

Transcervical occlusion of atrial septal defect complicating with absence of hepatic segment of inferior vena cava in a patient with dextrocardia

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<https://doi.org/10.26599/1671-5411.2024.12.006>

A 52-year-old female patient was admitted to our department primarily complaining of “palpitations and shortness of breath for one month following physical activity.” Upon admission, the physical examination revealed that the cardiac dullness border was expanded to the left, a grade 2/6 systolic murmur at the 2nd to 3rd left intercostal spaces along the left sternal border, and a fixed split second heart sound in the pulmonary valve area. An electrocardiogram revealed sinus rhythm with incomplete right bundle branch block. Echocardiography demonstrated a secundum type atrial septal defect (ASD) with a 16 mm interruption in the atrial septal echo (Figure 1B–1D), good marginal conditions, and enlargement of the right atrium and right ventricle, with internal diameters of 45 mm and 29 mm, respectively. A chest radiograph showed dextrocardia (Figure 1A & 1E). Abdominal color Doppler ultrasound indicated a normal position of the liver and spleen. The patient's laboratory test results were unremarkable. The admission diagnosis was congenital heart disease (CHD) with dextrocardia and a secundum type ASD. Based on the aforementioned auxiliary examination results, the patient met the criteria for interventional therapy, and a right heart catheterization examination along with ASD closure was planned.

The patient was placed in the supine position, and the bilateral inguinal areas were routinely disinfected and draped. After puncturing the right femoral vein, a guidewire and a 6F end-hole catheter were inserted. Under X-ray anteroposterior fluoroscopy, the majority of the heart was located to the right of the spine with the apex pointing downward and to the right (Figure 2A). The gu-

idewire and catheter were seen ascending along the left edge of the spine, crossing the overlapping area with the cardiac silhouette, and then returning to the right atrium from the anatomical position of the superior vena cava. Subsequently, the guidewire traveled downward along the right atrium to the base of the cardiac silhouette, entered the right ventricle through the tricuspid valve, and then sharply angled upward into the pulmonary artery. The pressures measured in the pulmonary artery and right ventricle were 40/12 (22) mmHg and 40/0 mmHg, respectively, with no pressure gradient observed in the continuous pressure curve from the pulmonary artery to the right ventricle. Based on the catheter trajectory and angiography, it was determined that there was an absence of hepatic segment of the inferior vena cava with an open azygos vein (Figure 2B & 2C). A stiff long guidewire was passed through the interatrial communication into the left upper pulmonary vein via the route of the right femoral vein-right iliac vein-azygos vein-superior vena cava-right atrium. An 80 cm 12F delivery sheath was then inserted, but due to the length limitation of the sheath, the tip could only just pass the atrial septum and could not be advanced into the left atrium, failing to provide sufficient support, thus the femoral vein approach for occlusion was abandoned. Subsequently, the right internal jugular vein was punctured and angiography confirmed the blood return pathway (Figure 2D). A 6F end-hole catheter was inserted through the right internal jugular vein-superior vena cava-right atrium pathway, passing through the interatrial communication into the left atrium, and a stiff guidewire was advanced into the left atrium-right upper pulmonary ve-

