A Standard for Terminology in Chronic Pelvic Pain Syndromes: A Report From the Chronic Pelvic Pain Working Group of the International Continence Society

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Aims: Terms used in the field of chronic pelvic pain (CPP) are poorly defined and often confusing. An International Continence Society (ICS Standard for Terminology in chronic pelvic pain syndromes (CPPS) has been developed with the aim of improving diagnosis and treatment of patients affected by chronic pelvic pain syndromes. The standard aims to facilitate research, enhance therapy development and support healthcare delivery, for healthcare providers, and patients. This document looks at the whole person and all the domains (organ systems) in a systematic way. **Methods:** A dedicated working group (WG) was instituted by the ICS Standardisation Steering Committee according to published procedures. The WG extracted information from existing relevant guidelines, consensus documents, and scientific publications. Medline and other databases were searched in relation to each chronic pelvic pain domain from 1980 to 2014. Existing ICS Standards for terminology were utilized where appropriate to ensure transparency, accessibility, flexibility, and evolution. Consensus was based on majority agreement. **Results:** The multidisciplinary CPPS Standard reports updated consensus terminology in nine domains; lower urinary tract, female genital, male genital, gastrointestinal, musculoskeletal, neurological aspects, psychological aspects, sexual aspects, and comorbidities. Each is described in terms of symptoms, signs and further evaluation. **Conclusion:** The document presents preferred terms and definitions for symptoms, signs, and evaluation (diagnostic work-up) of female and male patients with chronic pelvic pain syndromes, serving as a platform for ongoing development in this field. *Neurourol. Urodynam.* © 2016 Wiley Periodicals, Inc.

Key words: bladder pain syndrome; chronic pelvic pain syndromes; comorbidities; condition; disease; domain; female genital pain; gastrointestinal pain; Hunner lesion; hypersensitive bladder; interstitial cystitis; lower urinary tract pain; male genital pain; musculoskeletal pain; neurological aspects; phenotype; psychological aspects; sign; sexual aspects; symptom; syndrome

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INTRODUCTION

This is the first International Continence Society (ICS published Standard of Chronic Pelvic Pain Syndromes (CPPS). Global standardization of terms and clear definitions are essential for scientific and clinical progress. Furthermore, meaningful coding of diseases, nationally and internationally, depends on accepted terminology. Inappropriate and unclear coding and definitions have negative effects not only on diagnosis, but also on the patient's ability to obtain appropriate treatment, reimbursement, and social benefits. The International Continence Society (ICS has led the way in the development of Standards for terminology of lower urinary tract function and dysfunction,[1] and the need for a Standard in CPPS was identified by the ICS Standardisation Steering Committee (SSC).

Chronic pelvic pain (CPP) is the most common indication for referral to women's health services, and accounts for 20% of all outpatient appointments in secondary care.[2] This leads to a substantial burden on limited health care resources. For example, $881.5 million are spent per year on its outpatient management in the USA, while an estimated £158 million are spent annually on management in the United Kingdom National Health Service.[2] CPPS are multifactorial and multidisciplinary conditions, and terminology can vary according to which specialist is looking at the patient. This document is an endeavour to look at the whole person and to consider all the domains involved. Each domain is described separately.

Pain in the pelvic area potentially includes urologic, gynecologic, gastrointestinal, musculoskeletal, neurologic and/or rheumatologic etiology, with psycho-social aspects, and hence must be regarded as a multidisciplinary issue. A taxonomy of the relevant elements of CPP was provided by the International Association for the Study of Pain (IASP).[3,4] Complementing the taxonomy, the European Association of Urology (EAU) Guidelines on Chronic Pelvic Pain provide a comprehensive overview of basic science pertaining to pelvic pain, clinical workup and management of CPPS.[5] This ICS Standard should be seen as complementary to other CPPS standards and guidelines. Its aims are to:

1. Describe the nine clinical domains involved in CPPS (summarized in Tables I–XI).
2. Define terminology.
3. Develop an evaluation guideline for each domain.
4. Establish a process for evolving terminology in response to scientific and clinical development and patient need.

This Standard for CPPS Terminology should facilitate future research and therapy development, improve cost effectiveness, and ensure access by the patient to appropriate treatment, reimbursement, and social benefits.

METHODS

The CPPS Standard was developed according to the published methodology of the ICS Standardisation Steering Committee (SSC).[6] The Working Group (WG) and Chairperson were selected by an independent SSC sub-committee following an open advertisement. The WG comprised a multi-disciplinary group of health care providers, a basic science researcher, and a patient advocate. Activities of the WG and contributions of individual members were recorded in an open forum on the ICS website. The WG developed an outline of proposed content at an open workshop at the ICS annual scientific meeting in Beijing (2012). Successive iterations of the draft standard involved electronic communications, teleconferences, and face-to-face meetings. The WG reviewed documents that provided historical and research insight into the multidisciplinary approach to the evaluation of female and male CPPS.

A literature review covered the period 1980–2015 and extracted sources from electronic database searches, including MEDLINE and Cochrane. In addition, cross-referencing was done for existing relevant guidelines and consensus documents, notably:

- The EAU Guidelines on Chronic Pelvic Pain,[5,7] which place CPP in the clinical context.
- The American Urological Association (AUA) guidelines for the diagnosis and the treatment of IC/BPS.[8,9]
- The International Continence Society (ICS) joint/ICS joint report on the terminology for female pelvic floor dysfunction,[10] which covers terminology for female sexual dysfunction, genital pain, and pudendal neuralgia.
- The IASP Taxonomy, which classified pain on the basis of "organ + pain + syndrome" and applied it to pain of urogenital origin.[3]
- In 2008, the International Society for the Study of Bladder Pain Syndrome (ESSIC) published diagnostic criteria, classification, and nomenclature for bladder pain syndrome (BPS).[11]
- The East Asian IC Study Group/Society of Interstitial Cystitis of Japan (SICJ) guidelines, which revived the concept of hypersensitive bladder.[12]
- The Functional Gastro-Intestinal Disorder Society (FGIDS) Rome III Diagnostic Criteria[13] for gastro-intestinal disorders.

RESULTS

The WG identified the following nine domains, each of which are considered in terms of symptoms, signs, and further evaluation.

I. Lower Urinary Tract Domain

A. Bladder.
B. Urethra.

II. Female Genital Domain

A. Vulva, vestibule, and clitoris.
B. Intra-abdominal female genital pain.
C. Pelvic floor muscle pain.

III. Male Genital Domain

A. Prostate.
B. Scrotum.
C. Epididymis.
D. Testicle.
E. Penis.
F. Urethra.
G. Sexual Pain.

IV. Gastro-Intestinal Domain

A. Anorectum.
B. Colorectum.

V. Musculoskeletal Domain

A. Pelvic muscle pain.
B. Coccyx pain syndrome.
C. Pelvic Joint, ligament, or bony pain.

VI. Neurological Domain

A. Complex Regional Pain Syndrome (CRPS).
B. Somatic neuropathic pain.
C. Pain following mesh surgery.

VII. Psychological Domain

A. Worry, anxiety, and fear.
B. Depression and depressed mood.
C. Catastrophizing.

VIII. Sexual Domain

A. Sexual desire disorder.
B. Sexual arousal disorder.
C. Orgasmic disorder.
D. Sexual pain disorder.

IX. Comorbidities

A. Allergies.
B. Chronic pain and fatigue syndromes.
C. Systemic autoimmune syndromes/disease.
D. Extraintestinal manifestations of inflammatory bowel disease.

TAXONOMY

A. Pain—A subjective phenomenon described as an unpleasant sensory and emotional experience associated with actual or potential tissue damage, or described in terms of such damage.[14]

a. Nociceptive pain: arises from actual or threatened damage to non-neural tissue and is due to the activation of nociceptors.
b. Somatic pain: arises from bone, joints, muscles, skin, or connective tissue and is normally achy or throbbing and well localized.
c. Visceral pain: arises from visceral organs, with involvement of the organ capsule with aching, and is localized. There is obstruction of hollow viscus, causing intermittent cramping, which is poorly localized.[15]

   i. Nociceptive: direct injury or lesion of an internal organ such as: bladder stone, surgical injury.
   ii. Inflammatory: acute/chronic inflammation of an internal organ such as urinary tract infection, pelvic inflammatory disease, colitis, endometriosis.
   iii. Neuropathic: primary lesion of visceral nerves such as neuritis following mesh placement.

d. Centrally generated pain/deafferentation pain: may result from injury to either the peripheral or central nervous system, leading to burning pain below the level of the lesion. It can be sympathetic-nervous system maintained pain, which may result in chronic regional pain syndrome (CRPS). There is increased responsiveness of nociceptive neurons in the central nervous system to normal or sub-threshold afferent input.
e. Hypersensitivity: increased nerve activity from a standardized stimulus with an expected tissue/clinical response. The underlying mechanism remains to be defined.
f. Central sensitization:[16] nociceptor sensitization results in synaptic strengthening by incoming afferent volleys (sensitization) and is expressed as hyperalgesia (a form of non-associative learning characterized by an increase in responsiveness upon repeated exposure to a stimulus).[17]

B. Pain Experience—According to the most common views, pain constitutes the internal perception of bodily damage. It is unknown whether chronic pelvic pain syndromes (CPPS) are primarily an abnormal perception of a normal stimulus or a normal perception of an abnormal physiologic sensory stimulus.[14] [FN1]

