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Definition of pelvic venous disease and the new SVP classification

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ABSTRACT

Chronic pelvic pain (CPP) is often diagnosed by gynecologists. In recent years, the realization that venous insufficiency in the pelvic or retroperitoneal area can also cause CPP has led to the involvement of vascular physicians in the diagnosis and treatment. Differentiating pelvic venous disease (PeVD) from gynecological, internal or even orthopedic complaints poses a challenge. A comprehensive study of the patient's medical history and a detailed description of symptoms are of paramount importance. The symptoms of PeVD depend on the veins involved. Whereas pelvic pain is mainly an expression of an insufficiency of the ovarian vein and/or the internal iliac vein, left flank pain and hematuria are caused by an obstruction of the renal vein. Symptoms may also be associated with atypical varicose veins in the vulva, perineum, or legs. Anatomical gaps, pelvic escape points, allow them to communicate with the branches of the insufficient internal iliac veins and with the ovarian. As an effective tool for reporting PeVD patient populations the symptoms-varices-pathophysiology (SVP) classification was published in 2021. It is thought to be the initial point of both the treatment standard and identifying homogenous patient populations for clinical trials.

Keywords: Chronic pelvic pain, May-Thurner syndrome, Nutcracker syndrome, ovarian vein insufficiency, pelvic congestion syndrome, SVP classification.

Up to 10% of gynecological consultations are for chronic pelvic pain (CPP) defined as pelvic pain that persists for more than six months and is not directly related to menstruation, pregnancy, coitus, or intestinal diseases.[1,2] Gynecological factors such as endometriosis, ovarian tumors, fibroids and inflammatory changes are differentiated from extra-gynecological factors (e.g., interstitial cystitis, urolithiasis, irritable bowel syndrome, postoperative adhesions, as well as illnesses of neurological and psychosomatic origin). The precise cause cannot be identified with certainty in more than half of the cases.[1,2]

In addition to these factors, CPP can also be caused by pelvic congestion syndrome (PCS). In 2021, the term PCS was replaced in the international nomenclature by the term pelvic venous disease (PeVD) due to its etiological vagueness.[3]

Pelvic congestion syndrome was first described in 1831.[4] In 1949, the gynecologist Lindsay Watt[5] published a case series of 32 women who suffered from pelvic pain, back pain, an urge to defecate, and a sudden need to urinate. The cystoscopy showed congestion in the veins of the fundus of the bladder, leading to a conclusion that there was a relationship between the complaints and the detected increase in blood in the pelvis. The women reported an alleviation of discomfort after they stopped wearing corsets, resulting in a reduction in the intra-abdominal pressure.

Patients are often referred to vascular specialists by gynecologists after transvaginal ultrasound detects

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pelvic varicose veins. As this finding is not based on a non-specific compression of the pelvic organs, but on venous compression and often venous reflux, the term PeVD is a more appropriate term.

SYMPTOMS OF PeVD

A PeVD may cause a number of different symptoms. According to the 2009 VEIN-TERM Transatlantic Interdisciplinary Consensus Document, symptoms of the condition include pelvic pain, perineal heaviness, urinary urgency and postcoital pain, which are caused by ovarian or pelvic vein reflux and/or strong pressure as a result of an obstruction. Symptoms may also be associated with atypical varicose veins in the vulva, perineum, or legs.[6] The pain, itself, is described by patients as a shooting or pressing pain and can affect the lower abdomen or the lesser pelvis, but can also radiate to the hip. Typically, the women are still of childbearing age and there seems to be a correlation to the number of pregnancies they have had. In men, varicocele is an analogous diagnosis. Left flank pain and hematuria may also be a result of PeVD.

The non-specific symptoms often place the patient on a long and agonizing journey until the definite diagnosis is established. In addition to receiving referrals from gynecologists, vascular specialists often see patients referred by orthopedists and urologists.[7-10] Various manifestations suggest that different vascular regions may be compromised in PeVD.

The symptoms depend on the veins involved. Whereas pelvic pain is mainly an expression of an insufficiency of the ovarian vein and/or the internal iliac vein, left flank pain and hematuria are caused by an obstruction of the renal vein.

