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COMPLEX SYSTEMIC AND PULMONARY VENOUS ANOMALIES IN A CHILD WITH SITUS AMBIGUUS, INTERRUPTED INFERIOR VENA CAVA, AND ATRIOVENTRICULAR SEPTAL DEFECT WITH PRIOR PULMONARY ARTERY BANDING: A CASE REPORT

Amr Adel Shaheen[1]*, Walid Abdullah Abdelrazak[1], Ashraf Abdelhamed El midany[2], Samia I. Sharaf[2]
[1]Department of Cardiac Surgery, Egypt Children's Hospital, Sekat Hadid Al Mahger, Zeinhom, El Sayeda Zeinab, Cairo Governorate 4260102, Egypt
[2]Department of Congenital Cardiac Surgery, Ain Shams University, El-Khalyfa El-Mamoun Street in the Abbassia district of Cairo, Egypt 11517, Egypt
*amradelkamal953@gmail.com

ABSTRACT
Congenital heart disease associated with situs ambiguus is frequently characterized by complex and heterogeneous abnormalities of systemic and pulmonary venous return. These anomalies are associated with atrioventricular septal defects and interrupted inferior vena cava, particularly in the setting of heterotaxy syndromes. We report a 2-year-old male child (10.5 kg) with complex congenital heart disease and inconsistent findings across multiple echocardiographic studies. Initial assessments suggested atrioventricular septal defect with variable ventricular balance and systemic venous anomalies. Cardiac CT demonstrated situs ambiguus, levocardia, common atrioventricular canal features, anomalous hepatic venous drainage into the left atrium, and partial anomalous pulmonary venous drainage, and absent of left SVC. Intraoperatively, more complex anatomy was identified, including two large atrial septal defects or common atrium without ventricular septal defect, cleft mitral valve, left superior vena cava draining into the left atrium, interrupted inferior vena cava with hepatic venous drainage into the left atrium, and partial anomalous pulmonary venous return to the right atrium. Surgical repair included atrioventricular valve repair, baffling of left superior vena cava and hepatic venous drainage using bovine pericardium, closure of atrial septal defects. Tricuspid and mitral valve repair and finally removal of pulmonary artery band and augmentation of main pulmonary artery with bovine pericardium. The postoperative course was complicated by arrhythmias requiring temporary pacing, low cardiac output syndrome, and need for peritoneal dialysis, followed by full recovery and discharge after 10 days to home. This case highlights rare combination of systemic and pulmonary venous anomalies associated with atrioventricular septal defects and interrupted inferior vena cava which illustrate the limitations of echocardiography and CT in complex systemic and pulmonary venous anomalies with heterotaxy syndromes and underscores the importance of intraoperative assessment.

Keywords: anomalous pulmonary venous return; atrioventricular septal defect; congenital heart disease; heterotaxysyndrome; interrupted inferior vena cava; systemic venous anomaly

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INTRODUCTION

Congenital heart disease in the setting of situs ambiguus is commonly associated with highly complex and heterogeneous abnormalities of systemic and pulmonary venous return (Jacobs et al., 2007; Kim, 2011). These anatomical variations are often difficult to delineate preoperatively due to their variability and the frequent overlap of intracardiac and extracardiac structures (Applegate et al., 1999; Uemura et al., 1995). In the setting of heterotaxy syndromes, atrioventricular septal defects frequently coexist with interrupted inferior vena cava, anomalous systemic venous connections, and abnormalities of pulmonary venous return (Dillman et al., 2009; Kim, 2011; Redington et al., 1990).

Despite significant advances in echocardiography and computed tomography, precise preoperative anatomical characterization remains challenging in a subset of patients. Discrepancies between

imaging findings and intraoperative anatomy are well recognized, underscoring the importance of multimodality imaging combined with surgical correlation in complex congenital heart disease (Applegate et al., 1999; Dillman et al., 2009; Kogon et al., 2010).

METHOD

This study is a descriptive clinical case report documenting the diagnostic evaluation, intraoperative decision-making, and surgical management of a complex congenital cardiac anomaly in a single pediatric patient. Preoperative non-invasive anatomical delineation was performed using serial transthoracic echocardiography (TTE) and multislice cardiac computed tomography (CT) angiography. Surgical repair was executed via median sternotomy under cardiopulmonary bypass (CPB) with aortic cross-clamping and moderate hypothermia. Written informed consent was obtained from the patient's legal guardians for the surgical intervention and the publication of this report and accompanying imaging. All clinical and surgical interventions complied with the ethical standards of the institutional review committee and the Declaration of Helsinki.

