Case Report

# Mixed Inferior Sinus Venosus and Secundum Atrial Septal Defects with Mixed Partial Anomalous Pulmonary and Systemic Venous Drainage: Key Insights and Practical Implications

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Received 20 May 2024; Accepted 20 July 2024

## Abstract

*Mixed atrial septal defects (ASDs) involving inferior vena cava (IVC)-type sinus venosus and secundum types and mixed partial anomalous pulmonary, systemic, and hepatic venous drainage are rare. We describe a 3-year-old acyanotic boy who presented with a large mixed inferior sinus venosus and secundum-type ASD. He exhibited an abnormal connection between the right upper pulmonary vein and the right atrium. Additionally, the IVC and a hepatic vein drained abnormally into the left atrium. The patient also had valvular and supravalvular pulmonary stenosis, as well as a small patent ductus arteriosus.*

*The ASD was surgically closed using a pericardial patch, positioned lower than usual to reroute the IVC and hepatic vein flow into the right atrium. The surgery was successful, with no residual lesions or complications. The patient recovered without issues and was discharged smoothly. At the 6-month follow-up, the child's cardiac examination and oxygen saturation were normal. Furthermore, echocardiography confirmed normal drainage of the systemic and hepatic veins into the right atrium.*

*J Teh Univ Heart Ctr 2024;19(4):283-288*

***This paper should be cited as:*** Malakan Rad E, Radmehr H, Taghizadeh A, Pouraliakbar H, Radmehr E. Mixed Inferior Sinus Venosus and Secun-dum Atrial Septal Defects with Mixed Partial Anomalous Pulmonary and Systemic Venous Drainage: Key Insights and Practical Implications. J Teh Univ Heart Ctr 2024;19(4):283-288.

***Keywords:*** *Atrial septal defect; Pulmonary veins; Inferior vena cava; Hepatic veins; Congenital heart disease*

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## Introduction

The inferior vena cava (IVC) type sinus venosus atrial septal defect (ASD) is uncommon. The occurrence of mixed ASDs, which combine the IVC-type sinus venosus and secundum types, is even more unusual and has yet to be documented.[1] Furthermore, reports of mixed partial anomalous pulmonary, systemic, and hepatic venous drainage are exceedingly rare.[2] The co-occurrence of these conditions with valvular and supravalvular pulmonary stenosis and a patent ductus arteriosus (PDA) has not been previously reported.

We present the case of a 3-year-old boy diagnosed with a significantly large mixed inferior sinus venosus and secundum-type ASD. He exhibited an abnormal connection between the right upper pulmonary vein and the right atrium, along with anomalous drainage of the IVC and a hepatic vein into the left atrium (LA). Additionally, he had valvular and supravalvular pulmonary stenosis, as well as a small PDA.

This report discusses the complex interactions among these concurrent cardiac abnormalities, their impact on clinical presentation, and practical considerations for diagnosis and treatment.

## Case Report

A 3-year-old boy was referred to our center for surgical closure of a large ASD and repair of pulmonary stenosis. Upon gross examination, he appeared acyanotic and was in good clinical condition. Physical examination revealed normal vital signs. Pulses were normal, a right ventricular impulse was detected at the left lower sternal border, and a thrill was palpated in the suprasternal notch. Auscultation revealed a wide and fixed splitting of S2 and a grade 4/6 ejection systolic murmur at the left sternal border. Additionally, a grade 2/6 diastolic murmur was heard in the tricuspid area, and a grade 3/6 continuous murmur was noted at the upper left sternal border. His oxygen saturation, measured by pulse oximetry, was 93%. ECG showed right axis deviation with right ventricular hypertrophy, while the chest X-ray indicated an enlarged right atrium in the posteroanterior view. Echocardiography demonstrated that the atrial situs was solitus, the ventricles were d-loop, and the great arteries were normally related. Both the right atrium and ventricle were significantly enlarged. A sweeping examination from anterior to posterior in the 4-chamber view revealed a large mixed ASD of inferior sinus venosus and secundum types, with its size varying significantly during the anteroposterior sweep (Figure 1, Videos 1–3). The IVC and one of the hepatic veins drained into the LA (Figures 2 and 3). There was no evidence of a superior vena cava-type sinus venosus atrial septal defect (Figure 4, Video 4). The connection and drainage of the left pulmonary veins were normal (Videos 5–8). The right lower pulmonary vein was observed draining into the LA (Video 9), while the right upper pulmonary vein appeared to drain into the right atrium, a finding that was later confirmed by computed tomography angiography (Figure 5).[3]

