Pelvic Congestion Syndrome

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Pathophysiology

Chronic pelvic pain – defined as intermittent or constant pain of 3–6 months duration not associated with menses, intercourse, or pregnancy – affects up to 27% of women 18–50 years of age [1, 2]. This pain can cause severe disability with 15% of women missing work and 45% experiencing reduced productivity [3]. There is a vast differential diagnosis for chronic pelvic pain including musculoskeletal, neurologic, gastrointestinal, urinary, gynecologic, and even mental health disorders. Even after diagnostic laparoscopy, nearly 60% of patients remain without a diagnosis for the cause of their pain [4].

Pelvic congestion syndrome (PCS) was first described in 1857 by Richet [5]. In the second half of the twentieth century, a connection was made between pelvic varices and chronic pelvic pain [6]. Pelvic congestion syndrome is caused by pelvic or vulvar varicosities and can be a cause of chronic pelvic pain in 10–30% of women without other obvious pathologies [7, 8]. It tends to be positional; most women describe increasing discomfort with prolonged standing. PCS is caused by incompetence of valves in the gonadal veins or internal iliac veins which leads to pelvic venous insufficiency. Ovarian vein incompetence has been reported in up to 60% of women with PCS [9]. The male analogue is venous incompetence of the gonadal vein leading to varicoceles. Pelvic venous reflux has gained more attention in the past few decades for its potential causal relationship with lower extremity varicose veins. Pelvic venous hypertension is transmitted to the lower extremity venous plexus through the external iliac vein. In one study, pelvic venous reflux was detected in 17% of patients who had recurrence of varicose veins after treatment [10]. In women with pelvic venous insufficiency, 71% demonstrated pelvic venous reflux, and 58% demonstrated communication with the lower extremity superficial veins. Nearly two-thirds of these patients demonstrated recurrence of varicose veins after treatment indicating pelvic venous insufficiency as an important cause of both pelvic congestion syndrome and varicose veins [11].

Key Point
The presence of vulvar varicosities in a patient with lower extremity varicose veins should raise the concern for pelvic congestion syndrome.

Key Point
Risk factors for pelvic congestion syndrome

- Pregnancy
- Nutcracker syndrome
- Congenital vascular malformation
- Genetics
- Iliac vein compression (May-Thurner syndrome)
- Hormonal dysfunction

Similar to varicose veins in the legs, the pathophysiology of pelvic venous insufficiency is multifactorial. Pregnancy and genetics play a strong role. During pregnancy, the pelvic and ovarian veins become dilated in response to increased flow of nearly 50% to the fetus and are compressed by the gravid uterus. With multiple pregnancies and repetitive venous dilatation, there may be valve damage which results in reflux. The left ovarian vein and the right internal iliac vein are the most frequently involved [12]. Pelvic congestion

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syndrome is absent in postmenopausal women likely secondary to the decline of estrogen, which acts as a venous dilator [8].

Occasionally, PCS can be due to upstream occlusion, such as in cases of nutcracker syndrome or iliac vein compression syndrome. Nutcracker syndrome occurs when the left renal vein is compressed between the aorta and SMA resulting in increased pressure and subsequent reflux into the left gonadal vessel (see Chap. 29 for more information). In iliac vein compression syndrome, also known as May-Thurner syndrome, the left common iliac vein is compressed by the right common iliac artery. The left pelvic veins may then develop reflux and flow proceeds through cross-pelvic collaterals to the right internal iliac vein. Rarely, PCS can be secondary to congenital vascular malformations [12].

Clinically, patients are typically multiparous women of childbearing age who present to clinic with pelvic pain. They describe the pain as aching or heaviness in their pelvis worse in the standing position. The pain is exacerbated just before their menstrual period. Commonly they will describe feeling best in the morning, and as they move around, the pain/heaviness will continually increase. Many will describe increasing pain if they are sitting for long periods of time, either at work or on long car or airplane rides. Pain during intercourse is not a common complaint, but they do complain of pain which occurs following intercourse. Postcoital pain is particularly severe if they have intercourse in the morning.

