

# Current Clinical Management of Pelvic Congestion Syndrome

David A. Greuner, MD, and Danielle DeMarco

**Abstract.** After an extensive review of the current literature on the etiology, diagnosis, and treatment modalities for pelvic congestion syndrome (PCS), we have found that the issue of underdiagnosis can likely be attributed to the lack of a standard protocol for the clinical management and treatment of the disease. There have been consistencies across various studies, including accurate diagnostic testing and a successful treatment method, and it is our goal to propose a clear algorithm for the diagnosis and treatment of PCS, as well as to highlight the importance of differentiating between types of PCS when deciding on an appropriate treatment plan.

VASCULAR DISEASE MANAGEMENT 2020;17(2):E23-E28.

**Key words:** Pelvic congestion syndrome, pelvic congestion, chronic pelvic pain, pelvic venous insufficiency, venous embolization, contrast venography, ovarian veins, gonadal veins, iliac veins

Pelvic congestion syndrome (PCS) is a misunderstood and often underdiagnosed cause of chronic pelvic pain (CPP) in women of child-bearing age. CPP is estimated to account for up to 40% of all gynecological visits, yet an estimated 51% to 55% of women with pelvic pain remain untreated for years without receiving an accurate diagnosis.<sup>1</sup> Although PCS was previously assumed to be a rare disorder, its prevalence seems to be significantly greater than what was once assumed. More women with once unexplained CPP are being diagnosed with PCS, and this increase in diagnosis is likely driven by increased awareness of the syndrome during recent years.<sup>1</sup>

Despite the increase in the detection of PCS, there are still differences in opinion among members of the medical community regarding the best protocols for diagnosis and treatment. We believe that the existence of a clear, standard approach to the diagnosis and treatment of PCS would even further reduce the number of women who experience CPP.

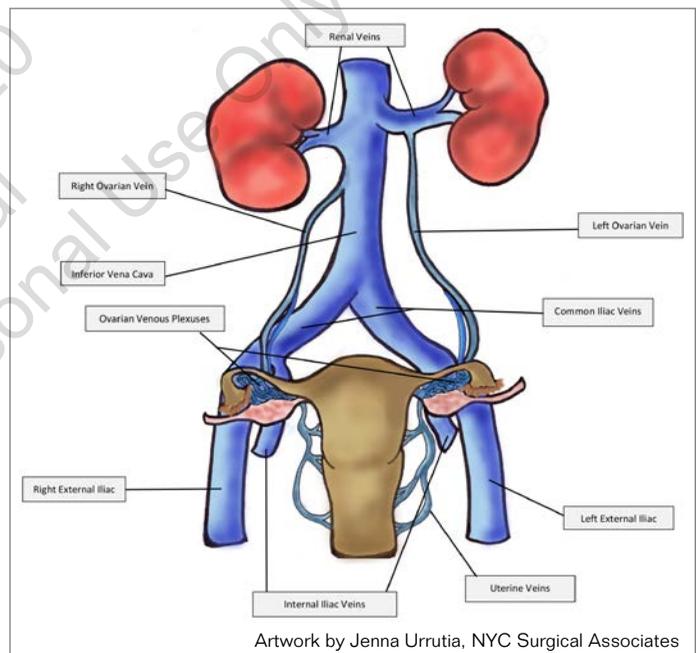
## METHODS

A literature review was performed using PubMed, EMBASE, UpToDate, and the Stony Brook University Library online database. The goal was to find all of the necessary information known about the diagnosis and treatment of PCS.

Articles were sorted by evidence level in order to assess their reliability, using the Oxford Centre for Evidence-Based Medicine (OCEBM)<sup>2</sup> Table of Evidence as a general guide. Forty-three articles were selected and sorted by article type (diagnosis-focused or treatment-focused) for further detailed evaluation by their title, abstract, and key words.

## CLINICAL FEATURES AND DIAGNOSIS

**Anatomy.** The major vessels involved in venous drainage of the



**Figure 1.** Female pelvic venous anatomy.

female pelvic area (**Figure 1**) are the inferior vena cava (IVC), left and right common iliac veins, left and right external iliac veins, left and right internal iliac veins, left and right ovarian veins, left renal vein, right renal vein, and the left and right uterine veins. Venous blood flows into bilateral common iliac veins from the internal iliac veins and their tributaries, the uterine, ovarian, and vesicovaginal plexuses. The bilateral common iliac veins and bilateral renal veins all drain directly into the IVC. The right ovarian vein also drains directly into the IVC, while the left ovarian vein drains into the left renal vein.

