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Case Report

An Intriguing Case of a Twisted and Tilted Heart

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Abstract

Background: Diagnosing congenital cardiac malpositions can be complex, especially when multiple anomalies coexist. We report an unusual case of supero-inferior (upstairs-downstairs) ventricular arrangement with a criss-cross inlet, in a neonate with D-looped transposition of the great arteries (D-TGA), double outlet right ventricle (DORV), and hypoplastic aortic arch with coarctation of the isthmus.

Case Presentation: The patient, diagnosed antenatally with DORV, D-TGA, and hypoplastic arch, was born in stable condition and started on prostaglandin E1 infusion. Postnatal echocardiography and CT angiography revealed a criss-cross heart (CCH) with supero-inferior ventricles. At 2-3 weeks, the patient underwent aortic arch repair and pulmonary artery (PA) banding. Subsequently, balloon atrial septostomy caused an intimal tear in the inferior vena cava (IVC), requiring IVC stenting. Over several months, PA band revision and selective left PA banding were necessary due to recurrent band loosening and stenosis. A Jatene arterial switch and ventricular septal defect (VSD) repair are planned after the infant reaches appropriate weight.

Investigations: Imaging confirmed situs solitus with atrioventricular concordance and ventriculoarterial discordance. A large VSD without inlet valve straddling, double outlet right ventricle, and plethoric lung fields were noted. The criss-cross inlet was characterized by a misaligned right atrioventricular connection, with the right ventricle positioned superiorly and anteriorly to the left ventricle (upstairs-downstairs arrangement).

Management and Outcome: Despite three sternotomies, including PA banding and multiple revisions, the child remains in high-dependency care, with plans for future surgery. Multidisciplinary team management was essential in navigating this challenging case.

Conclusion: This case highlights the intricate nature of criss-cross heart with supero-inferior ventricular arrangement. Diagnosing these rare congenital anomalies requires a combination of imaging modalities, and management involves tailored surgical interventions to address the associated malformations. Early recognition and multidisciplinary collaboration are key to improving outcomes in such complex cases.

Keywords: *Criss-cross heart, Supero-inferior ventricles, D-TGA, Double outlet right ventricle, Congenital heart disease.*

Abbreviations and Acronyms

CCH, Crisscross heart; **SIV/UDV**, Supero-inferior / upstairs-downstairs ventricle; **LV**, Left ventricle; **RV**, Right ventricle; **LA**, Left atrium; **RA**, Right atrium; **D-TGA**, D looped transposition of great arteries; **DORV**, Double outlet right ventricle

Introduction

Diagnosing Cardiac Malpositions are sometimes quite tricky and challenging, because many a times they don't follow any stipulated rules, plus the presence of more than one anomaly occurring simultaneously, makes things even more cumbersome. This is a case of similar topographical complexity, where we describe an ante-natal diagnosis of supero-inferior / upstairs-downstairs ventricular arrangement with a semi criss-cross inlet in a case of D-TGA with DORV and Hypoplastic Arch with Coarctation of the isthmus. (1-13)

Case Presentation

Antenatally known to have a DORV, d-TGA with hypoplastic Arch. Born in good condition, started on dinoprostone (prostin) soon after birth. Echocardiogram and CT Angiogram done soon after birth revealed (CCH) Criss-Cross Heart with an upstairs-downstairs / supero-inferior ventricular arrangement. (see Fig. 1a,1b & 1c)

This child subsequently experienced a highly complex inpatient course, during which the following events occurred in chronological order:

- At 2-3wks of age, underwent Arch Repair and PA banding.
- At 2months of age, balloon atrial septostomy attempted but the baby sustained an intimal tear in the IVC due to an unfortunate complication and an IVC stent was later put in
- At 3months of age, PA band was found to be lose, hence revision PA banding with atrial septectomy was done.
- At 4 months of age, Later PA band was again found to be relatively lose and the MPA and proximal segment of RPA was found to be mildly stenotic but with plethoric lung fields, hence a selective LPA banding was done.
- Planning for switching the great arteries (Jatene arterial switch operation) and repair the VSD, for now to let baby grow as much as possible in the meantime.

The Child currently remains in High dependency unit care, on Bilevel pressure support, gradual weight gain (Nasogastric feeds) whilst tolerating only 5-10min of pressure relief.

