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EDITORIAL

Chronic proctalgia and chronic pelvic pain syndromes: New etiologic insights and treatment options

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Abstract

This systematic review addresses the pathophysiology, diagnostic evaluation, and treatment of several chronic pain syndromes affecting the pelvic organs: chronic proctalgia, coccygodynia, pudendal neuralgia, and chronic pelvic pain. Chronic or recurrent pain in the anal canal, rectum, or other pelvic organs occurs in 7% to 24% of the population and is associated with impaired quality of life and high health care costs. However, these pain syndromes are poorly understood, with little research evidence available to guide their diagnosis and treatment. This situation appears to be changing: A recently published large randomized, controlled trial by our group comparing biofeedback, electrogalvanic stimulation, and massage for the treatment of chronic proctalgia has shown success rates of 85% for biofeedback when patients are selected based on physical examination evidence of tenderness in response to traction on the levator ani muscle-a physical sign suggestive of striated muscle tension. Excessive tension (spasm) in the striated muscles of the pelvic floor appears to be common to most of the pelvic pain syndromes. This suggests the possibility that similar approaches to diagnostic assessment and treatment may improve outcomes in other pelvic pain disorders.

Key words: Biofeedback; Chronic pelvic pain; Chronic proctalgia; Coccygodynia; Levator ani syndrome; Pudendal neuralgia

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INTRODUCTION

Chronic or frequently recurring pain in the anal canal, rectum, or pelvis is a prevalent symptom that affects an estimated 6.6% of the population[1]. Although only 1/3 of people with such pains consult physicians, they nev-

ertheless report significant impairment in quality of life, work absenteeism, and psychological distress. However, despite its prevalence and impact, relatively little research has been published which addresses its epidemiology, pathophysiology, and treatment; and pelvic and rectal pain is widely considered frustrating to diagnose and treat. This may soon change: Our group recently reported a randomized controlled trial[2] in which we compared different approaches to treating chronic proctalgia and showed that the results were excellent 85% success rate-if the patients were appropriately screened. This study also provided new insights into the pathophysiology of chronic proctalgia and stimulated renewed interest in anorectal pain syndromes. The aims of this review are to critically assess what is known about the diagnosis and treatment of the most common forms of anorectal and pelvic pain, namely chronic proctalgia, chronic pelvic pain, coccygodynia, and pudendal neuralgia. This review is meant to help Gastroenterologists and Colorectal Surgeons when dealing with complex consultation on pelvic pain syndromes. It is mostly focused on chronic proctalgia and issues of differential diagnosis with other pelvic pain syndromes.

One of the challenges in caring for patients with anorectal and pelvic pain is that a number of inflammatory and structural etiologies must be considered. The organic diseases that are most commonly involved in chronic anorectal and pelvic pain are cryptitis, fissure, abscess, hemorrhoids, solitary rectal ulcer, inflammatory bowel disease, and rectal ischemia[3]. One should also consider chronic prostatitis and pelvic endometriosis as potential contributors to chronic pelvic pain[3]. Although the differential diagnosis is large and unfortunately poorly standardized, our experience[2] suggests that no organic disease explanation will be found in approximately 85% of patients presenting to gastroenterologists with chronic anorectal or pelvic pain. We screened 227 patients referred for unremitting, chronic rectal pain with a diagnostic evaluation that included digital rectal examination, colonoscopy, pelvic ultrasound and surgical consultation in all patients, plus gynecology and urology referrals in selected cases[2]. This extensive work-up identified only 33 patients (15%) with a probable organic disease accounting for their symptoms. Thus, for most patients with chronic anorectal or pelvic pain, the origin of the pain is uncertain and the relevant pathophysiological mechanisms are unclear. These are commonly defined as "functional" chronic anorectal and pelvic pain syndromes since no structural and anatomical disease was found. These functional pain syndromes constitute the main subject of this review.

