Interstitial Cystitis/Painful Bladder Syndrome

Synonyms: interstitial cystitis, painful bladder syndrome, bladder pain syndrome, hypersensitive bladder syndrome, trigonitis

The common symptoms of interstitial cystitis/painful bladder syndrome (IC/PBS) include urinary urgency, frequency, nocturia and suprapubic or pelvic pain without any known aetiological factor. The term ‘interstitial cystitis’ is usually reserved for patients with typical cystoscopic features.

The European Association of Urology (EAU) guidelines refer to this condition as bladder pain syndrome (BPS). This condition is the occurrence of persistent or recurrent pain perceived in the urinary bladder region, accompanied by at least one other symptom, such as pain worsening with bladder filling and daytime and/or night-time urinary frequency. There is no proven infection or other obvious local pathology. It is often associated with negative cognitive, behavioural, sexual or emotional consequences, as well as with symptoms suggestive of lower urinary tract and sexual dysfunction.

Epidemiology

- The exact prevalence is unknown because of misdiagnosis and also underdiagnosis. Although once thought to be rare, IC/PBS is now believed to have a much higher prevalence and the incidence is increasing globally.
- This condition primarily affects women.
- However, many women are often reluctant to seek treatment[1].
- More recently, the prevalence of the condition has increased to at least 2% among females, due to broader and more inclusive diagnostic criteria[2].
- The average age at diagnosis is 40 years[3].
- However, around 35% of patients actually have symptoms that started in childhood[2].
- This condition can occur in children.

Aetiology

- This condition has a multi-factorial aetiology including epithelial dysfunction, subclinical infection, mast cell and vascular abnormalities, neurogenic inflammation, autoimmune phenomena and up-regulation of sensory nerves in the bladder[4].
- It is thought that there is injury or dysfunction of the glycosaminoglycan layer that covers the urothelium. This injury can be caused by bacterial cystitis, childbirth, pelvic surgery or urological instrumentation.
- These factors provoke sensory nerve activation, neurogenic inflammation, pain and fibrosis [5].
- Changes in the somatosensory grey matter occur which may have an important role in pain sensitivity as well as affective and sensory aspects of interstitial cystitis[6].

Presentation

Symptoms vary widely in severity and nature but the onset of symptoms is often acute and may be sudden.

- Recurrent symptoms similar to urinary tract infections (urgency, frequency, dysuria), lower abdominal pain, pressure in the bladder and/or pelvis, and dyspareunia.
- Characteristics of the pain include:
  - Pain, pressure or discomfort perceived to be related to the bladder, increasing with increasing bladder content.
  - Located suprapublically, sometimes radiating to the groins, vagina, rectum or sacrum.
  - Relieved by voiding but soon returns.
  - Aggravated by food or drink.
- In women the symptoms are often worse during menstruation.
- There is wide variation in symptoms between individuals and in any one individual over time.
- Examination may be normal apart from suprapubic tenderness.
- The severity of the symptoms often bears little correlation with the clinical findings.
- Trials of antibiotic treatment do not cure the condition.

Associated disorders

- An association has been reported between BPS and non-bladder syndromes such as fibromyalgia, chronic fatigue syndrome, irritable bowel syndrome, vulvodynia, depression, panic disorders, migraine, sicca syndrome, temporomandibular joint disorder, asthma and systemic lupus erythematosus[7].

Differential diagnosis

Other causes of urinary frequency, urgency of micturition and pelvic pain, including:
Infection or other inflammatory conditions - eg, recurrent urinary tract infection, urethral diverticulum, infected Bartholin's gland, tuberculous, bacterial or viral vaginitis, schistosomiasis.

Gynaecological - eg, pelvic malignancy, uterine fibroids, endometriosis, mittelschmerz (ovulation pain), pelvic inflammatory disease, genital atrophy.

Urological - eg, bladder cancer, radiation cystitis, overflow incontinence, chronic pelvic pain syndrome, bladder outlet obstruction, urolithiasis, urethritis, chronic prostatitis, prostate cancer.

Neurological - eg, detrusor overactivity, Parkinson's disease, lumbosacral disc disease, spinal stenosis, spinal tumour, multiple sclerosis, cerebrovascular disease.

Others include inflammatory bowel disease, gastrointestinal neoplasm, diverticulitis and adhesions from previous surgery.

Investigations
The diagnosis and management of this syndrome may be difficult in some patients. The diagnosis of interstitial cystitis is usually based on a thorough assessment and exclusion of other causes.

- Urinalysis and midstream urine for urine cultures: rule out urinary tract infection, including tuberculosis.
- Cervical swabs for herpes and chlamydia.
- Urodynamic studies: there are no specific findings but pain with bladder filling that reproduces the symptoms is very supportive of a diagnosis of IC/PBS.
- Most cases need cystoscopy to exclude bladder cancer. Hunner's ulcers (reddened mucosal areas often associated with small vessels radiating towards a central scar, sometimes covered by a small clot or fibrin deposit) may be seen in 10-50%.
- Men should have urethral swabs and prostatic secretion cultures (for chronic prostatitis).
- Investigations such as cystoscopy, hydro-distension and biopsy are often used for diagnosing IC/PBS in paediatric patients, as eliciting clinical symptoms can be difficult and unreliable.