Physical exam may be unrevealing; however a history of unusual varicosities such as gluteal, labial, and/or vulvar varices supports the diagnosis. Lower extremity varicose veins that originate in the upper inner thigh can be supplied by pudendal vein collaterals. Some patients can experience ovarian point tenderness. The combination of a history of postcoital pain and ovarian point tenderness is 94% sensitive and 77% specific for PCS [13].

Clinical Indication

The diagnosis of pelvic congestion syndrome is made through imaging as abnormal physical exam findings or lab values may be absent. Transvaginal ultrasound is typically the first-line imaging. Typically, a patient's primary gynecologist has already ordered a pelvic sonograph to rule out neoplasm and other ovarian or uterine pathologies. Sonographic diagnostic criteria include tortuous pelvic and ovarian veins which enlarge during Valsalva maneuver. Normal pelvic veins are straight tubular structures with a diameter of less than 4 mm [14]. There may be dilated arcuate veins in the myometrium. Slowed or reversed blood flow and polycystic changes of the ovaries may be identified. Polycystic ovaries are associated with PCS in 56% of cases, although these patients do not typically have hirsutism and amenorrhea [13].

Table 13.1 Imaging diagnostic criteria for pelvic congestion syndrome

Noninvasive imaging criteria	Venography criteria
≥4 ipsilateral tortuous parauterine veins	Uterine vein enlargement
Parauterine veins measuring ≥4 mm	Ovarian plexus congestion
Ovarian veins measuring ≥8 mm	Ovarian vein measuring ≥10 mm
Dilated myometrial arcuate veins filling bilateral pelvic varices	Cross-pelvic filling of pelvis, thigh, or vulvovaginal veins

CT and MR diagnostic criteria include the presence of four or more tortuous parauterine veins with diameter of more than 4 mm and ovarian vein diameter greater than 8 mm (Table 13.1, Fig. 13.1) [12]. MR venography has been gaining ground as a diagnostic modality of choice as it demonstrates the venous anatomy while avoiding radiation exposure. MR venography can be used to demonstrate retrograde flow of the ovarian veins [15]. On MR, varices will appear hyperintense on T1-weighted gradient echo sequences. CT and MR may also delineate other etiologies of PCS including compression of the renal or iliac veins as described above. However, both of these cross-sectional imaging techniques are performed with the patient in the supine position, and reflux into the gonadal and pelvic veins is best demonstrated in the standing position. Thus cross-sectional imaging may lead to missed diagnosis in less severe cases.

Key Point
Ovarian varices ≠ PCS, 10% of women with varices are asymptomatic.

The American Venous Forum and Society for Vascular Surgery developed clinical practice guidelines stating that symptomatic patients should undergo noninvasive imaging for evaluation of pelvic venous insufficiency. Ovarian and internal iliac venography should be performed prior to planned intervention and is the gold standard diagnostic test for pelvic congestion syndrome [16]. The first description of venographic visualization of PCS was in 1965 when the tip of the catheter, meant for the left renal vein, happened to lie in the left ovarian vein orifice [17]. In addition to venous diameter, venography can also demonstrate cross-pelvic collaterals on late phase imaging. The procedure is performed through a selected catheterization of the left renal vein with a forceful injection of contrast to evaluate for ovarian venous reflux (Fig. 13.2). Ideally the patient should be studied on a fluoroscopic table which has the capability of being tilted into at least a partial upright position, thus utilizing gravity to help elucidate the patient's pathology.

