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Conservative treatment vs thrombus removal for Iliofemoral vein thrombosis in patients with congenital abnormalities of the inferior vena cava: a case report and systematic review of the literature

Vangelis G. Alexiou[1,2] · Anna Ntanika[3] · Georgios Pappas[4] · Areti Vassiliou[1] · Konstantinos Palialexis[5] · George Geroulakos[4]

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Abstract

Congenital abnormalities of the Inferior Vena Cava (IVC) should be suspected in cases of Deep Venous Thrombosis (DVT), especially in young patients, with no other risk factors and apparent causes. Currently, there is no guidance regarding the management of such patients. We report a case of Iliofemoral vein thrombosis in a young patient with congenital absence of the IVC that was successfully treated with catheter-directed thrombolysis (CDT) and perform a systematic review of the literature to identify evidence about the epidemiology, clinical presentation, management, and prognosis of this rare cause of DVT. A total of 42 studies reporting on 56 cases were included in the review. The mean age of the patients at the presentation of their first DVT episode is 23.6 years, 83.9% of patients were males, conservative management with anticoagulation was used in 68% of the reported cases, and thrombolysis was used in 32% of the cases. Only 10.7% of patients presented with PE potentially justified by the abnormal anatomy of the deep veins which makes the propagation of thrombi into the pulmonary arteries less possible. Comparing the long-term outcomes of the two treatment groups; 42.3% of the patients treated conservatively vs 15.4% of the patients treated with thrombolysis developed chronic symptoms (residual heaviness, pain, swelling, and cramping). 11.5% of patients who received conservative treatment developed post-thrombotic syndrome. None of the patients treated with thrombolysis developed post-thrombotic syndrome. There were no procedure-related complications and thrombolysis was well tolerated by the entirety of the thrombolysis treatment group. Recurrence of DVT occurred in 13% of the patients treated conservatively and in 7.7% of patients treated with thrombolysis. Thrombus removal by means of thrombolysis is the recommended treatment and can offer excellent short and long-term results. Anticoagulation with NOACs may be prescribed for life to prevent recurrence or for at least 6 months and then reconsidered following further evaluation of patients' bleeding risk. It may be of value to organise an international registry for such patients. Guidelines issued by the relevant scientific societies will then be able to make a clear recommendation about the management of such patients.

Keyword Thrombolysis · Deep vein thrombosis · Congenital abnormalities · Inferior Vena Cava

✉ Vangelis G. Alexiou
v.alexiou@aibs.gr

[1] Department of Surgery - Vascular Surgery Unit, Faculty of Medicine, University of Ioannina, Niarxou Avenue, 45500 Ioannina, Greece

[2] Alfa Institute of Biomedical Sciences (AIBS), Athens, Greece

[3] Faculty of Medicine, University of Ioannina, Ioannina, Greece

[4] Department of Vascular Surgery, Medical School, National and Kapodistrian University of Athens, Attikon University Hospital, Athens, Greece

[5] Department of Interventional Radiology, Medical School, National and Kapodistrian University of Athens, Attikon University Hospital, Athens, Greece

Highlights

- Currently, there is no guidance regarding the management of patients with congenital abnormalities of the inferior vena cava who present with deep vein thrombosis
- More than 80% of the patients were males and the mean age at the presentation of their first DVT episode was 23.6 years
- 42.3% of the patients treated conservatively vs 15.4% of the patients treated with thrombolysis developed chronic symptoms
- None of the patients treated with thrombolysis developed post-thrombotic syndrome
- Recurrence of DVT occurred in 13% of the patients treated conservatively vs 7.7% of patients treated with thrombolysis

Introduction

Abnormalities of the inferior vena cava (IVC) are rare congenital conditions that occur during embryogenesis, from the 6th to 10th week of gestation, when the venous system develops [1, 2]. In rare cases, they are developed due to IVC thrombosis in a fetus or neonate with thrombophilia. These abnormalities are found approximately in 0.5% of the population and have been described as absence, atresia, agenesis, anomalous formation, or interruption of a particular segment of the IVC (hepatic, prerenal, renal, or infrarenal). There is no data on whether this condition is equally distributed between genders, however, the majority of patients with clinical presentations are young males [2–4]. Absence of the IVC has been associated with deep vein thrombosis (DVT) based on the pathophysiology of stasis of venous blood flow in the lower limbs that may not be adequately drained by the azygous venous system [2]. Compensatory enlargement of the collateral venous network may occur in some patients to outweigh the insufficient drainage [1]. The limited literature, which mainly consists of case reports and case series, has not been summarized and analyzed to provide adequate evidence for the management and clinical course of patients with DVT and abnormalities of the IVC.

Patients with this vascular defect may remain asymptomatic and their condition is mostly reported as an incidental finding in a CT angiography, MRI, or post-mortem examination. However, congenital abnormalities of the IVC should be suspected in cases of DVT, especially in young patients, with no other risk factors and apparent causes of DVT [5]. Current guidance of the European Society for Vascular Surgery (ESVS) 2021 Clinical Practice Guidelines on the Management of Venous Thrombosis suggests early thrombus removal for iliofemoral DVT for patients with low risk for bleeding and without comorbidity of cancer or pregnancy. For patients with low to high risk for bleeding, anticoagulation with Novel Oral Anticoagulants (NOACs) is recommended over Low Molecular Weight Heparin (LMWH) and Vitamin K antagonists (VKAs) along with early compression which will relieve symptoms and residual venous occlusion. Anticoagulation therapy may be discontinued at 3 months if recurrence is not present. In cases of recurrent DVT despite compliance with treatment, modifications in the therapeutic schedule are required including switching the type of anticoagulation, increasing the dose of LMWH or NOAC to therapeutic dose, or switching to VKAs with a higher international normalized ratio (INR) target. Anticoagulation should be extended beyond three months in cases of recurrent unprovoked DVT in accordance with the judgment of the treating physician. However, the ESVS guidelines have no specific recommendation on the management of patients with DVT and congenital absence of IVC [2].

We report a case of Iliofemoral vein thrombosis in a young patient with congenital absence of the IVC and also seek to review the literature to identify evidence about the epidemiology, clinical presentation, management, and prognosis of this rare cause of deep venous thrombosis.

Methods

Case report

Medical notes including detailed patient history, emergency department admission notes, medical charts, imaging (preoperative, intraoperative, and postoperative completion angiogram), lab tests, drug charts, and follow-up notes were retrieved and studied to describe the case in detail. Written informed consent was obtained from the patient for the publication of his case accompanied by respective images.

