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SHORT REPORT

Congenital Anomalies of the Inferior Vena Cava and their Clinical Manifestation

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Congenital anomalies of IVC are rare. They are seen more often in young males. They are latent for a long time. Peripheral venous thrombosis or CVI are often the first symptoms of a congenital IVC anomaly. We present 5 patients aged 20 to 43 with congenital anomalies of the IVC. The diagnosis and the level of hypoplasia have been determined by compression ultrasonography with color Doppler assessment, spiral computer tomography, pelvic phlebography and retrograde cavagraphy. In three out of five patients the disease presented as a deep venous thrombosis, in two by temperature rise, chills and subsequent edema of both legs. Two patients had hypoplasia of the infrarenal segment of the IVC. Two others had abnormal of infra-, renal and suprarenal regions of the IVC and one had almost complete vena cava aplasia. In case of DVT or CVI, especially in young males, a potential IVC abnormality should be excluded by ultrasonography of the infra-, renal and suprarenal areas of the IVC. In case of recognized abnormalities a spiral CT scan is indicated. Treatment should comprise vasotonic drugs, elastic compression stockings and use of anticoagulants in cases with peripheral thrombosis.

Keywords: Congenital anomaly; Inferior vena cava; Helical computer tomography.

Introduction

Hypo- and aplasia of the inferior vena cava is a relatively rare congenital anomaly. Despite being associated with deep vein thrombosis (DVT) or chronic venous insufficiency (CVI), the correct diagnosis is often made late during the assessment of patients with suspected peripheral venous thrombosis. It is sometimes suspected on a chest X-ray examination due to an enlargement of mediastinal shadow caused by azygos veindilatation.[1,2]

Congenital anomalies of the IVC are a problem due to development of CVI in such patients. The interest shown to it has increased due to improved diagnostic methods, such as compression ultrasonography with color Doppler assessment (US) and helical computer tomography (HCT).[3,4]

Differentiating congenital pathology of the IVC from acquired diseases such as thrombosis or compression by a tumor is important in choosing a treatment method and further prognosis.

We aim to present our results of investigation and treatment of patients with congenital IVC anomalies.

Materials and Methods

We present 5 cases of patients with congenital IVC abnormalities (Table 1). We performed an US of veins of lower extremities, pelvis and retroperitoneal space, a HCT scan of the abdominal cavity and chest, pelvic phlebography and retrograde cavagraphy, echocardiography and ultrasound investigation of the abdominal cavity in our patients in order to determine the

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Table 1. Basic clinical data

Age/ sex	DVT: primary/ relapse	Temperature rise, fever	Aplasia extension	Blood flow collateral tracts	Other abnormalities	Therapy type	Prognosis
20/m	Primary	–	Infrarenal, renal, partly suprarenal	Ascending lumbar veins, azygos and hemiazygos veins, vertebral veins, paravertebral plexus, left testicular, suprarenal and phrenic veins, veins of abdominal wall	–	Anticoagulants, phlebotonics, elastic compression	Recovery of working abilities
26/m	— // —	+	Infrarenal, renal, suprarenal, major part of retrohepatic	— // —	Pyelocaliceal duplication of the left kidney	— // —	Invalidi-zation
20/m	–	+	Infrarenal	Left testicular vein, ascending lumbar veins, paravertebral plexus, veins of abdominal wall	–	Phlebotonics, elastic compression	Recovery of working abilities
32/m	Relapse	+	Infrarenal, renal, partly suprarenal	Ascending lumbar veins, azygos and hemiazygos veins, vertebral veins, paravertebral plexus, left testicular, suprarenal and phrenic veins, veins of abdominal wall	–	Anticoagulants, phlebotonics, elastic compression	— // —
43/m	–	+	Infrarenal	Left testicular vein, ascending lumbar veins, vertebral veins, paravertebral plexus, veins of abdominal wall	Pulmonary artery stenosis, atrial septal defect	Phlebotonics, elastic compression	— // —

character and prevalence of the pathological process in the IVC and to identify other anomalies.

US was conducted by the same operator using an Acuson 128 XP 10 Scanner (Acuson Inc. Mountain View) with a 5–7 MHz probe in low-flow setting (minimum measurable velocity 1 cm/s). All veins were examined with the individual in a 45° sitting position, the knee flexed and the feet resting on a footstool. The veins examined were the posterior tibial veins, perforators, the short saphenous vein, the popliteal vein, the long saphenous vein, the superficial femoral vein, the common femoral vein the internal iliac vein, the external iliac vein, the common iliac vein, the inferior vena cava.

