

Horizontal sandwiched interventricular septum: Complex cardiac anatomy deciphered with DSCT

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Abstract

The superior-inferior ventricle is a rare anomaly with the unknown incidence owing to less number of reported cases. However, one of the studies has reported this incidence to be <0.1% of all congenital heart diseases. This has a characteristic imaging appearance with horizontal interventricular septum. Most of the previous reports of superior-inferior ventricles have been described on echo, catheter angiography and only a few with cross-sectional imaging. We report two cases of superior-inferior ventricles associated with complex congenital cardiac defects on dual-source CT.

Key words: Congenital heart disease; horizontal interventricular septum; superoinferior ventricles

Introduction

The term superior-inferior ventricles (SIV) implies that the interventricular septum (IVS) is horizontal with the ventricles lying on above each other instead of side by side. It is also known as upstairs-downstairs ventricles and depending on the morphology, it can be either right ventricle (RV) on top or left ventricle (LV) on top. It is a rare anomaly and the exact incidence is unknown. However, TaksandeAM^[1] has reported the incidence to be <0.1% of all congenital heart diseases [CHDs]. Most of the cases reported in the literature are of RV on top morphology and Porras *et al.*^[2] has infact described the LV on top morphology for the first time in 2003. Since then there are quite a few cases of SIV which are reported in the literature with LV on top morphology. While most of the reported cases in the literature have projected SIV on echocardiography and catheter angiography,^[1-5] we present two cases of complex

congenital heart diseases having associated SIV (RV on top morphology), deciphered on dual-source CT (DSCT).

Case History

Case 1

First patient was an 8-year-old boy with cyanosis. CT scan showed situs solitus with levocardia. Both the ventricles were situated supero-inferiorly with horizontal IVS [Figure 1]. The ventricle on top was of RV morphology with both great vessels arising from it (double outlet RV) [Figure 2]. There was a sub-aortic ventricular septal defect (VSD). Pulmonary stenosis with confluent pulmonary arteries was noted. There was malpositioned great arteries with aorta anteriorly and left of the main pulmonary artery [Figure 3]. Coronaries were normal.

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Case 2

Second patient was a 15-year-old male with cyanosis. CT scan showed situs solitus with levocardia. Both atria were connected to antero-inferiorly placed ventricle which was

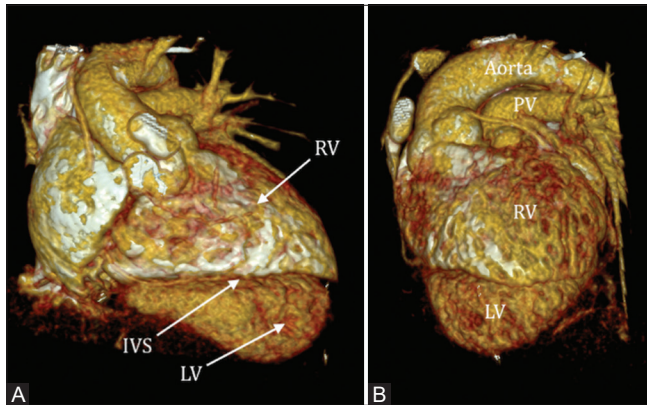


Figure 1 (A and B): Volume rendered CT images (A and B) depict the SIV morphology (RV on top type) with horizontal IVS. Malpositioned great arteries are also seen. SIV: Supero-inferior ventricles, RV: Right ventricle, IVS: Interventricular septum, LV: Left ventricle, PV: Pulmonary artery

of RV morphology. Morphological LV, which was placed postero-superiorly [Figure 4] had no atrial connection. There was complete transposition of great vessels with aorta arising from morphological RV and pulmonary artery from morphological LV. Sub-aortic VSD was present [Figure 5]. Pulmonary stenosis with confluent pulmonary arteries was noted [Figure 6]. The associated right-sided aortic arch was also seen.

