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Introduction

Bladder Pain Syndrome (BPS) is one of most disabling of the pelvic pain syndromes because of the urinary symptoms of urgency and frequency accompanying the condition. Pain perceived to be of bladder origin is the key symptom for the diagnosis of BPS, but in many patients the urinary symptoms might have an equally or even higher influence on the quality of life of the patient. Some patients have to void up to 50 times per day and night up making patients' life anchored to the toilet, destroying ability to work, destroying social and family life and often with severe economical consequences [1].

As the etiology(ies) of the disease is unknown, treatment is empiric and symptomatic and in many cases insufficient. Reconstructive surgery in the form of bladder augmentation or even urinary diversion with or without cystectomy might be an option in these patients not responding satisfactory to conservative treatment.

It must however be remembered that urinary diversion and cystectomy includes extensive and, in principle, irreversible procedures for a disease that otherwise imply a modest risk of death or life-threatening complications, but carries of a number of severe consequences and up to 48 % of early complications like UTIs, pyelonephritis, ureteroileal leakage, and anastomotic/stomal stenosis [2].

In a questionnaire to urologists in USA in 2001 Gershbaum and Moldwin found urinary diversion to be the most common surgical treatment for BPS underscoring the severity of the disease for the patients quality of life and the limitations of conservative treatments [3].

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Patient Selection

About 25 % of patients with BPS do not become pain free after urinary diversion (Burkes D, 2013, personal communication). It is essential that a patient has been through an extensive workup and trials of conservative treatment before irreversible surgery is undertaken. In these cases, a decision to perform major reconstructive surgery is relevant. It is a decision made in close cooperation between the patient and the doctor. Many patients are very reluctant to accept such major surgery for a “not life-threatening disorder” because of the change of body image and the risk of short and long-term complications. If the patient for various reasons does not want major surgery, this of course settles the question. But also for the doctor, this is a difficult decision.

The first question that arises in patient selection is whether the pelvic pain really comes from the bladder. In the case of evident objective bladder pathology like Hunner lesions or a contracted bladder, this question is easily answered. The use of bladder-related symptoms, such as pain associated with bladder filling and relieved by bladder emptying, is more speculative. As with most other chronic pain syndromes, the decision to move forward with aggressive treatment is based upon small retrospective studies and expert opinion. In this context, it must however be remembered that in these patients two major symptoms are the underlying cause of the patients poor quality of life: PAIN and FREQUENCY. At least it can be promised that a urinary diversion will solve the problem of frequency making daily life much easier to manage for some patients.

Choice of Procedure

It must be decided what sort of reconstructive surgery should be performed. Should the patient have a urinary diversion with or without a continent reservoir? Should the patient have a cystectomy? or a bladder augmentation? The literature seems to favor bladder augmentation to be reserved for patients with end stage Hunner lesion disease (classic IC or ESSIC type 3C) with a contracted bladder *and little or no pain* [4–7] although some report good results in other bladder pain groups including subtotal cystectomy in the procedure [8]. The relatively high percentage of patients needing to perform clean intermittent self-catheterization is also a limiting factor for choosing bladder augmentation, as many patients cannot tolerate the pain associated with urethral manipulation of any sort.

The amount of bowel surface to be incorporated into the urinary tract should be decided [9]. The risk of hyperchloremic metabolic acidosis must be considered making kidney function an important issue if a continent reservoir is considered. In this case a glomerular filtration rate in excess of 40 ml/min/1.73 m² body surface must be present. The use of a part of the ileum might compromise bile acid reabsorption leading to diarrhea and in patients with poor anal sphincter function to fecal incontinence. Also uptake of cobalamin/folic acid might be compromised.

Urinary Diversion

Urinary diversion in the form of a simple ileal conduit (Bricker procedure) or with a continent reservoir seems in most patients to be a reliable choice. A continent diversion requires a patient with an unspoiled cognitive ability and good manual dexterity. In a younger patient, meeting the above-mentioned prerequisites, a continent diversion might be preferable whereas an older patient probably is better served with a Bricker conduit due to risk of complications and difficulties handling self catheterization of a reservoir. However, it is important to note that it seems as patients with benign functional or inflammatory bladder diseases experience comparatively more problems with their reservoirs than patients with spinal cord injury or malignant bladder disease [10].

Recent reports seem to confirm the reasonably good clinical results after urinary diversion. In a Swedish study, 14 patients with Hunner lesions all became symptom free after diversion while only five of 13 BPS patients without Hunner lesion became symptom free after getting a continent reservoir [6]. In a study from Norway, 28 of 38 patients became pain free after either diversion (23 patients) or augmentation (15 patients). Two of the patients had Hunner lesions and became pain free [11]. Finally in a Danish report 19 of 23 patients became pain free after diversion with an ileal conduit. Also here two patients with Hunner lesions became pain free [12].

