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A Case Report of Situs Inversus Totalis with Ventricular Septal Defect: Flipped Physiology

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Abstract

Dextrocardia with situs inversus (situs inversus totalis) is a rare congenital condition characterized by the abnormal positioning of the heart and other organs in the thoracic and abdominal cavities. When combined with a ventricular septal defect (VSD), it presents unique challenges in diagnosis and management. Here, we present a case report of a patient with dextrocardia, situs inversus, and a peri-membranous VSD, highlighting the diagnostic process and discussing available treatment options.

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Introduction

Dextrocardia with situs inversus (also known as situs inversus totalis) is an uncommon congenital condition where the heart lies on the right half of the chest with its base to apex directed toward the right, and the abdominal organs are mirrored in their anatomical arrangement [1]. Among congenital heart defects in children, VSD stands as the most prevalent, ranking second only to the bicuspid aortic valve as the most prevalent congenital anomaly in adults. The combination of dextrocardia with situs inversus along with a VSD is even rarer [2]. Understanding the relative arrangements of the heart's various chambers is crucial to achieving a favourable result in surgery. This case report aims to provide insights into the diagnosis, management, and outcomes of a patient with this unique combination of cardiac anomalies.

Case Presentation

A 36-year-old female, weighing 57 kilograms, presented to us with recurring respiratory infections and exertional dyspnoea over the past two years, with an escalation in severity during her pregnancy and normal healthy baby delivery one year back. But now these symptoms gradually subsided by loop diuretics and Digoxin. At the time of during her first antenatal check-up she was diagnosed as having dextrocardia. Her growth, development, and developmental milestones throughout childhood were all normal, and she had no prior history of squatting, cyanosis, dyspnoea, syncope, or recurring lung infections. Upon examination, the patient was physically well-developed and showed no clubbing, cyanosis, venous distention, or oedema. With signs of mild dyspnoea, the respiration rate was 26 per minute, the pulse rate was 90 beats per minute, and the blood pressure was 136/76 mm Hg. Abdominal examination demonstrated situs inversus, with the liver dullness percussion on the left side and the stomach's tympanic note over the right hypochondrium. Cardiovascular system examination revealed cardiac dullness percussion with the apex beat palpable on the right half of the chest (right 5th intercostal space) in the mid-clavicular line, and on auscultation, heart sounds were more pronounced on the right half of the chest, and also revealed a loud, harsh holosystolic murmur best heard at the lower sternal border of the right side (right 2-4th intercostal spaces).

A chest X-ray revealed the heart on the right side, with the base-to-apex axis oriented in the right direction, flipped bronchial anatomy and the gastric bubble on the right side (figure 1).