Discussion

SIV is generally associated with other complex type of congenital heart diseases. However, Yang *et al.*^[4] described cases with SIV being the only abnormality. Morphogenesis of this anomaly is a debatable topic with most of the recent literature suggestive of post-septational defect. Angelini *et al.*^[3] suggested it to be a deformed mid-distal IVS and ventricles to a variable degree in addition to clockwise rotation of apical ventricular segments with normal atrioventricular valves and ventricular inlets, hence suggested the term “apical malrotation syndrome”. Angelini *et al.*^[3] also stated that criss-cross heart and SIV are

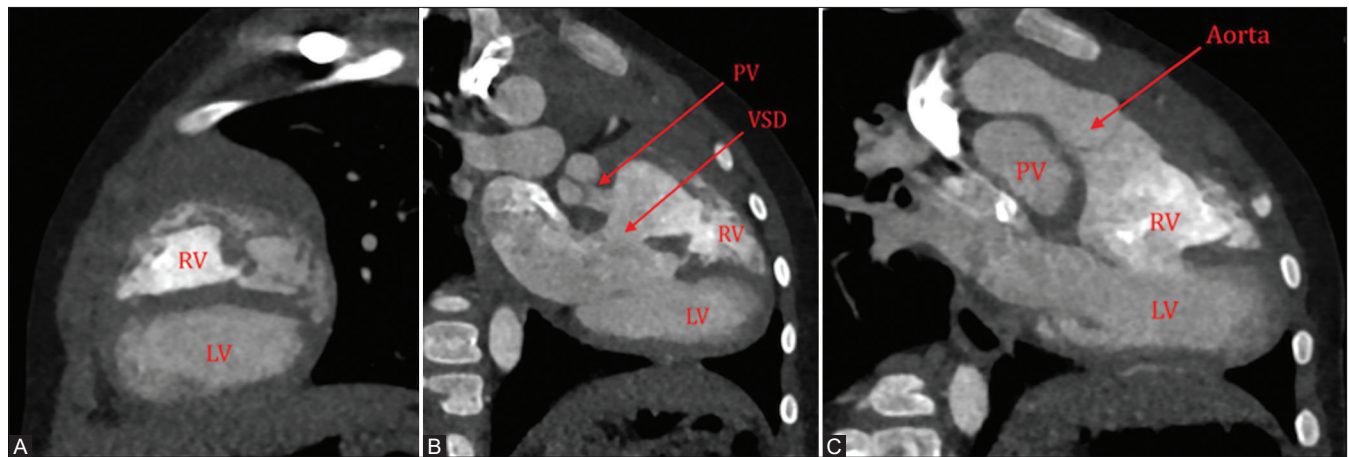


Figure 2 (A-C): CT short-axis (A) and oblique long axis (B and C) images show supero-inferior relationship of ventricles with large VSD, DORV and malpositioned great vessels. VSD: Ventricular septal defect, DORV: Double outlet right ventricle, RV: Right ventricle, LV: Left ventricle, PV: Pulmonary artery

