

Inferior Vena Cava Agenesis: an incidental diagnosis during electrophysiological study

Sérgio Ferreira de Ferreira Filho¹; Marco Antonio Vinciprova Dall Agnese^{2,*}; Tiago Luiz Luz Leiria¹; Luísa Rohr Schäfer²; Marcelo Kruse¹; Gustavo Glotz de Lima^{1,2}

ORCID ID

Ferreira Filho SF  <https://orcid.org/0000-0001-7374-1932>

Agnese MAVD  <https://orcid.org/0000-0001-5829-5954>

Leiria TLL  <https://orcid.org/0000-0002-3905-102X>

Schäfer LR  <https://orcid.org/0009-0003-1478-1338>

Kruse M  <https://orcid.org/0000-0002-2429-4491>

Lima GG  <https://orcid.org/0000-0003-0097-5206>

ABSTRACT

Inferior Vena Cava Agenesis (IVCA) is a rare congenital anomaly occurring in 0.0005% to 1% of the general population. Although often asymptomatic, IVCA significantly increases the risk of serious thromboembolic events, including chronic venous insufficiency, pulmonary thromboembolism, and deep vein thrombosis (DVT). This study investigates the incidence and clinical implications of IVCA discovered incidentally during electrophysiological studies (EPS) at the Instituto de Cardiologia do Rio Grande do Sul over the past 27 years. A cross-sectional review of 13,194 EPS reports from January 1997 to July 2024 identified 12 cases (0.09%) of incidentally diagnosed IVCA. Of the patients in question, 66.7% were female, with a mean age of 44.6 years. One patient had a history of complex heart disease, and arrhythmia induction was a potential outcome in 66.7% of cases. Atrioventricular node reentry tachycardia (AVNRT) was the most common arrhythmia observed. Catheterization of the right heart chambers via the azygos system was feasible in 83.3% of cases. One patient experienced deep vein thrombosis and pulmonary thromboembolism post-procedure, which was managed with anticoagulation. The study highlights the importance of recognizing IVCA during EPS as it can impact catheterization techniques and procedural outcomes. Although IVCA is a rare occurrence, it should be considered as a potential cause of difficulties encountered during catheterization procedures, and alternative access methods should be prepared. This awareness is crucial for preventing complications and ensuring effective treatment of cardiac arrhythmias.

KEYWORDS: Inferior Vena Cava Agenesis; Electrophysiological Techniques, Cardiac; Arrhythmias, Cardiac; Radiofrequency Ablation.

INTRODUCTION

Inferior Vena Cava Agenesis (IVCA) is a rare congenital malformation that can be caused by a defective embryological process or by perinatal complications, such as perinatal thrombosis. IVCA has an estimated prevalence of 0.0005% to 1% in the general population, and albeit generally asymptomatic, this condition has a vital is associated with an increased risk

1. Fundação Universitária de Cardiologia  – Instituto de Cardiologia – Porto Alegre - RS, Brazil.

2. Universidade Federal de Ciências da Saúde de Porto Alegre  – Porto Alegre – RS, Brazil.

*Correspondence author: marcoda@ufcspa.edu.br

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of thromboembolic events, including chronic venous insufficiency, pulmonary thromboembolism and deep vein thrombosis (DVT), being found in approximately 5% of young adults diagnosed with unprovoked proximal DVT¹.

An electrophysiological study (EPS) is an invasive test mainly used to diagnose heart rhythm and conduction abnormalities. Furthermore, this procedure has therapeutic and prognostic value due to the possibility of performing an ablation to treat reentrant circuits and ectopic foci. The examination is typically conducted with access to the heart through a femoral or subclavian venipuncture to facilitate the insert a catheter into the heart. Potential challenges with catheter progression may indicate the possibility of IVCA. As this possibility frequently manifests in asymptomatic patients, the IVCA can be incidentally diagnosed during an electrophysiological study, needing other manoeuvres to confirm the diagnosis².

The present study reports the occurrence of incidentally diagnosed IVCA during electrophysiological procedures and the success rate for treating the arrhythmia in these patients over a 14-year experience in our centre. Our data is based on what has been experienced at the Electrophysiology Laboratory at the Instituto de Cardiologia do Rio Grande do Sul during the last 28 years.

METHODS

A cross-sectional study was conducted, wherein reports of electrophysiological studies conducted between January 1, 1997, and July 31, 2024, at the Electrophysiology Laboratory of the Institute of Cardiology in Porto Alegre, were reviewed using the keyword “agenesis”. The selected reports were individually examined by the researchers to confirm the diagnosis, with particular attention paid to the following details: gender, age, structural heart disease, arrhythmia induction, and the possibility of reaching the right heart.