There was moderate to severe tricuspid regurgitation, with a peak velocity of 5.3 meters/second and a peak pressure gradient of 91 mmHg. Tricuspid annular plane systolic excursion (TAPSE) was 24.3 millimeters, and S' of the lateral annulus of the tricuspid valve was 15.6 centimeters/second, indicative of normal right ventricular systolic function. Doming of the pulmonary valve with an area of narrowing above the pulmonary valve was noted. The pulmonary valve was mildly thick. A small patent ductus arteriosus with a continuous shunt and an approximately 80 mmHg systolic and 65 mmHg diastolic pressure gradient across the PDA was indicative of normal pulmonary arterial pressure (Video 10).

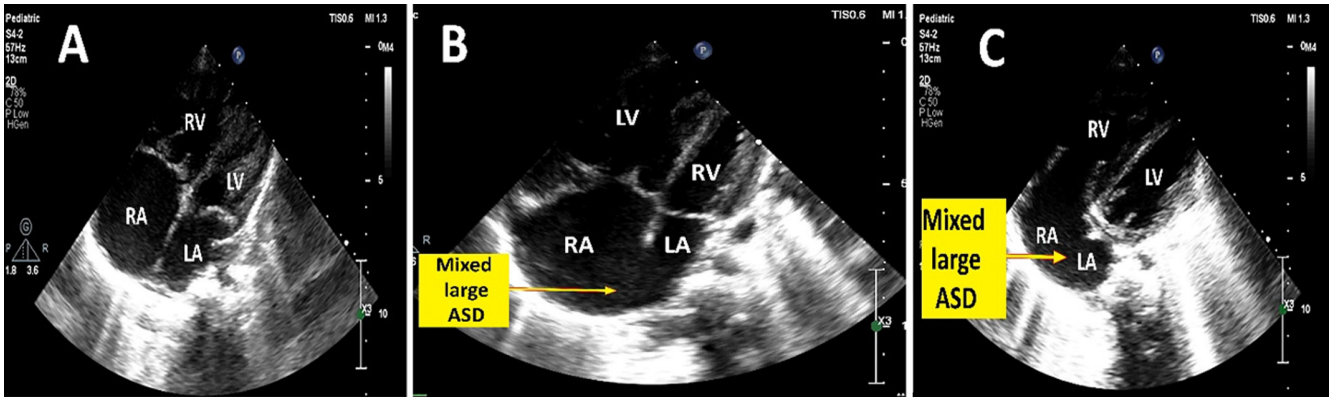


Figure 1. Figures A, B, and C demonstrate the importance of sweeping from the anterior (A) to the posterior (C) to accurately delineate the anatomy of the atrial septal defect using two-dimensional echocardiography. This approach reveals a notable variation in the size of the defect, which increases from the anterior to the posterior. It particularly highlights the near absence of a significant portion of the septum in the posterior plane at the level of the coronary sinus.

ASD, Atrial septal defect; LA, Left atrium; LV, Left ventricle; RA, Right atrium; RV, Right ventricle.

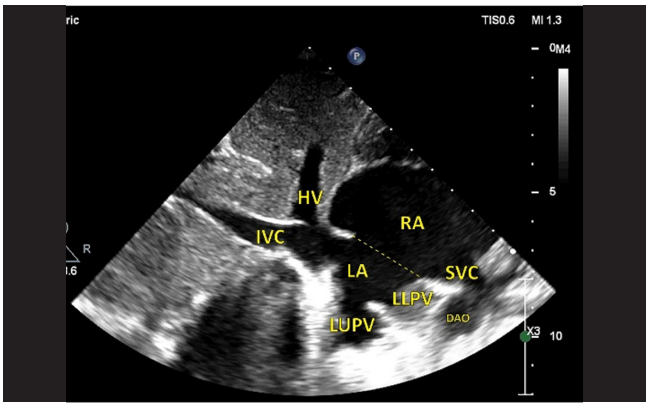


Figure 2. This subcostal echocardiogram depicts the normal drainage of the left-sided pulmonary veins and the drainage of the inferior vena cava and hepatic veins into the left atrium. The dotted line indicates the large atrial septal defect.
DAO, Descending aorta; LA, Left atrium; LLPV, Left lower pulmonary vein; LUPV, Left upper pulmonary vein; LV, Left ventricle; RA, Right atrium; RV, Right ventricle; SVC, Superior vena cava

