

CONGENITAL INTERRUPTION OF THE INFERIOR VENA CAVA WITH HEMIAZYGOS CONTINUATION

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Abstract

A case report of congenital interruption of the inferior vena cava with hemiazygos continuation is presented with an explanation of the development of this variation. Its potential consequences are pointed out and the most useful diagnostic methods are suggested. Although the azygos/hemiazygos continuation of the inferior vena cava has been described in the literature several times, we have not found any description of a case completely consistent with our finding. This type of congenital vascular variation has mostly been described in association with other variations or anomalies. However, in the case presented no other abnormalities were evident.

Key words

Inferior vena cava, Hemiazygos vein, Congenital variation, Embryology

INTRODUCTION

Congenital interruption of the inferior vena cava (IVC) with azygos/hemiazygos continuation is a rare developmental variation usually associated with other congenital anomalies, especially cardiac malformations (1–7). According to the latest data, the incidence of this variation is 0.6–2.0% in individuals with congenital heart malformations and less than 0.3% in individuals without any other variations or anomalies (5). Usually, the presence of only this vascular variation causes no clinically evident problems, and it is found incidentally (1–3, 6, 7) either during an X-ray examination or operation for some other reason, or at postmortem during dissection. However, in some cases it can become clinically important.

CASE REPORT

AUTOPSY FINDINGS

During routine anatomical dissection for teaching purposes, an anomalous IVC was found in a 40-year-old woman (Figs. 1, 2). The caudal part of the IVC was formed in the usual way, by the confluence of the two common iliac veins, posterolateral to the right common iliac artery. The IVC passed up along the right side of the abdominal aorta and, at the level of the second lumbar vertebra, it was joined by its tributaries, i.e., the right ovarian, renal and suprarenal veins.

At the level of the intervertebral disc between the first and the second lumbar vertebra, the IVC trunk turned to the left, running horizontally in front of the aorta. The tributaries to the horizontal part

were the left suprarenal and the left ovarian veins that, under normal conditions, drain into the left renal vein. After receiving the next tributary, the short left renal vein, the IVC trunk curved in the dorsocranial direction. Piercing the diaphragm, it continued as the hemiazygos vein. The part of the IVC between the common tributary of the right ovarian, renal and suprarenal veins and the liver was missing.

The anatomy of the azygos and hemiazygos veins was normal, but the whole hemiazygos vein and the terminal part of the azygos vein were enlarged. Also the cranial tributaries to the superior vena cava (SVC) showed dilatations in their distal parts.

The visceral surface of the liver had no relation to the IVC. A large venous trunk emerging from the liver in a cranial direction was formed inside the liver by the confluence of four hepatic veins. Its further course corresponded to the usual course of the suprahepatic part of the IVC including its termination in the right atrium. This venous trunk drained only venous blood from the liver.

No other variations or anomalies were found during the dissection.

DISCUSSION

EMBRYOLOGIC ORIGIN OF VASCULAR VARIATIONS

The development of the venous system makes it possible to explain the venous variation presented here. In the abdominal region, there is initially a symmetrical venous network composed of three pairs of vessels: the postcardinal, the subcardinal and the supracardinal veins with extensive anastomoses between them (*Fig. 3*). The further developmental process can be described in a simplified way as a successive transition of the venous drainage from the initially dominating postcardinal veins to the IVC finally formed on the right side of the body (2, 8). In the left lumbar region, there is a temporary homologue to the IVC, which gradually disappears (8). For the final form of the IVC three parts with their embryonic precursors have been described (8, 9, 10).

The prerenal part, above the level of the renal veins, is formed by the union between the cranial part of the right vitelline vein and the cranial part of the right subcardinal vein through the hepatosubcardinal anastomosis. The tributaries to this part of the IVC are the hepatic veins, right suprarenal and ovarian veins and the left renal vein. The hepatic veins develop from the middle part of the vitelline venous system (11). The right suprarenal vein has already been a tributary to the embryonic right subcardinal vein. The right ovarian vein develops from the caudal part of the right subcardinal vein under the subcardinal anastomosis. The finally formed left renal vein is composed of three parts: the subcardinal anastomosis, the medial part of the left sub-supracardinal anastomosis and the embryonic left renal vein. While the right subcardinal vein plays an important role in the formation of the final venous system, the left subcardinal vein disappears almost completely. Only two small parts of this vein close to the subcardinal anastomosis remain: the short part above the subcardinal anastomosis, where the embryonic left suprarenal vein enters, and the part under the subcardinal anastomosis transformed to the left ovarian vein. The described developmental changes determine the difference in position of the suprarenal and ovarian veins on the left and the right side of the body.

