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Solitary rectal ulcer syndrome

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INTRODUCTION

Solitary rectal ulcer syndrome is an uncommon rectal disorder that can present with rectal bleeding, straining during defecation, and a sense of incomplete evacuation [1-3]. The term solitary rectal ulcer syndrome is a misnomer. Endoscopic findings in patients with solitary rectal ulcer syndrome can range from mucosal erythema alone to single or multiple ulcers and polypoid/mass lesions. This topic will review the pathogenesis, clinical features, diagnosis, and management of solitary rectal ulcer syndrome. Other causes of chronic colonic ulceration, including ulcerative colitis, Crohn disease, and ischemic colitis, are discussed in detail, separately. (See "Clinical manifestations, diagnosis, and prognosis of ulcerative colitis in adults" and "Clinical manifestations, diagnosis, and prognosis of Crohn disease in adults" and "Colonic ischemia".)

EPIDEMIOLOGY

The incidence of solitary rectal ulcer syndrome is uncertain but has been estimated in one study to be 1 in 100,000 individuals per year [4]. In one retrospective study of 80 patients, the median age at diagnosis was 48 years with a range of 14 to 76 years [5]. Males and females appear to be affected equally, but a slight female preponderance has been suggested in some reports of adults. By contrast, in a series of 140 children, the majority (79 percent) were boys [6].

PATHOGENESIS

The pathogenesis of the solitary rectal ulcer is incompletely understood. However, a number of factors appear to have a causative role in individual reports. It is possible that different etiologies may contribute to the development of solitary rectal ulcer syndrome.

A common observation in a number of reports is rectal prolapse or rectal intussusception and paradoxical contraction of the puborectalis muscle. A case-control study compared anorectal physiology and defecation proctography in 25 patients with solitary rectal ulcer syndrome with matched controls with either outlet obstruction (25 patients), overt rectal prolapse without any mucosal change (25 patients), or overt rectal prolapse with mucosal changes (14 patients) [7]. Patients with solitary rectal ulcer syndrome more frequently had increasing anal pressure at straining, paradoxical puborectalis contraction, and prolapse of the inner circular smooth muscle of the rectum and less frequently had complete rectal emptying compared with controls. Compared with patients with overt rectal prolapse, mean resting and squeezing anal pressures were significantly higher.

Rectal prolapse and paradoxical contraction of the puborectalis muscle can result in rectal trauma by two different mechanisms [7,8]:

- Prolapsed rectal mucosa is forced downward due to the pressures generated by the rectum during defecation. The opposing force of the paradoxical contraction of the puborectalis muscle can generate high pressures within the rectum and lead to mucosal ischemia, thus predisposing to ulceration.
- Shear forces on the rectal mucosa are caused by the contraction of the puborectalis muscle.

However, excessive puborectalis contraction is not found in all patients with solitary rectal ulcer. Furthermore, it is unclear whether prolapse causes ulceration or whether they are both varying manifestations of a similar disease process. It is also possible that prolapse is an associated condition, rather than a causative one, a hypothesis supported by a study in which surgical correction of the rectal prolapse had no significant impact on symptoms [9].

Direct digital trauma has also been implicated since many patients have a history of constipation and report attempts at manual disimpaction. However, a number of lesions have been described that were beyond the reach of an inserted finger.

A possible hormonal cause has also been proposed. A case report documented a woman with solitary rectal ulcer syndrome that resolved during two pregnancies but recurred when she was https://www3.utdos.ir/contents/solitary-rectal-ulcer-syndrome/print?search=Solitary rectal ulcer syndrome&source=search result&selectedTitle=1~43... 2/17

not pregnant [10].

CLINICAL FEATURES

Clinical manifestations — Symptoms may be absent in up to 26 percent of patients [5,11-13]. In an illustrative series of 98 adults with a clinicopathologic diagnosis of solitary rectal ulcer syndrome, the most common symptoms were rectal bleeding (56 percent), straining with defecation (28 percent), and pelvic fullness (23 percent) [11]. The most common presenting features were bleeding (94 percent), small frequent stools (56 percent), and constipation (27 percent) in a series of 140 children [6]. Other less frequent symptoms include mucous discharge, incontinence, tenesmus, and rectal pain.