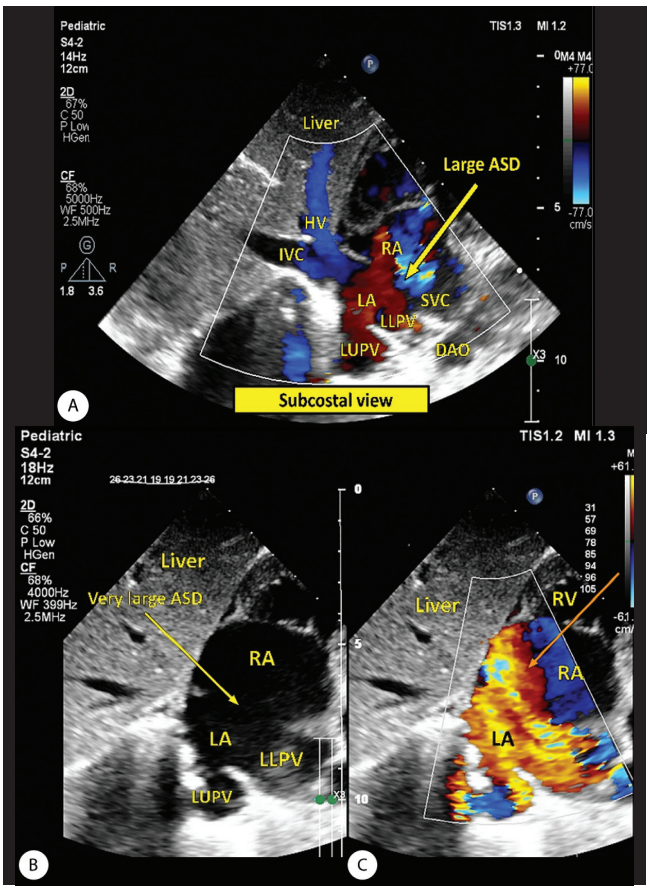


Figure 3. Figure A illustrates the drainage of the inferior vena cava (IVC) and hepatic vein (HV), as well as the left upper and lower pulmonary veins (LUPV and LLPV), into the left atrium. Figure B reveals the substantial size of the atrial septal defect (ASD), and Figure C depicts a significant left-to-right shunt across the ASD.
ASD, Atrial septal defect; DAO, Descending aorta; LA, Left atrium; LV, Left ventricle; RA, Right atrium; RV, Right ventricle

