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### Pelvic Congestion Syndrome: Current Diagnosis and Management

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#### Authors' contributions

This work was carried out in collaboration between all authors. Author YM designed, wrote the article. Author ME critiqued and applied logical reasoning to the published literature. Author AD located and retrieved scientific information relating to the article. Author RS edited and factually checked article. Author KW contributed to allocating scientifically related materials relating to the article. Author SAK read and approved the final manuscript.

#### Article Information

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**Review Article** 

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#### ABSTRACT

Pelvic Congestion Syndrome (PCS) is a common concern for premenopausal, multiparous women with chronic pelvic pain persisting greater than 6 months. It is defined as observable congestion of pelvic veins due to pelvic varicosities that cause reflux and dilation of ovarian veins, resulting in venostasis [1,2]. Although the etiology is unknown, PCS is associated with anterior, posterior or circumaortic location of left renal vein (vascular compression of the left renal vein between the aorta and the superior mesenteric artery) and Nutcracker syndrome. Another marker of PCS includes the absence of functional ovarian venous valves at the junction of left ovarian vein and left renal vein that increase venous pressure in left ovarian vein causing vulvar varicosities [2-4]. It is important to investigate the pathology, as well as the various diagnostic and therapeutic methods available to effectively manage patients with PCS.

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#### **1. EPIDEMIOLOGY**

The incidence is rather difficult to gather; however, the prevalence can be estimated as up to 30 percent in patients that present with abnormal chronic pelvic pain greater than 6 months with no evidence of inflammatory disease or pathology [5-7]. There is a higher prevalence of PCS associated with venous congestion in multiparous women. There are no documented cases of postmenopausal women with PCS [6,8].

#### 2. PATHOGENESIS

The etiology of PCS is unclear and routine investigations often produce normal findings [9,7]. However, Urologists and Gynecologists report findings of gross dilatation, incompetence, and reflux of ovarian veins that results in venous deficiency of the ovarian veins and/or internal iliac veins causing periovarian pelvic varicosities, similar to testicular varicocele present in males [4,5,10-12,7]. According to the literature, the dilation of ovarian veins is one of the biggest commonalities between all PCS cases. It is documented that chronic pelvic pain is a common symptom for women during their childbearing age [13]. The phenomenon results in left renal venous hypertension that causes the varicoceles [3]. These findings are not always suggestive of PCS and require further investigation, as most women with these findings are asymptomatic [5,14]. It is unclear why so findinas manv women with these are asymptomatic while others present with pain. In a prospective cohort study, 22 patients from ages 19-50 years with lower abdominal pain received sclerotherapy. The result was that 17 out of 20 patients displayed some form of pain relief within a 12-month period, suggesting an associative factor between venous dilatation and chronic pelvic pain [15]. It is common for PCS symptoms to present on the left side due to constriction and congestion of the left renal vein by the aorta and superior mesentery artery (Nutcracker Syndrome) [3,16,17].During pregnancy, the glomerular filtration rate increases by 40 to 65% and renal plasma flow increases by 50 to 85%. As a result, the renal vein is more prone to compression between the angle of superior mesenteric artery and aorta, and may be a contributing factor for PCS [18]. See Fig. 1, 1a, and 1b.



Fig. 1. Pelvic congestion syndrome: tortuous dilated and distended left ovarian vein with extensive anastomosis with pelvic, ovarian, uterine, vaginal and labial veins.Fig.1a. Anterior view of nutcracker syndrome (note: course of the left renal vein in the angle between aorta and superior mesenteric artery; left renal vein is compressed and angulated; risk factor for pelvic congestion syndrome.) Fig. 1b. Lateral view of nutcracker syndrome (note: renal vein between the angle of aorta and superior mesenteric artery)

This leads to a buildup of backpressure towards the ovarian vein due to this critical vascular compression. Left side presentation may also be common on the left simply because the valvular failure of the ovarian vein is more frequent on the left side than the right [3,19,20,15]. There have been no reported findings of PCS in postmenopausal women, which may be correlated with the decline in estrogen levels. Estrogen is a seminal hormone that acts as a venous dilator, who's levels start to decline after menopause. Dilated, tortuous pelvic veins may compress the pudendal nerves, which may explain the precipitation of postural pain.