C. Psychology of Pain—Pain is modulated by cognitive factors and emotional experience, memory, attention, and context represented in descending modulation of pain, affecting pain experience from moment to moment and longer term. Pain has an impact on many aspects of daily life, affecting mood, sleep, relationships, and activities. Therefore, attention to the psychological aspects of pain is an important part of effective assessment and treatment.[18,19]

D. Neurobiology of Pain—Alterations in gut and bladder motility, visceral perception and central processing of pain and motor function due to abnormalities in the visceral and central nervous systems may account for the symptoms.[18] [FN2]

E. Chronic Pelvic Pain—Chronic pelvic pain is characterized by persistent pain lasting longer than 6 months or recurrent episodes of abdominal/pelvic pain, hypersensitivity or discomfort often associated with elimination changes, and sexual dysfunction often in the absence of organic etiology.[20]

F. Symptoms and Signs of Chronic Pelvic Pain Syndromes

a. Symptoms: The subjective indicator of a disease or change in condition/syndrome/phenotype as perceived by the patient, caregiver or partner which may lead him/her to seek help from healthcare professionals.[21] The main symptom in CPPS is pain and will be described in relation to its domain and its perception. Complaint: what the patient describes when prompted by the physician.
b. Signs are observed by the physician including simple means to verify symptoms and quantify them. To evaluate and discover all the signs, a full evaluation of the pelvis and body is necessary as multiple intra and extra-pelvic domains (organ systems) are commonly involved. It is necessary to attempt to identify all of the pain generators.[11,21]

G. Condition, Disease, Syndrome

a. A condition is defined by the presence of observations associated with characteristic symptoms or signs and/or evidence of relevant pathological processes.[7]
b. A disease is a disordered or incorrectly functioning organ, part, structure, or system of the body resulting from the effect of genetic or developmental errors, infection, poisons, nutritional deficiency or imbalance, toxicity, or unfavourable environmental factors; illness; sickness; ailment.
c. A syndrome is a complex of concurrent symptoms and signs that is collectively indicative of a disease, dysfunction or disorder in the absence of obvious pathology. (NEW) Example: Interstitial Cystitis/Bladder Pain Syndrome (IC/BPS) is one of the Chronic Pelvic Pain Syndromes.[1] [FN3]

H. Characteristics

a. Duration of pain: Six months or more of persistent pain.[FN4]
b. Location of pain: Pelvis, lower abdomen, low back, medial aspect of thigh, inguinal area, perineum.
c. Perception of pain: Patients may describe the pain as sharp, burning, aching, shooting, stabbing, pressure or discomfort, sexual pain (dyspareunia).[22] [FN5]
d. Modality of pain (7): Persistent and/or continuous, recurrent and/or episodic and/or cyclic (related to menstrual cycle).

[FN1] Diagnosis is often based on the presence of clinical symptoms. The diagnosis of CPP is confirmed by applying symptom-based criteria and pursuing further diagnostic evaluation to exclude organic disease. Validation of symptom-based criteria is a process; it is not carved in stone and is easy to change as new data on its underlying pathophysiology emerge.[3]

[FN2] The brain-visceral axis and biopsychosocial model have been used to explain how intrinsic and extrinsic stimuli modulate disease expression.[14,18]

[FN3] This is an update of the ICS Standardisation Sub-committee report on the Standardisation of Lower Urinary Tract Function:[1]

[FN4] In different guidelines and standardisation documents, the duration varies from 6 weeks to 6 months.

[FN5] Some patients describe pain as an ache, soreness or simply discomfort, while cultural differences may influence perception of pain. For example, some patients describe an unpleasant sensation or pressure or discomfort, but do not consider these to be true pain. Memories, emotions, thoughts, expectations and culture are now believed to influence how people perceive pain.[22]

I. Phenotype—Subgroup of patients within a condition, disease, or syndrome that share similar expression of specific symptoms, signs and diagnostic parameters: Example: Irritable bowel syndrome has three phenotypes: constipation, diarrhea, or mixed constipation/diarrhea.[5,23,24] [FN6]

Phenotyping is currently in its infancy with regard to evidence and will increase in importance in the future to aid in identifying specific patient pools for research and treatment.[5,23]

J. Domain (organ system)—Lower urinary tract, female genital, male genital, gastro-intestinal, musculoskeletal, neurological, psychological, sexual, and comorbidities are domains involved in chronic pelvic pain syndromes (CPPS).

Tables I–IX are a summary of the appropriate domain for domains I–IX and appear in the Symptoms section.[25] [FN7]

INDIVIDUAL PATIENT ASSESSMENT

Section 1: Symptoms

The first and most important step is to obtain a thorough history of the patient's perception of her/his pain or discomfort. The common complaints are the most prevalent symptoms.

Ask about duration (at least 6 months), perception (identify inciting event and/or triggers), and modality (persistent/recurrent).

I. Lower Urinary Tract Domain (Table I)

A. Bladder

Common complaints include: increased urinary frequency day and night, urgency, hypersensitivity, pain, pressure, discomfort, pain with filling, hesitancy, intermittency, feeling of incomplete emptying. Pain/hypersensitivity related to the bladder provides an umbrella for hypersensitive bladder, interstitial cystitis/bladder pain syndrome, and interstitial cystitis with Hunner lesion.[26–28] Urgency: A compelling need to urinate which is difficult to defer (pain, pressure, discomfort).[1,5,11,21,30–33] [FN8] [FN9] The Working Group identified the following adjustments as applying more descriptively, for example, to Interstitial Cystitis/Bladder Pain Syndrome patients: a compelling need to urinate, due to pain or an unpleasant sensation, that is difficult to defer.[FN10] [FN11] [FN12]

As there are differences in symptoms as well as in the perception and experience of pain, the WG agreed to distinguish:

a. Hypersensitive Bladder (HSB)(Japanese and East Asian guidelines). Hypersensitive bladder symptoms (increased bladder sensation, usually associated with increased urinary frequency day and night, with or without bladder pain) in the absence of pathology explaining the symptoms.[29,30] [FN13]
b. Interstitial Cystitis/Bladder Pain Syndrome (IC/BPS). Persistent or recurrent chronic pelvic pain, pressure or discomfort perceived to be related to the urinary bladder accompanied by at least one other urinary symptom such as an urgent need to void or urinary frequency.[11]
c. Interstitial Cystitis (IC) with Hunner lesion has the same symptoms as IC/BPS.[31] [FN14] [FN15]. Pain in IC/BPS and IC with Hunner lesion may be pain, pressure or discomfort, which may increase with bladder filling. Possible locations of perceived discomfort and pain are the pelvis, lower abdomen/suprapubic area, low back, medial aspects of the thigh, inguinal area, or multiple pain sites.[11] Descriptors/Perception of pain[5] include: "Sharp, burning, ache, shooting, stabbing, pressure, discomfort."

B. Urethra

Urethral pain is perceived to be in the urethra, usually when voiding, with increased day- and night-time frequency. It may be combined with a feeling of dull pressure, and sometimes radiates toward the groin, sacrum and perineum.[1,7] The terms "chronic urethritis" and "urethral syndrome" are no longer recommended.[5,23]

i. Persistent or recurrent pain.
ii. No history of current infection or other obvious pathology.
iii. May be subsequent to a previous urinary tract infection.

[FN6] This has been adapted from the European Association of Urology Guidelines on Chronic Pelvic Pain.[5]

[FN7] Domains I–V involve intrapelvic organs, VI–IX involve overlying aspects including comorbidities.

[FN8] In the previous ICS LUTS document of 2002, urgency was defined as: the sudden complaint of a compelling desire to pass urine, which is difficult to defer.[1]

[FN9] This was a change from a previous definition in 1988 which stated that urgency may be associated with two types of dysfunction: (i) Overactive detrusor function (motor urgency), and (ii) Hypersensitivity (sensory urgency).[30]

[FN10] The change in definition in 2002 with introduction of the word "sudden" effectively restricted this term to overactive bladder syndrome and there was no mention of another sensation of urgency (urgent need to void) due to pain or hypersensitivity.

[FN11] The term hypersensitive bladder is a revival of an earlier ICS Document.[30]

[FN12] Hunner lesion is preferable to Hunnerś ulcer.

[FN13] There is currently global discussion as to whether Hunner lesion should/could be completely separated from IC/BPS and if so what it should be called. It is felt that more research is needed to provide sufficient evidence for such a step.[31]

[FN14] The term vulvodynia is no longer recommended.[3]

[FN15] The terms Dyesthetic vulvodynia and Essential vulvodynia are no longer recommended.[3,34]

TABLE I. Lower Urinary Tract Domain

Symptoms	Signs	Evaluation	Syndrome/disease
Increased daytime frequency	Suprapubic tenderness	Questionnaires	Hypersensitive bladder
Increased night-time frequency	Tenderness of the bladder	Voiding diary	Interstitial cystitis/bladder pain syndrome
Urgency	Tenderness of the pelvic floor muscles	Urine analysis	Interstitial cystitis/hunner lesion
Hypersensitivity		Optional: urine culture/cytology	
Pain, pressure, discomfort with bladder filling		Intravesical anesthetic challenge	
Hesitancy		Urodynamics	
Intermittency		Cystoscopy (biopsy)	
Feeling of incomplete bladder emptying			
Frequency/urgency painful urination	Tenderness of the urethra	Urine analysis	Urethral pain

II. Female Genital (Table II)

Common complaints: painful menstruation, abnormal bleeding, pain with intercourse (dyspareunia), discharge, burning, itching, stabbing pain, voiding/defecatory pain, abdominal/pelvic pain (unilateral or bilateral, persistent or cyclic).

