Review Article

Patient-centred standardization in interstitial cystitis/bladder pain syndrome—a PLEA

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Abstract: Standardization has an impact on all links in the healthcare chain and directly affects patients with regard to diagnosis, treatment, eligibility for reimbursement, social benefits and care. Furthermore, patients are also affected if research goes down inappropriate pathways due to inaccurate terminology and definitions. Complex healthcare coding and electronic medical record systems and global reliance on the internet and search machines mean that new terminology or changes now need in some way to be designed to last or to be updated in a way that will cause least disruption, while still allowing for evolution, since changes have far more ramifications today than they ever did in the past. Official recognition of a condition is vital, so coding must be correct and uniform across all authorities. Potential problems must be anticipated at an early stage in the process. In order to achieve global consensus, stakeholders from all parts of the world need to communicate and collaborate with all viewpoints taken into consideration. Patients and their advocacy groups should be involved in standardization processes to ensure that all aspects of a condition are covered, that no patients or symptoms are excluded and that there is no adverse impact on the patient in practical terms following implementation. The trend today is for patients to be more involved in healthcare and decision-making. We must ensure that standardisation and guidelines do not get left behind in this process of development either now or in the future. However, it is necessary to find some way of training patient representatives to enable them to participate fully in standardization and guidelines and also to encourage them to do so.

Keywords: Standardization; taxonomy; guidelines; interstitial cystitis (IC); painful bladder syndrome (PBS); bladder pain syndrome (BPS); hypersensitive bladder (HSB); Hunner lesion; urgency; patient-centred healthcare

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Introduction

Healthcare in the 21st century has evolved into an intricate chain with multiple links and participants. Standardisation of terminology and definitions can have an impact on every link and every participant in this chain. Participants in the healthcare chain include researchers, clinicians, national and international health authorities, regulatory bodies, health insurance companies, social security organizations, the pharmaceutical industry, hospitals, pharmacies, but above all the patients and their families, in other words the consumer. These different actors do not automatically have the same objectives, there may even be conflicts of interest, and the patients' interests may not necessarily be everyone's prime objective.

It goes without saying that new or changed terms or definitions should help the patient, not be a hindrance, and should never actually cause harm to the patient, whether medically, financially or otherwise. When terminology and definition standards are being developed or modified, every potential impact and scenario should be taken into account.

However, without meaningful patient representation in standardization processes, the practical impact on the patient of implementation of a standard may not even be taken into consideration.

Consequently, participation and input by patient representatives in standardization working groups should be considered an integral part of patient-centred healthcare including aspects such as patient risk and patient safety.

We live in an electronic age that is search engine- and Internet-driven, with patients, clinicians, researchers and multiple national and international authorities worldwide seeking and storing information electronically. We have healthcare systems in which many different authorities have a finger in the pie and each may have its own terminology, codes and electronic coding system. Repeated changes in terminology and many different names for a single disease can cause havoc in and between systems with potentially disastrous consequences for the patient further down the chain. However, few health professionals who are involved in standardisation and/or guidelines appear to have an adequate knowledge of coding and its consequences, either nationally or internationally.

The objectives of standardization of terminology and definitions include:

(I) Correct diagnosis of the patient;
(II) Official recognition of this diagnosis (nationally and globally);
(III) Appropriate treatment for the patient;
(IV) Reimbursement of appropriate treatment;
(V) Eligibility for social benefits (unemployment/disability etc.);
(VI) Recording/registering patient information: electronic records;
(VII) Comparison of data between centres and internationally;
(VIII) International scientific research;
(IX) International communication, collaboration, data collection and exchange of data;
(X) Meaningful prevalence statistics.

In order to achieve this, there must be international consensus: "*Standardized definitions of key medical terms with international consensus are increasingly needed as analysis and registration in healthcare become ever more automated and communication increasingly global*." as noted by Rosier *et al*. in 2012 (1).

