

# Editorial

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## PELVIC CONGESTION SYNDROME

**Abstract:** Pelvic congestion syndrome (PCS) is a common but poorly understood gynecological disorder characterized by noncyclical dull aching lower abdominal or pelvic pain usually in multiparous females in reproductive age group as a result of pelvic vein varicosities. This pain gets intensified during prolonged standing, menstruation and pregnancy due to congestion and dilatation of pelvic veins.

Transvaginal ultrasonography (TVS) with Doppler of pelvic veins is the investigation of choice for diagnosis of PCS and transcatheter embolization of the affected veins (generally ovarian vein) is the effective treatment for PCS.

**Keywords:** Embolization, Pelvic congestion syndrome (PCS), Pelvic venous reflux (PVR), Transvaginal USG (TVS), Varicosities

## Introduction

Chronic pelvic pain (CPP) is a common gynecological complaint among young women and accounts for 10–40% of OPD visits out of which 30% cases are attributed to PCS which is often ignored and misdiagnosed by the clinicians.<sup>1,2</sup> So, establishing the etiology of CPP is very important but is still a diagnostic challenge as it may be due to varied disorders related to gynecological as well as non-gynecological causes.

Pelvic congestion syndrome is a complex disorder defined as CPP resulting from reflux or obstruction of pelvic veins leading to pelvic varicosities involving gonadal, gluteal or periuterine veins. Patients with PCS may present with wide range of symptoms and signs leading to a battery of investigations and thus making the treatment difficult. It also has a great impact on the mental and social health of the young females affecting their daily activities.

## Clinical Features

Most women with PCS present with noncyclical dragging type of dull aching lower abdominal or pelvic pain that persist for more than six months which gets aggravated on sitting, or standing position and gets relieved on lying down particularly when the legs are elevated.<sup>3</sup> Pelvic congestion syndrome may also be associated with many nongynecological symptoms like, rectal discomfort, irritable bowel and bladder syndrome as pelvic varicose veins surrounds these structures. Patient may also present with vaginal discharge, vulval swelling, pain in lower extremities, lower backache, groin pain, gastrointestinal upset and sometimes psychological distress.<sup>4</sup> Very rarely, the sole manifestation may be persistent genital arousal and progressive hip pain mimicking osteoarthritis.<sup>5–7</sup> Though the symptoms of PCS are highly subjective in nature, the signs of PCS are objective. Varicosities around the external genitalia, i.e. vulva, perineum, buttocks, rectum (hemorrhoids) and lower extremities may be found on examination.

## Etiology

The etiopathogenesis of PCS is complex, multifactorial and poorly understood and various theories have been proposed.

- *Ovarian varicocoele:* Absent valves in the ovarian veins especially in the cranial portion and more common on the left side as compared to right side leads to retrograde venous reflux and may explain the development of PCS in nulliparous females.<sup>8,9</sup>
- *Pregnancy:* Ovarian vein flow increases up to 60% during pregnancy which results in venous ovarian vein dilatation and valve incompetence, aggravated by the pressure effect of the gravid uterus and relaxant effect of progesterone on the pelvic venous vasculature. This venous distension may remain persistent for months after delivery and can render the venous valves incompetent leading to retrograde flow, i.e. pelvic vein reflux (PVR).<sup>10</sup> It is more common on left side because the left ovarian veins drain at a sharp angle into the inferior vena cava.

- **Hormonal factors:** Estrogen weakens the veins by inducing venous dilatation through nitric oxide release whereas progesterone damages the venous valves of pelvic vasculature.<sup>1</sup> It is also seen that up to 50% of women with PCS have polycystic ovaries but no other features of polycystic ovarian syndrome.<sup>11</sup> All this suggests that hormones play a significant role in causation of PCS. Anatomically, there are mainly two distinct venous channels that give rise to PCS – the gonadal (ovarian) veins and internal iliac veins and both the venous systems are equally involved in PCS.<sup>12,13</sup>
- **Genetic:** There are some reports which links the association of genes like FOX C2 gene, TIE-2, NOTCH-3 gene, thrombomodulin and type-2 transforming growth factor  $\beta$  receptor<sup>13</sup> in the etiology of varicose vein in PCS.<sup>14,15</sup>

### Pathophysiology

As a result of changes in the valve structure chronic inflammation takes place due to prolonged and progressive venous dilation. This further induces a vicious cycle and promotes endothelial cell injury. This causes leaky valves and significant reflux which results in PCS. The engorgement of veins results in stretching of the intima and stimulation of pain receptors of pelvic venous system with the resultant release of vasoactive substances like substance P and neurokinins A and B responsible for neuropathic pain involved in PCS<sup>16</sup> (Flowchart 1). The supporting evidence for neuropathic pain is that gabapentin and amitriptyline are more efficient in pain management of PCS than opioids and nonsteroidal anti-inflammatory drugs.