Thrombophlebitis of the dilated pelvic veins may be responsible for pelvic pain. Past studies show that such pain is associated with endometriosis, PID, postoperative adhesion, uterine disease, and adenomyosis [21]. It is occasionally a mystery because there is no obvious reason for pelvic pain; however, the diagnosis of PCS must be acknowledged because it causes disability and distress for a large population, especially women in the reproductive age group [22]. There is not one risk factor that holds a greater weight than the others. Because the etiology remains undetermined, there are no current prevention measures for PCS.

## 3. SUSPECT DIAGNOSIS OF PELVIC CONGESTION SYNDROME

There are no definitive diagnostic criteria present. However, the following questions may suggest if PCS is present in the clinical diagnosis. Is there chronic pain without evidence of pelvic inflammatory disease or pathology? Does pelvic pain fluctuate in intensity, while standing, premenstrually or during coitus? Does fatigue accompany the pain? Did the pain arise after or during pregnancy? Is there a feeling of fullness of leg veins? Is there an increase in frequency of urination? Is there Perineal heaviness? Is the Uterus retroverted [7]? These questions may lead to the diagnosis of pelvic congestion syndrome. Differential diagnosis of pathology, PCS includes bowel cancer/metastases, endometriosis, fibroids, fibromyalgia, neurologic pathology, orthopedic pathology, ovarian cyst, pelvic inflammatory disorder, porphyria, urologic pathology, and uterine prolapse. [23] In order to distinguish PCS from the aforementioned disorders, emphasis is placed on complaints of persisting pelvic pain exacerbated standing. coitus. bv and premenstruation, and on the following diagnostic

tests: Ultrasound, Computerized Tomography (CT) scan, Magnetic Resonance Imaging (MRI), and Magnetic Resonance Venogram (MRV). MRI and MRV provide the highest sensitivity and optimal imaging [23].

#### 4. CLINICAL MANIFESTATIONS

The first reported case of chronic pelvic pain was documented in late the 1950s and is an overlooked symptom of PCS [24]. The presentation of this pain for greater than 6 months with variable intensity that arises during or after pregnancy with no other existing pelvic pathologies or pelvic inflammation is often indicative of PCS [7]. Peri vuval varices manifest in pregnant patients and later recede after delivery; however, cases are more predominant in later pregnancies [13].

Studies have shown that PCS encounters pathological situations premenstrual of syndrome, intermenstrual syndrome, and chronic pelvic congestion syndrome. Premenstrual syndrome and intermenstrual syndrome are cyclic and include psychological and physical symptoms, while chronic pelvic congestion syndrome includes physical symptoms and etiopathogenic issues [8]. Patients may complain of an increase in fatigue, and/or increase in pain premenstrually, while standing or during coitus. The pain is variable and may feel as a dull ache or can feel like a bursting and may take several hours to subside once onset begins [7]. Observations of atypical varicosities proximal to the pelvic region such as the upper thigh, or the buttocks are suggestive of PCS [25,7]. Testicular varicocele is the common problem for males that is associated with pain with reduced fertility rates [26]. Women who have persisting pain in the lower abdomen most likely have PCS caused by a retrograde flow in incompetent ovarian veins [26,27]. In addition to physical symptoms for male and females, it is suggested that there is a psychological factor that contributes to the disorder [24]. Overall, physical pain in the lower abdomen is a more common and accurate predictor of PCS than psychological symptoms.

#### 5. PHYSICAL EXAMINATION

Bilateral direct palpation must be performed during an abdominal physical examination to examine the ovaries, the cervix and the uterus to determine if tenderness is present in each structure [28]. Based on a recent study that observed tenderness on the abdominal examination with pelvic pain, there was a strong association with PCS and tenderness in the previously stated structures. Symptoms typically affect the lower extremity because of increased venous pressure and subsequent vein enlargement, which presents as aching, swelling, and throbbing to the lower appendages of the body [28,29]. This helps in distinguishing PCS from other pelvic pathologies [30].

#### 6. VULVAR VARICOSITIES

Vulvar Varicosities have a close association with PCS. Males develop testicular varicocele and females develop PCS from retrograde flow in gonadal veins, which are associated with pain and reduced fertility rates [31]. Vulvar varicosities often are the result of venous insufficiency and are often misdiagnosed as cysts or masses mainly in the Bartholin gland area [32]. Peri vulval varices are grossly dilated varices that may extend over the buttock and associated with other leg varices [7,27, 33]. Recent studies suggest that the primary cause of PCS is due to a dilation of the venous reflux [24]. Feeling of fullness of the upper thighs in one or both sides of the vulva can be indicative of vulvar varicosities [25,7,34].