Systematic literature review

Search strategy

We sought to review the literature to identify additional evidence about the epidemiology, clinical presentation, management, and prognosis of DVT in patients with congenital malformations of the inferior vena cava. To identify relevant articles, we systematically searched PubMed until November 2021 using the following search pattern: (absent IVC or absent inferior vena cava or inferior vena cava atresia or

inferior vena cava defect) and (thrombosis, DVT, iliofemoral thrombosis, thromboembolism, acute ischemia, lower extremities ischemia). No limitation on the year of publication or language was set. Furthermore, the references in the relevant articles, including review studies, were checked to identify additional resources. Abstracts of conference proceedings were not sought.

Inclusion criteria

Two of the authors (VGA and AN) independently performed the literature search to locate potentially eligible studies. All studies including reviews, case reports, and case series reporting on the incidence of DVT in patients with congenital anomalies of the IVC were retrieved.

Data extraction

The following data were extracted from all eligible articles: first author, year of publication, clinical details, treatment, outcomes, and follow-up information. The most important findings were tabulated.

The study adheres to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines [6].

Results

Case report

A 24-yrs-old male patient presented with a few hours history of sudden onset pain and swelling of his right limb. On clinical examination, the right calf appeared hot, erythematous, and painful on palpation with edema extending to his proximal thigh. DVT was verified by performing a venous duplex scan. The patient had no history of trauma, or prolonged immobilization and his blood workup including liver function tests (LFTs) and tumor markers appeared within normal limits and factors of protein thrombophilia (dosing antithrombin, homocysteine, protein C and S, factor V of Leiden, and prothrombin mutation) were investigated and found negative. CT of the abdomen and pelvis showed thrombosis at the confluence of the external iliac vein with the interior iliac vein (Image 1), congenital agenesis of the inferior vena cava, and multiple paraspinal collateral channels (Image 2). Angiography (Image 3) showed filling defects of the interior iliac vein which are an indicator of occlusion due to deep venous thrombosis. The patient underwent catheter-directed thrombolysis (CDT). Specifically, he received thrombolytic therapy with 5 mg Actilyse (rtPA) bolus, followed by an infusion of rtPA 0,5 mg/h for 30 h and 500 IU/h heparin. Completion angiogram (Image 4) showed adequate permeability of the internal iliac vein until the spinal external venous plexus which was interpreted as a successful postoperative outcome. Clinically, the patient made an excellent recovery and his limb edema resolved completely. He has been followed up for 9 months and he remains asymptomatic and continues receiving oral apixaban for life as thromboprophylaxis (see Image 5).

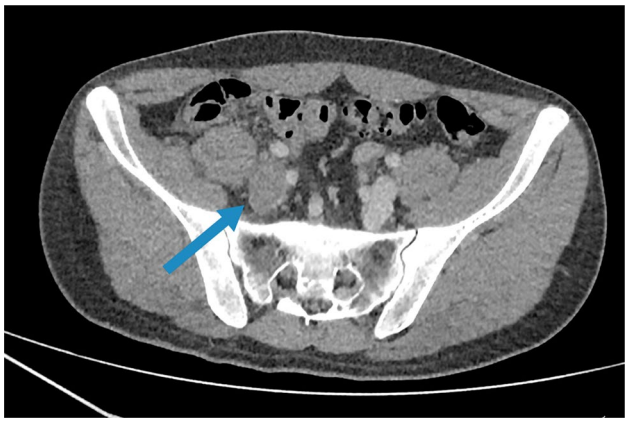

Image 1 Axial CT of the pelvis demonstrating filling defects as an indicator of thrombosis at the confluence of the external iliac vein with the interior iliac vein (red arrow)

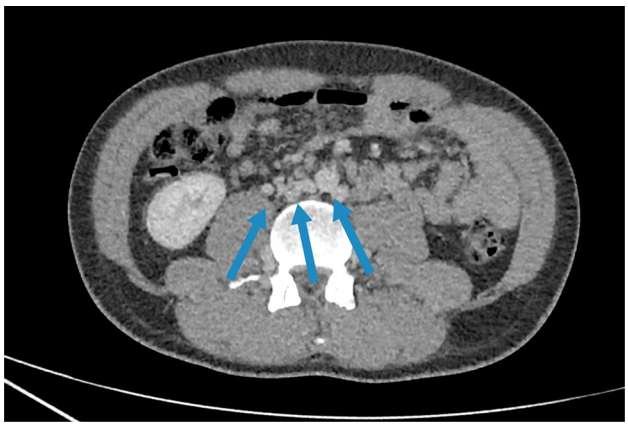

Image 2 Axial CT of the abdomen demonstrating congenital agenesis of the inferior vena cava and multiple paraspinal venous collateral channels

Systematic review

In Fig. 1, we present a flow diagram describing the selection process followed to identify reports included in this systematic review. The PubMed search yielded 320 potentially relevant articles published from 1996 to 2021. After screening the title and the abstract and reading the full text of the articles we decided that the inclusion criteria were fulfilled by 42 articles that presented 56 cases, in total [4,

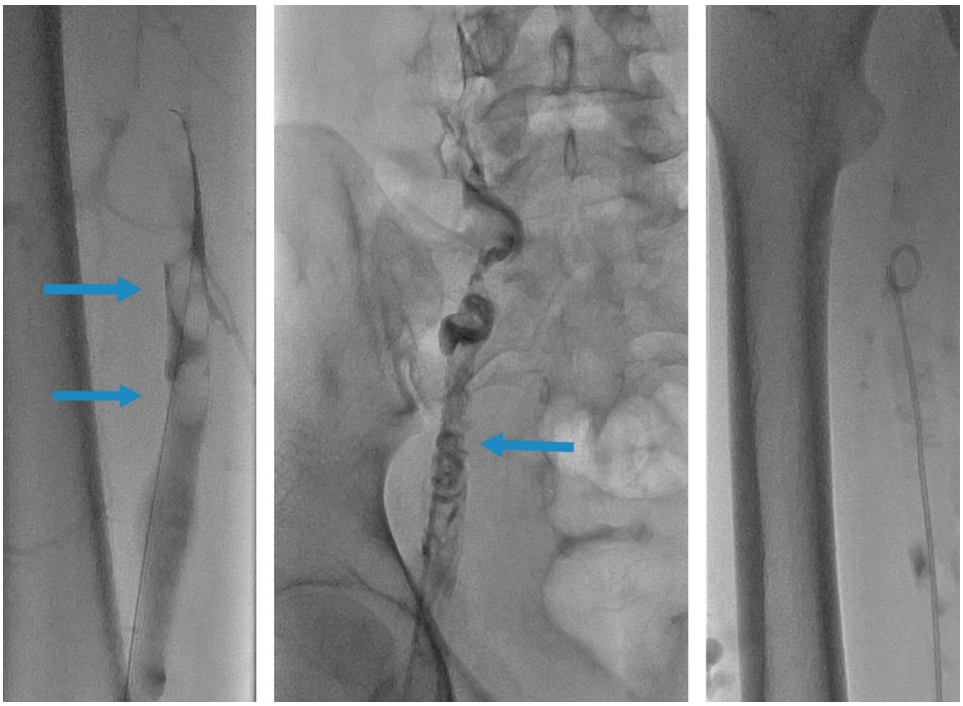

Image 3 Angiography showing filling defects of the interior iliac vein which are an indicator of occlusion due to deep venous thrombosis (first image from right to left) and the insertion of a venous catheter in the inferior iliac vein (second and third image from right to left)