**Types of PCS.** Although the precise etiology of PCS remains

unclear, nearly all of the current literature suggests that the disease is multifactorial. There are additional consistencies across studies regarding the possible presentation. However, only a small number of studies make specific mention of the different types of PCS.

Within the literature that does mention different types of PCS, authors do not agree on the exact number of types, and there are no clear definitions for each type. Articles mention anywhere from 2 to 5 types of PCS, and this range of types may add to the challenge in diagnosis.<sup>1,3,4</sup> To allow for a simple diagnostic and treatment protocol, we believe that the most logical solution would be to separate symptomatic PCS into two separate categories: primary and secondary.

Primary PCS is caused by pelvic venous insufficiency (PVI), which may be defined as an incompetence of ovarian veins and/or internal iliac veins.<sup>1,3</sup> It is likely that hormones, specifically estrogen acting as a vasodilator, play a role in weakening of the veins. High levels of estrogen cause an increase in the release of nitric oxide, which leads to the relaxation of smooth muscle. This smooth muscle relaxation may be a key contributor to vascular incompetence.

Secondary PCS is caused by extrinsic compression of the pelvic veins, resulting in obstructed outflow. It is often related to May-Thurner syndrome (MTS) or to compression of the left external iliac vein by the left external iliac artery, known as Nutcracker syndrome (NCS). Secondary PCS may also be related to compression of the left renal vein between the aorta and the superior mesenteric artery.

**Presentation.** Symptoms that have most consistently been found to accompany PCS include:<sup>1,3-4,3</sup>

- Pelvic pain lasting at least 6 months, usually described as a dull ache or heaviness in the lower abdominal/pelvic region;
- Presence of visible or non-visible pelvic varices;
- Dysmenorrhea;
- Dyspareunia;
- Bladder irritability and/or urgency;
- Worsening of symptoms after prolonged periods of standing;
- Lower-extremity varices;
- Back pain and/or leg pain.

It has also been reported that symptoms become more pronounced during and after pregnancy, and their severity may increase after each additional pregnancy.<sup>3,5-10</sup>

The limited knowledge and underdiagnosis of PCS can be attributed to both its atypical presentation of symptoms, and the complexity of the female pelvic-venous anatomy.

Some of the differential diagnoses include pelvic inflammatory disease, endometriosis, interstitial cystitis, adhesions, malignancy, adenomyosis, irritable bowel disease, and uterine fibroids.<sup>1,2,5-7,11-14</sup> The broad range of possible symptoms and atypical presentation often causes PCS to only be considered after all other differential diagnoses have been ruled out. While PCS shares many symptoms with other gynecologic, urologic,

and musculoskeletal disorders, it is possible that the exact location of symptoms may not be the key to diagnosis. If patients present with any of the common symptoms of PCS, primary care physicians and gynecologists may find it beneficial to inquire about the postural nature of those symptoms. Although more research needs to be done on the subject, it is likely that an increase in symptom severity due to postural changes is a strong indicator for PCS.<sup>5,10,13,14</sup>

**Non-invasive diagnostic imaging.** There are several types of non-invasive diagnostic examinations currently available that may be used to accurately predict the presence of PCS. Pelvic venous duplex ultrasound (DUS) using the Valsalva maneuver to accentuate venous filling seems to be the best option for the selection of patients who should be sent for further testing.

The diagnostic criteria for detection of PCS using DUS includes:<sup>1,8,15,16</sup>

- Tortuous pelvic veins measuring >6 mm in diameter;
- Dilated arcuate veins crossing the myometrium and communicating with bilateral pelvic varicosities;
- Reverse caudal or retrograde blood flow;
- Slow blood flow <3 cm/s.

