

## Case Report

# Electrophysiologic study and catheter ablation of a supraventricular tachycardia in a patient with inferior vena cava congenital anomaly

Stylianos Dragasis, Panagiotis Mililis, Athanasia Megarisiotou, George Bazoukis, Athanasios Saplaouras, Athena Mpatsouli, Fotios Toulgaridis, Efstathia Prappa, Konstantinos P Letsas, Michael Efremidis

*Second Department of Cardiology, Laboratory of Cardiac Electrophysiology, "Evangelismos" General Hospital of Athens, Greece*

Received May 30, 2020; Accepted August 18, 2020; Epub October 15, 2020; Published October 30, 2020

**Abstract:** Electrophysiologic procedures are performed widely nowadays, for the successful treatment of several cardiac arrhythmias. In this case report, we describe a rare congenital anomaly of the inferior vena cava, as an incidental finding during a scheduled electrophysiologic procedure for a supraventricular tachycardia ablation. The patient is a 32 year old male with an unremarkable medical history, suffering from sustained episodes of paroxysmal tachycardia. The electrophysiological maneuvers confirmed the presence of atrioventricular nodal reentry tachycardia, followed by a successful slow pathway ablation. We provide imaging details and guidance on the successful catheter positioning. In cases like this, the prognosis is excellent, while the follow up of our patient was unremarkable.

**Keywords:** Inferior vena cava, congenital anomaly, electrophysiologic study

### Introduction

Inferior vena cava (IVC) congenital anomalies stem from a variety of setbacks in its sequential development, such as abnormal regression or persistence of any of the three embryonic paired veins: sub-, supra- and postcardinal [1-3]. The cumulative prevalence of IVC congenital anomalies has been reported to be present in up to 8.7% of the general population worldwide [4]. While most of them are asymptomatic, there seems to be a correlation of their presence and other complex syndromes such as cardiac malformations, asplenia and polysplenia, which may constitute their timely identification crucial. Furthermore, awareness of their existence helps to avoid image misinterpretation and to improve the quality in preprocedural planning before a cardiac catheterization via the femoral vein. We must also take into account that there are limited data in the worldwide literature (mainly a few case reports) regarding patients with congenital anomalies of the IVC who have been subjected to cardiac catheterization and ablation.

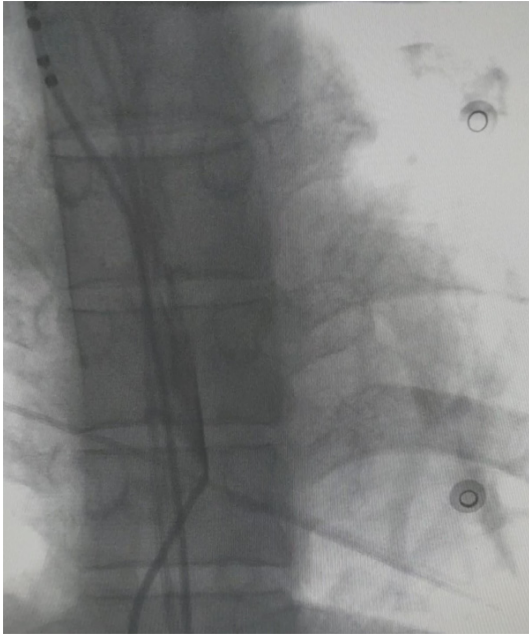
### Case report

A 32 year old man was referred to our electrophysiology laboratory due to episodes of sustained, symptomatic supraventricular tachycardia, non-responsive to oral antiarrhythmic medications. His past medical history was unremarkable. He did not smoke and reported no other drug use. Written informed consent was provided by the patient before the procedure.

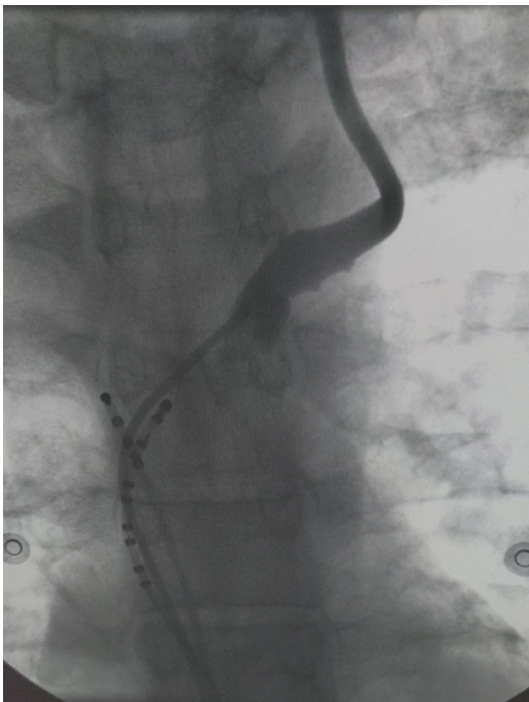
Physical examination upon presentation revealed a respiratory rate of 12/min, regular heart rate of 65/min and blood pressure of 110/70 mm Hg. His jugular veins were not distended, and lung fields were clear on auscultation. The patient's oxygen saturation was 99% on ambient air. Heart auscultation did not reveal any pathological sign.

