

Digestive System (Gut) Involvement in Scleroderma

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Digestive System Involvement

- **Esophagus-Food Pipe**
- **Stomach**
- **Small intestine**
- **Large intestine (Colon) and
rectum**
- **Liver**

Gut Involvement

- Esophagus
- Stomach
- Small intestine
- Large intestine and rectum
- Liver

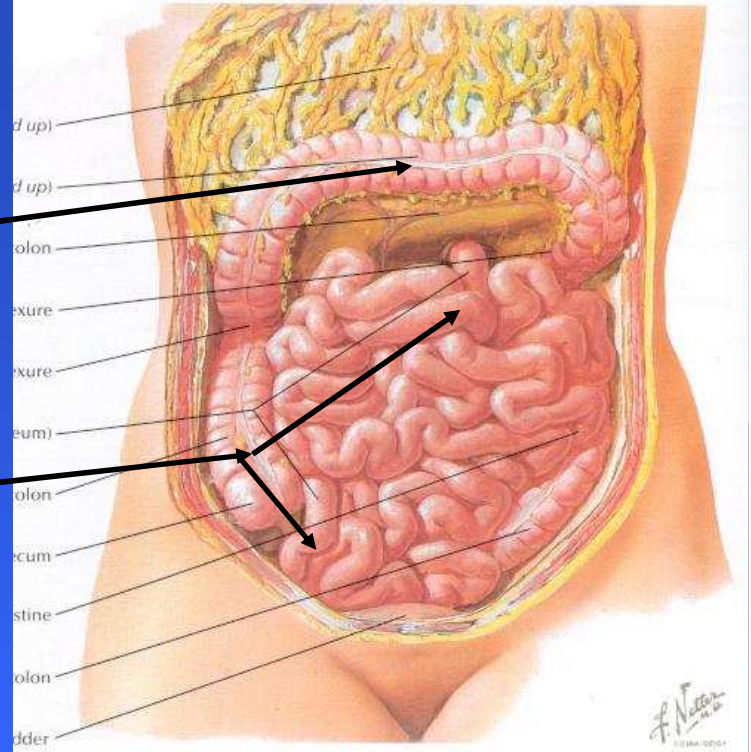
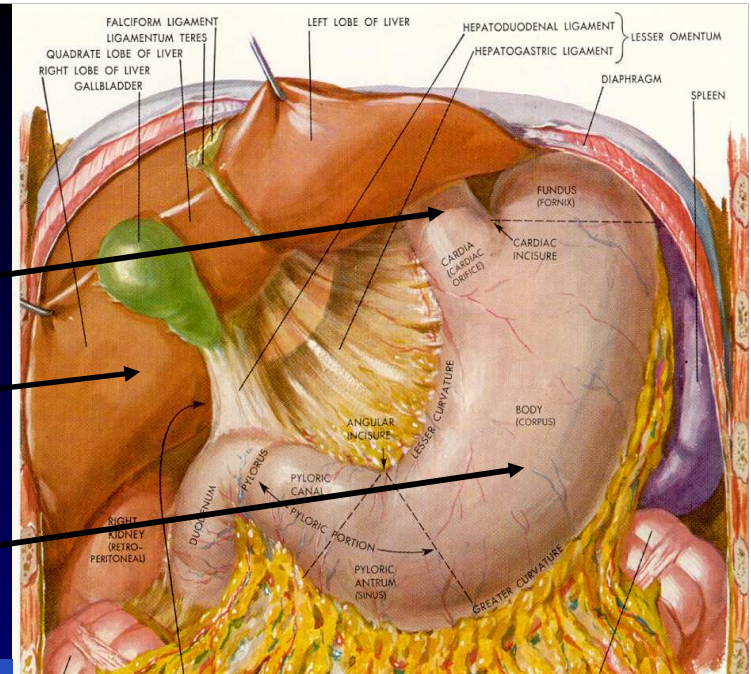
Esophagus

Liver

Stomach

Large intestine

Small intestine



Frequency of GI symptoms (209 SSc patients)

<u>Symptom</u>	<u>Prevalence (%)</u>
• Heartburn	71
• Trouble swallowing	52
• Bloating	80
• Diarrhea	51
• Constipation	51
• Fecal Incontinence	35

Scleroderma Foundation survey 2010

Function of the Gut

- To push the food down, extract and absorb nutrients!!
- Does it by well orchestrated motion of the gut **MUSCLE.**
- Also known as **PERISTALSIS.**

Gut Involvement

- The primary event that causes trouble in the gut is progressive weakening of muscle motion in the gut.
- Virtually every gut symptom is the result of weakening of the gut muscle.
- The weakening that starts in the esophagus and stomach, works its way down the small and large intestine.