Treatment should be based on definitive diagnosis and severity of symptoms. Nearly 10% of asymptomatic women

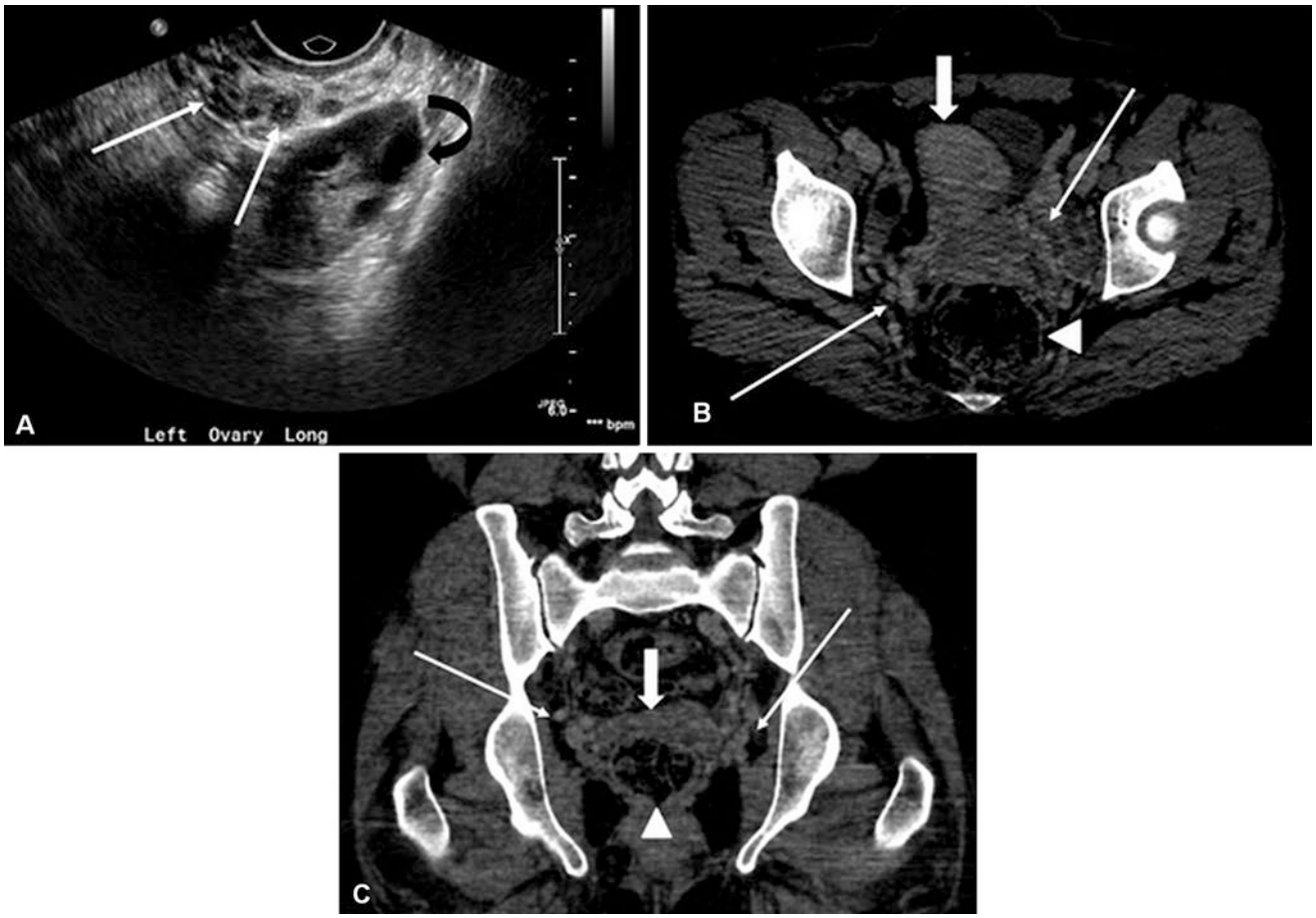

Fig. 13.1 33-year-old female with vulvar varicosities and pelvic heaviness following pregnancy. (**a**) Transvaginal ultrasound demonstrates multiple tubular structures (thin arrows) consistent with ovarian varices. The ovary demonstrates multiple simple cysts (curved black arrow). Axial (**b**) and coronal (**c**) views through the pelvis with IV contrast demonstrate extensive pelvic varicosities (thin arrow). The uterus (thick arrow) and rectum (arrowhead) are also visualized