During catheter manipulation and positioning at the level of the heart we noticed far field atrial and ventricular signals from the bipoles of all the catheters, along with the impression that they were not inside the chambers of the heart. **Figure 1** shows contrast agent administration

## Inferior vena cava congenital anomaly

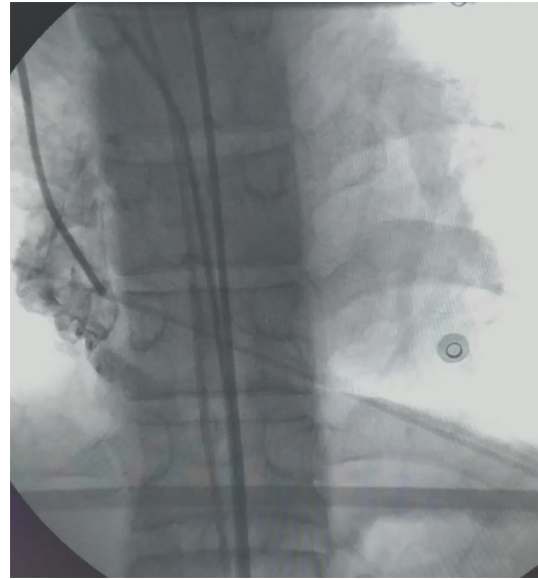


**Figure 1.** Contrast agent administration via a long sheath at the level of the heart, along with two electrophysiology catheters. Contrast administration proved that our catheters were positioned inside a vessel and not in the heart chambers.



**Figure 2.** Angiographic appearance of the left brachiocephalic vein.

via a long sheath at the level of the heart, along with two electrophysiology catheters. Contrast



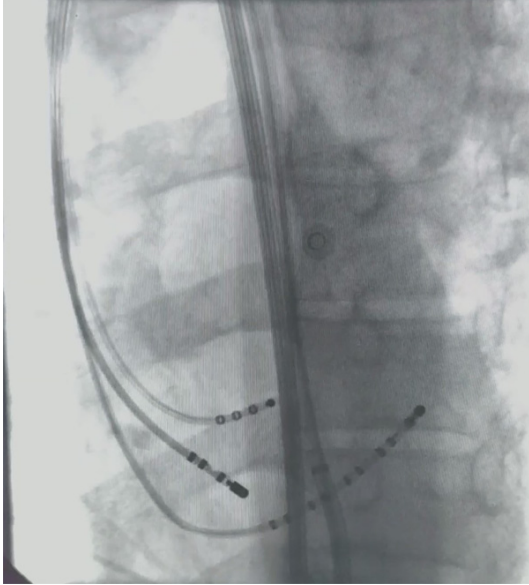
**Figure 3.** Angiographic appearance of the superior vena cava.

administration proved that our catheters were positioned inside a vessel and not in the heart chambers.

Further contrast agent administrations revealed more about the “curious” venous anatomy in our patient, such as the angiographic appearance of his left brachiocephalic vein (**Figure 2**) and his superior vena cava (**Figure 3**). It was then immediately clear that we were facing with a probable congenital anomaly of our patient’s IVC, in which instead of emptying blood directly into the right atrium, it seemed to drain it directly into the superior vena cava, bypassing the lower atrium. Using the long sheath, we then decided to place the decapolar coronary sinus catheter via this unexpected and rare route.

Following the decapolar coronary sinus catheter placement, the His catheter was also placed at the His-region. Final catheter positioning and routes, along with the ablation catheter, are clearly depicted in **Figure 4**. Our patient’s tachycardia was then easily induced, and after several electrophysiologic maneuvers, a diagnosis of typical (slow-fast) atrioventricular nodal reentry tachycardia (AVNRT) was made. Following successful slow pathway ablation at the typical area of the inferior triangle of Koch, the arrhythmia could not be induced, even after aggressive pacing protocols and isoproterenol administration.

## Inferior vena cava congenital anomaly



**Figure 4.** Final catheter positioning and routes, along with the ablation catheter located at the “slow pathway” area.