Oropharyngeal Involvement

- Facial involvement interfere with mastication
- 20% Sjogren's syndrome (dry eyes/ dry mouth)

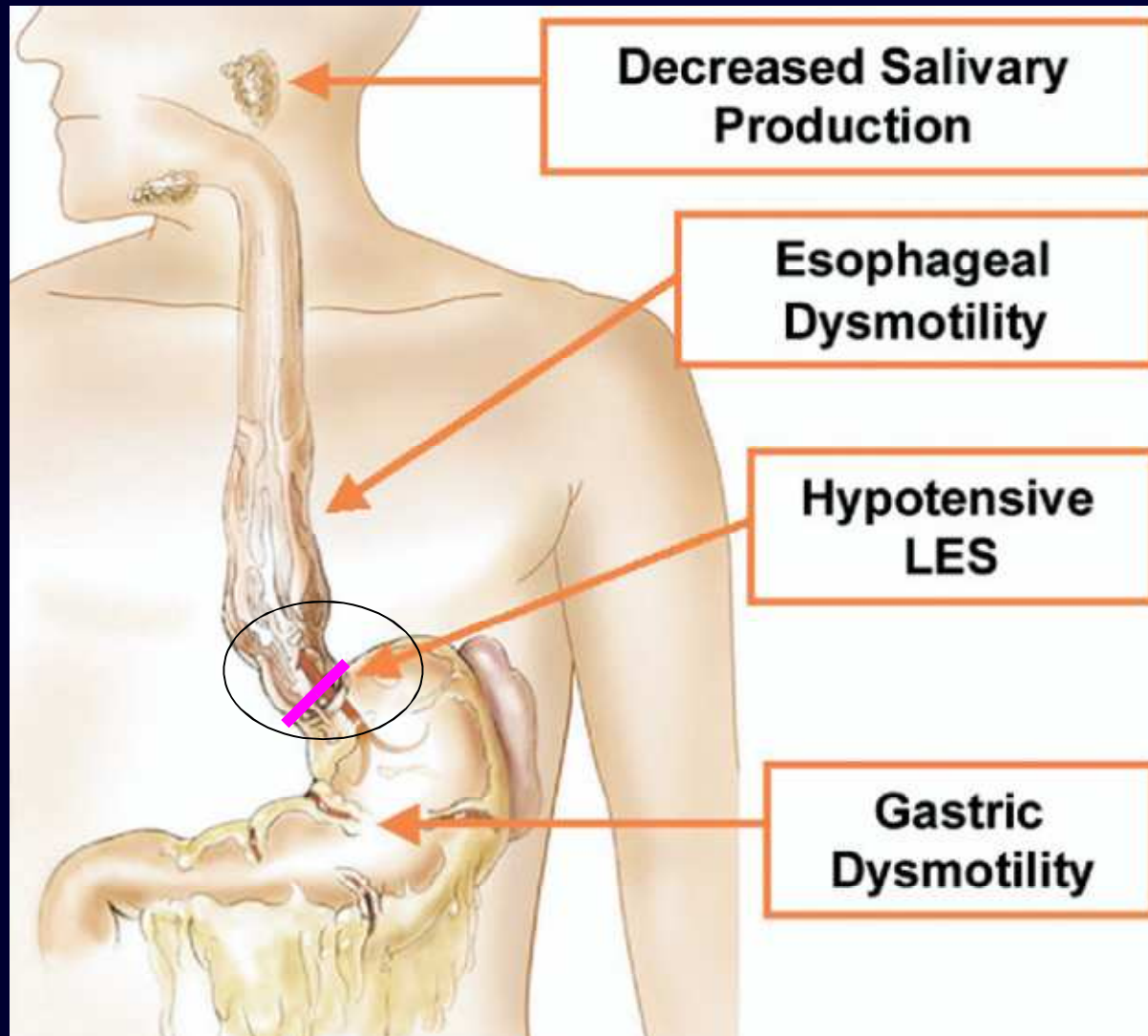
TREATMENT

- Small bolus
- Liberal fluid intake
- Sugar free gum and mouthwash (Biotene®)
- Regular dentist appointment
- Trial of pilocarpine and cevimeline therapy