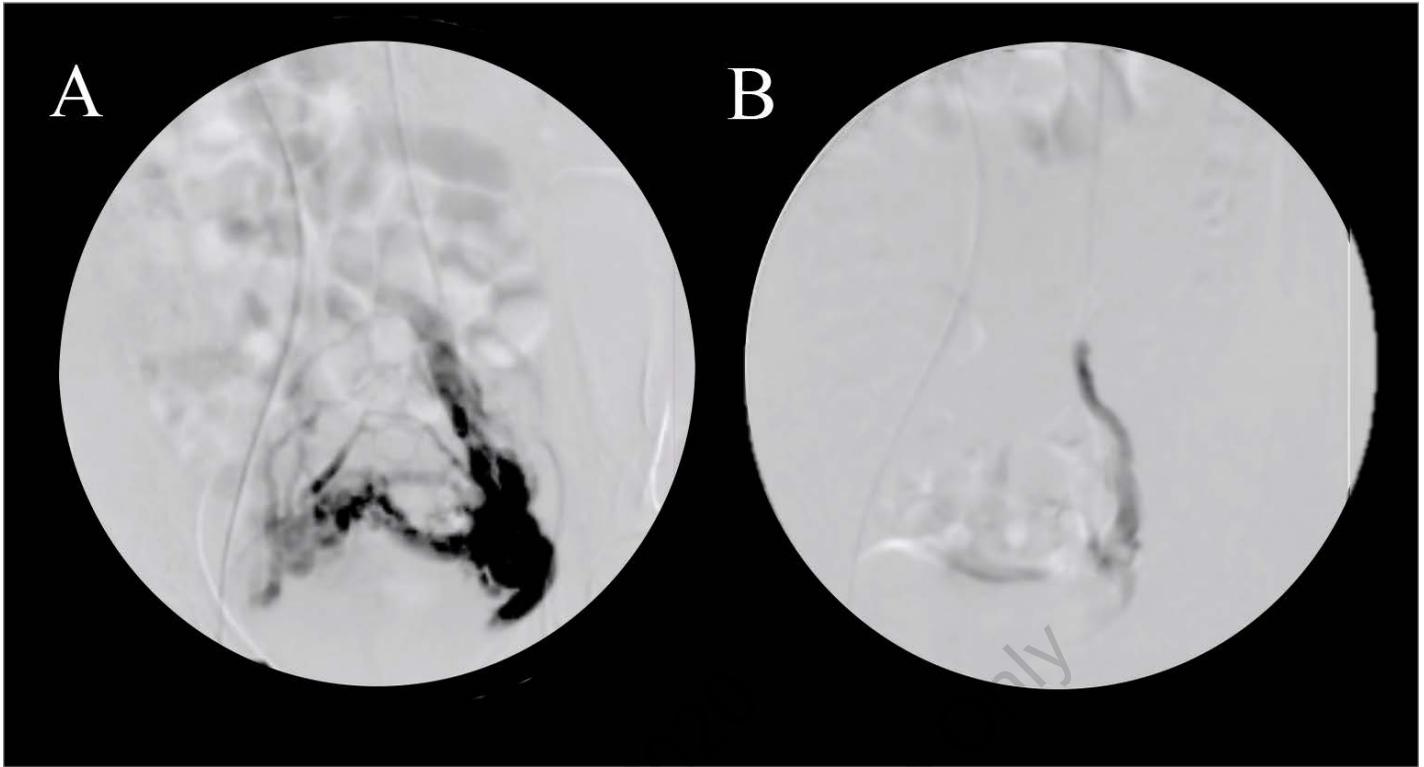
Some have advocated for the use of transvaginal ultrasound (TVUS) over DUS. While there is a high probability of visualizing the peri-uterine veins using TVUS, the examination is limited in its ability to follow the pathways of the ovarian veins and to visualize possible higher compressions/obstructions that may indicate the presence of stenotic lesions (MTS and NCS).<sup>17</sup>

Diagnostic testing that uses radiation, such as computed tomography, is not recommended as the first choice for patients (especially patients of child-bearing age) when there are other accurate modalities available. Research suggests that the findings most indicative of PCS include dilated ovarian veins and the presence of reverse caudal blood flow that can be seen using DUS.<sup>1,18,19</sup> The ability to take images using the Valsalva maneuver in order to accentuate blood flow, combined with its accuracy and cost-efficiency, make DUS the most reasonable choice when compared to other examinations such as magnetic resonance imaging and magnetic resonance venography.

**Invasive diagnostic imaging.** After symptomatic patients are screened using DUS, those who meet the criteria may be sent for contrast venography, which has been widely accepted as the gold standard in diagnosing PCS. The literature states that access may be achieved through either the femoral or jugular veins. However, in our experience, using the saphenous vein for access has resulted in less incidence of both hematoma and post-operative bleeding, given the compressibility of the vessel. Thus, the saphenous vein is our primary source of access.

