IntRODUCTION

Pelvic congestion syndrome (PCS) is probably one of the worst names for a medical condition. PCS covers a collection of symptoms and signs both inside and outside the pelvis, many of which have alternative causes. It is not surprising that most patients do not understand PCS as most doctors and nurses still have difficulty in comprehending this condition.

As PCS is made up of a diverse number of symptoms and signs, it is often ignored as a diagnosis by doctors and nurses. As will be noted below, gynecologists, in particular, seem to ignore PCS[1] despite it appearing to be a major cause of chronic pelvic pain (CPP) in women,[2,3] and midwives often tell women with vulvar varicose veins following pregnancy that they will get better by themselves, failing to inform the patients that these varicosities may well be linked to pelvic venous reflux (PVR).[4,5] Furthermore, even doctors and nurses who have some knowledge of PCS often think that the condition is restricted to females and do not diagnose PCS or PVR in males.[6]

The further problem is that conventionally, medical conditions are conventionally recognized by the anatomy of where the problem appears, and are usually treated by doctors specializing in that anatomical area. As such, hemorrhoids are usually treated by colorectal surgeons, CPP is treated by gynecologists and testicular varicocele and impotence are treated by urologists. However, with increased understanding of the pathophysiology of the venous system, it has been found that these conditions are due to venous pathology in PCS patients. With improved venous investigations, understanding and treatments, these may well be treated better either by phlebologists or at the very least, a combined approach between phlebologists and the traditional specialists in each area.

In this article, I will outline the current understanding of PCS and will hopefully identify some of the current areas that require research and development to improve on the current diagnosis and treatment of this condition.

SYMPTOMS AND SIGNS OF PELVIC CONGESTION SYNDROME

As noted in the introduction, PCS covers a wide range of symptoms and signs, many of which can be associated with other conditions. I have always found it useful to separate the symptoms from the signs, and to then think of the symptoms as internal or external to the pelvis and the signs as pelvic or leg [Table 1]. However, as can be seen from this Table, they can be a very wide range of symptoms and/or signs that might be attributable to PCS but also might be a symptom or sign of another underlying pathology.

The symptoms that are internal to the pelvis are probably those that are “classically” thought of as PCS. Primary of these is the “aching” or “dragging” type of pain that is present on sitting or standing when the varicose veins will be full, which then relieves on lying down particularly if the pelvis of legs are elevated.[7] The pelvic varicose veins surrounding the lower bowel and bladder can easily be thought of as causing irritable bowel syndrome (IBS) or irritable bladder, and in our experience, some patients find relief of these symptoms after treatment despite research suggesting that CPP and IBS are different conditions.[8]

Dilated pelvic veins due to PVR has been suggested as a cause of impotence in males[9] with reports of selective embolization of the pelvic veins restoring erectile function,[10] and there has also been a suggestion of an association between prostatic hypertrophy and pelvic congestion due to outflow obstruction in a rat model.[11]

Although low back pain, iliac fossa vein, and groin pain can easily be imagined to be a result of pelvic varicose veins, due to their anatomical association, a recent report of two patients

Address for correspondence: Prof. Mark S Whiteley, E-mail: info@whiteleyclinics.co.uk

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with severe hip pain being cured with pelvic vein embolization has shown that even pain in the hip severe enough to be diagnosed with osteoarthritis can be caused by PVR, and hence is an external symptom of PCS.\(^{[12]}\)

Whereas symptoms can be highly subjective by their very nature, signs are objective and therefore much easier to identify and investigate. Varicosities around the external genitalia, perineum, buttocks, and hemorrhoids have a very high chance of being associated with PVR and pelvic varicosities.\(^{[13]}\) Interestingly, this includes hemorrhoids\(^{[14]}\) even although these are conventionally treated by colorectal surgeons and thought of as a bowel condition.