Braking down the literature on the outcome after urinary diversion based upon the different ESSIC types [13] is rather difficult. As mentioned above, outcome seems almost always very good in the Hunner lesion group (ESSIC type 3) (Fig. 25.1), while outcome in ESSIC groups 1 and 2 is more difficult to judge. In the

ESSIC Classification of bladder pain syndrome (BPS) types

cystoscopy with hydrodistension

| | | not done | normal | glomerulations ¹ | Hunner's lesion ² |
|--------|-----------------------|----------|--------|-----------------------------|------------------------------|
| biopsy | not done | XX | 1X | 2X | 3X |
| | normal | XA | 1A | 2A | 3A |
| | inconclusive | XB | 1B | 2B | 3B |
| | positive ³ | XC | 1C | 2C | 3C |

¹ cystoscopy: glomerulations grade 2-3

² with or without glomerulations

³ histology showing inflammatory infiltrates and/or detrusor mastocytosis and/or granulation tissue and/or intrafascicular fibrosis.

Fig. 25.1 ESSIC classification of cystoscopic and morphological findings

Danish study, 17 of 21 patients with ESSIC groups 1C and 2C became pain free (81 %) [12]. The bladder capacity during bladder hydrodistention in general anesthesia had no predictive value [12], and glomerulations during or after bladder distension had no pathophysiological meaning and no influence on disease progression or results of treatment [14].

Bladder Augmentation with Subtotal Cystectomy

Many patients are very reluctant to accept a urinary diversion with a stoma of any sort. Bladder augmentation with subtotal cystectomy has therefore been frequently used as an alternative. Due to our own poor results with this procedure we do not offer it to patients with BPS except for the rare patient with end stage contracted bladder and little or no pain. We inform the patient that we very rarely do this procedure due to the high risk of both persistent bladder pain and need for intermittent self-catheterization and reserve the procedure for patients with small contracted bladder with little or no pain. Almost all bowel segments have been tried including ileum, ileocecum, coecum, right colon, sigmoid colon, and gastric segments. In the chapter on BPS from the 5th International Consultation on Incontinence it was concluded: *There is no significant difference between bowel segments with regard to outcome except for gastric tissue substitution that is associated with dysuria and persistent pain due to production of acids* [15].

Cystoplasty might be performed with an either subtrigonal or supratrigonal cystectomy. Subtrigonal cystectomy carries more complications with no better results and is therefore little used today. The results of supratrigonal approach reported by Nielsen et al. in 1990 were rather poor with two failures in eight patients. Van Ophoven reported 5-year results of orthotopic substitution enterocystoplasty in 18 patients with only two failures [8]. Rössberger reported excellent results in patients with Hunner lesion, but not so in patients with non Hunner BPS [6]. Finally, Kim had excellent results in 45 patients with Hunner lesions [7].

More favorable results have been reported in patients with small cystoscopic bladder capacity (<200 cc) [16, 17]. Cystoscopic low bladder capacity has in the literature been taken as a sign of end stage inflammatory fibrosis of the bladder. This is however not always the case. Sairanen et al. described the long-term effect of cyclosporine treatment in BPS patients [18], and in many of these patients functional bladder capacity after treatment far exceeded cystoscopic bladder capacity before treatment. Three patients with a cystoscopic bladder capacity of 200 cc, and an average functional bladder capacity of 70 cc, had a functional bladder capacity of 290, 220, and 350 cc after cyclosporine treatment. Two patients with a cystoscopic bladder capacity of 300 cc and a functional bladder capacity of 92 and 100 cc increased the functional bladder capacity to 490 and 350 cc.

Urinary Diversion with or Without Cystectomy?

Although supportive literature is scarce, it appears that pain relief after urinary diversion is comparable whether or not the bladder is also removed [19]. Andersen et al. [11] retrospectively evaluated 16 patients having subtotal cystectomy and bladder augmentation and 20 patients having diversion without cystectomy. Seven and eight patients respectively later had a cystectomy due to persistent pain, but it is unfortunately not reported if cystectomy relieved the pain. The final result was that there was no difference in pain improvement in patients with and without cystectomy. Sixteen of 20 non-cystectomized patients became pain free (80%), while 12 of 18 cystectomized patients became pain free (66%), ($p=0.85$).

Secondary cystectomy rarely adds anything in the form of pain relief [6, 12], but should be performed if recurrent bacterial bladder infections are a part of the disease spectrum due to the risk of developing pyocystis [12].

Conclusion

In BPS patients, reconstructive surgery is the ultimate treatment choice after failure of conservative or less invasive surgical treatments have failed. Nevertheless, reconstructive surgery often ends up being the treatment of choice due to the lack of efficacy of less invasive treatments and the often severely disabling symptoms.

Urinary diversion is the most frequently chosen procedure. Although pain relief is “only” seen in about 80% of the patients, the often equally disabling symptom of frequency is “cured” by this procedure. It is often clinically impossible to know if the pelvic pain is of bladder origin or not. More bladder-related symptoms like pain on bladder filling and relief by bladder emptying might help. Glomerulations at bladder distension and cystoscopic bladder capacity in general anesthesia are of no predictive value. Also objective findings of bladder pathology like Hunner lesion or increased mast cell count in the detrusor might be useful. The choice between a continent diversion and a conduit depends on patient preference, patient age, kidney function, and amount of intestine to be used.