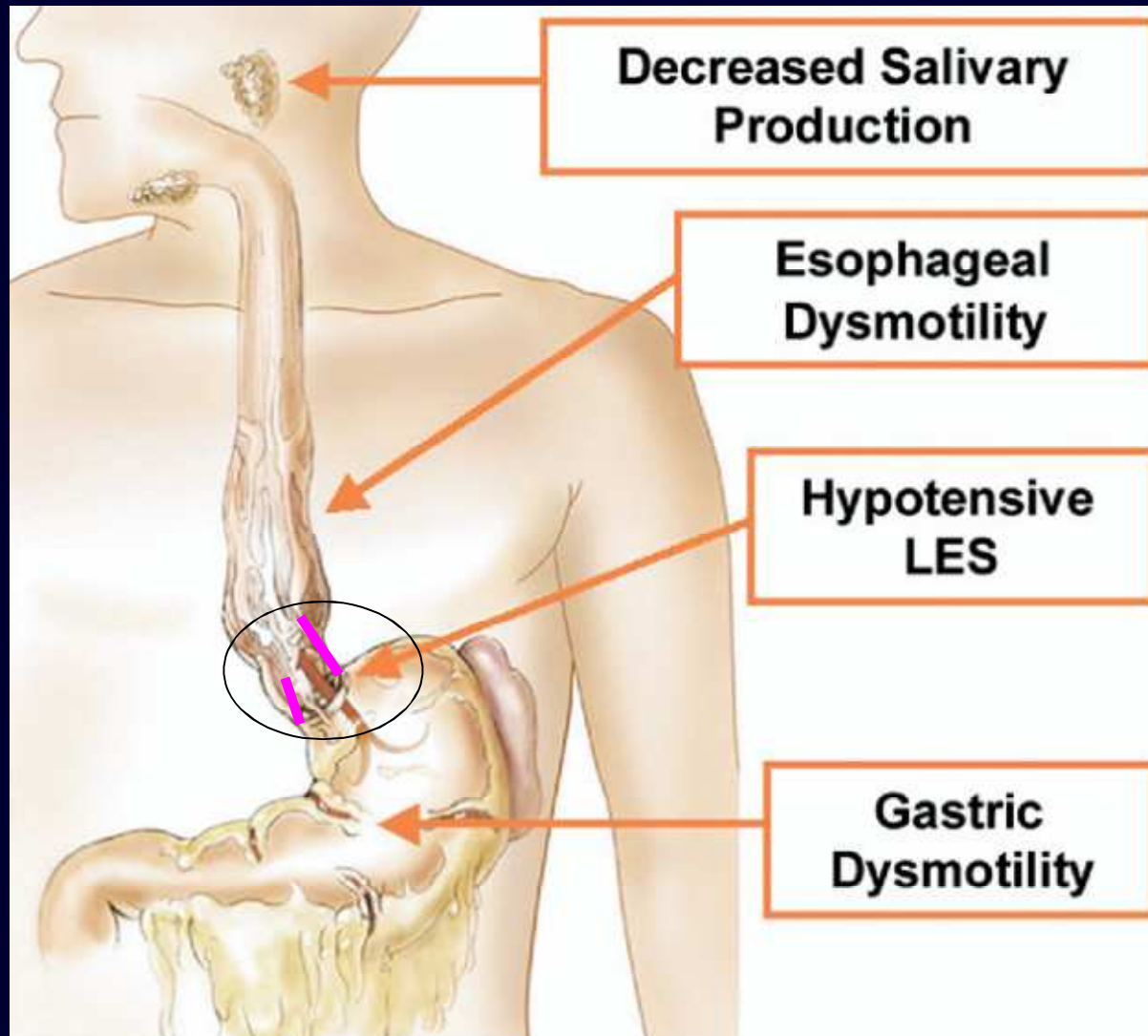


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Upper gut in scleroderma



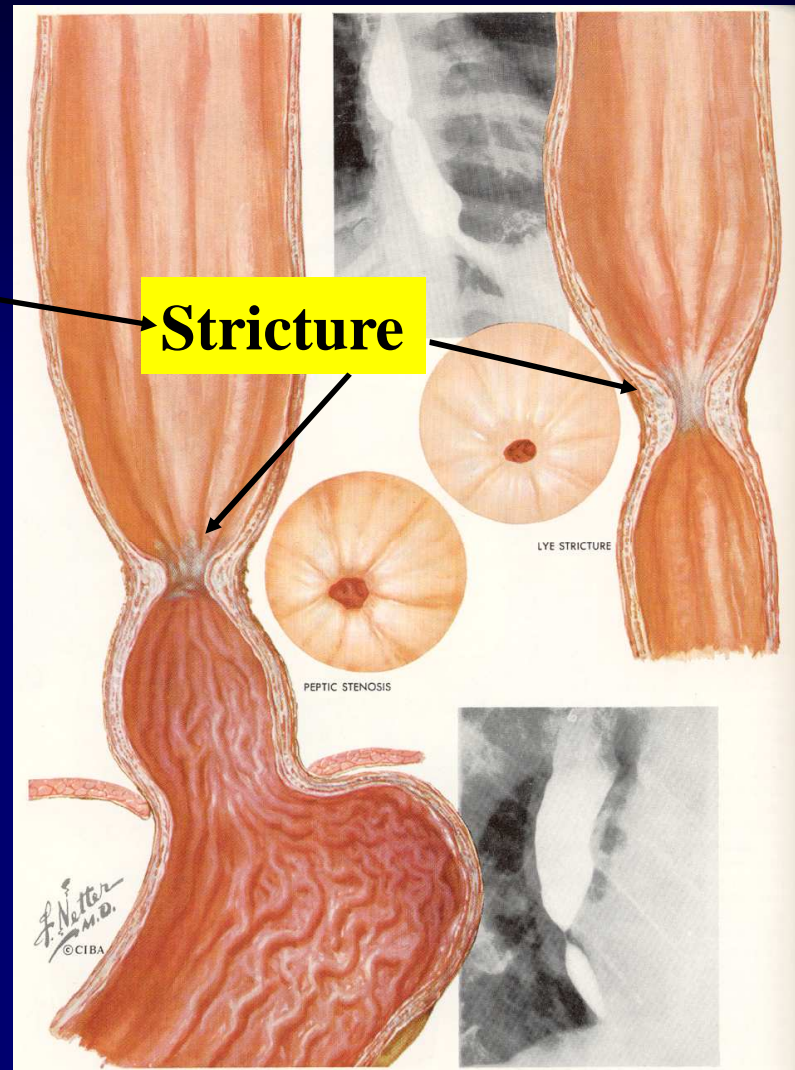
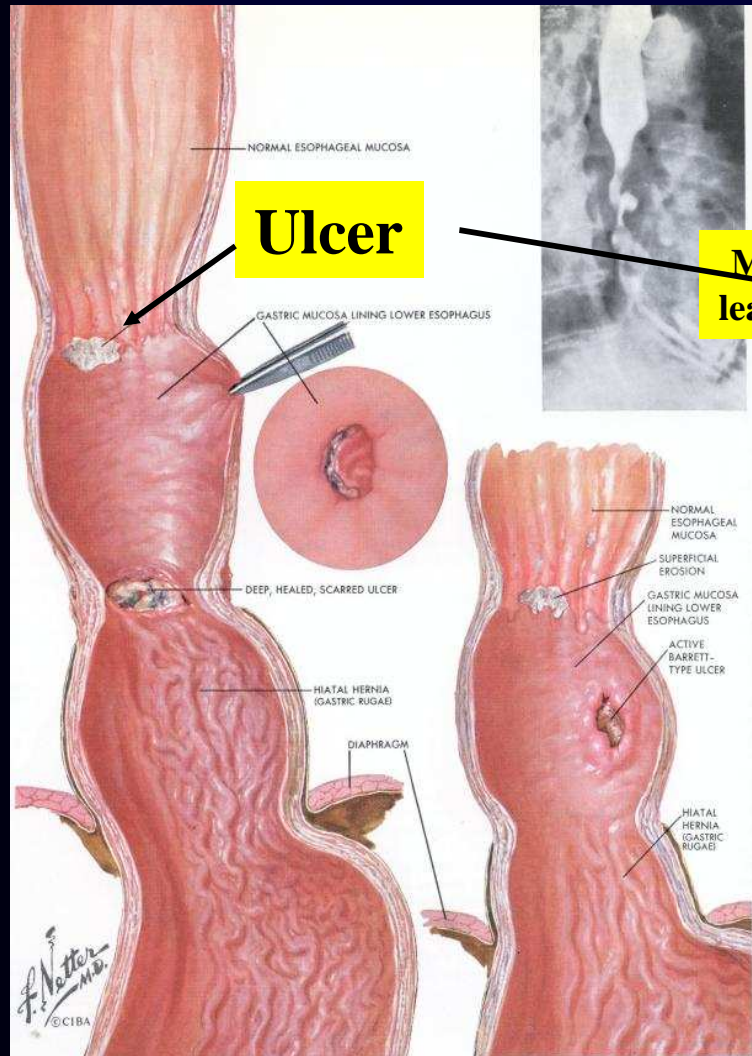
Upper gut in scleroderma



Symptoms of Reflux Disease

- **Heartburn**
- **Difficulty swallowing**
- **Chest pain**
- **Mouth ulcers/ burning**
- **Change in voice**
- **Chronic cough**
- **Asthma**

Acid reflux leads to ulcers and stricture (narrowing)



May lead to

Stricture

Why do we need to treat Reflux

- **To relieve symptoms and improve quality of life.**
- **Acid produced in the stomach may flow back while sleeping and get to the lungs causing lung inflammation.**
- **Continuous high acid production for a long time can cause inflammation and scarring of the esophagus, which may lead to a pre-cancerous lesion.**

Anti-reflux measures

- Head of the bed elevated (ie wedge pillow, blocks under head of bed, electric bed). NOT extra pillows.
- Biggest meal at noon, small meals otherwise.
- Do not eat late (after 6 pm); do not drink fluids late (after 8 pm) .
- Frequent small meals (5-6 per day).
- No tight garments around waist.