**Demography of Pelvic Congestion Syndrome**

It is usual when discussing medical conditions to start off discussing the demography of the condition. However, as PCS has such a wide variety of symptoms and signs, it is not surprising that there are also a wide variety of clinical presentations. Different clinical presentations of PCS have been studied by different specialists, some not even associating them with PCS of PVR, and demographic studies show very different populations for the different presentations. This is further confused by the fact that certain symptoms or signs can clearly also have other causes, which can confuse the demography further.

**Table 1: Signs and symptoms of pelvic congestion syndrome**

| Symptoms: Internal                       |
|-----------------------------------------|
| Aching or dragging pain on sitting or standing |
| CPP                                     |
| Deep dyspareunia (female)               |
| ? (IBS)                                 |
| ? Irritable bladder                     |
| ? Impotence (male)                      |
| ? Prostatic hypertrophy (male)          |
| **Symptoms: External**                  |
| Groin pain                              |
| Iliac fossa pain                        |
| Lower back pain                         |
| Upper thigh pain                        |
| Hip pain                                |
| **Signs: Pelvic**                       |
| Perineal varicosities                   |
| Buttock varicosities                    |
| Haemorrhoids                            |
| Suprapubic varicosities                 |
| Flank varicosities                      |
| Vulval varicosities (female)            |
| Vaginal varicosities (female)           |
| Varicocele (male)                       |
| **Signs: Legs**                         |
| Varicose veins of legs especially post thigh |
| Para-vulval varicosities (female)       |

CPP: Chronic pelvic pain, IBS: Irritable bowel syndrome

It is also very important to remember that PCS and PVR is found in males as well as females. Having made that point, I will consider some of the known demographic details relating to PCS and PVR, using the four headings featured in Table 1.

**Symptoms-internal**

Most of the internal pelvic symptoms reported in PCS are very nonspecific, and there have been many reports about CPP\(^{[15]}\) although as CPP has many causes, there is not much good demographic data for CPP due to PCS. A recent study performed in the UK using a freedom of information request for official statistics concluded that between 13% and 40% of women attending gynecological outpatients probably due to PCS.\(^{[2]}\) Unfortunately, in the UK, this is not a diagnosis that is currently recognized by gynecologists, and at the point that this article has been written, does not appear as a differential diagnosis for pelvic pain and the Royal College of Obstetricians and gynecology website.\(^{[16]}\) Clearly, in one-third of women presenting to gynecological outpatients with CPP are due to PCS, then the condition is prevalent in the UK.

**Symptoms-external**

The pains localized to the external pelvis such as the back, iliac fossa, inguinal regions, and upper thighs from PCS\(^{[17]}\) can clearly have other more common causes. As such it is impossible to be certain about demographic information relating to PCS with these presentations. However, hip pain due to PCS that was misdiagnosed with osteoarthritis of the hip has only recently been reported\(^{[18]}\) and now should be included in questionnaires and studies relating to PCS symptoms.

**Signs-pelvic varicosities**

Visible varicose veins that can be shown to be emanating from PVR are much easier to identify and hence quantify. As pointed out above, most medical conditions are identified and treated according to their anatomical location and by specialists who deal with that area. Hence, we have to look in nonvenous literature for some of the demographic data. Hemorrhoids are thought to be present in 5% of adults in the United States at any one time and to occur in approximately 50% of all adults sometime during their life.\(^{[19]}\) Vulval and vaginal varicosities are reported to occur in 4% of women, usually secondary to pregnancy.\(^{[20]}\) Varicoceles in males are probably the easiest sign of PVR to find and hence there is a huge amount of demographic data available in medical textbooks as well as in the literature. It has been reported to occur in up to 15% of children and adolescents\(^{[21]}\) is commonly thought that 95% present on the left due to the anatomy of the left testicular vein. Finally, varicosities associated with obstructive disease in the pelvis in the form of suprapubic or flank varicosities are much rarer, in keeping with the fact that obstructive venous disease in the pelvis is much rarer than PVR.