Investigations

Chest X-ray – cardiomegaly with plethoric lung fields

Echocardiogram / CT angiogram –

Situs solitus, levocardia.
Normal connection of pulmonary and systemic veins into the Left Atrium and Right Atrium, respectively.
Patent Foramen Ovale.
All four chambers could not be properly visualized in the apical four-chamber view.
Concordant Atrio-Ventricular arrangement, Discordant Ventriculo-Arterial arrangement with a nearly semi-criss-cross inlet.
The Left Atrium to Left Ventricle inlet is normally aligned.
However, the Right Atrium to Right Ventricle inlet appears diagonally misaligned, crossing the midline axis.
No significant Atrio-Ventricular valve regurgitation.
Transposition of the great arteries with the Aorta positioned anteriorly and to the right of the Pulmonary Artery.
Large Ventricular Septal Defect, comprising nearly half of the Inter-Ventricular Septum, with no straddling of the inlet valves.
Double Outlet Right Ventricle commitment of the great arteries, with no aorto-mitral continuity.
"Upstairs/downstairs" ventricular arrangement: Right Ventricle is superior and anterior, Left Ventricle is inferior and posterior, with subaortic and subpulmonary conus.
Overriding Pulmonary Annulus.
Normal coronary artery origins.
Single Right Superior Vena Cava.

Laminar flow through the aortic arch (post-arch repair), with mild stenosis of the Main Pulmonary Artery and Right Pulmonary Artery.
Left Pulmonary Artery banded – vmax 3.5 m/s.
Left-sided Aortic Arch.
Confluent pulmonary artery branches. (See Fig. 4a, 5a, 6a to 6k)

The tracheobronchial tree is patent and normal calibre.

There are features of pulmonary plethora and oedema, perihilar vessels are engorged and there is scattered posterobasal dependent atelectasis which is more pronounced and segmental in distribution in the left lower lobe.

Inferior Vena Cava stent partly imaged but patency cannot be determined.

Management and Outcomes

At first the aortic arch was repaired, and the PA was banded, then balloon atrial septostomy was attempted but ended up with an unfortunate complication when the balloon burst, an alternate sized balloon was not available for rescue, the burst balloon that was stuck inside the IVC could not be extracted and led to an intimal tear in the IVC, which later needed stenting. PA band was then found to be lose, so 2nd sternotomy performed for atrial septectomy and PA band revision, on follow up PA band was again found to be relatively lose, plus the MPA and the proximal RPA were mildly stenotic. CT thorax showed plethoric lungs with unprotected flow via the LPA, hence a discussion regarding placing a flow regulator in the LPA was made, but due to vessel calibre and potential risk of migration, a 3rd sternotomy performed to selectively band the LPA.

The next plan for now, is to allow the baby grow as much as possible and perform VSD repair and do a Jatene arterial switch operation.

This baby has experienced a tumultuous journey since birth, with numerous multidisciplinary team discussions and back and forth theatre visits and has taken the brunt of going through 3 sternotomies within the first 6 months of life. Such is the complexity of a twisted heart.

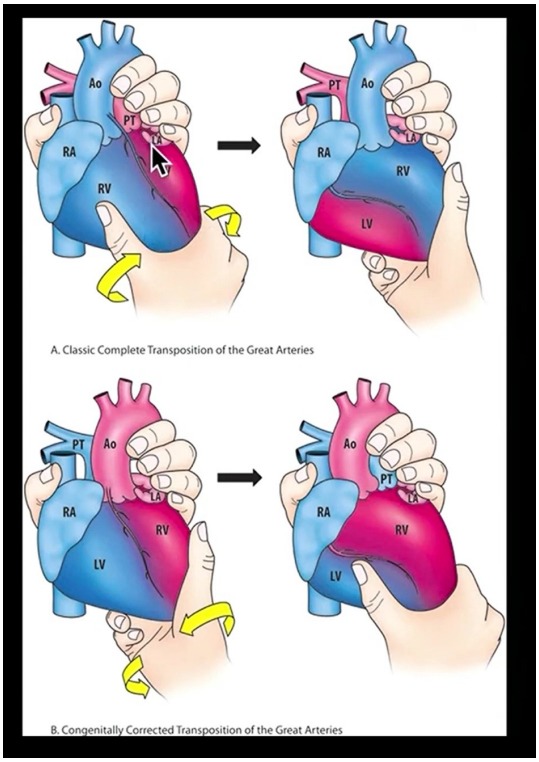

Figure 1a. *Anomalous cardio embroyogenetic mechanisms of 'TWISTING' along the long axis of heart, seen in cases of (CCH) Criss-Cross Heart in dTGA and l-TGA / ccTGA.*

