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Cardiac CT of a Criss-Cross Heart: A Case Report

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Abstract: ***A criss-cross heart is an extremely rare congenital anomaly that is commonly associated with complex cardiac defects. The literature on imaging of such a condition is limited. In particular, cardiac CT imaging of criss-cross heart is sparse. We present a case of a full-term infant with criss-cross heart, in which cardiac CT delineated the complex heart anatomy and confirmed known echocardiographic findings. Utilizing cardiac CT in criss-cross heart increases the treating physician's confidence before embarkin g on a final surgical approach.***

Keywords: Congenital heart disease, Cardiac CT, Criss-cross heart, Ventricular septal defect

1. Case Report

A full-term male infant that was transferred to our tertiary care center with progressive cyanosis and severe hypoxemia that started at the age of two days. The baby was intubated and managed with prostaglandin infusion, which mildly improved his condition. The initial transthoracic echocardiographic assessment revealed: malpositioned great arteries, a floppy interatrial septum, a 4 mm atrial septal defect (ASD) with left-to-right shunting of blood, a 5 mm perimembranous ventricular septal defect (VSD), atrioventricular concordance, ventriculoarterial discordance, mild subpulmonic stenosis, trivial tricuspid and pulmonary regurgitation jets, dextrocardia and a normal left-sided aortic arch.

Computed tomography (CT) was performed two days after echocardiography, after administration of intravenous contrast material, to delineate both the arterial and venous circulations. The CT was performed on a dual-source 128 multidetector scanner, using a non-electrocardiographic-synchronized high-pitch helical acquisition. A low estimated effective radiation dose of 0.2 millisieverts was administrated to conduct the study. CT clearly delineated the cardiac anatomy and confirming most of the echocardiographic identified abnormalities. The patient was confirmed to have atrial situs solitus (Figure 1). There was L-bulboventricular looping, with aleft-sided morphologic right ventricle resided on top of a right-sided morphologic left ventricle (Figure 2). The right atrial inlet had an opening that was directed to the left ventricle, while the left atrial inlet had an opening that was directed to the right ventricle (Figure 3). Malpositioned great arteries were noted (Figure 4). This arrangement resulted in atrioventricular concordance and ventriculoarterial discordance. The ASD and VSD were not identified on CT. However, a small patent ductus arteriosus (PDA) was seen (Figure 5).

At the age of 8 days, the infant underwent surgical correction. The surgery consisted of Jatene arterial switch procedure, with LeCompte maneuvering. The baby tolerated the procedure. The patient remained vitally stable throughout the hospital course and was discharged uneventfully.

2. Discussion

The criss-cross heart is an extremely rare congenital cardiac condition, in which the flow from the atria is directed to opposite ventricles. The ventricles are positioned in an unusual superior-inferior relationship to each other, instead of the typical side-by-side arrangement (1-3). Such an anomaly results from abnormal rotation of the ventricular components about their long axis due to unclear causes. With such an abnormality, the flow from the systemic and pulmonary venous systems intersects at the ventricular inlet level without mixing of blood(1).The criss-cross heart is typically associated with a multitude of congenital cardiac defects, such asVSD, transposition of the great arteries, right ventricular hypoplasia, pulmonary stenosis and several others (1,4-6).

A segmental approach in evaluating pediatric congenital heart disease is of utmost importance, especially those cases expected to have complex heart anatomy (7,8). This is especially true of criss-cross hearts, where there may be atrioventricular and ventriculoarterial concordance or discordance (1,3,9). Correct understanding of the segmental anatomical relationships of the cardiac structures in criss-cross hearts will lead proper understanding of the resultant hemodynamics, which is important in formulating a final management plan. In our presented case, and despite atrial situ solitus and L-bulboventricular looping, there was atrioventricular concordance, a result of the criss-cross configuration. With the great arterial malposition, there was ventriculoarterial discordance, resulting in classic transportation of the great arteries physiology.

The application of cardiac CT in pediatric congenital heart disease has been recently expanding (10,11). With the newer generations of CT, the cardiac assessment may be performed with very low radiation doses, much lower than what could be achieved by cardiac catheterization (10). Additionally, the new CT scanners are very quick in acquiring images, a feature that is advantageous in enabling the imaging of patients with no sedation, during free breathing and with fewer motion artifacts (11). The obtained CT images are currently of high quality and can be reviewed in multiple reconstruction planes.

Although most reported of criss-cross hearts have been assessed by echocardiography (1,4,12,13) and a few MRI reports of criss-cross hearts have been published(14,15), the

CT literature on this condition is very sparse. To our knowledge, only one small study of five criss-cross hearts cases was published; however, the patients in this study were imaged with older generation CT platforms (6). In our case, CT yielded more confidence in the pre-operative understanding of this complex anomaly, helping the surgical team in planning the most appropriate surgical approach.

3. Disclosures

None.