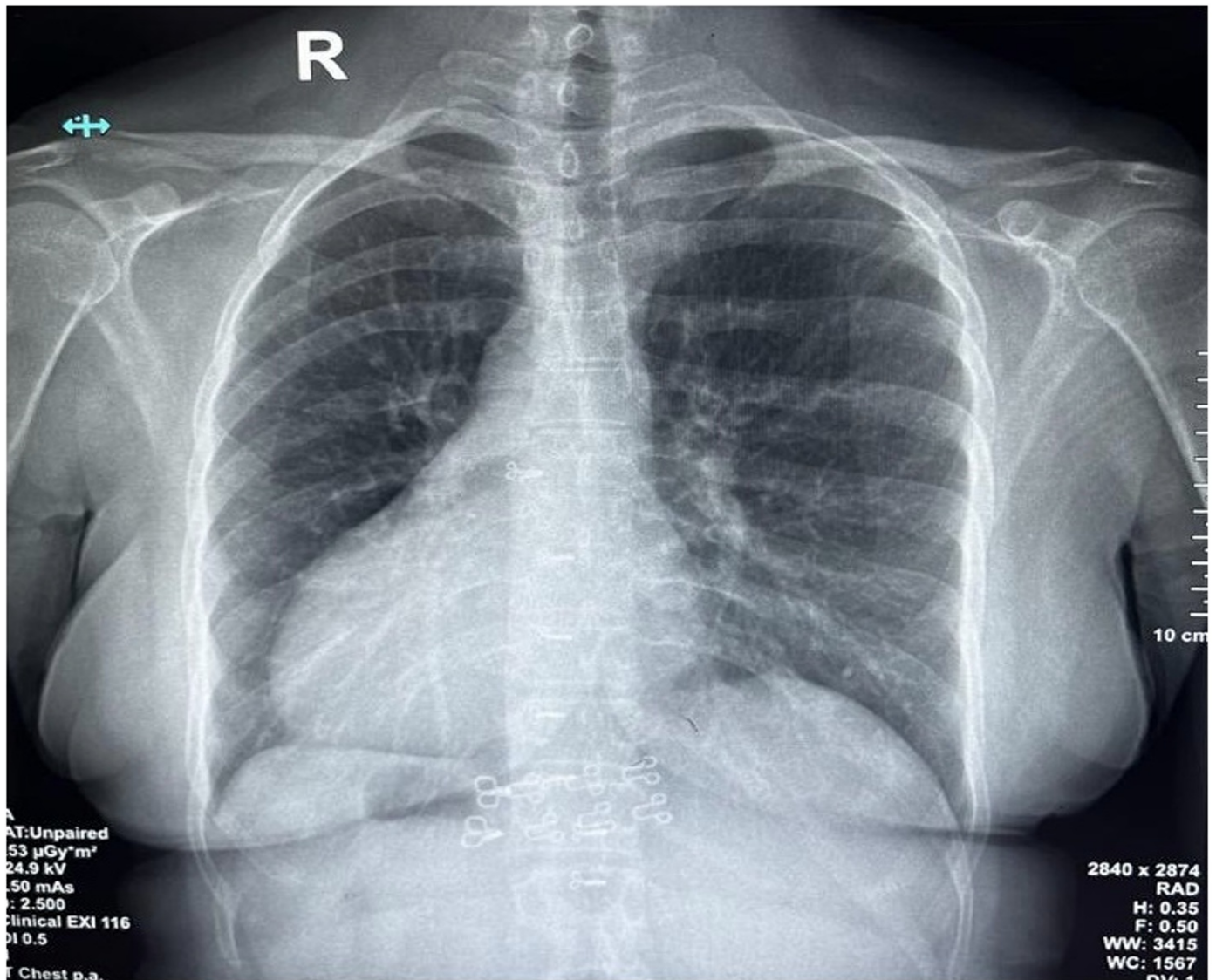


Figure 1. Chest radiograph showing the cardiac apex in the right hemithorax with the gastric bubble visible in the right upper quadrant.

A standard 12-lead electrocardiogram (ECG) demonstrated a normal sinus rhythm, a normal axis deviation, a negative QRS complex, and inverted P and T waves in lead I, with biphasic T waves and progressively decreasing R-wave amplitude from leads V_1 to V_6 (figure 2-a). Results of a repeat ECG obtained after right-sided repositioning of the precordial leads (V_1 – V_6) demonstrated a normal R-wave progression in the precordial leads (figure 2-b).

