

# The pattern of esophageal manometry in progressive systemic sclerosis

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## ABSTRACT

**Objectives:** The purpose of this paper is to study the esophageal motility pattern and the frequency of acid reflux in patients diagnosed to have progressive systemic sclerosis and compare the results to that of normal controls.

**Methods:** All consecutive patients diagnosed to have progressive systemic sclerosis between 1417-1419 (Hijra year) at King Khalid University Hospital, Riyadh, Kingdom of Saudi Arabia were included (Group I). History of heartburn, dysphagia and regurgitation was reviewed. Drugs that might suppress acid or alter motility were discontinued 2 weeks before inclusion. The results were compared to that of 21 symptomatic controls of similar age and sexes that were seen at the same period (Group II). Esophageal manometry and upper gastrointestinal endoscopy were performed in all patients. Ambulatory 24 hour-pH monitoring was carried out in 6 patients of group I and 20 patients of group II.

**Results:** Thirteen progressive systemic sclerosis patients (12

females) mean age was 38.7 years and 21 (19 females) mean age was 34.8 years were included. The symptom scores, lower esophageal sphincter pressure, esophageal contractions amplitude were significantly worse in patients compared to control, dysphagia was mostly due to aperistalsis. All progressive systemic sclerosis patients showed the typical esophageal manometry pattern of lower esophageal sphincter pressure and diminished amplitude with aperistalsis. Gastroesophageal reflux was detected in 83% of patients with progressive systemic sclerosis. Moreover, all 24-hour pH monitoring variables were significantly worse in group I.

**Conclusion:** Patients with progressive systemic sclerosis usually present with heartburn, dysphagia and regurgitation. Esophageal manometry typically shows lower pressure and aperistalsis. Gastroesophageal reflux is frequent.

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**P**rogressive systemic sclerosis (PSS) is a connective tissue disorder of unknown etiology.<sup>1</sup> Several organs may be affected by the disease. Gastrointestinal involvement occurs in approximately 90% of patients.<sup>1,2</sup> The esophagus is the most frequently affected part of the gastrointestinal tract.<sup>3</sup> Esophageal smooth muscle becomes atrophied and replaced by fibrous tissue leading to severe motility disturbance of distal esophagus.<sup>4,5</sup> Esophageal motility disturbance classically manifest as a reduced lower esophageal sphincter pressure (LESP) and loss of distal esophageal body peristalsis.<sup>6,7</sup> Consequently, PSS patients with esophageal involvement have impaired acid clearance which may be

complicated by erosive esophagitis and eventually by Barrett's esophagus.<sup>4,8</sup> The aim of this paper is to study the esophageal motility pattern and the frequency of acid reflux in patients diagnosed to have PSS and compare the results to that of normal control.

**Methods.** Thirteen patients diagnosed to have PSS were referred to the Gastroenterology Division at King Khalid University Hospital (KKUH) between 1417 and 1419 (Hijra year). Progressive systemic sclerosis diagnosis was based on the criteria of the American Rheumatism Association.<sup>9</sup> All patients were interviewed

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and history of heartburn, regurgitation and dysphagia was noted, drugs used were reviewed. All patient underwent upper gastrointestinal endoscopy and esophageal manometry. Twenty-four hour pH monitoring was performed in 6 patients (group I). Drugs known to suppress acid or alter esophageal motility were discontinued 2 weeks before inclusion. Endoscopic esophagitis was graded according to Savary-Miller classification.<sup>10</sup> Briefly, the endoscopic findings are divided into 4 stages: 1. One or more non-confluent mucosal lesion accompanied by erythema; 2. Confluent erosive exudative lesions not covering the entire circumference; 3. Erosive and exudative lesions covering the entire circumference, 4. Chronic mucosa lesions (ulcers, stricture). Their results were compared to those of 21 symptomatic control but negative upper gastrointestinal endoscopy, esophageal manometry and 24 hour pH studies. The control patients were seen during the same period (group II). The reasons for referral to endoscopy unit were heartburn with negative endoscopy, feeling of foreign body sensation in the throat, hoarseness in the voice and regurgitation. Exclusion criteria included presence of 1. Diabetes mellitus; 2. Other connective tissue disorders; 3. Concomitant use of acid suppressing drugs or drugs that alter gastrointestinal (GI) motility.