RESULT

Case Presentation

A 2-year-old male child weighing 10.5 kg was referred for evaluation of complex congenital heart disease after Palliation with pulmonary artery banding 16 month ago.

Echocardiographic Findings

Multiple echocardiographic studies showed variable findings including situs ambiguus, interrupted inferior vena cava, variable ventricular balance, common atrium, AV valve regurgitation, suspected pulmonary venous anomalies, and pulmonary artery band in situ.

CT Findings

Cardiac CT revealed situs ambiguus with levocardia, common AV canal features, multiple atrial septal defects, small ventricular septal defects, hepatic veins draining into the left atrium, partial anomalous pulmonary venous drainage, and a single right superior vena cava with no left superior vena cava.

Neurological History

Brain MRI demonstrated an almost stationary encephalomalacia involving the right fronto-parieto-temporal region in the right MCA territory, with established Wallerian degeneration.

Intraoperative Findings

Two large atrial septal defects or common atrium as described
No ventricular septal defect
Cleft mitral valve
Left superior vena cava draining into left atrium
Interrupted IVC with hepatic venous drainage into left atrium
Total anomalous pulmonary veins drainge

Surgical Procedure

Mitral valve repair with closure of cleft mitral
Tricuspid valve repair
Baffling of left superior vena cava with separate bovine pericardium patch
Baffling of hepatic vein with another bovine pericardium patch
Closure of atrial septal defects using generous bovine pericardial patch
De banding and augmentation of main pulmonary artery with bovine pericardial patch
CPB time: 120 minutes
Cross-clamp time: 90 minutes

Postoperative Course

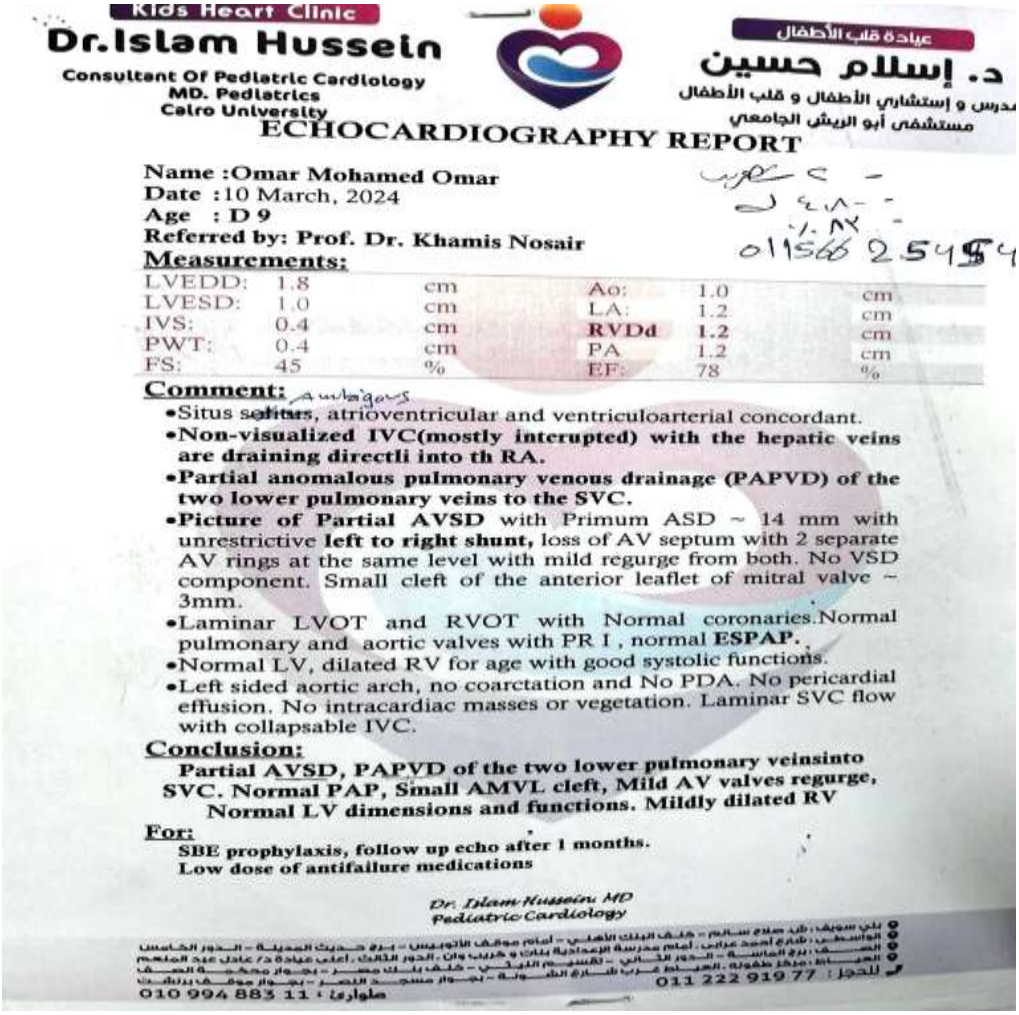
The postoperative coursewas complicated by arrhythmias requiring temporary pacing for 4 days. The patient developed low cardiac output syndrome requiring inotropic support (adrenaline, noradrenaline, and milrinone).