CHRONIC PROCTALGIA

Chronic proctalgia is a general term for chronic or recurring pain in the anal canal or rectum[3]. Other names considered synonymous with chronic proctalgia are levator ani syndrome, puborectalis syndrome, chronic idiopathic perineal pain, pyriformis syndrome, and pelvic tension myalgia. Thiele, one of the first researchers to investigate this pain syndrome, called it coccygodynia, although he acknowledged that the pain was not in the coccyx[4]. To provide greater consistency in the diagnosis and labeling of anorectal pain syndromes, the Rome Ⅲ criteria[3] define chronic proctalgia as chronic or recurrent rectal pain or aching lasting at least 20 min, in the absence of structural or systemic disease explanations for these symptoms[3]. Pain duration of at least 20 min is a key feature since shorter episodes of pain are suggestive of proctalgia fugax, which is defined as a sudden, severe pain in the anorectal region lasting less than 20 min and then disappearing completely[3]. Proctalgia fugax may recur, but episodes are rare. Proctalgia fugax is believed to have a different etiology to chronic proctalgia, although there is no consensus on what causes it. Its consideration is beyond the scope of this review, which is intended to deal with chronic unremitting diseases.

Chronic proctalgia is further divided by the Rome Ⅲ criteria into two subtypes-levator ani syndrome (LAS) and unspecified functional anorectal pain-based on the presence or absence of a sensation of tenderness when the levator muscle is palpated during digital rectal examination. This classification updates the previous Rome Ⅱ classification in which LAS was designated as "highly likely" if traction on the pelvic floor produced a report of tenderness and only "possible LAS" if no tenderness was elicited[5]. Subgrouping patients with chronic proctalgia is consistent with clinical experience of different response to treatment, but distinct epidemiology and pathophysiology data are lacking[5]. Therefore, data provided mostly refer to chronic proctalgia patients as a whole.

Pathophysiology

Chronic tension or spasm of the striated muscles of the pelvic floor is commonly assumed to be the pathophysiological basis for chronic proctalgia[3,5-7], although there is no definitive evidence for this hypothesis. Inflammation of the levator or arcus tendon of the levator ani muscle has also been suggested as a cause of chronic proctalgia, since tenderness on palpation is most commonly found on the left side where the muscle inserts into the pubic ramus of the pelvis. However, contrary to this tendinitis hypothesis, local steroid injection has not been shown to be an effective treatment for chronic proctalgia[8]. In retrospective studies, many patients reported prior pelvic surgery, anal surgery and even spinal surgery as significant in the development of their pain syndrome[6,9]. Childbirth can be another precipitating factor[9]. In addition, high rates of anxiety disorders, depression, and stress are frequently reported in chronic proctalgia, and may act as significant precipitating factors in some patients[6,10].

Except for the exclusion of organic diseases, tests of anorectal physiology and imaging studies were traditionally considered to be of little diagnostic or prognostic

value[11,12]. Increased anal canal resting pressures tested by anorectal manometry were sometimes reported, but results were inconsistent. Grimaud and coworkers reported that LAS was associated with anal sphincter hypertonia and disordered defecation on dynamic proctography in a study of 12 patients, but this was not confirmed in a larger prospective study of 60 patients by Ger and coworkers[11,13] . Ger *et al*[11] reported that LAS was associated with paradoxical contraction of the pelvic floor muscles on straining as evidenced by anal electromyography or defecography. However, all these studies were potentially biased by small size, mixed patient population, and poor patient selection[3,5]. In addition, a number of structural disorders (descending perineum, rectocele, mucosal prolapse and pelvic floor dyssyenrgia) have been reported in small studies[6,11-13].

In a recent study, Hompes *et al*[14] reported on 59 patients referred to a Pelvic Floor Clinic for chronic functional anorectal pain who were tested by means of defecating proctography, anorectal manometry, anal ultrasound, and in selected cases, rectal examination under anesthesia. The same diagnostic protocol was applied to 543 rectal prolapse patients complaining of obstructed defecation and to a control group of patients with fecal incontinence. In the control group with fecal incontinence, pain was reported in 50% of patients but was a non-dominant symptom. Anorectal manometry failed to show any difference among groups. Rectal morphology examinations demonstrated high grade internal rectal prolapse in 59% of pain patients, which was often associated with symptoms of obstructed defecation. The authors concluded that rectal prolapse commonly underlies chronic proctalgia, particularly when obstructed defecation is present. However, the severity of prolapse did not correlate with pain intensity, leaving pain pathophysiology unclear[14]. In addition, chronic idiopathic rectal pain is sometimes reported as a complication of corrective surgery for rectal prolapse[14].

