

Hard flaccid syndrome

BY KAYLIE HUGHES, ARIE PARNHAM AND MARC LUCKY

Chronic pelvic pain is defined as pain present below the level of the umbilicus with a duration of more than six months [1,2]. Chronic pelvic pain syndromes (CPPS) are highly prevalent in Western society, affecting both males and females. Studies have shown that CPPS significantly impacts upon a person's quality of life because of debilitating physical symptoms along with the psychological burden of the condition [1-5].

Until recently, non-bacterial chronic prostatitis has been widely accepted as the leading cause of chronic pelvic pain in men [4]. However, a new phenomenon, known as hard flaccid syndrome, is being recognised as an alternative cause of unremitting pelvic pain in men by specialist pelvic floor physiotherapists and some urologists.

Currently, no published literature exists of the syndrome. Using observed information obtained from men's health and pelvic floor physiotherapy forums (www.allthingsmale.com, <https://yourbrainrebalanced.com>, <https://pelvicpainrehab.com/>, <https://curehardflaccid.wordpress.com/>) we aim to raise awareness of this condition. We present the common signs and symptoms men exhibit and discuss how a multimodality holistic approach is the gold standard management.

What is hard flaccid syndrome?

Hard flaccid syndrome is a type of CPPS. The true prevalence is unknown, but cases have been reported in forums from men aged from their late teens to the seventh decade of life. Symptoms can be present for many years before correct diagnosis occurs.

Clinical features

Patients usually seek medical advice because of the following symptoms:

- Penile and perineal pain. This is most severe in the standing position, less so in the sitting position and absent when supine. The pain regresses when urinating.
- Patients report shortening of the penis associated with a constant cramp / clenching sensation in the pelvis.
- Penile sensory changes can be reported with the penis feeling numb to touch.
- Erectile dysfunction. There can be loss of morning and nocturnal erections. Excessive physical stimulation is often required to obtain an erection, in addition to visual stimulus or cognitive thought. The glans

can remain flaccid during erection.

- Pain on ejaculation and urination with a reduction in urinary flow.

At presentation, psychological symptoms are usually simultaneously present, such as insomnia, anxiety, depression, feelings of low self-worth leading to social isolation and suicidal thoughts.

Clinical signs

Abdominal and digital rectal examination are unremarkable. Examination of the penis in the flaccid state can reveal engorged corpora cavernous muscles like a semi-erect penis, with a rubbery texture. In the erect state, the penis feels very tight, with men reporting increasing levels of pain. The glans penis can remain flaccid unlike a normal erection.

Aetiology

The true aetiology of this condition is unknown. Like many CPPS the development of hard flaccid syndrome is thought to be multifactorial in nature. Biological, psychological and social influences all contribute to the development and severity of the condition by altering the neurovascular supply to the muscles of the pelvic floor and penis.

Stress is a key risk factor for the development of this condition by way of causing prolonged contraction of the muscles of the pelvic floor. Stress can be triggered by an injury directly to the penis during sexual intercourse or masturbation, or stress secondary to psychosocial distress in the absence of injury.

Within the body, psychological stress triggers the release of adrenalin with a primitive fight or flight response. The body goes into high alert; blood is directed away from certain visceral organs like the stomach to the muscles of the limbs and pelvis, so danger can be avoided.

One of the biological theories proposed to explain the relationship between stress and the development of hard flaccid syndrome is as follows:

- Initial stress, be it physical or psychological, triggers an abnormal fight or flight response resulting in increased sympathetic stimulation to the muscles of the pelvis via the perineal branch of the pudendal nerve.
- A surge of adrenaline, noradrenaline and cortisol is released from the afferent nerve fibres promoting increased blood flow to the bulbospongiosus, ischiocavernosus muscle and levator ani muscles as well as

sustained muscle contraction.

- Sustained contraction of the ischiocavernosus and bulbocavernosus muscle results in obstructed venous outflow from the penis via compression of the deep dorsal vein. This process is likely to be responsible for the semi-engorged penis in the flaccid state.
- Prolonged contraction of the muscles results in pelvic myoneuropathy secondary to neurogenic inflammation. The muscles physically lose their ability to relax, remaining contracted. The severe cramping, clenching sensation men describe in their pelvis, penile shortening, erectile and urinary dysfunction is likely attributed to this.
- Myofascial syndrome then occurs. Multiple painful trigger points develop in the muscles, resulting in the area becoming hypersensitive. Slight stimulation to the penis / pelvis results in a complex cascade of afferent and efferent nerve impulses, resulting in the secretion of neuropeptides which consequentially cause severe pain and inflammation disproportionate to the initial stimulus.

