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# Frequency of motor alterations detected through manometry in patients with esophageal symptoms and scleroderma<sup>☆</sup>



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### KEYWORDS

Scleroderma;  
Esophagus;  
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manometry;  
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### Abstract

**Background:** Scleroderma can present with esophageal involvement causing important morbidity.

**Aims:** To describe the manometric findings and clinical characteristics of patients with scleroderma and esophageal symptoms.

**Materials and methods:** Patients with scleroderma and esophageal symptoms were evaluated through esophageal manometry within the time frame of one year. Descriptive statistics were carried out and the continuous variables were expressed as means and standard deviation. Frequencies were expressed as percentages.

**Results:** The study included 24 female patients with a mean age of 53.5 years and mean disease progression of 7.84 years. The most frequent findings were short and hypotonic lower esophageal sphincter (mean length 1.58 cm and mean tone 9.49 mmHg) and ineffective esophageal motility (mean non-transmitted waves 92.91%, mean effective primary peristalsis 40.05%, and mean amplitude 13.11 mmHg). The most frequent symptom was dysphagia.

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**PALABRAS CLAVE**

Esclerodermia;  
 Esófago;  
 Esfínter esofágico inferior;  
 Manometría esofágica;  
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**Conclusions:** Scleroderma is associated with lower esophageal sphincter alterations and symptomatic ineffective esophageal motility.

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## Frecuencia de alteraciones motoras detectadas por manometría en pacientes con síntomas esofágicos y esclerodermia

**Resumen**

**Antecedentes:** La esclerodermia puede presentar compromiso esofágico, lo cual es causa de importante morbilidad.

**Objetivos:** Describir los hallazgos manométricos de pacientes con esclerodermia y síntomas esofágicos así como características clínicas.

**Materiales y métodos:** Pacientes con esclerodermia y síntomas esofágicos sometidos a manometría esofágica en un año. Se realizó estadística descriptiva, las variables continuas se expresaron como promedios y desviación estándar. Las frecuencias se expresaron como porcentajes.

**Resultados:** Se incluyó a 24 pacientes, 100% mujeres, con edad promedio de 53.5 años y un promedio de evolución de 7.84 años. Los hallazgos más frecuentes fueron esfínter esofágico inferior (EEI) corto e hipotónico (longitud promedio 1.58 cm y tono promedio 9.49 mmHg) y motilidad esofágica inefectiva (MEI) (ondas no transmitidas promedio 92.91%, peristalsis primaria efectiva promedio 40.05% y amplitud promedio 13.11 mmHg). El síntoma más frecuente fue disfagia.

**Conclusiones:** La esclerodermia se asocia con alteraciones del EEI y presencia de MEI sintomática.

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**Introduction**

Scleroderma is an autoimmune, multisystemic disease of the connective tissue characterized by inflammation, progressive fibrosis, and vascular alterations. It affects females more frequently and its annual incidence is 20 new cases per one million adults. The esophagus is one of the most compromised internal target organs. Esophageal involvement is defined as the presence of symptoms of esophageal origin in patients diagnosed with scleroderma.<sup>1-2</sup>

**Materials and methods**

The study included patients diagnosed with scleroderma that were referred by the Rheumatology Service to the Motility Laboratory to undergo esophageal manometry due to the presence of esophageal symptoms, within the time frame of June 2013 to June 2014. The patients had a panendoscopy study with no anatomic alterations or erosive reflux disease, in accordance with the Los Angeles endoscopic classification.

After signing statements of informed consent, all the patients underwent conventional esophageal manometry with Polygraf ID™ Model A500 equipment from Given Imaging. A solid-state Alpine Biomed catheter was used that consisted of 4 pressure sensors located 0, 5, 10, and 15 cm

from the tip of the probe, and analyzed with the Alpine Biomed GastroTrac™, version 4.4a.

Normal values were based on those described by Castell and Richter<sup>3</sup> for conventional solid-state esophageal manometry.

Ineffective esophageal motility (IEM) was defined as the presence of 50% or more non-transmitted waves.

A complete anamnesis was carried out for all the patients with an emphasis on esophageal symptoms and their characteristics.

The variables analyzed included: demographic characteristics of the population (distribution by sex and age), time of disease progression, reported symptoms, manometric characteristics of peristalsis (amplitude and velocity of progression), manometric characteristics of the lower esophageal sphincter (LES) (length, basal pressure, relaxation), and manometric characteristics of the upper esophageal sphincter (UES) (length, basal pressure, relaxation, peak pharyngeal pressure).

Descriptive statistics were carried out and the continuous variables were expressed as means and standard deviation. Frequencies were expressed as percentages.

**Results**

A total of 24 female patients diagnosed with scleroderma were included in the study. Their mean age was

**Table 1** Values of the manometric findings in the study group.

	EPP	NT	Wave amplitude	LES length	LES resting tone
MEAN	4.58 mmHg	92.91%	13.11 mmHg	1.58 cm	9.49 mmHg
Range	0-100 mmHg	0-100%	2.4-63.1 mmHg	1-3 cm	2.56-23.36 mmHg
SD	20.42	21.56	13.27	0.717	4.63

EPP: effective primary peristalsis; NT: non-transmitted waves; LES: lower esophageal sphincter; SD: standard deviation.

53.5 ± 11.7842 (range: 17-70 years). The most relevant manometric findings in the LES were: short length and hypotonia. The findings in the esophageal body were: poor effective primary peristalsis, high percentage of non-transmitted waves, and mean diminished wave amplitude (Table 1). Hypotonia and incomplete relaxation were found in the UES. There was reduced peak contraction in the pharynx.

The most frequently reported symptoms were: esophageal dysphagia (15 patients, 62.5%), heartburn (8 patients, 33.3%), and regurgitation (7 patients, 29.1%).

Mean disease progression time was 7.84 ± 6.73 (0.33-20) years.

## Discussion

The relation between the presence of esophageal motor disorders and scleroderma is well known. Esophageal alterations are found in 50 to 90% of the patients with scleroderma and the esophagus is the internal organ that is the most affected.<sup>4</sup>

Marked atrophy of the esophageal smooth muscle, including the LES, is observed in these patients. The resulting distal aperistalsis and LES incompetence can predispose to gastroesophageal reflux disease, as well as its complications.<sup>5</sup> The typical symptoms are heartburn and dysphagia.<sup>2</sup>

Previous studies have documented the presence of LES alterations, such as hypotonic sphincter in 39% of the patients, esophageal body hypomotility in 82%, and both alterations in 36%.<sup>6</sup> These alterations are more severe in patients with a longer time of disease progression and have been described in up to 16% of patients with scleroderma that do not complain of esophageal symptoms.<sup>7</sup> In our study, the most frequently found manometric alterations were hypotonic LES and symptomatic IEM (the most frequent symptom was esophageal dysphagia), and a trend to present with more severe motor alterations was observed in the patients with longer disease progression time.

The presence of patients with oropharyngeal dysphagia was striking, given that we did not find the description of UES alterations in scleroderma in other studies.

## Ethical disclosure

**Protection of human and animal subjects.** The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and with those of the Code of Ethics of the World Medical Association (Declaration of Helsinki).

**Confidentiality of data.** The authors declare that no patient data appear in this article.

**Right to privacy and informed consent.** The authors declare that no patient data appear in this article.

## Conflict of interest

The authors declare that there is no conflict of interest.

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