The renal part, at the level of the renal veins, arises from the right subsupracardinal anastomosis. The right renal vein, which develops from its embryonic precursor of the same name, drains into this part.

The postrenal part, under the level of the renal veins, develops from the caudal part of the right supracardinal vein. The iliac veins originate from the iliac anastomosis. According to Gray's anatomy (12), the most caudal part of the right postcardinal vein also takes part in formation of the caudal end of the IVC and the iliac veins.

The major parts of the azygos and hemiazygos veins develop from the right and left supracardinal veins. The horizontal terminal part of the hemiazygos vein, which turns to the right and joins the azygos vein, arises from the supracardinal anastomosis (*Fig. 4*). In relation to the further development, it is possible to distinguish three parts of the supracardinal veins. Their cranial parts give rise to the caudal parts of the azygos and hemiazygos veins. The caudal part of the right supracardinal vein forms the postrenal part of the IVC while the caudal part of the left supracardinal vein and the intermediate parts of both supracardinal veins disappear.

In contrast to this traditional concept, another pair of embryonic veins, the medial sympathetic line veins, has been described as the precursor of the azygos/hemiazygos systems (12). The medial sympathetic line veins run medial to the sympathetic trunk and parallel to the cranial part of the supracardinal veins running lateral to the sympathetic trunk. The cranial ends of the medial sympathetic line veins join the cranial part of the postcardinal veins.

The extensively established and initially symmetrical embryonic venous network, with the complexity of further developmental changes, provides the opportunity for many variations. In our case, only the caudal part of the IVC, under the level of the subcardinal anastomosis, and the cranial part above the hepatosubcardinal anastomosis developed in the usual way (*Fig. 5*). The middle part, which usually develops from the right subcardinal vein and the hepatosubcardinal anastomosis, was missing. Only the right ovarian and suprarenal veins from that part were present. The normally developed caudal part of the IVC was connected to the hemiazygos vein via the subcardinal and the left subsupracardinal anastomoses (*Fig. 5*). By this route, blood from the postrenal part of the IVC "bypassed" its missing part and flowed via the hemiazygos vein and further via the azygos vein into the SVC. All the venous blood, except that from the liver, entered the heart through the SVC. An overload of the hemiazygos vein and the terminal part of the azygos vein caused the enlargement of those two veins. The dilatation of the cranial tributaries to the SVC was a consequence of venostasis.

The formation of a congenital interruption of the IVC with azygos/hemiazygos continuation can be explained in two ways: as a failure in obliteration of the