Pain in PCS is also contributed by other neurotransmitters like calcitonin gene-related peptide (CGRP), adenosine triphosphate, endothelin, vasopressin and nitric oxide besides substance P as the use of medroxy progesterone acetate relieves the pain by inhibiting their release.<sup>4</sup>

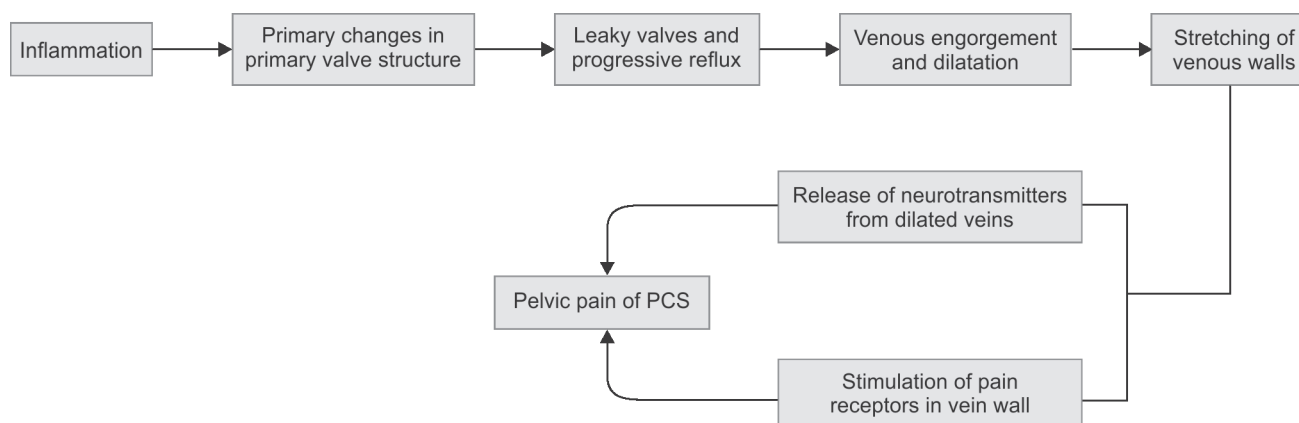
### Investigations

Radiological imaging is important in the evaluation and confirmation of pelvic congestion syndrome in patients presenting with chronic pelvic pain. Noninvasive modalities are recommended as a first-line investigation, however, the gold standard remains selective venography.

### Noninvasive Modalities

- **Ultrasonography:** Transvaginal sonography with color Doppler of pelvic vasculature is first-line preferred modality. It should be performed at 45° head-up position or in standing position or preferably the patient is asked to perform a Valsalva maneuver during the scan so that venous filling gets increased leading to better visualization of pelvic varicosities and also simultaneously providing information regarding pelvic anatomy and coexisting pathologies such as compressive tumours.<sup>12,17</sup> The most common ultrasound criteria used for evaluating PCS in the literature are:<sup>4</sup>
  - Tortuous pelvic veins with a diameter of >6 mm. mean diameter in PCS approximately 8 mm
  - Slow blood flow <3 cm/second or reversed caudal flow in the left ovarian vein shown in Doppler waveforms.

**Flowchart 1:** Pathophysiology of pelvic pain in PCS



- Dilated arcuate veins in the myometrium that communicate between the bilateral pelvic varicose veins.
- Polycystic changes within the ovaries. Features ranged from the typical polycystic ovaries appearance to the presence of clusters of four to six cysts of 5–15 mm in diameter in bilaterally enlarged ovaries.
- *Computed tomography (CT) and magnetic resonance imaging (MRI)*: These modalities have their own limitations as the patient has to be imaged in supine position resulting in missing of less severe cases. Both the imaging techniques demonstrate tortuous, dilated and enhancing tubular structures near the ovaries and uterus due to the PCS varices.<sup>18</sup> MRI phase contrast velocity mapping is preferred over venography if the expertise and facilities are available as the former is noninvasive.

