Case report

Duplication of the inferior vena cava: case report and a literature review of anatomical variation

Saad, KR.[1,2]*, Saad, PF.[3], Amorim, CA.[4], Armstrong, D.[5], Soares, BLF.[6], Neves, PCF.[7] and Razuk Filho, A.[8]

[1]Ph.D., Adjunct Professor of Morphology, School of Medicine, Federal University of the Vale do São Francisco – UNIVASF, Rua Jose de Sá Maniçoba, s/n, Centro, CEP 56304-917, Petrolina, PE, Brazil
[2]Member of the Brazilian Society of Anatomy
[3]Ph.D., Vascular Surgeon, Adjunct Professor of Emergency Medicine, School of Medicine, Federal University of the Vale do São Francisco – UNIVASF, Rua Jose de Sá Maniçoba, s/n, Centro,CEP 56304-917, Petrolina, PE, Brazil
[4]Resident in General Surgery, Petrolina Emergency and Trauma Hospital, Federal University of the Vale do São Francisco – UNIVASF, Rua Jose de Sá Maniçoba, s/n, Centro, CEP 56304-917, Petrolina, PE, Brazil
[5]Vascular Surgeon, Auxiliary Professor of Semiotics, Federal University of the Vale do São Francisco – UNIVASF, Rua Jose de Sá Maniçoba, s/n, Centro, CEP 56304-917, Petrolina, PE, Brazil
[6]Vascular Surgeon, Assistant Professor of Emergency Medicine, School of Medicine, Federal University of the Vale do São Francisco – UNIVASF, Rua Jose de Sá Maniçoba, s/n, Centro, CEP 56304-917, Petrolina, PE, Brazil
[7]Ph.D., Orthopedic Surgeon, Adjunct Professor of orthopedics, School of Medicine, Federal University of the Vale do São Francisco – UNIVASF, Rua Jose de Sá Maniçoba, s/n, Centro, CEP 56304-917, Petrolina, PE, Brazil
[8]Vascular Surgeon, Assistant Primary Physician, Assistant Professor, School of Medical Sciences, Irmandade da Santa Casa de Misericórdia de São Paulo, Rua Dr. Cesário Motta Jr., 112, Santa Cecília, CEP 01221-020, São Paulo, SP, Brazil
*E-mail: Karen.ruggeri@univasf.edu.br

Abstract

Anatomical variation in the inferior vena cava can result in misdiagnosis, making a better understanding of such variations crucial. Here we report the case of a 29 year-old male, victim of multiple trauma, who in the course of treatment presented with a pulmonary thromboembolism confirmed by tomography. Given the gravity of the situation and the need for additional surgeries, a decision was made to implant an inferior vena cava filter. During phlebography, prior to implantation of the filter, the duplication of the vena cava was detected and classified as a complete duplication. A review of the literature revealed various anatomical descriptions of duplicated inferior vena cava, the most common of which were incomplete cases showing greater variation in venous contion. All in vivo anatomical descriptions were done via phlebography, demonstrating the value of this test for the diagnosis of anatomical variation in the abdominal veins. While duplication of the inferior vena cava was not the cause of the venous thrombosis in our patient, a detailed phlebography test was needed to both identify the anatomical variation and facilitate the placement of the filter to prevent a new pulmonary thromboembolism.

Keywords: inferior vena cava, pulmonary embolism, anatomical variation, vena cava duplication, phlebography.

1 Introduction

Anatomical variation of the inferior vena cava occurs in 0.4-4% of the population (MAYO, GRAY, LOUIS et al., 1983). The most common variant is duplication of the inferior vena cava (ARTICO, LORENZINI, MANCINI et al., 2004).

While most variants are found via radiology or during post-mortem dissection of cadavers in university anatomy classes, their identification is important in the clinical realm because it may reduce misdiagnoses. It has been reported that these variants can be confused with lymphadenopathy, aortic aneurysm, and retroperitoneal cysts (EVANS, EARIS and CURTIS, 2001; SÉNÉCAIL, LEFEVRE, PERSON et al., 1987), which often leads to unnecessary interventions.