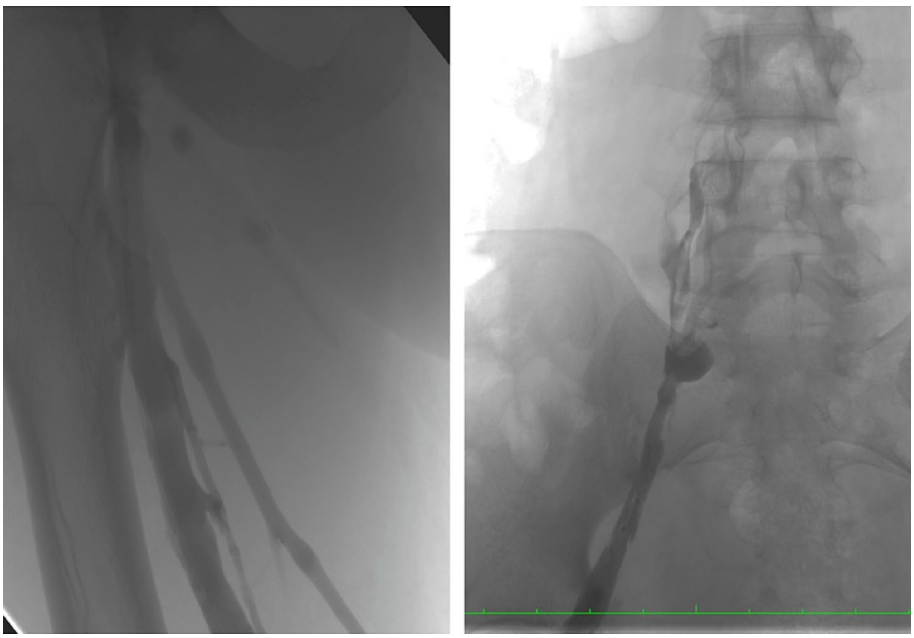

Image 4 Completion angiogram demonstrating adequate permeability of the interior iliac vein up to the spinal external venous plexus

5, 7–44]. Data extracted from the 56 cases are presented in Tables 1, 2. Regarding gender distribution of the reported cases; 83.9% (53/56) of patients were males. The mean age of the patients at the presentation of their first DVT episode is 23.6 years.

Comorbidities and other risk factors were also analyzed; 8.9% (5/56) of the patients had hereditary thrombosis risk factors (heterozygosity for factor V Leiden gene polymorphism, deficiency of antithrombin, protein C or protein S, heterozygous prothrombin gene 20210 mutation, positive lupus anticoagulant) and 19.6% (11/56) of the patients had non-hereditary risk factors (cancer, acute medical illness, surgery, trauma, immobility, obesity, inflammatory diseases and/or infection, hormone therapy, long-distance travel, recent hospitalization, use of contraceptive pills). Among females, 33.3% (3/9) were on oral contraceptives when the symptoms of DVT occurred. Only 10.7% of the reported cases presented with pulmonary embolism (PE). It should be

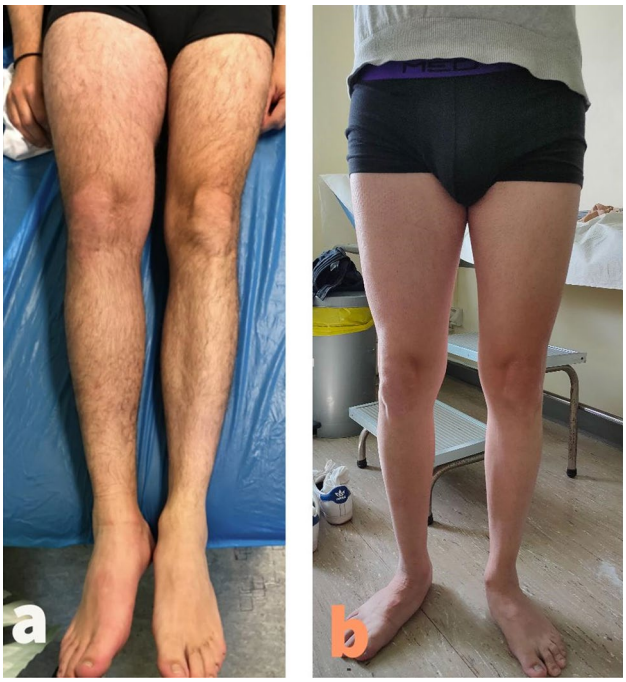

Image 5 Image of the patient at presentation (a) and a week following intervention with thrombolysis (b). The patient had immediate and complete resolution of the right limb edema

noted that, in most cases, there were no clinical symptoms of PE, thus no further investigation was undertaken.

Conservative management with anticoagulation was used in 68% of the reported cases (38/56). Thrombolysis was used in 32% of the cases (18/56) followed by anticoagulation therapy. Thrombolysis was combined with thrombectomy, angioplasty, and/or stent placement in 5 patients, and in 1 patient the treatment was completed with open surgery and the use of prosthetic IVC grafting.

Among cases treated conservatively with anticoagulation and cases treated with thrombolysis, 68.4% (38/26) and 72.2% (13/18) had follow-up information available, respectively. Comparing the long-term outcomes of the two treatment groups, recurrence of DVT occurred in 13% (3/26) of the patients treated conservatively and in 7.7% (1/18) of patients treated with thrombolysis. The mean follow-up time was 22.6 and 14.4 months, respectively. At follow-up evaluation, 19.2% (5/38) of the patients treated conservatively presented completely asymptomatic and 42.3% (11/38) developed chronic symptoms (residual heaviness, pain, swelling, and cramping) while 76.9% (10/18) of the patients treated with thrombolysis presented completely asymptomatic and 15.4% (2/18) developed chronic symptoms. Finally, 11.5% (3/38) of patients who received conservative treatment developed post-thrombotic syndrome. None of the patients treated with thrombolysis developed post-thrombotic syndrome.