Venous valvular incompetence was examined using the Valsalva test and manual calf compression. The increased abdominal pressure during the Valsalva manoeuvre provokes reversed (red color) blood flow if the venous valve is incompetent. Calf compression and decompression simulates the calf muscle pump. Valvular incompetence was shown by reversed (red color) blood flow during decompression. Blood flow velocity was registered also.

HCT scanning was performed with a helical scanner (HiSpeed; General Electric Medical Systems) with the patient in the supine position. Nonenhanced CT images were obtained from the pelvis to the Th_4 at a pitch of 1:1 using a slice thickness of 6 mm. For contrast-enhanced HCT, 100 ml of 350 mg I/ml iohexol (Omnipaque; Nycomed) was automatically injected with use of a power injector (Envision CT; Medrad) at a rate of 4.35 ml/sec through the antecubital brachial vein. After a 70-second and 5 minutes delay, the IVC system was imaged from the pelvis to the superior vena cava junction by means of a collimation of 6 mm and a 2.5–3.2-mm reconstruction interval at a pitch of 1:1. With use of three-dimensional reconstructions, including multiplanar reformation and volume rendering, the presence or absence of hypoplasia were evaluated.

For pelvic phlebography both femoral veins were catheterized just below the groin, contrast medium (Omnipaque; Nycomed) was injected and X-rays of the pelvis were made.

Retrograde cavagraphy was carried out in one patient with the patient in the supine position and a catheter was inserted through the superior vena cava tributaries and the right atrium into the IVC. Contrast was entered and X-ray examination of the chest and abdomen were performed.

The treatment of patients with congenital anomalies of IVC included vasotonic drugs (500 mg of diosmin – Daflon 500, Les Laboratoires Servier) two times per day and elastic compression. In case of

DVT we used of IV heparin for 5 days followed by warfarin for life.

The follow-up lasted from 0.5 to 3.5 years.

Results

All patients examined were male, aged from 20 to 43 years old. The medical history revealed that the disease first presented at 17 till 39 years of age respectively. One of the patients underwent a heart surgery for congenital malformation, pulmonary artery stenosis and atrial septal defect, at the age of 5. An other one suffered from varicose disease of the lower extremities from the age of 15.

In 2 out of 5 patients the pathology presented by symptoms of the right iliofemoral thrombosis, and in 2 others by a temperature rise, chills and subsequent edema of both legs, and one presented with symptoms of deep vein thrombosis of the right calf. Thus, in 3 out of 5 patients the disease presented with the clinical picture of a DVT.

Retrospective analysis of the US data of the patients with peripheral vein thrombosis in acute stage of disease showed thrombus in one or both common iliac veins. It extended to the superficial femoral and popliteal veins, and in one patient to the posterior tibial vein.

In one out of three patients the development of thrombosis was contributed to by surgery for a left-sided ureterohydronephrosis. In one other patient a cause of DVT was a lower leg fracture and in the third patient no explicit predisposing risk factors were identified.

All patients developed dilated superficial veins, tributaries of the great saphenous vein (vv. circumflexa ilium superficialis, epigastrica superficilalis, pudenda externa) on the anterior abdominal wall, mainly on the right side, in about three months up to 1 year after the acute stage of DVT. In three patients the CVI was complicated by the development of an ulcer of the lower legs and in one of the patients in both legs after 2 to 2.5 years.

US showed in all patients changes in the superficial and deep veins of lower extremities, as well as the IVC abnormalities of different levels and length.

US demonstrated marked transformation of the superficial veins, especially of the great saphenous vein, short saphenous vein and its tributaries. The common femoral veins with thickened walls were seen in all patients, two of them demonstrating hyperechoic masses in the vessel lumen at this level, with blood flow only partly mapped.

Stenoses, the diameter reduced to 5–8 mm, were seen in the external and common iliac veins. In two cases partial organized thrombotic masses were visualized in the lumen. The internal iliac veins were tortuous, distended to 13–15 mm, with retrograde blood flow.

The IVC was partially invisible on US of the retroperitoneal space in all patients. In 2 patients no blood flow could be determined in the infrarenal segment and in 3 patients in the infra-, renal and suprarenal segments. Dilated ascending lumbar veins were seen. In 3 patients dilated renal veins (18–20 mm in diameter, linear blood flow velocity up to 10 cm/sec) and their segmental tributaries in the portals were revealed. Also ultrasound showed duplication of the pyelocaliceal system of the left kidney in one of the patients.

