Increased risk of GI motility issues:
- Cerebral palsy
- Rett syndrome
- 22q- (velocardiofacial syndrome)
- Cornelia de Lange syndrome,
  Degenerative neuromuscular disorders
  (muscular dystrophies, mitochondrial cytopathies)
- Trisomy 18
  Chromosomal translocations

Gastro-esophageal Reflux
- Reflux of acid from the stomach into the esophagus
  Due to defective or stretched sphincter (LES) between the stomach & esophagus, a hiatal hernia or ↓ motility

GERD
- Reflux esophagitis: 5-7% in general population
  In persons w/ DD
  Overall prevalence is 10-15%
  But w/ known risk factors: 30-50%
  70% in those w/ severe/profound DD!

Risk factors
**In persons w/ DD**
- Scoliosis
- Cerebral palsy
- Severe & profound DD
- Non-ambulatory
- Certain Rx

**Additional factors**
- Hiatal hernia (90%)!
- Obesity
- Diabetes
- Pregnancy
- Scleroderma
- Older age

Symptoms
- Burning pain in throat & esophagus
- Abdominal pain
- Thoracic (chest) pain
  But … what if the person is non-verbal?
**GERD or CB?**

- Refuses to eat or food avoidance
- Refusal to sleep, lay down (on their back)
- Depressive behaviors (isolates oneself)
- Irritability after meals & during the night
- Screaming, aggression & SIB (head-banging, hand-mouthing/biting)
- Stripping (pants, belt, bra): too tight?
- Excessive water drinking (pica?)

**Signs & Symptoms**

- Wt loss, poor growth, FTT in children
- Arched back with difficulty sitting
- Recurrent vomiting/aspiration
- Recurrent episodes of pneumonia, asthma exacerbations
- Hoarseness
- Dental caries
- Aphthous ulcers

**Signs & Symptoms**

- Regurgitation and rumination 1-2 hours after meals (p.c.) or overnight
- N & V, burping, coughing, wheezing, throat clearing, yawning, hiccupps
- Sinusitis, pharyngitis, otitis
- Abdominal distension
- Constipation (as a contributing factor)

**When is it GERD?**

Predisposing risk factors along with occurrence of any of those behaviors, a minimum of 4 times per month:

**SHOULD RAISE SUSPICION TO HIGHEST LEVEL for GERD!**

MEDINA (2010)

**Monitoring**

- Scatter plot
- Calendar
- A-B-C sheets
- Food diary
- Breau pain scale

**Red Flags (Alarm symptoms)** (**notify MD ASAP!**)

- Chest pain (cardiac type)
- Choking (coughing, hoarseness & SOB)
- Dysphagia (?strictures)
- Vomiting (hematemesis)
- Pain on swallowing (odynophagia)
- Anemia d/t GI bleeding (melena stool)
  - Bloody vomit or coffee grains**
  - Weight loss >5%
Risk factors: Meds

- ASA
- Alpha-blockers
- Anti-cholinergics
- Anticonvulsants
- Benzodiazepines
- Beta-blockers
- Bisphosphonates
- Calcium channel blockers
- Narcotics
- Nicotine
- Nitrites
- NSAIDs & COX-2 inhibitors
- Theophylline

Complications

- Esophageal strictures
- Recurrent inflammation
  => erosion
  => Barrett's esophagitis (↑ if s/s > 5yrs)
- Increased risk of esophageal cancer with Barrett's (↑ if s/s > 5yrs)

Diagnosis

Mild
- S/S less often (<3/wk)
- Low intensity
- Not at night
- No interference w/ life
- Heartburn pain rated 1-3/10
- No major complications

Severe
- S/S more frequent
- Present for > 6 months
- More intense
- Wakes at night, or restless nights
- Interferes w/ life
- Pain rated 7/10 +
- Complications

Dx Tests

- Blood/urine testing (r/o anemia)
- X-ray (fluoroscopy: barium swallow) to check for hiatal hernia.
- Ambulatory 24-hour pH monitoring*
- Upper endoscopy (not conclusive?)*
- Esophageal motility studies*

*for refractory esophagitis

***Diagnosis can be made without endoscopy based on history.

Other possibilities?