Female genital pain is defined as pain perceived in the pelvis, pelvic organs, the vagina and/or the female external genitals.[5,34]

A. Vagina (Vulva, Vestibule, and Clitoris)

1. Pain in the vagina or the external genital organs (vulva, which includes the labia, clitoris and entrance to the vagina).[5 FN14]
2. Generalized vulvar pain syndrome.[4]
   i. Diffuse vulvar pain perceived to be in the vestibule or beyond.
   ii. Dyspareunia.
   iii. Provocation of pain with touch, pressure or friction.[3,34 FN15]
3. Localized vulvar pain syndrome.[4]

Pain is usually provoked with touch, pressure, or friction; example: tight clothing, bicycle riding, tampon use, sexual activity.

   i. Vestibular pain syndrome—Pain localized to one or more portions of the vulvar vestibule.[3,34 FN16 FN17]
   ii. Clitoral pain syndrome—Pain localized to or perceived in the clitoris.

B. Intra-Abdominal Female Genital

1. Ovary
   i. Unilateral or bilateral abdominal/pelvic pain.
   ii. Persistent.
   iii. Cyclic.
2. Pelvic Congestion Syndrome
   i. Pressure, heaviness, dull aching pain in the pelvis and/or in the back.
   ii. Dysmenorrhea.

C. Pelvic Floor Muscle[5] (See Domain V Musculoskeletal Pain)

   i. Urinary/defecatory dysfunction.
   ii. Dyspareunia (see also VIII sexual aspects).
   iii. Pain with sitting.
   iv. Bulging sensation.

D. Female Sexual Pain (See Domain VIII)

[FN16] The terms vulvar vestibulitis, vestibulodynia, and focal vulvitis are no longer recommended.[34]

[FN17] Differential diagnosis and treatable diseases: A history of infection (Pelvic Inflammatory Disease, sexually transmitted diseases, endometriosis, adenomyosis or fibroids, and Mullerian abnormalities should be excluded.

TABLE II. Female Genital Domain

Dyspareunia	Tenderness	Pain Mapping	Vaginal/vulvar/perineal pain
Sharp burning and/or stabbing	Erythema	Q-tip touch sensitivity test	
Provocation of pain with touch			
Dysmenorrhea	Tenderness: uterine, adnexal	Laboratory testing	Intra-abdominal:
Abnormal menstrual bleeding		Pelvic ultrasound	Ovarian Pain
Dyspareunia		Laparoscopy/biopsy	Pelvic congestion
Itching, stabbing, burning pain		CT-scan	Uterine pain
Cyclic, (episodic or persistent)			Tubal pain

III Male Genital Domain (Table III)

Male genital pain syndromes are often associated with symptoms suggestive of lower urinary tract and sexual dysfunction. Common complaints: genital pain, uncomfortable urination, dysuria, sensation of residual urine, increased daytime frequency, slow stream, urgency, dyspareunia.[4,5] Absence of infection, previous operations, or other obvious pathology.

A. Prostate

Persistent or recurrent prostate pain, associated with symptoms suggestive of urinary tract and/or sexual dysfunction. No proven infection or other obvious pathology is present to account for the symptoms.[3,35 FN18 FN19 FN20]

Bladder, perineal, testicular, penile and/or groin pain.

i. Perception of pain: variable.
ii. Persistent or recurrent.
iii. Dyspareunia or erectile dysfunction.
iv. Voiding and post micturition symptoms (for example: hesitancy, intermittency, feeling of incomplete emptying).

B. Scrotum

Chronic scrotal pain (generic term used when site of pain is not clearly in the testis or epididymis.[23 FN21]

i. Persistent or recurrent episodic pain, unilateral or bilateral.
ii. Spontaneous, or reproduced by digital pressure and physical activities.
iii. Pain is not in the skin of the scrotum but perceived within its contents.
iv. Lower urinary tract symptoms or sexual dysfunction.

C. Epididymis

Pain is specific/localized to the epididymis.[FN22]

i. Persistent or recurrent episodic pain.
ii. Spontaneous, or reproduced by digital pressure and physical activities.
iii. Lower urinary tract symptoms or sexual dysfunction.

D. Testicle[23 FN23]

i. Persistent or recurrent episodic pain.
ii. Spontaneous, or reproduced by digital pressure and physical activities.
iii. Lower urinary tract symptoms or sexual dysfunction.

E. Penis

Pain within the penis that is not primarily in the urethra[FN24] and may be:

i. Persistent or recurrent.
ii. Spontaneous, or reproduced by digital pressure and physical activities.
iii. Lower urinary tract symptoms or sexual dysfunction.

F. Urethra

(See Domain I Lower Urinary Tract)

[FN18] Using the National Institutes of Health classification system, prostate pain syndrome may be subdivided into type A (inflammatory) and type B (non-inflammatory).[35] Nickel JC. Prostatitis and related conditions, orchitis and epididymitis. In: Campbell-Walsh, editor. Urology. Philadelphia: Elsevier; 2012.

[FN19] Based on a more general definition, the term prostate pain syndrome (PPS) is used by the European Association of Urology (EAU) instead of the National Institute of Diabetes and Digestive and Kidney Diseases (NIDDK) term chronic prostatitis/chronic pelvic pain syndrome.

[FN20] The terms "Chronic Prostatitis" and "Prostatodynia" are no longer recommended.

[FN21] It may be associated with a congestive epididymitis. Example: after vasectomy. It can result from a mechanical pressure more often than an inflammatory process.

[FN22] It may occur at any age, but the majority of cases are in the mid to late thirties and it may be disabling and associated with anxiety about cancer.[36]

[FN23] Pain is localized to the testis and could be explained by neural plasticity when subsequent to a trauma or disease and this phenomenon can result from the amplification of the pain messages at all levels of nervous system.[22] The previous terms "Chronic Orchitis," Orchalgia," or "Orchiodynia" are no longer recommended.

[FN24] The most common site for referral to the penis is from the bladder outlet.

TABLE III. Male Genital Domain

Symptoms	Signs	Evaluation	Syndrome/disease
Pain	Tenderness on rectal/genital examination	Questionnaires	Prostate pain
LUTS	Urethral discharge	Culture	
Dyspareunia		PSA/biopsy	
Erectile dysfunction		Cystoscopy/biopsy	
Persistent or episodic		Ultrasound	
	Tenderness on physical examination	Questionnaires	Scrotal pain
	Scars	Ultrasound	Epididymal pain
			Testicular pain
			Penile pain

G. Sexual Pain
(dyspareunia; (see Domain VIII)
i. Penile

1. Prior to penetration (example: pain with erection).
2. With penetration.
3. Post coital.

ii. Perineal

1. During intercourse.
2. After intercourse.

iii. Orgasmic Pain (during ejaculation)

1. Penile.
2. Anorectal.
3. Pelvic.

IV Gastro-Intestinal (Table IV)
Common complaints: constipation, diarrhea and obstructive defecation, pain with defecation, bleeding, discharge, cramping abdominal pain, recurrent rectal pain, rectal pressure, burning sensation or aching episodes.[37] [FN25]

A. Anorectum (7) (4)

1. Chronic Proctalgia—rectal pain, more than 20 min of duration per episode, for at least 3 months with symptom onset at least 6 months prior to diagnosis.
   i. Persistent or recurrent rectal pain.
   ii. Rectal pressure or aching episodes.
   iii. In the absence of other causes of rectal pain.
2. Levator Ani Syndrome (the term may refer to the same syndrome as "pelvic floor muscle pain syndrome"/"tension myalgia of the PFM"—see Domain V).
   i. Pain with sitting.
   ii. Pain with defecation.
3. Proctalgia Fugax
   i. Severe recurrent episodic pain localized in the anus or lower rectum.
   ii. Duration seconds to minutes.
   iii. No pain between episodes.

Consider the Symptoms of the Following Treatable Diseases, as They Need to Be Excluded
4. Anal Fissure[38]
   i. Bright red bleeding with bowel movements.
   ii. Anal pain or spasms that can last hours after bowel movements.[38]
   iii. Pain with sitting.
5. Abscess
   i. Pelvic rectal pain.

[FN25] Chronic Gastro-Intestinal pain includes syndromes and diseases that have obvious pathologies, but similar symptoms.

TABLE IV. Gastro-Intestinal Domain

Symptoms	Signs	Evaluation	Syndrome/disease
Pain with defecation	Tenderness on rectal examination	Questionnaire	Ano-rectal pain
Evacuation dysfunction		Culture	
Pain/pressure with sitting		Colonoscopy/biopsy	
Abdominal pain	Abdominal tenderness		Colo-rectal pain
Nausea	Bloating	Ultrasound	
Constipation/diarrhea		CT/barium enema/MRI	
Persistent or episodic			

ii. Tenesmus[39] (persistent painful need to defecate despite an empty colon).
iii. Pain with sitting.