In all patients US demonstrated dilatation of the anterior abdominal wall veins with the diameter of the superficial epigastric vein measuring 9 mm in one of the patients.

Pelvic phlebography showed in all patients postthrombotic stenosis of external and common iliac veins with obscure margins (Fig. 1), as well as dilated internal iliac veins. Retrograde cavagraphy was performed in one patient for clear illustration of retrohepatic IVC area and its data showed the constricted (7.7–9.4 mm in diameter) part of retrohepatic segment of IVC (Fig. 2).

Echocardiography did not identify heart- or vessels-related pathology in any of the patients.

The most thorough information about the character of the congenital IVC pathology was obtained from contrast-enhanced HCT. These methods helped to

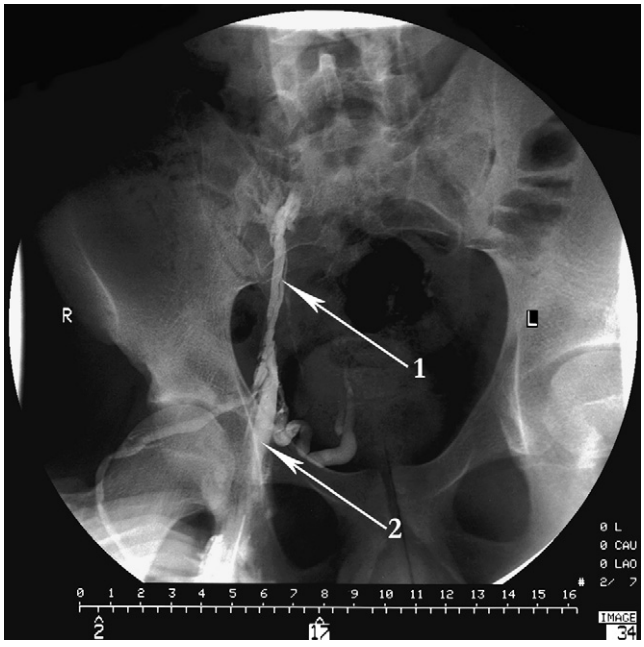

Fig. 1. Pelvic phlebography. Stricture formation of the right iliac vein. Common (1) and external (2) iliac vein.

clearly visualize the level and extension of the lesion (Fig. 3), and the routes of the collateral drainage (Fig. 4) and to diagnose concomitant congenital pathology such as duplication of the pyelocaliceal system in 1 patient.

The infrarenal region of the IVC was hypoplastic in two patients, in two others hypoplasia of infra-, renal and suprarenal regions of the IVC was documented, and one had an almost complete aplasia of the IVC except for a small part of the suprahepatic area where hepatic veins and the right superior renal vein drained into.

The most important collateral vessels were the ascending lumbar veins, azygos and hemiazygos veins (its diameter was 15–18 mm), the paravertebral venous plexus, vertebral veins, left testicular, suprarenal and phrenic veins.

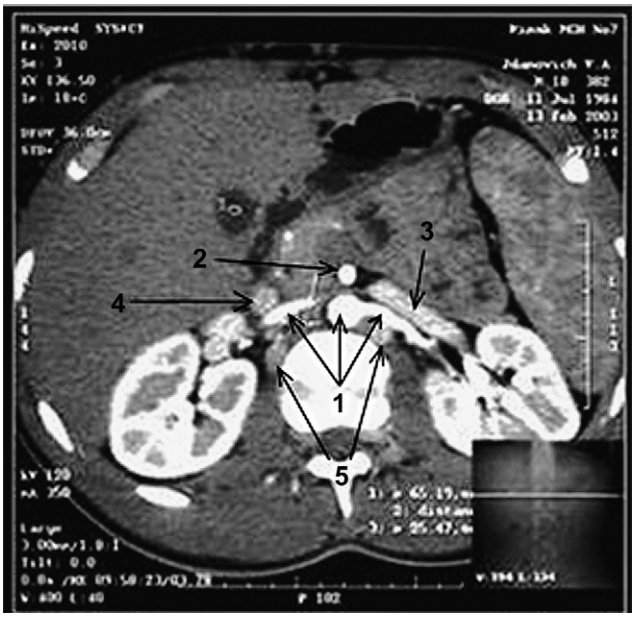

Fig. 3. Spiral CT (transverse section at the L_2 level). 1 – aorta and renal arteries, 2 – superior mesenteric artery, 3 – left renal vein, 4 – stria corresponding to IVC, 5 – ascending lumbar veins.