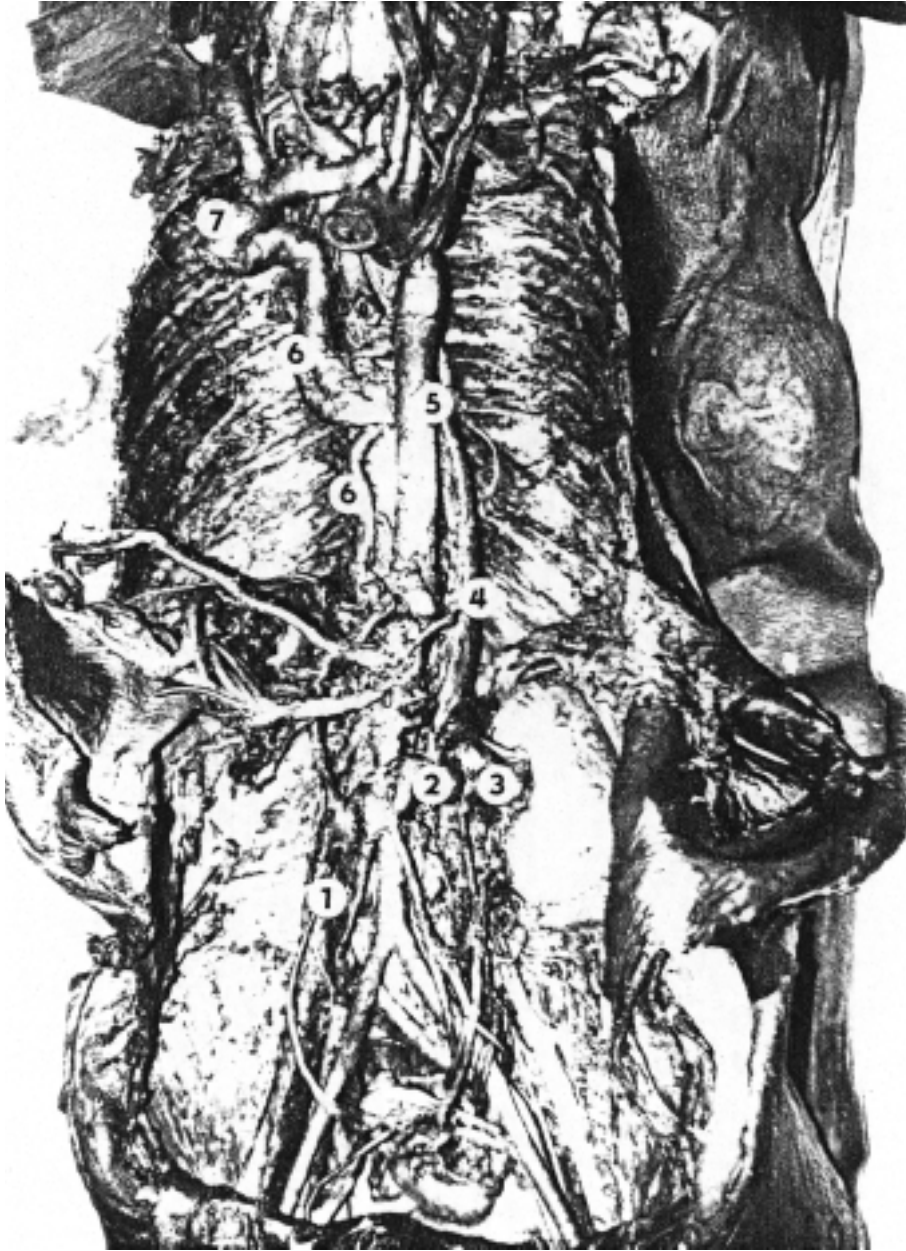


Fig. 1

Anomaly in the anatomy of the inferior vena cava (IVC). 1, IVC; 2, abnormal transversal connection of IVC with the vena hemiazygos; 3, vena renalis sinistra; 4, vena hemiazygos; 5, aorta; 6, vena cava superior.