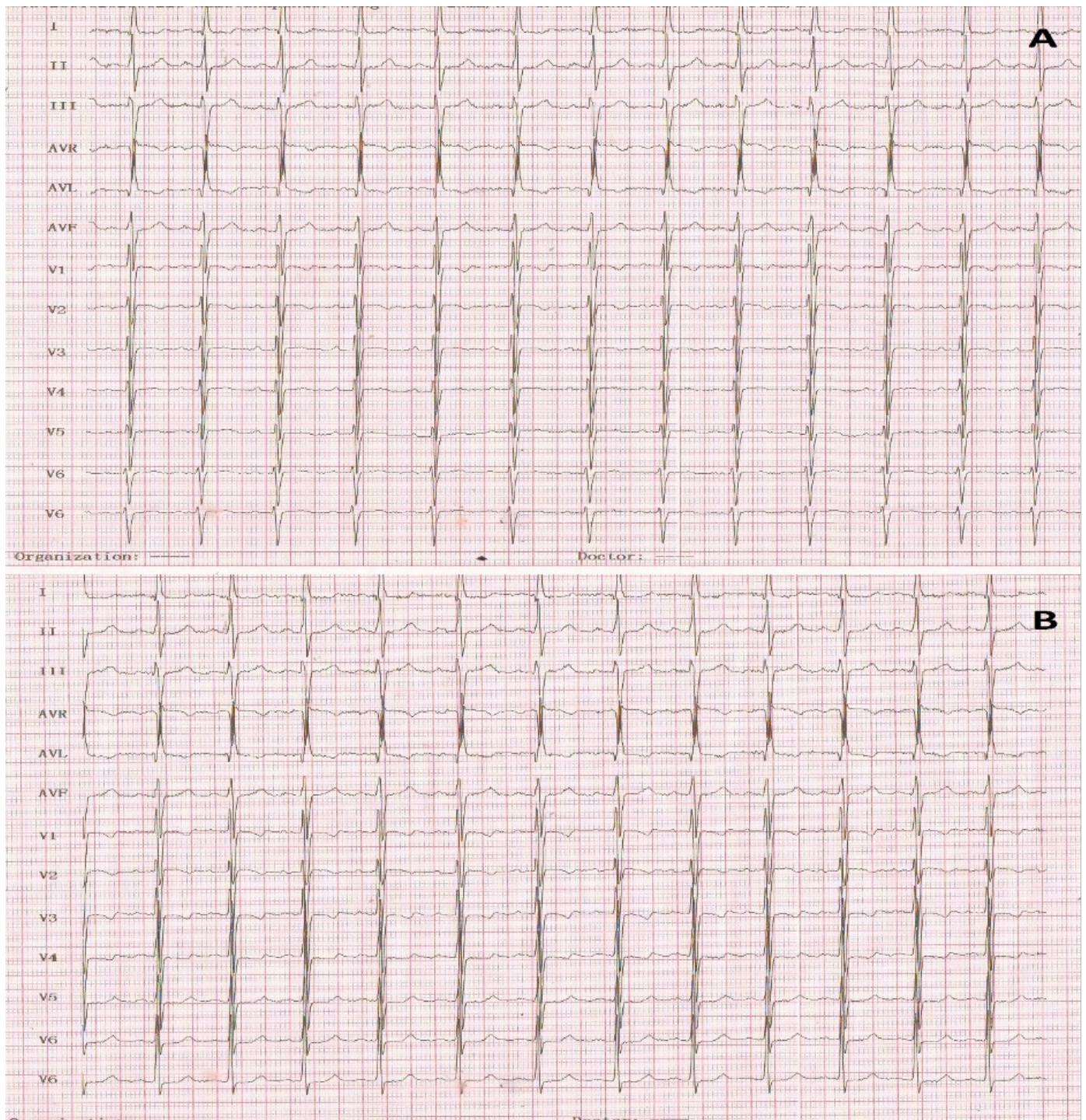


Figure 2. 12-lead ECG showing A: normal sinus rhythm, a normal axis deviation, a negative QRS complex, and inverted P and T waves in lead I, with biphasic T waves and progressively decreasing R-wave amplitude from leads V1 to V6, B: normal R-wave progression from V1 to V6 on repeat ECG obtained after right-sided repositioning of the precordial leads.

ECG and chest X-ray confirmed the dextrocardia and situs inversus. An ultrasound of the entire abdomen revealed a reverse positioning of the abdominal organs (situs inversus). Echocardiography demonstrated a small peri-membranous VSD (figure 3-a) with left-to-right shunting (figure 3-b), a gradient of 115 mm Hg, and normal biventricular function.