**Signs-leg varicose veins**

Most venous surgeons first come across PCS and PVR due to the link between PVR and leg varicose veins. A study from our own unit, performed in our local NHS hospital
and also in our specialist private vein clinic, showed that the prevalence of women with primary varicose veins having a significant contribution from PVR to their leg reflux pattern was approximately 20%.\[22\] We performed the study across both units to see if there was a selection bias in our private clinic, as we are known to specialize in this area. However, we found that there was no evidence for such bias. Furthermore, we have recently published our results showing that 3% of males presenting with leg varicose veins have a significant contribution to their leg varicose veins from PVR.\[23\] It is not surprising that as most vascular (arterial) surgeons who operate on veins are not looking for PVR (and many will not even be aware of the link), we have found that unidentified and untreated PVR is a major cause of recurrent leg varicose veins. PVR was found to be a significant cause of recurrence in 1/4th–1/3rd of women who had undergone their primary varicose vein surgery elsewhere.\[24\]

**CAUSES OF PELVIC CONGESTION SYNDROME**

In order for us to have any logical starting point in investigating PCS, we have to know what the underlying pathological process is that we are describing. PCS is venous congestion of the pelvis. Indeed, many authors have called this condition “pelvic venous congestion” rather than PCS. Simply, we can call PCS “varicose veins of the pelvis” or “pelvic venous stasis.” To a large extent, we can draw parallels with varicose veins in the legs.

All medical students learn about one form of PCS at medical school, but then seem to promptly forget it as soon as they talk about PCS in women. One of the first conditions, we learn about at medical school is varicocele in the male. We will get taught that a testicular varicocele is a varicose vein of the testicular vein, due to the failure of the valves in this long truncal vein. We get taught that it is more common on the left than the right, due to the left testicular vein having a longer course in the retro-peritoneum where it drains into the left renal vein rather than on the right, where it drains into the inferior vena cava. We learn that this is a common condition and is easily treated by ligation, and more recently, embolization. Therefore, we have an underlying comprehension that testicular varicocele is caused by PVR of the testicular vein or veins.

However, when we qualify as vascular surgeons, and come across women presenting with PCS, we seem to forget what we have learnt as medical students. Rather than think of females with PCS as “varicocele of the ovary,” with a similar etiology of PVR is found in the male, we find that some authors are convinced that PCS is frequently caused by compression syndromes such as nutcracker syndrome or May Thurner syndrome rather than simple reflux. Furthermore, it is amazing how many articles on PCS tribute the condition to pregnancy, despite pregnancy being exceptionally rare in males with testicular varicocele!

Contrary to some authors who seem to find compression syndromes regularly in such patients, in our own figures, we have found the primary cause of PCS to be PVR in the vast majority of patients, with compression only being suggested as a cause in some 1%–2% of cases.\[25\]

Furthermore, although many authors still concentrate on reflux in the gonadal veins, our figures have shown that the internal iliac veins are more commonly affected by PVR than the gonadal veins. Indeed, we have shown that the most common pattern of PVR is reflux in the left ovarian vein and both internal iliac veins.\[25\]

It is also interesting to note that in our investigations of PVR when associated with leg varicose veins, although approximately 20% of females with leg varicose veins show a pelvic origin for their venous reflux, only some 16.5% show truncal venous reflux, with the others showing pelvic varicose veins with no truncal reflux from the gonadal veins or internal iliac veins underlying it.\[25\] This suggests an ascending pattern of reflux in the majority of patients\[26\] as has been found in leg varicose veins,\[27\] and this also goes against compression being a primary cause in these patients. In addition, it also shows that although most authors accept that dilated veins in the pelvis when associated with the signs or symptoms of PCS are diagnostic of the condition; PVR in the truncal veins does not need to be present. This becomes highly relevant when considering the pathophysiology of the symptoms found in patients with PCS.