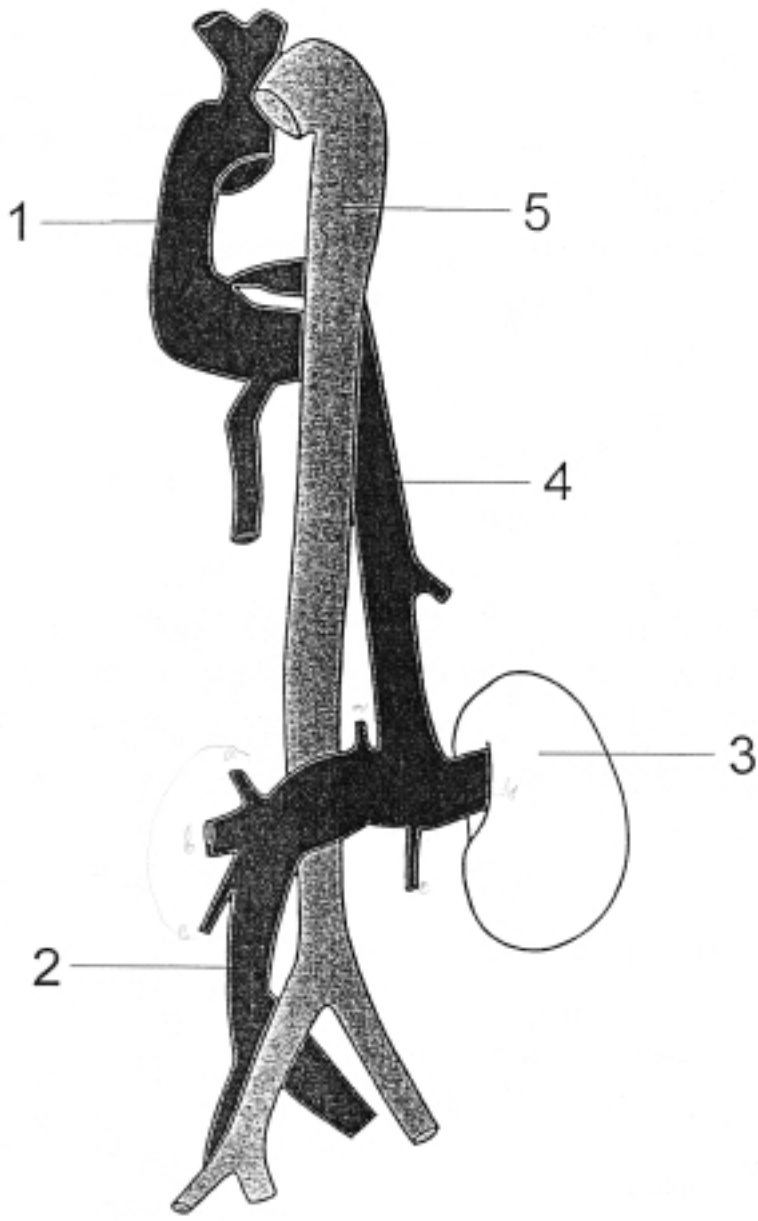


Fig. 2
Schematic drawing of the anomalous anatomy of IVC. 1, vena azygos; 2, IVC; 3, ren sinister;
4, vena hemiazygos; 5, aorta.

intermediate parts of the supracardinal veins (7), or as an absence of establishment of the hepatosubcardinal anastomosis (2, 3, 5). In the latter situation, the persistence of continuity of the right or/and left supracardinal vein is required in order to transport venous blood towards the heart. If the IVC is interrupted in its prerenal part, the venous system may develop in several ways. A direct azygos continuation of the IVC through the persistent intermediate part of the right supracardinal vein is the most common finding (2, 3). When a hemiazygos continuation is present, there are three possible arrangements in the prerenal region (6, 13). First, which occurs most frequently, the enlarged hemiazygos vein drains into the dilated sinus coronarius via the dilated accessory hemiazygos vein and the persistent left SVC (6). Second, which was consistent with our finding, the hemiazygos vein drains into the azygos vein approximately at the level of Th 8–9 and the hemiazygos vein and the terminal part of the azygos vein are enlarged. Third, the hemiazygos vein drains into the right-sided SVC via the dilated accessory hemiazygos, left superior intercostal and left brachiocephalic veins.

The hemiazygos continuation usually occurs in association with a left-sided postrenal segment of the IVC (13). In our case, however, the right-sided postrenal segment was present. No condition similar or identical to our finding has so far been reported in the literature .

CLINICAL IMPLICATIONS

Although the variation described here is usually found incidentally in asymptomatic individuals, it is important in terms of clinical implications.

It may point towards other associated variations or anomalies, especially heart defects, which should be taken into account (1–7).

This variation can cause venous insufficiency of the lower limbs with a potential thromboembolic disease. This should be considered especially in young patients with deep venous thrombosis when no other reason for thromboembolism is evident (1, 4).

The dilated azygos/hemiazygos system shown by chest or abdominal X-ray films can be misinterpreted as a mediastinal or retroperitoneal neoplasm, lymphadenopathy or aortic dissection (2, 5–7, 10). Venostasis due to pathological conditions such as acquired obstruction of the IVC or SVC, the right heart failure, portal hypertension or due to pregnancy can have the same clinical presentation as a azygos/hemiazygos continuation of the IVC (5, 6, 10, 13).

In the case of an azygos/hemiazygos continuation of the IVC, the hepatic veins can drain directly into the right atrium (3, 10). An incidental finding of this condition during venous cannulation for cardiopulmonary bypass complicates the procedure, since no solid IVC trunk for placing the cannula is present. Separate cannulation of the SVC and the right atrium should be used in this case (3).