It has also been shown that such anatomical variation can cause illness, in which cases it is considered an anomaly. Wang, Lo, Yu et al. (2005) described a case in which duplication of the inferior vena cava caused obstruction of the left ureter, leading to moderate hydronephrosis in a 21 year-old patient. Wartmann, Kinsella Junior, Tubbs et al. (2011) described an 82 year-old woman whose post-prandial abdominal pain was attributed to the presence of a duplicated inferior vena cava positioned anterior to the aorta, at the level of the celiac trunk, which caused compression of the celiac trunk.

The duplication of the inferior vena cava can also be associated with the recurrence of pulmonary thromboembolism (PTE) when the anatomical variation goes undiagnosed (MILANI, CONSTATINOU, BERZ et al., 2008; ANNE, PALLAPOTHU, HOLMES et al., 2005, JONES, OWEIS, GEDELA et al., 2010; CHEE, CULLIGAN and WATSON, 2001; GAYER, LUBOSHITZ, HERTZ et al., 2003) and surgical procedures to prevent new episodes of PTE treat just one vein. Here we report the case of a patient with duplication of the inferior vena cava and provide an anatomical description of the variation.

We also review the main types of such anatomical variation reported to date in the literature.

2 Case report

C.B.S., male, 29 years old, previously healthy but with a history of smoking, was hospitalized after suffering trauma in a traffic accident in which the right leg was fractured. While awaiting surgical treatment he remained hospitalized with the right leg immobilized by splints and no prophylaxis for venous thromboembolism. On the fifth day of hospitalization, the patient was transferred to our ward for orthopedic surgery. On the day he was admitted, he suddenly presented with dyspnea and intense thoracic pain on the right side. X-rays of the thorax revealed veiling of the lower 1/3 of the right lung. This was interpreted at the time as PTE. The patient was transferred to the intensive care unit (ICU) and treatment with Clexane® (80 mg, 12/12 hours) initiated. A venous EcoDoppler exam of the right lower member confirmed the diagnosis of femoropopliteal Deep Vein Thrombosis (DVT) with a recent appearance, while contrast-enhanced Computed Tomography (CT) confirmed PTE, suggesting extensive infarction of the right pulmonary lobe. The patient remained in the ICU for two days and transferred to the orthopedic ward, treated with anticoagulants, once the dyspnea and thoracic pain improved.

In preparation for surgery to treat the fractured leg we planned to implant the inferior vena cava filter. A prior phlebography (cavography) revealed duplication of the inferior vena cava, with a confluence of the left inferior vena cava with the inferior vena cava immediately after receiving the left renal vein. No venous thrombosis was observed in the inferior cava system. A TULIP (cook) inferior vena cava filter was implanted in the suprarenal position (Figure 1).

The two inferior venae cava originate in the pelvis and run parallel to each other, bordering the aorta. The left inferior vena cava has a slightly smaller caliber until the level of the L2 vertebra, when the left inferior vena cava converges into the inferior right vena cava, crossing the aorta anteriorly, and immediately after receiving the left renal vein. The left inferior vena cava discharges into the right lower vena cava at the level of the L1 vertebra, immediately after the right inferior vena cava has received the right renal vein.

Computed tomography (CT) of the abdomen done after the procedure confirmed duplication of the inferior vena cava (Figure 2).

After receiving a vena cava filter the patient was operated on and later discharged from the hospital 24 hours after surgery.

3 Discussion

The formation of the inferior vena cava is a complex event that has been well described. It is formed between the sixth and eighth weeks of embryonic development via a series of anastomoses and regressions of the following primitive trunk veins: posterior cardinal veins, subcardinal veins, and supracardinal veins. The prerenal segment is derived from the right subcardinal vein, while the renal segment is derived via anastomoses between the subcardinal and supracardinal veins, and the postrenal segment is derived from the right supracardinal vein (MOORE, DALLEY and AGUR, 2011; ROMERO, SALCEDO, HOFMANN et al., 1999).