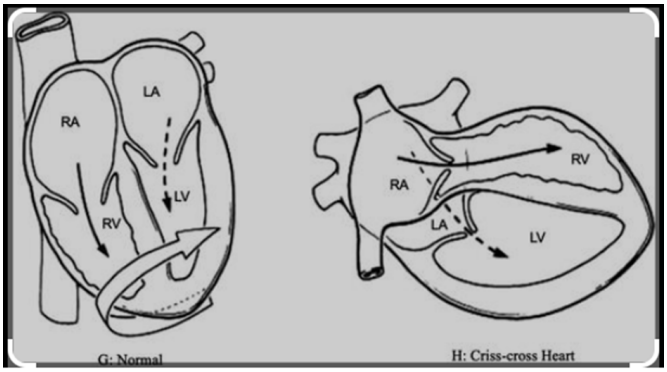

Figure 1b. *Criss-Cross Heart – misaligned inlet valves that diagonally crossing midline.*

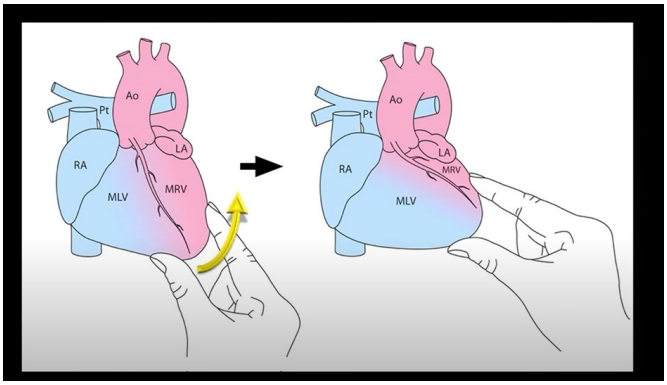

Figure 1c. *Mechanism of 'TILTING' along the long axis of heart, seen in cases of (UDV) Upstairs-Downstairs Ventricles / (SIV) Supero-Inferior Ventricles.*

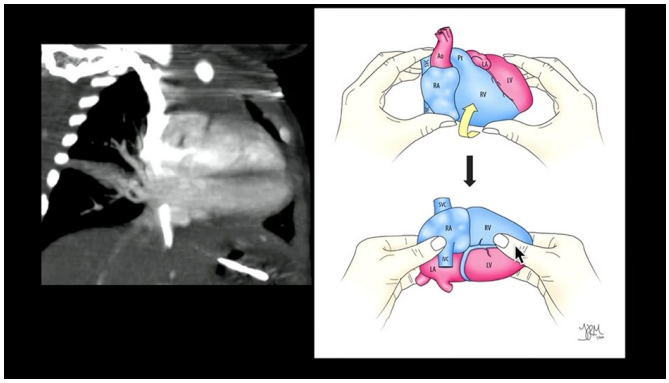

Figure 2a. *(TTH) Topsy Turvy Heart – image depicting 'ORGANO-AXIAL MALROTATION' of the heart in the postero-inferior direction.*

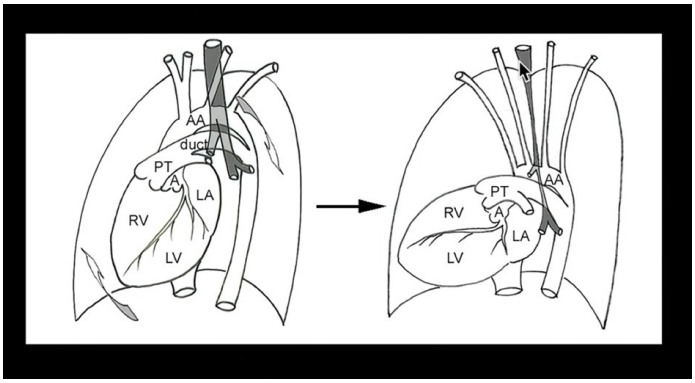

Figure 2b*. Due to malrotation, causes elongation of the neck veins and airways.*