### *Invasive Modalities*

- *Venography*: It remains the gold standard imaging modality for evaluation, confirmation and interventional treatment in patients with PCS. Presence of one or more of the following venographic appearances is suggestive of PCS:<sup>19</sup>
  - Ovarian vein diameter >10 mm
  - Uterine venous engorgement
  - Congestion of the ovarian plexus
  - Filling of the pelvic veins across the midline and/or filling of the vulvovaginal thigh varicosities.It can be done by guiding a catheter to the ovarian or internal iliac veins via jugular or femoral vein and the jugular approach is preferred if embolization has to be done simultaneously. Complications like contrast extravasation, venepuncture site hematoma and vessel injury can occur rarely.
- *Laparoscopy*: It may be useful in visualization of engorgement of the pelvic venous system directly as well as ruling out other causes of CPP associated with endometriosis or adhesions. It may also give false-negative results due to the supine position and intraperitoneal insufflation of pressurized carbon dioxide which is due to possible venous decompression.

## **Treatment**

A multidisciplinary approach should always be considered in evaluation as well as in treatment of patients of PCS with the involvement of gynecology, gastroenterology, the pain team, psychology, and the interventional radiologist.

### *Medical Treatment*

The pelvic pain of PCS may be temporarily ameliorated by pharmacological agents viz. NSAIDs, gabapentin, amitriptyline, medroxyprogesterone acetate (30 mg for 6 months), gonadotropin-releasing hormone analog (GnRH analog) along with add-back therapy (6 months). Both medroxyprogesterone and GnRH agonists have shown subjective and objective improvement of pain associated with PCS as well as reduction in anxiety and stress levels.<sup>1</sup> Recently micronized flavonoid fraction (daflon 500 mg BD for 6 months) has been increasingly used with improvement in pelvic pain scores without any significant side effect by improving the lymphatic drainage and reducing the capillary permeability thereby reducing the venous stasis.<sup>20</sup> Medical management proves to be beneficial for women but there is insufficient evidence on their long-term benefits on checking the enervating symptoms.

### *Surgical Treatment*

Laparoscopic bilateral ligation of ovarian veins transperitoneally has shown reports of complete remission of pain in PCS for up to 12 months along with resolution of pelvic varicosities.

Hysterectomy and bilateral oophorectomy with hormone replacement therapy is another alternative in multiparous females, and has proved to be efficacious in treatment of chronic pelvic pain occurring in PCS.

### *Endovascular Treatment*

Embolization being minimally invasive, superior and very effective, is considered as gold standard therapy for PCS. Transcatheter embolization of ovarian veins, after accessing through femoral or jugular veins, can be done by using metal coils (more commonly), foam sclerosants glue or any of the above combinations with

significant relief of symptoms about 60–100%, has been reported.<sup>23</sup> Complications are rare and mainly coil migration, vessel perforation and local thrombophlebitis.<sup>24</sup>

### Psychotherapy

Cognitive behavioral therapy (CBT) has been proved to be beneficial when combined with surgical and medical management in patients with PCS as it enables the patient to cope up with fear, anxiety and stress.

### Conclusion

PCS is a controversial, unfamiliar clinical entity and is rarely considered in diagnosis of CPP due to lack of diagnostic criteria and acceptable noninvasive tests. Different surgical and medical modalities are available but cognitive behavioral therapy is of cardinal importance in the management of these patients. Pelvic congestion syndrome will continue to exist as a conundrum if well-designed randomized controlled trials are not done to define the optimal diagnosis as well as management of PCS.

**Dr Jaideep Malhotra**  
President FOGSI 2018, India  
Editor in Chief

**Ruchika Garg**  
Associate Professor  
Department of Obstetrics and Gynecology  
SN Medical College, Agra, Uttar Pradesh, India

**Pavika Lal**  
Assistant Professor  
GSVM Medical College, Kanpur, Uttar Pradesh, India

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