Categorical variables were demonstrated through absolute and relative frequencies in a descriptive study. Continuous variables were presented as mean and standard deviation.

RESULTS

Over the course of 28 years, a total of 13,194 EPS were completed. Among them, 12 (0.09%) were conducted in patients with incidentally diagnosed IVCA (Fig. 1).

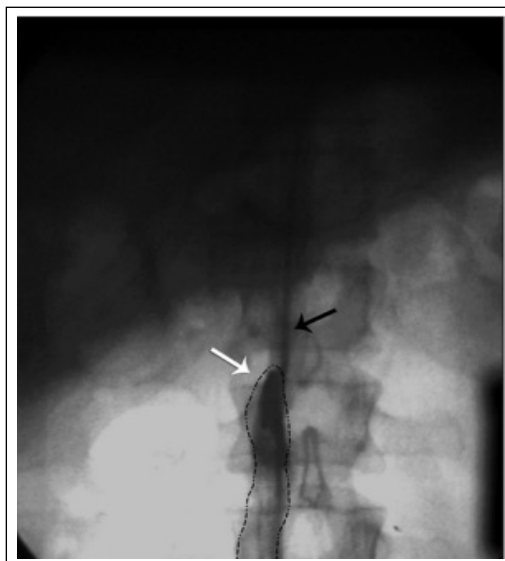


Figure 1. Venous angiography showing inferior vena cava agenesis (white arrow) and catheter progression via hemiazygos vein (black arrow).

Source: Elaborated by the authors.

Of the 12 patients, 8 (66.7%) of them were female with an average age of 44.58 (+- 16.2). One patient (8.3%) had a corrected complex heart disease. Arrhythmia induction was possible in 8 (66.7%) of the patients. Atrioventricular node reentry tachycardia (AVNRT) was the most common induced arrhythmia, present in 5 (41.6%) of the patients, followed by atrial tachycardia (AT) and atrioventricular reentry tachycardia (AVRT) (Table 1).

Table 1. Patients characteristics and procedure complications.

Patient	Sex	Age	Arrhythmia	Possibility to reach the heart via Azygos System	Complications
M.A.T.S	F	46	AVNRT	Yes	No
L.L.A	F	49	AVNRT	Yes	No
M.M	M	60		Yes	No
C.M.C.M	F	51	AT	Yes	No
C.C.L.R	F	32	AVNRT	Yes	No
B.P.F	M	64	AVNRT	Yes	No
J.S.S	F	21	-	No	No
J.C.S	F	12	-	Yes	No
R.P.B	F	55	AT	Yes	No
C.G.R	F	40	AVNRT	Yes	DVT and PE
R.A.F	M	62	AVRT	Yes	No
G.O.M.B	M	43	AVRT	No	No

Source: Elaborated by the authors.

The right chambers were able to be catheterized via the azygos system in 10 out of 12 (83.3%) patients (Fig. 2). A single patient (8.3%) experienced a complication following the procedure. A 40-year-old female, with no known comorbidities developed deep vein thrombosis and subsequent pulmonary thromboembolism on the day following the procedure. Anticoagulation was initiated and the patient remained stable. The last patient of our sample had impossibility of catheterization via the azygos system. Consequently, access to the right chambers was achieved through the Right Internal Jugular Vein (Fig. 3).



Figure 2. Venous Angiography of patient number 12 showing occlusion of IVC.

Source: Elaborated by the authors.

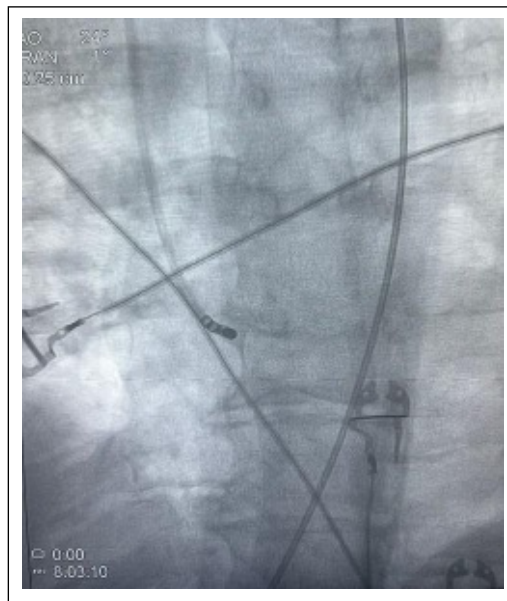


Figure 3. Access to the heart through the Right Internal Jugular Vein in patient number 12.
Source: Elaborated by the authors.