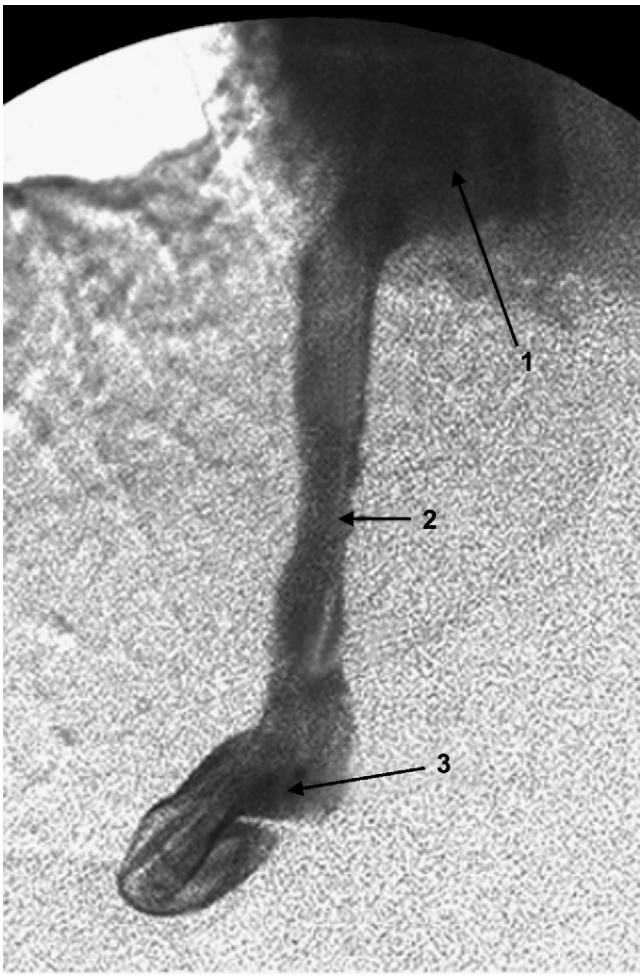

Fig. 2. Retrograde cavagraphy. Retrohepatic part of IVC. 1 – right atrium, 2 – retrohepatic IVC segment, 3 – part of the suprarenal IVC region.

Discussion

The development of the inferior vena cava is a complicated process that includes, at early embryogenesis stages, formation, confluence, and involution of the three primary longitudinal venous systems: subcardinal veins draining the kidneys; sacrocardinal veins draining the lower extremities, and supracardinal veins gathering blood from the body walls.

The different ways of the IVC development, as well as formation of transversal anastomoses between the veins of the right and left body parts, serve as a precondition for anomalies.

The IVC hypo- and aplasia are the most common congenital venous anomaly.

The IVC hypoplasia as a development defect has been described by many authors.[5–7] According to publications, this pathology was first diagnosed in the end of the 18th century during autopsy.[8] Due to recently developed examination methods such as US, HCT the detection frequency of IVC anomalies has improved, and is even found in healthy individuals. This pathology is found in 0.6–2% of patients with congenital heart malformations and in 0.3% of healthy individuals.[9]

The result of our investigation show that the most frequent type of the congenital IVC anomaly is underdevelopment of its subhepatic part from the common iliac veins confluence to the retrohepatic

