

# Welcome!



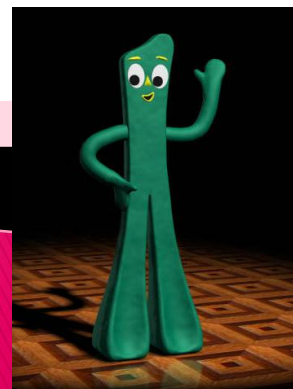
Capital Area  
Ehlers–Danlos Syndrome  
Support Group

# Capital Area Ehlers–Danlos Syndrome Support Group

## Gastrointestinal Issues

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Gayle Yankee, MSN, RN, FNP–BC



# Gastrointestinal Issues

- ▶ Hypermobile EDS is the most common hereditary non-inflammatory disorder of connective tissue. A majority of EDSers also have gastrointestinal (GI) symptoms.

# Gastrointestinal Issues

- ▶ Why? Nobody knows – research is ongoing.
  - ▶ May be related to abnormal collagen formation or abnormality in the proteins of the extracellular matrix (ECM).
  - ▶ The genetic basis is unknown.
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# Structural Gastrointestinal Issues

- ▶ Abdominal hernias in 1 / 5, increasing risk with aging
- ▶ Rectal prolapse in more than 1 / 10 women with EDS, increased with prior episiotomy
- ▶ Pelvic prolapse
- ▶ Obstructed defecation
- ▶ Distension of bowel, megacolon

# Structural GI Issues

- ▶ Decreased strength and/or increased elasticity of connective tissues causes displacement or dysfunction of organs.
  - Visceroptosis –displacement of organs, downward in abdomen and pelvis
  - Intussusception –portion of bowel sliding into adjacent segment
  - Volvulus –twisting of bowel around fixed point
  - Diverticulosis –pocket in bowel wall

# Structural GI Issues

- ▶ Hernias (from weakened connective tissue):
  - Hiatal (hiatus)
  - Abdominal wall/inguinal
  - Cystocele
  - Rectocele
  - Rectal prolapse

# Structural Gastrointestinal (GI) Issues

- ▶ Elasticity leading to increased distension of hollow organs (stomach, bowel, bladder, etc.)
  - Bloating, distension, pain
  - Direct interference with gut mechanoreceptors, causing decreased (or increased?) motility, increased pain
  - Stasis → increased gassiness from fermentation



# Structural GI Issues

- ▶ Decreased motility/strength of muscle walls
  - Delayed gastric emptying, bowel motility
    - But some studies show increased motility
  - Impaired swallowing, globus, esophageal dysmotility (spasm, uncoordinated contractions)

# Structural GI Issues

- ▶ Problems of the ECM in the lining of the gut, and alterations of the gut microbiome from those problems, may affect the permeability of gut mucosa

# Structural GI Issues

- ▶ **Slow transit/motility**
  - Nausea/vomiting, gastroparesis (delayed emptying)
  - Dyspepsia
  - Reflux
  - Constipation (more on this topic another evening)
    - Obstipation/diarrhea
- ▶ **Rapid transit/motility**
  - Diarrhea
  - Incomplete absorption of nutrients
  - Weight loss

# Structural GI Issues

- ▶ Lax sphincters
  - Gastroesophageal reflux (GERD –acid)
  - Enterogastric reflux (bile –alkaline)
    - Which can then cause bile GERD (which may be why your PPI isn't working)!

# Structural GI Issues

- ▶ Dysautonomia (autonomic nervous system dysfunction) and POTS complicate these structural issues
- ▶ POTS, independent of EDS status, is associated with GI symptoms, and dysmotility of stomach, small and large intestines

# Structural GI Issues

Tonight we'll focus on the  
upper GI tract.