An innovative pathophysiology explanation for chronic proctalgia was recently reported by our group in a large, prospective, randomized controlled trial comparing biofeedback, electrogalvanic stimulation (EGS), and digital massage of the levator muscles for the treatment of chronic proctalgia. In this study, 157 patients with chronic proctalgia (confirmed by Rome Ⅱ criteria) were studied by anorectal manometry and a balloon evacuation test at baseline and again after 3 mo of treatment[2]. Based on a priori exclusion criteria, patients reporting symptoms consistent with either irritable bowel syndrome or functional constipation were not enrolled in the study. In patients reporting tenderness on palpation of the levator muscles (Rome Ⅱ: highly likely LAS, Rome Ⅲ: LAS), physiologic features of dyssynergic defecation (i.e., paradoxical contraction or failure to relax the pelvic floor on straining) were seen in approximately 85% of subjects in the absence of symptoms of constipation. Conversely, in patients who denied tenderness when the levators were palpated during digital rectal examination, inability to relax pelvic floor muscles when straining was an uncommon finding (19%). Dyssynergic defecation was a strong predictor of successful treatment outcome. These observations led us to conclude that the physiologic mechanisms responsible for LAS and dyssynergic defecation are similar[2].

This study also showed that the inability to relax pelvic floor muscles when straining to defecate may occur without symptoms of constipation, even though it is commonly assumed that dyssynergic defecation invariably results in obstructed defecation. Factors that interact with pelvic floor physiology to determine which symptoms develop-either pain or constipation-are left unanswered by our study and deserve further investigations. Also, we were not able to provide a physiological explanation for unspecified functional anorectal pain (i.e., anorectal pain without tenderness on digital palpation), which may represent a heterogeneous group of patients. Our study suggests, however, that adding a simple balloon evacuation test with a disposable Foley catheter to the diagnostic work up of chronic proctalgia patients enables one to select subjects that are more likely to benefit from pelvic floor rehabilitation.

Clinical presentation

Chronic proctalgia is often described by patients as a dull ache or pressure sensation in the rectum that is exacerbated by prolonged sitting and relieved by standing or lying down[3,5]. This pain rarely occurs at night; rather, it usually begins in the morning and increases in severity throughout the day. The pain may be precipitated by long-distance car travelling, stress, sexual intercourse and defecation[6,7]. During digital rectal examination, the examining finger is moved from the coccyx posteriorly to the symphysis pubis anteriorly[5,7]. For unexplained reasons, tenderness is often non-symmetric, being greater on the left side than on the right[5]. When performing digital rectal examination, the examiner should pause after inserting their finger into the rectum before applying traction on the levator muscles to avoid false positive results. In our experience, repeating the posterior traction on the levator muscle on the same exam is also useful to check for reproducibility and to avoid false positive results.

Diagnostic assessment

Although our recent study provides new insights into the pathophysiology of chronic proctalgia, these observations require validation by other laboratories before they can be incorporated as diagnostic criteria. The diagnosis of chronic proctalgia still relies on (1) clinical symptoms of recurring or chronic pain or aching in the anal canal or rectum with episodes lasting 20 min or longer[3], and (2) exclusion of alternative disease explanations for these symptoms by multiple diagnostic tests and consultations by other specialists. In addition, digital rectal examination should be performed to ascertain whether the patient reports tenderness when traction is applied to the levator ani muscles because this diagnostic sign

is a strong predictor of whether the patient is likely to benefit from treatments directed at relaxing pelvic floor muscles.

Treatment

No single treatment has been reported to be consistently effective in chronic proctalgia[3,7], and management can be a frustrating endeavor for both patients and physicians[11]. The first-line treatment most commonly provided is reassurance that the pain is of benign origin and is not suggestive of malignancy[7,9]. No data are available on the impact of reassurance, but education and counseling are often incorporated as a component of treatment.

Digital massage of the puborectalis sling, intended to relax tense muscles, was one of the first treatments proposed for chronic proctalgia[9]. Massage of the puborectalis muscle should be performed in a firm manner with the affected side massaged up to 50 times, depending on the patient's tolerance. Some claim that if the massage is not uncomfortable to the patient while being performed, it may not be effective[9]. Massage of the levator ani muscle is rarely performed as the sole therapy, with the most common adjunctive treatments being hot sitz baths or a short-term course of oral Diazepam, both of which are assumed to have myorelaxant properties. Earlier open-label studies suggested that digital massage combined with hot sitz baths and/or Diazepam were effective for relieving pain in 68% of 316 chronic proctalgia patients[15]. However, benefits seemed to fade away during long-term follow-up, and the addictive potential of Diazepam discourages long-term treatment[7].