Fig. 13.2 Consecutive venography pictures demonstrate (**a**) a sheath within the left renal vein (arrow head) with a catheter advanced through a collateral vessel off of the left ovarian vein. Extensive collateral vessels are noted (thin arrows). (**b**) Filling of the distal ovarian vein collaterals (thin arrow) result in rapid shunting of contrast into the dilated left internal iliac vein (thick arrow). (**c**) Extensive cross-pelvic collateral vessels (thin arrow) can be seen

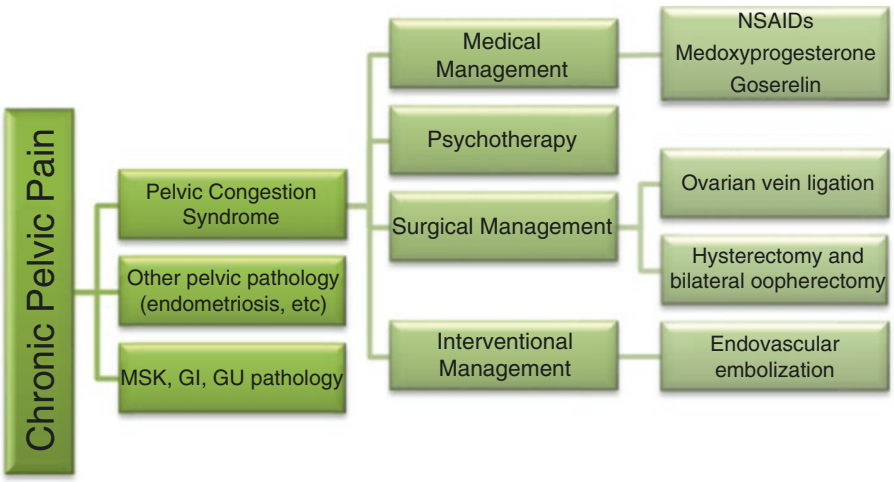

Fig. 13.3 Chronic pelvic pain treatment algorithm

can have ovarian varices, and therefore it is important to exclude other pathologies as the cause of the patients' pain (Fig. 13.3) [7]. Chronic unremitting undiagnosed pain such as that caused by PCS can often lead to depression. Frequently, these patients can experience incomplete relief of their symptoms if there are several pathologic causes to their pain. When venous incompetence is the sole contributing factor of their symptoms however, patients can do extremely well following treatment.

Key Point
Ovarian and internal iliac venography is the gold standard for PCS diagnosis.

Conventional Therapy

Historically, pelvic congestion syndrome was treated with hysterectomy and commonly oophorectomy. However, there are nonsurgical options which may be tried prior to performing an invasive procedure. The goal of medical therapy is to suppress ovarian function or cause vasoconstriction of the dilated veins. Medroxyprogesterone acetate and the GnRH analogue goserelin have both been used for the treatment of PCS. Both of these drugs demonstrated mild relief of symptoms with short-term results. When combined with psychotherapy, the effects seemed to last longer [18]. This further demonstrates the link between psychological and somatic symptoms of PCS. Side effects of progestins include weight gain and bloating, while side effects of GnRH analogues include depression, night sweats, and vaginal dryness. Given the limited efficaciousness and side effect profile, medical therapy is not indicated for long-term use.