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Figures and Figure Legends

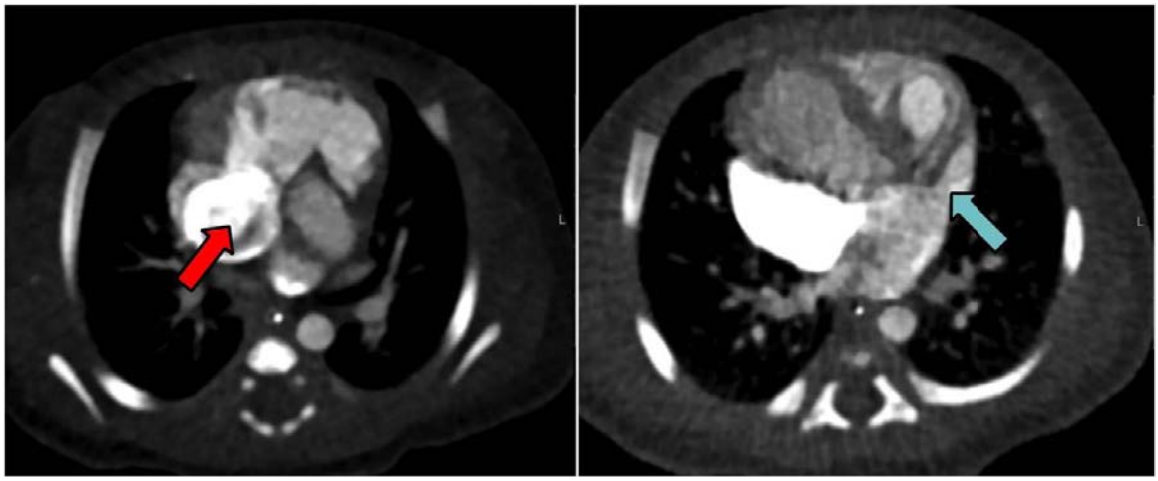

Figure 1: There is a normal position of the right atrium (red arrow) and left atrium (blue arrow), consistent with atrial situs solitus

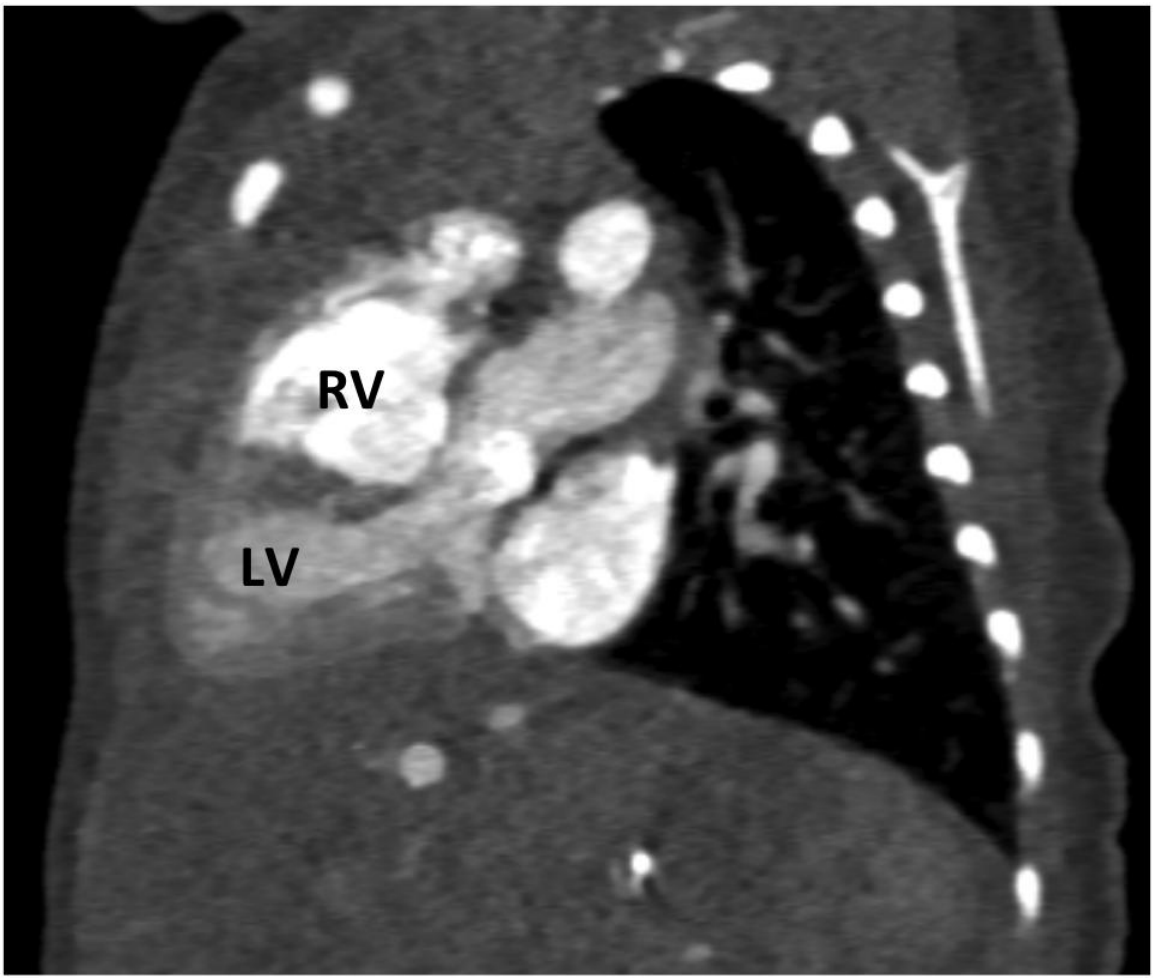

Figure 2: This sagittal oblique image shows that the left-sided morphologic right ventricle (RV) resides on top of a right-sided morphologic left ventricle (LV)