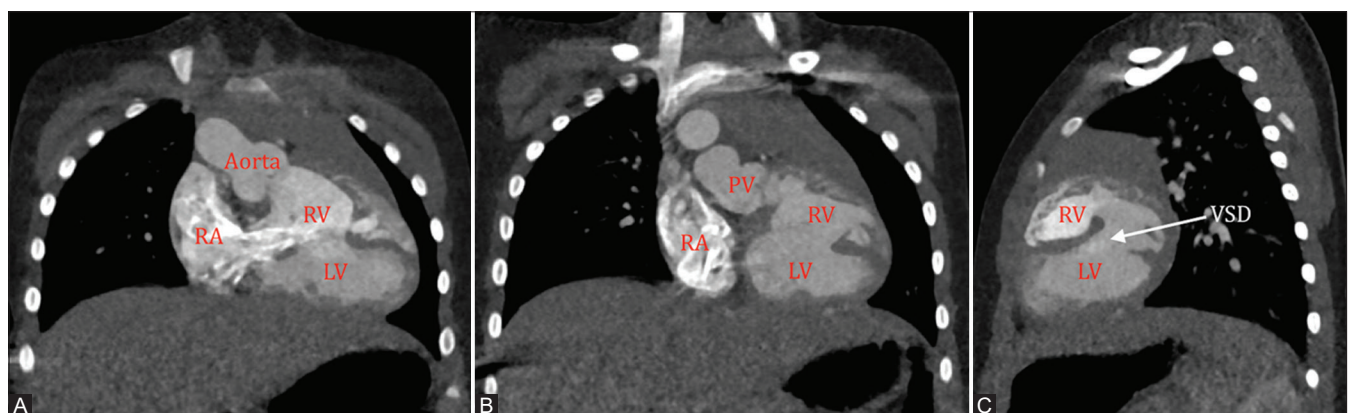


Figure 3 (A-C): True coronal (A and B) and sagittal images (C) show superior-inferior ventricles with VSD and DORV. RV: Right ventricle, LV: Left ventricle, RA: Right Atrium, PV: Pulmonary Artery, VSD-Ventricular septal defect, DORV - Double outlet right ventricle

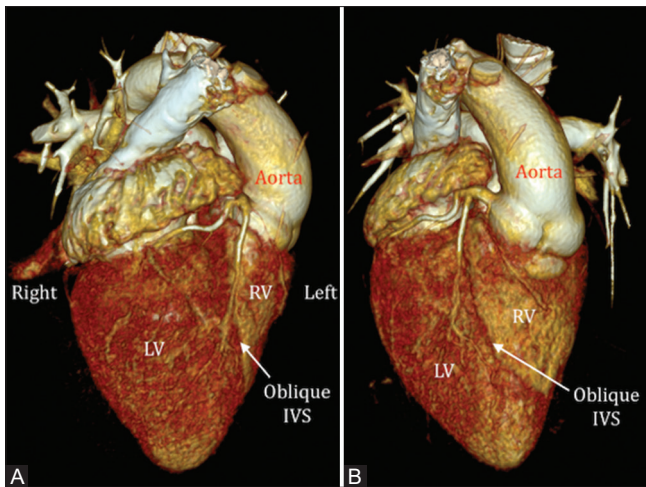


Figure 4 (A and B): Volume rendered CT images (A and B) show obliquely oriented IVS with posterior-superior RV and anteroinferior LV. Right-sided aortic arch is also seen. IVS: Interventricular septum, RV: Right ventricle, LV: Left ventricle

different structural anomalies that may co-exist depending on the clockwise or anti-clockwise rotation of the apical structures.

Earlier generation CTs often produced images of sub-optimal diagnostic quality. Newer generation CTs with improved temporal resolution produces excellent quality images so that all the associated defects can be depicted accurately. Better quality volume rendered CT images (VRT) can be produced, which depict this particular morphology accurately. In these cases, the images were acquired on DSCT (Siemens Somatom Force) with retrospective ECG gating and phase-wise reconstruction. VRT images were generated with a bone subtraction algorithm. No premedication or sedation was required in these patients. These images accurately depict the complex anatomy as shown in the images and VRT images nicely show the SIV morphology.

