Pelvic congestion syndrome (PCS) was first described in 1857 and was associated with chronic pelvic pain in 1949. Despite this history, PCS is a condition that is often not considered during the evaluation of a patient with chronic pelvic pain. Even when it is considered, PCS is a condition that is difficult to diagnose. This does not diminish its importance, however, because PCS is a condition that causes distress to a significant number of women. There are many options for treating PCS; some are more successful than others, but none are completely successful. In this respect, embolotherapy is no different but the results of endovascular treatment are at least equivalent to those of surgery and better than conventional medical therapy.

Chronic pelvic pain accounts for up to 40% of female patients attending gynecology outpatient clinics and is said to occur in 15% of all women between the ages of 18 and 50 years. Fifteen percent of all hysterectomies and 35% of diagnostic laparoscopies are performed to diagnose and treat patients with chronic pelvic pain. Many of these patients will undergo clinical and ultrasound examinations that are normal and will ultimately get better without any intervention. However, approximately one-third of these patients will have endometriosis and another third will have some other structural abnormality of the fallopian tubes, bowel, or bladder. It is important that these significant pathologies are excluded before a diagnosis of PCS is considered. The diagnosis of PCS should be on every gynecologist’s list of causes of chronic pelvic pain, but unfortunately often it is not.

### Etiology

Pelvic congestion syndrome has a mixed etiology. The demands made on venous return by the constant hormonal changes associated with pregnancy leads to variable increases in intraluminal pressure. This can ultimately result in weakening of the ovarian vein walls and valvular incompetence. In some patients, there is a congenital absence of valves in the ovarian veins. These conditions can all result in reflux of blood down the ovarian vein into the tributaries of the internal iliac veins within the pelvis. This occurs more frequently on the left than the right because of the anatomic configuration of these veins.

The left ovarian vein drains into the left renal vein, which then takes a roughly perpendicular course to drain into the inferior vena cava. The right ovarian vein usually drains directly into the inferior vena cava, just below the right renal vein. The resulting congestion of the infundibulopelvic and broad ligaments leads to swelling and engagement, which, in part, leads to the pain experienced by the patient. However, it is not just the physical blood volume and back pressure that causes the pain: venous ischemia, both in the vessel wall and the end organ also plays a role and most interestingly the stretching and shear stress on the inner surface of the ovarian vein distorts both the endothelial and the smooth muscle cells that respond by releasing vasodilator substances. These include neuropeptide transmitters such as substance P and the neurokinins A and B. The nutcracker syndrome has a different etiology and implications and is discussed separately.

### Patient Presentation

One of the reasons why a diagnosis of PCS is often not considered is because it can present with a variety of symptoms (Table 16.1). These findings tend to be nonspecific and are therefore potentially indicative of several of the conditions that can cause chronic pelvic pain. PCS occurs most often in women who are multigravida and in their 20s and 30s. It classically presents with pelvic pain or a

<table>
<thead>
<tr>
<th>Table 16.1 Symptoms Associated with Pelvic Congestion Syndrome</th>
</tr>
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<tbody>
<tr>
<td>Pain brought on by an increase in intraabdominal pressure</td>
</tr>
<tr>
<td>Pain relieved by lying down</td>
</tr>
<tr>
<td>May be unilateral or bilateral</td>
</tr>
<tr>
<td>Dull aching pain</td>
</tr>
<tr>
<td>Dysfunctional bleeding and dysmenorrhea common</td>
</tr>
<tr>
<td>Low back pain</td>
</tr>
<tr>
<td>Postcoital ache that may last for hours or days</td>
</tr>
<tr>
<td>Acute episodes of pain common</td>
</tr>
<tr>
<td>Menstrual disorders</td>
</tr>
<tr>
<td>Bladder irritability (24 to 45% of patients)</td>
</tr>
<tr>
<td>Functional gastrointestinal symptoms</td>
</tr>
<tr>
<td>Family history of varicose veins and associated with vulval varicosities</td>
</tr>
</tbody>
</table>
postcoital ache that is worse on the left side, may last for hours or days, and may be relieved by lying down. The pelvic fullness experienced by these patients can be acute and severe or chronic and dull. It can be unilateral or bilateral. These symptoms, when combined with ovarian point tenderness, are said to be 94% sensitive and 77% specific for pelvic congestion syndrome. Such specificity and sensitivity figures indicate that women do have a variety of other symptoms including pain brought on by increased intraabdominal pressure, bladder irritability (which occurs in 24 to 45% of all patients), functional gastrointestinal (GI) symptoms (such as constipation), and low back pain. A history of varicose veins and associated vulval varicosities is often present in patients with PCS. Other features on physical examination that may point to the diagnosis include cervical motion tenderness and an engorged blue-looking cervix.