In patients with a rectal prolapse, a painless, dark red mass at the anal verge with or without mucous may be demonstrated during straining. Palpation of the prolapsed mucosa between the thumb and forefinger permits the examiner to distinguish between mucosal (Type I) or complete (Type II) rectal prolapse (figure 1). (See "Overview of rectal procidentia (rectal prolapse)", section on 'Definition'.)

Imaging findings — Barium enema may be normal or may have nonspecific findings, including nodularity of the distal rectal mucosa, thickened rectal folds, rectal ulcers, and a rectal stricture [14-16].

EVALUATION

Diagnosis — Solitary rectal ulcer syndrome is usually diagnosed incidentally during the course of evaluation of the patient's symptoms of rectal bleeding, tenesmus, or straining during defecation. The endoscopic appearance of solitary rectal ulcer syndrome, while supportive of the diagnosis, is not specific. The diagnosis is made on the basis of histologic finding of fibromuscular obliteration of the lamina propria.

Endoscopy and histology — The endoscopic findings range from mucosal erythema to single or multiple ulcers and polypoid/mass lesions [5,14]. In one retrospective study of 51 patients with solitary rectal ulcer syndrome, only 35 percent of patients had a solitary rectal ulceration, 22 percent had multiple ulcers, and 43 percent had non-ulcerative lesions [4]. Ulcers are usually superficial and 1 to 1.5 cm in diameter, but they can range in size from 0.5 to 4 cm. In the majority of patients, the lesions are located in the anterior rectal wall within 10 cm of the anal verge, but they can also be located in the anal canal or the sigmoid colon [5,15]. On histology, solitary rectal ulcer is characterized by fibromuscular obliteration of the lamina propria that leads to hypertrophy and disorganization of the muscularis mucosa and regenerative changes with distortion of crypt architecture [2]. In polypoid cases, the mucosa has a villiform configuration, and in some cases, the glands may be trapped in the submucosa (colitis cystica profunda) [16].

Additional evaluation in selected patients — In patients with solitary rectal ulcer syndrome with persistent symptoms despite initial management, we perform an additional evaluation to diagnose a rectal intussusception, if a true rectal prolapse cannot be reproduced on physical examination. (See 'Subsequent management for persistent symptoms' below.)

Defecography — Defecography can demonstrate a rectal prolapse, rectal intussusception, or rectocele, as well as incomplete or delayed rectal emptying [17]. In a study of 53 patients with solitary rectal ulcer syndrome, 40 (75 percent) had abnormalities on defecography. The most common findings were internal and external rectal prolapse and delayed rectal emptying [18]. Magnetic resonance defecography can identify and grade both rectal intussusception and dyssynergia in solitary rectal ulcer syndrome, and also depict associated anterior and/or middle compartment descent [19]. (See "Etiology and evaluation of chronic constipation in adults", section on 'Defecography' and "Overview of gastrointestinal motility testing", section on 'Anorectum'.)

Endoscopic ultrasound — Endoscopic ultrasound is generally not required for the diagnosis of solitary rectal ulcer syndrome. Compatible findings include a thick hyperechoic submucosa and hypoechoic band in the muscularis propria with an intermediate hyperechoic layer [20,21]. (See "Endoscopic ultrasound for evaluating patients with rectal cancer".)

DIFFERENTIAL DIAGNOSIS

In order to diagnose solitary rectal ulcer syndrome, it is important to distinguish it from other disorders that can have similar clinical presentation and endoscopic appearance [11,22]. The distinction between them can be made by histopathological evaluation of biopsies of the colon. The presence of collagen infiltration of the lamina propria on histology can help distinguish solitary rectal ulcer syndrome from other etiologies [16,23].

• **Inflammatory bowel disease** – Ulcerative colitis and Crohn disease can have a similar endoscopic appearance to solitary rectal ulcer syndrome. In addition, prolapsed mucosa in patients with solitary rectal ulcer syndrome may contain inflamed and distorted mucosa and may be misdiagnosed as inflammatory bowel disease. However, the presence of

smooth muscle between crypts on biopsy can help differentiate solitary rectal ulcer syndrome from inflammatory bowel disease. (See "Clinical manifestations, diagnosis, and prognosis of ulcerative colitis in adults", section on 'Endoscopy and biopsy' and "Clinical manifestations, diagnosis, and prognosis of Crohn disease in adults", section on 'Endoscopy'.)