International consensus on terminology, definitions and criteria has often been notably lacking in the urological field, particularly with regard to the lower urinary tract where in the past committees took decisions without consulting the rest of the medical world, let alone the patients themselves. Even today, we still see "local" rather than "global" thinking: local in the sense of either national or regional criteria or in the sense of taxonomies, standards or guidelines developed by individual specialty societies. Indeed, almost every society is currently producing its own (different) standard terminology or guidelines, often in competition with each other, resulting in considerable confusion and controversy.

In the field of interstitial cystitis (IC)/bladder pain syndrome (BPS)—to use one of its many names—we have in recent times seen classifications, terminology, definitions, taxonomies, guidelines and consensus documents emerging from a multitude of sources including the NIDDK, the International Continence Society (ICS) standardisation document on lower urinary tract symptoms, the European Association of Urology (EAU) Guidelines on Chronic Pelvic Pain, the International Association for the Study of Pain (IASP) taxonomy on chronic pelvic pain, diagnostic criteria, classification and nomenclature from the International Society for the Study of Bladder Pain Syndrome (ESSIC), a Japanese guideline for diagnosis and treatment of IC, a Japanese and East Asian guideline on hypersensitive bladder (HSB) and IC, an AUA Guideline on IC/BPS, in addition to a report from a multidisciplinary consensus meeting in Washington DC in 2007, new research definitions from the NIDDK for its MAPP project and of course the World Health Organization's International Classification of Diseases (ICD) (2-11).

When Guy Hunner [1868-1957], gynaecologist and obstetrician at the Johns Hopkins Hospital in Baltimore, wrote in some desperation when faced with multiple different names for the painful bladder disorder: "*It is apparent to any one reviewing the literature of bladder ulcer that some organization such as the American Urological Association or the American Association of Genito-Urinary Surgeons, could perform a valuable service by appointing a small commission to review the subject from every point of view, and adopt a nomenclature that would serve as a working basis for our present knowledge*," he could not have imagined that this would ultimately be taken up by so many societies, or that it would generate even more confusing new terms including: (chronic) IC, painful bladder syndrome (PBS), BPS, HSB, along with Hunner's ulcer and Hunner lesion, and multiple combinations of these terms (12).

These numerous criteria, definitions and terms have made a mockery of research studies and expensive drug trials with their varying study populations based on different diagnostic criteria and methodology and often with lesion and non-lesion types lumped together. This naturally also means that prevalence data for IC/BPS/HSB are unreliable and cannot be compared from centre to centre, let alone country to country.

Above all, it is greatly regrettable that such a huge amount of time and energy (and consequently money) over a number of years has been poured into defending/attacking controversial new terminology and definitions which could have been resolved by all parties involved—including the patient organizations—working together in a collaborative manner.

Global consensus can only be achieved by involving researchers and clinicians from different parts of the world and different cultures, together with the input of patient representatives and other relevant stakeholders. However, in order to achieve this, there must be a willingness to work together, to accept that other parties may have a valid point of view and to incorporate these points of view in any new standards. In the past 15 years in particular, this has been lacking in the IC/BPS/HSB world.

Impact on the patient

New or changed terminology and definitions have an impact on the patient in many ways, including:

(I) A research impact, ultimately affecting the patient;
(II) A medical impact on the patient;
(III) A social and financial impact on the patient and family or carers.

If the terminology and definitions are not right, research will go down the wrong pathways, reach wrong conclusions, thereby ultimately leading to wrong treatment for the patients and potentially the exclusion of what may be very important groups of patients who should be studied and of course treated.

Right terminology and definitions on the other hand mean right diagnosis, right treatment, reimbursement of that treatment, eligibility for social security, unemployment and disability benefits and entitlement to all kinds of social services and care, many of which may have an impact on the entire family not just the patient. This ultimately leads to improved quality of life for the patient.

There is also a cost aspect involved here. Since wrong terminology and definitions can lead to a wrong or inadequate diagnosis and consequently inappropriate treatment, this can lead to tremendous wastage of drugs that have been prescribed but prove ineffective. The "trial and error" approach to IC/BPS/HSB is enormously wasteful and costly, both for the patient and for the state and/or insurance authorities.