Wedge Pillow



Anti-reflux measures

- **Take anti-secretory and pro-motility agents.**
- **Stop smoking (if currently smoking).**
- **Avoid or minimize reflux producing foods (fat, chocolate, coffee).**
- **Certain medications such as Nifedipine (taken for Raynaud's) can weaken GE sphincter causing reflux symptoms.**

Anti-secretory Agents

<u>Agent</u>	<u>Frequency</u>
• Prilosec	20-40mg 1-2x per day
• Prevacid	15-30mg 1-2x per day
• Aciphex	20 mg 1-2 x per day
• Protonix	40 mg 1-2 x per day
• Nexium	20-40 mg 1-2 x per day
• Kapidex/ Dexilant	60 mg 1 x per day

Pro-motility agents

<u>Agent</u>	<u>Frequency</u>	<u>Part of Gut</u>
• Reglan	10 mg TID-QID	Whole
• Erythromycin	100-123 mg TID	Stomach
• Domeperidone*	10-20 mg QID	Stomach and Small Bowel
• Propulsid	10-20 mg TID	Whole

*Domeperidone not approved in USA--can obtain in Canada or Mexico (Clayman Pharmacy – Phone: 204-261-4214; Fax: 204-261-6390)

US CALL 1-800-JANSSEN (Restricted use)

How to use it?

- PPI and prokinetics 30 to 60 minutes before each meal
- Start PPI agent once a day
- *Increase to twice a day
- *Add Domperidone 1 hour before the meals
- *Add Pepcid or Tagamet at bedtime

* If the heartburn or other symptoms continue for 2 weeks

Barrett's esophagus

- Barrett's esophagus (a pre-cancerous lesion) is a complication of long-standing GERD.
- Present in 7%-13% consecutive people with scleroderma receiving chronic therapy with PPI.
- Similar incidence seen in non-SSc GERD.
- Barrett's esophagus is associated with adenocarcinoma in SSc.
- If Barrett's esophagus is diagnosed, very close follow-up with a gastroenterologist and regular endoscopies.

Surgery

- Surgery for GERD is relatively contraindicated.
- Surgery result in severe dysphagia secondary to the surgically induced narrowing of the GE junction, coupled with distal esophageal hypomotility.
- Generally restricted to patients with severe, poorly controlled, and complicated reflux.
- Fundoplication and laproscopic modified Roux-en Y gastric bypass (RYGB) both are effective.
- **IF SURGERY NEEDED, GO TO AN EXPERIENCED SURGEON.**

Stomach Motility (movement)

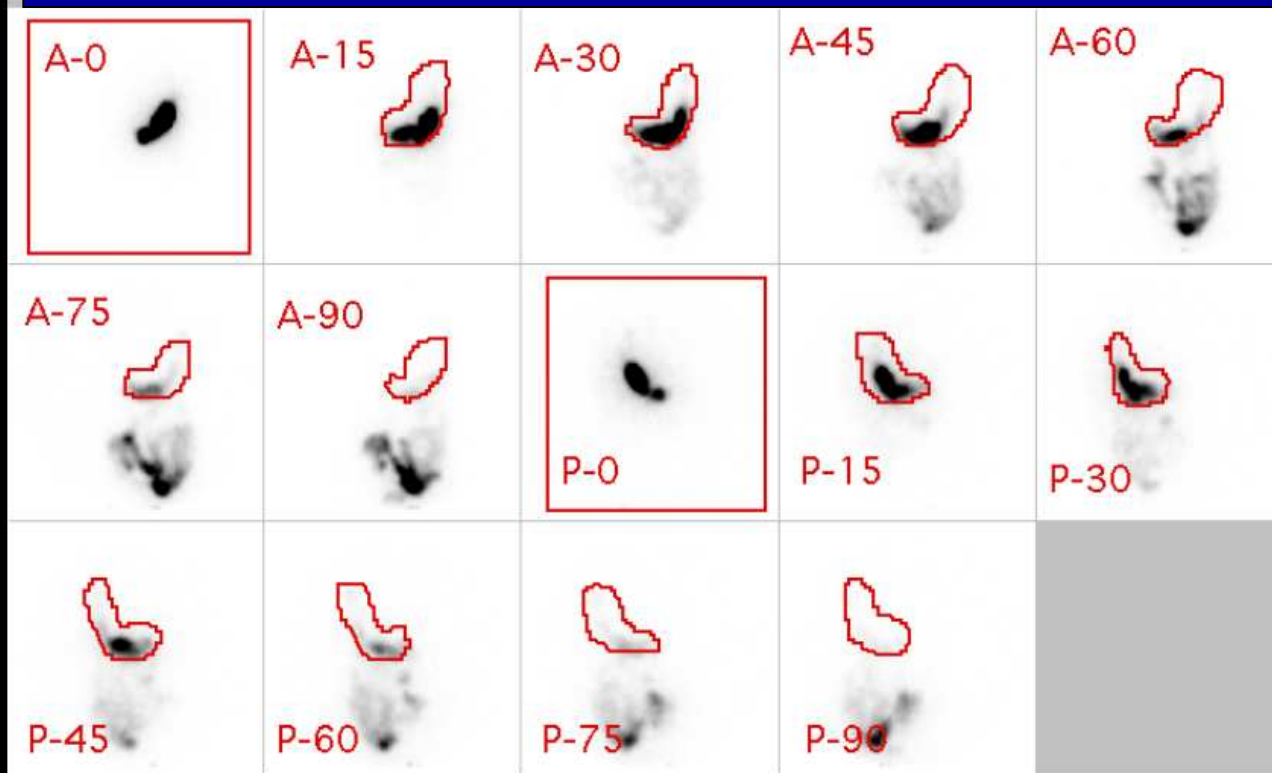
- **The main problem encountered is slow emptying of the stomach into the small intestine.**
- **Leads to symptoms of**
 - Bloating
 - Nausea and vomiting
 - Abdominal pain
 - Excessive flatulence
 - Result in weight loss

