Radiofrequency ablation of typical atrial flutter with access through the azygos vein in a patient with heterotopia utilizing high-density electroanatomic mapping

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Introduction
Discontinuation of the inferior vena cava (IVC) is a rare congenital anomaly and is most often caused by the absence of the pars hepatica. Additional congenital heterotopias may be present. In these cases, the venous blood of the lower body part is often drained via the azygos vein, which reaches the right atrium in the upper part. The interventional catheter ablation of typical atrial flutter seems uncomplicated unless a previously unknown interruption of the IVC suddenly presents the electrophysiologist with a challenge.

Case report
A 77-year-old woman presented with acute heart failure, dyspnea, and first-time typical atrial flutter. A few days before, the patient suddenly experienced palpitations and shortness of breath. The left ventricular ejection fraction was moderately reduced. No history of heart disease was known at the time of admission. The physical examination was unremarkable except for slight lower leg edema. Owing to known good long-term success of the procedure, interventional therapy for atrial flutter was discussed with the patient. The preinterventional transesophageal echocardiography was inconspicuous. This was followed by the electrophysiological examination. Under sedation with propofol and local anesthesia of the right groin, 3 sheaths were placed in the right femoral vein. Via this femoral access, the coronary sinus (CS) catheter (Webster CS, Biosense Webster, Diamond Bar, CA) was advanced, but the right atrium could only be accessed from the superior vena cava. Angiographic examination confirmed the suspicion of a missing liver segment of the IVC with continuity via the azygos vein. We decided to ablate the atrial flutter via this superior access. Using the CARTO 3 electroanatomic mapping system (Biosense Webster, Diamond Bar, CA) the thoracic part of the azygos vein and the right atrium as well as the CS were reconstructed. With additional use of the CARTO UNIVU module (Biosense Webster, Diamond Bar, CA), the radiation exposure for the patient and staff was kept low. After positioning of the CS catheter and detection of a CS activation from proximal to distal, an entrainment maneuver was performed at the cavo-tricuspid isthmus using a Thermocool SmartTouch SF (Biosense Webster, Diamond Bar, CA) contact force–measuring catheter, which was stabilized with a steerable sheath (MobiCath, Biosense Webster, Diamond Bar, CA).

KEY TEACHING POINTS
- Using high-density electroanatomic mapping and contact force technology for ablation of typical atrial flutter in patients with discontinuation of the inferior vena cava and venous drain via the azygos vein can help to reduce radiation exposure, improve lesion quality, and allow visualization of anatomic abnormalities, scar regions, and final conduction block.
- Congenital cardiac abnormalities should be considered in situations where the projected catheter position does not match with expected positional relationship.
- Radiofrequency ablation of typical atrial flutter and final proof of ablation success in this rare congenital anomaly is feasible using a femoral approach and catheter access via the azygos vein.

KEYWORDS
Azygos vein; Catheter ablation; Congenital heart disease; High-density mapping; Inferior vena cava discontinuity; Isthmus-dependent flutter; Radiofrequency ablation; Typical atrial flutter

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This confirmed isthmus-dependent atrial flutter with a cycle length of 320 ms. The 3-dimensional high-density voltage map, which was created with a multielectrode mapping catheter (PentaRay, Biosense Webster, Diamond Bar, CA), helped identify the nonconducting stump region of the interrupted IVC (Figure 1). As a nonconducting structure, this region allows the use of a classic inferior ablation line. The ablation procedure was performed with a power of 40 W and led to termination of atrial flutter during energy delivery. Subsequently, the conduction block was examined and confirmed under stimulation from the proximal CS segment, which is clearly recognizable in the local activation time (LAT) map (Figure 2). The result was stable throughout a waiting period of 20 minutes. The total examination time was 155 minutes with a total dose area product of 844 cGy·cm², which was predominantly driven by the loops taken after administration of contrast agent in the right femoral vein to confirm the suspicion of IVC discontinuity. On the following day, the standard variant of an IVC with missing liver segment and continuity via the extended retrocrural vena azygos was confirmed by computed tomography (CT) diagnosis. In addition, other heterotopias were noted: a double kidney on the left side, a retroaortic left renal vein, and the right upper pulmonary vein opening into the right atrium.

Figure 1  Three-dimensional reconstruction and bipolar voltage map of the azygos vein and right atrium with coronary sinus. Visitags visualize the inferior line connecting the tricuspid valve annulus and the nonconducting region of the interrupted inferior vena cava. A: Right anterior oblique projection 30°. B: Left anterior oblique projection 45°.

Figure 2  A: Local activation time (LAT) map created during stimulation from the coronary sinus in left anterior oblique projection (45°) for demonstration of cavotricuspid isthmus block. B: Inferior view on the right atrium with LAT map and visualization of conduction block.
with concomitant enlargement of the pulmonary arteries. Owing to suspect partially calcified 9 mm focus retromamillary on the left side, further gynecologic treatment is planned. Renewed administration of contrast agent via the femoral vein for 3-dimensional reconstruction of vena cava inferior and vena azygos was not desired by the patient. The patient remained symptom-free and had no further arrhythmias.

Discussion
The IVC arises between the sixth and eighth week of embryonic development out of 3 paired embryonic veins (supracardinal, subcardinal, and posterior cardinal veins). Aberrations in this developmental step lead to a large number of variations in the venous drainage of the lower body. A lack of the pars hepatica of the IVC leads to azygos continuation, as in the case described here. The prevalence of this variant is 0.6% in the population with congenital heart defects. In our patient, there was no structural conspicuousness of the heart except for a partial anomalous pulmonary venous connection. The hepatic segment did not drain directly into the right atrium. This was evident in the 3-dimensional reconstruction as well as in CT examination. In transesophageal echocardiography prior to ablation, no anatomic peculiarity was assumed in the bicaual view. Potentially, the stump of the IVC was misinterpreted as a regular junction of the inferior vein. Nevertheless, CT diagnosis revealed several heterotopias, already described above. Apart from the newly occurring cardiac arrhythmias, the patient was free from physical complaints. Ablation of typical atrial flutter requires venous access to the right atrium. It has recently been published that ablation with access via the subclavian vein can be successful. This is necessary if several segments of IVC are missing and femoral access is not possible. Also conceivable is venous access via the jugular vein. In the few previously published case reports of similar anatomic conditions with ablation procedures owing to typical atrial flutter, no high-density mapping was used, but predominantly conventional fluoroscopic approach was employed.

We decided to use high-density mapping to confirm the usual IVC estuary as a nonconductive structure via a bipolar voltage map, thus not only to achieve termination of atrial flutter under ablation, but also to confirm a block along the ablation line for long-term freedom from recurrence (Figure 2). A final LAT map under CS stimulation could visualize the blocked isthmus. It is important to emphasize that cases such as this can be undertaken, even when venous anomalies are diagnosed at the time of catheterization. However, it is important to keep in mind the recommendation that the care of adults with more complex congenital heart disease should be coordinated with a regional adult congenital heart disease specialist. To the best of our knowledge, this is the first description of an ablation of typical atrial flutter with use of high-density mapping in a patient with IVC discontinuation and venous drainage via the azygos vein.

Conclusion
Radiofrequency ablation of the cavotricuspid isthmus in patients with typical atrial flutter and interruption of IVC is feasible using an access via the azygos vein. The target region is accessible. Sufficient tissue contact during ablation and final proof of conduction block can be verified by using high-density electroanatomic mapping and contact force technology.

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