- Dyspepsia
- Pyloric stenosis
- Peptic ulcer disease
- Hirschsprung's
- Esophagitis secondary to esophageal motility or structural issue

- Cardiac pain
- Lactose intolerance
- Esophageal cancer
- Biliary tract disease

Dyspepsia or GERD

**Dyspepsia**

For at least 3 months
- chronic/recurrent epigastric pain
- postprandial fullness
- Or early satiety

Other s/s:
- Bloating
- Nausea

**GERD**

- Frequent regurgitation or heartburn
- Epigastric pain
- Nausea
- Dysphagia
- Odynophagia
Planning

- Review & modify risk factors (if possible)
- Have MD assess for comorbid dx
- Have MD assess if problematic Rx can be adjusted or D/C’ed
- Adopt an individualized plan

Therapeutic Goals

- Relieve S/S
- Improve QoL
- Promote healing of esophagus
- Prevent complications (strictures, bleeds & Barrett’s)
- Prevent recurrences

Lifestyle modifications

- Avoid exercising/bending on a full stomach
- Stop smoking
- Limit ETOH
- Attain healthy weight
- Avoid lying down for 3 hours p.c. or eating before hs
- Avoid large meals
- Fill in food diary
- Avoid trigger foods: chocolate, tomato-based foods, alcohol, peppermint, onions, caffeinated products, citrus fruits and drinks, high fat meals
- Avoid tight clothing around waist/chest
- Elevate head of bed (10 cm, 6-8 inches, or use 2-3 pillows)

Treatment: Rx

Mild
- Antacids (Aluminum hydroxide, Magnesium salts, Calcium carbonate or combos: Rolaids, Tums, Maalox)
- Alginates (Gaviscon)
- H2-RA (histamine receptor antagonist) (see next slide)

Moderate/severe
- PPI (proton pump inhibitor) (see next slide for examples) (heal up to 90% by 12wks)
- Step-down Tx (PPI first, then H2RA after healed)
- May even require Sx: fundiplication

Constipation:

a symptom
NOT
a disease!
How serious a problem?

- 50-85% of older people w/ DD suffer from constipation (2005, Australia, Management Guidelines: Developmental Disability)
- Up to 70% of persons with dx of moderate to profound MR have it (2001, Netherlands, JIDR)
*Especially for non-ambulatory residents

What exactly is Normal?

Frequency:
- 3 X per week to 3X per day!

Average passage time:
- 50 (men) to 57 hrs (women), but can vary from 20-100hrs!

Average weight:
- 100g = 3.5 oz.

Transit times

- Mouth: 1 min
- Esophagus: 4-8 secs
- Stomach: 2-4 hrs
- Small intestine: 3 to 5hrs
- Colon: 10 hrs to several days

What exactly is Normal?

Usual color:
- Brown but can vary from reddish or greenish dark brown to lighter mustard brown

Consistency:
- 70-75% actually H2O!
  (in bacteria & undigested plant cells)
- 50-66% bacteria
- 33-50% undigested plant foods (fiber)

Constipation: how it can happen

Cause & Effect: Risk factors

- Female > Male (2-3X); Age over 65
- IQ < 50 (moderate/severe/profound MR)
- Diagnoses: CP, hypothyroidism (DS), WS, DM, comorbid GERD
- Inadequate food/caloric & fluid intake
- Inadequate dietary fiber
- Immobility/non-ambulatory
- Medications
**Meds**

- Anticholinergics; or meds w/ antiCH SE
- Anti-psychotics, antidepressants, Anticonvulsants, benzodiazepines, Antiparkinsonians, antispasmodics
- Ca+ channel blockers
- Diuretics
- Antacids (Al hydroxide, Tums, Maalox)
- Fe+, Ca carbonate
- Opioids (narcotic analgesics) & NSAIDs