Discussion

The main finding of this study is that thrombolysis in patients with DVT and congenital abnormalities of the IVC can offer definitive treatment. Most of the patients are free of long-term symptoms and complications and rarely do they have recurrent DVT, especially if they are given long-term thromboprophylaxis. The appropriate dosing regimen or selection of thromboprophylaxis agents is not clear. Most contemporary studies report the use of lifelong prophylaxis with NOACs. Among cases that were identified by our systematic review and were treated with thrombolysis, only 1 suffered recurrent DVT. Furthermore, all patients reported complete resolution of their symptoms including pain and edema. Most importantly, none presented post-thrombotic syndrome. Finally, it should be noted that there were no procedure-related complications and thrombolysis was well tolerated by the entirety of the thrombolysis treatment group.

On the other hand, 42.3% of the patients treated with anticoagulation only developed at least mild chronic symptoms (residual heaviness, pain, swelling, and cramping), and a significant 11.5% of this group had post-thrombotic syndrome. Furthermore, 13% of the patients in this treatment group had recurrent DVT. It should be noted that most of the cases of patients with DVT and congenital malformations of the IVC that were treated conservatively were reported prior to 2010 when CDT was not available in most institutions. The contemporary guidelines for the management of iliofemoral venous thrombosis suggest early thrombus removal for patients with low risk for bleeding and without comorbidity of cancer or pregnancy [2]. Based on the above guidance and the findings of this study, we believe that it is imperative to perform CDT in patients with iliofemoral venous thrombosis and congenital malformations of the IVC.

It is really important to be able to recognize the presence of DVT in patients with IVC defects. The clinical presentation of thrombosis due to the absence of IVC varies according to the acuity and the extent of the thrombosis. Many patients may present with subtle symptoms that are difficult to recognize and correlate with DVT. Symptoms like leg heaviness, pain, swelling, and cramping may be accompanied by nonspecific abdominal/pelvic pain and scrotal swelling. Other symptoms like dyspnea and oliguria may occur due to clot migration and/or embolization into the lungs and renal veins respectively. Most patients with chronic IVC abnormalities have already developed collateral drainage networks and may tolerate an extensive iliofemoral thrombosis better than other patients. There are also reports of less common symptoms like lumbar radicular pain, sciatica, and cauda equina syndrome due to compression of peripheral nerves by dilated veins [45]. If not promptly and adequately

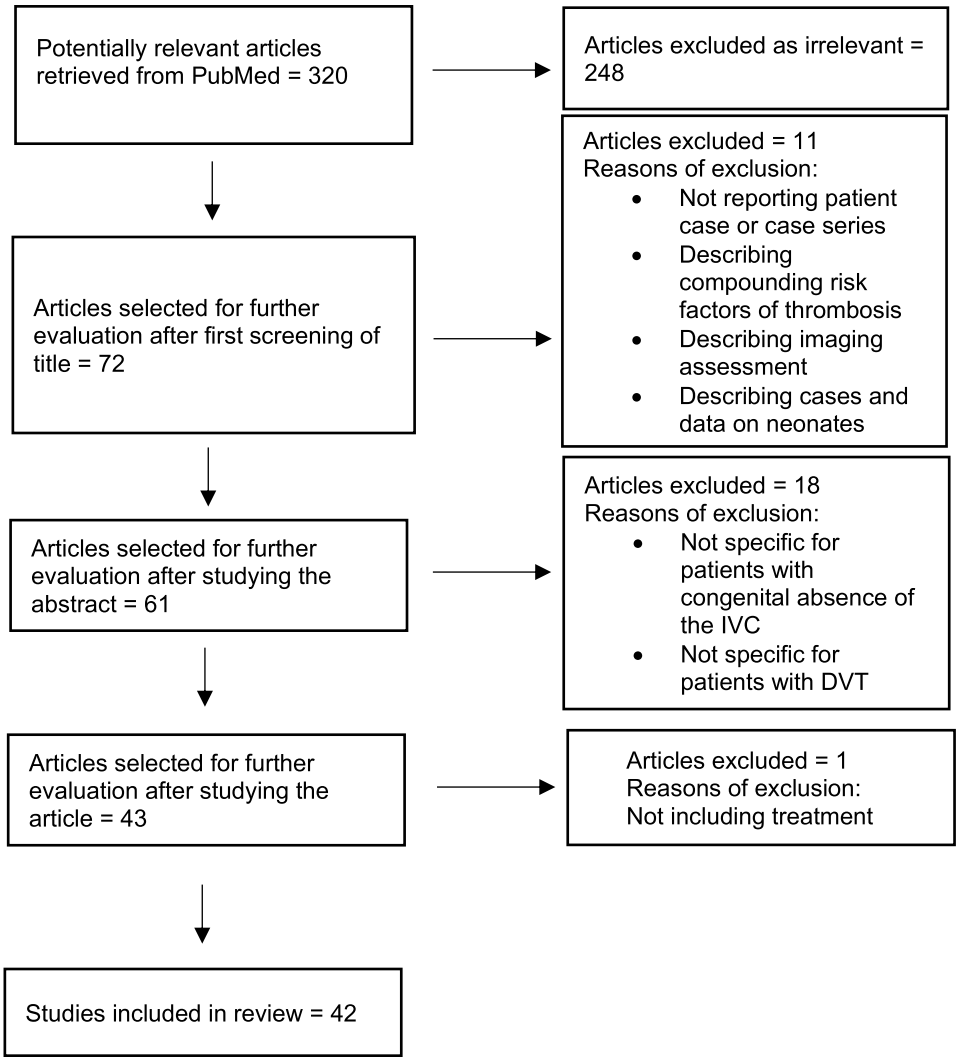

Fig. 1 Flow diagram of the systematic review

treated, thrombosis can result in severe post-thrombotic syndrome with leg cramping, skin pigmentation, venous ulceration, and disabling claudication [2, 45].

The suspicion of IVC congenital malformations is based on the patients' clinical symptoms that are not in keeping with their young age and the absence of other thrombosis risk factors. Duplex ultrasound is usually the modality of choice for initial screening but it should be followed by a CT angiography and/or MRI. Furthermore, it has been reported that in 30–40% of patients with DVT, PE is also present. The finding of our study that only 10.7% of patients with congenital IVC malformations present with PE may be justified by the abnormal anatomy of the deep veins which makes the propagation of thrombi into the pulmonary arteries less possible. Finally, it should be noted that the results of this study confirm that there is an uneven distribution among genders. More than 80% of the cases of venous thrombosis associated with malformations of the IVC occurred in males.