Augmentation cystoplasty should be reserved for patients with contracted bladder and in the absence of pain, which in practice often is end stage Hunner lesion patients. It should be kept in mind that decreased bladder capacity during cystoscopy in general anesthesia might be due to inflammation and not fibrosis and thereby be reversible.

Primary cystectomy should be reserved for patients with a component of recurrent bacterial cystitis due to the risk of pyocystis. Secondary cystectomy has seemingly a limited symptomatic effect in patients having had a previous urinary diversion.

References

1. Koziol JA, Clark DC, Gittes RF, Tan EM. The natural history of interstitial cystitis: a survey of 374 patients. *J Urol.* 1993;149(3):465–9.
2. Nieuwenhuijzen JA, de Vries RR, Bex A, van der Poel HG, Meinhardt W, Antonini N, et al. Urinary diversions after cystectomy: the association of clinical factors, complications and functional results of four different diversions. *Eur Urol.* 2008;53(4):834–42. discussion 42–4.
3. Gershbaum D, Moldwin R. Practice trends for the management of interstitial cystitis. *Urology.* 2001;57(6 Suppl 1):119.
4. Nielsen KK, Kromann-Andersen B, Steven K, Hald T. Failure of combined supratrigonal cystectomy and Mainz ileoceceocystoplasty in intractable interstitial cystitis: is histology and mast cell count a reliable predictor for the outcome of surgery? *J Urol.* 1990;144(2 Pt 1):255–8. discussion 8–9.
5. Peeker R, Aldenborg F, Fall M. The treatment of interstitial cystitis with supratrigonal cystectomy and ileocystoplasty: difference in outcome between classic and nonulcer disease. *J Urol.* 1998;159(5):1479–82.
6. Rossberger J, Fall M, Jonsson O, Peeker R. Long-term results of reconstructive surgery in patients with bladder pain syndrome/interstitial cystitis: subtyping is imperative. *Urology.* 2007;70(4):638–42.
7. Kim HJ, Lee JS, Cho WJ, Lee HS, Lee HN, You HW, et al. Efficacy and safety of augmentation ileocystoplasty combined with supratrigonal cystectomy for the treatment of refractory bladder pain syndrome/interstitial cystitis with Hunner's lesion. *Int J Urol.* 2014;21 Suppl 1:69–73.
8. van Ophoven A, Oberpenning F, Hertle L. Long-term results of trigone-preserving orthotopic substitution enterocystoplasty for interstitial cystitis. *J Urol.* 2002;167(2 Pt 1):603–7.
9. Mills RD, Studer UE. Metabolic consequences of continent urinary diversion. *J Urol.* 1999;161(4):1057–66.
10. Jonsson O, Olofsson G, Lindholm E, Tornqvist H. Long-time experience with the Kock ileal reservoir for continent urinary diversion. *Eur Urol.* 2001;40(6):632–40.
11. Andersen AV, Granlund P, Schultz A, Talseth T, Hedlund H, Frich L. Long-term experience with surgical treatment of selected patients with bladder pain syndrome/interstitial cystitis. *Scand J Urol Nephrol.* 2012;46(4):284–9.
12. Norus T, Fode M, Nordling J. Ileal conduit without cystectomy may be an appropriate option in the treatment of intractable bladder pain syndrome/interstitial cystitis. *Scand J Urol.* 2014;48(2):210–5.
13. van de Merwe JP, Nordling J, Bouchelouche P, Bouchelouche K, Cervigni M, Daha LK, et al. Diagnostic criteria, classification, and nomenclature for painful bladder syndrome/interstitial cystitis: an ESSIC proposal. *Eur Urol.* 2008;53(1):60–7.
14. Wennevik GE, Meijlink JM, Hanno P, Nordling J. The role of glomerulations in bladder pain syndrome: a review. *J Urol.* 2015;195(1):19–25.
15. Hanno P, Dinis P, Lin A, Nickel C, Nordling J, van Ophoven A, Ueda T. Bladder pain syndrome. In: Abrams P, Cardozo L, Khoury S, Wein A, editors. *Incontinence.* 5th ed. Paris: ICUD-EAU; 2013.
16. Messing EM, Stamey TA. Interstitial cystitis: early diagnosis, pathology, and treatment. *Urology.* 1978;12(4):381–92.
17. Hohenfellner M, Black P, Linn JF, Dahms SE, Thuroff JW. Surgical treatment of interstitial cystitis in women. *Int Urogynecol J Pelvic Floor Dysfunct.* 2000;11(2):113–9.
18. Sairanen J, Forsell T, Ruutu M. Long-term outcome of patients with interstitial cystitis treated with low dose cyclosporine A. *J Urol.* 2004;171(6 Pt 1):2138–41.
19. Nordling J, Blaivas JG. Should urinary diversion for bladder pain syndrome/interstitial cystitis include cystectomy? *J Urol.* 2014;191(2):293–5.