In a search of the English-language literature using the terms “inferior vena cava” and “duplication”, we found a wide variety of case-reports dating to 1912 that describe cases of duplicated inferior vena cava (GIVENS, 1912). Unfortunately, some authors were unable to describe the venous variants anatomically, because their patients were diagnosed during exploration of various symptoms via imaging tests that do not always provide a complete visualization of the abdominal veins. The most detailed descriptions are typically from anatomical studies of cadavers dissected during medical school anatomy courses. Chart 1 shows the types of variation reported by various authors and their classification under the system Natsis, Apostolidis, Noussios et al. (2010) have proposed for duplication of the inferior vena cava.

The literature strongly suggests that the non-regression of the left supracardinal vein during embryonic development leads to the occurrence of a second abdominal vessel, which is generally positioned to the left of the aorta in adults. As shown by Natsis, Apostolidis, Noussios et al. (2010), however, the configuration of this supernumerary vessel and thus its embryonic origin is quite variable. These authors have proposed that duplications of the inferior vena cava be classified as complete or incomplete. Where complete, the most likely etiological cause is the persistence of the left suprasubcardinal anastomosis, of the postsubcardinal anastomosis, and probably of the intersubcardinal anastomosis, which in turn results in the persistence of the left supracardinal vein. Such a case may also be associated with an absence of iliac anastomosis (posterior distal intercardinal anastomosis). In cases of incomplete duplication, in which the supernumerary vena cava to the left is smaller and sometimes irregular, the likely cause is inadequate regression of the supracardinal vein.

Although it is an invasive procedure, phlebography is indicated by several authors (ZUBAIR, HASHMI and SMAROFF, 2007) as the gold standard for diagnosing duplication of the inferior vena cava. Other authors have suggested that the combination of ultrasound and CT are sufficient for an adequate diagnosis. However, Evans, Earis, and Curtis (2001) reported misdiagnosis and unnecessary intervention in a patient following the use of these tests. In our case, phlebography was done in preparation for implanting a filter in the vena cava, and when the filter was implanted it was easy to diagnose the variant, make a detailed in vivo description, and otherwise improve our understanding of this type of anatomical variation. Phlebography was used for every in vivo anatomical description reported in the literature.

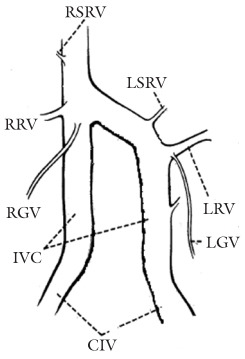
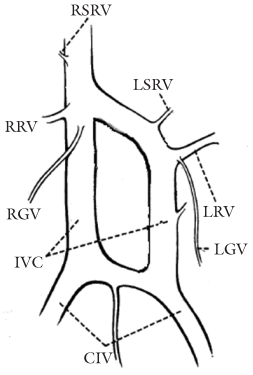
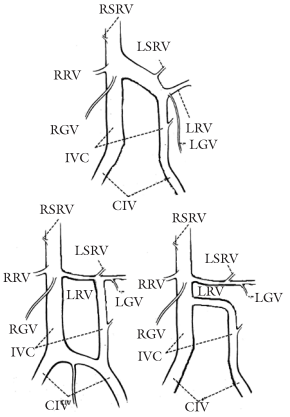
In 33% of the published descriptions we surveyed and in our case, the left common iliac vein gives rise to the supernumerary vena cava, without, however, any anastomosis between the common iliac veins. This may have facilitated our diagnosis via phlebography. However, 41.7% of published descriptions refer to incomplete duplications, which have a greater variety of configurations and thus appear more difficult to diagnose.