Electrogalvanic stimulation, traditionally used by physiatrists to treat muscle spasticity[9], has also been advocated for the treatment of LAS when conservative therapy is ineffective. A low frequency oscillating current applied to the pelvic floor muscles through an anal probe, induces fasciculation and prolonged fatigue, which breaks the spastic cycle and may produce sustained symptom relief. Low frequency current has no thermal effect. No side effects have ever been reported other than mild worsening of pain on the first days of treatment. Sohn and coworkers were the first to test EGS in an open study of 80 chronic proctalgia patients[16]. They recommended a pulse frequency of 80 cycles per second with the voltage being gradually increased from zero to the point of discomfort (250-300 Volts according to patient's tolerance). Recommended treatment duration is one hour per day for 3 sessions in a ten-day period. In the Sohn study[16], 91% of patients reported good to excellent pain relief from EGS in the short-term, but no long-term follow-up was reported. This high percentage of success was never replicated by subsequent open label studies, although approximately two-thirds of patients did report short-term pain relief. Treatment protocols varied widely in terms of number and duration of sessions. Authors claimed that non-responders showed features of psychology disturbances, but no evidence was provided on the issue. However, three additional studies that investigated the long-term benefits of EGS treatment in chronic proctalgia found that only 25%-38% of patients reported persistent pain improvement[17-19].

Biofeedback treatment of LAS was first described in 1991 by Grimaud and coworkers[13]. They treated 12 patients with biofeedback techniques focused on voluntary relaxation of external anal sphincter tone. Pain disappeared in all patients after a mean of eight sessions. Subsequent studies using biofeedback were not able to replicate these results, with success rates varying from 35% to 87.5%[6,11,19]. All studies were small, none was controlled, and treatment modalities varied.

Botulinum Toxin A (BoTox A) was tested in a randomized controlled trial run in 12 patients, and no differences in rectal pain were observed between patients injected with active BoTox *versus* those injected with saline[20]. The average amount of time required to defecate a rectal balloon was actually increased after BoTox injection. The tendinitis (inflammation) hypothesis for chronic proctalgia was tested by steroid caudal block and by pelvic tender point injection of a mixture of Triamcinolone Acetonide and Lidocaine with negative results[8,11]. Sacral nerve stimulation was also reported to be beneficial in an open study involving 27 chronic proctalgia patients. However, when benefits were assessed by intent to treat analysis, pain relief was reported in less than 50% of subjects[21].

A major drawback in assessing the literature on chronic proctalgia treatment is the huge variation in inclusion criteria, outcome criteria, and follow-up intervals. Additional limitations are small sample sizes and lack of an appropriate control group. The few quasi-randomized studies had control groups that included subjects who received more than one treatment and patients not responding to a former therapy[7]. To overcome these limitations, Chiarioni and coworkers recently reported a prospective, randomized controlled trial of 157 chronic proctalgia patients to investigate the comparative effectiveness of the 3 most commonly prescribed treatments: biofeedback to teach pelvic floor muscle relaxation, EGS, and digital massage of the levator muscles[2]. A physiological assessment including manometry and balloon defecation was carried-out at baseline and at 1-3 mo follow-up. In addition, self-reported stool frequency was assessed at baseline and at 6-mo follow-up. The primary outcome was subjective reporting of adequate pain relief by the patient. Secondary outcomes included subjective pain improvement on an ordinal scale, number of days per month with rectal pain, and visual analog scale ratings of pain. According to Rome II criteria, proctalgia patients were subgrouped into highly likely LAS and possible LAS based on the presence or absence of levator tenderness at digital rectal exam, and randomization to treatment groups was stratified so that each treatment group contained a similar number of patients with a highly likely diagnosis of LAS.

At one-month follow-up, biofeedback was significantly more effective than EGS and massage by intent-to-treat analysis, with adequate relief of pain reported by 59.6% *vs* 32.7% *vs* 28.3% for biofeedback, EGS, and massage, respectively. Benefits were maintained throughout follow-up (12 mo) and no side effects were reported with any treatment. When results were further investigated in subgroups of patients, no treatment was effective in possible LAS patients (Rome Ⅲ unspecified functional anorectal pain). However, among patients with highly likely LAS (Rome Ⅲ levator ani syndrome) adequate relief was reported by 87% for biofeedback, 45% for EGS and 22% for massage at 1 mo follow-up. Improvements were maintained for the whole follow-up. The superiority of biofeedback was supported by all the secondary outcome measures including number of days per month with pain, which decreased from 14.7 per month to 3.3 per month for biofeedback, 8.9 for EGS, and 13.3 for massage[2].