Peritoneal dialysis was initiated with subsequent clinical improvement.

The patient was extubated after 48 hours. Chest drains were removed without complications. The patient was transferred from CCU after 72 hours and discharged after 10 days with complete recovery.

Follow-up

Echocardiography before discharge showed good biventricular function, secure ASD closure, competent mitral valve, trivial tricuspid regurgitation, no pericardial effusion, and hypertrophied left ventricle.



Kids Heart Clinic
Dr.Islam Hussein
Consultant Of Pediatric Cardiology
MD. Pediatrics
Cairo University

عيادة قلب الأطفال
د. إسلام حسين
مدرس و إستشاري الأطفال و قلب الأطفال
مستشفى أبو الريش الجامعي

ECHOCARDIOGRAPHY REPORT

Name :Omar Mohamed Omar
Date :10 March, 2024
Age : D 9
Referred by: Prof. Dr. Khamis Nosair

Measurements:

LVEDD:	1.8	cm	Ao:	1.0	cm
LVESD:	1.0	cm	LA:	1.2	cm
IVS:	0.4	cm	RVDd	1.2	cm
PWT:	0.4	cm	PA	1.2	cm
FS:	45	%	EF:	78	%

Comment:
- Situs solitus, atrioventricular and ventriculoarterial concordant.
- **Non-visualized IVC(mostly interupted) with the hepatic veins are draining directli into th RA.**
- **Partial anomalous pulmonary venous drainage (PAPVD) of the two lower pulmonary veins to the SVC.**
- **Picture of Partial AVSD** with Primum ASD ~ 14 mm with unrestrictive **left to right shunt,** loss of AV septum with 2 separate AV rings at the same level with mild regurge from both. No VSD component. Small cleft of the anterior leaflet of mitral valve ~ 3mm.
- Laminar LVOT and RVOT with Normal coronaries.Normal pulmonary and aortic valves with PR I , normal **ESPAP.**
- Normal LV, dilated RV for age with good systolic functions.
- Left sided aortic arch, no coarctation and No PDA. No pericardial effusion. No intracardiac masses or vegetation. Laminar SVC flow with collapsable IVC.

Conclusion:
Partial AVSD, PAPVD of the two lower pulmonary veinsinto SVC. Normal PAP, Small AMVL cleft, Mild AV valves regurge, Normal LV dimensions and functions. Mildly dilated RV

For:
SBE prophylaxis, follow up echo after 1 months.
Low dose of antifailure medications