Not surprisingly, phlebography is widely preferred over other imaging tests in the presence of symptoms associated with deep vein thrombosis. We found eight studies in the

English-language literature (LEONG, OISIN, BARRY et al., 2010; MILANI, CONSTATINOU, BERZ et al., 2008; HASHMI and SMAROFF, 2007; ANNE, PALLAPOTHU, HOLMES et al., 2005; SÉNÉCAIL, LEFEVRE, PERSON et al., 1987; REINUS and GUTIERREZ, 1986; RICHARDSON, KINARD and LEVESQUE, 1983, WAGNER and MARK, 1969) that reported cases in which a diagnosis of duplicated vena cava was made during attempts to prevent PTE. Milani, Constatinou, Berz et al. (2008) argued that patients with duplication of the inferior vena cava may have a propensity for developing thrombosis. They described two clinical cases in which the supernumerary vein to the left was irregular and had a smaller caliber. They also noted a fifth case study reported by Sénécail, Lefevre, Person et al. (1987) in which complex anatomical variation was apparent in the abdominal veins and thrombosis present in the lower limbs. In all the other published studies and in our own case, we observed no relationship between DVT and duplication of the inferior vena cava. In these studies DVT appears to be associated with other pre-existing illnesses, such as lower limb fractures, cancer, the use of hormonal contraceptives, prior abdominal surgeries in elderly patients, genetic predisposition to thrombosis, and others.

On the other hand, Gayer, Luboshitz, Hertz et al. (2003) highlight the possibility of an association between DVT and anomalies of the inferior vena cava other than simple duplication. The authors reviewed nine cases in which DVT coincided with anomalies in which part or all of the superior

Chart 1. Cases of inferior vena cava duplication described in the literature and classification.

Study	Descriptive method	Schematic illustration of venous anatomy	Classification (NATSIS, APOSTOLIDIS, NOUSSIOS et al., 2010)
Our case	Phlebography		Complete (type I or II)
Leong, Oisin, Barry et al. (2010)	Phlebography		
Natsis, Apostolidis, Noussios et al. (2010)	Cadaver dissection		
Richardson, Kinard and Levesque (1983)	USG and Phlebography		
Sénécail, Lefevre, Person et al. (1987) Case 7	Cadaver dissection		
Sénécail, Lefevre, Person et al. (1987) Case 4	Scanography and Phlebography		Complete (type I or II) with iliac anastomosis
Wagner and Mark (1969)	Phlebography		
Lewis (1992)	Cadaver dissection		
Hashmi and Smaroff (2007)	Phlebography and TC		Incomplete (type III)
Milani, Constatinou, Berz et al. (2008)	Phlebography		
Sénécail, Lefevre, Person et al. (1987) Case 2	Phlebography		
Sénécail, Lefevre, Person et al. (1987) Case 8	Cadaver dissection		
Bay, Tay and Yee-Kong (1997)	Cadaver dissection		

RSRV – Right supra-renal vein; LSRV – Left supra-renal vein; RRV – Right renal vein; LRV – Left renal vein; RGV - Right gonadal vein; LGV – Left gonadal vein; IVC – Inferior vena cava; CIV – Common iliac vein.