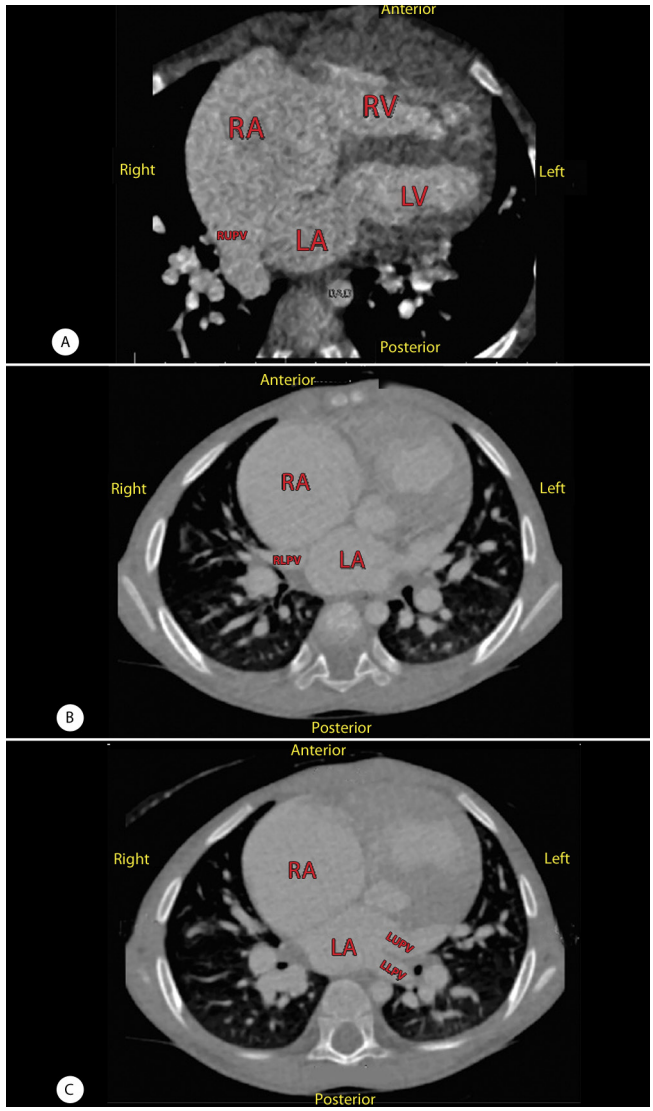


Figure 5. Computed tomographic angiography images of the patient revealed a normal connection of the right lower pulmonary vein and the left upper and lower pulmonary veins into the left atrium. Additionally, these images identify anomalous drainage of the right upper pulmonary vein into the right atrium, occurring at the site of the atrial septal defect.
LA, Left atrium; LLPV, Left lower pulmonary vein; LUPV, Left upper pulmonary vein; LV, Left ventricle; RA, Right atrium; RLPV, Right lower pulmonary vein; RV, Right ventricle; RUPV, Right upper pulmonary vein

The patient underwent elective cardiac surgery. A mid-sternotomy incision was made and total thymectomy was done for exposure. The pericardium was opened. RA and right ventricle (RV) were dilated. The PDA was ligated and transfixed. After heparin injection, total cardiopulmonary bypass was established with systemic hypothermia to 28° centigrade. The aorta was x-clamped, and custodial cardioplegia was given via the aortic root. RA was opened. There was a large secundum ASD with an extension to the ostium of the IVC (Figure 6).

ASD was closed using a sizeable pericardial patch. The

patch was sewn lower than usual to direct the IVC ostium into the RA. The IVC and hepatic vein were directed toward the right atrium and the right pulmonary vein toward the right atrium. (Figure 7).

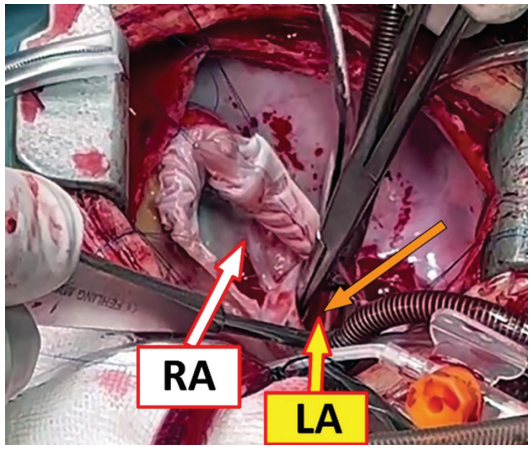


Figure 6. This image, captured in the operating room before surgical correction, shows the drainage of the inferior vena cava into the left atrium.
LA, Left atrium; RA, Right atrium

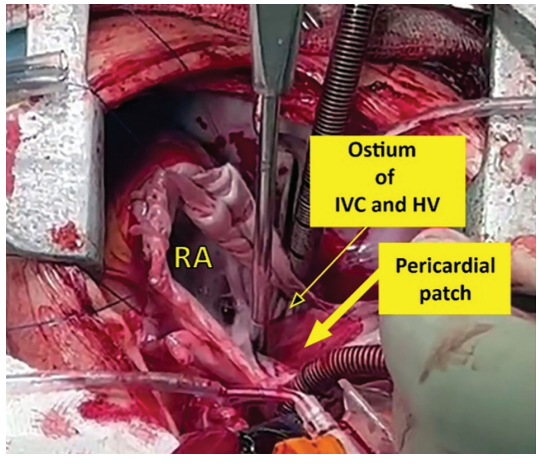


Figure 7. This post-repair image illustrates that the surgeon has placed the patch below the ostium of the inferior vena cava and the hepatic vein to redirect its flow into the right atrium.
IVC, Inferior vena cava; HV, Hepatic vein; RA, Right atrium

Limited commissurotomy and supra-annular pericardial patch reconstruction were used to resolve the valvar and supravalvar PS. Tricuspid valve annuloplasty was performed.

Interestingly, the day after the operation, the size of the RA and RV had returned to normal (Video 11). There was trivial tricuspid regurgitation and no obstruction to the route of IVC and hepatic vein into the RA and left upper pulmonary vein into the left atrium (Video 12).

The child had an uneventful and short stay in the open heart intensive care unit and was discharged with complete parent education on care at home. One week later, he was visited in the outpatient pediatric cardiology clinic. He was in good clinical condition, and the echocardiographic examination revealed no abnormal findings. At the six-month follow-up, the child's cardiac examination and oxygen saturation were normal. Furthermore, echocardiography confirmed normal drainage of the systemic and hepatic veins into the right atrium.