VENOUS COMPRESSION AND VENOUS INSUFFICIENCY - THE PATHOPHYSIOLOGY OF PeVD

Meissner and Glovicki[11] defined three basic pathomechanisms that can lead to the development of a PeVD: compression of the left renal vein (L-RV; so-called nutcracker syndrome), reflux of the left gonadal vein, and compression of the left iliac vein (Figure 1, Table 1).

1. Compression of the L-RV

The L-RV crosses the abdominal aorta before joining the inferior vena cava (IVC). It also crosses under the superior mesenteric artery. External compression of the L-RV is possible due to its topographical proximity, although this depends on the angle which it exits the aorta, but also on an increasing displacement of the L-RV as retroperitoneal adipose tissue increases. This is also referred to "anterior nutcracker phenomenon" or "renal vein entrapment" in the literature. In rare cases, the L-RV runs behind the aorta and can, then, be compromised by the spine (i.e., posterior nutcracker phenomenon).

The resulting stenosis leads to a congestion of venous outflow from the left kidney and also causes venous hypertension. This can result in the development of left flank pain and hematuria. Venous hypertension in the renal reservoir is compensated, when left ovarian vein (L-OV) insufficiency develops, thereby leading to retrograde filling of the pelvic reservoir with the clinical implications described below.

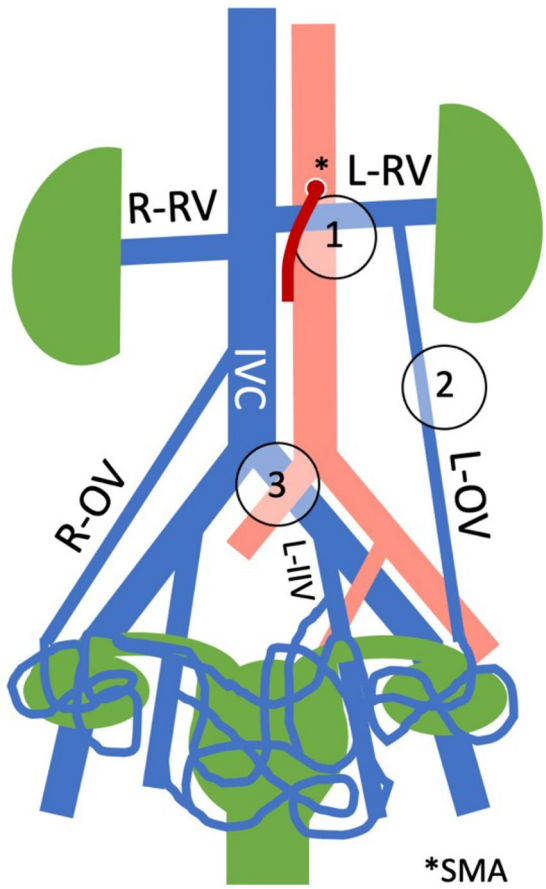

Figure 1. Illustration of the anatomical structures whose pathological changes underlie pelvic venous disease (modified from Hirsch and Wohlgemuth[12]).
1: Compression of the left renal vein (nutcracker phenomenon); 2: Insufficiency of the left ovarian (or testicular) vein; 3: Compression of the left iliac vein (May-Thurner syndrome); L-RV: Left renal vein; R-RV: Right renal vein; L-OV: Left ovarian vein; R-OV: Right ovarian vein; L-IIV: Left internal iliac vein; SMA: Superior mesenteric artery.

Table 1. Pathomechanisms of pelvic venous disease with leading symptoms and compensation mechanisms		
Compression of the left renal vein	• Flank pain • Hematuria	• Drainage in the pelvic reservoir via an insufficient gonadal vein
Insufficiency of the left gonadal vein and/or internal iliac vein	• Lower abdominal pain • Feeling of heaviness and fullness • Dyspareunia • Varicocele in men	• Drainage in the extrafascial venous system of the legs via pelvic escape points
Compression of the left iliac vein	• Edema of the leg • Atypical varicose veins	• Drainage in the pelvic reservoir via the internal iliac vein

2. Reflux of the L-OV and/or testicular vein and the left internal iliac vein (L-IIV)

In contrast to the right ovarian vein, the L-OV does not drain directly into the IVC, but into the L-RV, which in turn is drained by the IVC. It often has two to three valves. The caliber of the L-OV is approximately 3 mm and increases with the aging. The L-OV and the L-IIV drain the venous blood of the urogenital organs, as well as the surrounding venous plexuses (parametrium, mesosalpinx in females, pampiniform plexus in males), which form the pelvic reservoir.