Diagnosis

- **A gastric emptying study is performed to diagnose abnormal emptying of food from the stomach.**
- **The food and radioactive material remain in the stomach longer than normal (usually hours) before emptying into the small intestine.**

Gastric Emptying Study

Se:0
Im:1 (F1)
B



PT HAD 1/2 JELLY SANDWICH WITH 4 OZ
OF APPLE JUICE.
1.0mCi TC-Sq

UCLA Gastric/Adult/14 7

C128
W258

NORMAL EMPTYING STUDY Less than 50% Remaining (or Greater than 50% Emptying) at 90 minutes

Pro-motility agents

<u>Agent</u>	<u>Frequency</u>	<u>Part of Gut</u>
• Metoclopramide	10 mg TID-QID	Whole
• Erythromycin	100-123 mg TID	Stomach
• Domeperidone*	10-20 mg QID	Stomach and Small Bowel
• Propulsid	10-20 mg TID	Whole

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Watermelon Stomach

- **Telangiectasia (dilated blood vessels) in the stomach.**
- **Can cause anemia due to slow (or rapid) blood loss.**
- **Can be without stomach symptoms or may be only feeling VERY tired and fatigued.**
- **Suspect when anemia on the blood count.**
- **Confirmed using endoscopy.**
- **Treated with laser coagulation.**

Watermelon Stomach



Greater omentum (turned up)