Women with PCS are often described as being depressed and anxious. This is partly due to the fact that they have very real symptoms, which are chronic and potentially debilitating. In addition, they have often gone through an extensive diagnostic workup and in many cases, the cause for their discomfort is difficult to find. There are also good pharmacologic and physiologic reasons why these patients might be suffering from psychological stress. Substance P and the neurokinins A and B, released in response to stretching by endothelial and smooth muscle cells described earlier, play a key role in the regulation of emotions and are an integral part of central nervous system pathways involved in psychological stress.

## Imaging

Following the performance of a detailed history and physical examination, most patients presenting with chronic pelvic pain will undergo some type of imaging evaluation. This most commonly includes an ultrasound examination and laparoscopy. Sonographic findings of pelvic congestion syndrome include a dilated left ovarian vein with reversed caudal flow, dilated arcuate veins crossing the uterine myometrium, polycystic changes of the ovary, and variable duplex waveform during a Valsalva maneuver. Computed tomography (CT), magnetic resonance imaging (MRI), and magnetic resonance angiography (MRA) (Fig. 16.1) can all aid in the diagnosis of PCS, but in all of these investigations the patient is likely to be supine. Therefore, although all of these studies may demonstrate venous engorgement in the pelvis in severe cases, in less severe cases this same venous engorgement might not be seen, which will result in a false-negative examination.

Cross-sectional imaging studies remain important because they may reveal associated abnormalities that suggest a diagnosis of PCS. For instance, polycystic ovaries may be found on ultrasound and these are associated with pelvic congestion syndrome in 56% of patients. Similarly, imaging may reveal an increase in uterine cross-sectional area and an endometrial thickness, which is marginally greater than normal. On CT, these findings may cause confusion with adenomyosis. None of these imaging modalities, however, should be considered as a primary tool for diagnosing PCS.

Presently, ovarian venography remains the definitive modality for evaluating patients for PCS (Fig. 16.2). Ideally, a percutaneous venogram should be performed on a tilting table so that the patient is partially upright during the examination. This will maximize the ability of venography to detect reflux and perhaps address the false-negative cases seen on cross-sectional imaging or laparoscopy performed in the supine position. Ovarian venography can be performed from a jugular or femoral vein approach. The jugular vein is often preferred because the angles favor selective catheterization of the ovarian veins from this approach. In addition, it is preferred by many if embolization is being considered at the same sitting for the same reason. Selective catheterization of the left renal vein (LRV) can usually be performed with 4 French cobra or multipurpose catheters. The same catheters can be used to catheterize the right ovarian vein (ROV), though a sidewinder is often required if the study is being performed via femoral vein access. The LRV should be catheterized first because it is typically more straightforward and often has the greatest yield for a positive finding.

Once the LRV is catheterized, a venogram should be performed with the patient in a semierect position or while the patient is performing a Valsalva maneuver (if a
tilting table is not available). This can be achieved using an old-fashioned sphygmomanometer. With this technique, the patient is asked to blow into the rubber tubing and to hold the mercury at ~20 mm. The venogram should be performed prior to catheterization of the LOV for two reasons. First, it is important not to miss the diagnosis of nutcracker syndrome (see below). Second, if significant reflux is present, an injection of contrast into the LRV (under the above-described conditions) will immediately determine whether or not the patient has reflux down the ovarian vein. If no reflux is seen then it is highly unlikely that the patient has PCS. An attempt can be made at this point to catheterize the ROV, but if this is also not seen or is not quickly catheterized, it is probable that it is small and not refluxing. The examination can be terminated at this point. The diagnostic criteria for PCS on venography include a minimum ovarian diameter of 8 to 10 mm, uterine venous engorgement, congestion of the ovarian plexus, filling of the pelvic veins across the midline, and/or filling of vulvo-vaginal and thigh varicosities.9,10

The LOV can be catheterized with a hydrophilic guide wire and the chosen catheter. Great care should be taken in performing this selective catheterization because in its distended state, the ovarian vein is fragile and can easily be ruptured. In addition, it can go into spasm very quickly if it is overly irritated and this can prevent treatment and occasionally prevent the removal of the catheter until it relaxes. The guide wire and catheter should be carefully advanced further into the ovarian vein until it is positioned just above the pelvic brim. At that point, additional contrast should be injected to confirm reflux into the pelvic veins. In addition, this injection can demonstrate cross filling to the right side of the pelvis (if present) and reflux into any vulvar or thigh varicosities that may be present.

**Nutcracker Syndrome**

The so-called nutcracker syndrome is caused by LRV compression between the superior mesenteric artery and the aorta (Fig. 16.3), with associated retrograde hypertension within the LRV (and its tributaries).11,12 The renal venous
hypertension found in patients with nutcracker syndrome leads to the development of collaterals around the renal pelvis that may cause hematuria and retrograde flow in the LOV.13 This can result in the symptoms of PCS.