## *Discussion*

We reported an acyanotic boy with an exceedingly rare combination of huge mixed inferior sinus venous and secundum ASDs and mixed partial anomalous pulmonary and venous drainage associated with valvar and supravalvar PS and PDA with normal right ventricular function and normal pulmonary arterial pressure.

Inferior-type sinus venosus ASD is very rare.[1] Five septal zones can be distinguished anatomically within the normal interatrial septum. A mixed defect is defined as one that affects two or more atrial septal zones. Just 7% of ASDs are mixed defects.[4] Although the coexistence of inferior sinus venosus ASD and pulmonary stenosis has already been reported, the combination of the anomalies present in this case has not been reported to date.[5]

The diagnosis of IVC-type sinus venosus ASD is both uncommon and frequently overlooked. Snarr et al. reported the absence of the posterior rim on the parasternal short-axis view, the so-called "bald posterior wall," as a consistent finding that aids in diagnosing IVC-type ASD and differentiates it from secundum ASD with inferior extension.[6]

The deceptive aspect of this case was the absence of cyanosis and systemic arterial desaturation despite anomalous drainage of IVC and hepatic vein.[7] This finding can explained by the rare coexistence of congenital cardiac lesions. In cases where both the IVC and hepatic vein drain into the LA, coupled with a huge ASD that facilitates nearly complete blood mixing between the right and left atria and a partial anomalous pulmonary venous connection, systemic arterial desaturation may not be observed (Figure 8).

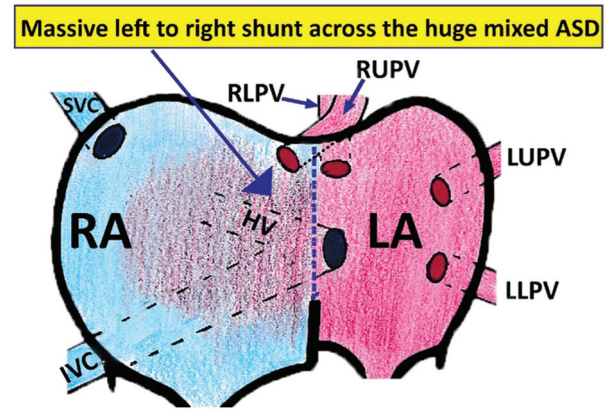


Figure 8. This figure illustrates why the patient's oxygen saturation remained

within the normal range despite the anomalous drainage of the inferior vena cava and hepatic vein. The dotted line indicates the presence of a large atrial septal defect.
ASD, Atrial septal defect; IVC, Inferior vena cava; LA, Left atrium; LLPV, Left lower pulmonary vein; LUPV, Left upper pulmonary vein; RLPV, Right lower pulmonary vein; RUPV, Right upper pulmonary vein; SVC, Superior vena cava

Consequently, the absence of arterial desaturation should not prevent the identification of anomalous drainage of the IVC and hepatic vein into the LA under these circumstances.

This case offers several crucial insights and practical implications.

1. In patients with ASD and anomalous drainage of the IVC and hepatic vein, achieving an accurate pre-operative diagnosis is crucial to avoid unexpected cyanosis following ASD closure.[8] The pediatric cardiac surgeon should be fully apprised of the patient's specific condition to correctly tailor the surgical patch placement to ensure the proper redirection of the IVC flow to the RA and the pulmonary veins to the LA. In our case, the patch was sewn lower than usual to achieve this result. Risk factors and causes of inadvertent diversion of IVC to LA after ASD closure are depicted in Table 1.[9-11]

Table 1. Causes and risk factors for accidental diversion of inferior vena cava into the left atrium during surgical closure of the atrial septal defect

| |
|---|
| Large secundum atrial septal defect |
| Low-lying atrial septal defect |
| Sinus venosus atrial septal defect of inferior vena caval type |
| Partial anomalous pulmonary venous connection to the right atrium |
| Mistaking Eustachian valve with the margin of the atrial septal defect |

2. Mixed ASDs require a more diligent echocardiographic examination, focusing on the integrity of pulmonary and systemic venous drainage and associated congenital heart anomalies, such as pulmonary stenosis and patent ductus arteriosus, as demonstrated in this case. In large mixed-type ASDs involving the IVC, sinus venosus, and secundum types, it is crucial to examine the potential for abnormal drainage of the IVC into the LA. The notable lack of atrial septum, coupled with the embryological predisposition for IVC flow towards the LA, predisposes the abnormal diversion of the IVC into the LA under these conditions. Additionally, it is crucial to elucidate the drainage pattern of the hepatic veins. The IVC and hepatic veins entered the LA through a single opening in the case presented.

