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CASE REPORT / ПРИКАЗ БОЛЕСНИКА

# Association of bilateral inferior vena cava with azygos and hemiazygos continuation and aortic coarctation in a child

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**SUMMARY**

**Introduction** Developmental variations of the inferior vena cava (IVC) rarely cause symptoms, and they are usually detected during routine examinations performed for other reasons. The prevalence in the general population is between 0.07% and 8.7%. Various anomalies of the IVC can be seen depending on abnormal regression or abnormal persistence of embryonic veins. They are usually associated with more complex intracardiac and atrioventricular septal defects, partial anomalous pulmonary venous connection, and pulmonary atresia.

**Case outline** We presented an 18-month-old patient with double IVC, IVC interruption, azygos, and hemiazygos continuation associated with aortic coarctation. The vein malformation was discovered during percutaneous balloon angioplasty of the aortic re-coarctation and confirmed by using cross-sectional imaging modalities. Our patient had no symptoms of IVC malformation.

**Conclusion** In clinical practice, double IVC should be suspected in patients with recurrent pulmonary emboli. Another important point in practice is the identification of those anomalies to avoid potential complications of retroperitoneal surgery and cannulation during cardiac surgery.

**Keywords:** double inferior vena cava; interruption of inferior vena cava; hemiazygos continuation; azygos continuation

## INTRODUCTION

Developmental variations of inferior vena cava (IVC) are described with the incidence in the general population between 0.07% and 8.7%, and they are primarily asymptomatic [1]. They are usually associated with more complex intracardiac and spleen lesions (polysplenia/asplenia syndrome), but in some patients, they were detected during routine examinations performed for other reasons [2–6]. The most frequent congenital abnormalities include circumaortic left renal vein in 1.5-8.7% of patients, 2.1% patients had retroaortic left renal vein, 0.2–3% had double IVC, 0.6% had azygos or hemiazygos continuation of IVC, and isolated left-sided IVC was found in 0.2–0.5% [7]. These anomalies reflect the complicated and multisegmental character of the IVC development during embryogenesis [3–6].

We presented a two-year-old child with aortic coarctation, IVC duplication, intrahepatic agenesis and azygos and hemiazygos continuation.

## CASE REPORT

A newborn was admitted to our Institute due to a prenatal diagnosis of the hypoplastic left chamber and aortic arch. At the admission, the echocardiographic finding pointed out a poorly developed left ventricle, bicuspid aortic valve (BAV), aortic coarctation and pulmonary hypertension. Minor atrial and perimembranous ventricular septal defects and intrahepatic IVC agenesis were described. At six days old, aortic coarctation surgery was performed. During the follow-up period, on echocardiography examinations, re-coarctation was observed. At 18 months old, percutaneous balloon angioplasty was performed. During the heart catheterization, IVC malformation with intrahepatic IVC agenesis was observed (Figure 1).

MDCT examination of the abdomen was performed for better evaluation of venous anomalies. The double IVC was formed by the confluence of the ipsilateral right and left common iliac veins, and the duplication was seen on both sides of the aorta (Figure 2A). The right renal vein joins into the right IVC and the left into the left IVC. The IVCs had two communications: at the level of common iliac veins and renal veins with retroaortic course (Figure 2B). The intrahepatic agenesis of the IVC was described, with azygos and hemiazygos continuation of the right and left IVC. The hemiazygos vein joins the azygos vein at the usual place (Figure 2C), and the azygos vein drains into the superior vena cava (SVC). Hepatic veins drain into the right atrium.

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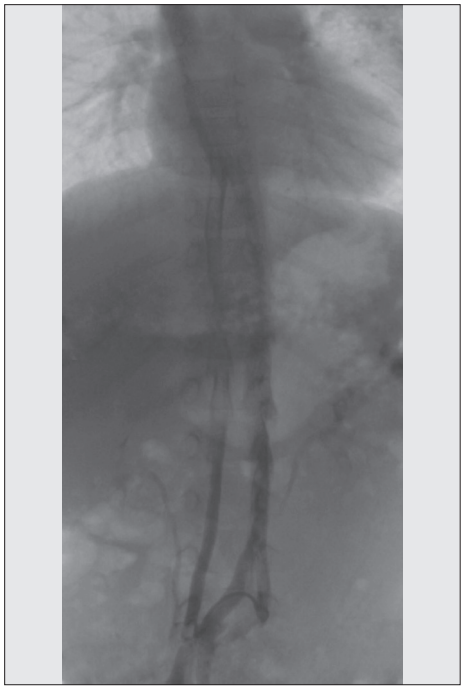