DISCUSSION

Abernethy et al. were the first to describe the congenital variations in the structure of the inferior vena cava (IVC) and its many alternative structures³. The embryonic process of IVC formation consists of the regression and anastomosis of posterior cardinal, subcardinal, and supracardinal veins between the sixth and eighth weeks of gestation. Because of the complexity of their developmental stages, the venae cava may present with many congenital anomalies, such as duplication of the inferior vena cava, agenesis of the inferior vena cava, and the interruption of a certain segment. Given the considerable variation possible in the normal adult structure, congenital deviations of the IVC structure can present significant challenges for many clinical conditions, diagnoses, and procedures⁴.

IVCA has a known relation to deep vein thrombosis (DVT). Tufano et al. observed 31 (0.06%) patients with IVCA in a sample of 50.744 patients with DVT. Their findings indicated an increased risk of IVCA in patients aged < 30 years (odds ratio [OR]: 17.9), or patients with unprovoked DVT (OR: 2.49), proximal DVT (OR: 2.81) or bilateral DVT (OR: 11.5). In our sample, one patient (9%) displayed DVT post-procedure, and no patients had a history of DVT prior to the EPS⁵.

The current literature provides strong evidence of the prevalence of IVCA in samples that are not related to electrophysiological procedures or specific groups. Examples of such samples include pediatric patients with congenital heart disease and patients with DVT. Koc et al. described a sample of 7972 patients who had undergone computed tomography, with a prevalence of 12 (0.15%) patients with IVCA⁶. In their institution, Cordina et al enrolled 80 patients with congenital heart disease who were subjected to EPS, resulting in a prevalence of 10 (13%) patients with an anomaly in their venous systems⁷. As far as we know, the eventuality of accidentally diagnosed IVCA in a general set of patients submitted to EPS has not been closely researched in Brazil and South America.

Al-Sinan et al. published a systematic review of electrophysiological procedures in patients with obstruction of the inferior vena cava, with a sample of 142 patients, and in 54% of them the obstruction was not known before their first procedure. The authors evaluated four primary approaches for EPS in patients with IVCA: the superior approach via superior vena cava, the inferior approach via the Azygos System, the transhepatic approach, and retrograde trans-aortic approach. The first was the most used in their sample, with important benefits such as applicability for all arrhythmic substrates and the reported familiarity with catheter manipulation. It was preferred over the others despite the greater exposure to radiation and increased risk of pneumothorax⁸.

In our sample, the most used approach was the inferior one, using the Azygos System, with low incidence of complications during and post procedure. In the review, atrial fibrillation was the most common arrhythmia, followed by AVNRT. In our study, however, we found a predominance of AVNRT (36%), followed by AT (18%).

The success of transcatheter operations in cardiac electrophysiology depends on access to the heart via the IVC. In cases where femoral approaches are employed, catheter progression difficulties should raise the possibility of undiagnosed IVCA. Venous access is directly impaired in these patients, and the electrophysiologist must know the possible solutions for this problem⁹. In our total sample of EPS executed in the described period, we observed a relatively small yet statistically significant percentage of patients with incidentally diagnosed IVCA during the procedure.

CONCLUSION

As previously documented in our sample, any patient who is scheduled for an electrophysiological study or ablation may come to the electrophysiology laboratory without prior diagnosis of different conditions, including IVCA. The electrophysiologist should be aware that these morphological alterations can occur, aiming to avoid complications of vascular access and catheter handling. In such context, other alternative ways of access to the heart must be considered to ensure a safe and adequate procedure during the correction of cardiac arrhythmias.

CONFLICT OF INTEREST

The authors declare that there are no conflicts of interest.

AUTHOR CONTRIBUTIONS

Conceptualization: Ferreira Filho SF, Leiria TLL; **Methodology:** Ferreira Filho SF, Agnese MAVD, Leiria TLL; **Investigation:** Ferreira Filho SF, Leiria TLL, Kruse M, Lima GG; **Writing Original draft:** Ferreira Filho SF, Agnese MAVD, Schäfer LR; **Review and Editing:** Ferreira Filho SF, Leiria TLL, Kruse M, Lima GG; **Supervision:** Leiria TLL.

DATA AVAILABILITY STATEMENT

The data will be available upon request. All data sets were generated or analyzed in the current study.

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