There are two peaks of incidence at young and middle age, which correspond to two different pathophysiological stages of the nutcracker syndrome. At an early stage, LRV compression leads to LRV hypertension and the development of venous collaterals around the renal capsule. Because incompetence of the LOV veins has presumably not yet occurred, no reflux is seen in these vessels at this stage; furthermore, high pressure in the LRV potentially leads to the development of direct communications between the dilated veins and adjacent calyces with consequent hematuria and flank pain.14,15

At a later stage, the persistence of LRV hypertension causes valvular incompetence and massive reflux of blood into ovarian or lumbar veins leading to pelvic or vulvar varices with the symptomatology of PCS. This is mainly encountered in middle-age women.16,17 The diagnosis of nutcracker syndrome is based on an association of symptoms of hematuria, left-sided flank pain, pelvic congestion, pelvic and vulvar varices, and imaging findings of LRV compression on cross-sectional imaging confirmed by venography and documentation of a pressure gradient of more than 3 mm Hg between the LRV and IVC.18

A variety of surgical procedures, including LRV bypass with polytetrafluoroethylene (PTFE) graft interposition, and autotransplantation of the left kidney into the left iliac fossa have been described to alleviate the LRV compression associated with the nutcracker syndrome.19 Recent reports describe endovascular procedures such as renal venous stenting from a femoral vein approach (Fig. 16.4).20–22 Follow-up in these series, which total nine patients only, ranges from 4 to 54 months. These few case reports and series are encouraging, especially in light of the fact that venous stenting is proving successful in the short treatment of venous diseases such as vena cava syndromes and Budd–Chiari syndrome. However, it is my experience that venous stenting is successful only for a short time in patients with nutcracker syndrome. Studies require much longer follow-up if intravascular stenting is to become a treatment option.

### Treatment

There are few medical options to treat patients with PCS, all of which revolve around addressing the pain associated with this syndrome. Nonsteroidal antiinflammatory drugs may help with pain relief in the short term, but they are not a long-term solution to this problem. Medroxyprogesterone acetate has been shown to relieve symptoms in ~40% of patients and a combination of this and psychotherapy may be effective in ~60% of patients. However, Farquhar et al23 performed a study in which patients were assigned to psychotherapy alone, medroxyprogesterone acetate alone, these two in combination, or placebo and found that placebo use was effective at one year in 50% of patients. Analgesia is a good first line treatment, but patients symptoms

![Fig. 16.4](image)

**Fig. 16.4** Left renal vein stenting has been suggested as a treatment option in nutcracker syndrome as in this case where a 10 mm self-expanding stent has been inserted successfully. However, longer-term studies are required to evaluate the real efficacy of this treatment.
should not be ignored if it is not completely effective or pain recurs when it is stopped.

Surgical treatment utilizing hysterectomy and bilateral oophorectomy with hormone replacement is said to effectively cure, or at least bring symptomatic improvement in two-thirds of patients. Despite the definitive nature of a hysterectomy, it does not appear to be as effective as simply ligating the ovarian vein, which is reported as having a 73% cure rate with 78% of patients demonstrating symptomatic improvement. Ligation of the ovarian vein(s) can be done using an open or laparoscopic approach. However, ligation of the ovarian vein can also result in transaction to nerves of the pelvis and still leaves open the possibility of establishing collateral channels leading to symptomatic recurrence. Surgery is rarely used because endovascular treatments are more effective and less invasive.

Since Edwards et al. reported the first case in 1993, embolization used to treat ovarian varices has become an effective treatment option for these patients. Unlike ovarian vein ligation, embolization of the ovarian vein leaves the nerves that accompany the vein intact. Though it does potentially allow for the establishment of collaterals, these are limited by occlusion of collaterals that drain into the high ovarian vein. Embolization is reported to have a 73 to 78% rate of symptomatic improvement or cure. There is still controversy among interventional radiologists regarding the best technique to use and it is clear from the literature that the technique is still evolving. Specific questions that remain unanswered include the following:

- Should only refluxing veins be embolized (usually the left)?
- Should both ovarian veins be embolized regardless of whether they are both refluxing?
- Should both ovarian veins and the internal iliac veins be embolized?
- If the latter, should this be done all at once or should treatment be staged?
- What embolization material should be used – coils, sclerosants, glue, or Gelfoam (Pfizer, Inc., New York, NY)?

Initially, patients were treated with unilateral ovarian vein embolization. This was invariably the left ovarian vein given the anatomic predisposition of this vessel toward causing PCS. In this early experience, coils were the main embolization material used. Results were variable, with significant relief being described in ~66% of all patients. It is not always clear in these articles how “significant relief” was defined. Approximately 33% of patients were described as having partial or no relief.