Finally, the patient’s clinical course was uneventful, without any episodes of tachyarrhythmia 1 year after his initial presentation. The electrophysiology procedure was approved by the Ethics Committee of Evangelismos hospital.

### Discussion

In this case report, we demonstrate a relatively rare IVC congenital anomaly, in an otherwise healthy adult male suffering from sustained episodes of AVNRT. This anomaly has been previously reported as “infrahepatic interruption of the IVC with azygos continuation”, with a general prevalence of 0.5-2.1% [1, 5, 6]. The embryological division of the IVC is characterized by 4 segments (hepatic, prerenal, renal and postrenal segment) formed between the 4<sup>th</sup> and 8<sup>th</sup> week of gestation, with different development for each one. These segments derive from anastomoses and regression of several fetal veins, such as posterior cardinal, anterior cardinal, common cardinal, superior cardinal and subcardinal vein. Given the fact that the last portion of the posterior cardinal veins gives rise to the azygos and hemiazygos vein, anomalies in the development of these embryonic veins such as persistence, pathological anastomosis or regression, give rise to several IVC congenital anomalies.

Discontinuation of IVC with azygos continuation can be the result of a failure in the anastomosis between the right subcardinal and hepatic embryonic vein, with consequent atrophy of the subcardinal vein [1, 5]. In our case, the infrarenal IVC continues through the azygos vein. Left IVC can also be observed in cases where the infrarenal IVC continues as the hemiazygos vein [7]. Although this anomaly has been reported as an incidental finding in typically asymptomatic individuals, the large azygos or hemiazygos vein may be easily misinterpreted as adenopathy or aortic pathology (such as dissection, rupture etc.) due to its close proximity and course to the descending thoracic aorta. The enlarged azygos vein could also result in enlarged mediastinum or give the false impression of a paratracheal mass. All the aforementioned scenarios highlight the importance of preoperative knowledge of this anomaly, especially in patients undergoing cardiopulmonary bypass surgery, or other surgical procedures involving the mediastinum.

Other congenital anomalies of the IVC include double IVC (0,2-3% prevalence) [3], portocaval shunt, retroaortic and circumaortic left renal vein, membranous obstruction of the intrahepatic IVC, absence of infrarenal IVC and retrocaval ureter. Ultrasonography with Doppler flow imaging may constitute the initial evaluation, but usually a CT or even better a MRI scan is required to depict the exact anatomical variant of the IVC anomaly.

### Conclusion

IVC congenital anomalies are usually an incidental finding in otherwise healthy subjects. Depending on the anomaly, their reported prevalence ranges between 0.2-8.7% in the general population. Correct and timely identification of these anomalies may prove essential in order to avoid false diagnoses and facilitate correct preprocedural planning. These anomalies may prove particularly challenging in catheter manipulation and positioning, during electrophysiology procedures.

### Disclosure of conflict of interest

None.

**Address correspondence to:** Dr. Konstantinos P Letsas, Second Department of Cardiology, La-

## Inferior vena cava congenital anomaly

boratory of Cardiac Electrophysiology, Evangelismos General Hospital of Athens, Athens 10676, Greece. Fax: +302132041344; E-mail: k.letsas@gmail.com

### References

- [1] Kandpal H, Sharma R, Gamangatti S, Srivastava DN and Vashisht S. Srivastava and sushma vashisht. imaging the inferior vena cava: a road less travelled. *Radiographics* 2008; 28: 669-689.
- [2] Sheth S and Fishman EK. Imaging of the Inferior Vena Cava with MDCT. *AJR Am J Roentgenol* 2007; 1243-1251.
- [3] Bass JE, Redwine MD, Kramer LA, Huynh PT and Harris JH Jr. Spectrum of congenital anomalies of the inferior vena cava: cross-sectional imaging findings. *Radiographics* 2000; 20: 639-652.
- [4] Kellman GM, Alpern MB, Sandler MA and Craig BM. Computed tomography of vena caval anomalies with embryologic correlation. *Radiographics* 1988; 8: 533-556.
- [5] Awais M, Rehman A, Baloch NU and Salam B. Multiplanar imaging of inferior vena cava variants. *Abdominal Imaging* 2014; 40: 159-166.
- [6] Blanchard DG, Sobel JL, Hope J, Raisinghani A, Keramati S and DeMaria AN. Infrahepatic interruption of the inferior vena cava with azygos continuation: a potential mimicker of aortic pathology. *J Am Soc Echocardiogr* 1998; 11: 1078-1083.
- [7] Minniti S and Procacci C. Congenital anomalies of the venae cavae: embryological origin, imaging features and report of three new variants. *Eur Radiol* 2002; 12: 2040-2055.