**Esophageal motility study.** The study was performed in fasted patients in the supine position using 8-lumen polyvinyl catheter (outer diameter is 4.5 mm; internal diameter is 0.8 mm; esophageal synetics medicals 3R, Arndorfer Medical specialties). The distal 4 openings were spaced at 5 cm distant at 90° angle. The catheter was connected to external pressure transducers (Novadome MX 860 medex Rossenda, United Kingdom). The catheter was continuously perfused with distilled water at a rate of 0.5 ml/min by a low-compliance pneumohydraulic capillary infusion system (Arndorfer Medical specialties). The catheter assembly was passed through the nose until all recording orifices were in the stomach. The station pull-through of the lower esophageal sphincter (LES) was performed at one cm intervals. The LES pressure recorded for each patient represented the calculated average of four individual pressures (distal ports), measured at end-expiratory variation to the mean gastric baseline pressure. Esophageal body recording were performed by positioning the 4 proximal ports 2, 7, 12 and 17 cm above the LES. At least, 10 wet swallows (10 ml water each) were administered; each separated by 30 seconds period. The amplitude of pressure wave was measured from the mean intraesophageal baseline pressure to the peak of the wave.

**Ambulatory 24-hour pH monitoring.** In an overnight fasting patient, an antimony pH probe with an outer diameter of 2.1 mm, was positioned 5 cm above the upper border of the LES and a reference electrode was attached to the anterior chest wall. Both electrodes were connected to a recording device (Synectics Medical, Inc. Irving, Texas, United States of America).

Alkaline and acidity electrodes were calibrated using buffers of pH 1 and 7. Patients were instructed to keep records of their upright and supine positions. The pH tracings were analyzed by a commercial computer software program and reviewed by the author. Reflux disease was considered pathological if any of the following criteria were exceeded: 1. Percentage of total time with pH <4 (normal <5.5%); 2. Percentage upright with pH <4 (normal <8.2%); 3. Percentage supine with pH <4 (normal <3%).<sup>7</sup>

Symptoms of dysphagia, regurgitation and heartburn were scored according to their frequency on a scale of 0-3. The highest obtainable score was 9 (Table 1). The 2 groups were compared using Stat Gold Package and Epistate Package. Values are expressed as mean ± SD. P value of <0.05 was considered statistically significant.

**Results.** The study included 2 groups, Group I consisted of 12 females and one male. Mean age was 38.7 (14.5) years. All patients were symptomatic. Ninety-two percent of patients complained of heartburn of variable severity, 85% had dysphagia and 77% regurgitation. Distal esophageal aperistalsis was present in 12 patients (92.3%). The remaining patient showed low amplitude contractions, which were propagating, normally in 50% of wet swallows. Group II consisted of 19 females and 2 males. Mean age was 34.8 (8.8) years. They presented with heartburn in 76%, mild dysphagia in 52%, regurgitation in 24%, hoarseness of voice in 19% and foreign body sensation in the throat in 19%. Esophageal manometry and upper GI endoscopy were normal in all control. Twenty-four hours pH monitoring was performed in 20 patients and in all instances, the study was normal. There was no significant difference between the groups regarding age and sex distribution. However, Group II patients were significantly more symptomatic; P values for dysphagia regurgitation and heartburn were 0.01, 0.004 and 0.04. Endoscopically, grade I esophagitis was found in 3 patients, grade II in 7 patients, grades III and IV in one patient each. Normal gastrointestinal endoscopy was reported in one patient; mean reflux esophagitis score was 2.8. Esophageal manometry revealed a significantly lower LES pressure, proximal, middle as well and distal esophageal amplitude; P values were 0.001, 0.02, 0.001 and 0.0001. Ambulatory esophageal pH monitoring were performed in 6 patients and 20 controls. Similarly, all mean pH values were significantly worse in patient compared to control (Table 2).

**Discussion.** Esophageal involvement by progressive systemic sclerosis is characterized manometrically by reduced LES pressure and loss of distal esophageal peristalsis.<sup>6,7</sup> As consequences of this involvement, patients usually manifest with heartburn, dysphagia and regurgitation.<sup>10</sup> Heartburn and regurgitation are due to reflux of gastric juice across an incompetent LES,<sup>11,12</sup> whereas dysphagia may result

**Table 1** - Symptom score.

Symptom	Score
Dysphagia	0 No dysphagia 1 Dysphagia to solids 2 Dysphagia to semisolid 3 Dysphagia to liquid
Regurgitation	0 No regurgitation 1 Occasional on straining or lying down 2 Predictable on positioning or straining 3 Occurrence of pulmonary aspiration
Heartburn	0 None 1 Minimal 3 Moderate - reason for medical visit 3 Severe - Constant interference with activities