Although these figures are slightly disappointing, they are at least as good as those described for surgical treatment. By the late 1990s, bilateral ovarian vein embolization was becoming the standard of care for this patient population. Many operators were embolizing the refluxing left ovarian vein and the right ovarian vein whether it was refluxing or not. This is not always as easy as it sounds. Occasionally, the right ovarian vein cannot be found or is extremely small and therefore difficult to catheterize. In these circumstances only the left ovarian vein is embolized because it is reasonable to conclude that a small vein that is difficult to catheterize may not be responsible for significant reflux and symptoms.

The use of foam sclerotherapy has also been applied to the ovarian veins when treating patients with PCS. A total dose of 3 to 6 cc of 3% sodium tetradecyl sulfate (STD), mixed with either air or Gelfoam and then passed rapidly back and forth across a three-way stopcock between two syringes, can be injected into the ovarian vein below the level of the pelvic brim to treat PCS. This can then be followed by coil embolization to within one centimeter of the vein origin. In addition, the internal iliac

![Fig. 16.5](image) A catheter in the left ovarian vein has been advanced to the pelvic brim in this patient with classical pelvic congestion syndrome. Venography reveals multiple large varicosities with cross filling to the right. Following injection of 3 cc of 3% sodium tetradecyl sulfate and Gelfoam (Pfizer, Inc., New York, NY), mixed into a foam between two syringes there is stasis in the varicosities. This was followed by coil embolization of the left and right ovarian veins.
veins can be embolized with either coils or sclerosant. Results using this approach seemed to improve, though it is difficult to determine objectively how this improvement was being measured.

The largest series to date describes 127 patients, 106 of whom underwent bilateral ovarian vein sclerotherapy followed by coil embolization.30 The remaining patients underwent unilateral embolization and sclerotherapy. All patients underwent a venogram of the internal iliac veins 4 to 6 weeks after ovarian vein embolization. The internal iliac veins were then occluded proximally with a balloon and contrast was injected into the internal iliac vein to determine if and when contrast refluxed into the contralateral internal iliac vein. If residual varices were seen, the same volume of sclerosant/Gelfoam slurry was then injected. After 5 minutes, the balloon was deflated and the procedure was repeated in the opposite internal iliac vein. One hundred eight of the 127 patients in this study underwent bilateral internal iliac vein embolization in this way. A visual analogue score similar to that used for pain relief in cancer was used to assess outcome in this study. Eighty three percent of patients demonstrated relief of symptoms, 13% were no better, and 4% were worse. It would seem from these results that there can be a significant improvement in the success rate of embolization by embolizing the internal iliac veins.

My approach is to embolize the incompetent left ovarian vein by using a slurry of Gelfoam and a sclerosing agent together with coils. The same approach can then be utilized on the right if there is obvious reflux or to just use coil embolization if the reflux is not obvious (assuming that the vessel can be easily catheterized). Patients are seen 6 weeks later in the outpatient setting, and if they are still complaining of significant symptoms, the internal iliac veins are embolized in the manner described by Kim et al.30

Complications

Significant pulmonary embolic disease does not appear to be a complication of ovarian or internal iliac vein embolization. The use of coils in the internal iliac veins in some patients can lead to coil displacement with migration into the lungs; this can potentially require retrieval of these coils. This problem, however, has not been reported in association with the use of a Gelfoam/sclerosant slurry. Occasionally a patient’s symptoms can be made worse by embolization, but this is exceptional, occurring in only 4% of patients.30 Spasm and occasional rupture of the thin-walled ovarian vein can occur, but this rarely leads to any sequelae and can be treated with simple analgesia. There have not been significant changes in basal follicle-stimulating hormone, luteinizing hormone, or estradiol levels associated with this procedure.30

Conclusions

Patients with pelvic congestion syndrome are generally unhappy and have often been through a plethora of investigations to exclude other causes of pelvic pain. Pelvic venography performed in an upright or semiupright position, should be the procedure of choice when evaluating these patients because other investigations can misdiagnose subtle pelvic congestion syndrome. If pelvic congestion is caused by the nutcracker syndrome it can be treated by embolization and renal vein stenting, but this may fail in the medium to long term. In classic pelvic congestion syndrome, embolization, if done carefully, is not a difficult or time-consuming procedure, especially if performed from the jugular approach. The evolution of the embolization technique would now dictate that both ovarian veins are embolized if the right ovarian vein can be found and easily catheterized. Gelfoam sclerosant slurry is very effective for this purpose, followed by coil embolization. The internal iliac veins can be embolized as described if the patient’s symptoms persist. Finally, it is very important not to build the patient’s expectations too high as treatment may fail to relieve symptoms on a long-term basis.
References
2. Taylor HC. Vascular congestion and hyperemia; their effects on structure and function in the female reproductive system. Am J Obstet Gynecol 1949;57:637–653