Anorectal Dysfunction and Rectal Prolapse in Progressive Systemic Sclerosis

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Our aim was to characterize the clinical spectrum of anorectal dysfunction among eight patients with progressive systemic sclerosis (PSS) who presented with altered bowel movements with or without fecal incontinence. The anorectum was assessed by physical examination, proctosigmoidoscopy, and anorectal manometry. There was concomitant involvement of the other regions of the digestive tract in all patients as determined by barium studies, endoscopy, or manometry: eight esophageal, three gastric, four small bowel, and two colonic. Seven patients had fecal incontinence, and four also had second-degree complete rectal prolapse. Abnormal anorectal function, particularly abnormal anal sphincter resting pressures, were detected in all patients; anal sphincter pressures were lower in those with rectal prolapse. Rectal capacity and wall compliance were impaired in seven of seven patients. Successful surgical correction of prolapse in three patients resulted in restoration of incontinence for six months and seven years in two of the three patients. We conclude that rectal dysfunction and weakness of the anal sphincters are important factors contributing, respectively, to altered bowel movements and fecal incontinence in patients with gastrointestinal involvement by PSS. Rectal prolapse worsens anal sphincter dysfunction and should be sought routinely as it is a treatable factor aggravating fecal incontinence in patients with PSS. [Key words: Systemic sclerosis; Anorectal dysfunction; Prolapse; Incontinence; Manometry; Rectal compliance; Rectal capacity]

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The digestive tract is affected in about half the patients suffering from progressive systemic sclerosis (PSS). Although esophageal symptoms (dysphagia or heartburn) are most common, a number of patients present with altered bowel movements, abdominal distention, or pain. Classically, altered bowel movements are attributed to bacterial overgrowth in the small bowel. Over the last few years, we have seen patients who present with altered bowel movements in whom careful review identified an associated history of either fecal incontinence or rectal prolapse. The spectrum of anorectal dysfunction in patients with PSS is unclear. Hamel-Roy *et al.*[1] previously reported the lack of the rectoanal inhibitory reflex (RAIR) in patients with PSS and drew attention to the fact that esophageal motility was very often abnormal in such patients without a RAIR. Our aim was to characterize the clinical spectrum of anorectal dysfunction in patients with PSS who presented to us during a three-year period because of altered bowel movements and fecal incontinence.

MATERIALS AND METHODS

Patients

We identified eight patients, all of whom had undergone rectal examination, proctosigmoidoscopy, and anorectal manometry. Clinical evidence of rectal prolapse was sought during rectal examination or proctosigmoidoscopy. Severity of prolapse was graded clinically.[2] We reviewed the case records of these patients and determined whether other regions of the gastrointestinal tract were shown to be affected by barium studies, endoscopy, or manometric studies of the stomach and small bowel.[3]

Anorectal Function Testing

Using a standardized approach,[4] we measured the following parameters of anorectal function: rectal capacity; rectal compliance; anal canal length and the presence of the RAIR; resting and maximum squeeze pressures in the proximal, middle,

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and distal levels of the anal canal; ability to expel a 50-ml latex balloon placed in the rectum, with or without added weights suspended from the balloon; and perianal and rectal sensation (volume at first sensation; volume inducing urge to defecate).

Data and Statistical Analysis

These data were compared with published control data previously acquired in our laboratory using these methods. [4] We subsequently separated patients into two groups on the basis of the presence or absence of rectal prolapse. Anal sphincter pressures were compared between these two groups using Student's *t*-test. Since sphincter pressures were compared at three levels of the anal canal, we applied Bonferroni's correction and accepted $P < 0.017$ as significant.

RESULTS

Clinical Data

There were eight female patients with PSS, altered bowel habits, and fecal incontinence or prolapse. The mean age was 61 years, with a range of 49 to 74 years. Four patients had diarrhea, two had constipation, and two had diarrhea alternating with constipation. Seven patients had fecal incontinence; second-degree complete rectal prolapse (that is, prolapse externally visible on straining) was identified in four patients, all of whom complained of fecal incontinence. All patients had evidence that other regions were involved by PSS: eight esophageal, three gastric, four small bowel, and two colonic. These clinical data are summarized in Table 1.