Vulvar varicosities usually appear during pregnancy arising in 4 percent of pregnant women and often recede after delivery [7]. Usually after pregnancy vulvar varicosities resolve spontaneously, however if they persist they can lead to venous insufficiency and reflux [35,21]. It is suggested that vulvar varicosities can arise from an incompetent great saphenous vein and/or insufficiency of the internal iliac and ovarian veins [36,7,26]. This leads to venostasis and can result in pelvic pain that is associated with PCS [34].

#### 7. IMAGING

Imaging techniques have been an effective means of determining if atypical varices are present while avoiding surgical intervention. As often varicosities radiate from the perivulval region it is important to use such methods to reveal the clinical manifestations associated with venous insufficiency such as the gonadal veins [2,37]. Techniques such as ultrasonographic, tomographic venography, and Magnetic Resonance Imaging (MRI) were reported to be the most noninvasive three-dimensional study and useful technique for diagnosis of unusual lower extremity varicose veins and ovarian reflux

[34,35,38]. The most preferred methods are noninvasive however, when results are inconclusive more invasive means are present to yield more accurate findings [7,13,37]. It is proven that magnetic resonance venography (MRV) with time-resolved imagine of contrast kinetics (TRICKS) is an accurate imaging technique for evaluating ovarian vein dilation, reflux, and direction of flow in patients with suspected PCS [36].

#### 8. ULTRASOUND WITH DUPLEX DOPPLER IN SUPINE AND UPRIGHT POSITIONS

Ultrasound is a non-invasive imaging method used to assess pelvic venous changes. Ultrasound provides detailed conditions about the uterus, ovaries, cysts, and dilation of veins[1, 39, 15]. This is important in order to rule out other pathologies that may contribute to pelvic pain. Ultrasound with duplex doppler not only aids in the detection of a dilated ovarian vein, but it provides insight towards whether reversed caudal flow is present and whether the dilated pelvic venous plexus is tortuous [40]. One study looked at 32 control subjects presenting with pelvic vein dilation between 4.9 and 7.9 mm displaying symptoms associated with PCS [15]. One of the main limitations with ultrasound is that it is performed in the supine position and the upright position is significant in detection of venostasis in varicosities.

#### 9. VENOGRAPHY

Venography, although more invasive. is recommended to locate varicoceles and incompetent gonadal veins [4, 31]. It can provide a clearer representation of venous structures where ultrasound fails. Venography is also recommended in adjunction of PCS symptoms in the presence or absence varicosities [7, 41]. Ovarian venograms suggestive of PCS present with abnormally dilated ovarian veins (>10mm in diameter), sluggish blood flow, reflux which causes retrograde filling, congestion of the ovarian venous plexus, tortuosity of venous plexuses, uterine venous engorgement, and filling of pelvic veins across the midline [42,43,12,7,15]. Findings of pelvic varicosities via venogram are not always indicative of chronic pelvic pain, as most women are asymptomatic [6,15,20]. Venography allows for more sensitive results, as it can be performed in the semiupright position.

#### 10. COMPUTED TOMOGRAPHY AND MAGNETIC RESONANCE IMAGING (MRA and MRV)

CT scan is a noninvasive method that can be useful to detect tortuous, dilated pelvic ovarian veins, ovarian veins, broad ligament vascular congestion and ovarian varicosities better than ultrasound imaging. CT scans also can reveal unusual sites of compression other than the left renal vein due to nutcracker syndrome [3,19,14,17,44] MRA and MRV are important in providing a 3D image to detail the condition of pudendal veins and dilation of the venous plexus behind the bladder [43, 45]. The drawback is that these techniques are more expensive and may not provide more information than ultrasound imaging. They do not allow for concurrent therapeutic intervention.

# 11. DIAGNOSTIC LAPAROSCOPY AND VEIN LIGATION

Laparoscopy and vein ligation are not recommended for diagnostic purposes as they are significantly invasive and often do not reveal new information and lack specificity unlike the noninvasive imaging techniques described above [46,12,7]. It is important to note that although vein ligation is not an effective means for diagnosing PCS, it is very effective in reducing pain associated with vein dilatation and/or vulvar varicosities [47].

#### 12. MANAGEMENT OF PCS

There is a wide debate on how management of PCS should be conducted. Efforts on developing methods that are minimally invasive or noninvasive are ideal in relieving the pain associated with PCS. In the following we will discuss some of the researched methods and their effectiveness in the treatment and management of PCS.