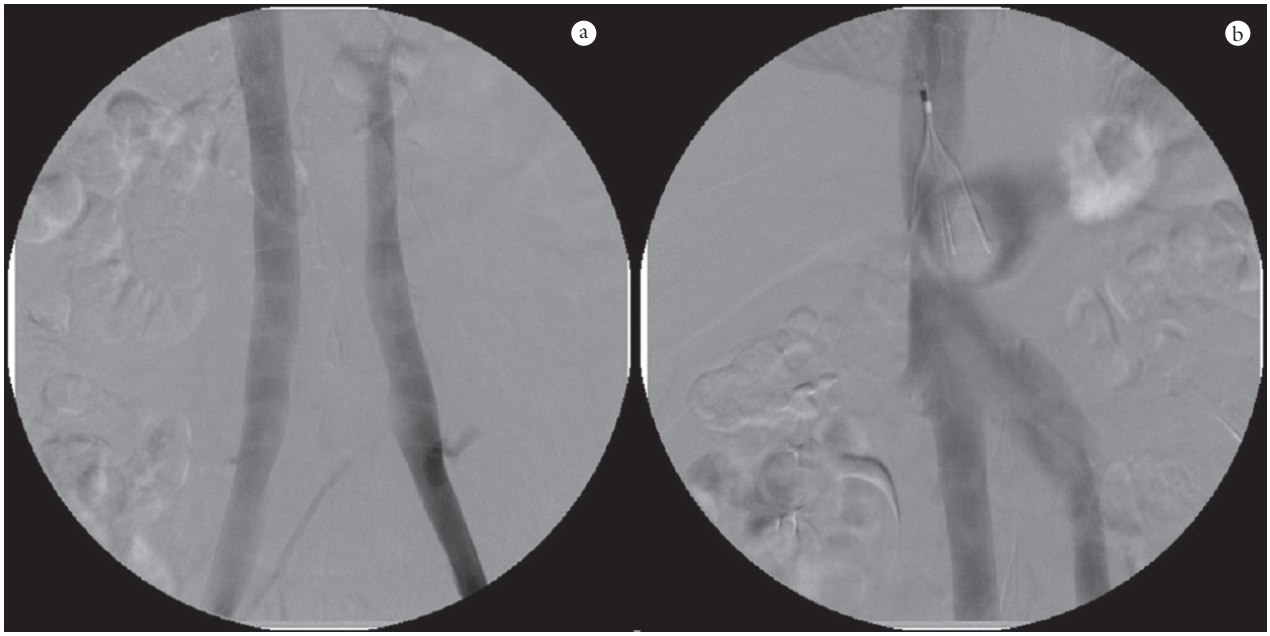


Figure 1. a) Phlebography demonstrating duplication of inferior vena cava. b) A Inferior vena cava filter was implanted in the suprarenal position (contrast was injected in the both femoral veins for this exam).

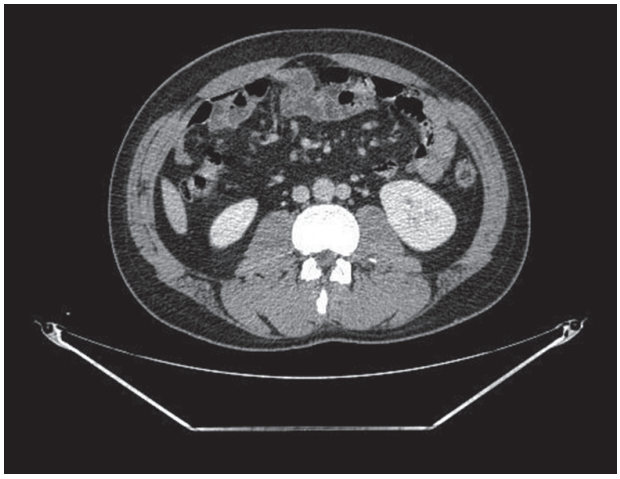


Figure 2. Abdominal CT demonstrating duplication of inferior vena cava.

or inferior vena cava was absent. In these cases, venous return was via the azygos or collateral system, which caused stasis and thereby increased the probability of thrombosis.

The investigation of the abdominal vein anatomy of our patient allowed us to select the most appropriate site to place a filter in the inferior vena cava in order to prevent a new PTE. Our decision to place a single filter at the confluence of the two venae cavae and above the renal veins was largely determined by conditions in our ward, as in the case reported by Zubair, Hashmi and Smaroff (2007). Other authors (LEONG, OISIN, BARRY et al., 2010, MOUBARAK, SCHLEICH and DAUBERT, 2009, MALGOR, OROPALLO, WOOD et al., 2011) chose to implant two filters, one in each vena cava. Both procedures have been considered effective in preventing PTE.