Surgical treatment for PCS directly addressing the gonadal veins was first described in 1984 with extraperitoneal resection of the left gonadal vein which demonstrated improvement or resolution of symptoms of 73% of patients [19]. With advances in technology, laparoscopic transperitoneal ovarian vein ligation became the mainstay therapy in the late 1990s. A study of 23 patients demonstrated complete resolution of patients' symptoms at 1-year follow-up [20]. Although potentially useful, the procedure is not without its drawbacks. Serious complications include DVT, retroperitoneal hematoma, ileus, and bowel obstruction secondary to adhesions. With the development of interventional transcatheter therapies, this surgical technique has largely been supplanted. In difficult-to-treat patients and patients with recurrent disease, hysterectomy and bilateral oophorectomy are still employed by some gynecologists, but they are less common than previously [21].

Interventional Therapy

Transcatheter embolotherapy of incompetent ovarian veins was first performed in 1993 by Edwards on a single patient who experienced prolonged symptomatic relief of symptoms [22]. Since that time, the technique has become widely available and now is the mainstay for treatment for pelvic congestion syndrome. Numerous studies have demonstrated varying success rates for the reduction of pelvic pain ranging from 47% to 98% [11, 23–25]. Of note, patients with isolated ovarian vein reflux had improved outcomes compared to patients with isolated iliac vein reflux or combined disease [11]. No large studies have been performed to date on outcomes of patients with vulvar and lower extremity varicosities after pelvic embolotherapy. Several small studies reported a more than 80% reduction in vulvar varicosities with limited improvement of lower extremity varicosities after treatment [5]. Ovarian vein embolization is a relatively straightforward outpatient procedure.

Key Point
Contraindication to embolization

- Active infection
- Contrast allergy
- Severe coagulopathy

Once a patient has decided to seek treatment for PCS, preoperative imaging with a venous phased MRI or CT (MRV or CTV, respectively) may be performed. This can both determine the extent of varicosities as well as assist with procedural planning. If the patient's symptoms are very consistent with pelvic congestion syndrome, and she has not had previous cross-sectional images, then some interventional radiologists will proceed directly to venography and embolization. If there is a question as to the diagnosis, then cross-sectional imaging may be helpful, although it is important to recognize that less severe reflux could be missed since the patient will be supine.

The How To

1. Venous access is gained through the femoral, jugular, or arm vein approach.
2. An optional venogram of the IVC can be performed first to establish anatomy. Where possible, venography should be performed with the patient in a reverse Trendelenburg position. A cavogram can demonstrate reflux into the internal iliac veins and usually demonstrate the position of the renal veins.
3. Catheterization of the left renal vein with an injection near the renal hilum to seek spontaneous reflux into an incompetent left ovarian vein. The catheter is then advanced into the left ovarian vein. Injection of contrast will demonstrate a dilated vein with reflux of contrast into the pelvic vasculature.
4. The catheter is advanced down the ovarian vein terminating just above the pelvic brim. Injection of contrast will demonstrate reflux of contrast material into the pelvic veins, cross-pelvic collaterals, and any varices (Fig. 13.4a).
5. Embolotherapy varies based on user preferences and degree of reflux. Numerous agents can be used including glue, coils, sclerosing agents and Gelfoam, or a combination of various embolics [26]; frequently, a sclerosing agent such as sodium tetradecyl sulfate (Sotradecol, SDS) is used, followed by coils (Fig. 13.4b). In order to use a sclerosing agent, the accessed vein may be occluded using a balloon occlusion technique. This can help keep the sclerosing agent from crossing the cross-pelvic collaterals and moving into systemic circulation through the contralateral ovarian vein.
6. In order to perform the balloon occlusion technique, a balloon is advanced into the distal ovarian vein. A microcatheter is advanced distal to the balloon. The balloon is inflated preventing reflux of contrast up around the ipsilateral ovarian or iliac vein.
7. Sclerosing agent is injected into the pelvic veins until near occlusion. Completion of embolization can be demonstrated by increased resistance to further injection of contrast which can be seen as slow flow, stasis, or reflux of contrast.
8. Embolization coils or plugs are used to embolize the ovarian vein prior to deflation of the balloon. The microcatheter and balloon are then retracted halfway in the ovarian vein, and the procedure is repeated with both deployment of SDS and coils. This is typically repeated three times in one ovarian vein before moving to the contralateral side and repeating the process (Fig. 13.4c).
9. Alternatively, one can advance a catheter into the distal ovarian vein, and inject contrast, measuring the amount of contrast it takes to fill the cross-pelvic collaterals and begin to see filling of the internal iliac, and then use an amount of sclerosant that is less than what it took to fill the cross-pelvic collaterals. Another method is to mix the Sotradecol with Gelfoam and air to make a foam slurry and inject this mixture. The Gelfoam helps to hold the Sotradecol into the veins perhaps increasing the contact with the vein wall. Then coil embolization can be performed through the catheter, which allows the use of larger .035 inch coils.
10. There remains debate as to the optimal technique for ovarian vein embolization; however a combination of coils and sclerosis has been demonstrated as the most efficacious through multiple published studies. The decision to treat one or both ovarian veins is likely patient specific. Those with significant reflux bilaterally may benefit more from bilateral embolization, whereas a patient with unilateral dilatation and only moderate varicosities may only need unilateral embolization.
11. If the internal iliac veins demonstrate evidence of reflux, they can be treated with sclerosing agent, coils, or a combination. Some interventionalists feel that refluxing internal iliac veins should be treated at the same session as the ovarian vein(s) treatment, and other interventionalists feel that the ovarian vein(s) should be treated first. They would then perform another procedure with embolization if the patient continues to have symptoms.