Dr. Islam Hussein MD
Pediatric Cardiology

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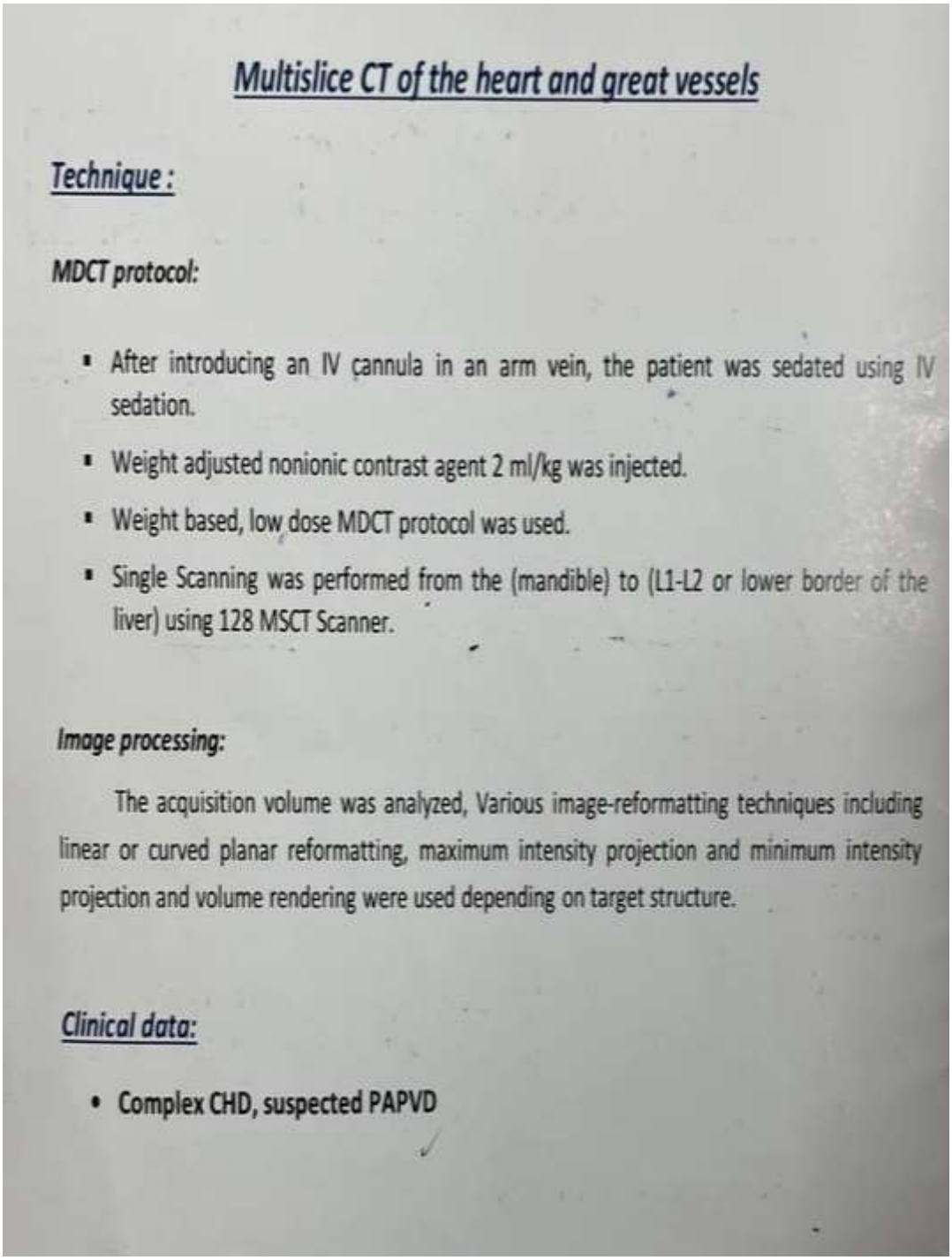
011 222 919 77 : للحجز

Figure 1. Echo Summary of the final diagnosis showing complex congenital heart disease (CHD) characterized by situs ambiguus with levocardia, complete atrioventricular canal (CAVC) with large primum ASD and moderate secundum ASD, multiple ventricular septal defects (VSDs), anomalous systemic and pulmonary venous drainage, balanced ventricles, and associated aortic arch narrowing with single right superior vena cava (SVC).



Multislice CT of the heart and great vessels

Technique :

MDCT protocol:

- After introducing an IV cannula in an arm vein, the patient was sedated using IV sedation.
- Weight adjusted nonionic contrast agent 2 ml/kg was injected.
- Weight based, low dose MDCT protocol was used.
- Single Scanning was performed from the (mandible) to (L1-L2 or lower border of the liver) using 128 MSCT Scanner.

