Transcatheter secundum atrial septal defect closure using intracardiac echocardiography in adult patient with azygos/hemiazygos continuation of the Inferior Vena Cava

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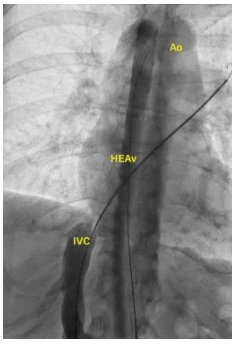
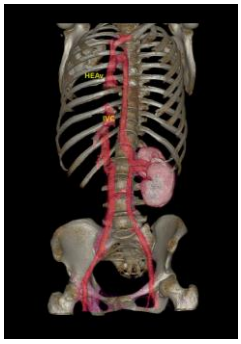
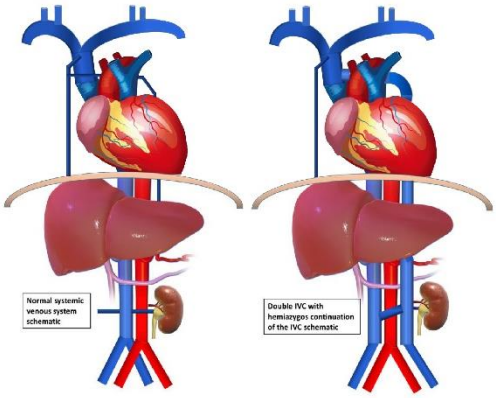
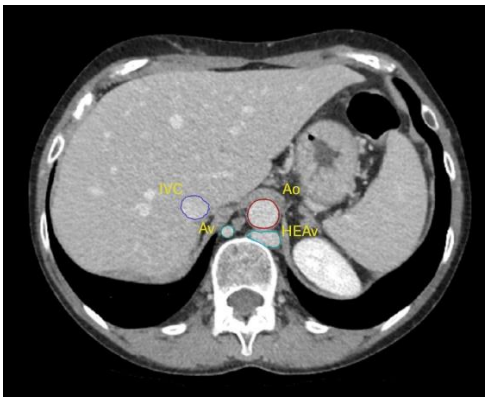
Introduction. Percutaneous closure of an isolated secundum atrial septal defect (ASD) has became the first-line treatment in patients with sufficient rim. In patients with secundum ASD, the procedure is mostly performed through the femoral vein. Standard techniques include the use of balloon-size technique and intracardiac echocardiography (ICE), the latter as an accepted alternative to the transesophageal echocardiography (TEE). Congenital anomalies of the deep thoracoabdominal venous system are caused by variations in the development during embryogenesis. As described in the literature, knowledge of this anatomic arrangement is important in percutaneous cardiopulmonary procedures.





Case summary. A 53-year-old female patient was admitted to our cardiology department with recent complaint of dyspnea. Physical examination revealed a fixed splitting second heart sound. Chest radiography showed prominent pulmonary vasculature. Transthoracic echocardiography revealed enlarged right heart cavities and TEE confirmed a 20 x 12 mm secundum ASD, with just enough rims for percutaneous interventional closure, except the anterior rim (2 mm).
The patient was admitted to catheter laboratory for percutaneous closure of secundum ASD through the right femoral approach.





Results. The ICE was inserted via 9 F long sheath from the left femoral vein and advanced to attempt through the Inferior Vena Cava (IVC) to enter into the right atrium. Venography was performed because of the abnormal course of the catheter and an azygos (Av)/hemiazygos (HEAv) continuation of the left iliac vein, up to the Superior Vena Cava (SVC).
For this double IVC with HEAv continuation of the left-side IVC and for a very acute angle of the draining A/HEA veins into the SVC, it was not possible to advance the ICE into the right atrium through this venous system. The right femoral vein was therefore catheterised twice: proximally with an 7 F sheath, which was used for preoperative right cardiac catheterization and pulmonary pressure measurement and replaced with 12 F long sheath for device implantation; distally with 9 F sheath for the ICE study in the right atrium.
The interatrial septal defect was passed through using a superstiff-0.035' Amplatzer wire and directed toward the left superior pulmonary vein. The dimensions of the ASD was confirmed by the sizing balloon, and compared with the ICE. For the small anterior rim, a 37 mm Gore Cardioform Occluder was loaded and advanced through the sheath, and the defect was closed using the routine protocol. The position and stability of the device were evaluated on ICE. No post-procedure residual shunt was detected.
Postoperatively, computed tomography angiography was performed through the lower extremity. The patient was discharged 2 days later with oral aspirin.

Conclusions. The interventional endovascular maneuvers normally coded for vascular access, materials, and technique, must be carefully re-evaluated in case of congenital anomalies involving the systemic venous return to the right atrium. These anomalies are usually asymptomatic and incidentally found, and the major clinical significance of their recognition is prevent misdiagnosis, which can have implications on invasive procedures and the abdominal hemorrhage (a highly feared complication).

The authors declare absence of conflicts of interest