The diagnostic criteria for PCS using contrast venography vary slightly across studies.<sup>6,9</sup> However, the majority of current articles describe the criteria for PCS as having one or more of the following:<sup>1,3,6,9,20</sup>

- Incompetent pelvic veins measuring >5-10 mm in diameter;
- Moderate-severe engorgement of the ovarian plexus



**Figure 2A–B.** Pre and post embolization. Left gonadal vein embolization of female patient with PCS. **A)** Angiographic image pre-Gelfoam (Pfizer) embolization. **B)** Angiographic image post-Gelfoam embolization.

- Venous reflux indicated by (slow) proximal injection into the ovarian veins with distal filling of the ovarian venous plexus;
- Filling of veins across the midline;
- Filling of vulvar or thigh varicosities.

During diagnostic venography, it is important to perform a full evaluation in order to find all possible venous pathologies that may be contributing to symptoms. Full evaluation includes visualization of the 1) internal and external iliac system, 2) bilateral ovarian veins, and 3) left renal vein.

### TREATMENT

Medical management of PCS is an option for treatment with the use of drugs such as medroxyprogesterone acetate (MPA) and gonadotropin-releasing hormone (HnRH)]. However, the use of those drugs is not a favored long-term method due to the high reports of symptom reoccurrence after 6 months, possibility of adverse events, and inability to reduce symptoms caused by secondary PCS.<sup>9,12,13,15</sup>

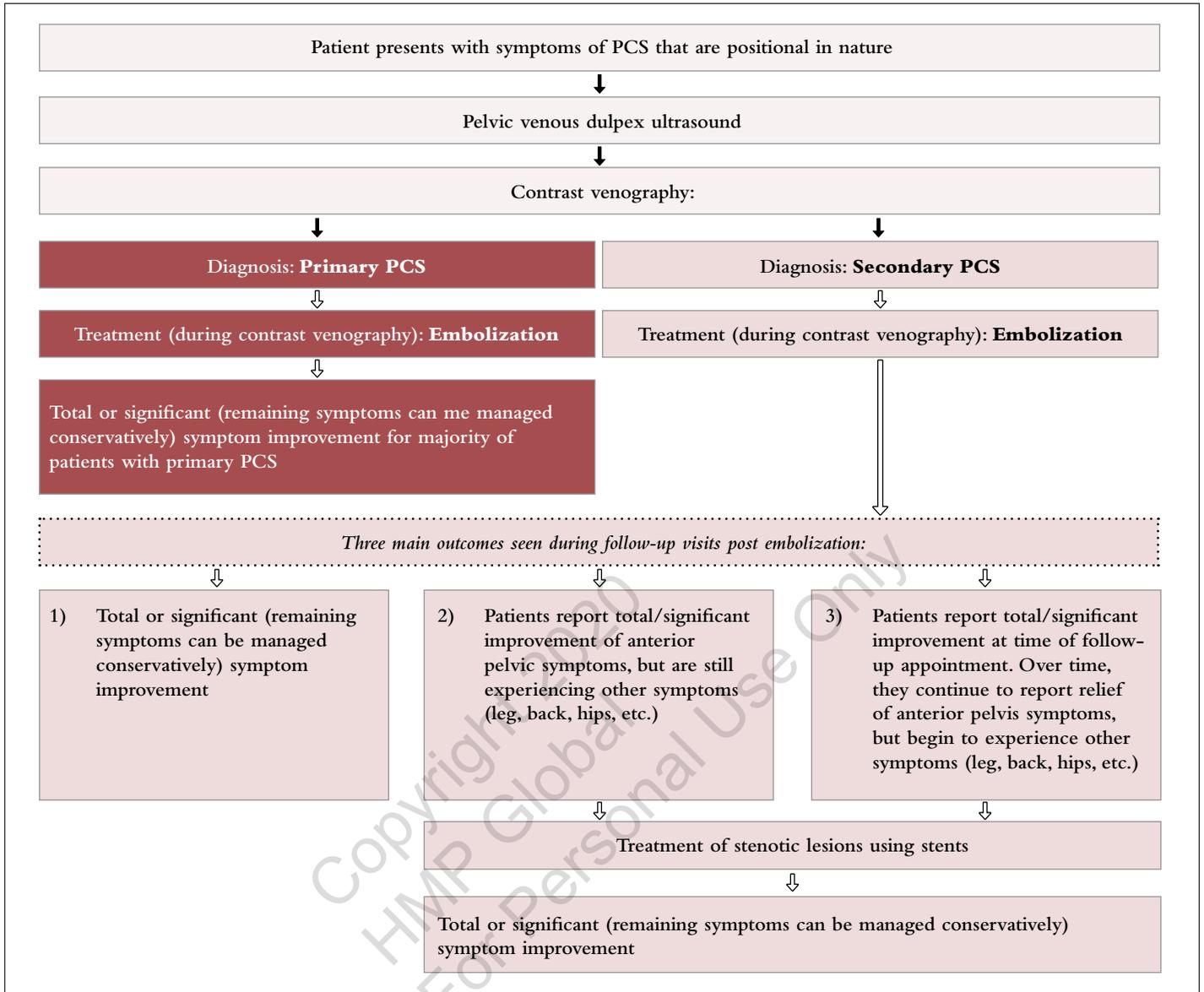
There are several traditional and laparoscopic surgical options with reported success in management of PCS symptoms, but these surgeries often involve the removal of uninvolved bystander structures and the risk of surgical complications and possible fertility issues, which is why endovascular treatment has been the most agreed-upon treatment for PCS.<sup>12,21,22</sup> Trans-