Management
There are few clinical urogynaecological conditions that cause greater frustration than IC/PBS. This is due to its chronicity, serious impairment of quality of life, diagnostic difficulties and unsatisfactory treatment.

- Management is often difficult and only partially effective. Early diagnosis and management are important.
- A comprehensive assessment of patients is required to identify treatment options that are tailored to the needs of individual patients.
- Multimodal behavioural, physical and psychological techniques should always be considered alongside oral or invasive treatments of IC/BPS.
- Treatment is mainly symptomatic and supportive.
- Oral or intravesical therapies are the mainstay of treatment whilst surgical procedures are reserved for refractory cases.
- This condition usually warrants a multidisciplinary approach for optimum outcome.
- The following are important to consider regarding managing these patients:
  - Self-awareness of the condition.
  - Immediate urine culture test.
  - Specifying the location of pain.
  - The type of urinary incontinence.
  - Helpful dietary control.
  - Complementary and alternative medicine may be considered.
  - Finding an expert early is ideal but not always possible.

Non-drug
- Behavioural therapy: biofeedback, pelvic floor exercises and bladder training programmes may be effective.
- Diet: certain foods and drinks such as alcohol, tomatoes, spices, chocolate, caffeinated and citrus drinks and acidic foods may contribute to bladder irritation and inflammation. Many patients find benefit in keeping a food diary to try to assess which foods exacerbate and worsen their symptoms.
- Some people report a reduction in symptoms following distension of the bladder during diagnostic cystoscopy. There is belief that distending the bladder causes the nerve cells to be stretched and thus less sensitive for a time.
- Transcutaneous electrical nerve stimulation (TENS) can help in conjunction with other therapies.

Drugs

Oral medications
- Analgesics should be used preferably in collaboration with a pain clinic.
- There is no good evidence for corticosteroids, duloxetine or prostaglandins and they are not recommended for long-term treatment.
- Cimetidine can be considered for some patients as a valid oral option before invasive treatments.
- Amitriptyline may be beneficial for some patients.
- Oral pentosan-polysulfate sodium is available, although it is not currently licensed in the UK.
- Oxybutynin or gabapentin might be considered for some patients.
- Oral treatment with ciclosporin has been shown to be a beneficial therapeutic strategy for some patients with IC/PBS. However, adverse effects are significant and should be carefully considered.

Intravesical drugs
- It is recommended that intravesical lidocaine plus sodium bicarbonate can be administered prior to more invasive methods.
- Intravesical pentosan-polysulfate sodium or intravesical heparin are sometimes used.
- Intravesical hyaluronic acid or intravesical chondroitin sulfate is also sometimes used and can be considered before more invasive measures.
- However, intravesical therapy with BCG is not recommended for these patients, nor is intravesical therapy with clorpactin.
- Bladder distension is also not recommended as a treatment.

Other treatments
Botulinum toxin type A is sometimes effective in managing this condition. However, it is not consistently effective in all patients.

Surgical
All ablative organ surgery should be the last resort for experienced and knowledgeable surgeons only.

Major surgery (subtotal cystectomy and bladder augmentation or supravesical urinary diversion with intact bladder) is associated with good symptom relief in strictly selected patients with disabling IC/PBS, where conservative treatment has failed.

When all other treatment options fail to relieve disabling symptoms, surgical removal of the diseased bladder is the ultimate option, for which three major techniques are common: supratrigonal (trigone-sparing) cystectomy, subtrigonal cystectomy and radical cystectomy and radical cystectomy with excision of the urethra.

Prognosis
- The prognosis is very variable. The condition can:
  - Have complete resolution of symptoms within months.
  - Follow a waxing and waning course.
  - Be completely asymptomatic with intermittent flares.
  - Follow a chronically progressive course of increasing symptoms over several years.

Some people do recover spontaneously, but individuals may have the condition for many years and there may be spontaneous resolution only to return days or months later.

Short-term (up to one year) cure rates range from 50% to 75% for non-invasive or minimally invasive therapies but repeat administration of a therapeutic agent is required. Although definitive surgical intervention is associated with greater long-term cure rates (>80%), significant short-term and long-term adverse effects occur more frequently.

Interstitial cystitis can have a significant and even profound effect on quality of life.

Further reading & references

- Management of Bladder Pain Syndrome; Royal College of Obstetricians and Gynaecologists (2016)
- Interstitial Cystitis Diet; Confidence Choices

11. Interstitial cystitis: oral pentosan polysulfate sodium - Key points from the evidence; NICE advice, April 2015

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