Physiological measurements revealed that the mechanism for achieving adequate pain relief was an improvement in pelvic floor function from being unable to relax anal canal pressures on straining to being able to do so and/or an improvement on the balloon evacuation test from being unable to pass a 50 mL balloon to being able to do so[2]. This interpretation of the mechanism of action was confirmed by a post-hoc analysis showing that 94.2% of those who improved pelvic floor dysfunction on one or both of these measures reported adequate pain relief, while only 13.6% of those who did not improve pelvic floor function reported positive therapy outcome regardless of the treatment provided. In addition, stool frequency increased from baseline to post-treatment in responders, even in the absence of a former complaint of constipation. This study led us to conclude that biofeedback is an effective treatment for LAS, and EGS is somewhat effective. However, the minority of proctalgia patients affected by unspecified functional anorectal pain are still left without a satisfactory treatment option. In this regard, depression and anxiety are both frequently reported in non-responsive proctalgia patients[6,10]. Brain processing of pain may be altered in functional gastrointestinal disorders, but data in proctalgia patients are lacking[22]. In addition, no trial has actually evaluated the effect of either psychotherapy intervention or psychotropic drugs in proctalgia patients. Finally, there is no evidence that surgery can help these severely disabled patients. Invasive interventions should be avoided in the absence of a clearer etiologic understanding of non-responsive proctalgia patients[3].

COCCYGODYNIA

Coccygodynia is defined as pain arising in or around the coccyx, usually triggered by prolonged sitting on hard surfaces[23]. The pain is considered chronic when it lasts more than two months and it is commonly reported after repetitive trauma or childbirth[23,24]. Coccygodynia may also be of idiopathic origin or secondary to lumbar disc degeneration[23-25]. It is also rarely reported as a complication of epidural injection of anesthetic or of various rectal and spine surgery[25].

Pathophysiology

It is up to five times more common in women than in men, and obesity seems to be a predisposing factor due to the associated pelvic rotation[23-25]. The female pelvic anatomy may also predispose to coccygodynia by leaving the coccyx more exposed to traumatic injury. The exact etiologic mechanism/s associated with coccygodynia are still obscure. Chronic spasm of the pelvic floor exerting a painful tension on a stiff coccyx has been traditionally considered a relevant etiologic factor, with accidental trauma acting as a trigger[23-25]. However, instability of the coccyx potentially correlated with symptom severity was then discovered in a high percentage of patients by dedicated X-Ray examination[26]. In addition, it is unclear whether pre-existing spine alterations play a role by predisposing patients to develop post-traumatic coccygodynia[26,27]. Inflammation of structures (i.e., bursitis) in close proximity to the spine has also been described as a causative factor in a minority of patients complaining of coccygodynia[27]. Depression and anxiety disorder have been reported to amplify coccygeal pain symptoms[23]. Some authors do not diagnose coccygodynia when there is an ongoing medicolegal litigation, even if it occurs following a traumatic injury[24,25].

Clinical presentation

Pain in the coccyx and in close anatomical regions (sacrum, perineum, anorectum) is the main reported symptom[23]. Epidemiologic data on coccygodynia in the general population are lacking, but coccygodynia is considered to be a rare disorder. Retrospective data suggest that coccygodynia accounts for less than 1% of all reported cases of lower back pain[24,25]. Diagnosis of coccygodynia relies heavily on history and clinical exam. Questioning the patient about previous trauma to the coccyx or childbirth trauma is a must, since according to Salvati the absence of a previous trauma makes the diagnosis unlikely[9]. In addition, patients should report worsening of pain by prolonged sitting, bending, lifting or having a restricted poor posture for long intervals[23-25]. Some patients may report that standing from a sitting position triggers the pain[23].