4 Conclusion

Anatomical descriptions in the literature reveal great variation in the duplication of the inferior vena cava, and a higher incidence of 'incomplete' duplication. Understanding this variation is important in the clinical realm, both for avoiding misdiagnosis of retroperitoneal illnesses and for facilitating procedures to prevent PTE in patients with DVT who cannot be medicated with anticoagulants. Anatomical variation of this kind should be studied via imaging tests such as phlebography that are capable of both identifying such variants and revealing the anatomy of the relevant vessels.

References

ANNE, N., PALLAPOTHU, R., HOLMES, R. and JOHNSON, MD. Inferior vena cava duplication and deep venous thrombosis: case report and review of literature. *Annals of Vascular Surgery*, 2005, vol. 19, p. 740-743. PMid:16034513. http://dx.doi.org/10.1007/s10016-005-5674-6

ARTICO, M., LORENZINI, D., MANCINI, P., GOBBI, P., CARLOIA, S. and DAVID, V. Radiological evidence of anatomical variation of the inferior vena cava. *Surgical and Radiologic Anatomy*, 2004, vol. 26, p. 153-6. PMid:14600789. http://dx.doi.org/10.1007/s00276-003-0192-0

BAY, BH., TAY, SSW. and YEE-KONG, NG. Duplication of inferior vena cava with right renal hypoplasia. *Congenital Anomalies*, 1997, vol. 37, p. 149-155.

CHEE, YL., CULLIGAN, DJ. and WATSON HG. Inferior venacava malformation as a risk factor for deep venous thrombosis in the yang. *British Journal of Haematology*, 2001, vol. 114, p. 878-880. PMid:11564079. http://dx.doi.org/10.1046/j.1365-2141.2001.03025.x

EVANS, JC., EARIS, J. and CURTIS J. Thrombosed double inferior vena cava mimicking paraortic lymphadenopathy. *The British Journal of Radiology*, 2001, vol. 74, p. 192-194. PMid:11718395.

GAYER, G., LUBOSHITZ, J., HERTZ, M., ZISSIN, R., THALER, M., LUBETSKY, A., BASS, A., KORAT, A. and APTER, S. Congenital anomalies of the inferior vena cava reveled on CT in patients with deep vein thrombosis. *American Journal of Roentgenology*, 2003, vol. 180, p. 729-732. PMid:12591684.

GIVENS, MH. Duplication of the inferior vena cava in man. *The Anatomical Record*, 1912, vol. 6, p. 475-486. http://dx.doi.org/10.1002/ar.1090061203

HASHMI, ZA. and SMAROFF, GG. Dual inferior vena cava: two inferior vena cava filters. *The Annals of Thoracic Surgery*, 2007, vol. 84, p. 661-663. PMid:17643661. http://dx.doi.org/10.1016/j.athoracsur.2007.03.076

JONES, O., OWEIS, D., GEDELA, VR. and MASON, PF. *Duplicated inferior vena cava with associated ilio-femoral deep vein thrombosis*. BMJ Case Reports, 2010. Available from: <http://casereports.bmj.com/content/2010/bcr.03.2010.2838.full.pdf>.

LEONG, S., OISIN, F., BARRY, JE., MAHER, MM. and BOGUE, CO. Bilateral Inferior vena cava filter insertion in a patient with duplication of the infrarenal vena cava. *Irish Journal of Medical Science,* 2010. Epub ahead of print.

LEWIS, SJ. Observations on a double inferior vena cava: case report with a literature review. *Clinical Anatomy*, 1992, vol. 5, p. 227-233. http://dx.doi.org/10.1002/ca.980050307

MALGOR, RD., OROPALLO, A., WOOD, E., NATAN, K. and LABROPOULOS N. Filter placement for duplicated cava. *Vascular and Endovascular Surgery*, 2011, vol. 45, p. 269-73. PMid:21278173.