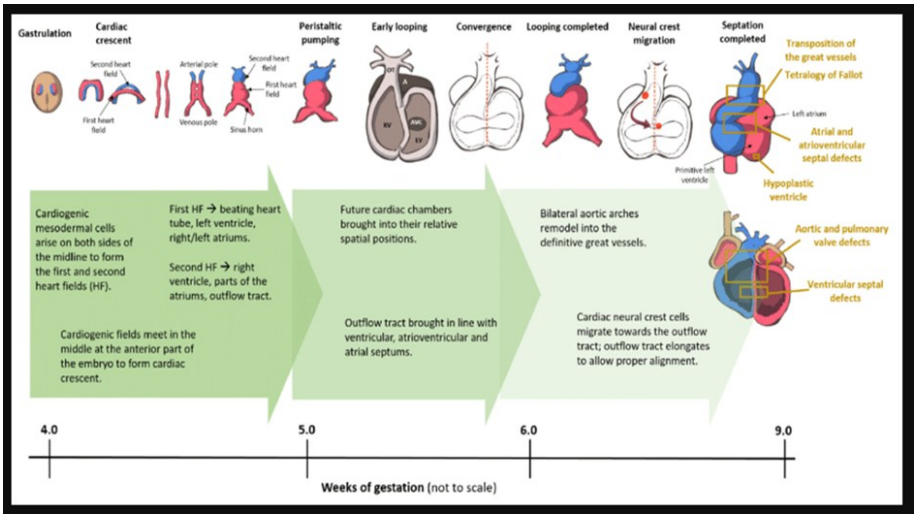

Figure 3. *Cardiac Embryology timeline.*

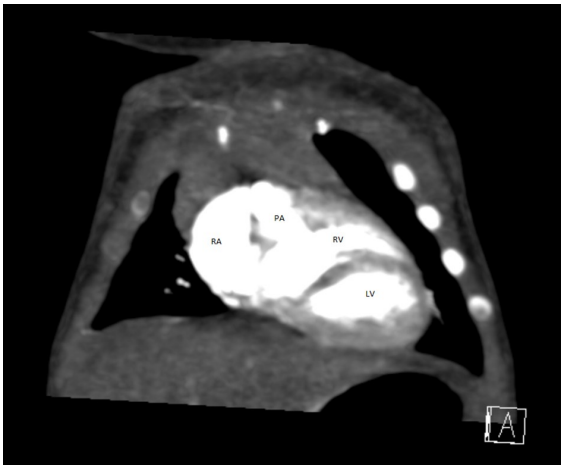

Figure 4a. *CT angiogram demonstrating upstairs- downstairs arrangement of Right Ventricle & Left Ventricle.*