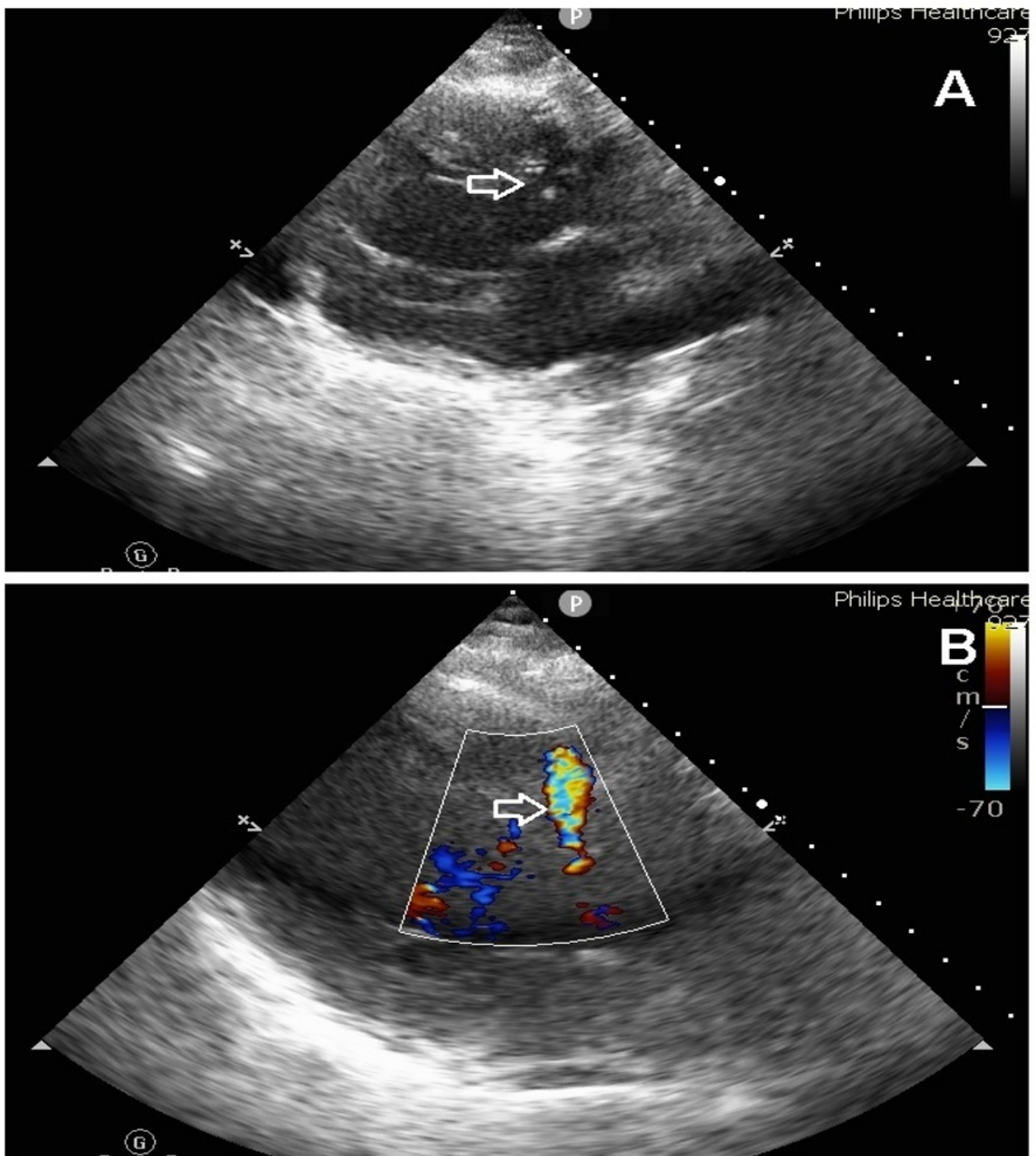


Figure 3. Trans thoracic 2D-echocardiography images A: small peri-membranous VSD, B: Left-to-right shunting and gradient of 115 mm Hg and normal biventricular function.

Cardiac magnetic resonance imaging (MRI) was also done, which revealed a small peri-membranous VSD, situs inversus with dextrocardia, right-sided aortic arch, and left ventricle ejection fraction (LVEF) of 45% (figure 4). This reduced EF was due to peripartum cardiomyopathy at that time and the coronary angiography (CAG) was done now, which showed it near normal.