**Rome II criteria:**

2 or > for at least 12 weeks in last year:

For >25 % of defecations:
- Straining
- Lumpy or hard stools
- Sensation of incomplete evacuation
- Sensation of anorectal obstruction
- Use of manual maneuvers to facilitate evacuation of stool

Less than 3 defecations per week
Loose stools are not present
Insufficient criteria for IBS Dx

**Behavioral Signs:**

- SIB/aggression
- Irritability
- Positioning: legs bent at the knee, with thighs elevated to ↑ pressure on the abdomen (crouching) (rocking on the toilet seat)

**Red Flags (Alarm Symptoms)**

- Abdominal pain
- N & V
- Melena, rectal bleeding, rectal pain
- Fever
- Weight loss

**Monitoring**

- Scatter plot
- Calendar
- A-B-C sheets
- Food diary
- Bristol Stool form

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**Drugs Associated with Constipation**

<table>
<thead>
<tr>
<th>Drug</th>
<th>Category</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amantadine</td>
<td>Anticonvulsants</td>
</tr>
<tr>
<td>Antidiarrheal agents</td>
<td>Antihistamines</td>
</tr>
<tr>
<td>Antipsychotics (e.g., phenothiazines)</td>
<td>Barbiturates</td>
</tr>
<tr>
<td>Benzodiazepines</td>
<td>Beta-blockers</td>
</tr>
<tr>
<td>Cholestyramine</td>
<td>Clonidine</td>
</tr>
<tr>
<td>Calcium channel blockers</td>
<td>Diuretics</td>
</tr>
<tr>
<td>Calcium channel blockers</td>
<td>Diuretics</td>
</tr>
<tr>
<td>Lithium</td>
<td>SHT2 antagonists (e.g., ondansetron)</td>
</tr>
<tr>
<td>Non-steroidal anti-inflammatory drugs</td>
<td>Opioids</td>
</tr>
<tr>
<td>Polystyrene sodium sulfonate</td>
<td>Pseudoephedrine</td>
</tr>
<tr>
<td>Vinca alkaloids</td>
<td>Tricyclic antidepressants (e.g., amitriptyline)</td>
</tr>
<tr>
<td>Cotton-containing agents (aluminum, calcium, iron, bismuth, barium)</td>
<td></td>
</tr>
</tbody>
</table>
Planning

- Review & modify risk factors (if possible)
- Have MD assess for comorbid dx
- Have MD assess if problematic Rx can be adjusted or D/C’ed
- Adopt an individualized plan

Therapeutic Goals

- Regular BM within 48-72 hours
- Appropriate laxative use
- Avoid complications:
  - From straining: hemorrhoids, hernia, GERD, coronary & cerebrovascular dysfunction in elderly
  - Long Term: hemorrhoids, incontinence, impaction, obstruction, rectal prolapse, anal fissures, megacolon

Hemorrhoids (symptoms)

- Intermittent pain in rectum, during bowel movements
- Blood stains in underwear, shorts
- Various complaints such as burning, itching, swelling
- Protrusion of internal hemorrhoid noted

Hemorrhoids

Causes: unknown, but several aggravating factors (heredity):
- Diarrhea
- Spicy foods
- Alcohol
- Dehydration
- Pregnancy
- Chronic constipation

Hemorrhoids

Interventions:
- Try to eliminate constipation
  - Cream to ↓ pain & inflammation
- Observe for any bleeding**

Tx Options: laxatives

- Bulk-forming
- Hyper-osmotic agents
- Lubricants
- Osmotic saline agents
- Stool softeners
- Stimulants

Please see chart...
Health Education

- Normal BM: frequency, type
- Dietary modifications:
  - Increase calories
  - Increase fiber, fruits & vegetables
  - Increase fluids
- Lifestyle modifications
  - Increase exercise

Continued monitoring

- Frequency: scatter plot
- Bristol Stool form
- Improvements in behavior
- Breau pain scale
- Food diary

Questions?

Thank you!