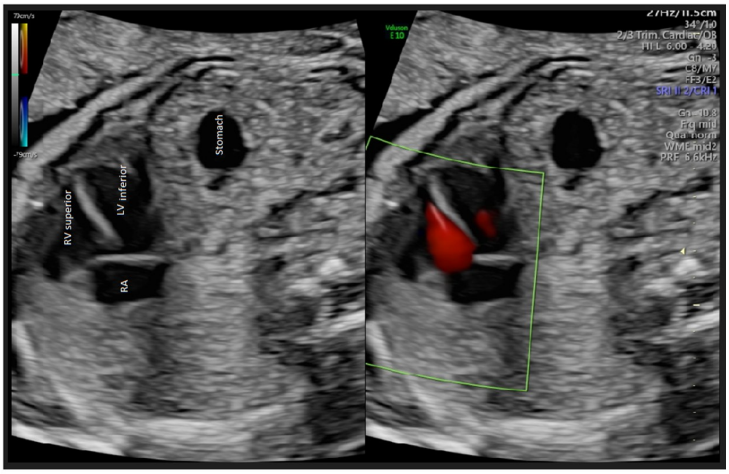

Figure 5a. *Fetal Echo demonstrating RV-LV supero-inferior arrangement*

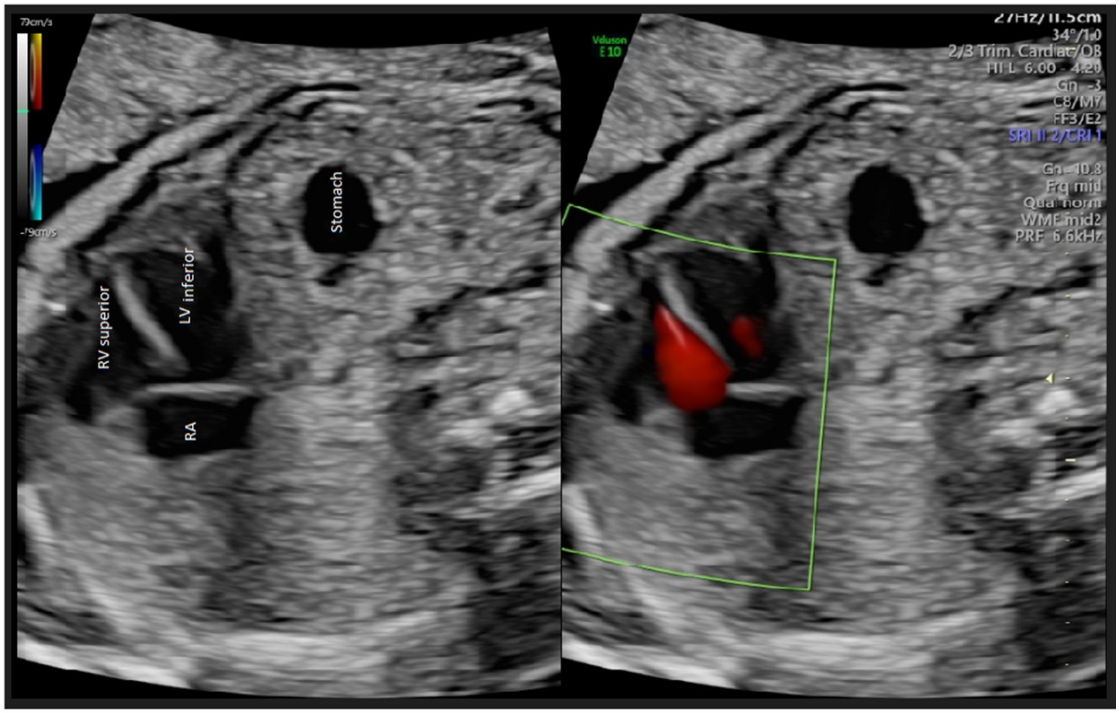

***Figure 5b**. Fetal Echo demonstrating RV-LV supero-inferior arrangement.*

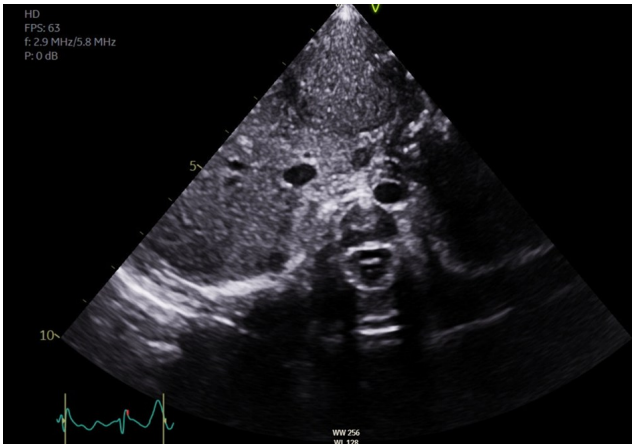

***Figure 6a**. 2D TTE situs solitus.*

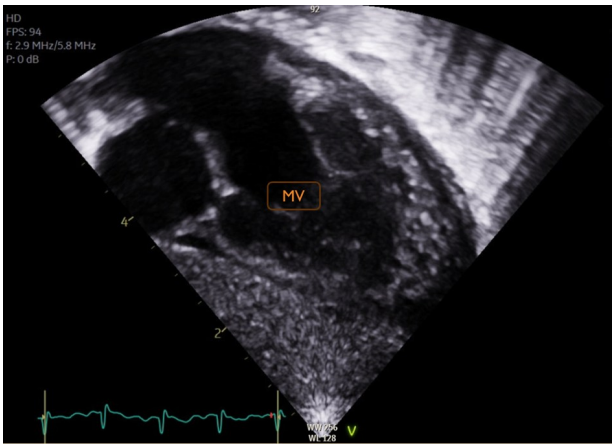

***Figure 6b.** Subcostal view 2D TTE – showing normally aligned LA inlet into LV.*

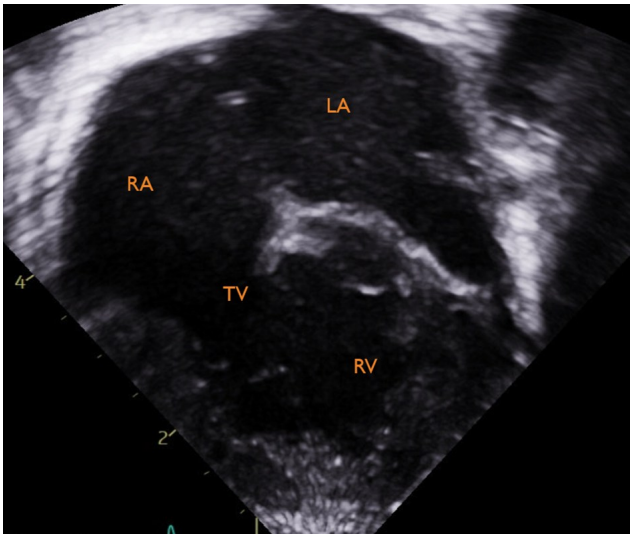