Nearly half of women have an insufficiency or reflux in the L-OV, while 13 to 15% have no valves in the L-OV.[13] Both the insufficiency and the caliber of the L-OV increase with the number of pregnancies. An L-OV diameter of >6 mm is associated with a predictive value of 96% for the presence of intrapelvic varices.[14]

Insufficiency of the L-IIV may also develop. The insufficiency causes venous hypertension in the lower abdominal organs which can trigger the alluded to discomfort in women or a varicocele in men. This high pressure may be relieved via collaterals to the epifascial veins through the pelvic escape points, leading to the formation of atypical varices, when there is no or only mild pelvic discomfort.

3. Compression of the left common iliac vein (L-CIV)

Another cause of a PeVD may be a compression or occlusion of the L-CIV. It lies ventral to the bony structures of the fourth or fifth vertebrae and dorsal to the right iliac artery, before it joins the IVC. In addition to the mechanically induced compression that this alone can cause, an intraluminal fibrotic ribbon- or mesh-shaped vessel change is often located near the junction, which can obstruct the venous outflow. This condition is called May-Thurner syndrome after the Austrian pathologists May and Thurner[15] first described the phenomenon. In this case, a congestion of the deep veins in the left leg can be expected, causing the usual symptoms. This condition can also be compensated. Additional insufficiency of the L-IIV causes the pelvic reservoir, the venous plexus in the lesser pelvis to fill, producing the symptoms described in Section 2.

Pelvic escape points

Pelvic escape points (also known as pelvic leak points) are anatomical gaps through which branches of the venous plexus of the pelvis are connected to the epifascial veins of the legs. This allows them to communicate with the branches of the internal iliac veins and with the ovarian veins (pudendal and gluteal veins). They represent the pre-selection points of atypical varicose veins on the vulva, perineum or scrotum, and in the groin area. At the same time, these pelvic escape points may relieve venous hypertension in the pelvis. The topography varies widely between individuals, as does the terminology. Meissner and Gloviczki[11] described four pelvic escape points on

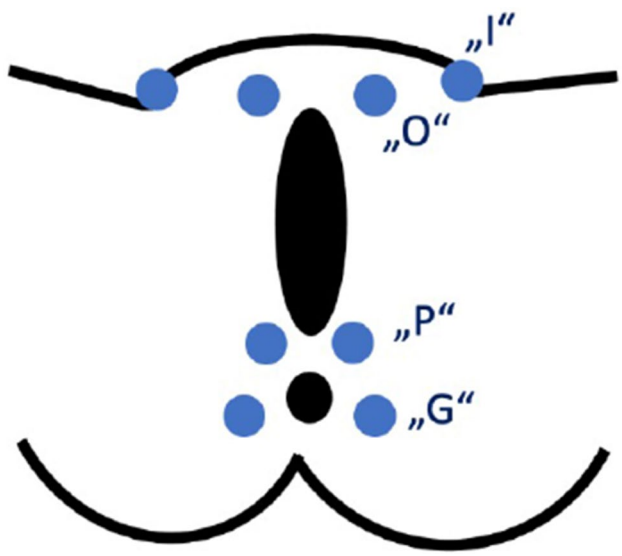

Figure 2. Illustration of the pelvic escape points.

Table 2. Anatomical connections between branches of the external iliac veins and branches of the internal iliac veins via the pelvic escape points

I point (inguinal escape point)	• Inguinal canal	• Ovarian vein-labial vein-great saphenous vein (GSV)
O point (obturator escape point)	• Canalis obturatorius	• Obturator vein-epifascial veins of the medial compartment of the thigh
P point (perineal escape point)	• Foramen infrapiriforme	• Pudendal veins-medial compartment of the thigh, GSV
G point (gluteal escape point)	• Foramen infrapiriforme	• Gluteal veins-veins of the posterior compartment of the thigh

each side (Figure 2, Table 2). Delfrate and Mendoza[16] additionally define a clitoral venous escape point on both sides and distinguish between two gluteal escape points each.