Diagnostic assessment

Reproducing the usual pain by pressure or manipulation of the coccyx is key to diagnosis[23]. Patients may also report mild tenderness on puborectalis posterior traction on digital rectal examination and a differential diagnosis of chronic proctalgia needs to be entertained[9,28]. However, this maneuver should never be able to provoke the usual pain. Abnormal movement of the coccyx on palpation is an additional sign to confirm the clinical suspicion of coccygodynia[25]. Dynamic X-Ray investigation may support the clinical diagnosis. The standard lateral

X-Ray investigation of the coccyx in the standing position should be supplemented with a second film taken while the patient is sitting on a hard surface possibly in a posture worsening the pain[26]. More than 50% of patients would show features of coccyx instability (either exaggerated flexion or luxation) that seem to correlate with pain severity and previous traumatic events. An additional 15% of coccygodynia patients would show features of an abnormal bone spur at the end of the tailbone (so called spicule)[26]. Spine magnetic resonance imaging (MRI) could be performed to exclude tumors or disc disease, but do not seem to add significantly to the diagnosis in coccygodynia[25].

Treatment

The initial treatment of coccygodynia is focused on avoiding potentially offending factors and includes sitting on a donut-shaped pillow or a gel cushion to reduce pressure, posture ameliorating interventions, sitz bath and on demand nonsteroidal anti-inflammatory drugs[23,24]. This treatment is commonly applied for 6-8 wk. No controlled study has investigated the therapeutic outcome of these simple measures. When initial treatment fails most authors recommend adding digital manipulation of the coccygeal ligaments as well as intrarectal manipulation of the pelvic floor muscles. Various massage and manipulation techniques have been described in open studies to decrease coccygeal pain in up to 85% of patients, particularly when combined with local steroid injection or physiotherapy[29]. A recent prospective, randomized, controlled study aimed to compare intrarectal pelvic floor muscles manipulation (3 sessions) *vs* placebo physiotherapy (sacral short wave magnetic field applied at marginal power) in 102 chronic coccygodynia patients[30]. Primary outcome was subjective decrement of more than 50% in pain intensity on a visual analog scale score at follow-up intervals of 1-6 mo. At 1 mo follow-up, 22% of patients in the manipulation group reported a significant pain decrement compared to only 12% of patients in the placebo group. Benefits persisted throughout follow-up in both groups. Manipulation was more effective in recent onset coccygodynia of post-traumatic origin not associated with instability of the coccyx. Psychosocial factors seemed to predict a poorer treatment outcome. The authors concluded that intrarectal manipulation is at least mildly effective in chronic coccygodynia and suggested either to increase the number of therapeutic sessions or to add local steroid injection to improve outcome. However, no randomized study has actually evaluated both treatment options for coccygodynia. In selected patients with severe and unresponsive coccygodynia, surgery may be considered[31].

A recent review on surgical treatment of coccygodynia reported on 24 studies, but 22 of them were retrospective case series[32]. Surgery was a treatment option in a minority of patients (approximately 19%), but mean satisfaction rate for pain relief was high (over 80% of treated patients). Some series reported a satisfactory outcome of just 54% which was attributed to patient selection bias. Mean overall complication rate was 10.9% with wound infection being the most commonly reported complication. Surgeon expertise seemed to play a role since the smallest series reported the highest procedure-related complication rates (up to 50%). The type of surgery chosen was either total or partial removal of the coccyx and this did not seem to influence outcome. However, the worst outcomes were reported in patients with a history of rectal or spinal diseases and ongoing compensation issues[32].

PUDENDAL NEURALGIA

Pathophysiology

Pudendal neuralgia is a chronic pain in the perineal area secondary to entrapment and injury to the pudendal nerve in its musculo-osteo-aponeurotic tunnel between the sacrotuberal and sacrospinal ligaments, in the absence of organic diseases that may explain this symptom[23]. Pudendal neuralgia has been rarely described as secondary to herpetic neuropathy, stretch neuropathy, and post-radiotherapy neuropathy, but pudendal nerve entrapment is by far the most common etiology[33,34]. Pudendal neuralgia is also called Alcock's canal syndrome, or pudendal canal syndrome[23].

