# Pelvic Congestion Syndrome: Etiology of Pain, Diagnosis, and Clinical Management

Darci Phillips, MD, PhD, Amy R. Deipolyi, MD, PhD, Richard L. Hesketh, MB, BCh, MPhil, Mehran Midia, MD, FRCPC, and Rahmi Oklu, MD, PhD

## ABSTRACT

Pelvic congestion syndrome is associated with pelvic varicosities that result in chronic pelvic pain, especially in the setting of prolonged standing, coitus, menstruation, and pregnancy. Although the underlying pathophysiology of pelvic congestion syndrome is unclear, it probably results from a combination of dysfunctional venous valves, retrograde blood flow, venous hypertension, and dilatation. Asymptomatic women may also have pelvic varicosities, making pelvic congestion syndrome difficult to diagnose. This article explores the etiologies of pain, use of imaging techniques, and clinical management of pelvic congestion syndrome. Possible explanations for the spectrum of pain among women with pelvic varicosities are also discussed.

## ABBREVIATIONS

CGRP = calcitonin gene-related peptide, CPP = chronic pelvic pain, IVC = inferior vena cava, PCS = pelvic congestion syndrome

Chronic pelvic pain (CPP) affects approximately one third of all women and accounts for 20% of outpatient gynecology appointments (1). The causes of CPP are varied and can involve endometriosis, pelvic inflammatory disease, pelvic varicosities, and many other conditions (**Table 1**). Even with extensive diagnostic testing and exploratory laparoscopic studies, the etiology of CPP generally remains elusive. Pelvic congestion syndrome (PCS) occurs when varicose veins develop around the ovaries in a setting of CPP. Similar to varicose veins in the legs, pelvic varicosities are thought to result from a combination of dysfunctional venous valves, retrograde blood flow, and venous engorgement. Congested pelvic veins can be very painful and account for approximately one third of cases of CPP (1). Although enlarged pelvic veins and pain are the hallmark features of PCS, asymptomatic women also have been found to have pelvic varicosities (2–4), making PCS a challenging disorder to diagnose. Within this context, the clinical efficacy of ovarian vein embolization for treating PCS has yielded mixed results suggesting that there is a clear imperative to identify patient subpopulations for which endovascular intervention would be most beneficial. In this article, we explore the mechanisms by which pelvic varicosities can lead to pelvic pain, the imaging criteria used to confirm venous dilatation, and the clinical management of PCS.

## CLINICAL MANIFESTATIONS OF PCS

PCS typically affects multiparous women of reproductive age. Most women present with noncyclic lower abdominal or pelvic pain, usually described as a dull ache or fullness that persists for > 6 months. This pain is often exacerbated by prolonged standing, coitus, menstruation, and pregnancy. Associated symptoms are nonspecific and include headache, bloating, nausea, vaginal discharge, vulvar swelling, feeling of leg fullness, lower backache, rectal discomfort, urinary urgency, generalized lethargy, and depression. Some women with PCS present with minimal to no pelvic pain but instead have progressive hip pain (5), lower extremity varicose veins (6), or persistent genital arousal (7) as the sole manifestation. Most commonly, varicose veins of the vulva, perineum, buttocks, and lower extremities are found on physical examination. One study found that the combination of

From Harvard Medical School (D.P., A.R.D., R.O.), and Department of Imaging, Division of Interventional Radiology (A.R.D., R.O.), Massachusetts General Hospital, 55 Fruit Street, 290 Gray/Bigelow, Boston, MA 02114; West Middlesex University Hospital (R.L.H), London, United Kingdom; and Division of Interventional Radiology (M.M.), McMaster University Medical Center, Hamilton, Ontario, Canada. Received September 25, 2013; final revision received January 14, 2014; accepted January 24, 2014. **Address correspondence to** R.O.; E-mail: roklu@mgh.harvard.edu

None of the authors have identified a conflict of interest.



*J Vasc Interv Radiol 2014; 25:725–733*

http://dx.doi.org/10.1016/j.jvir.2014.01.030

**Table 1**. Differential Diagnosis of Chronic Pelvic Pain

| | |
|---|---|
| Gynecology | Gastroenterology |
| Endometriosis | Irritable bowel syndrome |
| Chronic pelvic inflammatory disease | Inflammatory bowel disease |
| Pelvic varicosities (pelvic congestion syndrome) | Diverticular disease |
| Fibroids | Chronic constipation |
| Ovarian cysts | Hernia |
| Adhesions | Hematology/oncology |
| Uterine prolapse | Cancer or metastases |
| Adenomyosis | Porphyria |
| Urology | Musculoskeletal |
| Interstitial cystitis | Pelvic floor myalgia |
| Recurrent urinary tract infections | Myofascial pain (trigger points) |
| Urethral diverticulum | Piriformis syndrome |
| Neurology | Psoas inflammation |
| Neuralgia of ilioinguinal, genitofemoral, or pudendal nerves | Sacroiliac joint inflammation |
| Neuropathic pain | Hip joint pathology |
| Herniated nucleus pulposus | Fractured coccyx |
| Abdominal epilepsy/migraine | Fibromyalgia |
| Psychiatry | |
| Major depression | |
| Somatization | |
| Sleep disorders | |
| Physical, sexual, or substance abuse | |

ovarian point tenderness and a history of postcoital pain is 94% sensitive and 77% specific for PCS (8).