The result of this systematic review should be considered in view of certain limitations. First, it should be noted that IVC abnormalities are rare and the available evidence comes from case reports and case series. Thus the quality of evidence is low. Furthermore, the "file-drawer effect" should be acknowledged when interpreting the results of this review. Meaning that authors tend to publish cases of patients who had good outcomes, and put the cases with complications in the file-drawer. Finally, the available follow-up information for the reported cases is rather limited.

Due to the rarity of the condition, it may not be possible to organize a clinical trial to further investigate the management and treatment that should be provided to these patients. However, it is of value to organize a national or international registry for rare venous diseases like this. It is imperative to record and analyze all relevant cases and provide data about long-term follow-up with comprehensive and clear outcomes. This way, we will be able to summarise more patients and provide stronger evidence about the appropriate

Table 1 Studies reporting cases of patiens with DVT and congenital malformations of IVC treated with thrombolysis

Publication details	Age, Sex	Commorbidities / risk factors	DVT anatomy	PE	Treatment	Discharge	Additional Procedures	Recurrence	Follow-up	Absence of symptoms	Chronic symptoms	Duration of treatment	PTS
Tarazi et al., 2020 [7]	28 M	Recent trip, factor V Leiden	Bilateral iliofemoral	No clinical symptoms	Thrombolysis	Oral warfarin, compression stockings	Left sided thrombectomy, 14 mm balloon angioplasty	–	27 months RE asymptomatic, LLE swelling and ankle pigmentation	No	Yes	27 months	No
Tarazi et al., 2020 [7]	25 M	Elevated calcium and PTH, treated for parathyroid carcinoma	Iliofemoral vein, extending to the LCIV at level L4-5	No clinical symptoms	Thrombolysis with alteplase	Apixaban 5 mg twice daily	–	–	24 months, asymptomatic	Yes	No	24 months	No
Tarazi et al., 2020 [7]	35 M	–	No information	CTPA, left lower lobe segmental PE	Thrombolysis, LMWH	Apixaban long-term, compression stockings	–	–	20 months, asymptomatic	Yes	No	20 months	No
Tarazi et al., 2020 [7]	21 M	Factor V Leiden, nephrectomy 10 years ago	Bilateral iliofemoral	No clinical symptoms	LMWH, thrombolysis	Oral warfarin	Residual thrombus in RLE required thrombectomy	–	No information	No information	No information	No information	No information
Ramos Aranda et al., 2018 [8]	23 M	–	Bilateral extending to both CIVs	No clinical symptoms	LMWH, Thrombolysis (EKOS) with alteplase	Rivaroxaban, compression stockings	Stent placement across the left CIV	–	12 months, asymptomatic	Yes	No	12 months	No
Ramos Aranda et al., 2018 [8]	30 M	Prior use of anabolic steroids, extreme physical activity	Left CIV, bilateral renal veins and the infrarenal segment of the IVC	CTA, bilateral PE	LMWH, Thrombolysis (EKOS) with alteplase	Oral rivaroxaban, compression stockings	Balloon angioplasty, stent placement across the left iliac vein	–	6 months, asymptomatic	Yes	No	6 months	No

Table 1 (continued)

Publication details	Age, Sex	Commorbidities / risk factors	DVT anatomy	PE	Treatment	Discharge	Additional Procedures	Recurrence	Follow-up	Absence of symptoms	Chronic symptoms	Duration of treatment	PTS
Khalid et al., 2018 [9]	27 M	Recent trip, KILT syndrome	Extensive DVT of the right CFV, left CIV, left internal and external iliac veins	CT negative	LMWH, thrombolysis, thrombectomy, balloon angioplasty	Rivaroxaban, compression stockings, lifestyle changes	–	No information	No information	No information	No information	No information	No information
Kim et al., 2018 [10]	24 M	Prior DVT episode	Iliac through popliteal veins extending into the posterior tibial vein, anterior tibial vein and small saphenous vein	No clinical symptoms	Thrombolysis (EKOS), heparin infusion,	Enoxaparin, warfarin	–	No information	No information	No information	No information	No information	No information
Man et al., 2016 [11]	19 F	–	Bilateral CIV	No clinical symptoms	Bilateral iliac vein thrombolysis	Oral apixaban, compression stockings	–	–	6 weeks, improved edema	No?	Yes?	6 weeks	No
Reslan et al., 2015 [12]	22 F	Use of oral contraceptive pills	CFV and external iliac vein	No clinical symptoms	LMWH, warfarin, thrombolysis (EKOS), compression stockings	Coumadin for life	–	–	1 year, asymptomatic	Yes	No	1 year	No

Table 1 (continued)

Publication details	Age, Sex	Commorbidities / risk factors	DVT anatomy	PE	Treatment	Discharge	Additional Procedures	Recurrence	Follow-up	Absence of symptoms	Chronic symptoms	Duration of treatment	PTS
Koppisetty et al., 2015 [13]	36 M	Previous physical exertion	Right external iliac vein, right and left CFV, femoral vein, greater saphenous vein, small saphenous vein, popliteal vein, gastrocnemius vein, peroneal veins, posterior tibial veins, soleal vein	CT negative	Thrombolysis, (EKOS), balloon angioplasty, mechanical thrombectomy	Rivaroxaban, compression stockings	Final venogram showed patency of the iliac veins and lower IVC with persistent but improved sluggish flow	No	1 year, no recurrent DVT	Yes?	No	1 year	No
Parsa et al., 2015 [14]	19 M	–	Left CFV	No clinical symptoms	Thrombolysis, heparin	Coumadin		No	No information	Yes	No information	No information	No
Ali et al., 2015 [15]	28 M	Prior lower extremity DVT	Extensive bilateral greater saphenous reflux, unilateral right femoral vein incompetence, bilateral iliofemoral DVTs	No clinical symptoms	Stripping of right great saphenous vein, laser ablation of left great saphenous vein, bilateral microphlebectomies, Thrombolysis	Oral anticoagulation for 6 months, continues on low-dose aspirin	Prosthetic reconstruction of the IVC with 14 mm ringed polytetrafluoroethylene graft	–	24 months, no recurrent symptoms	Yes	No	6 months Aspirin 24 months	No

Table 1 (continued)