An important aspect of terminology where the patient is concerned is official recognition of a condition. The name of the disease or complaint as clinically diagnosed must match the officially recognized name and coding and consequently the name of the disease for which a specific treatment is authorised. And this is vital in today's electronic official administrative world where repeated changes in terminology can cause havoc in and between systems, leading in some cases to refusal by insurance authorities to reimburse the cost of appropriate treatment or to recognise a diagnosis because the name of the diagnosis does not synchronize with the name in the system, as we have seen happening recently in Germany due to the change from the name IC (considered a disease) to BPS (considered a syndrome or collection of symptoms). Drugs developed and registered for a specific disease name may not necessarily be reimbursed if the name is changed. This can have a huge financial impact on the patients, causing great hardship to patients and their families and can result in the patient being compelled to use inappropriate and ineffective treatment which in turn leads to more wastage of resources and money and inevitably poorer quality of life.

A question we face is how can we ensure that potential consequences for the patient of new or changed terminology or definitions can be anticipated before publication, rather than being discovered after the event when it may be too late or extremely difficult to rectify? Some of the pitfalls in the development of standard terminology can be avoided by involving patient organizations or their representatives. This was emphasized by Rosier *et al.* in a paper on developing evidence-based standards for diagnosis and management of lower urinary tract or pelvic floor dysfunction published in 2012 by the ICS Standardisation Steering Committee, stating that the composition of working groups needs to represent the most important stakeholders including patient representatives (1).

Patient input can be invaluable. The IC/BPS/HSB patient organizations—who run helplines and may hear from not just hundreds but thousands of patients—have contact with patients from the entire spectrum of this bladder disorder and its comorbidities, while clinicians may only see restricted groups of these patients and therefore may have a more limited overall perspective. Patient organisations are experts on the challenges and issues facing patients in their daily lives. They can provide health professionals engaged in standardization or guidelines with supplementary information, helping to create a far more comprehensive picture of each condition and everything this may involve along the healthcare chain.

Patients can help to ensure that terminology and definitions correspond to the disease (IC/BPS/HSB or

Hunner lesion) or symptom (e.g., pain, urgency, frequency, etc.) as experienced by the patient and cover the full spectrum of the disorder with all its variations. In this way, one can avoid square patients being forced into round holes simply to fit in with an unproven pet theory.

Patient support groups, usually on tight budgets with limited manpower and often run voluntarily, play a huge role in the patient care chain by providing information, websites, helplines, patient-to-patient counseling, raising awareness, raising research funds; and they cannot simply be informed repeatedly that terminology has changed (again) and that they will (again) be expected to change all their information and even the name of their organization without having been consulted at any step of the way. Practical commonsense tells us that this is simply not tenable, including from a financial point of view, and therefore solutions will need to be sought in partnership.

A further aspect that should not be ignored is that nomenclature changes can also create stress and uncertainty among patients who may have waited years for a diagnosis and now suddenly find that the name of the diagnosis they received is no longer "valid" or that their medical condition now appears to be known under at least four different names. One change is difficult, but repeated changes within a period of a few years are psychologically harmful for patients.

In the past, standardization, taxonomy and guideline committees have tended to go over the heads of patients and have looked at terminology issues from research and clinical points of view, but failed to look further along the chain at the potential impact on the patient in practical terms once the changes have been implemented. In the past, while patient advocates may occasionally have been invited to work with guideline committees, this has often been limited and these patient representatives have frequently only been "consulted" at the very end when the work is virtually finished, in order to "rubber stamp" the document as being patient-approved, without the patient representative having had the opportunity for any real input.

Meaningful patient participation means involvement at all stages in the development process. However, in order to be able to undertake this successfully, patients may need prior training about not only standardization and guideline processes, but also aspects such as patient safety, insurances and reimbursement, and some insight into how coding systems operate.

Researchers are therefore urged to ensure that patient representatives are given every opportunity—and training if necessary—to play a full role in national or international discussions on guidelines, taxonomy, definitions and nomenclature, including of course the ICD, to ensure that these are a true reflection of all the key components of a disease or complaint, that they make practical sense and do not cause the patient any harm.