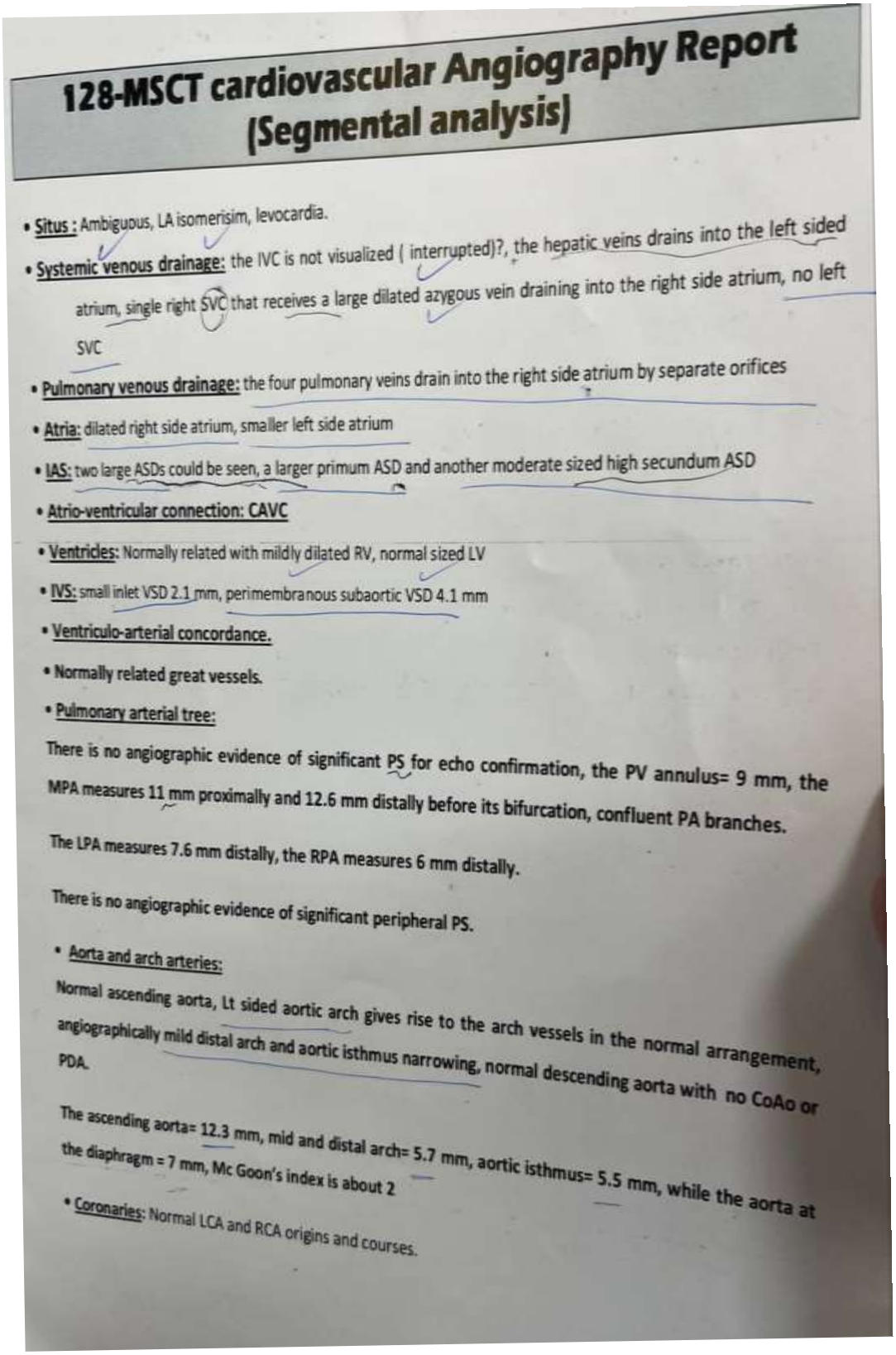
Image processing:

The acquisition volume was analyzed, Various image-reformatting techniques including linear or curved planar reformatting, maximum intensity projection and minimum intensity projection and volume rendering were used depending on target structure.