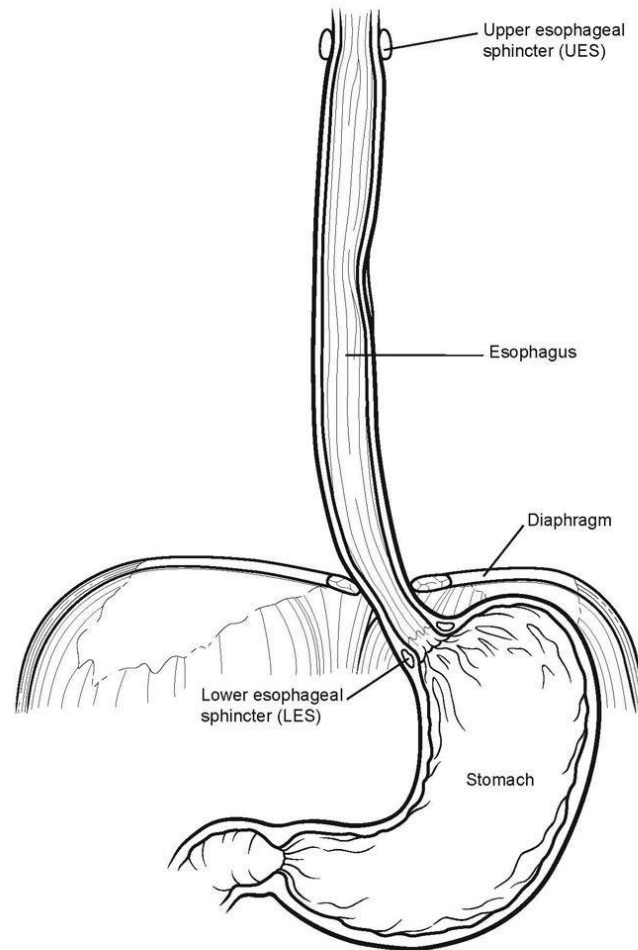
# Upper GI Anatomy



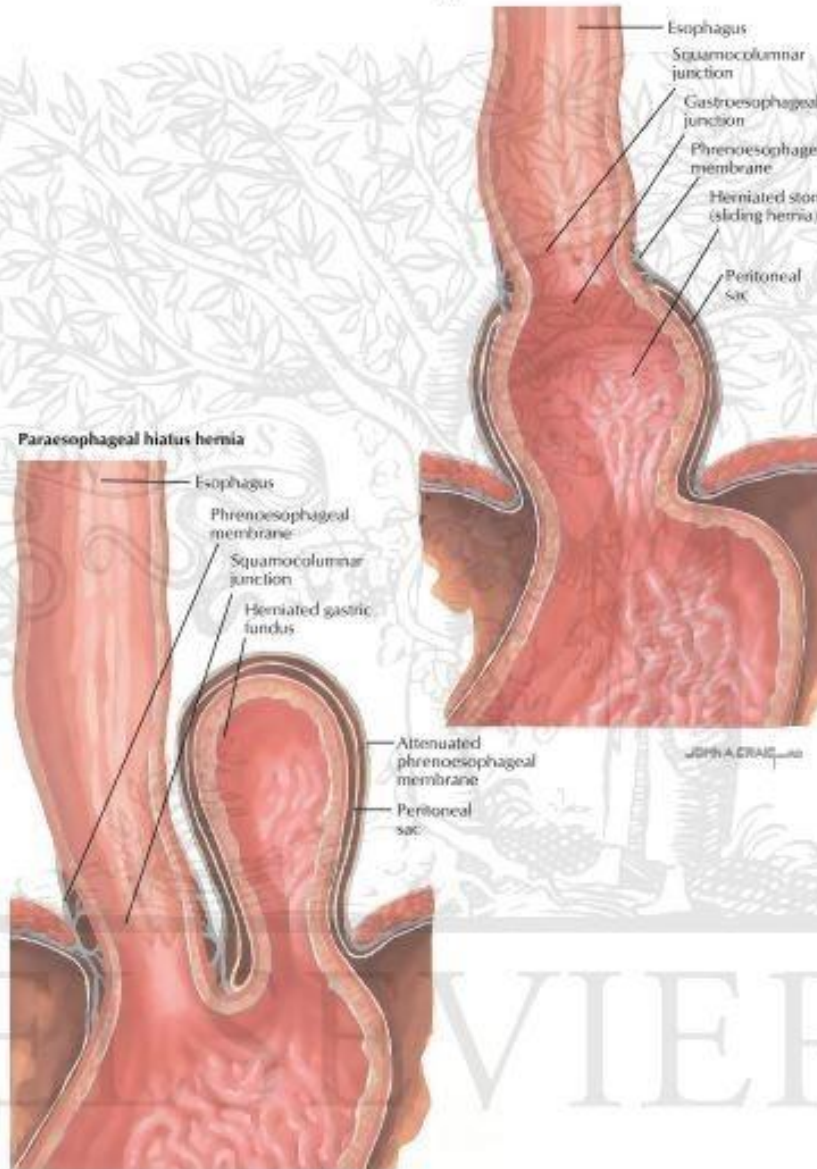
Gastrointestinal System

Upper GI Tract - Coronal Section

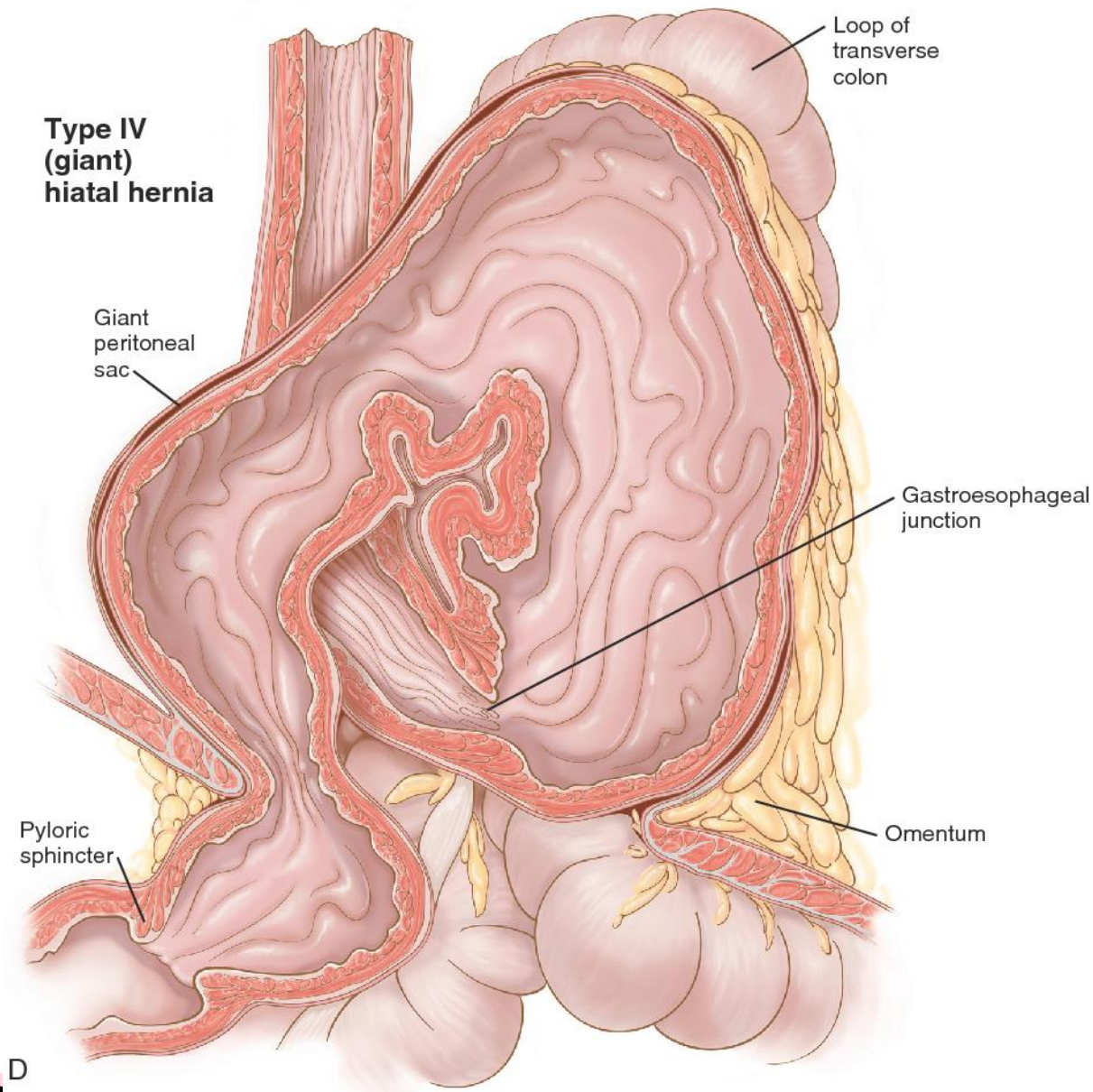
Anatomical Line Drawings



## Hernia Types







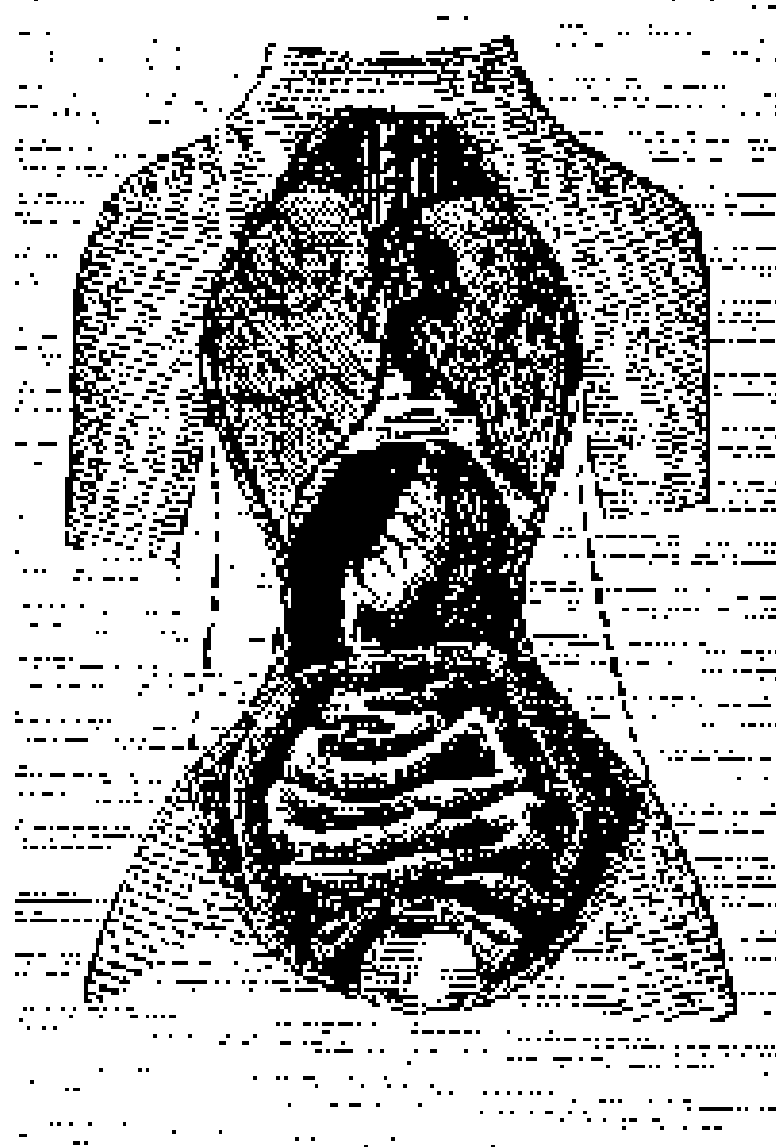
# GERD – Management

- ▶ Heartburn
  - If lying down, sit up and drink water/milk to wash down reflux
  - TUMS, bicarbonate (baking soda, Alka Seltzer Gold) for quick relief
- ▶ Acid reduction for GERD
  - Proton pump inhibitor (PPI), H<sub>2</sub>-receptor antagonist (H<sub>2</sub>RA) to decrease acid production
  - Long term acid suppression may have adverse effects; cycling or using medications short term may help
- ▶ Bile acid sequestrant for bile reflux
  - Cholestyramine (Questran)
  - Sucralfate (Carafate) to coat stomach and esophagus

# GERD – Management

- ▶ Antispasmodic for esophageal spasm
  - Hyoscyamine sublingual very helpful, fast-acting
- ▶ Reduce hiatal hernia if possible
  - Heel drop
  - Manual reduction
  - Surgical reduction
- ▶ Avoid food and fluids two to three hours before bedtime
- ▶ Avoid carbonated beverages, mint
- ▶ Elevate the head of the bed

How I feel after ten minutes in  
pantyhose...



# Structural GI Issues

## ▶ Compression


- Historically, compression from corsets and other torture devices devised by fashion caused visceroptosis, as seen in the previous slide.
- Visceroptosis occurs in EDS, noted in case reports.
- I believe it is under-reported, as most abdominal imaging is done in the supine position.