catheter embolization is a safe and reliable treatment modality for PCS that may conveniently be performed at the same time as diagnostic venography. During transcatheter embolization, a catheter is advanced to the ovarian and/or hypogastric vein, and the symptomatic vessel is embolized using Gelfoam (Pfizer) (Figure 2), sclerotherapy, coil, or a combination of materials.

The reported success rate of transcatheter embolization for the treatment of PCS varies due to the wide range of inclusion parameters, variable measurement strategies, and amount of time that constitutes “long-term,” with studies reporting anywhere from 47% to 100% of patients experiencing significant symptom relief 18 months to 5 years post embolization.<sup>1,11,21,23,24</sup>

Complications with embolization are not common, but possible risks include coil migration, vessel perforation, local phlebitis, deep-vein thrombosis, occlusion of non-target vessels, and reaction to a contrast agent.<sup>6,9,14,15,21-24</sup> While there have been no studies comparing different embolization materials, the incidence of coil migration has been reduced over recent years, since more providers have been choosing to use other methods such as Gelfoam or sclerotherapy.<sup>5</sup>

The majority of the current literature does not differentiate between primary and secondary PCS when discussing treatment, but it is important to remember that embolization alone may not be successful in reducing all symptoms in patients with stenotic lesions resulting from MTS or NCS. Greiner and colleagues<sup>7</sup> highlight the importance of looking into the specifics of the pathophysiology of each case of PCS before deciding on an appropriate treatment plan.



**Figure 3.** Diagnosis and treatment algorithm for PCS.

While patients with secondary PCS may feel relief post embolization, some may experience reoccurrence of symptoms, or they may develop new symptoms if stenotic lesions remain untreated. It may be necessary to perform an additional contrast venography with intravascular ultrasound (IVUS) to measure any lesions caused by MTS and/or NCS, and to treat the stenosis with stent placement in order to relieve all remaining/new symptoms.

Although the research is limited, some studies have been done on stenting and angioplasty of iliac veins to improve symptoms of PCS. The results of these studies imply that embolization and stenting together are more effective in the treatment of secondary PCS than embolization alone.<sup>4</sup> Daughtry and Gillespie<sup>41</sup> reported that treatment with stents and angioplasty resulted in complete relief of pelvic pain, dyspareunia, and left lower-extremity pain in 79%, 82%, and 87% of patients, respectively.

**DISCUSSION**

While there have been significant advances in recent years, lack of a standard protocol for diagnosis and treatment of PCS remains an issue. After a thorough review of the literature, we propose a simple algorithmic approach for clinical management of PCS (Figure 3).

We would like to highlight the importance of remembering the different types of PCS during diagnosis and treatment. There is a chance that the atypical presentation of symptoms may be attributed to the specific type of PCS (primary vs secondary), which is why the positional nature of symptoms may be a better indicator for PCS than the specific symptom location.

It is also possible that a large number of patient reports of symptom reoccurrence or presentation of new symptoms post embolization are due to failure to treat all of the symptom-causing venous pathologies in patients with secondary PCS. ■

*Disclosure: The authors report no financial relationships or conflicts of interest regarding the content herein.*

*Manuscript submitted August 30, 2020; accepted September 24, 2020.*

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