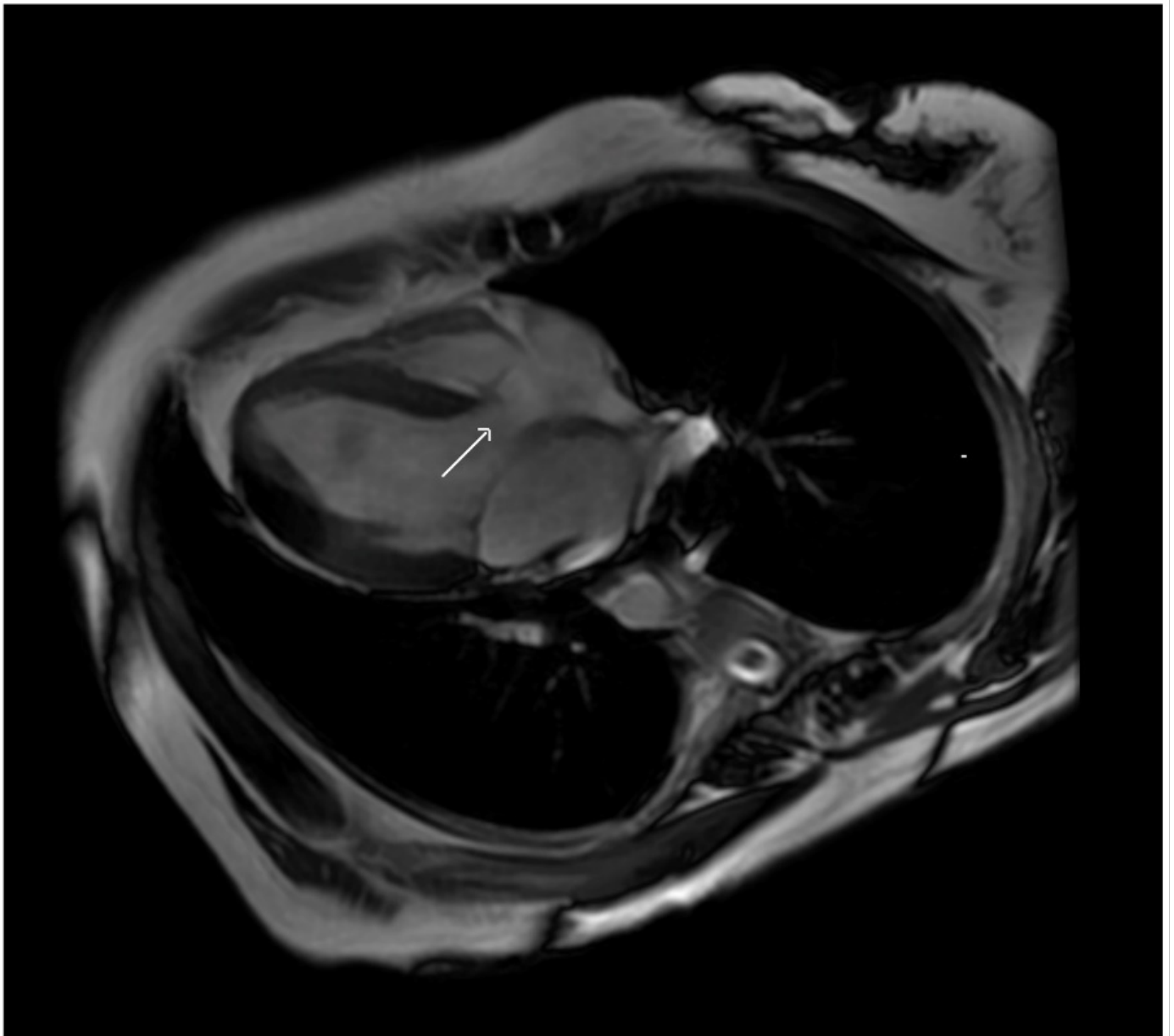


Figure 4. Cardiac MRI showing a small peri-membranous VSD, situs inversus with dextrocardia, with right-sided aortic arch.

A cardiac catheterization study was conducted via the right femoral vein to evaluate the hemodynamic importance of the VSD and strategize subsequent treatment. The catheter traversed the inferior vena cava (IVC), proceeded through the left-oriented right atrium (RA), the left-sided right ventricle (RV), and entered the pulmonary artery (PA). The PA was located to the right of the ascending aortic shadow. The results were aligned with the presence of a moderate-flow VSD accompanied by mild pulmonary hypertension and mirror-image dextrocardia. (Table 1 and figure 5). And pulmonary capillary wedge pressure (PCWP) is 12 mmhg, calculated cardiac output (CO) is 2.81 l/min, and ratio between pulmonary (Q_p) to systemic flow (Q_s) is 1.52.