#### 13. MANAGEMENT OF PCS THROUGH MEDICATION

It is clear that one of the main factors that contributes to the pelvic pain and is associated with PCS is pelvic varices [6,7]. Researchers have observed that the administration of medroxyprogestestrone acetate (MPA) or Goserelin (GnRH agonists) for 6 months is thought to reduce pelvic pain by decreasing the size of the pelvic varices [7]. This is the ideal treatment as it is noninvasive, however Goserelin or MPA are not permanent solutions as they can affect hormonal balance [21]. In a 1-year study a comparison was made between MPA and Goserelin to observe which of the two drugs was more effective. Based on a score system, Goserelin was observed to reduce pain, improve sexual functioning, reduce anxiety, and lower depressive states better than MPA [21]. Studies by Charles et al. show that premenstrual, intermenstrual women suffering from PCS were treated with combinations of progesterones and venotonics because the complication was hormonal and circulatory [8].

#### 14. SELECTIVE EMBOLIZATION

For patients who are not suitable or fail hormone therapy treatment, selective embolization of the superficial veins proves to be a simple, useful nonsurgical method in treating patients with PCS [4]. Transcatheter embolotherapy is a minimally invasive technique that is performed by an interventional radiologist that provides immediate and variable signs of pain reduction and shorthospitalization post operatively for men and women who suffer from varicocele [4,6,24,25,44,7,33]. Embolization aims to eliminate ovarian vein and/or internal iliac vein reflux in the presence of gross dilation of pelvic veins [7,17,24,25,48,49,]. Studies have shown that embolotherapy was more efficient in reducing pain associated with pelvic congestion syndromes compared to laparoscopy and venography when selective catherization targets and internal the ovarian iliac veins [4,6,17,24,33,49]. Patients saw good results after they were treated with a left phlebectomy and ligation of the supplying veins as a result of varicosities undergoing coil-embolization and liquid therapy [11]. It is noted that there was no statistically significant difference in relief outcome for patients presented with bilateral insufficient ovarian veins [50]. Although oophorectomy with hysterectomy provides a permanent cure, embolization is a much better technique because the surgery is most likely premature and genital organs are essential for the group of women [24].

#### **15. VALVULAR SCLEROTHERAPY**

Another method that is often used with beneficial results when patients are unresponsive to hormone therapy is sclerotherapy [51,52]. The most effective means of sclerotherapy is ultrasound-guided foam sclerotherapy [20,41].

This method can be used alone or in conjunction with pelvic vein embolization to yield a greater benefit for prolonged pain relief in PCS patients [6,41]. Not only does this method provide pain relief but it also allows for improved pelvic circulation in most patients [53].

#### **16. OPEN SURGICAL LIGATION**

Open surgical ligation is not recommended for most PCS patients [54]. This option is available when all other nonsurgical options have failed. However, open surgical ligation maybe a viable option to treat nutcracker syndrome through the insertion of stents and reducing venous dilatation of ovarian veins [3,16,12,20]. Although surgical ligation is not recommended as the top choice of treatment, it is important to understand the risks and benefits with any surgical procedure to most effective ensure the treatment is administered to the patient [33].

#### **17. CONCLUSION**

PCS has no known etiology, it is very difficult to diagnose, and appropriate treatment is still under investigation. Multifactorial treatment options are available to treat PCS syndrome.

There is a higher prevalence of PCS in premenopausal, multiparous women with chronic pelvic pain greater than 6 months. Investigation of Nutcracker syndrome, ovarian reflux, and ovarian vein dilatation may suggest PCS. Some patients with these conditions are asymptomatic while others display symptoms of chronic pain.

Although there are no diagnostic criteria present, various questions can be answered that may lead towards diagnosing PCS. Presence of a dull aching pain, tenderness, and heaviness in the upper thighs is suggestive of PCS.

Imaging techniques such as ultrasound, venography, computed tomography and magnetic resonance (MRA-MRV), are important methods in ruling out other pathologies associated with pelvic pain and confirming atypical venous changes.

Although most treatments are observed to reduce PCS symptoms it is important to administer primary non-invasive treatment options before administering more invasive procedures. Hormone therapy and selective embolization are the most useful treatment for PCS. Ultrasound-guided foam sclerotherapy and

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laparoscopic surgical ligation are viable options when the previous treatments fail. Pain consultation for pelvic pain syndrome is another option. These are all methods of improving patients' quality of life.

#### CONSENT

Not applicable.

#### ETHICAL ISSUES

Authors have declared that no ethical issues exist.

#### COMPETING INTERESTS

Authors have declared that no competing interests exist.

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