

# Vein thrombosis in a patient with agenesis of the inferior vena cava

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## SUMMARY

We report a 29 year-old woman with bilateral deep vein thrombosis. CT-scan was performed and an agenesis of the inferior vena cava was found. We provide a brief review of the embryogenesis of the inferior vena cava, and of anomalies in the genesis of this vessel. In young patients with idiopathic deep venous thrombosis the existence of an abnormality in the inferior vena cava should be considered.

**Key words:** Agenesis – Inferior vena cava – Embryogenesis – Deep venous thrombosis

## INTRODUCTION

Agenesis of the inferior vena cava (IVC) is an uncommon disorder and can be misdiagnosed. From an anatomical point of view, an anatomical variation can be considered as "normal" as long as it is within a certain range of structure, form, size and position. Any deviation beyond these limits is considered "anomalous" or "malformed". The anomalies are classified as *major*, when they constitute a common cause of illness or death, or *minor*, when they do not cause any relevant clinical complication (Sañudo et al., 2003). IVC has a complex embryonic development which involves

the formation of three pairs of anastomosis between embryonic veins, and which is complete by the 8th week of pregnancy (Malaki et al., 2012). Congenital abnormalities of the IVC vena cava often yield little clinical importance, and are usually an incidental radiological finding, but some of them may cause morbidity, mainly thromboembolic complications. Thromboembolic complications are often the clinical manifestation of the major anomalies of the IVC. Reported prevalence of the disorder varies greatly, from 0.0005 to 1% of the general population (Castro et al., 2003), although some authors report that the prevalence of anomalies of the IVC rises to 8.7% (Malaki et al., 2012; Yigit et al., 2006). Regression or persistence of the embryonic veins can lead to many variations that can result in different types of anomalies which include the left-sided IVC, double IVC or complete absence of the IVC, among others. This last is a rare condition and its etiology is controversial. We report a case of a 29-year-old woman, placed on oral contraceptive therapy, affected by multiple sclerosis, who presented a bilateral iliofemoral deep vein thrombosis, in whom complete agenesis of the inferior vena cava was documented.

## CASE REPORT

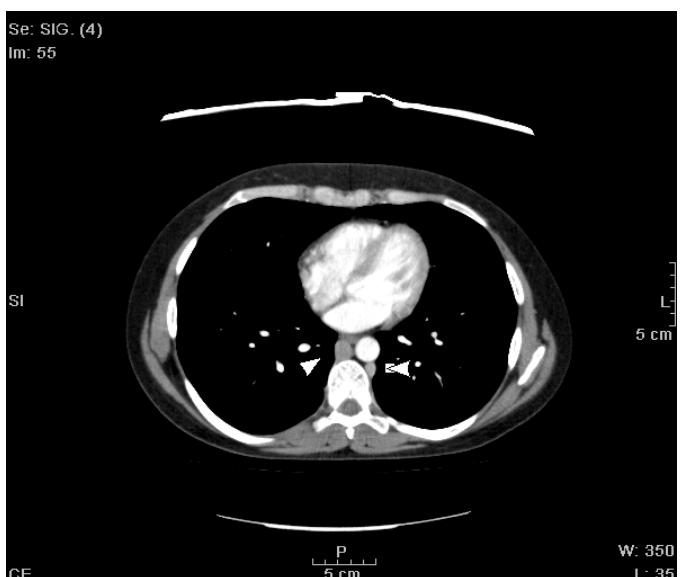
A 29-year-old woman presented with bilateral back pain, loss of strength in the legs and numbness, especially on the right one. Her medical history included: chronic venous insufficiency, oral contraceptive use and multiple sclerosis di-

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**Fig. 1.** A coronal CT reconstruction of the abdomen and pelvis shows the aorta (arrows) with absence of the inferior vena cava and development of the lumbar veins.



agnosed 7 years ago. There was no history of smoking. On physical examination she presented a body temperature of 38.5°C; the lower abdomen was slightly painful at deep palpation, but without signs of peritonism. The right leg was swollen and inflamed. Doppler ultrasonography confirmed the presence of iliofemoral deep vein thrombosis from the external iliac to the popliteal vein. Two days after admission the patient presented pain in left iliac fossa and fever up to 38°C again. Doppler ultrasonography of the left leg was performed showing iliofemoral deep vein thrombosis from the external iliac to the popliteal vein.

A Computed Tomography AngioScan (CT) (Aquileon 64, Toshiba) was performed, showing the absence of the inferior vena cava. Drainage of the lower limbs and kidneys was achieved by large flow lumbar veins, which ultimately connected with the azygos-hemiazygous system. A coronal reconstruction of abdomen and pelvis shows the aorta (arrows) with absence of the inferior vena cava and development of the lumbar veins (Fig. 1); an axial CT section shows increased size of both external iliac veins, which are occupied by hypodense material corresponding to thrombi (Fig. 2). In an axial slice of the chest CT scan we see the aorta and the venous system (azygos vein on the right and left hemiazygous vein). This system is well developed due to the agenesis of the intrahepatic vena cava (Fig. 3). Furthermore, a coronal slice of CT chest shown the azygous vein (less contrast) and the descending aorta (left, with more density by arterial phase contrast) (Fig. 4).