Clinical presentation

It is commonly described as a superficial pain, burning sensation, numbness, or paresthesia in the gluteal, perineal, and/or genital areas[23]. It may be homolateral or bilateral, radiate to the pelvis and the thighs, and be associated with deep pelvic discomfort[33,34]. Pain may be worsened by sexual intercourse and initially reported as sciatic pain[33,34]. The epidemiology of pudendal neuralgia in the general population is unknown. The diagnosis is rarely considered except in highly focused Pelvic Floor Units or in specialized Urogynecologist practices. It is usually considered to be a rare entity, but it may be overdiagnosed due to the functional comorbidities associated with pudendal nerve dysfunction[33,34]. Recently, a multidisciplinary Committee reported that pudendal neuralgia may be simply diagnosed by default in the presence of pelvic, perineal, and buttock pain without evidence of organic disease at diagnostic workup[35]. Particularly controversial is its association with rectal pain, the presence of which requires differential diagnosis with chronic proctalgia[23].

Diagnostic evaluation

Clinical neurophysiology has improved our knowledge of this disorder, but a definitive diagnostic test is still not available. As in many neuropathic pain syndromes, the diagnosis of pudendal neuralgia remains primarily clinical and should be reviewed in the light of the course of the disease. In 2006, a multidisciplinary working party on pudendal neuralgia held in Nantes, France, concluded that only the operative finding of nerve entrapment

and post-operative pain relief can formally confirm the diagnosis, provided the placebo effect of surgery is excluded[35]. However, this panel of experts identified four domains of diagnostic criteria for pudendal neuralgia: (A) essential criteria, (B) complementary diagnostic criteria, (C) exclusion criteria, (D) associated signs not excluding the diagnosis. Essential criteria are particularly relevant and will be discussed in detail. (1) Pain should be limited to the innervation territory of the pudendal nerve. This excludes any pain that is limited to the coccygeal, pelvic or gluteal areas; (2) Pain is predominantly experienced while sitting, in accordance with the nerve compression etiology hypothesis. In long-standing pudendal neuralgia, pain may become continuous, but it is still worsened by the sitting position; (3) The pain rarely awakens the patient at night; (4) On clinical examination, no objective sensory impairment can be found even in the presence of paresthesia. The presence of a sensory defect should prompt investigations to exclude diseases of the sacral nerve roots and the cauda equina; and (5) Pain should be relieved by anesthetic infiltration of the pudendal nerve. This is an essential criterion, but it lacks specificity as pain related to any perineal disease may be relieved by pudendal nerve block. Moreover, a negative block does not exclude the diagnosis of pudendal neuralgia because it may have been performed inadequately (e.g., too distally). The complementary diagnostic criteria include the sensation of a rectal foreign body and the worsening of pain during defecation, both of which should prompt the physician to entertain the differential diagnosis of chronic proctalgia. Exclusion criteria for pudendal neuralgia are pain in a territory unrelated to the pudendal nerve, symptomatic pruritus instead of paresthesia, exclusively paroxysmal pain, and imaging abnormalities that could explain the symptom[35].

Treatment

Pudendal neuralgia is treated by pudendal nerve block, which is both diagnostic and therapeutic. However, data on the long-term benefits of pudendal nerve block are lacking[35]. In addition, only the operative demonstration of nerve entrapment and post-operative pain relief can formally confirm the diagnosis of pudendal neuralgia secondary to it, except for a potential placebo effect of surgery[35].

CHRONIC PELVIC PAIN IN WOMEN

Chronic pelvic pain (CPP), which is diagnosed only in women, is commonly defined as noncyclic, nonmalignant pain in any organs related to the pelvis, in the absence of pregnancy and inflammatory bowel disease, that has lasted for at least six months[36]. Pain occurring exclusively in association with menstruation (dysmenorrhea) and sexual intercourse (dyspareunia) are generally not considered to be CPP, but general agreement is lacking. Other definitions include a pain severity sufficient to cause functional disability or to require medical care[36]. Since the definition of CPP varies, it is difficult to ascertain its exact prevalence. However, the prevalence of CPP in the general population assessed by mail questionnaires among women aged 18-50 has been reported to be as high as 15% in the United States and 24% in the United Kingdom[37,38]. CPP has been estimated to account for 10% of all outpatient referrals to gynecologists and 40% of diagnostic laparoscopies, so it constitutes a significant economic burden[39,40]. No organic disease is found on laparoscopy in at least a third of women with CPP[40]. In the community, 32% of patients who consult for this symptom report high rates of anxiety and quality of life impairment as measured by the SF-36[37,38]. Consulting behavior is directly influenced by the severity of pain[39].