# Structural GI Issues

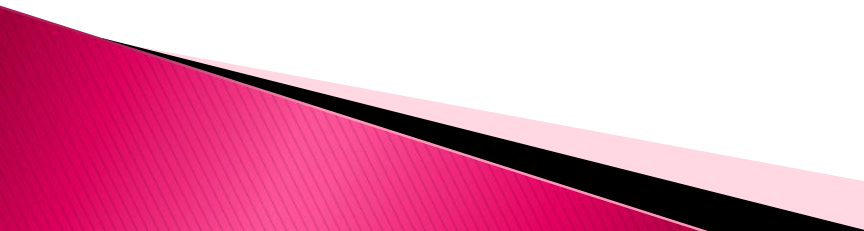
## ▶ Compression

- Organs are more susceptible to compression from the outside (clothing, positional) or other organs/structures (e.g., ribs, stool in bowel) because the walls are thin and elastic.
- Function can be disrupted by compression.
- Compression of the stomach can increase reflux, of the duodenum can delay gastric emptying
- Prolonged compression may cause ischemia
- May contribute to hernia formation or strangulation of hernias

# Compression – Management

- ▶ Avoid tight clothing, especially waistbands.
    - Hip-hugger styles may be more comfortable.
  - ▶ If a tight waistband is unavoidable, try to change its position regularly.
  - ▶ Monitor your body position to be sure you're not compressing anything.
  - ▶ Change position frequently, especially avoid prolonged sitting.
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# Dietary Management – General

- ▶ Small, frequent meals to decrease transit time
  - ▶ Chew food thoroughly to mix with saliva
  - ▶ Put down utensil between bites of food
  - ▶ No additional fluids with meals
  - ▶ No activity other than conversation at the table
  - ▶ Try to achieve/maintain ideal body weight
- 



# Dietary Management – General

- ▶ Elimination diets may be helpful
- ▶ Low FODMAP (short-chain carbohydrates: Fructose, Oligosaccharides, Disaccharides, Monoamines, and Polyols) diet may be helpful for abdominal bloating, pain, and diarrhea
  - Known benefit for IBS
- ▶ Keep a detailed diet and symptom diary.
- ▶ Change only *one* thing at a time!

# FODMAP carbohydrates and their richest food sources

- ▶ Fructo-oligosaccharides (fructans)\*: Wheat, rye, onions, garlic, artichokes
- ▶ Galacto-oligosaccharides (GOS)\*: Legumes
- ▶ Lactose: Milk
- ▶ Fructose: Honey, apples, pears, watermelon, mango
- ▶ Sorbitol: Apples, pears, stone fruits, sugar-free mints/gums
- ▶ Mannitol: Mushrooms, cauliflower, sugar-free mints/gums

\*incompletely absorbed in the human GI tract

# Dietary Management

- ▶ If oral diet is not tolerated, enteral feeding via gastostomy tube or jejunostomy tube may be tried
- ▶ If enteral feeding not tolerated, total parenteral nutrition (TPN) may be tried

# Thank You!



-and good night!

# Bonus Slide!

- ▶ Results of gastrointestinal physiological studies were reported in a retrospective observational study from the Mayo clinic in EDS patients, 71.7% with hEDS [Nelson et al., 2015].
- ▶ About 13 out of 46 (28%) patients who underwent colonic transit studies had abnormal results.
  - nine with slow transit, four with fast transit.
- ▶ A total of 60% of these patients with abnormal colonic transit had hEDS.
- ▶ In the same study, 17 out of 76 (22%) patients had abnormal gastric emptying, half being fast and half being slow.
- ▶ Abnormal oesophageal manometry was present in 5 out of 11 (31%) patients. About 7 out of 16 patients (44%) had pathological acid reflux on reflux testing.

# Bonus Slide!

- ▶ GI symptoms of nausea, reflux, bloating, constipation, diarrhea may be r/t POTS or dysautonomia.
- ▶ Current literature suggests an association between all subtypes of EDS and GI symptoms. This association is common and has hitherto been underestimated.
- ▶ The group observed that evidence for GI symptoms to be included as a major EDS diagnostic criteria is compelling. However, a causative relationship between abnormalities in connective tissue and GI symptoms has not yet been established.

# Gastrointestinal Issues

- ▶ In EDS, more common to have celiac disease, eosinophilic esophagitis and Crohn's disease, but not ulcerative colitis.