- **Ischemic colitis** Chronic ischemic colitis may manifest as colonic strictures, mucosal atrophy, or areas of granulation tissue. Biopsy of a post-ischemic stricture is marked by extensive transmural fibrosis that makes it difficult to distinguish from solitary rectal ulcer syndrome. However, patients with ischemic colitis generally have a history of underlying vascular disease or other risk factors (eg, aortic surgery, cardiopulmonary bypass, myocardial infarction, hemodialysis, dehydration, thrombophilia). The clinical presentation and diagnosis of ischemic colitis are discussed in detail, separately. (See "Colonic ischemia", section on 'Lower endoscopy'.)
- **Infectious proctitis** Amebiasis, lymphogranuloma venereum, herpes simplex virus, cytomegalovirus, and secondary syphilis may present with proctitis and rectal ulceration. However, on biopsy, features of solitary rectal ulcer syndrome are absent, and acute inflammation in the lamina propria with neutrophilic infiltration of crypts may be seen.

INITIAL MANAGEMENT

The management of solitary rectal ulcer syndrome depends upon the severity of symptoms and the presence and type of a concomitant rectal prolapse (algorithm 1) [24-26].

General measures in all patients — All patients with solitary rectal ulcer syndrome should first be treated conservatively [5,27,28]. Patients are educated about avoidance of straining or anal digitation, minimization of time on the commode, and consumption of a high-fiber diet and bulk laxatives. These dietary and behavioral modifications are especially effective in patients with mild to moderate symptoms and absence of significant mucosal prolapse.

In a retrospective study of 80 patients with solitary rectal ulcer, conservative therapy with bulk laxatives and bowel retraining was associated with a symptomatic improvement in 11 of 59 patients (19 percent) at a median follow-up of 25 months [5]. None of the 22 patients with rectal prolapse had an improvement in symptoms with conservative management. Persistent symptoms led to surgery in 27 (34 percent) patients.

Biofeedback — Patients whose symptoms are persistent despite general measures should receive biofeedback therapy [8,28-30]. Biofeedback reduces excessive straining associated with

defecation by correcting pelvic floor dyssynergia [30]. Biofeedback has been associated with symptomatic improvement in patients with solitary rectal ulcer syndrome in several small studies [8,28,29,31]. However, the durability of the benefit of biofeedback therapy is unclear, and repeat treatment may be needed. In one study that included 13 patients with solitary rectal ulcer syndrome, approximately one-half of the seven who initially responded to biofeedback relapsed within 36 months after training [29]. (See "Management of chronic constipation in adults".)

SUBSEQUENT MANAGEMENT FOR PERSISTENT SYMPTOMS

Surgery — Surgery is reserved for patients with severe symptoms that are unresponsive to initial management including biofeedback (algorithm 1) [32]. Options include excision of the ulcer, treatment of rectal prolapse or rectal intussusception, and defunctioning colostomy [33]. The choice is determined by the presence and type of a concomitant rectal prolapse or rectal intussusception [34]. (See 'Additional evaluation in selected patients' above.)

Patients with rectal prolapse or intussusception — For patients with a full-thickness rectal prolapse or rectal intussusception who can tolerate abdominal surgery, we suggest ventral rectopexy, rather than a perineal procedure. Ventral rectopexy can correct not only rectal prolapse, but also rectoceles and rectal intussusception [35-38]. (See "Surgical approach to rectal procedure)", section on 'Ventral mesh rectopexy'.)

For patients with a concomitant partial-thickness rectal prolapse, we suggest a perineal procedure such as mucosal resection (Delorme's procedure) or perineal proctectomy (Altemeier's procedure) [34]. Although perineal procedures resect the rectal ulcer while fixing the rectal prolapse, the recurrence rate of rectal prolapse is higher than after rectopexy. (See "Surgical approach to rectal procidentia (rectal prolapse)".)

Patients without rectal prolapse or intussusception — In patients without evidence of a rectal prolapse, local excision or suturing can provide symptomatic relief but with uncertain long-term benefit [39]. It is typically only performed for significant rectal bleeding.