MAYO, J., GRAY, R., LOUIS, ES., GROSSMAN, H., MC LOUGHLIN, M. and WISE, D. Anomalies of inferior vena cava. *American Journal of Roentgenology*, 1983, vol. 140, p. 339-45. PMid:6336872.

MILANI, C., CONSTATINOU, M., BERZ, D., BUTERA, JN. and COLVIN, GA. Left sided inferior vena cava duplication and venous thromboembolism: case report and review of literature. *Journal of Hematology & Oncology*, 2008, vol. 1, p. 24-28.

MOORE, KL., DALLEY, AF. and AGUR, AMR. *Anatomia orientada para a clínica*. Rio de Janeiro: Gen-Guanabara Koogan, 2011. 1136 p.

MOUBARAK, G., SCHLEICH, JM. and DAUBERT, JC. Long-term efficacy of two vena cava filter implants for congenital duplicated inferior vena cava. *Archives of Cardiovascular Disease*, 2009, vol. 102, p. 77-78. PMid:19233113. http://dx.doi.org/10.1016/j.acvd.2008.06.009

NATSIS, K., APOSTOLIDIS, S., NOUSSIOS, G., PAPATHANASIOU, E., KYRIAZIDOU, A. and VIZAS V. Duplication of the inferior vena cava: anatomy, embryology and classification proposal. *Anatomical Science International*, 2010, vol. 85, p. 56-60. PMid:19330283. http://dx.doi.org/10.1007/s12565-009-0036-z

REINUS, WR. and GUTIERREZ, FR. Duplication of the inferior vena cava in thromboembolic disease. *Chest*, 1986, vol. 90, p. 916-918. PMid:3780336. http://dx.doi.org/10.1378/chest.90.6.916

RICHARDSON, ML., KINARD, RE. and LEVESQUE, PH. Inferior vena cava duplication: Demonstration by sonography. *Journal of Clinical Ultrasound*, 1983, vol. 11, p. 225-228. PMid:6408135. http://dx.doi.org/10.1002/jcu.1870110411

ROMERO, MEC., SALCEDO, PG., HOFMANN, PG., DORADO, AMD. and ORTIZ, PGT. *Embriologia - biologia do desenvolvimento*. São Paulo: Iátria, 1999.

SÉNÉCAIL, B., LEFEVRE, C., PERSON, H. and MERIOT, P. Radiologic anatomy of duplication of the inferior vena cava: A trap in abdominal imaging. A report of 8 cases. *Surgical and Radiologic Anatomy*, 1987, vol. 9, p. 151-157. PMid:3120333. http://dx.doi.org/10.1007/BF02086600

WAGNER, M. and MARK, L. Duplication of the inferior vena cava and Its role in recurrent pulmonary emboli. *The Journal of the American Medical Association*, 1969, vol. 209, p. 108-109. http://dx.doi.org/10.1001/jama.1969.03160140064017

WANG, LT., LO, HC., YU, DS., SUN, GH., WU, CC. and FONG, CJ. Ureteral obstruction caused by a duplicated anomaly of inferior vena cava. *International Journal of Urology*, 2005, vol. 12, p. 842-84. PMid:16201983.

WARTMANN, CT., KINSELLA JUNIOR, CR., TUBBS, RS. and LOUKAS, M. A rare case of a complete left inferior vena cava associated with the symptoms of Dunbar syndrome. *Clinical Anatomy*, 2011, vol. 24, n. 2, p. 262-5. http://dx.doi.org/10.1002/ca.21078

ZUBAIR, A., HASHMI, ZA. and SMAROFF, GG. Dual inferior vena cava: two inferior vena cava filters. *The Annals of Thoracic Surgery*, 2007, vol. 84, p. 661-663. PMid:17643661. http://dx.doi.org/10.1016/j.athoracsur.2007.03.076

Received January 7, 2012
Accepted March 19, 2012