ea

Colon

Transverse mesocolon

Left colic (splenic) flexure

Right colic (hepatic) flexure

Small intestine

Ascending colon

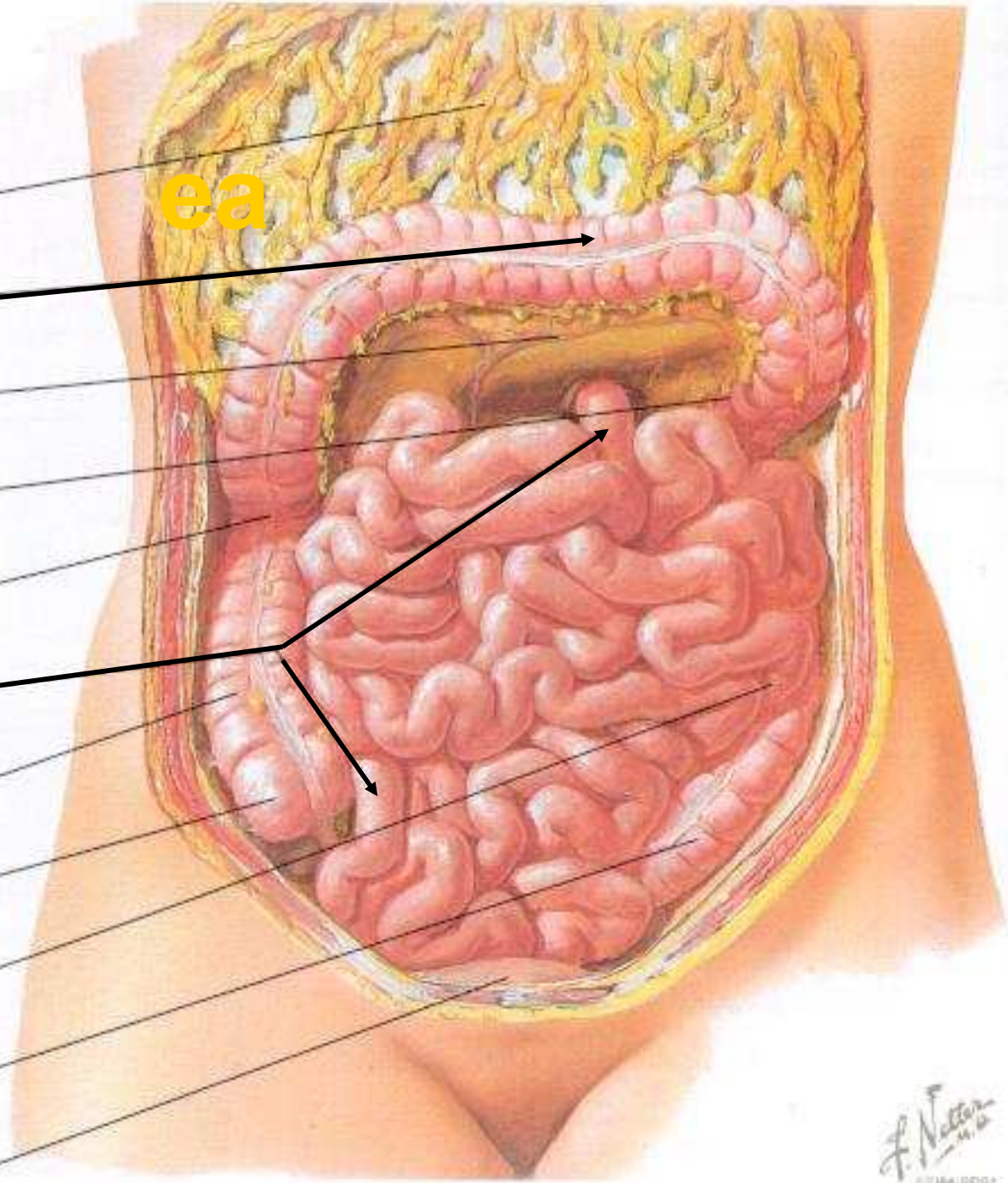
Cecum

Descending colon covered by small intestine

Sigmoid colon

Urinary bladder

F. Netter
M.D.
© 1989



Small Intestine

- **The small bowel is the part of the gut that absorbs nutrients from food.**
- **Lack of muscle tone leads to stagnation of food, bacterial overgrowth, diarrhea, cramping, feeling of bloating.**

Bacterial overgrowth

- The most frequent cause of diarrhea in SSc is the migration of bacteria, which normally live in the distal (lower) colon, into the small intestine—**bacterial overgrowth**.
- They compete with you for nutrition.

Bacterial overgrowth

- Can cause weight loss and inability to gain weight.
- Can cause malabsorption of essential vitamins and minerals.
- Diagnosed with lactulose or hydrogen breath test.
- Suppressing the bacteria with antibiotics often reduces the diarrhea and bloating.

Some Antibiotics for suppressing bacteria

Antibiotic

Dosing schedule

- | | |
|----------------|--------------------|
| • Augmentin | 875 mg 2x per day |
| • Cipro | 500 mg 2x per day |
| • Flagyl | 500 mg 3 x per day |
| • Doxycycline | 100 mg 2x per day |
| • Tetracycline | 250 mg 4x per day |
| • Rifaximin | 400 mg 2x per day |

Pro-motility agents

<u>Agent</u>	<u>Frequency</u>
• Reglan	10 mg 3-4 x per day
• Erythromycin	250-333 mg 3-4 x per day
• Domperidone	10-20 mg 3-4 x per day
• Octreotide	50 mcg sub q 2 x per day

NB Domperidone not approved in USA--
can obtain in Canada or Mexico

How to use it?

- **The antibiotics can be used every 2 weeks ON/ 2-4 weeks OFF.**
- **Some people need initial 4-6 weeks of antibiotics.**
- **Same or different antibiotics are used depending upon the physician's preference / person's symptoms.**
- **Take multivitamin and calcium (500 mg) twice a day.**

Probiotic/FODMAP diet

- Probiotics are bacteria that have a beneficial effect in the prevention and treatment of gut problems when they are ingested.
- Lactobacillus has been evaluated with positive results in one study.
- "Live and Active Cultures" seal on the yogurt label.
- FODMAPs are sugars in foods that we eat that are poorly absorbed by the gut. These FODMAPs can be easily fermented by the intestinal bacteria to cause abdominal pain, gas, bloating.
- More information:
www.med.umich.edu/scleroderma