6. Hemorrhoids[40–42]

i. Anal discomfort with engorgement.
ii. Pain and itching.
iii. Lump in perianal area.
iv. Pain with defecation.
v. Internal hemorrhoids—Painless bleeding, mucus discharge, incomplete evacuation.
vi. External hemorrhoids—Anal discomfort with engorgement, pain, and itching.
vii. Thrombosed External Hemorrhoids- Exquisitely painful lump in the perianal area. The pain tends to be acute at onset. Typically following straining at the time of bowel movement or physical exertion.

7. Anorectal Crohn's Disease—May be asymptomatic, with possible anal pain during flare.[43]

B. Colorectum (ROME III Criteria)

Rome III Criteria are a standard for functional gastrointestinal disorders. The Rome III Criteria are a system developed to classify the **functional gastrointestinal disorders (FGIDs)** of the digestive system, in which symptoms cannot be explained by the presence of structural or tissue abnormality, based on clinical symptoms. Some examples of FGIDs include irritable bowel syndrome, functional dyspepsia, functional constipation, and functional heartburn.[13]

1. Irritable Bowel Syndrome (IBS) Functional (non-inflammatory)

i. Recurrent episodes of abdominal pain.
ii. Changes in frequency, form or consistency of the stool.
iii. Sensation of incomplete evacuation, straining, fecal urgency.[44]
iv. Sensation of nausea, fatigue, fullness, vomiting.
v. Recurrent abdominal pain or discomfort at least 3 days/month in the last 3 months associated with two or more of the following:
   1. Improvement of pain with defecation.
   2. Onset associated with change in frequency of stool.
   3. Onset associated with a change in the form (appearance) of stool.[45]

Note: Consider the Symptoms of the Following Disease

Inflammatory Bowel Disease (IBD)—Complaint of recurrent abdominal pain and discomfort of at least 3 days per month in the last 3 months. The majority of IBD patients experience periods of flares and remission.

i. Abdominal and anal pain, diarrhea which may be associated with blood, suggestive of ulcerative colitis.
ii. Abdominal pain, fatigue, prolonged diarrhea with crampy abdominal pain, weight loss, and fever, with or without gross bleeding. Irregular bowel habits, with possible blood in the stool, are suggestive of Crohn's disease.[45]

V. Musculoskeletal Domain (Table V)

Musculoskeletal pain may originate from muscles, fascia, ligaments, joints, or bones.

TABLE V. Musculoskeletal Domain

Symptoms	Signs	Evaluation	Syndrome/disease
Abdomino-pelvic-perineal pain	Altered muscle tone	Questionnaires	Pelvic muscle pain syndrome
	Tension; muscle spasms and muscle compliance	Pain mapping	Coccyx pain syndrome
Pain at rest, with movement, with sitting, with sexual activity	Stiffness muscle tightness	Ultrasound	Pelvic joint, ligament or bony pain
Pain with voiding or bowel evacuation	Trigger point tenderness		
Unilateral or bilateral pain	Tender taut band		
Persistent or episodic	Twitch response, referred pain		

Common complaints: abdominal/pelvic pain, pain with sitting or with movement or with change of posture, with sexual activity, unilateral or bilateral pain. Possible pain with voiding or bowel evacuation.

A. Pelvic Muscle Pain[10,46] (See also Domain IV)

1. Pelvic Floor Muscle Pain (Pelvic Floor Myalgia)[FN26]

   i. Pain in the muscles of the pelvic floor (perineal or levator ani).

2. Intra-pelvic Muscle Pain

   i. Pain in the pelvic side wall muscles (obturator internus, piriformis, coccygeus).

3. Anterior Pelvic/Lower Abdominal Muscle Pain

   i. Pain in the rectus abdominus, oblique or transverse abdominus muscles, described below the umbilicus.

4. Posterior Pelvic/Buttock Muscle Pain

   i. Pain in the gluteal muscles.

B. Coccyx Pain Syndrome

   i. Complaint of chronic or recurrent pain in the coccyx or sacro-coccygeal joint.

C. Pelvic Joint, Ligament, or Bony Pain

1. Joint pain

   i. Sacroiliac or pubic symphysis joint.

2. Ligament pain

   i. Sacro-spinous or Sacro-tuberous ligament.

3. Bony pain

   i. Pain described in or along the margins of the pubic ramus, ilium, ischial spine or ischial tuberosity.

VI Neurological Aspects Domain[47] (Table VI)

Common complaints: Burning, throbbing, stabbing, electric shock-like sensation, tingling, stinging and/or paresthesia pain in the pelvis and/or perineal region.

A. Complex Regional Pain Syndrome[48] (CRPS)

Sympathetic, centrally generated pain.

1. CRPS 1- Triggered by tissue injury with no underlying nerve injury.
2. CRPS 2- Associated with nerve injury.

   i. Burning pain.
   ii. Increased skin sensitivity.
   iii. Changes in skin temperature, color, and/or texture.

Note: Consider Differential Diagnosis:

B. Somatic Neuropathic Pain—Nerve injury (stretching, blunt trauma, compression, entrapment, suture ligature).

1. Sacral nerve (disease)[49]

   iv. Pudendal neuralgia is a disabling form of pelvic pain. It is related to a ligamentous nerve compression mechanism. This pain is associated with the second stage of labor, sacrospinous vault suspension, vaginal laceration repairs, prostatectomy, straddle injuries, prolonged motorcycle riding, and laser treatment to the vulva, scrotum and/or perineum.

      1. Unilateral or bilateral.
      2. Lancinating burning pain in the clitoris, penis, urethra, labia, scrotum, perineum and/or anus.
      3. Worse with sitting.
      4. Relieved by standing or supine position.

3. Thoracolumbar nerve (disease)[50] [FN27]

[FN26] International Urogynecological Association (IUGA)/International Continence Society (ICS) joint report on the terminology for the conservative management of female pelvic floor dysfunction (under review).[51]

[FN27] Irritation of the thoracolumbar facet joints causes pain referred to the distribution of nerves T12, L1, and L2. This results in pain to the iliac crest and buttock. Frequently seen after abdominal and/or pelvic surgery.[50]

TABLE VI. Neurological Aspects Domain

Symptoms	Signs	Evaluation	Syndrome/disease
Characteristic sensation descriptions: burning, throbbing, stabbing, tingling, stinging, shooting, electric shock-like sensation paresthesia, atrophy, persistent or episodic	Tenderness (nerve distribution)	Questionnaires	Somatic neuropathic pain
	Referred pain	Quantitative sensory testing	Complex regional pain syndrome
	Possible skin change (color, temp, texture)	Pain mapping Nerve block imaging Ultrasound MRI	

i. Ilioinguinal nerve.

1. Pulling or throbbing that limits physical activity (groin, labia scrotum inner thigh).

ii. Iliohypogastric nerve.

1. Pulling or throbbing sensation that limits physical activity (suprapubic area and groin).

iii. Genitofemoral nerve[52]

1. Burning, paresthesia and pain (groin, labia or scrotum, medial thigh).

iv. Obturator nerve

1. Medial thigh or groin pain.
2. Weakness with adduction of the thigh.

C. Pain Following Mesh Surgery[34,53]

i. Pain during physical activity.
ii. Dyspareunia.
iii. Vaginal discharge.
iv. Exposure of mesh in vagina or elsewhere.

VII Psychological Aspects Domain (Table VII)

Common complaints: worry, anxiety, low mood, frustration, sleep disturbance, helplessness, hopelessness, difficulty in concentrating, pain impairing enjoyment. These all have biopsychosocial aspects.[54]

The **biopsychosocial model** in pain medicine was introduced with the publication of the Gate Control Theory of Pain. It is suggested that in the perception of pain three different inputs are involved: the sensory-nociceptive, the affective-motivational and the cognitive-evaluation input. These could differ within individuals, but all of these are involved in the human experience of pain.[55]

Psychological distress as a biopsychological aspect is most often a consequence of persistent pain, although existing distress is likely to exacerbate the experience of pain and difficulties dealing with it. Findings support growing evidence that the negative affective, cognitive and psychosocial state of chronic pain is universal, regardless of a neuropathic, or nociceptive nature. Emotions, thoughts and behavior involve many different locations in the brain and multiple psychological processes are involved in neuromodulation of pain.[18,54]

A. Worry, anxiety, fear: Pain is interpreted as a message of something seriously wrong with the body at the point where the pain is felt, consistent with models of severe acute pain. Without an explanation for chronic pain, anxiety is likely to persist and results in attempts to avoid activities which exacerbate the pain or are expected to do so.

B. Depression and depressed mood: This is predominantly pain-related and concerns loss of valued activities and roles as a result of pain. Difficulty sleeping, difficulty concentrating, helplessness, and hopelessness about finding a solution to the pain or a way of living a worthwhile life despite pain are common.

C. Catastrophizing: a tendency to overattend (magnification) to pain stimuli, with overestimation of the threat value and underestimation (hopelessness and helplessness) of the capacity to deal with the threat.

TABLE VII. Psychological Aspects Domain

Symptoms	Signs	Evaluation	Syndrome/disease
Worry, anxiety, fear	Helplessness	Formal psychological assessment	Worry/anxiety/fear
Catastrophizing	Hopelessness	Asking the patient what is wrong and what worries her/him about pain	Depression
Persistent or episodic	Avoidance of certain activities	Questionnaires	

VIII Sexual Aspects Domain (Table VIII)

Common Complaints: Low sex drive, inability to become aroused, pain with intercourse, difficulty reaching orgasm.