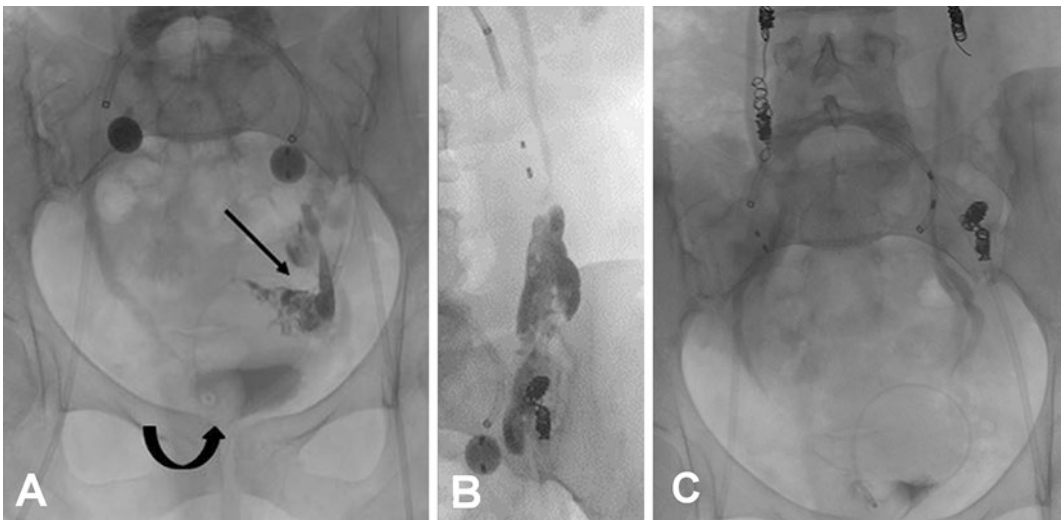

Fig. 13.4 (**a**) Due to the rapid internal iliac vein shunting and extensive cross-pelvic collaterals, the decision was made to obtain bilateral femoral vein access and inflate a balloon in bilateral internal iliac veins. Contrast injection demonstrates decreased shunting (thin arrow) compared to Fig 13.1 without balloon occlusion. A Foley catheter (curved arrow) can be seen with a small volume of contrast within the bladder. (**b**) Gelfoam embolization followed by coiling was performed in the distal left ovarian vein. This was then performed subsequently in a retrograde fashion up the ovarian vein. The same procedure was performed on the contralateral side. (**c**) Following completion of embolization, venography with the balloons deflated demonstrates no contrast filling the ovarian veins