Moreover, these pathologies can occur together. Atypical localizations for venous disorders in the lesser pelvis are possible, if they are topographically adjacent to bony or fibrous structures (ligaments) or arteries. Manifestations on the right side can also be observed. They usually require special diagnostic finesse. Surprisingly, no correlation with the body mass index can be confirmed, unlike in the case of varicose veins in the leg and chronic venous insufficiency.[17]

THE SYMPTOMS-VARICES-PATHOPHYSIOLOGY (SVP) CLASSIFICATION OF PeVD

Although the etiology of PeVD varies, it is associated with hypertension and dilation of the pelvic venous plexus due to primary and/or secondary pelvic vein incompetence. Before the importance of venous disorders of the abdomen and pelvis has become recognized, progress in the field has been hindered due to using previously syndromic terminology such as May-Thurner, pelvic congestion, and nutcracker syndromes. This historical nomenclature fails to describe pathophysiology of PeVD, due to nature of complex and interrelated pelvic venous circulation.

The first description of the venous congestion syndrome of the pelvis was symptoms consisting of pelvic pain, pelvic varicosities, dysmenorrhea and dyspareunia in 1948.[18] Classifications for PeVD have been suggested in the literature since then.[19,20] However, previous classification systems for PeVD were insufficient to describe a homogeneous patient population for similar clinical presentation resulting from different underlying pathophysiologies. Therefore, recently published the SVP classification issued by the American Vein and Lymphatic Society International Working Group was designed as an initial point to clarify the classification of PeVD.[3] The main goals of SVP classification are to identify a homogeneous patient population, develop equipment to investigate treatment, and plan scientific clinical trials to optimize management of patients with PeVD.[3]

Clinical symptoms and radiological signs have been also described for SVP classification system to minimize interobserver variability. The PeVD, venous origin of renal symptoms, CPP, pelvic origin of extrapelvic symptoms, venous claudication were defined as clinical symptoms. The L-RV obstruction, pelvic varicose veins, gonadal vein reflux, iliac venous obstruction, internal iliac venous reflux, pelvic origin of extrapelvic varices were defined as radiological signs. Four anatomic zones of the abdomen and pelvis were defined for explaining the symptoms, signs, and pathophysiologic manifestations of PeVD as follows:

Zone 1: Renal hilum

Zone 2: Venous plexuses of the pelvis

Zone 3: Extra-pelvic vessels of pelvic origin.

Zone 4: Deep and superficial veins of lower extremity

Zone 1, Zone 2, and Zone 3 are included in the SVP classification. However, lower extremity varices (Zone 4) are best described by the Clinical-Etiological-Anatomical-Pathophysiological (CEAP) classification and are not included in the SVP classification. Therefore, the authors have attempted to provide these definitions compatible with the CEAP classification.

The SVP classification consists of the three main domains: symptoms (S), varices (V), and pathophysiology (P). Patient's clinical symptoms (“S”) domain is determined by subscripts ranging from 0 to 3. This section is arranged according to anatomical regions descending from renal veins to lower extremities (Table 3). Symptoms in the table is defined in this classification system separately. Moreover, radiological signs are also defined for

Table 3. SVP classifications: Symptoms ("S")

S0	No symptoms of a PeVD (no renal, pelvic, or extra-pelvic symptoms)
S1	Renal symptoms of venous origin
S2	Chronic pelvic pain of venous origin
S3	Extra-pelvic symptoms of venous origin
a	Localized symptoms (pain, discomfort, tenderness, itching, bleeding, and superficial venous thrombosis) associated with veins of the external genitalia (vulva and scrotum)
b	Localized symptoms associated with pelvic origin nonsaphenous veins of the leg. These include those related to pelvic origin varices of the posteromedial thigh (pain, discomfort, tenderness, itching, superficial venous thrombosis) as well as those related to sciatic/tibial nerve varices (pain, paresthesia). More generalized lower extremity symptoms and signs, such as heaviness and swelling, are classified with CEAP not SVP
c	Venous claudication*

SVP: Symptoms-Varices-Pathophysiology; PeVD: Pelvic venous disorder; * Must include Clinical-Etiological-Anatomical-Pathophysiological (CEAP) classification for full characterization of lower extremity symptoms.