Of course, in parallel with leg varicose veins, the presence of reflux in one or more pelvic veins is not enough to diagnose PCS, as the reflux may be minor or even asymptomatic. It is for this reason that I have recently suggested that we work toward producing a grading system of PVR to see if this correlates with the severity of symptoms or signs in PCS.\[28\]

**PATHOPHYSIOLOGY OF PELVIC PAIN IN PELVIC CONGESTION SYNDROME**

For the diagnosis of PCS, two criteria have to be met. First, the patient needs to have symptoms or signs of PCS. Second, there needs to be proven PVR and/or dilated veins identified in the pelvis. A patient who presents with symptoms or signs of PCS but who does not have any abnormal veins in the pelvis cannot be diagnosed with having PCS.

However, although there is a clear association between PCS and PVR/dilated veins, the mechanism of how the venous reflux or stasis causes symptoms in the pelvis is unknown.

In 2014, Phillips et al. authored a review of PCS, in which they outlined the current theories on how pelvic varicose veins could cause the symptoms of PCS.\[29\] One theory is that the engorgement of the veins in the pelvis causes stretching of the vein walls and venous stasis, both of which can stimulate pain receptors in the vein wall. The supporting evidence for this is that gabapentin and amitriptyline which are used for neuropathic pain get better results in PCS than opiates or nonsteroidal anti-inflammatory drugs. Furthermore, they
reported that studies have shown that patients with pelvic and leg varicose veins have more pain than patients with leg varicose veins alone.

Alternatively, they reported research suggesting that the symptoms of PCS could be due to the release of neurotransmitters from the vein walls in the pelvis such as substance P, calcitonin gene-related peptide, adenosine triphosphate, endothelin, vasopressin, and nitric oxide. Supporting evidence for this is that medroxyprogesterone acetate reduces pain in PCS and this is known to act by inhibiting a neurotransmitter release.

A further potential mechanism, and much more simple to contemplate is the suggestion that the very dilation of the veins in the pelvis mechanical stress on the surrounding viscera in the pelvis, causing inflammation, direct pressure on local nerves or ischemia of pelvic organs.

Of course, it is likely that there will be a combination of different reasons that pelvic varicose veins cause the symptoms of PCS, and indeed it is also known that some patients with proven dilated veins in the pelvis do not have any symptoms of PCS at all.

Therefore, it is impossible at the moment to know which of these mechanisms of primary and which are secondary. Identification of the primary mechanism is useful if we are aiming for a pharmacological solution to PCS. However, if treatment strategies are aimed at physically preventing PVR and removing the dilated veins in the pelvis, then understanding these mechanisms are largely irrelevant.

**Investigation of Pelvic Congestion Syndrome and Pelvic Venous Reflux**

To diagnose PCS, there have to be both the symptoms and/or signs associated with PCS and a positive diagnosis of pelvic varicosities and/or PVR. As previously noted, although some 20% of females with leg varicose veins show reflux from pelvic veins into the leg veins duplex ultrasonography, only 16.5% show truncal vein PVR. Therefore, approximately 3.5% have reflux in pelvic varicosities without any truncal PVR. This should not surprise any venous surgeon as all venous surgeons have seen patients with leg varicose veins who, on duplex ultrasonography, do not have any truncal vein reflux causing them.

It is natural to think of venogram as a first-line investigation for PVR and pelvic varicosities. However, venous surgeons learned in the 1990s that venogram was very second-rate compared to have venous duplex ultrasonography in the assessment of leg varicose veins and truncal reflux in the legs. It, therefore, seems amazing that we have to learn the same lessons all over again 20 years later in the pelvic veins.

The same arguments still hold as they did for trying to assess leg varicose veins with venography. Venography involves the introduction of radiopaque contrast which is denser than blood, is often injected colder than blood and is usually injected under pressure. It only opacifies the veins that it flows through, giving no image of veins that are present which are bypassed by the contrast. The flow of the contrast is not physiological for the above reasons. Venography can be performed at different angles, and to have any chance of seeing venous reflux, the patient needs to be tilted into a head-up angle.