Pathophysiology

The etiology of CPP is considered to be complex and multifactorial[36,40]. Some gynecological diseases such as endometriosis, pelvic inflammatory disease, and interstitial cystitis may cause CPP, but gastrointestinal comorbidities are also reported in up to 1/3 of CPP patients in primary care[38,40]. A psychosomatic component of pain has also been hypothesized[36,40]. The common association of CPP with irritable bowel syndrome has led some to question whether these two diseases are actually a single clinical entity that is diagnosed differently according to the specialist consulted[41]. The etiology of CPP is poorly understood.

Diagnostic assessment

Initial evaluation should include a history and physical examination to narrow the differential diagnosis[40]. When this examination does not identify another explanation for the pain, limited laboratory testing and transvaginal ultrasound scanning is often employed to rule out organic disease and reassure the patient[40]. The laboratory workup should include: complete blood count, beta human chorionic gonadotropin level, erythrocyte sedimentation rate, vaginal swabs for Chlamydia and Gonorrhea, and urinalysis with urine and culture[40]. Additional magnetic resonance imaging should be considered when in doubt for organic disease and diagnostic laparoscopy may be eventually performed in selected cases[40]. A tense pelvic floor is often reported during vaginal examination in CPP and spasm of the pelvic floor muscles is considered a relevant etiologic factor[36]. In addition, up to 60% of patients may report symptoms of either voiding dysfunction or dyschezia[42].

Treatment

Physiotherapy to relax the pelvic floor is often prescribed as first-line treatment for CPP, but randomized, controlled trials to confirm its effectiveness are lacking. Vaginal electrical stimulation was retrospectively reported to decrease pain in 52% of 66 chronic pelvic pain patients when coexistent levator ani spasm was also diagnosed by clinical exam[43]. Benefits were generally sustained during a 30-wk follow-up. Nonetheless, few studies have evalu-

ated pelvic floor function in women with CPP. Abbott and coworkers reported pelvic floor myalgia in 68 out of 118 patients referred for long-standing, unresponsive CPP[44]. They diagnosed pelvic floor myalgia based on objective evidence of contracted, painful pelvic muscles on palpation and elevated resting intraluminal pressures as measured by vaginal manometry. This study was a double blind, placebo controlled trial to test the efficacy of BoTox A in patients who had CPP with pelvic floor spasm. BoTox injection was associated with a significant reduction in vaginal resting pressure compared to placebo, but pain was only partially relieved and was not significantly different between BoTox and placebo. The authors concluded that pelvic floor spasm can cause CPP and that improvement in some symptoms occurs following reductions in muscle spasm[44]. In open studies, a number of treatment modalities have been reported to be effective for decreasing symptoms in CPP. These options include either oral or intramuscular hormone therapy, levator ani trigger point steroid injections, and sacral neuromodulation[36,40,42,45]. Most studies are open, retrospective, single center experiences with poor generalizability of treatment outcome.

Tricyclic antidepressants and Sertraline seem to work no better than placebo in CPP[40]. Surgery should be limited to patients with an organic cause for pelvic pain[36,40]. In non-responsive, severely disabled patients a multidisciplinary approach is advocated to fit with a biopsychosocial model of pain[40].

CONCLUSION

Chronic anorectal and pelvic pain syndromes receive little research attention despite the fact that they are prevalent, often disabling pain syndromes which are associated with significant health care costs and quality of life impact. Their frequency in the general population may be as high as 24% for chronic pelvic pain in women and 6.6% for chronic proctalgia. It is common for these patients to be referred to multiple specialists. Etiology is poorly defined, but chronic tension (spasm) in the striated muscles of the pelvic floor is often considered to be the pathophysiological mechanism for most of them. A recent randomized, controlled trial provided evidence that dyssynergic defecation (i.e., paradoxical contraction or failure to relax the pelvic floor muscles when straining to defecate) is the primary cause of pain for the majority of patients with chronic proctalgia, even for patients who do not complain of constipation. Biofeedback to treat dyssynergic defecation was an effective treatment for the subset of patients with chronic proctalgia who reported tenderness when traction was applied to the levator ani muscles during digital rectal exam (a sign of excessive tension in these muscles). This finding should prompt researchers to look for features of dyssynergic defecation in other pelvic pain syndromes and to try a similar treatment. A multidisciplinary and tailored approach to treat anorectal and pelvic pain patients without pelvic floor dysfunction is strongly suggested.

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