Table 4. SVP classifications: Varices ("V")

V0	No abdominal, pelvic, or pelvic origin extra-pelvic varices on clinical or imaging examination
V1	Renal hilar varices
V2	Pelvic varices
V3	Pelvic origin extra-pelvic varices.
a	Genital varices (vulvar varices and varicocele)
b	Pelvic origin lower extremity varicose veins arising from the pelvic escape points and extending into the thigh. Includes visible varicosities, typically over the posteromedial thigh, as well as sciatic varices and other refluxing veins transitioning the pelvic floor which are visualized only with ultrasound.

SVP: Symptoms-Varices-Pathophysiology; Must include Clinical-Etiological-Anatomical-Pathophysiological classification for full characterization of lower extremity varices.

standardization. Nonetheless, some authors have suggested that some definitions, particularly for radiological signs, should be revised in the future.[21] Patient's varices ("V") domain is determined ranging from 0 to 3 like symptoms ("S") domain (Table 4).

The pathophysiology (P) domain is consisted of three subdomains called anatomical (A), hemodynamic (H), and etiological (E). Anatomical subdomain indicates with anatomical abbreviations of abdomen and pelvis venous structures. Underlying hemodynamic (H) irregularities-reflux (R), obstruction (O) or both (R, O) are shown (Table 5). The etiology of pelvic venous pathology (PE) is defined as thrombotic (T), non-thrombotic (NT), or congenital (C) (Table 6). All three components are indicated by a subscript in "P" category.[3]

Finally, the classification of patients is made with the SVP classification scoring table (Table 7). According to this classification, the pelvic disease score of an individual is shown as SVPA, H, E.[3] A smart phone application has also been developed to classify patients easily using the SVP tool.[3,22]

Some causes of CPP and pelvic varices in women are illustrated by the SVP classification below.[22]

Primary bilateral ovarian vein reflux: $S_2\ V_2\ P_{BGV,R,NT}$

Left renal vein obstruction and secondary L-OV reflux: $S_2\ V_{1,2}\ P_{LRV,O,NT;\ LGV,R,NT}$

Left common iliac vein obstruction and L-IIV trunk and tributary reflux: $S_2\ V_2\ P_{LCIV,O,NT;\ LIIV,R,NT}$.

In conclusion, SVP classification is a new tool for reporting PeVD patient populations. It is thought to be the initial point of standard treatment indications,

Table 5. Hemodynamics ("H")

Obstruction (O)	Thrombotic or non-thrombotic (venous compression) venous obstruction
Reflux (R)	Thrombotic or non-thrombotic reflux

Table 6. Etiology ("E")

Thrombotic (T)	Venous reflux or obstruction arising from a previous episode of DVT
Non-thrombotic (NT)	Reflux arising from a degenerative process of the vein wall or proximal obstruction; Obstruction arising from extrinsic compression
Congenital (C)	Congenital venous or mixed vascular malformations

DVT: Deep vein thrombosis.

Table 7. SVP classification scoring table

Symptoms (S)		Varices (V)		Anatomy/pathophysiology (P)			
No symptoms of a PeVD	0	No pelvic varices	0		A	H	E
Renal	1	Renal	1				
Pelvic	2	Pelvic	2				
Extra-pelvic	3	Extra-pelvic	3				T
Genital	3_a	Genital	3_a				NT
Lower extremity symptoms	3_b	Lower extremity varicose	3_b	L	RV	O	C
Venous claudication	3_c			R			T
				L	GV	O R	NT
				B			C
				R			T
				L	CIV	O R	NT
				B			C
				R			T
				L	IIV	O R	NT
				B			C
				R			T
				L	EIV	O R	NT
				B			C
				R			T
				L	PELV	O R	NT
				B			C
S		V		Psegment1, H,E; segment 2, H,E			

SVP: Symptoms-varices-pathophysiology; PeVD: Pelvic venous disorder; RV: Renal vein; GV: Gonadal vein; CIV: Common iliac vein; IIV: Internal iliac vein; EIV: External iliac vein, PELV: Pelvic escape veins; R: Right; L: left; B: Bilateral; O: Obstruction; R: Reflux; T: Thrombotic; NT: Non-thrombotic; C: Congenital

methods, technique, and uniform reporting. The SVP instrument accurately defines the diverse patient populations with PeVD, an important step in improving clinical decision making, developing disease-specific outcome measures, clinical communication, and identifying homogenous patient populations for clinical trials.

Declaration of conflicting interests

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