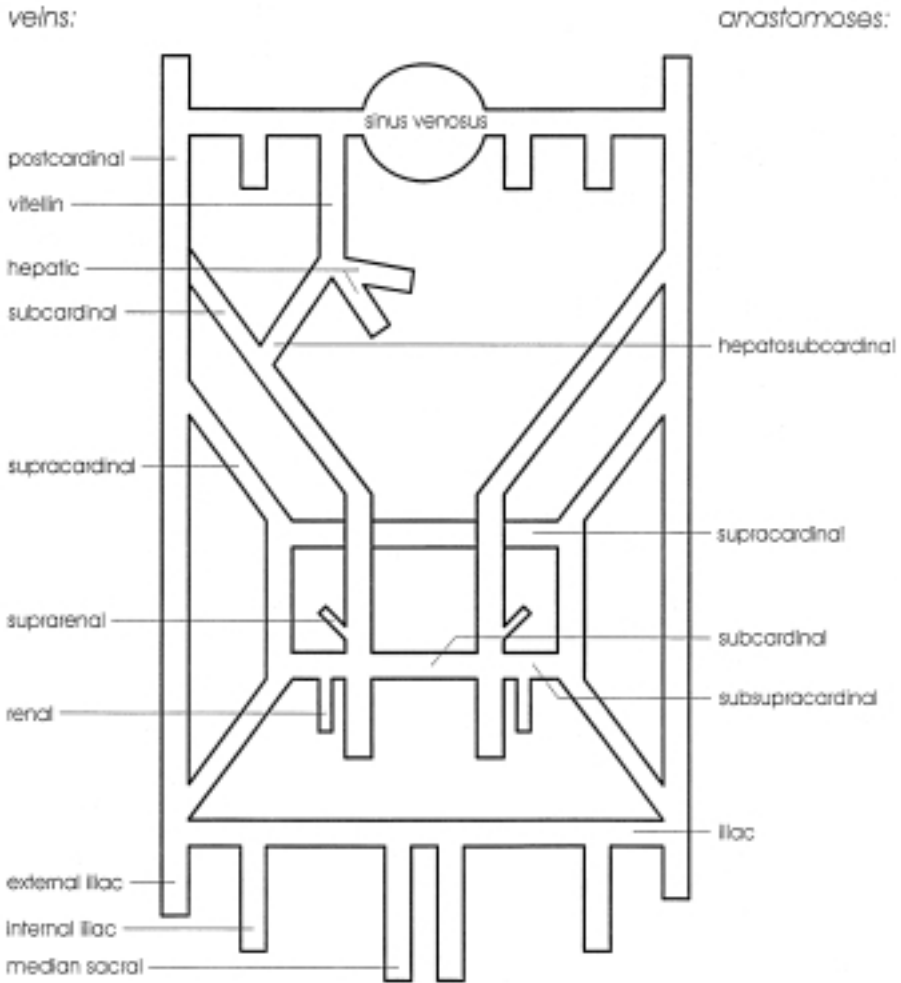


Fig. 3
Schematic drawing showing precursors of the visceral venous system of a human embryo.

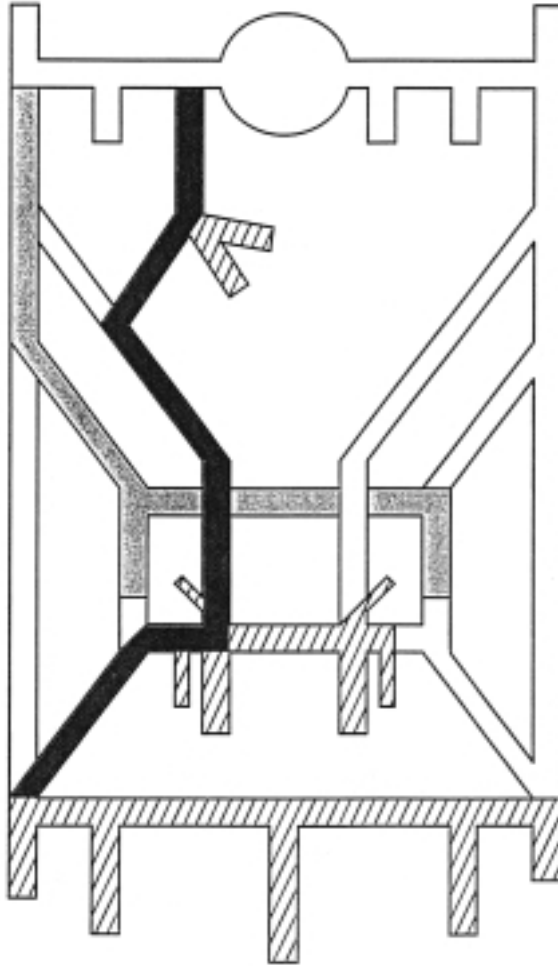


Fig. 4

Schematic drawing of the normal adult human anatomy of the visceral venous system.