Sexual dysfunction is a disturbance in the sexual response cycle or pain associated with sexual intercourse, and can take a heavy psychological toll; it is associated with depression, anxiety, and debilitating feelings of inadequacy.[56] It is appropriate to investigate for possible history of sexual/physical abuse.

Dyspareunia is a biopsychosocial phenomenon that can have physical and psychosocial implications for the individual as well for the relationship.[57] Decrease in self-esteem, depression, anxiety, fatigue, and the need to use pain medication and other medications increase the likelihood of one or more of the disorders.

Superficial or entry dyspareunia is often associated with provoked vaginal-vulvar pain syndrome. Deep or thrusting dyspareunia often occurs in association with lower urinary tract pain, musculoskeletal pain, gastrointestinal pain, as well as abdominal/pelvic pain.[58,59]

Female and male sexual function is adversely affected in most patients with chronic pelvic pain, resulting in more than one comorbid disorder. More than 50% of partners are significantly affected and develop sexual dysfunction.

A. Sexual Desire Disorder

The following definitions form part of the DSM IV TR[60,61] [FN28]

1. Hypoactive Sexual Desire Disorder (HSDD)

   i. Low sex drive.
   ii. An absence of sexual fantasizing or erotic thoughts.
   iii. No longer feeling aroused or excited during sex.
   iv. A substantial decrease in sexual activity with partner, persisting for more than 6 months.

2. Sexual aversion disorder

   i. Persistent or recurrent aversion to, or avoidance of, sexual activity.
   ii. When presented with a sexual opportunity, the individual may experience panic attacks or extreme anxiety.

B. Sexual Arousal Disorder

i. Persistent or recurrent inability to become sexually aroused.
ii. Often characterized by inadequate vaginal lubrication for penetration (female).
iii. Inability to achieve or maintain an adequate erection for penetration (male).
iv. Symptoms present for more than 6 months.

C. Orgasmic Disorder

i. Difficulty or delay in reaching orgasm, after sufficient sexual stimulation (female).
ii. Premature or delayed ejaculation (male).
iii. Present for more than 6 months.

D. Sexual Pain Disorder

1. Dyspareunia

   i. Female sexual pain: Burning, ripping, tearing, or aching sensation associated with penetration. The pain can be at the vaginal opening, deep in the pelvis, or anywhere between. It may also be felt throughout the entire pelvic area and the sexual organs and may occur only with deep thrusting.
   ii. Male sexual pain: Sexual activity may induce a central sensitization process characterized by hypersensitivity or hyperalgesia.

History should include duration of symptoms, identification of disorder, impact on quality of life, and partner relationship. Partner interviews may be very helpful as erectile dysfunction, delayed or premature ejaculation in males with hypoactive sexual desire disorder result in a 4–30 times increased risk of female partner desire, arousal or orgasmic disorder.

TABLE VIII. Sexual Aspects Domain

Symptoms	Signs	Evaluation	Syndrome/disease
Lack of desire, arousal, orgasm Dyspareunia Persistent or episodic	Depression Relationship issues	Questionnaires Laboratory (hormonal and complete metabolic panel) Doppler ultrasound	Sexual dysfunction

IX Comorbidities (Table IX)

Patients with chronic pelvic pain syndromes, and in particular those with interstitial cystitis/bladder pain syndrome (IC/BPS), have a higher prevalence of one or multiple comorbid syndromes and diseases than the general population. These include: allergies, non-cancer chronic pain, fatigue syndromes and systemic autoimmune diseases. The risk of a comorbidity in patients

[FN28] Minimal data are available utilizing DSM 5 criteria, DSM IV TR was thus utilized. The Diagnostic and Statistical Manual of Mental Disorders, published by the American Psychiatric Association, offers a common language and standard criteria for the classification of mental disorders.[60]

TABLE IX. Comorbidities

Symptoms	Signs	Evaluation	Syndrome/disease
Allergies	Fatigue	General medical evaluation	Allergies
Fatigue	Skin lesions	Laboratory	Chronic pain and fatigue syndrome
Widespread muscular and joint pain	Dry eye	Imaging	Systemic autoimmune diseases
Irritation of the eyes	Muscular skeletal tenderness		
Dryness			
Sleep disorder			

affected by IC/BPS is usually between two and ten times higher than in a healthy population. However, data from studies on comorbidities in chronic pelvic pain patients are difficult to interpret as the composition of study populations and methodology are highly variable. Information on the prevalence of comorbidities is therefore often obtained from studies on IC/BPS.[62–66]

A. Allergies

Allergies are a heterogeneous group of diseases with involvement of the airways, skin, and sometimes of other organs.

Symptoms are caused by an immunologic reaction to some kind of trigger (e.g., inhaled allergens such as dust mite allergen, pet dander, pollen, mold, food, drugs). Nonallergic reactions to drugs or food may cause symptoms similar to allergic reactions.[67]

Examples include allergic asthma, allergic rhinitis (hay fever), atopic dermatitis (eczema), allergic drug reactions and allergic food reactions (tingling mouth, swelling of the lips, tongue, face or throat), hives, anaphylaxis, and atopic dermatitis.[68]

B. Chronic Pain and Fatigue Syndromes

Chronic pain and fatigue syndromes are characterized by pain, often widespread; fatigue; sleep disturbances; and disability. The symptoms are usually medically unexplained, have no known pathophysiology or organic basis and show no abnormal laboratory or imaging investigations. The literature suggests that many of these conditions share demographic characteristics, clinical course and psychosocial profiles.[69] Examples are:

1. Fibromyalgia: symptoms are widespread musculoskeletal pain, fatigue, non-restorative sleep, psychological distress, and regions of localized tenderness.
2. Temporomandibular Joint Disorders: symptoms consist of complaints of facial, jaw, neck, or shoulder pain. The pain is experienced in or around the ear with chewing, speaking, or opening the mouth, with or without migraine.
3. Chronic Fatigue Syndrome: is defined as clinically evaluated, unexplained, persistent or relapsing fatigue plus four or more specifically defined associated symptoms (self-reported impairment in short term memory or concentration; sore throat; tender cervical or axillary nodes; muscle pain; pain in multiple joints without redness or swelling; headaches of a new pattern or severity; unrefreshing sleep).[70]

C. Systemic Autoimmune Syndromes/Diseases

Systemic or generalized autoimmune diseases are a heterogeneous group of diseases with multi-organ involvement and evidence indicating a role played by the immune system in the pathogenesis. Examples are systemic lupus erythematosus (SLE), Sjögren's syndrome, and rheumatoid arthritis (RA). Many patients can be diagnosed with more than one of these diseases, or also with fibromyalgia and irritable bowel syndrome.

1. Systemic Lupus Erythematosus (SLE). Most frequent symptoms are debilitating fatigue, arthritis, red skin lesions after sun exposure such as a red butterfly lesion of the face, pericarditis and pleuritis, glomerulonephritis. The prevalence is 10× higher in females than in males and 2× more frequent in non-white people.
2. Sjögren's Syndrome is a systemic autoimmune disease characterized by a functional disorder of the tear and salivary glands, with or without signs of inflammation. The most common symptoms are irritation of the eyes, a dry mouth, muscle and joint pain, (debilitating) fatigue and Raynaud phenomenon.
3. Rheumatoid Arthritis (RA) is a disease characterized by chronic symmetric polyarthritis resulting in painful swelling of the joints. Other symptoms are morning stiffness, rheumatoid nodules and typical changes on hand and wrist radiographs.

D. Extraintestinal Manifestations of Inflammatory bowel disease (IBD) include non-destructive arthritis of large joints or axial arthritis such as sacroiliitis, inflammation of the eyes (uveitis, scleritis), or inflammation of the skin (erythema nodosum, pyoderma gangrenosum).[71]

Section 2: Signs

Generalized Physical Examination

A comprehensive physical examination should be performed, including palpation of the lower abdomen for bladder fullness and tenderness, and a complete pelvic exam to identify pain generators and referred pain patterns:

1. Observe posture, gait and protective behavior (avoiding sitting on flat surface or standing to avoid sitting, neck folding posture).
2. Standing: kyphosis, scars, hernia.
3. Supine: abduction/adduction of the hips, hyperaesthetic areas, scars, hernia.
4. Comprehensive pelvic examination for female and male.
5. Pain mapping (identification of pain generators/trigger points and referred pain).[72]

I Lower Urinary Tract

A. Bladder/Urethra

1. Suprapubic tenderness.
2. Tenderness of the bladder.
3. Tenderness of the urethra.
4. Tenderness of the pelvic floor muscles and identification of trigger points.[46] (See Domain V).

II Female Genital[5]

A. Vulva, Vestibule, and Clitoris

Generalized vulvar pain syndrome refers to a vulvar pain syndrome where the pain/burning cannot be consistently and precisely localized by point-pressure "mapping" via probing with a cotton-tipped applicator or similar instrument. Tenderness is diffuse and may affect all locations of the vulva.[FN29]

1. Localized and Generalized Vulvar Pain Syndrome

   i. Tenderness, Q-Tip touch sensitivity test.[72]
   ii. Erythema (localized or generalized).
   iii. Fissures.
   iv. Ulcers.