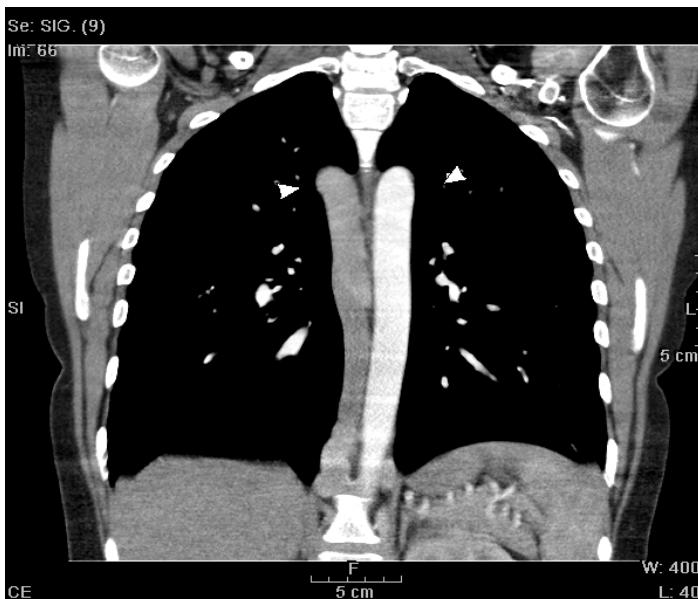
After fibrinolysis the clinical symptoms resolved; the patient was placed on acenocumarol and oral contraceptives were discontinued. She remains asymptomatic one year later.

## DISCUSSION

Embryogenesis of the IVC is a complex process involving the formation, involution and fusion between three longitudinal pairs of veins: the posterior cardinal veins, the subcardinal veins, and the supracardinal veins.

**Fig. 2 (Left-Top).** An axial CT section shows increased size of both external iliac veins which are occupied by hypodense material corresponding to thrombi (arrows).

**Fig. 3 (Left-Bottom).** In an axial CT slice of the chest the aorta and the venous system (azygos vein on the right and left hemiazygous vein, arrows) can be seen. This system is well developed due to the agenesis of the intrahepatic vena cava.



**Fig. 4.** This CT coronal slice of the chest shows the azygous vein (less contrast, horizontal arrow) and the descending aorta (left, with more density by arterial phase contrast, oblique arrow).

These three pairs of primitive veins appear in a chronological order between the fourth and eight weeks of gestation (Bass et al., 2000; Jiménez-Gil et al., 2006; Malaki et al., 2012). The posterior cardinal veins are the first to appear. These veins do not form any part of the definitive IVC, but their abnormal development may lead to a permanent morphological anomaly. By the seventh week of gestation, the right subcardinal vein forms the pre-renal/suprarrenal segment of the IVC. The supracardinal veins predominate by the eighth week and then differentiate into cranial (azygos) and caudal (lumbar) ends. The right supracardinal vein forms the infrarenal segment and the left supracardinal vein regresses (Malaki et al., 2012). The renal segment of the definitive IVC develops from the right supracardinal and subcardinal vein anastomoses. To summarize, the final adult form of the IVC is composed of four segments: the hepatic segment (derived from the vitelline vein); suprarenal segment (from the subcardinal-hepatic vein anastomosis); renal segment and infrarenal segment which are formed as described above.

As noted in the introduction, there are many anomalies in the development of the IVC, and variations can be classified according to the abnormal persistence or abnormal regression of the embryonic veins. These variations include the left-sided IVC, double IVC, azygos and hemiazygos continuation of the IVC, anomalies of the renal segment of the IVC (such as a circumaortic renal vein, left retroaortic renal vein), anomalies of the infrarenal segment of the IVC (which causes the development of a retrocaval ureter), or complete absence of the IVC. The women described in this

case represents an example of agenesis of IVC. There are three entities which may be classified according to Jiménez-Gil as follows (Jiménez-Gil et al., 2006):

1. Failure of the development of the right subcardinal vein, which leads to the absence of the suprarenal IVC. In this case, blood from the infrarenal cava vein returns to the hemiazygos and azygos veins while the hepatic segment of the cava drains into the right atrium. The failure to develop the right subcardinal vein can be associated with cardiac malformations or malformations of other organs (Ramanathan et al., 2001).
2. Failure of the development of the right supracardinal vein, which leads to the absence of the infrarenal cava, with preservation of the suprarenal segment.
3. Total absence of the IVC. In this rare anomaly an alteration in the development of the three pairs of primitive veins takes place. This is the anomaly we found in our patient. Renal veins drained to azygos and hemiazygos, and caudally, to lumbar veins and primitive iliac veins. This bizarre drainage system probably causes venous stasis and propensity to thrombosis, especially in young adults (O'Connor et al., 2011).

The reason why these anomalies occur is unclear. Most authors do not describe the agenesis of the inferior vena cava as an anatomical variation (Milner and Marchan, 1980; Kandpal et al., 2008), and although some reports are controversial (Bass et al., 2000), it is considered that the agenesis of the IVC is the result of an intrauterine or perinatal thrombosis and degeneration of the developing vein (Ramanathan et al., 2001; Jiménez-Gil et al., 2006), as probably happened in this case.

As a summary, we present the case of a young patient with agenesis of the IVC, which was diagnosed during the evaluation of a bilateral iliofemoral deep vein thrombosis. This case constitutes a rare, life-threatening example of a developmental abnormality of the inferior vena cava, whose multiple variants are briefly reviewed in this paper.

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