Complications of embolotherapy are thankfully rare but do occur. The major complication is migration of coils into the pulmonary arteries, reported in 2% of patients following internal iliac vein embolization [27]. Migration can also occur from coils deployed in the gonadal vein. This risk is increased in vessels >12 mm. Some people advocate for the use of detachable coils to mitigate this risk. Detachable coils are coils that can be deployed but remain attached to the deployment device. If the coil is felt not to be in satisfactory positioning, it can be retracted and redeployed; once it is in appropriate positioning, it can be detached from the deployment device. To prevent coil migration, the diameter of the coil used should be 30–50% larger than the diameter of the internal iliac vein or the gonadal vein [27]. Another complication includes perforation of the ovarian vein; this is not a serious problem since the vein is being embolized and perforation does not cause significant extravasation. Some patients can experience flank pain, thrombophlebitis, postprocedural fevers, and puncture site hematomas also [25]. Recurrence is rare but has been reported in the literature [28]. When nutcracker syndrome is present concurrently, it may be necessary to relieve compression of the left renal vein as well to assure durable relief of PCS symptoms.

Key Point
Complications of ovarian vein embolization

- Nontarget embolization
- Coil migration
- Vessel perforation

Chronic pelvic pain can be a diagnostic challenge for practitioner and patient alike. In those patients with signs and symptoms of pelvic congestion syndrome, embolization can provide a minimally invasive means of relieving pain and improving quality of life.

References

1. Ahangari A. Prevalence of chronic pelvic pain among women: an updated review. Pain Physician. 2014;17(2):E141–7.
2. Latthe P, Latthe M, Say L, Gülmezoglu M, Khan KS. WHO systematic review of prevalence of chronic pelvic pain: a neglected reproductive health morbidity. BMC Public Health. 2006;6:177.
3. Mathias SD, Kuppermann M, Liberman RF, Lipschutz RC, Steege JF. Chronic pelvic pain: prevalence, health-related quality of life, and economic correlates. Obstet Gynecol. 1996;87(3):321–7.
4. Daniels J, Gray R, Hills RK, Latthe P, Buckley L, Gupta J, et al. Laparoscopic uterosacral nerve ablation for alleviating chronic pelvic pain: a randomized controlled trial. JAMA. 2009;302(9):955–61.
5. O'Brien MT, Gillespie DL. Diagnosis and treatment of the pelvic congestion syndrome. J Vasc Surg Venous Lymphat Disord. 2015;3(1):96–106.
6. Liddle AD, Davies AH. Pelvic congestion syndrome: chronic pelvic pain caused by ovarian and internal iliac varices. Phlebology. 2007;22(3):100–4.
7. Borghi C, Dell'Atti L. Pelvic congestion syndrome: the current state of the literature. Arch Gynecol Obstet. 2016;293(2):291–301.
8. Hobbs JT. The pelvic congestion syndrome. Br J Hosp Med. 1990;43(3):200–6.
9. Ganeshan A, Upponi S, Hon LQ, Uthappa MC, Warakaulle DR, Uberoi R. Chronic pelvic pain due to pelvic congestion syndrome: the role of diagnostic and interventional radiology. Cardiovasc Intervent Radiol. 2007;30(6):1105–11.
10. Perrin MR, Labropoulos N, Leon LR Jr. Presentation of the patient with recurrent varices after surgery (REVAS). J Vasc Surg. 2006;43(2):327–34. discussion 334