***Figure 6c**. Apical 4C view 2D TTE – not feasible to see all 4 chambers in this view.*

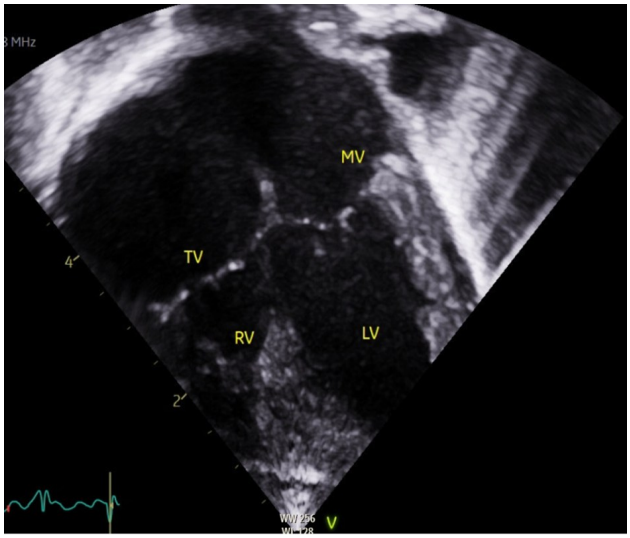

***Figure 6d**. A4C 2D TTE - not able to visualise all 4 chambers properly in A4C view.*

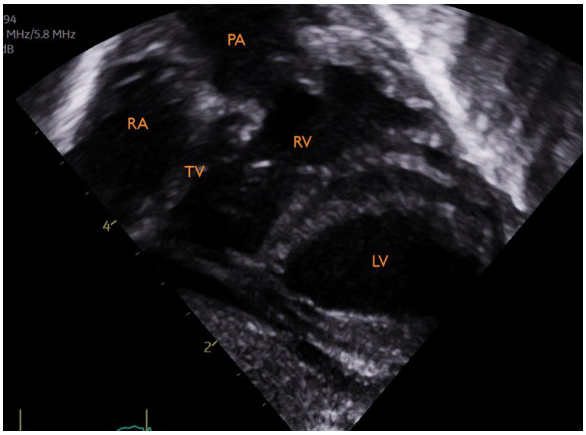

Figure 6e. *2D TTE - misaligned RA to RV inlet (criss inlet) with RV sitting on top of LV (upstairs-downstairs ventricular arrangement)*