Site	O2 Saturation (%)	Pressure (mm. Hg)
SVC	81.4	7/1 (6)
IVC	80.2	6/2 (6)
RA	83.6	7/2 (6)
RV	87.4	48/2 (14)
PA	86	35/13 (17)
LV	99.5	158/12 (82)
Aorta	99.3	170/97 (119)
RFA	98.6	158/88 (97)

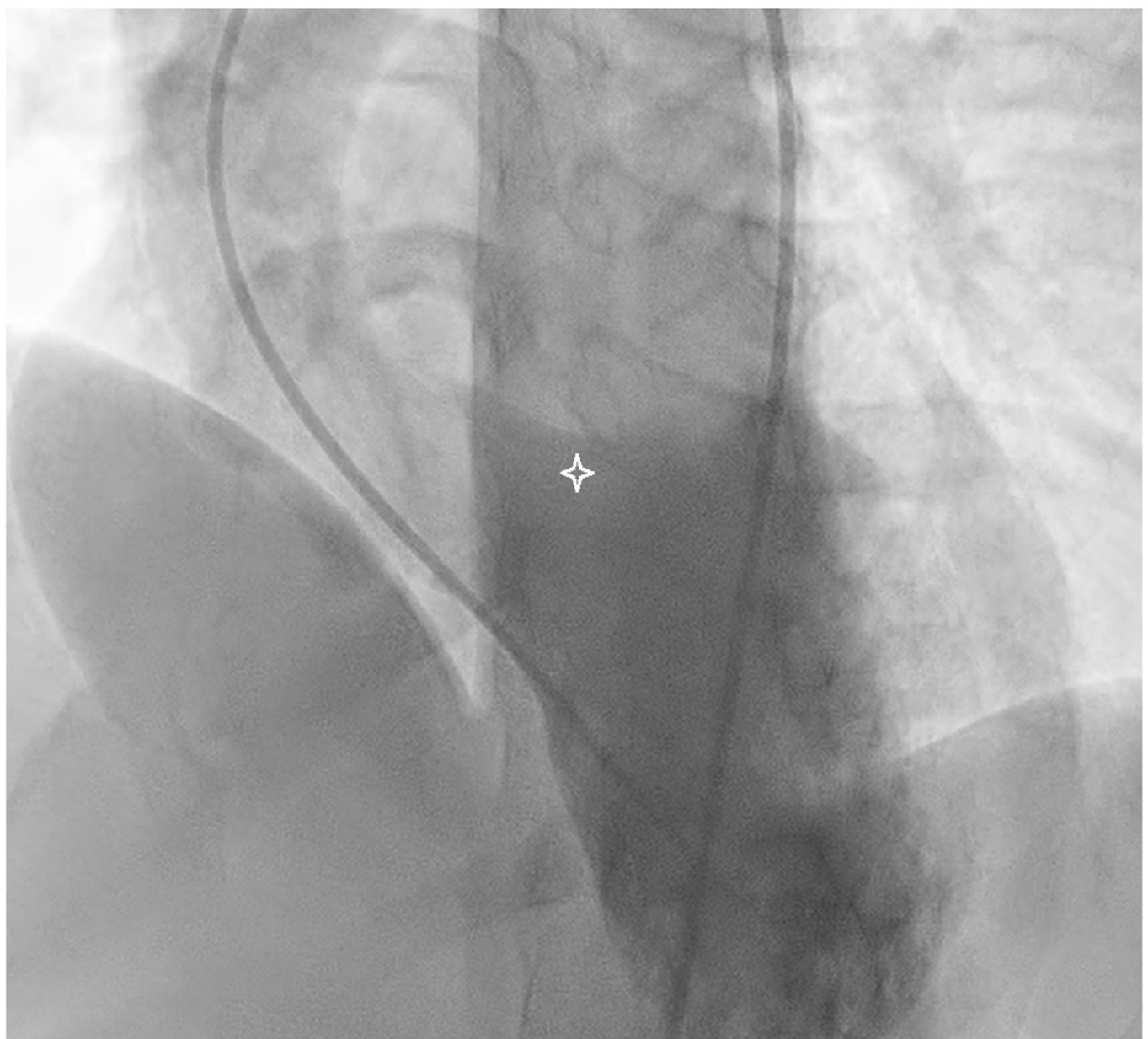


Figure 5. LV angiogram image of cardiac cath study showing a regurgitation of dye from LV to RV via VSD.