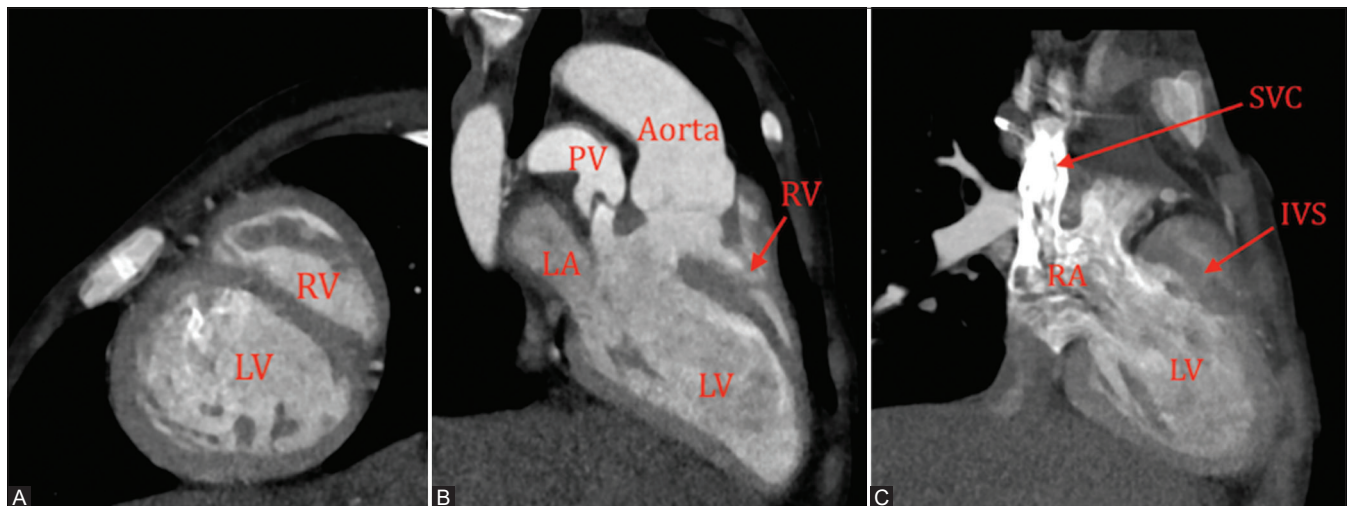


Figure 5 (A-C): CT two-chamber short-axis (A) and oblique long-axis images (B and C) show obliquely oriented IVS with SIV. Aorta is seen arising from hypoplastic RV and pulmonary artery from LV with large sub-aortic VSD. Both the atria are seen connected to morphological LV. IVS: Interventricular septum, SIV: Supero-inferior ventricles, RV: Right ventricle, PV: Pulmonary artery, LV: Left ventricle, VSD: Ventricular septal defect, RA: Right atrium, LA: Left atrium, SVC: Superior vena cava

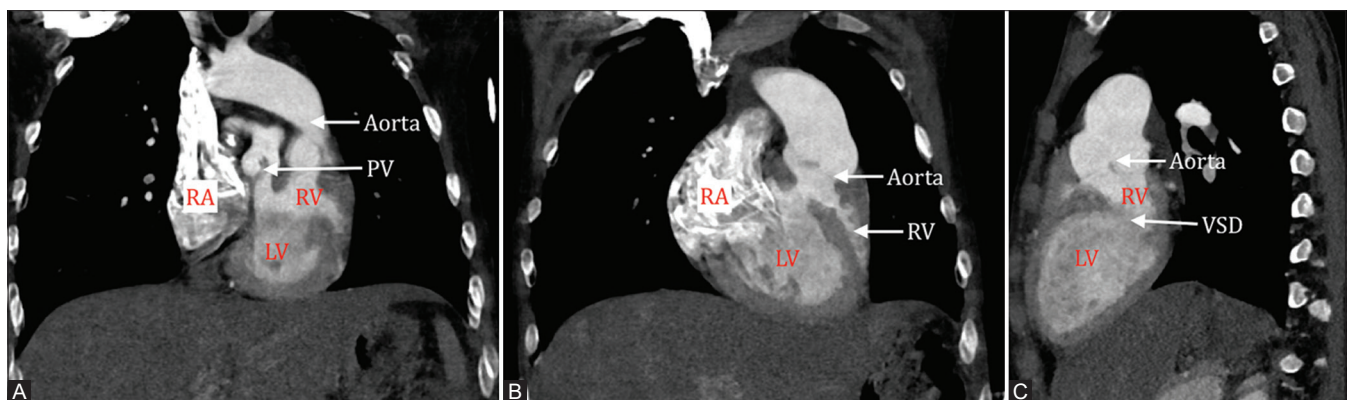


Figure 6 (A-C): True coronal (A and B) and sagittal (C) CT images with a wider view showing complex cardiac anatomy with SIV and orientation of the IVS. Note is also made of VSD with transposition of great arteries and pulmonary stenosis. SIV: Supero-inferior ventricles, IVS: Interventricular septum, VSD: Ventricular septal defect, RA: Right atrium, RV: Right ventricle, PV: Pulmonary artery, LV: Left ventricle

SIV is a rare form of CHD, which is generally associated with another complex type of cardiac defect. With the latest development of CT scanners, accurate depiction of cardiac anomalies can be demonstrated and the latest reconstruction algorithms enable us to generate good quality VRT images as seen in our cases.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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