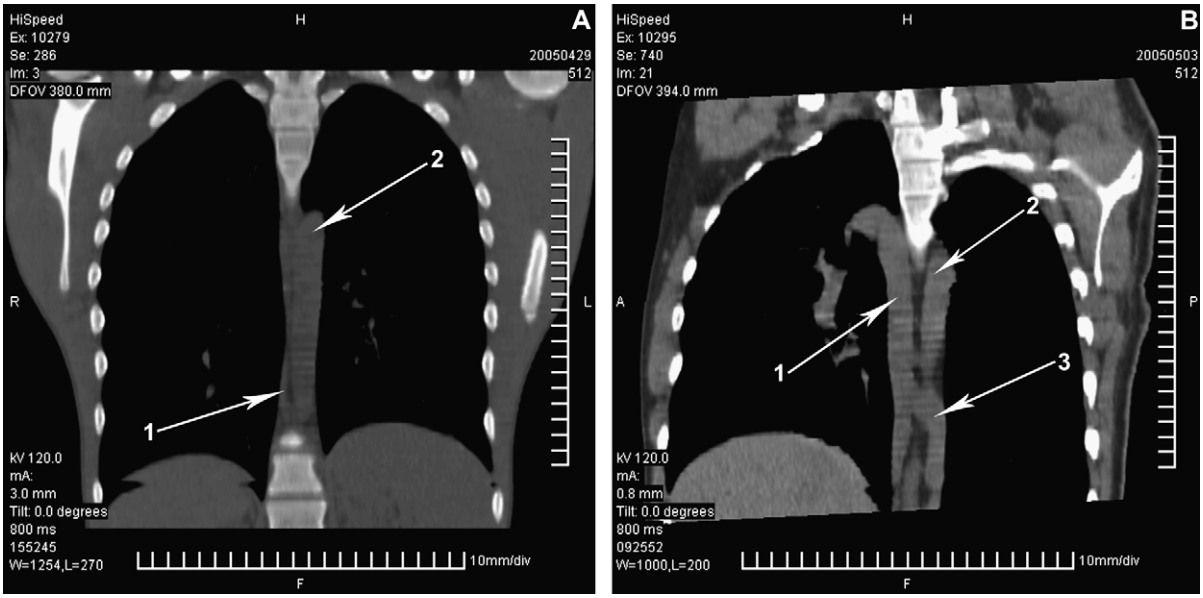

Fig. 4. Spiral CT of the thoracic cavity (longitudinal section). Dilatation of azygos and hemiazygos veins. A – azygos vein (1), aorta (2) in healthy patient, B – azygos vein (1), aorta (2), hemiazygos vein (3) in patient with IVC agenesis.

segment. Abnormality of the retro- and subhepatic IVC regions develops when the right subcardinal vein does not join the hepatic sinusoids. In such cases, the blood from the lower body part reaches the heart through the azygos and hemiazygos veins draining into the superior vena cava. According to our investigations, dilatation of the hemiazygos and azygos veins in such cases amounted to 15–18 mm. The terminal part of azygos vein near its drainage into the superior vena cava was particularly dilated. We believe that the reason for this dilatation of the azygos vein is not only the increased blood flow, but also insufficient diameter of the venous confluence.

The hepatic veins can drain into the right atrium independently in cases of a complete IVC aplasia.[10]

The three patients with extended hypoplasia developed more marked disorders of venous hemodynamics, as demonstrated by trophic ulcers in 2 of 3 patients than the patients with limited hypoplasia of the infrarenal IVC.

There is no consensus regarding most likely cause of the hypoplasia of the infrarenal IVC. It could be due to embryonic dysgenesis of the right supracardial (synonyms – lateral sympathetic, thoracolumbar, paraureteral) vein,[8] or due to an intrauterine or perinatal thrombosis of a formed IVC.[11]

The non-closed right postcardinal vein, a precursor of the supracardinal vein, leads to a pathology called retrocaval ureter.[12] The most part of the ureter in such patients lies behind the IVC. This anomaly is diagnosed during excretory urography or retrograde pyeloureterography. The compression of the ureter by the vena cava leads to dilated pyelocaliceal system and its proximal one-third.

In our case series we found a dilated ureter compressed by a dilated and thrombosed testicular vein. It led to a dilatation of the pyelocaliceal system of the left kidney and acute pyelonephritis, which led to a surgical operation in this patient.

The IVC aplasia is often complemented by congenital heart diseases (dextracardia, septal defects, transposition of the great vessels, pulmonary artery stenosis, common atrium), sometimes with abnormalities of other viscera: situs inversus, duplication or absence of the spleen.[13–16] In our series one out of five patients was diagnosed with an atrial septal defect and stenosis of pulmonary artery, for which he had an open heart operation in childhood. One other patient had a duplication of the pyelocaliceal system.

Our study and literature data show that congenital IVC abnormalities do not present clinically for a long time due to compensated blood outflow via a system of developed collaterals. In case of adverse conditions (trauma, surgery or other) that lead to thrombosis of the outflow tracts, this pathology is manifested by CVI or DVT. The literature shows it is bilateral in more than 50% patients.[15]

The treatment of congenital IVC anomalies is currently conservative for a majority of patients. Our investigation showed that anticoagulant therapy conducted during the acute DVT stage or its relapse

and later vasotonic medications together with elastic leg compression demonstrate a slight improvement in hemodynamics.

At the same time, publications report results of surgical operations on patients with this pathology. In particular, M.J. Dougherty *et al.* described a case of shunting when an anastomosis was made between the azygos and right common iliac veins.[7]

Conclusion

Congenital anomalies of IVC are rare. They are seen more often in young males. They are latent for a long time. Peripheral venous thrombosis or CVI are often the first symptoms of a congenital IVC anomaly. In case of DVT or CVI, especially in young males, a potential IVC abnormality should be excluded.

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