Confusion, controversy and guesswork

A bladder disorder of unknown cause that has had well over 20 different names in the past 200 years can be expected to give rise to a certain amount of confusion and controversy. Since even today little is known or understood about this enigmatic bladder disorder, it has led to multiple hypotheses and pet theories about its cause or causes. In recent decades, this "guesswork" has spilled over into classifications, nomenclature, definitions, taxonomies and guidelines to such an extent that these theories have begun to lead a life of their own, causing many people to assume that they are based on fact. Much of the confusion could have been avoided by simply talking to a wide spectrum of patients and involving the patient organizations in standardization projects. The situation has not been helped by bundling all the patients together for decades and, until recently, failing to take subtyping or phenotyping seriously.

Nomenclature controversies and the impact on the patient

As we know from Guy Hunner and his colleagues a century ago, nomenclature has always been a problem in this field. For several decades, the term IC was used; however, this was largely referring to patients with bladder lesions. Patients with symptoms but no lesions were still often considered hysterical and neurotic, a hangover from the anti-feminist theories of Sigmund Freud who believed that women faced by adversity would become hysterical and develop imaginary illnesses. Unexplained pain fell into this category and from the beginning of the 20th century female IC patients with no visible lesions were considered by many to be suffering from psychosomatic bladder neurosis. A few physicians such as Albert Meads in 1934 incorporated the non-lesion type in their classification as an early pre-ulcer form of the lesion disease, but the psychosomatic label continued for years and still hovers like a phantom from the past behind the scenes.

Controversy reared its head in 2002 when the ICS standardisation sub-committee published a standardisation document on lower urinary tract symptoms, now using the term PBS with IC reserved for a specific diagnosis requiring confirmation by typical cystoscopic and histological features, but not stating exactly what these typical features were (4,13,14).

This was followed by the EAU Guidelines on Chronic Pelvic Pain which introduced the term BPS with IC no longer recommended (5).

The controversy here lies in the use of the word "pain" or "painful" in the name and has particularly been challenged by the East Asian countries on the grounds that there are patients who have no sensation of pain but rather of pressure, discomfort or fullness even when the bladder is nearly empty.

While the definition of pain by the IASP does indeed cover unpleasant sensations: "an unpleasant sensory and emotional experience associated with actual or potential tissue damage, or described in terms of such damage", this definition is very much a researcher's definition, since a patient with an unpleasant sensation or discomfort in the bladder does not consider this to be pain and will therefore deny having pain when questioned by the doctor (6).

While many milder (or perhaps a different phenotype of) IC/BPS/HSB patients have urgency/frequency with discomfort rather than pain, the names containing pain (PBS and BPS) create the impression that these patients must have pain (for a diagnosis), although a substantial number do not. These terms could therefore be considered restrictive and confusing. Perhaps not for the experts, but potentially for primary care, the general urologist and certainly for the patient, whereas standard terminology and definitions must be crystal clear to non-experts too, particularly if patients are to play a full role in their healthcare today and in the future.

In practical terms, PBS or BPS as a name left a group of patients—potentially undiagnosed—in a "grey area" who were then neglected, excluded from research studies, swept under the carpet and may have ended up wrongly diagnosed as overactive bladder (OAB), leading one to wonder just how many patients grouped under OAB may actually have a hypersensitive rather than hyperactive bladder.

Even though the term "unpleasant sensation" was included in some new definitions of IC/BPS, it made little difference because the name suggested that pain was essential. This led to objections by Japanese/East Asian IC researchers who then revived the old ICS term "hypersensitive bladder", a name that in fact not only covers all the patients in the spectrum, but also matches definitions that include symptoms such as unpleasant sensation, discomfort or pressure. While the West has been brushing this aside as an "oriental cultural difference", it most definitely is not. But the "pain-in-the-name" issue continues to be a major cause of failure to reach global consensus regarding nomenclature. Furthermore, patient organizations were naturally offended that nobody saw fit to invite them around the table for discussions about their disease, and this led to a disintegration of the hitherto good relations between the physician experts and the patient support groups (8,15).

It was soon realised in the United States and Europe that use of the term BPS or PBS alone would potentially create huge problems in electronic databanks, including for reimbursement and social benefits, since IC was the term commonly used for the coding, with chronic IC used in the ICD, while no code existed for either BPS or PBS, let alone HSB. Nobody had thought of this impact on the patient in advance. Why not?