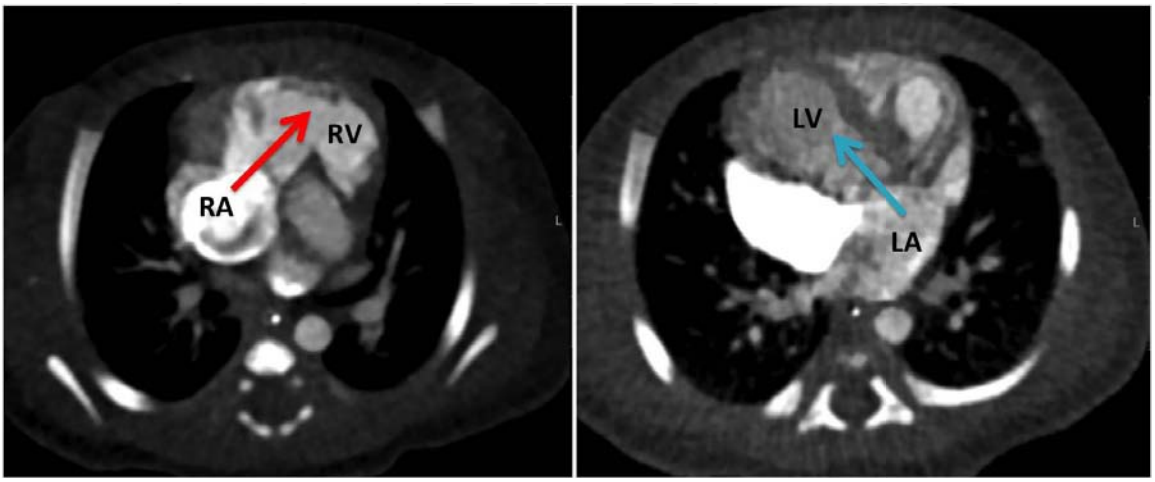

Figure 3: The right atrium (RA) opens on (red arrow) the left-sided morphologic right ventricle (RV), while the left atrium (LA) opens on (blue arrow) the right-sided morphologic left ventricle (LV). This arrangement results in atrioventricular concordance.

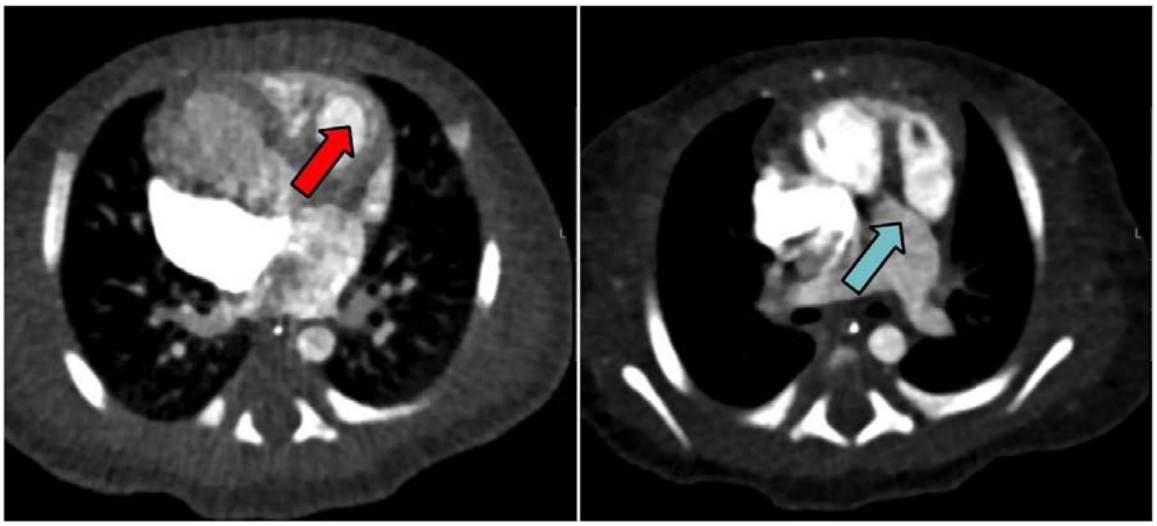

Figure 4: The aorta (red arrow) arises from the left-sided morphologic right ventricle, while the pulmonary artery (blue arrow) arises from the right-sided morphologic left ventricle (LV). This arrangement results in ventriculoarterialdiscordance.

Figure 4: There is a short and small communicating channel that connects the descending aorta with the proximal left pulmonary artery (circle), consistent with a patent ductus arteriosus (PDA)