3. Partial anomalous right upper pulmonary vein drainage can occur without a superior vena cava-type sinus venosus ASD.

4. When using two-dimensional echocardiography to examine the atrial septum, complete sweeps in the anterior-posterior planes are necessary to assess its integrity completely. When available, three-dimensional echocardiography is the preferred imaging modality for evaluating the atrial septum.[12] Additionally, it is crucial that during echocardiographic evaluations and surgical procedures, the Eustachian valve is not misidentified as the edge of the ASD.[9]

5. Long-term clinical monitoring is advisable to detect sinus node dysfunction in patients with repaired sinus venosus ASD.[13]

## Conclusion

In patients presenting with large mixed inferior sinus venosus and secundum ASDs, accompanied by anomalous drainage of the IVC and hepatic vein into the LA, arterial desaturation might not be a consistent finding. Thus, the absence of systemic arterial desaturation should not lead to the dismissal of partial anomalous systemic venous return. In the context of mixed ASDs, conducting a comprehensive evaluation of both pulmonary and systemic venous drainage is imperative, along with a meticulous search for any related congenital heart defects. Achieving an accurate pre-operative diagnosis is paramount, as it guides the correct positioning of the surgical patch for ASD closure, aiming to avert postoperative hypoxemia caused by the anomalous drainage of the IVC and hepatic vein into the LA. When three-dimensional echocardiography is not accessible, performing a thorough examination in the anteroposterior plane is critical to accurately identify the presence and size of the atrial septal defects.

## To watch the following videos, please refer to the relevant URLs:

https://jthc.tums.ac.ir/index.php/jthc/article/view/2099/1149
Video 1. This 4-chamber view shows right atrial and right ventricular enlargement and a large mixed atrial septal defect.
https://jthc.tums.ac.ir/index.php/jthc/article/view/2099/1150
Video 2. This video demonstrates that the size of the atrial septal defect increases upon posterior tilting of the probe, as highlighted by the visualization of the coronary sinus.

https://jthc.tums.ac.ir/index.php/jthc/article/view/2099/1151
Video 3. Similar to Video 2, posterior tilting of the probe, confirmed by the appearance of the coronary sinus, reveals an enlarged atrial septal defect.

https://jthc.tums.ac.ir/index.php/jthc/article/view/2099/1152
Video 4. This subcostal echocardiogram reveals the absence of a superior vena cava-type sinus venosus atrial septal defect.

https://jthc.tums.ac.ir/index.php/jthc/article/view/2099/1153
Video 5. This echocardiogram, taken from the subcostal view, displays the

hepatic vein and the inferior vena cava draining into the left atrium through a single opening, with the left-sided pulmonary veins also draining into the left atrium. For structural annotations, please see Figure 2.

https://jthc.tums.ac.ir/index.php/jthc/article/view/2099/1154
Video 6. This echocardiogram, captured from the subcostal view, is identical to Video 4 but includes the addition of color Doppler imaging.

https://jthc.tums.ac.ir/index.php/jthc/article/view/2099/1156
Video 7. Captured from a modified subcostal view, this echocardiogram more clearly depicts the entry of the inferior vena cava and the hepatic vein into the left atrium via a single opening.

https://jthc.tums.ac.ir/index.php/jthc/article/view/2099/1157
Video 8. This echocardiogram, captured from the subcostal view, is identical to video 6 with the addition of color Doppler imaging.

https://jthc.tums.ac.ir/index.php/jthc/article/view/2099/1158
Video 9. This subcostal echocardiogram demonstrates the normal connection of the right lower pulmonary vein to the left atrium.

https://jthc.tums.ac.ir/index.php/jthc/article/view/2099/1159
Video 10. Captured from the parasternal short-axis view, this echocardiogram displays the flow from the patent ductus arteriosus and the aliasing caused by pulmonary stenosis.

https://jthc.tums.ac.ir/index.php/jthc/article/view/2099/1160
Video 11. This postoperative echocardiogram, viewed from the 4-chamber perspective, shows the normalized sizes of the right atrium and ventricle, along with trivial tricuspid regurgitation.

https://jthc.tums.ac.ir/index.php/jthc/article/view/2099/1161
Video 12. This postoperative echocardiogram, viewed from the subcostal perspective, illustrates the direction of the inferior vena cava and the hepatic veins into the left atrium.

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