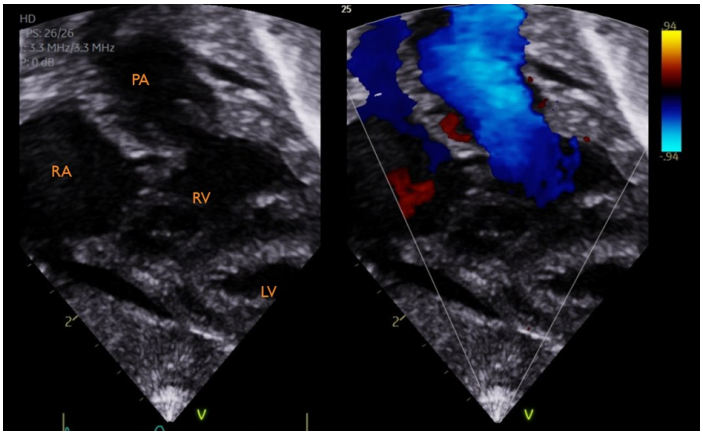

Figure 6f*. 2D TTE - MPA dilated seen posteriorly with no LVOT .*

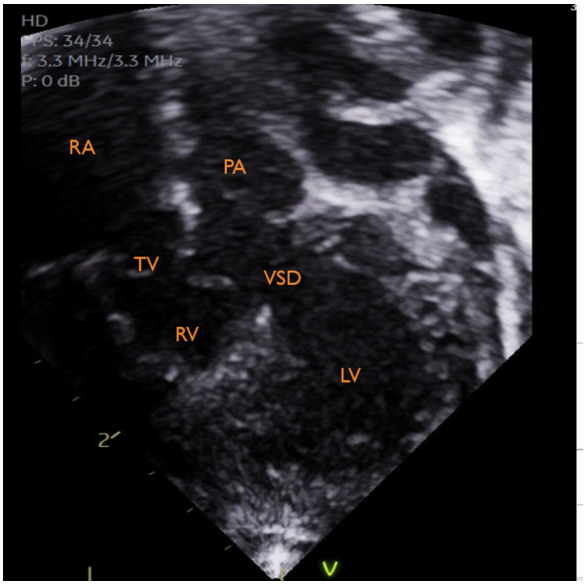

Figure 6g*. 2D TTE - LVOT is via the large VSD.*

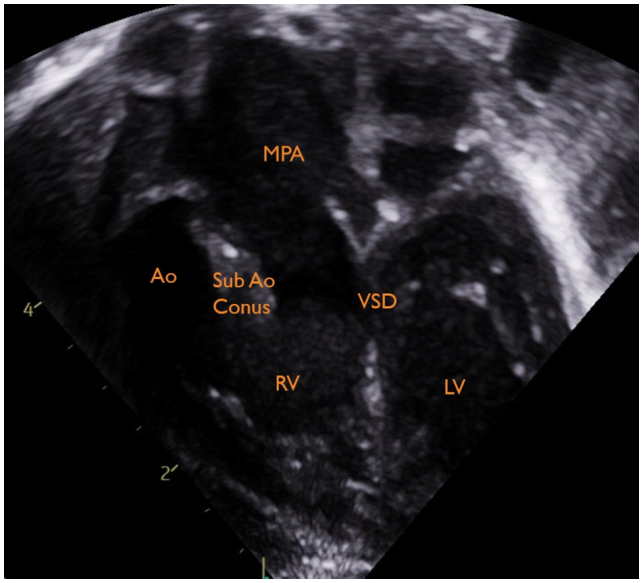

Figure 6h. *2D TTE – Double commitment of great vessels into the RV with no aorto-mitral continuity.*

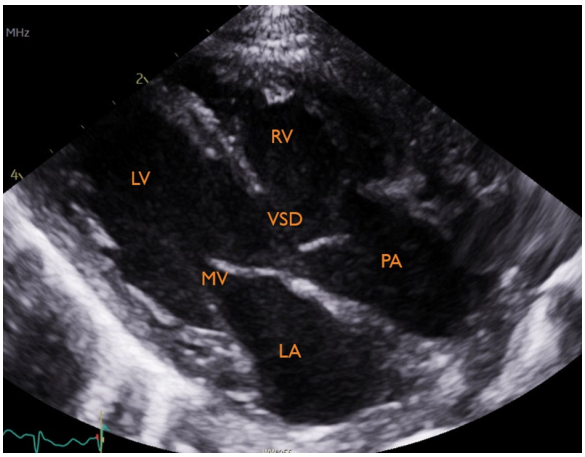

Figure 6i*. PLAX 2D TTE – demonstrating no aorto-mitral continuity and over-riding PA .*

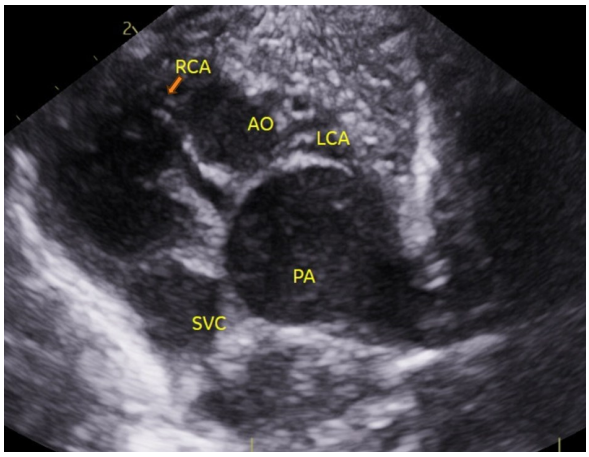

Figure 6j*. PSAX 2D TTE – demonstrating D-TGA configuration of great vessels with the aorta arising anterior and right side of PA.*