**Figure 1.** Coronary plane – double inferior cava vein with intrahepatic agenesis, hemiazygos and azygos continuation

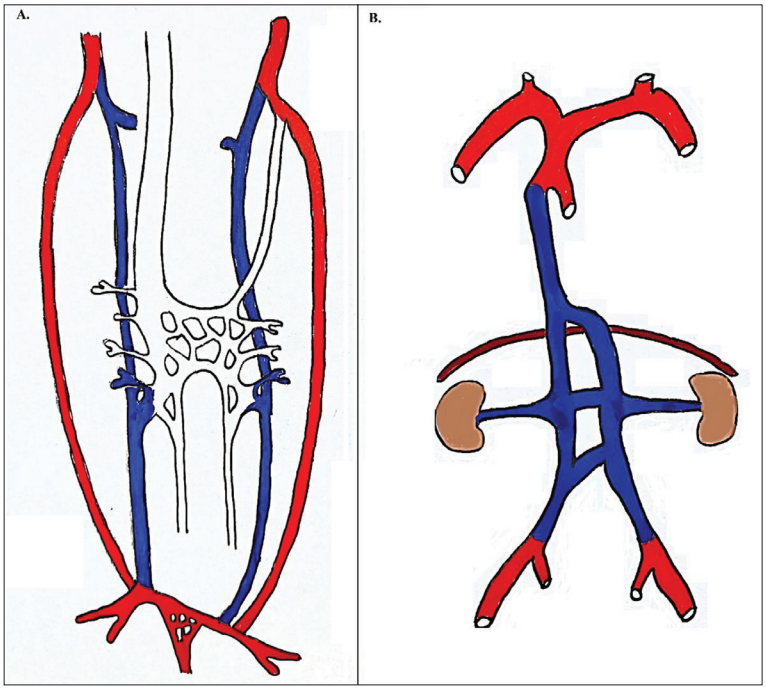


**Figure 3.** A – schematic diagram showing early formation of the posterior cardinal veins (red), subcardinal veins (white), and supracardinal veins (blue); B – interruption of the hepatic segment of double inferior vena cava (right and left) with hemiazygos and azygos continuation; the left inferior vena cava reaches the azygos vein through the hemiazygos vein; the subcardinal part is missing; supracardinal veins (blue) persist bilaterally

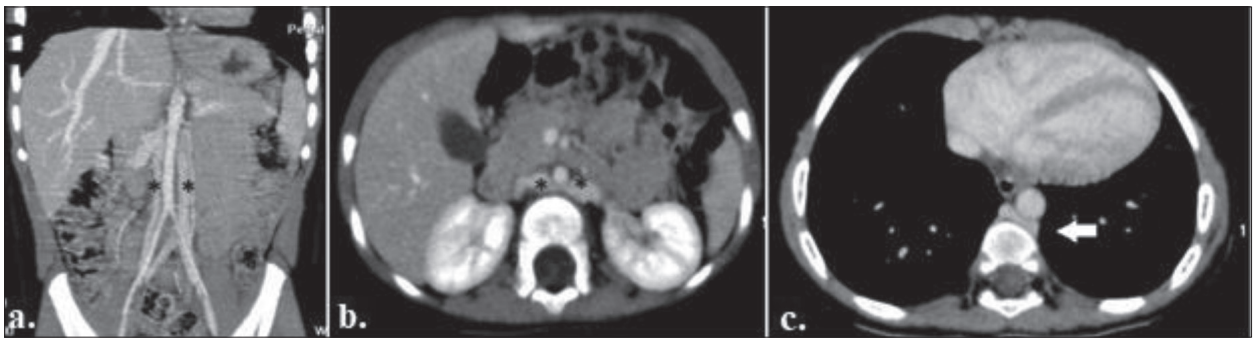


**Figure 2.** a – Inferior vena cava duplication, with interiliac communication from the right common iliac vein (type 2c); b – retroaortic communication at the level of renal veins; c – hemiazygos vein drain into azygos vein at the usual place; asterisk – inferior vena cava; arrow – hemiazygos – azygos drain place

The local ethics committee approved the manuscript. Written informed consent was obtained from the patient's parents to publish any potentially identifiable images or data in this article.

## DISCUSSION

The retroperitoneal venous system undergoes development between the 4th and 8th gestational week. The IVC development is a complicated embryological process including the growth, regression, midline anastomoses, and replacement of three pairs of embryological veins (posterior cardinal, subcardinal, and supra cardinal veins) and the vitelline veins [1–5]. The infrahepatic IVC develops from a set of three paired parallel cardinal veins, while the cranial segment of the right vitelline vein emanates suprahepatic IVC. The retrohepatic segment is derived from an anastomosis between the cranial segment of the right subcardinal vein and the right vitelline vein (Figure 3 A) [2–6]. Varied IVC anomalies can be seen pivoting on abnormal regression or persistence of embryonic veins [3].