Publication details	Age, Sex	Commorbidities / risk factors	DVT anatomy	PE	Treatment	Discharge	Additional Procedures	Recurrence	Follow-up	Absence of symptoms	Chronic symptoms	Duration of treatment	PTS
Epperla et al., 2014 [16]	21 M	Prothrombin gene 20210 mutation heterozygosity, positive lupus anticoagulant and elevated factor VIII level	Lower IVC and iliac vessels	CTPA negative	IV heparin, thrombolysis, mechanical thrombectomy, balloon angioplasty	sc enoxaparin, compression stockings		–	6 months, switched to rivaroxaban, remains stable	Yes	No	6 months	No
Singh et al., 2010 [17]	26 M	-	Left distal external iliac vein, left CFV, proximal superficial femoral vein and the ascending lumbar vein	No clinical symptoms	LMWH, mechanical thrombectomy, catheter-directed thrombolysis, warfarin	Oral warfarin	Venography confirmed that all the thrombotic veins were patent	–	No information	No information	No information	No information	No
Rogers et al., 2010 [18]	26 M	Underwent eft nephrectomy due to Wilms tumor at the age of 2, treated with neoadjuvant and adjuvant chemotherapy	Extensive DVT from the popliteal to the left external iliac vein	No clinical symptoms	IV LMWH, limb elevation, thrombolysis (EKOS)	Oral warfarin for 6 months	–	–	6 months, asymptomatic	Yes	No	6 months	No

Table 1 (continued)

Publication details	Age, Sex	Commorbidities / risk factors	DVT anatomy	PE	Treatment	Discharge	Additional Procedures	Recurrence	Follow-up	Absence of symptoms	Chronic symptoms	Duration of treatment	PTS
Dean et al., 2006 [19]	16 M	Previous physical exertion	Extensive DVT from the popliteal to the distal external iliac vein	Spiral chest CT displayed acute pulmonary emboli	Thrombolysis balloon angioplasty in the right CIV, placement of two stents, anticoagulation	Oral warfarin long-term	–	–	6 months, minimally swollen RLE, remained asymptomatic	Yes	No	6 months	No
Dean et al., 2006 [19]	18 F	Use of oral contraceptive pills	Right distal external iliac and CFV, infrarenal IVC and right CIV	No clinical symptoms	Thrombolysis,anticoagulation		Recurrence treated with thrombolysis, adjunctive mechanical thrombectomy, placement of 3 venous stents within the IVC and right CIV	Recurrence of thrombosis in the IVC and right iliofemoral veins 10 days after treatment,	6-month, RLE exhibited trivial swelling and she was ambulating on a treadmill without venous claudication	No	No	6 months	No

IVC: Inferior Vena Cava, *CFV*: Common Femoral Vein, *CIV*: Common Iliac Vein, *DVT*: Deep Venous Thrombosis, *PE*: Pulmonary Embolism, *PTS*: Post-thrombotic Syndrome, RLE: Right Lower Extremity, *LLE*: Left Lower Extremity, *CTPA*: Computed Tomography Pulmonary Angiogram, *CT*: Computed Tomography, *LMWH*: Low Molecular Weight Heparin, *PTH*: Parathyroid Hormone, *KILT*: Kidney and Inferior vena cava abnormalities with Leg Thrombosis, *EKOS*: EkoSonic Endovascular System

Table 2 Studies reporting cases of patiens with DVT and congenital malformations of IVC treated conservatively

Publication details	Age, Sex	Commorbidities / risk factors	DVT anatomy	PE	Treatment	Discharge	Complications	Recurrence	Follow-up	Absence of symptoms	Chronic symptoms	Eventual duration of treatment	PTS
Tarazi et al., 2020 [7]	19 M	–	CFV extending to the CIV	No clinical symptoms	LMWH, rivaroxaban long term, compression stockings	–	–	–	42 months, asymptomatic	Yes	No	42 months	No
Chew et al., 2016 [20]	21 M	Ureterocele, partial nephrectomy as a neonate	Right CFV extending into and throughout the external iliac vein	No clinical symptoms	Apixaban	Lifelong anticoagulation			No information	No information	No information	No information	No
Mustafa et al., 2015 [21]	24 M		Extending from the left CIV, external iliac vein, internal iliac vein, CFV, popliteal vein, great saphenous vein and small saphenous vein	No clinical symptoms	Enoxaparin, warfarin	Oral warfarin for 6 months		6 months after stopping warfarin, the patient developed DVT	Continue oral anticoagulation for life	No	Yes	6 months	No
Haddad et al., 2015 [22]	55 M	Obstructive sleep apnea	Extending from the CFV to the popliteal vein. Second DVT in the deep profunda vein	No clinical symptoms	Enoxaparin, warfarin	Warfarin for life, compression stockings			6 months, no recurrent DVT	No information	No information	6 months	No

Table 2 (continued)

Publication details	Age, Sex	Commorbidities / risk factors	DVT anatomy	PE	Treatment	Discharge	Complications	Recurrence	Follow-up	Absence of symptoms	Chronic symptoms	Eventual duration of treatment	PTS
Alderman et al., 2015 [23]	34 M	RLE DVT 14 years previously, oral warfarin for life was perscribed which he stopped 3 years ago of his own accord	External iliac vein, common, deep and superficial femoral veins. Partial occlusion of the popliteal vein, posterior tibial veins, peroneal veins and anterior tibial veins	Negative CTPA	LMWH	Oral warfarin for life, compression stockings			No information about duration, ongoing swelling in the LLE, pain improved	No	Yes	No information	No
Halparin et al., 2015 [4]	14 M	Atrophic left kidney, hypertrophy of the right kidney	Left CIV and external iliac vein, left superficial vein and CFV	No clinical symptoms	Low-dose aspirin or unfractionated heparin, LMWH, warfarin on discharge	Oral anticoagulation for 18 months	Initially some progression of thrombus	No	1 year, asymptomatic, mild pain on exersice	No	Yes	18 months	No
Halparin et al., 2015 [4]	15 M	Physical exertion prior to admission	Bilateral CIV and CFV, infrarenal IVC, bilateral renal veins, inferior mesenteric vein	No clinical symptoms	Low-dose aspirin or unfractionated heparin, LMWH and then warfarin on discharge	Oral anticoagulation for 14 months	2 weeks following initial presentation symptomatic extension of thrombosis on the RLE. Treated with bridging heparin infusion	No	4 years and 2 months, asymptomatic, mild pain on exercise	No	Yes	14 months	No

Table 2 (continued)