Whenever doctors think of imaging deep inside the body, they automatically think of cross-sectional imaging such as computer tomography scanning or magnetic resonance imaging. However, these cross-sectional imaging techniques are usually performed with the patient lying flat, preventing any physiological reflux. Moreover, lying flat pelvic varicosities may not be dilated. Although some workers in the field are trying to formulate protocols that show venous reflux following certain maneuvers such as valsalva, the diagnosis of abnormal gonadal veins is usually made on diameter alone.

However, we have recently published a study where we measured the diameter of ovarian veins, both competent and incompetent at venography compared with reflux seen on transvaginal venous duplex ultrasonography performed at 45° using the techniques invented by Judy Holdstock and outlined by Angie White and Judy Holdstock in a paper in this journal. This technique has already been shown to be likely to be the gold standard investigation for PVR. This study showed that diameter the vein was irrelevant when trying to diagnose competent from incompetent, casting huge doubt on the use of cross-sectional imaging to make a diagnosis of PVR.

Duplex ultrasonography is already widely accepted to be the gold standard test for venous reflux in the legs. Labropoulos and his group have published their criteria for investigating PVR using external transcutaneous duplex ultrasound. However, this technique depends on the size of the patient, the presence of bowel gas and images the gonadal veins on the posterior abdominal wall with limited views of the right ovarian vein, internal iliac veins, and veins within the true pelvis.

As there is evidence that PVR is an ascending condition, this makes external transcutaneous duplex ultrasonography of limited value in the investigation of PVR. The addition of transvaginal venous duplex ultrasonography, with the patient, elevated in the head-up position at 45° allows physiological reflux to be seen in the lower pelvic veins, as well as the presence of pelvic varicosities and any exit points from the pelvis. The addition of maneuvers such as the Valsalva maneuver and Kegel squeeze enhance this test further, as in the paper by Angie White and Judy Holdstock in this journal.

**Treatment of Pelvic Congestion Syndrome and Pelvic Venous Reflux**

Pharmacological treatments for PCS have been widely reported, including simple analgesia, or the use of...
gabapentin/amitriptyline or medroxyprogesterone acetate as in the section pathophysiology above. However, in parallel with our understanding of leg varicose veins, pharmacological treatments may take away symptoms temporarily, but of course, will not have any effect on the pathophysiology that is causing the problem in the first place. As such, these “treatments” can be thought of as temporary symptom relief cannot really be thought of as a curative treatment.

Logically, the only way to reverse the underlying pathophysiological abnormality is to stop the PVR and to reduce the dilation of any pelvic varicosities.

Recently, there has been a very interesting study reported from Russia that has suggested that compression from external compression pants can reduce the symptoms of PCS. This could be paralleled with the use of compression stockings for leg varicose veins. Of course in the pelvis, the compression is much harder to transmit pressure directly to the refluxing and dilated pelvic veins, due to the rigid bony pelvis and also the bowel in the peritoneal cavity. As such, it will be interesting to see whether other centers can repeat the results found in this innovative study. Moreover, if the improvement is confirmed with compression, just as with pharmacological treatments, it would only be a temporary symptomatic relief rather than a cure of the condition. In the same way, that we know that compression stockings are not a cure for varicose veins but merely a temporary improvement was there being worn, such compression pants might be used for those unwilling to go ahead with definitive treatment or unable to do so.

Definitive treatment of PVR and/or pelvic varicosities is the ablation of incompetent pelvic truncal veins and the reduction of pelvic varicosities. Probably, the most important part of this is the identification of which trunks are incompetent, and also the recognition that the internal iliac veins are much more commonly involved than the gonadal veins. However, due to the difficulty in treating internal iliac veins, these are often ignored.

Currently, the gold standard treatment for pelvic truncal venous incompetence is embolization, usually using metal embolization coils. In our own practice, we use this via a transjugular route under local anesthetic as a walk-in walk-out procedure, although a more traditional route is via the groin. However, as both the gonadal veins and the internal iliac veins pass upward and therefore have origins superiorly, a transjugular route allows easier cannulation into the relevant vein. Dilated veins in the pelvis can also be treated with coil embolization, although this can end up being very expensive and also leave a large mass of coils if the veins are large and numerous. Therefore, in our own practice, over the years we have developed the technique of using foam sclerotherapy to fill the dilated veins, using coil embolization proximal to this in the truncal veins to prevent reflux from re-opening them.