11. Asciutto G, Asciutto KC, Mumme A, Geier B. Pelvic venous incompetence: reflux patterns and treatment results. Eur J Vasc Endovasc Surg. 2009;38(3):381–6.
12. Koo S, Fan CM. Pelvic congestion syndrome and pelvic varicosities. Tech Vasc Interv Radiol. 2014;17(2):90–5.
13. Nicholson T, Basile A. Pelvic congestion syndrome, who should we treat and how? Tech Vasc Interv Radiol. 2006;9(1):19–23.
14. Kennedy A, Hemingway A. Radiology of ovarian varices. Br J Hosp Med. 1990;44(1):38–43.
15. Pandey T, Shaikh R, Viswamitra S, Jambhekar K. Use of time resolved magnetic resonance imaging in the diagnosis of pelvic congestion syndrome. J Magn Reson Imaging. 2010; 32(3):700–4.
16. Gloviczki P, Comerota AJ, Dalsing MC, Eklof BG, Gillespie DL, Gloviczki ML, et al. The care of patients with varicose veins and associated chronic venous diseases:clinical practice guidelines of the Society for Vascular Surgery and the American Venous Forum. J Vasc Surg. 2011;53(5 Suppl):2S–48S.
17. Ahlberg NE, Bartley O, Chidekel N. Circumference of the left gonadal vein. An anatomical and statistical study. Acta Radiol Diagn (Stockh). 1965;3(6):503–12.
18. Farquhar CM, Rogers V, Franks S, Pearce S, Wadsworth J, Beard RW. A randomized controlled trial of medroxyprogesterone acetate and psychotherapy for the treatment of pelvic congestion. Br J Obstet Gynaecol. 1989;96(10):1153–62.
19. Rundqvist E, Sandholm LE, Larsson G. Treatment of pelvic varicosities causing lower abdominal pain with extraperitoneal resection of the left ovarian vein. Ann Chir Gynaecol. 1984; 73(6):339–41.
20. Gargiulo T, Mais V, Brokaj L, Cossu E, Melis GB. Bilateral laparoscopic transperitoneal ligation of ovarian veins for treatment of pelvic congestion syndrome. J Am Assoc Gynecol Laparosc. 2003;10(4):501–4.
21. Smith PC. The outcome of treatment for pelvic congestion syndrome. Phlebology. 2012;27(Suppl 1):74–7.
22. Edwards RD, Robertson IR, MacLean AB, Hemingway AP. Case report: pelvic pain syndrome--successful treatment of a case by ovarian vein embolization. Clin Radiol. 1993;47(6):429–31.
23. Kwon SH, JH O, Ko KR, Park HC, Huh JY. Transcatheter ovarian vein embolization using coils for the treatment of pelvic congestion syndrome. Cardiovasc Intervent Radiol. 2007;30(4):655–61.
24. Venbrux AC, Chang AH, Kim HS, Montague BJ, Hebert JB, Arepally A, et al. Pelvic congestion syndrome (pelvic venous incompetence): impact of ovarian and internal iliac vein embolotherapy on menstrual cycle and chronic pelvic pain. J Vasc Interv Radiol. 2002;13(2 Pt 1):171–8.
25. Kim HS, Malhotra AD, Rowe PC, Lee JM, Venbrux AC. Embolotherapy for pelvic congestion syndrome: long-term results. J Vasc Interv Radiol. 2006;17(2 Pt 1):289–97.
26. Lopez AJ. Female pelvic vein embolization: indications, techniques, and outcomes. Cardiovasc Intervent Radiol. 2015;38(4):806–20.
27. Yamasaki W, Kakizawa H, Ishikawa M, Date S, Tatsugami F, Terada H, et al. Migration to the pulmonary artery of nine metallic coils placed in the internal iliac vein for treatment of giant rectal varices. Acta Radiol Short Rep. 2012;1(6):1–4.
28. Freedman J, Ganeshan A, Crowe PM. Pelvic congestion syndrome: the role of interventional radiology in the treatment of chronic pelvic pain. Postgrad Med J. 2010;86(1022):704–10.