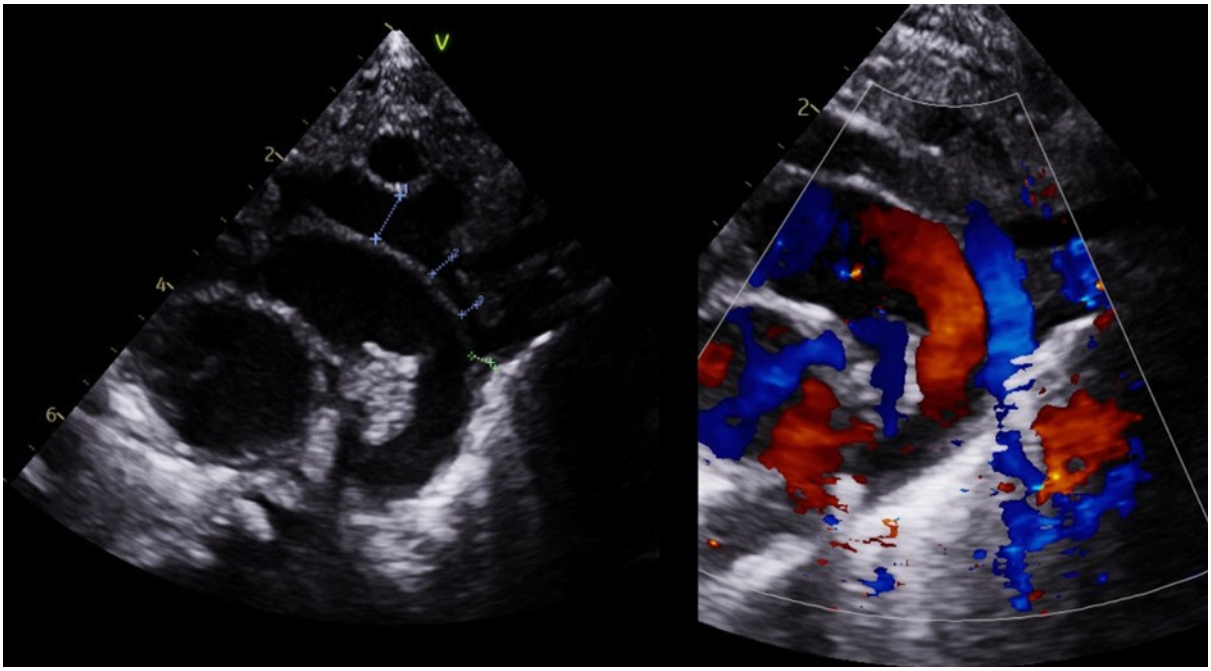

Figure 6k. *Suprasternal 2D TTE – demonstrating hypoplastic aortic arch and CoA at the isthmus.*

Discussion

(CCH) Criss-Cross Heart and (SIV) Superior-Inferior ventricle / (UDV) Upstairs-Downstairs ventricle, are the result of a certain anomalous mechanisms of cardiac embryogenesis, such as 'twisting' and 'tilting' along the long axis of the heart, respectively. (1) CCH is an extremely rare congenital heart anomaly characterized by crossed ventricular inlet without atrioventricular discordant connections. It was first identified by Lev and Rowlatt in 1961. (2) To date, there have been only 150-250 documented cases in the literature. It accounts to around 0.1% of all congenital heart defects. (2,3) The precise developmental mechanisms remain unclear. However, one hypothesis proposed by Anderson et al. suggests that it may result from either clockwise or counter-clockwise rotation of the heart during the post-septational phase. (4,5,11) In approximately 80% of cases, atrioventricular concordance and ventriculoarterial discordance are observed. A ventricular septal defect is present in nearly all instances. Additional anomalies may include pulmonary stenosis, straddling of the atrioventricular valves, tricuspid atresia, double outlet right ventricle, and congenitally corrected transposition. (6)

In Upstairs-Downstairs / Supero-Inferior ventricle - the interventricular septum lies in the horizontal plane, thereby making anteriorly placed RV to sit on top of posteriorly based LV. Not all CCHs have superior-inferior ventricles and vice versa is also true. However, both conditions can co-exist depending on the degree to which the heart twists and tilts. (7)

These terminology of 'twisting' causing 'CCH' and 'tilting' causing SIV / UDV; has been coined by the international Society Of Nomenclature of Paediatric & Congenital Heart Diseases (established in the year 2000. (8)

Recent literature also describe another similar but even more rare anomaly called (TTH) Topsy-Turvy heart (see Fig. 2a), where there is an organo-axial rotation along the long axis of the heart (approx. 25-30 cases only documented up till now).(9) CCH and SIV/UDV have normal origin and course of great arteries but, in cases of TTH, due to their posterior inferior malorientation we see elongation of the neck veins and airways (see Fig.2b).(9,10) Another major distinguishing feature of TTH is the association of Aortopulmonary windows (APW), which are quite often seen in such cases.(10) Due to its rarity, there hasn't been any generalization about a possible diagnostic criteria and associations for this condition. (9,10)

In the present case, all 4 chambers could not be shown in the same plane and the axis of the atrioventricular valves were not parallel to eachother. The right atrium was inferior and along the diaphragm with a superiorly placed right ventricle. Plus the RV sat on top of LV with the apex pointing a bit horizontally. Thus, this heart had more than one pathology, i.e., twisting (CCH) and tiling (upstairs-downstairs, supero-inferior)

Magnetic resonance imaging is useful when echocardiogram cannot confirm the diagnosis. (11)

Management depends upon the associated anomalies. A small right ventricle and pulmonary stenosis may require a staged Fontan repair. In other cases with simple ventricular septal defect or patent ductus arteriosus alone may just require closure of the defects. In case of isolated transposition of great vessels, arterial switch operation may be the only requirement. (12,13)

Conclusion

The suspicion of a criss-cross heart should arise when it is not feasible to visualize all four cardiac chambers in the apical four-chamber view. The crossing of inlets becomes apparent during a posterior-to-anterior sweep with the probe. The criss-cross heart provides insight into the concepts of situs concordance and alignment. Although no definitive mechanism has been established for this condition, one proposed explanation by Robert H.Anderson et al involves the idea of post-septational rotational abnormality (see Fig. 3), which posits that additional rotation of the heart after looping determines the position of the apex.

Conflict of Interest

The authors declare that they have no competing interests.

Acknowledgement

None.

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