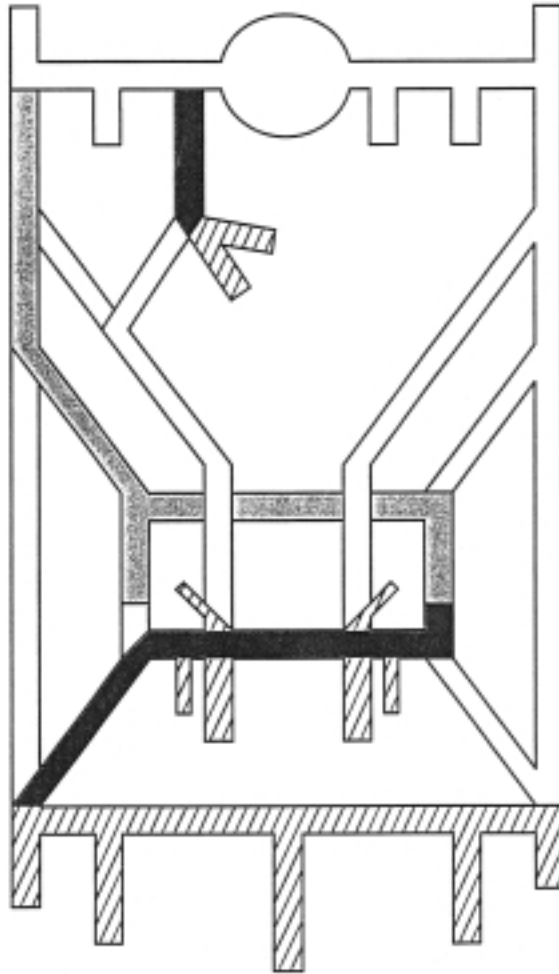






Fig. 5

Schematic drawing showing a congenital anomaly, i.e., the absence of part of IVC, as found in our case.

-  inferior vena cava
-  azygos/hemiazygos veins
-  tributaries of vena cava inferior
-  embryonic precursors of venous system

The presence of this variation affects decisions concerning shunt placement for portal hypertension and ligation or clipping of the IVC for thromboembolism (10).

A fatal outcome after ligation of the azygos vein in a patient with an azygos continuation of the IVC has been reported (14).

Angiography and cardiac catheterisation can be difficult when this variation is present (10).

DIAGNOSIS OF VASCULAR VARIATIONS

Conventional phlebography, intravenous contrast-enhanced CT scan, cross-sectional echocardiography and MRI are the appropriate diagnostic techniques used to diagnose the azygos/hemiazygos continuation of the IVC (1–5, 10, 13). MRI should be preferred as it provides an excellent differentiation between blood vessels and solid structures without using contrast material and ionising radiation (5,10).

Some useful signs have been described to differentiate the azygos/hemiazygos continuation of the IVC from other findings by conventional X-ray examination. The shadow of the dilated azygos or hemiazygos vein has the appearance of a tubular structure contiguous with a shadow paralleling the descending aorta (6). The typical line shadow of the posterior border of the IVC on the lateral chest roentgenogram is absent (3, 6, 10). The width of the normal azygos vein in adults should not exceed 6mm on the frontal chest roentgenogram (10). The size of the shadow of the enlarged azygos/hemiazygos system changes according to the position of the body or by performance of some maneuvers (6, 10); it increases when the patient is a prone or a supine position or during Müller's maneuver (forced inspiration against the closed glottis), and decreases in an upright position or by performance of the Valsalva maneuver (forced expiration against the closed glottis).

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VROZENÉ PŘERUŠENÍ DOLNÍ DUTÉ ŽÍLY S POKRAČOVÁNÍM CESTOU VENA HEMIAZYGOS

S o u h r n

Předkládáme případovou studii vrozeného přerušení dolní duté žíly s pokračováním cestou vena hemiazygos s vysvětlením vývoje této variety. Zdůrazňujeme její možné klinické důsledky a navrhuje nejužitečnější diagnostické metody. Ačkoli pokračování dolní duté žíly cestou vena azygos či hemiazygos bylo popsáno již mnohokrát, nenalezli jsme popis případu, který by se plně shodoval s našim nálezem. Tento typ vrozené cévní variety je většinou popisován ve spojení s dalšími varietami či anomáliemi. Nicméně v předkládaném případě nebyly žádné další abnormality zjevné.

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