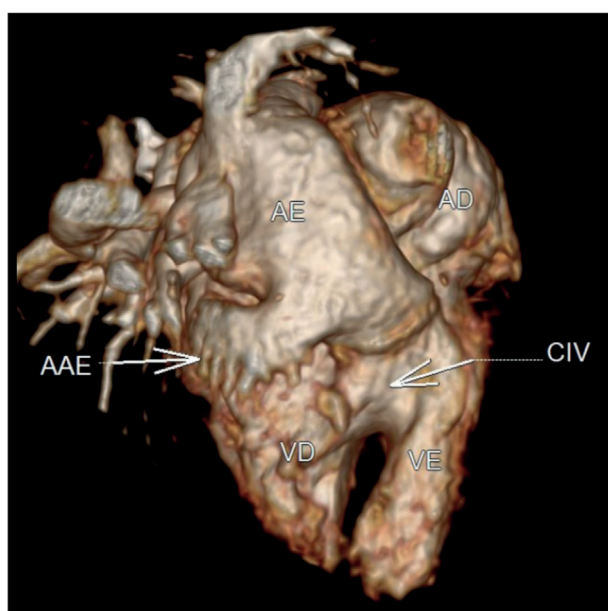


Figure 7 – Three-dimensional tomography reconstruction showing the rotation of the ventricles in the major axis. RV: right ventricle; VSD: ventricular septal defect; LV: left ventricle; LA: left atrium; RA: right atrium; LAA: left atrial appendix.

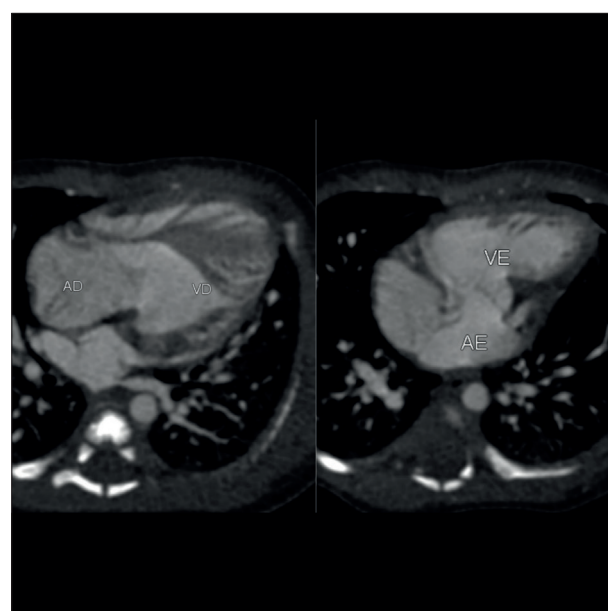


Figure 8 – Tomography image showing normal AV connections. RV: right ventricle; RA: right atrium; LV: left ventricle; LA: left atrium.

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