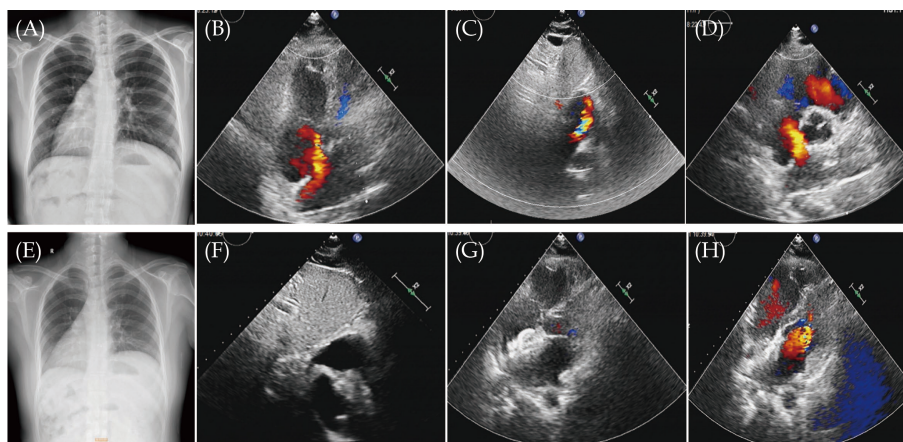


Figure 1 Preoperative and postoperative echocardiography and chest X-ray images. (A): The preoperative chest X-ray; (B–D): the images of the apical four-chamber view, subxiphoid biatrial view, and great artery short-axis view before surgery, showing a secundum atrial septal defect with a 16 mm interruption in the atrial septal echo and good marginal conditions; (E): the postoperative chest X-ray; (F & G): the images of the subxiphoid biatrial view and apical four-chamber view after occluder implantation, showing good positioning of the occluder; and (H): the postoperative apical four-chamber view showing no residual shunt across the atrial septum.

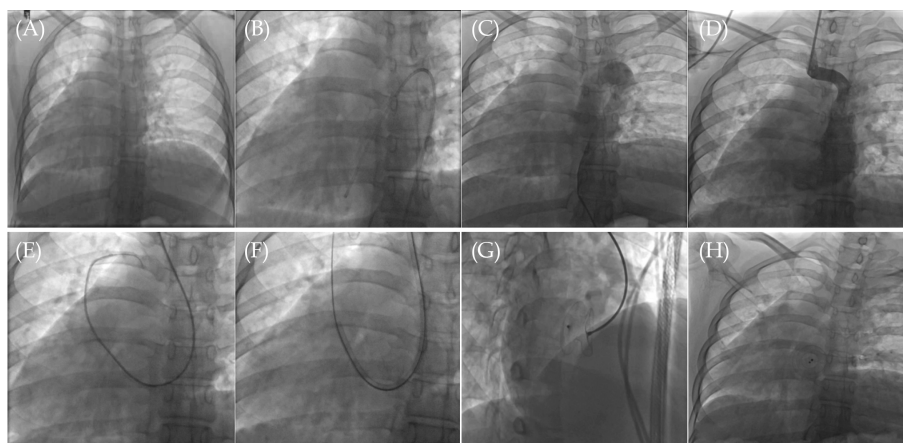


Figure 2 The entire process of transcatheter atrial septal defect occlusion. (A): Preoperative and postoperative anteroposterior fluoroscopic images; (B): end-hole catheter is advanced from the right femoral vein and the open azygos vein through the superior vena cava into the right atrium; (C): angiography confirms the absence of the hepatic segment of the inferior vena cava; (D): an end-hole catheter is inserted via the right internal jugular vein, and angiography shows blood returning through the superior vena cava to the right atrium; (E): the end-hole catheter is advanced via the right internal jugular vein to the right upper pulmonary vein; (F): after exchanging for a stiffer guidewire, the delivery sheath is introduced; (G): the occluder is successfully implanted through the sheath; and (H): post-release anteroposterior image of the occluder.

in (Figure 2E). A 12F delivery sheath was then inserted, and a 22 mm ASD occluder (Shenzhen Lifetech Scientific Co., Ltd., Shenzhen, China) was used for occlusion (Figure 2F). Under X-ray fluoroscopy, the sheath was retracted, and the left and right atrial discs were sequentially opened, observing the good morphology of the occluder and performing repeated traction tests to ensure secure fixation (Figure 2G). Intraoperative re-examination with transthoracic echocardiography showed no residual shunt across the atrial septum, normal valve activities, and no impact on the coronary sinus or pulmonary vein

flow, and the occluder was released (Figure 2H).

The patient was followed up at 1, 3, 6, and 12 months postoperatively, with good general condition, no palpitations, shortness of breath, or other discomforts, the original cardiac murmur disappeared, echocardiography showed good position of the occluder, no residual shunt across the atrial septum (Figure 1F–1H), and normal valve activities; electrocardiogram showed sinus rhythm, incomplete right bundle branch block, and no other types of arrhythmias.