Considering the patient's clinical symptoms and diagnostic results, surgery was advised. However, the patient has chosen not to proceed with the operation at the moment for personal reasons. As a result, the patient is currently on angiotensin receptor/neprilysin inhibitor (ARNI), diuretics, beta blockers with Digoxin and being monitored through follow-up care for the last one year.

Discussion

Dextrocardia with situs inversus, also referred to as situs inversus totalis, is a rare finding, occurring in approximately 1 in 12,000 live births (ranging from 1/6000 to 1/35000 live births), and it affects both males and females equally [3][4].

This condition results from abnormal embryological development in which the heart tube and internal organs fail to migrate to their usual positions. Patients with dextrocardia may remain asymptomatic, while others, like the case presented, may experience symptoms related to associated cardiac anomalies. Dextrocardia, when combined with a normal abdominal position, often presents a high occurrence of linked congenital heart defects, such as transposition of the great vessels, ASD, and VSD, and is found in 90 to 95% of cases, among other conditions [5][6]. Conversely, dextrocardia paired with situs inversus is linked to a reduced occurrence of congenital heart disease (0 to 10%). While the precise cause of dextrocardia remains uncertain, several conditions have been associated with it, such as conjoined twinning, cocaine usage, maternal diabetes, and an autosomal recessive gene with inadequate penetrance [7].

The small peri-membranous VSD in this patient further complicates the clinical picture. VSDs are among the most common congenital heart defects, but the combination of dextrocardia, situs inversus, and this type of VSD is exceedingly rare. Management options must consider the anatomical and physiological variations of this unique presentation. Treatment strategies for VSDs vary based on the size of the defect, its hemodynamic significance, and the presence of other cardiac anomalies. In this case, the patient's small peri-membranous VSD was closely monitored for signs of progression and hemodynamic compromise. Surgical closure of the VSD was considered as a treatment option, and the potential benefits and risks were weighed in consultation with the patient.

Dextrocardia with situs inversus poses a surgical challenge, not primarily due to concurrent cardiac malformations, but rather because attaining sufficient exposure for surgery proves to be challenging. This is particularly evident when performing procedures on the left side of the heart, as normally RA, RV, and tricuspid valve (TV) are situated posteriorly in the right hemithorax, in contrast to dextrocardia with situs inversus. In all usual cardiac cases, the surgical team follows a standard arrangement, where the primary surgeon is positioned on the right side, and the first assistant, perfusionist, and cardiopulmonary bypass (CPB) machine are on the left side of the patient. However, in cases of dextrocardia with situs inversus, we may adopt the opposite configuration (Figure 6) for the operative procedure.