Publication details	Age, Sex	Commorbidities / risk factors	DVT anatomy	PE	Treatment	Discharge	Complications	Recurrence	Follow-up	Absence of symptoms	Chronic symptoms	Eventual duration of treatment	PTS
Halparin et al., 2015 [4]	16 M	Physical exertion prior to admission	Right femoral vein, right common, external, and internal iliac veins	No clinical symptoms	Low-dose aspirin or unfractionated heparin, LMWH, warfarin on discharge	Oral anticoagulation for 14 months		No	6 years and 10 months, no evidence of recurrence	No	Yes	14 months	Yes
Halparin et al., 2015 [4]	14 M		Bilateral common iliac and femoral veins, right external iliac vein	No clinical symptoms	Low-dose aspirin or unfractionated heparin, LMWH and then warfarin on discharge	Oral anticoagulation for life	6 weeks following initial presentation symptomatic extension of DVT while on anticoagulation, two right knee hemarthroses while on anticoagulation	Recurrence of DVT 5 weeks after discontinuation of anticoagulation confirmed with MRV		No	Yes	for life	No
Halparin et al., 2015 [4]	14 M	Physical exertion prior to admission, elevated creatinine and hypertension	Bilateral external iliac vein, CIV, CFV and right superficial femoral vein	No clinical symptoms	Low-dose aspirin or unfractionated heparin, LMWH and then warfarin on discharge	Oral anticoagulation for 12 months		No	18 months, no recurrence	Yes	No	12 months	No

Table 2 (continued)

Publication details	Age, Sex	Commorbidities / risk factors	DVT anatomy	PE	Treatment	Discharge	Complications	Recurrence	Follow-up	Absence of symptoms	Chronic symptoms	Eventual duration of treatment	PTS
Paddock et al., 2014 [24]	18 M	Abdominal distention, grunting and acidosis on day 4 of life, formation of a descending loop colostomy, anastomotic leak, defunctioning colostomy closed at 4 years of age	left external iliac vein, CFV and superficial femoral vein	Chest radiograph could not exlude PE	Elevation of the limb, LMWH	Anticoagulation for 6 months		No information	No information, received 6 months of anticoagulation therapy	No information	No information	No information	No
Bami et al., 2014 [25]	14 M	Type 1 diabetes mellitus, hypertension, receiving enalapril,	Left CFV, left superficial femoral, and left popliteal vein		LMWH	Oral warfarin		No information	No information	No information	No information	No information	No

Table 2 (continued)

Publication details	Age, Sex	Commorbidities / risk factors	DVT anatomy	PE	Treatment	Discharge	Complications	Recurrence	Follow-up	Absence of symptoms	Chronic symptoms	Eventual duration of treatment	PTS
Bolocan et al. 2014 [26]	41 M	Obesity, neglected type II diabetes melitus, untreated hypertension, multiple surgeries from the age of 4 for varicose veins and recurrece, protein C deficiency, anticardiolipin antibodies	DVT in the right parietal and femoral popliteal vein	No clinical symptoms	Unfractionated heparin, Fondaparinux	Warfarin	The patient developed thrombocytopenia, required replacement therapy with fondaparinux	No information	No information	No	Yes	No information	No
Namisaki et al., 2013 [27]	44 M		Mural thrombus in the left popliteal vein	Present in the enhanced CT	iv unfractionated heparin	Warfarin		No	5 years, no recurrence	No information	No information	No information	No
Tribe et al., 2013 [28]	11 M	Partial protein C deficiency	Superficial femoral veins extending cranially to the end of the aberrant IVC	No clinical symptoms	LMWH, compression stockings	Oral warfarin		No information	No information	No information	No information	No information	No

Table 2 (continued)

Publication details	Age, Sex	Commorbidities / risk factors	DVT anatomy	PE	Treatment	Discharge	Complications	Recurrence	Follow-up	Absence of symptoms	Chronic symptoms	Eventual duration of treatment	PTS
O' Connor et al., 2011 [29]	20 M	No	Left long saphenous vein and its tributaries, extended through collaterals into superficial varicosities in the anterior abdominal wall across the midline into the right long saphenous vein tributaries	No clinical symptoms	Unfractionated heparin	Oral warfarin for 6 months, compression stockings		No	6 months, no recurrent DVT or lower limb venous ulceration	No information	No information	6 months	No

Table 2 (continued)

Publication details	Age, Sex	Commorbidities / risk factors	DVT anatomy	PE	Treatment	Discharge	Complications	Recurrence	Follow-up	Absence of symptoms	Chronic symptoms	Eventual duration of treatment	PTS
Nseir et al., 2011 [30]	33 M	Recurrent episodes of idiopathic DVT for the last 15 years, recently stopped prophylactic enoxaparin, receiving allopurinol and colchicine for presumed diagnosis of gout	RLE	No clinical symptoms	sc enoxaparin 160 mg daily, antibiotic therapy for skin infection	warfarin		No	3 months, swelling of left ankle, redness with trophic skin changes and a mild improvement of the skin ulcers	No	Yes	3 months	No
Kogias et al., 2011 [31]	21 M	Heterozygous factor V mutation	Major iliofemoral thrombosis with associated thrombophlebitis and massively dilated iliac vessels	No clinical symptoms	Anticoagulation and antiinflammatory therapy	Anticoagulation		No	5 months, asymptomatic	Yes	No	5 months	No

Table 2 (continued)

Publication details	Age, Sex	Commorbidities / risk factors	DVT anatomy	PE	Treatment	Discharge	Complications	Recurrence	Follow-up	Absence of symptoms	Chronic symptoms	Eventual duration of treatment	PTS
Konopka et al., 2010 [32]	43 F	Family history of varicose veins	Acute iliofemoral DVT on the right side	No clinical symptoms	IV unfractionated heparin adjusted by by activated partial thromboplastin time followed by oral anticoagulation with antivitamin K			No	6 months, no recurrence, mild edema of the right ankle and almost complete venous recanalization	No	Yes	6 months	No
Iqbal et al., 2006 [33]	54 M	Mild chronic renal impairment, Perthe's disease, and non-healing venous ulcers on the medial aspect of the right ankle	Right iliac and superficial femoral veins	No clinical symptoms	LMWH, warfarin			No information	No information	No information	No information	No information	No

Table 2 (continued)

Publication details	Age, Sex	Commorbidities / risk factors	DVT anatomy	PE	Treatment	Discharge	Complications	Recurrence	Follow-up	Absence of symptoms	Chronic symptoms	Eventual duration of treatment	PTS
Kelly et al., 2008 [34]	24 M	No	Extensive thrombus within the right common femoral, external iliac, right common iliac vein extending retrogradally into the left common iliac vein	No clinical symptoms	Warfarin	Warfarin		Readmitted with a further DVT of the right leg	No information	No information	No information	No information	No
Jung Suh et al., 2008 [35]	62 M	Had been treated for DVT of the lower extremity seventeen years earlier, right hepatic lobe agenesis	Occlusive thrombosis of the right external iliac and femoral vein	No clinical symptoms	LMWH			No information	No information	No information	No information	No information	No
Klessen et al., 2008 [43]	29 M	No	RLE and pelvis	No clinical symptoms	Heparin, compression stockings, ibuprofen,	Oral phenprocoumon		No	3 months, partial recanalization of the right pelvic and deep leg veins	No	Yes	3 months	No