B. Intra-Abdominal Female Genital

1. Uterus and Fallopian Tube

   i. Uterine tenderness.
   ii. Cervical discharge, cervical excoriation, tenderness, adnexal tenderness, erythema.
   iii. Extrauterine tenderness, decreased uterine mobility, adnexal mass.
   iv. Enlarged uterus, nonspecific tenderness.
   v. Abdominal or pelvic scars, neuroma.

2. Ovary; adnexal mass, tenderness, abdomino-pelvic scar.
3. Pelvic Congestion Syndrome: Labia varicosities, non-specific abdominal tenderness.
4. Cervix: Erosion, Nabothian follicles, discharge, bleeding.

C. Pelvic Floor Muscle (See Domain V)

1. Perineal scarring, neuroma, dermal cutaneous allodynia.
2. Tenderness (local and/or referred to another pelvic location).
3. Vaginal discharge, mesh extrusion.[53]
4. Bulging.
5. Mass, radiation changes.

III Male Genital

A comprehensive physical examination should be performed in standing (example: exclusion of varicocele) and supine positions, including observation and palpation with pain mapping (identification of pain generators) of the external male genitals, and rectal examination.[FN30]

A. Prostate

1. Prostate tenderness on rectal examination.
2. Possible urethral discharge.

B. Scrotum

1. Tenderness on physical examination.
2. Change in color.
3. Masses on palpation.
4. Scars post-vasectomy.
5. Allodynia (increased perception of pain.

C. Epididymis

1. Tenderness.

[FN29] The vulvar vestibule (part of the vulva which lies between the labia minora into which the urethral meatus and vaginal introitus open) may be involved, (but the discomfort is not limited to the vestibule and may include referred pain from the other CPPS domains).

[FN30] Tenderness might be graded as mild, moderate, or severe.

2. Masses, nodules.

D. Testicle

1. Tenderness.
2. Masses, nodules.

E. Penis

1. Tenderness.
2. Curvature.
3. Nodules/plaque.

F. Urethra

1. Tenderness.
2. Discharge.

IV Gastro-Intestinal

A. Anorectum

1. Chronic Proctalgia—Identification of tenderness on rectal exam[3]
2. Levator Ani Syndrome—Identification of tenderness during posterior traction on the puborectalis.
3. Proctalgia Fugax—Usually there is no evident sign on physical examination.[40]
4. Anal Fissure—Identification of separation of the anoderm, sentinel tag at the external apex, exposed internal sphincter muscle, hypertrophic anal papilla at the internal apex.[38]
5. Abscess—Identification of fluctuant collections in the perianal tissues, drainage (fistula).[39]
6. Hemorrhoids—Identification of skin tags, thrombosis, prolapse on straining (reducible and irreducible).
    i. Internal: Located proximal to the dentate line and covered by columnar epithelium.
    ii. External: Located distal to the dentate line and covered by modified Squamous epithelium (anoderm)
    iii. Thrombosed: Painful lump in the perianal area.[41,42]
7. Anorectal Crohn's Disease—Identification of skin tags, hemorrhoids, fissures, anal ulcers, strictures, abscess, fistula, severe proctitis.[43]

B. Colorectum (IBS, IBD)

1. Abdominal tenderness.
2. Watery or bloody diarrhea.
3. Rectal bleeding.
4. Weight loss.
5. Fever.

V. Musculoskeletal

The musculoskeletal structures are examined for signs of tenderness and altered tension or abnormal movement.[73–76] [FN31]

1. Muscle tone: State of the muscle, usually defined by its resting tension, clinically determined by resistance to passive movement. Muscle tone has two components: (i) the contractile component, created by a low-frequency activation of a small number of motor units; (ii) the viscoelastic component, which is independent of neural activity and reflects the passive physical properties of the elastic tension of the muscle fiber elements and the osmotic pressure of cells.[46] In normally innervated skeletal muscle, tone is comprised of both "active" (contractile) and "passive" (viscoelastic) components.[46,77,78] [FN32]
    a. Hypertonicity is a general increase in muscle tone that can be associated with either elevated contractile activity and/or passive stiffness in the muscle.[5,77–79] [FN33]
    b. Hypotonicity is a general decrease in muscle tone that can be associated with either reduced contractile activity and/or passive stiffness in the muscle.[FN34] As the cause is often unknown, the terms neurogenic hypotonicity and non-neurogenic hypotonicity are recommended.

[FN31] Varying reliability has been found from pelvic floor muscle (PFM) studies assessing pain and tension using digital palpation scales.[73–76]Patients who present with alteration in the musculoskeletal structure need to be referred to a Physical Therapist well trained in the treatment of CPPS.

[FN32] Muscle tone is evaluated clinically as the resistance provided by a muscle when a pressure/deformation or a stretch is applied to it[46,77,78]Muscle tone may be altered in the presence or absence of pain. There is no single accepted or standardized way of measuring muscle tone, and there are no normative values.

[FN33] As "hypertonicity" can also be used to describe increased muscle tone of neurogenic origin, the term "increased tone" is preferred when the cause is non-neurogenic.

[FN34] As "hypotonicity" can also be used to describe decreased muscle tone of neurogenic origin, the term "decreased tone" is preferred when the cause is non-neurogenic.

2. Stiffness: Stiffness is the resistance to deformation.[80,81 FN35]
3. Compliance: Passive compliance is defined as the reciprocal of muscle stiffness.[80,81 FN36]
4. Tension: may have a similar meaning to tone and stiffness.[FN37]
5. Spasm: persistent contraction of striated muscle that cannot be released voluntarily.[82 FN38]

   a. Contracture: is an involuntary tightening of a muscle. Clinically, a muscle cramp and contracture may appear similar, however contractures are electrically silent.[83]

6. Cramp: a muscle cramp is a painful involuntary muscle contraction that occurs suddenly and can be temporarily debilitating.[83,84 FN39]
7. Fasciculation: A fasciculation is a single, spontaneous, involuntary discharge of an individual motor unit.[83 FN40]
8. Tender point: tenderness to palpation at soft tissue body sites.[46]
9. Trigger point (TrP): a tender, taut band of muscle that can be painful spontaneously or when stimulated.[85] The taut band is electrically silent. Local or referred pain may be reproduced.[86 FN41]

VI Neurological Aspects[4]

1. Tenderness on palpation corresponding to the nerve distribution.
2. Pain mapping (reproduce pain on palpation).
3. Identify referred pain by palpation.
4. Possible skin changes (color, blistering, temperature).

VII Psychological Aspects

Observation by the provider may reveal:

1. Anxiety and/or depressed mood, and avoidance or reduction of activities which exacerbate pain, or are believed by the patient to carry a risk of increasing the pain or causing harm.
2. Expression of helplessness and hopelessness (feeling of despair and representing 'the internal belief that one cannot manage one's pain').[18,54]

VIII Sexual Aspects (59)

A patient with sexual pain often has one or more other sexual dysfunctions including desire disorder, arousal disorder or orgasm disorder.

In most cases the physical examination will not identify the specific etiology of sexual dysfunction. However, a focused and comprehensive pelvic examination in females and males is mandated. In addition, assessment of the secondary sexual characteristics should be performed.[60 FN42] For the specific assessment, see the relevant Domains.

Section 3: Further Evaluation

Pain Evaluation and Measurement (7)

Pain rating(s) are essential in patient evaluation, including; Baseline and ongoing regular evaluation of severity, quality of life, questions about thoughts, emotions and behavior associated with the pain (questionnaires).

Pain Measurement

1. One of the most commonly used tools is the visual analogue scale (VAS)(85), which is a 10 cm line from ''0'' no pain to ''10'' extreme pain.[5 FN43]

0	1	2	3	4	5	6	7	8	9	10
No pain Not unpleasant										Extreme pain Extremely unpleasant

[FN35] Passive elastic stiffness is defined as the ratio of the change in the passive resistance or passive force (ΔF) to the change in the length displacement (ΔL) or ΔF/ΔL.[81] The term should only be used if stiffness is measured quantitatively such as with instruments like dynamometry or myotonometry.

[FN36] It represents the compressibility of a muscle, clinically assessed by pressing a finger into it (palpation) to determine how easily it is indented and how ''springy'' it is.

[FN37] Muscle tension can be increased or decreased due to exogenous factors such as the amount of pressure applied and endogenous factors such as thickness/ cross sectional area of the muscle itself, fluid present within the muscle (swelling, inflammation), position (e.g., standing vs. sitting) or increased neural activity.

[FN38] Occurs at irregular intervals with variable frequency and extent. Spasm over days or weeks may lead to a contracture.

[FN39] Pain is intense and localized. It tends to occur when the muscle is in the shortened position and contracting, is generated by the motor unit, and displays a high firing rate (20–150 Hz).[83] Muscle cramp either during or immediately after exercise is commonly referred to as ''exercise- associated muscle cramping.''[84] However, cramps are not specific to exercise.

[FN40] The source generator is the motor unit or its axon, prior to its terminal branches. Fasciculations display an irregular firing pattern of low frequency (0.1–10 Hz).[83] Clinically, fasciculations are recognized as individual brief twitches. They may occur at rest or after muscle contraction and may last several minutes.