Colon

- **Main function is to reabsorb water and salts that have been secreted by the rest of the gut.**
- **This helps the formation of stools and if the mechanism is impaired may lead to constipation (or diarrhea).**

Constipation

- **<3 bowels/ week**
- **Hard or lumpy stools**
- **Incomplete evacuation**
- **Straining during bowel movement**

Colon

- **Constipation**

- Caused by weakening of the gut muscle and slow contractions.
- Use of stimulant laxatives-acts on nerve endings in the gut wall that make the muscles in the intestine contract with more force.
- Liberal use of fluids.
- Avoid high-fiber diet and bulk-forming laxatives; may make constipation worse.
- Take medication every other day to maintain a healthy bowel regimen.

Stimulant Laxatives

- Laxatives

- Colace
- Dulcolax
- Senna
- Milk of Magnesia
- Lactulose
- Amitiza

- Dosage

- 100 mg once-twice/day
- 10-15 mg once a day
- 2-4 tablets once a day
- 30-60 mg/day
- 15-30 ml/day
- 8-24 ucg twice a day

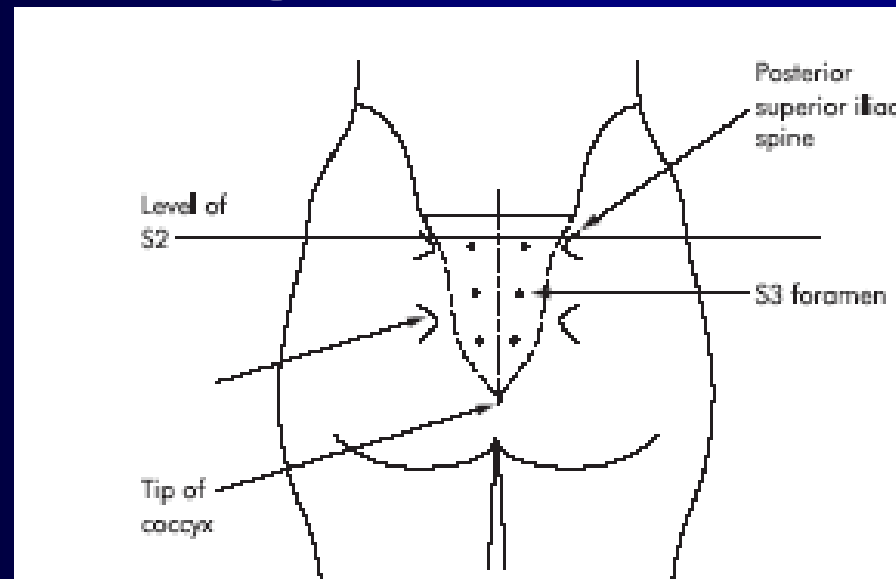
Do not use these laxatives if symptoms of bowel obstruction!!

Rectum

- Stool incontinence occurs in up to a third of patients.
- Weakening of the rectal muscle and poor control over rectal sphincter.
- Biofeedback therapy- **strengthen the rectal muscle by volunteer squeezing of the muscle.**
- Bulk agents such as Citrucel.
- Anti-diarrheal agents such as Imodium.

Sacral nerve stimulation

- Low-level electric stimulation
- Using electrodes to the nerves in the sacrum
- Done under general anesthesia



Liver

- **Liver plays an important role in detoxification of drugs in our body.**
- **About 10% of patients may have liver involvement-Primary Biliary Cirrhosis.**
- **Usually occurs 10-15 years after Scleroderma onset.**
- **Symptoms: Itchy skin and fatigue.**
- **BUT 39% HAVE no symptoms.**

Primary Biliary Cirrhosis

- **Diagnosis:** Increase bilirubin, decrease albumin, and increase alkaline phosphatase.
- **Blood test for anti- mitochondrial and anti-smooth muscle antibody.**
- **Treatment:** Ursodeoxycholic acid 500-1,000 mg three-four times/day.
- **May need liver transplantation.**

Scleroderma Gastrointestinal Tract 2.0 Instrument

- **Captures gut involvement in people with SSc.**
- **34-item instrument for clinical care and clinical trials.**
- **Takes approximately 8 minutes to complete.**
- **SSC-GIT has 7 scales**
 - **Reflux**
 - **Distension/Bloating**
 - **Diarrhea**
 - **Constipation**
 - **Fecal Soilage**
 - **Emotional well-being**
 - **Social functioning**

Conclusions

- **Scleroderma frequently effects the gut.**
- **Symptoms can be distressing and can cause impairment of person's quality of life.**
- **Good tests are available to find the location and extent of involvement.**
- **Appropriate treatments are very effective.**