Table 2 (continued)

Publication details	Age, Sex	Commorbidities / risk factors	DVT anatomy	PE	Treatment	Discharge	Complications	Recurrence	Follow-up	Absence of symptoms	Chronic symptoms	Eventual duration of treatment	PTS
Gil et al., 2006 [36]	14 M	No	Thrombosis of the left iliac and femoral veins	No clinical symptoms	Heparin	oral acenocoumarol for life, compression stockings, elevated bed		No	2 years, CT scan showed a reduction of the initial thrombosis, swelling of the LLE almost absent	No	Yes	2 years	No
Sakellaris et al., 2005 [37]	10 M	No	CIV, external iliac veins, CFV and superficial extention to the IVC	No clinical symptoms	IV heparin	Oral warfarin for life		No	18 months, asymptomatic	Yes	No	18 months	No
Cho et al., 2004 [38]	32 M	The aortic arch was in the opposite orientation to the norm	Right popliteal vein	CT showed extensive thrombosis in the bilateral pulmonary arteries	IV unfractionated heparin	Oral warfarin for life		No	15 months, no recurrence of symptoms	Yes	No	15 months	No
Simsek et al., 2004 [39]	33 M	DVT of the left iliac vein 8 years earlier, treated with anticoagulation for 6 months	Right deep femoral and the external iliac veins	No clinical symptoms	Oral acenocoumarol	Oral acenocoumarol for life		No information	No information	No information	No information	No information	No

Table 2 (continued)

Publication details	Age, Sex	Commorbidities / risk factors	DVT anatomy	PE	Treatment	Discharge	Complications	Recurrence	Follow-up	Absence of symptoms	Chronic symptoms	Eventual duration of treatment	PTS
D' Aloia et al., 2003 [40]	40 F		Infrarenal IVC, CIVs, external iliac veins, right femoral vein	Perfusion lung scintigraphy indicated high probability for bilateral PE	IV unfractionated heparin, oral warfarin, compression stockings, venous pump, leg exercise	Oral warfarin		No	1 year, improvement of thrombosis	No	No information	No information	No
Chee et al., 2001 [41]	23 M	No	Iliofemoral vein	No clinical symptoms	Heparin	Oral warfarin long-term		No	22 months, no recurrence	No information	No information	22 months	No
Chee et al., 2001 [41]	18 F	Oral contraceptive use, factor V Leiden heterozygosity	Bilateral iliofemoral veins	No clinical symptoms	Heparin	Oral warfarin long-term, discontinued contraceptive pill		No	22 months, no recurrence, warfarin withdrawn after 6 months	No information	No information	6 months	No
Chee et al., 2001 [41]	40 M	No	Bilateral iliofemoral veins	No clinical symptoms	Heparin	Oral warfarin long-term		No	22 months, no recurrence	No information	No information	22 months	No
Chee et al., 2001 [41]	26 F	No	Bilateral iliofemoral veins	No clinical symptoms	Heparin	Oral warfarin long-term		No	22 months, no recurrence	No information	No information	23 months	No
Ruggeri et al., 2001 [5]	20 M	No	Right femoral-iliac	No clinical symptoms	IV heparin	Oral warfarin long-term for 19–38 months		No	19 months, no recurrence	No information	No information	19 months	No
Ruggeri et al., 2001 [5]	22 M	No	Bilateral femoral	No clinical symptoms	IV heparin	Oral warfarin long-term for 19–38 months		No	38 months, no recurrence	No information	No information	38 months	No
Ruggeri et al., 2001 [5]	15 M	No	Right femoral	No clinical symptoms	IV heparin	Oral warfarin long-term for 19–38 months		No	48 months, warfarin withdraw after 15 months because of poor compliance	No information	No information	24 months	No

Table 2 (continued)

Publication details	Age, Sex	Commorbidities / risk factors	DVT anatomy	PE	Treatment	Discharge	Complications	Recurrence	Follow-up	Absence of symptoms	Chronic symptoms	Eventual duration of treatment	PTS
Ruggeri et al., 2001 [5]	19 F	No	Bilateral femoral-iliac	No clinical symptoms	IV heparin	Oral warfarin long-term for 19–38 months		No	20 months, no recurrence	No information	No information	20 months	No
Ramanathan et al., 2001 [42]	12 F	No	Common femoral and superficial femoral veins	Negative	IV heparin,			No information	No information	No information	No information	No information	No
Shah et al., 1996[44]	30 M	Three episodes of DVT involving the LLE during the preceding 8 years and an episode of DVT of the RLE, taking coumadin on a long term basis	Reduced flow and caliber in the main deep venous system were detected down to the left knee	No clinical symptoms	The patient was admitted for evaluation of lower extremity venous insufficiency	Raised dose of oral coumadin, compression stockings, leg elevation, exercise, venous pump		Multiple episodes of DVT	No information	No information	No information	No information	No information

IVC: Inferior Vena Cava, *CFV*: Common Femoral Vein, *CIV*: Common Iliac Vein, *DVT*: Deep Venous Thrombosis, *PE*: Pulmonary Embolism, *PTS*: Post-thrombotic Syndrome, *RLE*: Right Lower Extremity, *LLE*: Left Lower Extremity, *CTPA*: Computed Tomography Pulmonary Angiogram, *CT*: Computed Tomography, *LMWH*: Low Molecular Weight Heparin, *PTH*: Parathyroid Hormone, *KILT*: Kidney and Inferior vena cava abnormalities with Leg Thrombosis, *EKOS*: EkoSonic Endovascular System

management of patients with IVC abnormalities who present with DVT. Guidelines issued by the relevant scientific societies will then be able to make a clear recommendation about the management of such patients.

In conclusion, it is really important to have high suspicion and be able to recognize congenital IVC malformations in young males who present with DVT and do not have significant risk factors or comorbidities. Thrombus removal by means of thrombolysis is the recommended treatment and can offer excellent short and long-term results. Anticoagulation with NOACs may be prescribed for life to prevent recurrence or for at least 6 months and then reconsidered following further evaluation of patients' bleeding risk. In asymptomatic individuals, with an incidental finding of congenital absence of IVC, thromboprophylaxis may be considered to prevent DVT.

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Declarations

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