[FN41] An active TrP is said to have a characteristic ''twitch'' response when stimulated; however, the twitch response to palpation has been shown to be unreliable. The most reliable sign of a TrP is sensitivity to applied pressure.[86]

[FN42] Blood pressure, heart rate, peripheral pulses, edema, lower extremity strength, and vibratory sensation is almost always helpful.[60]

[FN43] A simple verbal rating scale can also be used, for example, ''none,'' ''mild,'' ''moderate,'' ''severe.''

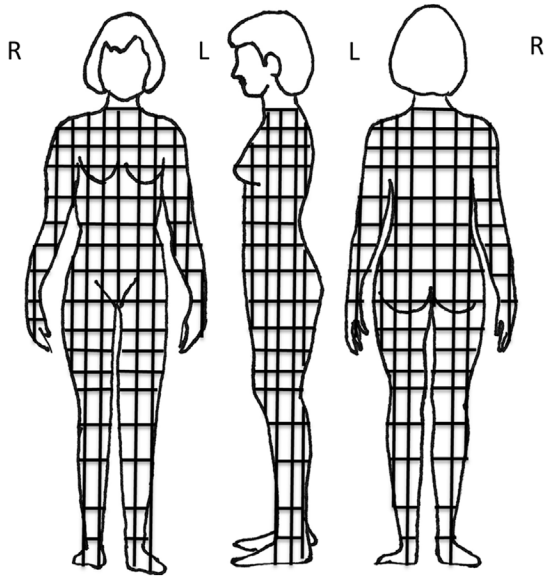
2. Pain Mapping Utilizing a Pain Body Chart[87] [FN44]

PAIN MAPPING

Instructions

Put highest intensity in each square that is applicable

May use color codes for different pains



Pain evaluation involves additional pain mapping by identifying pain generators through diagnostic procedures.[FN45] These include EMG, Q-tip touch sensitivity testing,[88,89] trigger point injections, nerve blocks and imaging.[88] [FN46]

I Lower Urinary Tract

A. Questionnaires[FN47]

a. Voiding diary with volume intake and output for 3 days at initial evaluation. Patient sensation at voiding might be recorded. At follow-up only the number of voids during day and night time is necessary. Morning volume might be recorded as a help to monitor highest functional capacity.[90]
b. Basic symptom severity Questionnaires (condition specific):
    i. The O'Leary–Sant Symptom Index.[91]
    ii. International Prostate Symptom Score.[92]
c. Visual Analogue Scale (VAS)[87] or a Likert scale for pain during the last 24 hr and over the last month (to fit with the voiding diary).[FN48]

B. Laboratory Testing

a. Urine Dipstick (red blood cells, pH, leucocytes, nitrite).
b. Urine Culture.[FN49]
c. Urine Cytology in high risk patients.
d. Investigations for Ureaplasma and Chlamydia are optional.

[FN44] Patients color the pain sites on the body chart.

[FN45] As pain is multidimensional, it can be helpful to assess separately pain intensity, pain distress, and interference of pain with activities of daily life.

[FN46] Kaufman Q-tip touch sensitivity test. This involves touching all four quadrants of the vulvar and vestibular Skene's gland ostia to evaluate for vestibulodynia, using a visual analog scale to document the level of pain and sensitivity the patient is experiencing.[89]

[FN47] Many of the questionnaires have not been studied and validated in patients with CPPS. The main assessment is still a thorough history and a full and accurate physical examination followed by pain mapping and other studies as indicated.

[FN48] Separate scores for the average, mildest and worst pain might be obtained.

[FN49] If sterile pyuria, culture for tuberculosis, in high risk patients.

e. C.T. Urogram for Hematuria.

C. Intravesical Anesthetic Challenge

An Anesthetic Challenge may be useful in pain mapping to identify the bladder and/or the urethra as a pain generator.[93] [FN50]

D. Urodynamic Evaluation (1)

a. Flowmetry and Post-void Residual
b. Filling Cystometry[94,95] [FN51]
c. Pressure-Flow Study

It is recommended to perform filling cystometry and pressure flow study if the flowmetry suggests voiding dysfunction. The demonstration of pain may identify the bladder and/or urethra as a pain generator.

In males, bladder outlet obstruction might be a differential diagnosis[96] and it is recommended to perform flowmetry in all males and consider pressure-flow studies. In males with a peak flow below 20 ml/second. In females, flowmetry and post void residual urine volume should be considered, and pressure-flow study is optional.

E. Cystoscopy

Needs to be done for patients with hematuria[97] and to identify Hunner lesions.

a. ESSIC standardized the procedure for cystoscopy and hydrodistension.[11] [FN52]

Cystoscopic findings by hydrodistension are important in subclassification of IC/BPS, see for example the ESSIC classification.[11,31,98,99] [FN53]

i. Glomerulation
   During cystoscopy with hydrodistension, glomerulations, with or without waterfall lesions (blood trickling downwards), may often be observed.[100] [FN54]
ii. Hunner Lesion Figure 1

A Hunner lesion is not an ulcer, but an inflammatory infiltrate.[11,101] [FN55]

i. Morphologic findings in Hunner Lesion

1. Inflammatory infiltrate on examination of biopsy taken with electro-resection or by cold cup biopsy.
2. Lymphocyte-like cells dominate in the infiltrate, but neutrophilic and eosinophilic granulocytes as well as plasma cells and mast cells are also found.
3. Perineural and perivascular arrangement of lymphocyte-like cell infiltrates
4. Granulation tissue.[101–104] [FN56]

F. Differential Diagnosis (Confusable, treatable diseases):

Criteria for diagnosis are needed as the target disease may be confused with other treatable diseases (confusable diseases) because of similar features.[11]

a. Ketamine Cystitis

[FN50] A solution of lidocaine and sodium bicarbonate administered intravesically results in reduction of pain. Alkalinized lidocaine instillation has not been validated, but may be useful.[93]

[FN51] The NIDDK criteria excluded patients with detrusor overactivity at filling cystometry in order not to confuse the picture in clinical trials.[94] However, this does not mean that detrusor overactivity cannot coexist with interstitial cystitis/bladder pain syndrome. In the interstitial cystitis database, approximately 14% of IC/BPS patients had detrusor overactivity.[95]

[FN52] A rigid cystoscope is preferred to facilitate taking adequate biopsies. Glycine or corresponding filling fluid should be used to allow for coagulation after biopsies. Infusion height should be approximately 80 cm above the Symphysis Pubis. A dripping chamber is used and the bladder is filled until fluid dribbling stops. If necessary, a digital block is applied around the urethra to prevent leakage. Pre-distension inspection includes observation for radiating vessels, coagulum or fibrin deposits, white spots, hyperaemia, edema, cracks, scars or any other mucosal changes. Continuous inspection while filling the bladder is advised. When maximum capacity is reached, the distension is maintained for 1–3 min. The bladder is emptied and the color of the fluid checked for the degree of bleeding. The total volume drained is the measured maximum bladder capacity. During a second filling, the bladder is filled to approximately 1/3rd to 2/3rds of the bladder capacity to achieve optimal vision for inspection and biopsies. The bladder should not be filled to maximum capacity or distended again to avoid further provocation of changes with doubtful reproducibility.[33]

[FN53] The finding of a Hunner lesion is important because effective treatment is available.[98,99] The presence of Hunner lesions may be the diagnostic finding of the proposed disease "Interstitial Cystitis."[31]

[FN54] The significance of the presence of glomerulation remains to be determined.[100]

[FN55] A Hunner lesion is a distinct cystoscopic finding and typically presents as a circumscript, reddened mucosal area with small vessels radiating towards a central scar, with a fibrin deposit or coagulum attached to this area. This site ruptures with increasing bladder distension, with petechial oozing of blood from the lesion and the mucosal margins in a waterfall manner. A rather typical edema may develop post-distension with varying peripheral extension. Usually, lesions are multiple but occasionally they may be single. It is not unusual for more lesions to be detected at re-inspection than seen at the initial phase of distension.[11]

[FN56] To a large extent, the detection rate and the findings on distribution of mast cells have been dependent on laboratory routines and staining as well as fixation techniques. Tryptase staining methods provide a stable result that is not sensitive to laboratory variations.[103,104]

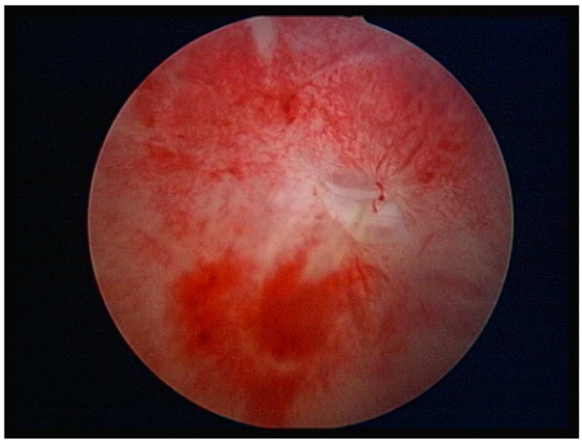


Fig. 1. Hunner lesion.

Ketamine Cystitis is a new condition not previously described. Caused by recreational ketamine abuse, ketamine cystitis includes increased voiding, frequency, dysuria, bladder pain and hematuria.[105,106] [FN57]

II Female Genital

A. Vulva, Vestibule and Clitoris

1. Questionnaires

   i. Visual Analog Scale for pain.[86]
   ii. Female Sexual Function Index (FSFI).[61]
   iii. Female Sexual Distress Scale (FSDS).[107]

2. Laboratory Testing

   i. Culture.
   ii. Biopsy.