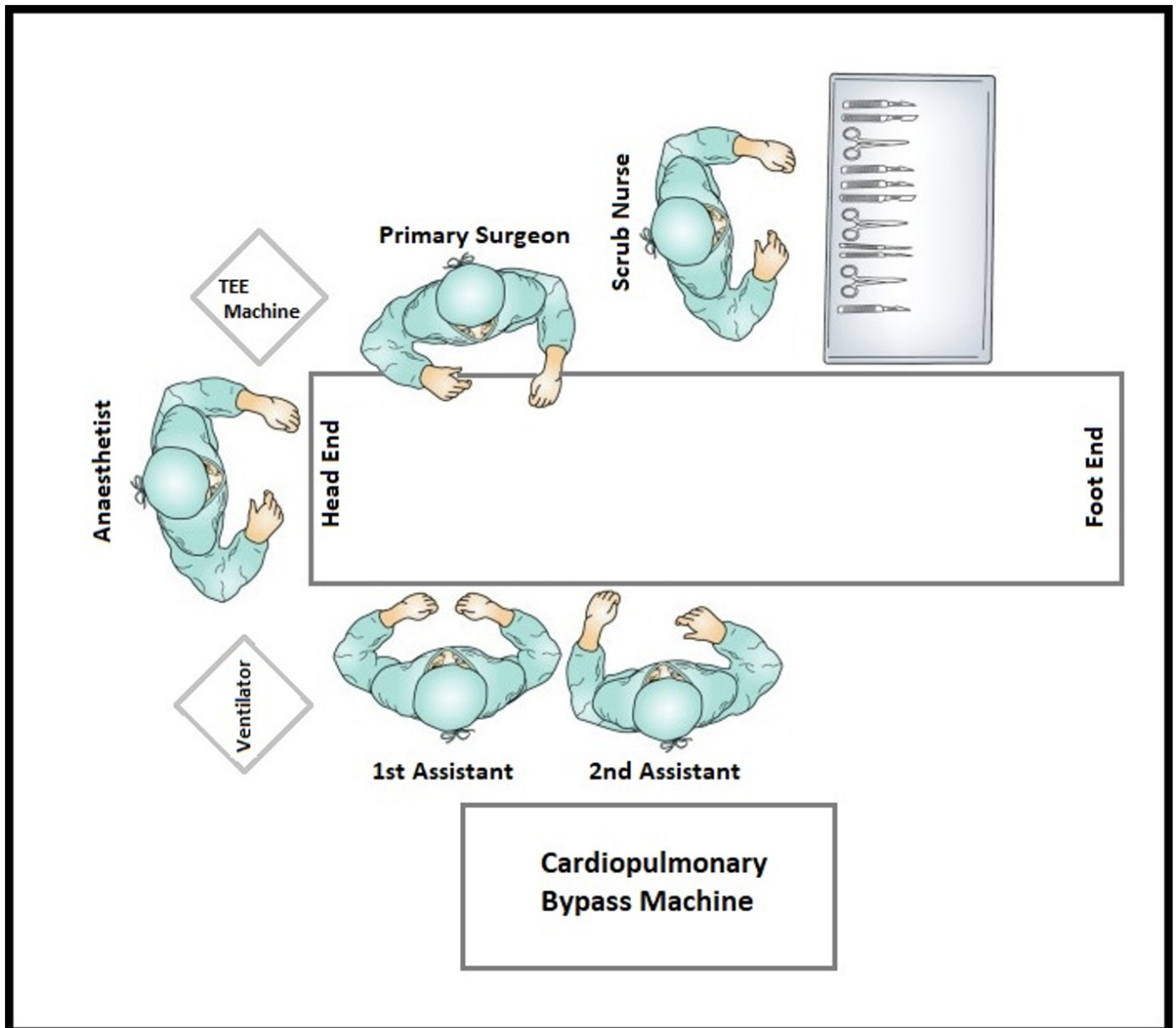


Figure 6. Schematic diagram of the operating table along with the surgical team in case of dextrocardia.

Proper consideration must be given to the placement of cannulas for CPB. In this particular morphology, the IVC is located in the midline. To address this, we should introduce the IVC cannula into the operative field from the midline, beneath the transverse bar of the retractor. This adjustment is documented to avoid potential interference with the retraction of the RA and RV in cases of dextrocardia [8].

In the course of the surgical correction, the VSD should be closed using a patch through an atriotomy made in the left-oriented right atrium. The changed spatial arrangement of cardiac structures can pose a challenge when closing the VSD with a patch. The surgeon must address the VSD from a different angle in a patient with dextrocardia compared to a patient with levocardia. Consequently, despite the conduction system being along the inferior margin of the VSD, there could be an increased risk of post-surgical heart block [9].

Percutaneous device closure of VSD is designated for patients who face high surgical risks due to factors such as severe

pulmonary arterial hypertension, multiple comorbidities, or a history of previous cardiothoracic surgeries, including instances with residual or recurrent VSD. This procedure is especially appropriate for muscular VSDs, but it can be difficult to apply in cases where defects are located near the inlet valves [10].

Such VSDs that present in adulthood are typically restrictive or resemble a pseudo-Gerbode defect. They carry a risk of infective endocarditis. For individuals with an unrepaired VSD, approximately 87% will survive 25 years post-diagnosis. The size of the VSD significantly impacts survival chances [11].

Conclusion

This case report highlights the rarity and complexity of dextrocardia with situs inversus and a small peri-membranous VSD, serving as a valuable addition to the medical literature and encouraging further discussion and investigation of such unique congenital cardiac presentations. Further research and long-term follow-up of such cases can contribute to our understanding of this rare combination of congenital anomalies and guide clinical practice.

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