Refractory symptoms — Patients with refractory or recurrent symptoms after surgery should be offered biofeedback again, because for many of them the symptoms are attributable to pelvic floor dyssynergia. Patients who fail to respond to biofeedback again may be offered salvage surgical treatment depending on the severity of symptoms. Mucosal sleeve resection with coloanal pull-through or fecal diversion is reserved for patients who have failed other surgical modalities [40]. Patients with intractable rectal pain and/or bleeding require proctectomy.

Other therapies with limited or unclear efficacy — There are insufficient data to support the use of other treatment options in solitary rectal ulcer syndrome. Treatment with sucralfate enemas, topical glucocorticoids, and mesalamine has been described in small series of patients [12,41-43]. Argon plasma coagulation has been used to control bleeding in patients with bleeding solitary rectal ulcers. A reduction in size and depth of ulcer was noted between sessions [44-48]. However, long-term follow-up data are lacking. Endoscopic mucosectomy was described in a case report of a patient with a polypoidal variant who had failed medical therapy and argon plasma coagulation [49].

PROGNOSIS

Solitary rectal ulcer syndrome is a chronic condition that significantly impacts quality of life. Many patients continue to have symptoms of anorectal dysfunction regardless of the treatment approach [50].

SOCIETY GUIDELINE LINKS

Links to society and government-sponsored guidelines from selected countries and regions around the world are provided separately. (See "Society guideline links: Solitary rectal ulcer syndrome".)

SUMMARY AND RECOMMENDATIONS

- Solitary rectal ulcer syndrome is a rare disorder with an estimated prevalence of 1 in 100,000 and a median age at diagnosis of 48 years. (See 'Epidemiology' above.)
- Patients with solitary rectal ulcer syndrome can present with rectal bleeding, straining during defecation, tenesmus, pelvic fullness, and passage of mucus. However, symptoms may be absent in up to 26 percent of patients. (See 'Clinical features' above.)
- Solitary rectal ulcer syndrome is a misnomer. Endoscopic findings in patients with solitary rectal ulcer syndrome can range from mucosal erythema alone to single or multiple ulcers and polypoid/mass lesions. In the majority of patients, the lesions are located in the anterior rectal wall within 10 cm of the anal verge. (See 'Endoscopy and histology' above.)

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- Solitary rectal ulcer syndrome is usually diagnosed incidentally during the course of evaluation of the patient's symptoms of rectal bleeding, tenesmus, or straining. The endoscopic appearance of solitary rectal ulcer syndrome, while supportive of the diagnosis, is not specific. The diagnosis is made on the basis of histologic finding of fibromuscular obliteration of the lamina propria. (See 'Evaluation' above.)
- In patients with solitary rectal ulcer syndrome, we perform an additional evaluation with defecography to diagnose associated anatomic and functional abnormalities such as rectal prolapse, rectal intussusception, rectocele, and pelvic floor dyssynergia, and to guide management. (See 'Additional evaluation in selected patients' above.)
- Initial management of patients with solitary rectal ulcer syndrome includes patient education, bulk laxatives, and a high-fiber diet. We suggest biofeedback in patients who remain symptomatic despite these measures (**Grade 2B**). (See 'Initial management' above.)
- We suggest surgery in patients with persistent severe symptoms that are unresponsive to initial management rather than continued conservative therapy (Grade 2C). The choice of procedure is determined by the presence and type of anatomic abnormalities. (See 'Surgery' above.):
 - For patients with a full-thickness rectal prolapse or rectal intussusception who can tolerate abdominal surgery, we perform ventral rectopexy, rather than a perineal procedure. Ventral rectopexy can correct not only rectal prolapse, but also rectoceles and rectal intussusception.
 - For patients with a partial-thickness rectal prolapse, we perform a perineal procedure such as mucosal resection (Delorme's procedure) or perineal proctectomy (Altemeier's procedure). Perineal procedures resect the rectal ulcer while fixing the rectal prolapse, but the recurrence rate of rectal prolapse is higher than after rectopexy.
 - For patients without a rectal prolapse or rectal intussusception, local excision or suturing is typically only performed for significant rectal bleeding.
 - Salvage surgical therapy may be offered for refractory or recurrent symptoms after initial surgery. Mucosal sleeve resection with coloanal pull-through or fecal diversion is reserved for patients who have failed other surgical modalities. Patients with intractable rectal pain and/or bleeding require proctectomy.
- Solitary rectal ulcer syndrome is a chronic condition that significantly impacts quality of life. Many patients continue to have symptoms of anorectal dysfunction regardless of the

treatment approach. (See 'Prognosis' above.)