Clinical data:

- **Complex CHD, suspected PAPVD**

Figure 2. Computed tomography (CT) of the chest showing finding of CT



128-MSCT cardiovascular Angiography Report (Segmental analysis)

- **Situs :** Ambiguous, LA isomerisim, levocardia.
- **Systemic venous drainage:** the IVC is not visualized (interrupted)?, the hepatic veins drains into the left sided atrium, single right SVC that receives a large dilated azygous vein draining into the right side atrium, no left SVC
- **Pulmonary venous drainage:** the four pulmonary veins drain into the right side atrium by separate orifices
- **Atria:** dilated right side atrium, smaller left side atrium
- **IAS:** two large ASDs could be seen, a larger primum ASD and another moderate sized high secundum ASD
- **Atrio-ventricular connection: CAVC**
- **Ventricles:** Normally related with mildly dilated RV, normal sized LV
- **IVS:** small inlet VSD 2.1 mm, perimembranous subaortic VSD 4.1 mm
- **Ventriculo-arterial concordance.**
- **Normally related great vessels.**
- **Pulmonary arterial tree:**

There is no angiographic evidence of significant PS for echo confirmation, the PV annulus= 9 mm, the MPA measures 11 mm proximally and 12.6 mm distally before its bifurcation, confluent PA branches.

The LPA measures 7.6 mm distally, the RPA measures 6 mm distally.

There is no angiographic evidence of significant peripheral PS.

- **Aorta and arch arteries:**

Normal ascending aorta, Lt sided aortic arch gives rise to the arch vessels in the normal arrangement, angiographically mild distal arch and aortic isthmus narrowing, normal descending aorta with no CoAo or PDA.

The ascending aorta= 12.3 mm, mid and distal arch= 5.7 mm, aortic isthmus= 5.5 mm, while the aorta at the diaphragm = 7 mm, Mc Goon's index is about 2

- **Coronaries:** Normal LCA and RCA origins and courses.