## RISK FACTORS FOR PCS

A combination of environmental, anatomic, and genetic risk factors contributes to the pelvic varicosities associated with PCS. Environmental factors include pregnancy, previous pelvic surgery, estrogen therapy, obesity, phlebitis, and engaging in careers that involve prolonged standing or heavy lifting. During pregnancy, pelvic vein capacity increases by 60% owing to the mechanical compression of the gravid uterus and the vasodilator action of progesterone (9). This venous distention persists for months after delivery and can render the venous valves incompetent, leading to venous hypertension and retrograde flow. Additionally, the weight gain and positional changes of the gravid uterus that occur during pregnancy can cause kinking of the ovarian veins and subsequent venous congestion.

Anomalies in pelvic venous anatomy also contribute to the development of PCS (**Fig 1**). In a normal individual, the ovarian veins originate from the pampiniform venous plexus in the broad ligament and communicate with the uterine plexus. The right ovarian vein drains into the inferior vena cava (IVC),

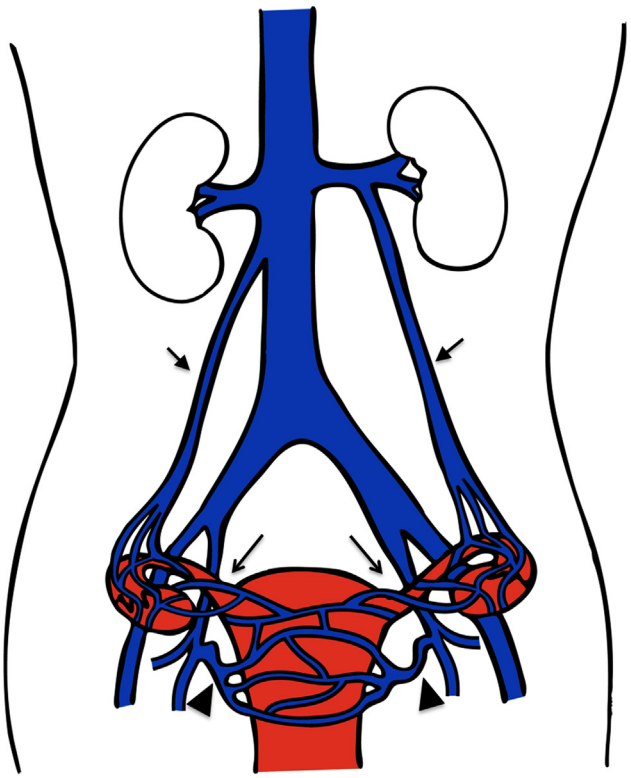

**Figure 1.** Gonadal veins (short arrows), uterine venous plexus (long arrows), and uterine veins (arrowheads) originating from the internal iliac veins. (Available in color online at *www.jvir.org*.)

whereas the left ovarian vein drains into the left renal vein. These veins are usually 3–4 mm in diameter. A rich anastomotic venous plexus is responsible for draining the pelvic viscera, including connections between ovarian, uterine, vulvar, rectal, vesicle, and upper thigh venous systems. Valves are absent from ovarian veins in 15% of women and incompetent on the left and right in 40% and 35%, respectively (10). Only 10% of internal iliac veins have valves (11), leading to a degree of reflux in normal, healthy individuals.

In PCS, the left ovarian vein is most commonly dilated, presumably because it joins the left renal vein at a right angle facilitating reflux. When the right ovarian vein is affected, its junction with the IVC is usually anomalous (4). PCS has also been associated with mechanical compression that leads to obstruction in draining veins, including nutcracker syndrome (13) and May-Thurner syndrome (1). Additionally, obstruction of flow (ie, IVC thrombosis) or external forces (ie, endometriosis, fibroids, postsurgical or infectious adhesions) can increase ovarian vein pressure and subsequently cause reflux. Hypervascular pelvic tumors, including uterine leiomyomas, gestational trophoblastic neoplasms, ovarian solid tumors, and mesenteric tumors, can also cause compression and increase pelvic venous return via collateral vessels (14).

Although a genetic basis for PCS has not been established, the fact that this syndrome affects young