3. Diagnostic Testing

   i. Vulvoscopy, with or without biopsy.
   ii. Quantitative Sensory Testing (Q-tip touch sensitivity test).[72,89,108]

B. Intra-abdominal Female Genital

1. Questionnaires

   i. Visual Analog Scale(85) for pain.

2. Laboratory Testing

   i. Culture.
   ii. Complete blood count.

3. Laparoscopy (with or without biopsy)
4. Ultrasound (US)
5. MRI
6. Venography (to rule out Pelvic Congestive Syndrome)[109]

C. Pelvic Floor Muscle

1. Questionnaires.

   i. Visual Analog Scale for pain.[86]
   ii. Pelvic Floor Distress Inventory (PFDI).[110]

[FN57] The molecular mechanism for ketamine-induced cystitis is unknown. The affected bladder exhibits a denudation of the urothelium with inflammatory cell infiltration. The upper urinary tract is also damaged in patients who use a higher dose and with a longer duration. Attention by both medical organizations and social workers for this increasing social phenomenon particularly among young people is now urgently needed.[105,106]

iii. Prolapse and Incontinence Sexual Questionnaire (PISQ).[111]

2. Laboratory Testing

i. Wet Mount, Culture.
ii. Biopsy.

3. Imaging References

i. Ultrasound (4D if available for visualization of mesh, where applicable).
ii. MRI (with or without defecography).
iii. Defecography.

III Male Genital

A. Prostate Pain

1. Quantitative assessments.

i. Bladder diary.[90,112]
ii. CPSI (Chronic Prostatitis Symptom Index).[113]
iii. Visual Analog Scale for Pain (VAS).[86]

2. Laboratory Testing

i. Urinalysis (including post prostate massage).
ii. Urine Culture post prostate massage.
iii. Semen Culture.

3. Uroflowmetry, Post voiding residual volume, pressure flow study
4. Cystoscopy
5. Ultrasonography, with or without biopsy.

B. Scrotum, Epididymis, Testicle, Penis

1. Quantitative assessments

i. VAS for Pain.[86]

2. Ultrasonography

C. Urethra Pain

1. Quantitative assessments

i. Bladder diary.
ii. VAS for Pain.[86]

2. Laboratory Testing

i. Urinalysis (including post prostate massage, Ureaplasma/Chlamydia as appropriate).

3. Urethroscopy/Urethrography
4. Ultrasonography

D. Sexual Pain (See Domain VIII)

1. Questionnaires

i. VAS for Pain.[86]
ii. International Index of Erectile Function (IIEF).[114]

IV Gastro-Intestinal (40)

1. Questionnaires

i. Rome III Criteria Questionnaire.[115]
ii. Colorectal Rectal Distress Inventory.[116]

2. Laboratory Testing

i. Culture.

ii. Stool Evaluation for ova and parasites.
iii. Antibody testing.
iv. Biopsy.

3. Diagnostic Testing

i. Anorectal Manometry (paradoxical contraction of the pelvic floor muscles when instructed to strain during defecation).
ii. Rigid or flexible endoscopy (Anorectal sigmoidoscopy) with or without biopsy.
iii. Anorectal/Pelvic US, 3D.
iv. Barium Enema.
v. CT Scan, Defecography, MRI defecography.

V Musculoskeletal[117]

1. Questionnaires

i. McGill Pain Questionnaire.[118]
ii. Pelvic Floor Distress Inventory (PFDI).[110]
iii. Female Sexual Function Index (FSFI).[61]
iv. Female Sexual Distress Scale (FSDS).[107]

2. Pain Location Drawing (Pain Mapping)

i. Pain Chart body map.[87]

3. Evaluation of Muscle Tension There is no single tool which is able to measure all components of muscle tone. Some tools may be able to measure aspects of tone such as contractility, stiffness or elasticity. Instrumented methods may have a role in the valid and reliable evaluation of muscle tone, for example, surface electromyography, dynamometry, real-time ultrasound, elastometry, myotonometry.

i. Pressure manometry is the measurement of resting pressure or pressure rise generated during contraction of the pelvic floor muscles using a pressure device (a manometer) inserted into the urethra, vagina or anus.[119,120] [FN58]
ii. Surface electromyography (sEMG) refers to the bioelectrical activity generated by muscle fibres.[121,122] [FN59]
iii. Dynamometry is the measurement of pelvic floor muscle resting and contractile forces using strain gauges mounted on a speculum (a dynamometer), which is inserted into the vagina.[123]
iv. Real-time ultrasound measures pelvic floor muscle morphology and function via a non-invasive (trans-abdominal or trans-perineal) probe.[124] [FN60]
v. Elastometry measures the elasticity of a tissue.[125] [FN61]

4. Trigger point injection or needling has been used as a diagnostic test to identify pain generators.[125] [FN62]
5. Imaging

i. X-Ray.
ii. Ultrasound.
iii. MRI.

VI Neurological Aspects

A. Neuropathic Pain Questionnaires

1. VAS Pain Score.[86]
2. Pain DETECT (Validated for CPPS evaluation).[129]
3. Leeds Assessment for neuropathic symptoms and signs (not validated for chronic pelvic pain).[130]
4. Douleur Neuropathique 4 Questionnaire.[131]

[FN58] The tool has been used as an outcome measure in intervention studies of pelvic floor pain.[119,120] However, the tool has not been tested for reliability in this population.
[FN59] Pelvic floor muscle surface electrodes use either flat interface perineal electrodes or intra-vaginal/intra-anal probes to record sEMG either at rest or during a PFM contraction. Surface EMG is considered to be non-specific to the PFM. Because of the large surface area covered by the electrode, cross-talk from adjacent muscles often occurs.[121,122] It is therefore not considered reliable as a measure.
[FN60] Trans-perineal measures of ano-rectal angle and levator plate angle have been tested for reliability in a male pelvic pain population.[124] Therefore, this tool shows promise as an instrumented method to evaluate pelvic floor muscle changes in pelvic pain.
[FN61] It has recently been applied to measure the passive stiffness of puborectalis in asymptomatic women and shown to be reliable in this pilot study[125] However, it requires testing to establish application in a pelvic pain cohort.
[FN62] The taut band(s) of sarcomeres within the TrP can be identified by ultrasonography.[126] and magnetic resonance elastography.[127] A tissue compliance meter which measures stiffness in the taut band has been shown to confirm the hardness of the discrete band of muscle that harbors the tender region in peripheral skeletal muscle.[128]

B. Quantitative Sensory Testing

1. 1Q-tip touch sensitivity.
2. Sensory pain mapping.[72]
3. Reflex evaluation.
4. Electromyography.

C. Nerve Blocks

1. May/may not be done under Computed Tomography, Ultrasound or EMG guidance.[132,133]

D. Imaging

1. Ultrasound
2. Magnetic resonance Imaging (MRI)

VII Psychological Aspects[18,54]

The chief purpose of psychological assessment is to get a complete picture of the pain syndrome with all affected dimensions: somatic, affective, cognitive and behavioral, and the individual consequences for the patient. Direct questioning about the patient's view of what is wrong or what worries him/her is more helpful than questionnaires.[FN63]

1. Questionnaires

i. SF-12 or SF-36.[134]
ii. Brief Pain Inventory.[135]
iii. Catastrophizing Questionnaire can be considered in certain cases.[136]

VIII Sexual Aspects

1. Questionnaires.

i. Female Sexual Function Index (FSFI).[61]
ii. Female Sexual Distress Scale (FSDS).[107]
iii. International Index of Erectile Function (IIEF).[137]

2. Laboratory Testing

i. Hormone Panel.
ii. Complete Metabolic Panel.
iii. Culture.

3. Imaging

i. Doppler US to assess blood flow.

It is also particularly important to work up the partner's potential sexual dysfunction. Early referral to a sexual counsellor is optimal.

IX Evaluation of Comorbidities

If patients have symptoms and signs of comorbidities, evaluation should be undertaken according to relevant guidance, and may be appropriate to refer to the relevant specialist.

SUMMARY

This first ICS Standard for Terminology in Chronic Pelvic Pain Syndromes aims to improve understanding of these syndromes and patient diagnosis. It is hoped that this will help develop the field, through facilitating phenotyping of patients, development of pertinent animal models and new preclinical development of therapeutic strategies.

Evaluation of patients based on the nine domains should be individualized, taking into consideration the patient's personal perception of pain, and also the biopsychosocial aspects of CPPS.

Discussions on nomenclature partly focused on the risk of inadequate patient care if diagnostic terminology is changed without taking into account the practical impact of its application on the patient's access to appropriate treatment, reimbursement, and social benefits.

This Standard for Terminology in CPPS will be reviewed in the future as continuing research, such as the Multidisciplinary Approach to the Study of Chronic Pelvic Pain (MAPP) Research Network, generates new insights.[138–140] Working with the guideline bodies, such as the AUA, East Asian IC Study Group/SICJ, EAU, ESSIC, FGIDS, and IASP, the ultimate aim should be to achieve international consensus.

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[FN63] Early referral to a psychological healthcare provider should be considered. Patients with sexual dysfunction may need sexual counseling.

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Any products/companies referred to in this document are not necessarily recommended or endorsed by the ICS.

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