Anorectal Function

Some of these data are summarized in Table 2. Abnormal anorectal function was present in all eight patients. Rectal capacity was 204 ± 28 ml (SEM), compared with the norm of >400 ml; similarly, rectal compliance was markedly reduced (5.7 ± 1.1 ml/mm Hg) compared with the norm, which is >17 ml/mm Hg. The anal canal length was normal (data not shown). RAIR was absent in

Table 1.
Clinical Features of Patients with Scleroderma

Case No.	Age (yr)	Dysphagia/ Heartburn	Nausea/ Vomiting	Diarrhea	Constipation	Fecal Incontinence	Rectal Prolapse	Manometry*	Endoscopy	X-Rays
1	73	+	−	+	+	+	+	E, St, SB	−	SB, C dilated, pneumoperitoneum
2	69	+	−	+	+	+	+	−	E stricture	Previous partial colectomy
3	51	+	−	−	+	+	+	−	−	E, SB dilated
4	49	+	−	+	−	+	+	−	Esophagitis	E dilated
5	49	+	−	+	−	+	−	E, St, SB	Esophagitis	−
6	66	+	+	+	−	+	−	E, St, SB	Esophagitis	−
7	57	+	+	−	+	−	−	E, LESP	E dilated	−
8	74	+	−	+	−	+	−	E, LESP	E dilated	−

+ = yes; − = no; E = esophagus; St = stomach; SB = small bowel, C = colon; LESP = lower esophageal sphincter pressure.
* Manometry shows low amplitude contractions.

Table 2.
Anorectal Manometry Results in Eight Patients with Progressive Systemic Sclerosis

	n	Resting Pressures (mmHg)			Squeeze Pressures (mmHg)			Rectal Capacity (ml)	Rectal Compliance (ml/mmHg)
		Proximal	Middle	Distal	Proximal	Middle	Distal		
Lab control range		40–60	50–80	30–50	115–150	100–150	80–120	≥400	≥17
PSS with rectal prolapse	4	8 ± 0.3	10 ± 1	16 ± 2*	24 ± 6	29 ± 9*	34 ± 11	173 ± 35	5.2 ± 1.2
PSS without rectal prolapse	4	15 ± 35	38 ± 11	46 ± 8	48 ± 8	87 ± 13	99 ± 24	247 ± 36	6.3 ± 2.3

All data are expressed as mean ± SEM.
* $P < 0.017$ (applying Bonferroni's correction) compared with PSS patients without rectal prolapse.

four of eight patients. Three of these four patients without a RAIR had rectal prolapse. Anal sphincter resting pressures were lower than the control range: at the proximal level in eight patients, at the middle level in six patients, and at the distal level in four patients. In contrast, maximal squeeze pressures were abnormal at the proximal level of the sphincter in eight patients, at the middle level in seven patients, and at the distal level in six patients. Resting pressures at the distal level and maximum squeeze pressures at the middle level of the sphincter were significantly ($P < 0.017$) lower in patients with rectal prolapse. Three patients with rectal prolapse underwent surgical correction, which relieved the prolapse in all. The operations performed were transanal excision of prolapsed mucosa in one patient and anterior resection of the sigmoid and portion of the rectum in two patients. Improvement in continence lasted one week, six months, and seven years in these three patients. Recurrent incontinence in the three operated-on patients was unassociated with prolapse.

DISCUSSION

This study documents the spectrum of anorectal dysfunction among a subgroup of patients with PSS who present with altered bowel movements and fecal incontinence. Half of our patients did not have a RAIR as described previously in the literature.[1,4] Other not previously reported disturbances of anorectal function were frequently observed, including reduced rectal capacity and compliance, anal sphincteric weakness (particularly at rest), and rectal prolapse. Rectal prolapse may result in reduced capacity and compliance of the rectum and probably contributes to the recording of low anal canal pressures as the sphincter is kept open by the prolapsing mucosa. However, rectal capacity and compliance were also abnormal in PSS patients without rectal prolapse, suggesting that anorectal dysfunction is not exclusively due to prolapse but results from the effects of PSS on the rectal wall and internal anal sphincter. Weakness of the submucosa and chronic straining contribute to the development of rectal prolapse. Squeeze pressures in the anal canal were lower in all patients (with or without rectal prolapse) than in the control group. Rectal prolapse itself may, in fact, impair the ability of the manometric probe to measure anal canal pressure. In the group with rectal prolapse, it is unclear from our study whether external anal sphincter function was impaired, and further studies of squeeze pressures after relief of prolapse are necessary. However, among those PSS patients without rectal prolapse, squeeze pressures in the proximal part of the canal were low and may have contributed to the fecal incontinence in our patients. In one previous study in patients with PSS, only 1 of 13 had incontinence and anal canal squeeze pressures were normal.[1] In contrast, Chiou *et al.*[5] found normal squeeze pressures in their 17 patients with PSS, nine of whom were incontinent. The anal canal squeeze pressure data in the patients reported here are in complete agreement with the preliminary report of Basilisco *et al.*,[6] who found reduced squeeze pressures in the orad part of the anal canal of patients with PSS and constipation.