The psychosocial factors contributing to the development of this syndrome are related to the psychosexual nature of the condition. Men report deterioration or recurrence of symptoms at times of elevated stress. It is not uncommon at presentation for men to be trapped in a vicious cycle. They can often agonise about the future leading to extreme levels of stress and anxiety, triggering more sympathetic stimulation to the pelvis. Men can become fixated, performing regular attentive self-examination to the area which may be hypersensitive. This results in further muscle spasm and exacerbation of symptoms. A possible explanation why men catastrophise is that they frequently blame themselves for past events such as excessive masturbation or sexual techniques, which may be totally unrelated. Their overall perception of body image changes as they become hyperaware of their sexual organ. They feel suboptimal and this in turn results in altered libido and an aversion to sexual encounters. Over time chronically elevated levels of cortisol can lead to lowered testosterone levels which can alter libido and sexual performance. This can lead to the perception of loss of masculinity [6-11].

Investigation / diagnosis

As with all chronic pain syndromes hard

flaccid syndrome is a diagnosis of exclusion [3,4]. The majority of men will have seen several doctors and may have been misdiagnosed with alternative conditions such as Peyronie's disease before the correct diagnosis is made. It is not uncommon for men to have received intensive investigation such as: blood tests including hormonal profiling and prostate specific antigen (PSA), urinalysis and urine culture, sexually transmitted infection screen, flexible cystoscopy, Doppler ultrasound of the penis and MRI of the pelvis / penis, all of which yield normal results.

Clinical history and examination can help direct towards a diagnosis. Key signs and symptoms may be shortening of the penis (although this is highly subjective), chronic pain which regresses in the supine position and on micturition, and men typically describing the penis as "feeling hollow". As mentioned previously, on examination the penis can be semi engorged in the flaccid state and have a rubbery texture.

Management

Hard flaccid syndrome is difficult to treat due to the fact it is poorly understood and not yet widely recognised as a condition by urologists. Like with any CCPS, adoption of a multimodality holistic approach is paramount when managing these men [4].

The most important step in the management of this condition is continued reassurance that physically there is nothing functionally wrong with the penis and that this is a chronic pain syndrome. The patient should be guided to the conclusion that they are not to blame for their symptoms which may alleviate some of the anxiety surrounding the disorder [12-14]. Simple analgesia and medication for neuropathic pain such as nortriptyline can be used for symptom relief [4]. Supervised pelvic floor, biofeedback and trigger point physiotherapy with reverse Kegel exercises and Z-wands, allow men to learn techniques which they can perform and continue to practise at home to successfully relax their pelvic floor [15,16].

As anxiety is the main risk factor for development and persistence of this condition, early referral for counselling and cognitive behavioural therapy (CBT) is of upmost importance in management. Patients are taught to address any underlying emotional issues whilst promoting the growth and development of stress management techniques. Keeping a symptom diary may help with this. Promotion of good sleep, hygiene, healthy eating and exercise, all factors which are known to help reduce stress and improve wellbeing, is important. Regular exercise is also beneficial as it strengthens core muscles, taking pressure off the pelvic floor muscle.

Relaxation techniques like breathing exercises and encouragement of mindfulness

with meditation, yoga or exercise are all vital in the successful management of this somewhat debilitating condition [17-20]. CBT may also help patients come to terms and adapt to life living with a chronic condition.

Conclusion

Hard flaccid syndrome should be suspected in men of all ages who present with chronic pelvic pain associated with urinary and erectile dysfunction. At present this condition is not universally accepted by urologists because of the lack of published literature on the subject, despite the debilitating physical and emotional symptoms it may cause. By raising awareness of the syndrome, we hope to provide a better understanding of the disorder with a view to enabling earlier diagnosis, less misdiagnosis and quicker access to appropriate therapy.