Dextrocardia is a relatively rare congenital cardiac po-

sitional anomaly, with an incidence of approximately 1 in 12,000 pregnancies, where the heart is located in the right chest. Although the cardiac apex points to the right, the relationships between the cardiac chambers are not mirror-imaged but are due to the displacement and rotation of the heart. It is usually associated with other cardiac malformations, such as ASD and ventricular septal defects. In recent years, with the continuous advancement of interventional treatment techniques for CHD and the increasing maturity of operational skills, even patients with dextrocardia combined with simple CHD, such as ASD or ventricular septal defects, can be treated with interventional occlusion.^[1-3] The absence of the hepatic segment of the inferior vena cava is usually related to the abnormal opening of the azygos or hemiazygos vein. In the general population, its incidence is about 0.1% to 0.6%, while in patients with CHD, this proportion significantly increases to 1% to 3%.^[4] Traditional ASD interventional occlusion surgery requires the construction of a track through the complete femoral vein-inferior vena cava-right atrium pathway to deliver the occluder. However, for patients with ASD combined with the absence of the infrahepatic segment of the inferior vena cava, the lack of a normal inferior vena cava return system poses a significant challenge to interventional occlusion treatment.

For patients with dextrocardia and the absence of the hepatic segment of the inferior vena cava who also have an ASD, the difficulty of interventional treatment is extremely high, with only a very small number of successful percutaneous interventional treatment cases reported internationally, and there are reports of both femoral vein and internal jugular vein approaches.^[4-7] This case is the first reported internationally of a successful occlusion via the internal jugular vein approach in a patient with dextrocardia and the absence of the hepatic segment of the inferior vena cava. After failing to perform interventional treatment through the femoral vein approach, we constructed a new treatment track via the internal jugular vein-superior vena cava pathway and successfully implemented the interventional occlusion treatment.

In patients with the absence of the hepatic segment of the inferior vena cava and an open azygos vein, the origin of the azygos vein is located in the right lumbar ascending vein. It ascends through the right crus of the diaphragm into the posterior mediastinum and forms the azygos arch by bending forward at the level of the fourth thoracic vertebra, ultimately draining into the superior

vena cava and thus returning to the right atrium. For percutaneous interventional occlusion treatment in such patients, there are two major technical challenges: the 90° bend at the azygos arch and another 90° bend when passing through the ASD from the right atrium to the left atrium. The former mainly hinders the smooth delivery of the sheath, while the latter affects the smooth passage of the guidewire and sheath into the pulmonary vein to establish the track.^[4] Although there have been reports of ASD occlusion via the hepatic vein approach, this method is technically challenging and has a higher rate of complications, making it unsuitable as the preferred interventional treatment method.^[8,9] Additionally, Truong, *et al.*^[7] reported a case of successful occlusion via the jugular vein approach, in which a sheath with a large curvature was used to smoothly pass through the severed end of the atrial septum. These reports indicate that despite technical challenges, even complex ASD cases can be successfully treated with interventional occlusion, providing valuable experience and confidence for future treatments.

In this case, the patient's left and right iliac veins were directly connected to the azygos vein, which drained into the superior vena cava. Based on the experience of interventional treatment for this patient, it was observed that when operating through the femoral vein approach, the presence of two large bends in the track made the passage of the sheath painful for the patient under local anesthesia due to the sheath traction, and the intraoperative manipulation was more difficult. It is recommended to be gentle during the procedure, use general anesthesia, and select specially made extended sheaths. When operating through the jugular vein approach, the shorter distance and avoidance of the azygos arch bend make it easier to introduce a sheath with a large curvature for occlusion, and it is recommended as the preferred pathway for such patients. Before CHD interventional treatment, it is important to examine the position of the inferior vena cava to clarify any anomalous connections. For the interventional treatment of dextrocardia with ASD, due to the rightward rotation of the heart, the occluder release and observation of its morphology during the occlusion procedure require a left anterior oblique 30° plus cranial 20° (or even posteroanterior, adjusted according to the actual situation to observe the best position of the occluder) instead of the conventional left anterior oblique 50° plus cranial 20°. In dealing with such complex cases, the medical team needs to consider the



patient's specific situation comprehensively, formulate a personalized treatment plan, and detailed preoperative examination, thorough preoperative preparation, and rich interventional operation experience are the keys to successful treatment.

ACKNOWLEDGMENTS

All authors had no conflicts of interest to disclose.

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Please cite this article as: XIAO JW, WANG ZC, GENG JS, WANG JM, WANG QG. Transcervical occlusion of atrial septal defect complicating with absence of hepatic segment of inferior vena cava in a patient with dextrocardia. *J Geriatr Cardiol* 2024; 21(12): 1149–1152. DOI: 10.26599/1671-5411.2024.12.006