It was the AUA that put forward a possible solution to this in its guideline of 2011 (amended 2014), suggesting the term IC/BPS, with the IC always at the front, in the hope that it would be picked up when searching electronic systems. However, only time and experience will show whether this actually works in practice, both in clinical healthcare and when searching databases. And the whole experience with this has demonstrated how essential it is to take into account the potential impact on the patient following implementation (9,13).

A "symptom definition" controversy and the impact on the patient: urgency

The definition of "urgency" has become one of the hot topics in lower urinary tract standardization in the past decade and a group of patients who have been most adversely affected are the IC/BPS/HBS patients.

In the ICS Standardization of Terminology of Lower Urinary Tract Function in 1988 (16), the term urgency was defined as follows:

"Urgency may be associated with two types of dysfunction:
(I) Overactive detrusor function (motor urgency);
(II) Hypersensitivity (sensory urgency)" (16).

While this was never considered an ideal definition, it did at least indicate that an urgent need to void manifests itself in more than one form. However, in its 2002 LUTS Standardization document, the ICS now redefined this term as: "the complaint of a 'sudden' compelling desire to pass urine which is difficult to defer" (4). Introduction of the word "sudden" effectively restricted this term to urge urinary incontinence as found in OAB and there was no longer any mention of any other kind of sensation of urgency due to pain or hypersensitivity.

In practice this meant that the term urgency could no longer be used in definitions for other bladder conditions with an urgent need to void without the threat of incontinence, including all the IC and painful or HSB patients.

Practical consequences of this ICS re-definition included the following:

(I) Many researchers and physicians were now claiming that IC/BPS patients do not suffer from urgency/an urgent need to void;

(II) This meant that no progress was made in research into urgency in IC/BPS patients;

(III) It was questioned whether IC/BPS patients even needed treatment for a symptom that according to the ICS they did not have.

This started to escalate and at a meeting of the Association of Reproductive Health Professionals (ARHP) in 2007 in the USA, the term "persistent urge" was coined to replace the term urgency in definitions for this group of patients, with a definition of IC/PBS (published May 2008) now reading as: "*Pelvic pain, pressure or discomfort related to the bladder, typically associated with persistent urge to void or urinary frequency, in the absence of urinary infection or other pathology*" (17). However, persistent urge does not mean an urgent need to void; at most it is another description of frequency or hypersensitivity, whereas an urgent need to void in IC/BPS/HSB patients is due to the presence of overwhelming pain or other unpleasant sensation.

Consequently, the controversy continued, the term urgency was omitted from all new definitions of IC/PBS/BPS/HSB and patients were quite rightly most indignant, since urgency has been a key symptom of their disease for well over a hundred years and they tended to feel that their symptoms were being "stolen" from them! Fortunately, there is now an increasing realization that there are indeed several types of urgency sensation and hopefully this problem will be resolved in new standards.

Summary

Standardization of terminology and definitions impacts on every link in the healthcare chain, directly affecting the patient with regard to diagnosis, treatment, eligibility for reimbursement, social benefits and care. Complex healthcare coding and electronic record systems and global reliance on the internet and search machines mean that new terminology or changes now need in some way to be designed to last or to be updated in a way that will cause least disruption, while still allowing for evolution, since changes have far more ramifications today than they ever did in the past. Official recognition of a condition is vital, so coding must be correct and uniform across all authorities. Potential problems must be anticipated at an early stage in the process.

In order to achieve global consensus, stakeholders from all parts of the world need to sit around the table together and everyone's viewpoint should be taken into full consideration. Patient representatives and organizations should be involved in the standardization process to ensure that all aspects of a condition are covered, that no patients or symptoms are excluded and that there is no adverse impact on the patient in practical terms following implementation. The trend today is for patients to be more involved in healthcare and decision-making. We must ensure that standardisation and guidelines do not get left behind in this process of development either now or in the future. However, we need to find some way of training patient representatives to enable them to participate fully in standardization and guidelines and also encourage them to do so.

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Footnote

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