The prevalence of bilateral IVCs is 0.2–3%, resulting from the persistence of both right and left supracardinal veins (failure of regression) [1–6]. The most typical setup implies two distinct IVCs originate from each iliac vein, as in our case. However, in most cases, the left IVC terminates at the level of the left renal vein, crossing over to join the right IVC [2–7]. Wolfhard et al. [7] presented a patient with bilateral IVC with azygos continuation and the supradiaphragmatic join of hemiazygos and azygos vein. This patient was like ours, but our patient had two connections between IVCs and associated congenital anomalies (double IVC type 2c [8]).

Interruption of IVC is distinguished by the lack of suprarenal IVC, with a preponderance of 0.6–2.1%. It is the consequence of ineffectual anastomosis of the right subcardinal and hepatic vein, and on the other side atrophy of the right subcardinal vein (maldevelopment). Consequently, owing to agenesis of the hepatic tract of the IVC, the blood circulating in the caudal segments of the vessel reaches the azygos system, partially emanating from the right supra cardinal vein [1–5, 8, 9]. Both azygos and hemiazygos continuation occur, but azygos continuation is more frequent [4]. Although our patient had double IVC with azygos and hemiazygos continuation, the supracardinal system is entirely persistent (Figure 3B). Interruption of IVC is characterized by the absence of suprarenal IVC, with a prevalence of 0.6–2.1%. It is caused by failure of the right subcardinal-hepatic anastomosis and atrophy of the right subcardinal vein (loss of development). Consequently, owing to agenesis of the hepatic tract of the IVC, the blood circulating in the caudal segments of the vessel reaches the azygos system, partially derived from the right supra cardinal vein [1–5, 8, 9]. Both azygos and hemiazygos continuation occur, but azygos continuation is much more common [4]. Although our patient had double IVC with azygos and hemiazygos continuation, the supracardinal system is entirely persistent (Figure 3B).

Hemiazygos and azygos continuation of the IVC with SVC interruption has been commonly related to *situs inversus*, asplenia, polysplenia syndromes, atrioventricular septal defects, partial anomalous pulmonary venous connection, and pulmonary atresia [1, 3, 4, 5, 8]. Our patient had BAV, aortic coarctation and a poorly developed left ventricle; however, vein malformation was detected during balloon angioplasty. A case with BAV, aortic coarctation and uninterrupted left-sided IVC was described in the literature, but this patient had unilateral IVC [10]. Karadeniz et al. [11] presented an adult patient with aortic coarctation, double SVC, a left-hand side IVC with hemiazygos vein continuation, and a right retroaortic renal vein with polysplenia/heterotaxy syndrome.

The prevalence of thromboembolic complications in patients with double IVC is unexplored, but the literature data showed that those patients had an increased incidence of thrombosis. In patients with recurrent embolic disease, IVC filter placement may be impacted by anomalies of the IVC and may require different techniques for each anomaly [1, 2, 7, 10]. Patients with azygos continuation might also develop sick sinus syndrome [1].

Congenital anomalies such as double IVC are rare and require no intervention. This case demonstrates an 18-month-old patient with double IVC, IVC interruption, azygos, and hemiazygos continuation associated with aortic coarctation. The vein malformation was discovered in the catheterization room and confirmed using cross-sectional imaging modalities. Our patient had no symptoms of IVC malformation. In clinical practice, double IVC should be suspected in patients with recurrent pulmonary emboli. Another important point in clinical practice is the identification of those anomalies to avoid potential complications of retroperitoneal surgery and cannulation during cardiac surgery.

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# Повезаност билатералне доње шупље вене са азигос и хемиазигос континуацијом и коарктацијом аорте код детета

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**САЖЕТАК**

**Увод** Развојне аномалије доње шупље вене (ДШВ) ретко изазивају симптоме, а откривају се углавном током рутинских прегледа из других разлога. Преваленција у општој популацији је између 0,07% и 8,7%. Аномалије ДШВ могу бити последица абнормалне регресије или перзистирања ембриоланих вена. Обично су удружени са сложенијим урођеним срчаним манама, као што је атриовентрикуларни септални дефект, парцијалним аномалним уливом плућних вена и атрезијом плућне артерије.

**Приказ болесника** Приказали смо 18-месечног болесника са двоструком ДШВ, прекидом ДШВ, азигос и хемиазигос континуацијом са коарктацијом аорте. Малформација ДШВ је откривена током перкутане балонске ангиопластике рекоарктације аорте, а потврђена је компјутеризованом томографијом. Болесник није имао симптоме малформације ДШВ.

**Закључак** У клиничкој пракси треба сумњати на дуплу ДШВ код болесника са рекурентном плућном емболијом. Такође, важна је идентификација ових аномалија како би се избегле могуће компликације ретроперитонеалне хирургије и канулације током кардиохируршких процедура.

**Кључне речи**: дупла доња шупља вена; прекид доње шупље вене; азигос и хемиазигос континуација