References

1. Labat JJ, Robert R, Delavierre D, et al. Anatomy and physiology of chronic pelvic and perineal pain. *Prog Urol* 2010;**20**(12):843-52.
2. Dybowski C, Löwe B, Brünahl C. Predictors of pain, urinary symptoms and quality of life in patients with chronic pelvic pain syndrome (CPPS): A prospective 12-month follow-up study. *J Psychosom Res* 2018;**112**:99-106.
3. Passavanti MB, Pota V, Sansone P, et al. Chronic pelvic pain: assessment, evaluation, and objectivation. *Pain Res Treat* 2017;**9**:472925.
4. Sandhu J, Tu HYV. Recent advances in managing chronic prostatitis/chronic pelvic pain syndrome. *F1000Res* 2017;**6**:pii:F1000 Faculty Rev-1747.
5. Quaghebeur J, Wyndaele JJ. Prevalence of lower urinary tract symptoms and level of quality of life in men and women with chronic pelvic pain. *Scand J Urol* 2015;**49**(3):242-9.
6. Jansen AS, Nguyen XV, Karpitskiy V, et al. Central command neurons of the sympathetic nervous system: basis of the fight-or-flight response. *Science* 1995;**270**(5236):644-6.
7. Jantos M. Understanding chronic pelvic pain. *Pelvipereineology* 2007;**26**:66-9.
8. Hubbard DR. Chronic and recurrent muscle pain: Pathophysiology and treatment, and review of pharmacologic studies. *J Musculoskeletal Pain* 1996;**4**:123-43.
9. Hannibal KE, Bishop MD. Chronic stress, cortisol dysfunction, and pain: a psychoneuroendocrine rationale for stress management in pain rehabilitation. *Physical Therapy* 2014;**94**(12):1816-25.
10. Krsmanovic A, Tripp DA, Nickel JC, et al. Psychosocial mechanisms of the pain and quality of life relationship for chronic prostatitis/chronic pelvic pain syndrome (CP/CPPS). *Can Urol Assoc J* 2014;**8**(11-12):403-8.
11. Montenegro ML, Gomide LB, Mateus-Vasconcelos EL, et al. Abdominal myofascial pain syndrome must be considered in the differential diagnosis of chronic pelvic pain. *Eur J Obstet Gynecol Reprod Biol* 2009;**147**(1):21-4.
12. Tripp DA, Curtis Nickel J, Katz L. A feasibility trial of a cognitive-behavioural symptom management program for chronic pelvic pain for men with refractory chronic prostatitis/chronic pelvic pain syndrome. *Can Urol Assoc J* 2011;**5**(5):328-32.
13. Fry RP, Crisp AH, Beard RW. Sociopsychological factors in chronic pelvic pain: a review. *J Psychosom Res* 1997;**42**(1):1-15.
14. Franco JVA, Turk T, Jung JH. Non-pharmacological interventions for treating chronic prostatitis/chronic pelvic pain syndrome: a Cochrane systematic review. *BJU Int* 2018;doi:10.1111/bju.14492 [Epub ahead of print].
15. Anderson R, Wise D, Sawyer T, et al. Safety and effectiveness of an internal pelvic myofascial trigger point wand for urologic chronic pelvic pain syndrome. *Clin J Pain* 2011;**27**(9):764-8.
16. Van Alstyne LS, Harrington KL, Haskvitz EM. Physical therapist management of chronic prostatitis/chronic pelvic pain syndrome. *Phys Ther* 2010;**90**(12):1795-806.
17. Schaffer SD, Yucha CB. Relaxation & pain management: the relaxation response can play a role in managing chronic and acute pain. *AJN* 2004;**104**(8):75-82.
18. Hofmann SG, Sawyer AT, Witt AA, et al. The effect of mindfulness-based therapy on anxiety and depression: a meta-analytic review. *J Consult Clin Psychol* 2010;**78**(2):169-83.
19. Sutar R, Yadav S, Desai G. Yoga intervention and functional pain syndromes: a selective review. *Int Rev Psychiatry* 2016;**28**(3):316-22.
20. Chiesa A, Serretti A. Mindfulness-based stress reduction for stress management in healthy people: a review and meta-analysis. *The Journal of Alternative and Complementary Medicine* 2009;**15**(5):593-600.

AUTHORS



Kaylie E Hughes,

Specialist Registrar in Urology, St Helens and Knowsley NHS Foundation Trust, Prescot, Merseyside.



Marc A Lucky,

Consultant Urologist, Aintree University Hospital NHS Foundation Trust, Liverpool.

E: marc.lucky@aintree.nhs.uk

Declaration of completing interests: None declared

SECTION EDITOR



Arie Parnham MBChB, FRCS(Urol)

Consultant Urologist, The Christie NHS Foundation Trust, Manchester, UK

E: arie_parnham@hotmail.com