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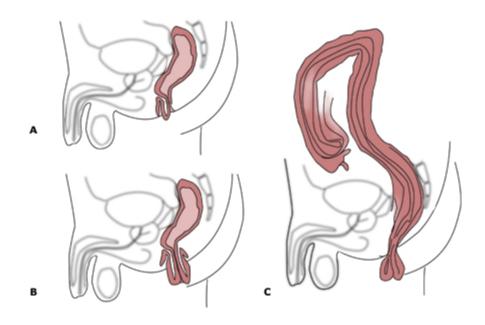
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GRAPHICS

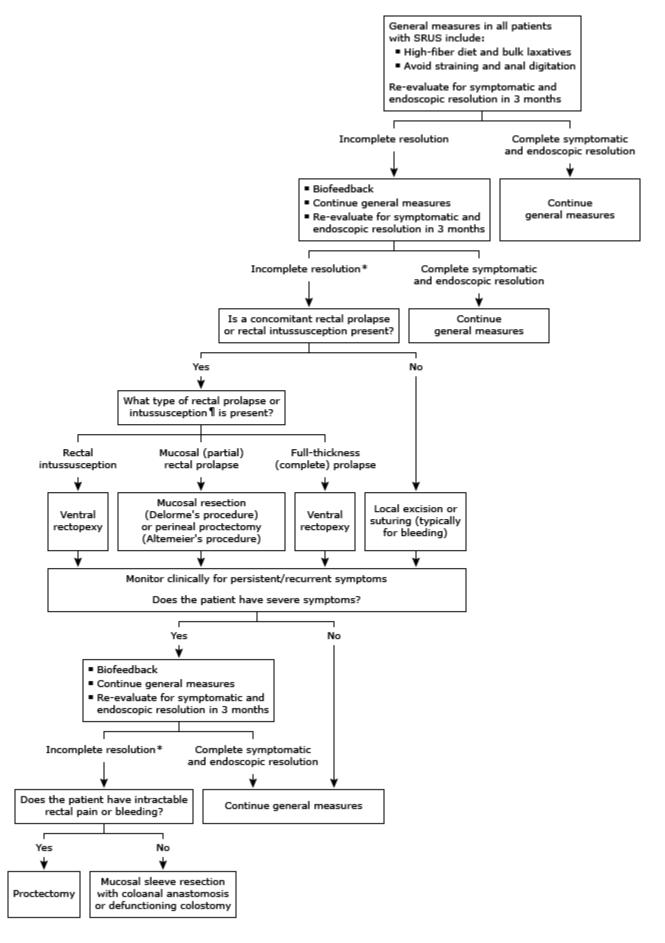
Anatomy of rectal prolapse



- (A) Partial prolapse.
- (B) Complete prolapse.
- (C) Prolapsed intussusception.

Graphic 65201 Version 2.0

Suggested approach for management of solitary rectal ulcer syndrome in adults



SRUS: solitary rectal ulcer syndrome.

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* In patients with incomplete resolution of symptoms, it is important to consider alternative diagnoses and consider the severity of symptoms before proceeding with additional interventions. Surgery is reserved for patients with severe symptoms that are unresponsive to initial management including biofeedback.

¶ A full-thickness (complete) rectal prolapse is the protrusion of all layers of the rectum through the anus, manifesting as concentric rings of rectal mucosa. A mucosal (partial) rectal prolapse is protrusion of only the rectal mucosa through the anus. If a prolapse cannot be reproduced on physical examination, we perform an additional evaluation to diagnose a rectal intussusception (eg, defecography). A rectal intussusception, also termed occult or internal rectal prolapse, is a "telescoping" of the bowel on itself internally, without protruding through the anal verge. Refer to UpToDate content on solitary rectal ulcer syndrome.

Graphic 122554 Version 1.0

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