Figure 3. Computed tomography (CT) of the chest showing finding of CT

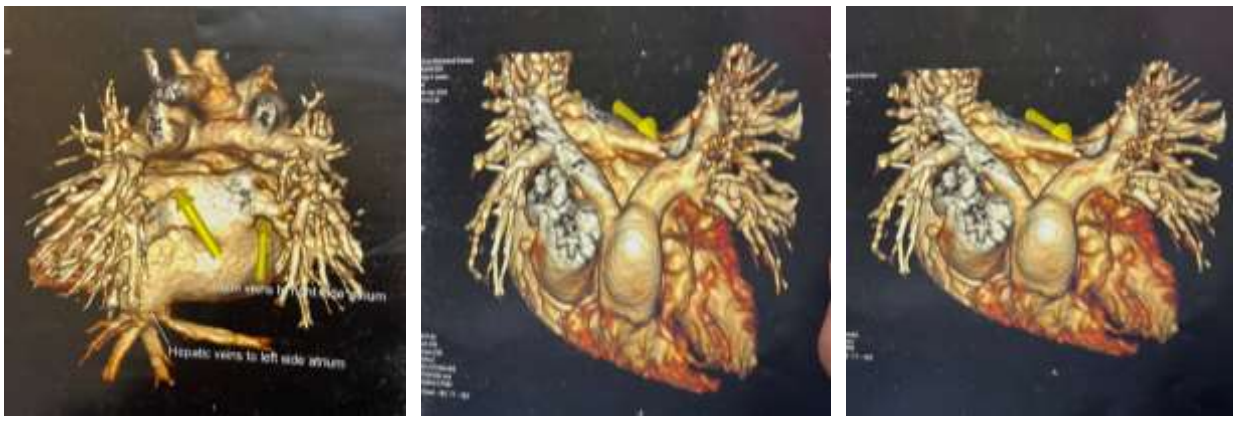


Figure 4. Computed tomography (CT) of the chest showing finding of CT

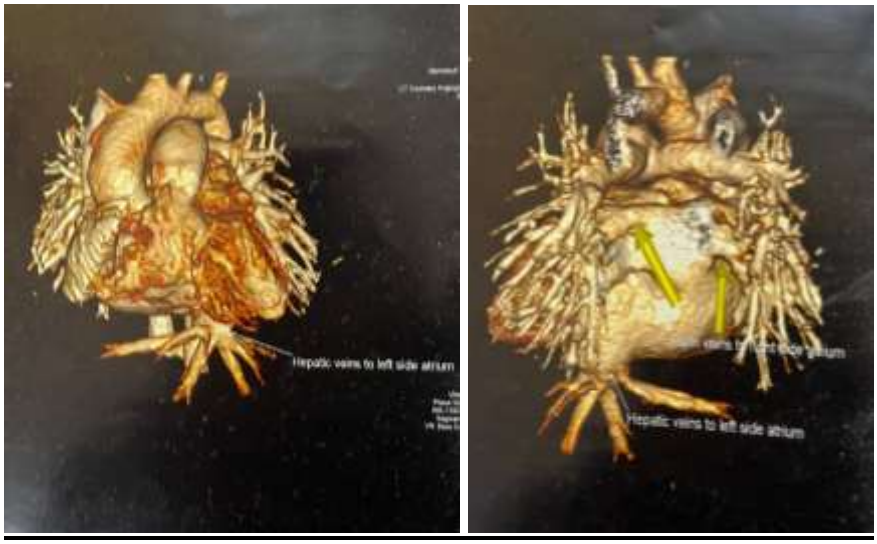


Figure 5. Computed tomography (CT) of the chest showing finding of CT

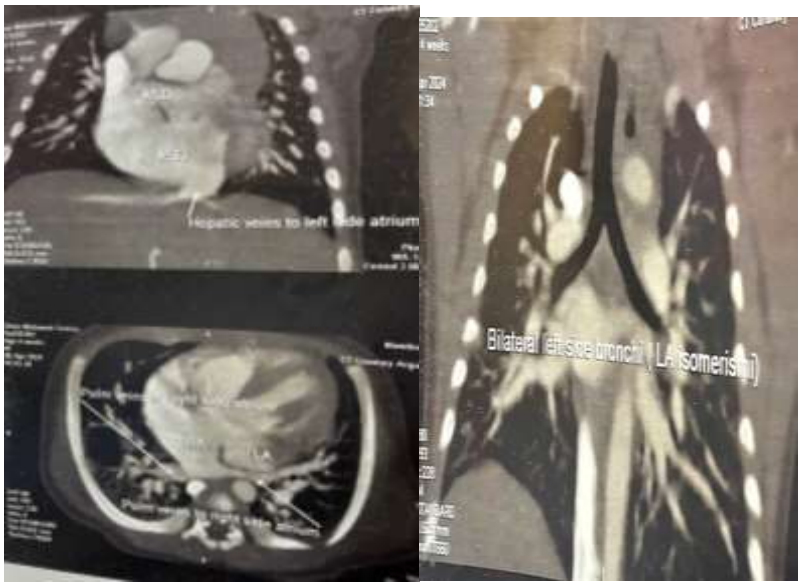


Figure 6. Conclusion of MSCT showing situs ambiguus with levocardia, common AV canal features, multiple atrial septal defects, small ventricular septal defects, hepatic veins draining into the left atrium, partial anomalous pulmonary venous drainage, and a single right superior vena cava with no left superior vena cava .

DISCUSSION

The coexistence of interrupted inferior vena cava, anomalous hepatic venous drainage to the left atrium, left superior vena cava to the left atrium, and partial or total anomalous pulmonary venous return represents an extremely rare and surgically challenging constellation (Dillman et al., 2009; Kim, 2011; Uemura et al., 1995). Systemic and pulmonary venous anomalies are well-recognized components of heterotaxy syndrome and are frequently associated with atrioventricular septal defects (Jacobs et al., 2007; Moses et al., 2021; Redington et al., 1990). This case illustrates the limitations of current imaging modalities and reinforces the importance of intraoperative assessment in complex congenital cardiac surgery (Applegate et al., 1999; Kogon et al., 2010). Significant discrepancies between echocardiography, CT imaging, and intraoperative findings have been previously reported, particularly in patients with complex venous anatomy (Applegate et al., 1999; Dillman et al., 2009). While CT improves anatomical understanding, certain anomalies such as anomalous systemic venous connections may still be missed (Applegate et al., 1999). From a surgical perspective, tailored approaches including atrial baffling techniques are often required to redirect anomalous venous return and achieve physiological circulation (Kogon et al., 2010; Perens et al., 2025). Outcomes in patients with heterotaxy syndrome remain variable and depend on anatomical complexity and surgical strategy (Alsoufi et al., 2007; "Early and Long-Term Outcomes," 2024; "The Complex and Hazardous Course," 2025).

CONCLUSION

Congenital heart disease in situs ambiguus is oftenassociated with complex systemic and pulmonary venous anomalies, frequently coexisting with atrioventricular septal defects and interrupted inferior vena cava. From a surgical perspective, tailored approaches including atrial baffling techniques are often required to redirect anomalous venous return and achieve physiological circulation. Despite advances in imaging, accurate preoperative delineation remains challenging, and discrepancies with intraoperative findings may occur, highlighting the importance of combined imaging and surgical assessment.

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