**Table 2** - Age, sex, symptomatic score, manometric and pH findings in patients and control.

Variable	Group I (N=13)	Group II (N=21)	P value
Age	38.69 ± 14.54	34.81 ± 8.8	p=NS
Sex (F/M)	12/1	19/2	0.66
Dysphagia score	1.076 ± 0.95	0.52 ± 0.51	P=0.017
Regurgitation score	1.076 ± 1.115	0.2857 ± 0.56	P=0.0048
Heartburn score	1.846 ± 1.14	1.238 ± 0.83	P=0.041
LES	7.538 ± 3.79	15.14 ± 0.761	P=0.011
Proximal amplitude/ mm Hg	33.38 ± 20.99	47.619 ± 18.74	P=0.024
Middle amplitude/ mm Hg	6.769 ± 2.166	60.23 ± 24.31	P<0.001
Distal amplitude/ mm Hg	5.0 ± 0.0	66.57 ± 21.03	P<0.0001
Endoscopy score	2.889 ± 0.01	0	P<0.0001
% pH <4	13.33 ± 10.94	1.03 ± 0.98	P<0.001
Reflux >5minute	7.5 ± 6.94	0.35 ± 0.587	P<0.001
Eosophageal clearance/minute	3.18 (2.2)	1.04 (0.78)	0.0006
% reflux in supine	18.38 ± 18.66	0.43 ± 0.576	P<0.001
% reflux in upright	11.4 ± 9.82	1.39 ± 1.73	P<0.001
N of reflux	38.167 ± 28.29	12.3 ± 10.8	P=0.001
Time pH <4	184.67 ± 150.9	14.2 ± 12.6	P<0.001
Longest episode of reflux/min	47.0 ± 29.05	5.55 ± 7.85	P<0.001
LES - lower esophageal sphinter NS - not significant, F - female, M - male			

from esophageal peptic stricture or disturbed esophageal peristalsis.<sup>12</sup> Upper gastrointestinal endoscopy and esophageal manometry can differentiate between these possibilities.

In our experience, almost all patients suffered from heartburn, regurgitation or dysphagia, either separately or in combination. Moreover, disturbed distal esophageal peristalsis was the cause of dysphagia in 92% of our patients. Our study clearly demonstrated a significantly reduced LES pressure and absent or diminished esophageal peristalsis in patients as compared to control; P value 0.001 and 0.0001. Furthermore, calculated esophageal clearance in group I was markedly prolonged compared to control P=0.0006.

The frequency of acid exposure and the esophageal acid clearance time are the 2 factors that influence esophageal acid exposure time.<sup>13,14</sup> Adequate esophageal body peristaltic waves are a critical determinant of esophageal acid clearance, whereas LES function is an important barrier of acid reflux.<sup>13,14</sup> Progressive systemic sclerosis patients are deficient in both mechanisms as demonstrated by diminished LES pressure, peristaltic amplitude and esophageal clearance compared to control P=0.001, 0.0001 and 0.0006. Abnormal distal esophageal acid exposure was documented in 5 of the 5 PSS patients tested. Similar results have been reported by others.<sup>1,15</sup> It has been proposed that loss of distal esophageal peristalsis is the critical factor in predicting the presence of erosive esophagitis in PSS patient.<sup>1,15</sup> To evaluate the value of LES pressure or poor clearance from decreased peristalsis in the distal esophagus, Murphy et al<sup>6</sup> compared 2 groups of patients, the first (N=7) were PSS patients and the 2nd group of patients (N=9) with similar endoscopic findings but not suffering from PSS. All patients underwent pH monitoring and scintigraphy; 4 PSS patients and all the non-PSS patients had simultaneous manometry. Their results showed that PSS patients had significantly fewer episodes but the reflux events had significantly longer duration. Indicating the more important role of decreased distal esophageal peristalsis in the pathogenesis of gastroesophageal reflux in PSS patients. Majority of PSS patients with defective LES and impaired body peristalsis has abnormal gastroesophageal reflux, which is more evident when recumbent.<sup>16</sup> This compares favorably with our results, as 83% of group I patients had abnormal gastroesophageal reflux. Furthermore, percent reflux in supine position was higher than upright posture.

In conclusion, this paper describes the esophageal manometry and 24 hour pH monitoring in PSS patients and symptomatic control. Majority of our PSS patients were symptomatic. The manometry typically showed low LES pressure and reduced contraction amplitude. Upper GI endoscopy and pH monitoring were pathological in most PSS patients. Dysphagia and abnormal gastroesophageal reflux were mainly due to distal esophageal aperistalsis.

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#### Abstract

Oesophageal motility disorders are recognized causes of oesophageal symptoms. Although adults are more commonly affected, reports indicate that all types of motility disorders may occur in children. In this article, a comprehensive review is presented in order to update physicians on recent developments in the diagnosis and treatment of oesophageal motor dysfunction.