Other techniques of embolization have been reported, some more logical than others. Glue has been used either by itself or in combination with sclerosant, with very good results and this is certainly a good alternative to coil embolization. Conversely, there have been reports of “plugs” to occlude the PVR. Although this is also a recommended embolization method by the AVF guidelines, it is an illogical way of trying to treat PVR as it assumes that reflux is a problem that emanates from the top of the vein in a descending pattern. As increasing evidence has shown that venous reflux disease is an ascending problem, both in the legs as well as in the pelvis, ablation should start distally and proceed approximately both to stop reflux in the future and to reduce the risk of pulmonary embolism of any device left high the vein.

There have been verbal reports at meetings of using thermoablation devices in the truncal pelvic veins. My personal view on this is that this is court disaster and should not be undertaken unless good evidence has been obtained to show how surrounding structures such as ureter and bowel can be protected from the heat generated by the device. As we have suggested previously that to get successful ablation it is necessary to get the transmural death of the vein wall, and increasing evidence seems to point to this being correct, then using thermoablation to get adequate ablation will by necessity cause thermal spread to surrounding tissues and organs. If low total energies are used to prevent such surrounding thermal spread, then it is likely that there will be inadequate energy to ablate the target vein resulting in immediate thrombosis and apparent occlusion, followed by recanalization and recurrence in the future.

One of the most frustrating problems with the treatment of PCS is that, despite improved imaging criteria and improving embolization techniques, patient-reported the success of symptom relief after treatment remains variable at 60%–100%. There is always a difference between technical success and patient-reported subjective success in all areas of medicine, but some studies showing lower success rates are surprising if the refluxing veins have been successfully ablated. Hence, there is a need for further research into this area to identify optimal patient selection.

### Scoring Systems for Pelvic Congestion Syndrome

It is clear that PCS is a condition that can cause severe morbidity and reduction in quality-of-life in many women and probably also in some men. The association with PVR is strong, although the mechanism of how PVR causes symptoms is not currently clear. It is much easier to see how PVR is associated with varicose veins externally as this can be mapped by duplex ultrasonography. The role of venous compression syndromes in PCS appears to be overstated in the past and is probably only relevant in a small minority of cases.

Recently, I published an article calling for the classification of PVR based on the number of truncal pelvic veins involved, and the severity of venous reflux in these veins. The optimal way...
to identify both which veins are involved and to assess reflux appears to be by transvaginal venous duplex ultrasound scanning using the Holdstock protocol (outlined in another paper from our clinic in this journal). I made the point that papers were starting to appear comparing groups of women with or without PCS. As suggested in my article, if we want to understand any disease process, we need to upgrade the severity of the disease rather than just say if it is present or not. Hence, we are working on a grading system to use objective measurements to define the severity of PVR. However, whether this relates to the symptoms of PCS only further research will show.

There has been a push recently for a “CEAP” type scoring system that can be used for PCS and two American doctors, Mel Rosenblatt and Mark Meissner, have been touring many venous meetings presenting their alternative views as to how such a scoring system would work.[39] However, both proposals are cumbersome and inelegant at present, and it is unlikely that there will be a simple “CEAP type” scoring system for PCS in the near future. It is likely that as we start to understand the disease process better, we may be able to identify a scoring system that will be relevant and easy-to-use. Indeed, just as most doctors are aware of the CEAP scoring system that actually only use the “C” or clinical part of it, a similar system for PCS will probably end up with a simple part of the scoring system that most people will use regularly, and a more complex part of it that can only be used in certain circumstances such as research projects and papers.

However, Kathy Gibson has published some early work on a disease-specific assessment tool for patients with PCS. This should provide a good basis for understanding more about PCS in the future.[40] The symptomatic improvements of 60%–100% after treatment suggest that there is a need for better diagnosis, investigation, and treatments.

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