Anorectal dysfunction is invariably associated with involvement of other parts of the digestive tract by PSS. In this series of patients, the spectrum of involved organs is similar to that in previous reports.[7-9] Abnormal rectal compliance and capacity probably reflect the infiltrative process affecting the muscularis propria of the rectum[10]; Whitehead *et al.*[11] have also reported on abnormal rectosigmoid motility reflecting rectal neuromuscular dysfunction in PSS patients.

Our study suggests that there is temporary improvement of continence after surgical treatment of rectal prolapse; full continence lasted six months in one patient and seven years in another. Recurrent incontinence may reflect progression of anal sphincter dysfunction over time. Relief of rectal prolapse can be achieved relatively safely by anterior resection of portions of the rectum and sigmoid or, alternatively, transanally. Even if incontinence recurs, the quality of life of individual patients can be significantly enhanced by such treatment since prolapse is relieved and continence may be restored for months or years. We believe that rectal prolapse should be sought in all patients with PSS who are experiencing altered bowel movements or fecal incontinence, particularly as it is a potentially treatable cofactor of their anorectal symptoms.

In summary, a wide spectrum of motor and sensory dysfunctions occurs in the anorectum of patients with PSS, altered bowel movements, and incontinence. Our study suggests that rectal mucosal prolapse may be a significant complication, particularly since it may be associated with external anal sphincter dysfunction, thereby further aggra-

vating fecal incontinence. Prolapse can be easily sought during anorectal examination and proctoscopy, and its correction may significantly alter the patient's quality of life.

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REFERENCES

1. Hamel-Roy J, Devroede G, Arhan P, *et al.* Comparative esophageal and anorectal motility in scleroderma. Gastroenterology 1985;88:1–7.
2. Nivatvongs S. Anorectal diseases. In: Phillips SF, Pemberton JH, Shorter RG, eds. The large intestine: physiology, pathophysiology and disease. New York: Raven Press, 1991:775–831.
3. Malagelada J-R, Camilleri M, Stanghellini V. Manometric diagnosis of gastrointestinal motility disorders. New York: Thieme Medical Publishers, 1986.
4. Pemberton JH. The clinical value of anorectal motility. Surg Annu 1990;22:185–214.
5. Chiou AW-H, Lin J-K, Wang F-M. Anorectal abnormalities in progressive systemic sclerosis. Dis Colon Rectum 1989;32:417–21.
6. Basilisco G, Barbera R, Vanoli M, Bianchi P. Delayed colonic transit in constipated patients with progressive systemic sclerosis (PSS) (abstract). Gastroenterology 1992;102:A421.
7. Cohen S, Lanfer I, Snape WJ, Shiau YF, Lavine GM, Jimenez S. The gastrointestinal manifestations of scleroderma: pathogenesis and management. Gastroenterology 1980;79:155–66.
8. Poirier TJ, Rankin GM. Gastrointestinal manifestations of progressive systemic sclerosis based on a review of 364 cases. Am J Gastroenterol 1972;58: 30–44.
9. Greydanus MP, Camilleri M. Abnormal postcibal antral and small bowel motility due to neuropathy or myopathy in systemic sclerosis. Gastroenterology 1989;96:110–5.
10. D'Angelo WA, Fries JF, Masi AT, Shulman LE. Pathologic observations in systemic sclerosis (scleroderma). A study of fifty-eight autopsy cases and fifty-eight matched controls. Am J Med 1969;46:428–40.
11. Whitehead WE, Taitelbaum G, Wigley FM, Schuster MM. Rectosigmoid motility and myoelectric activity in progressive systemic sclerosis. Gastroenterology 1989;96:428–32.