Two Common Gastrointestinal Problems in Persons with DD: Gastroesophageal Reflux & Constipation

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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?)

A word about food refusal..

- Breathing vs eating?
- Taste & texture
- Nausea & vomiting
- Food & Rx interaction?
- Medical procedures (spoon vs. scope!)
- Dental pain
- Dysphagia
- Constipation, obstruction (pica/feces)

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, hiccups
- 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
- Nitrates
- 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

Gray, p.721

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

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)
  - (BID dosing of H2RAs relieves s/s in 60%, heals tissue in 40%)

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

Meds: considerations

- CYP450 interactions & AEDs (cimetidine & omeprazole)
- Prokinetic (domperidone?)

Table 2
Acid Suppression Medications Used in the Treatment and Maintenance of GERD

Drug	Doses
Histamine receptor antagonists (H_2RA)	
cimetidine (Tagamet®)	800mg q.h.s. to 600mg b.i.d.
famotidine (Pepcid®)	20mg b.i.d.
ranitidine (Zantac®)	150mg b.i.d.
nizatidine (Axid®)	150mg b.i.d.
Proton pump inhibitors (PPIs)	
esomeprazole (Nexium®)	20mg o.d. to 40mg o.d.
lansoprazole (Prevacid®)	30mg o.d.
omeprazole (Losec®)	20mg o.d. to 20mg b.i.d.
pantoprazole (Pantoloc®)	40mg o.d.
rabeprazole (Pariet®)	20mg o.d.

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?

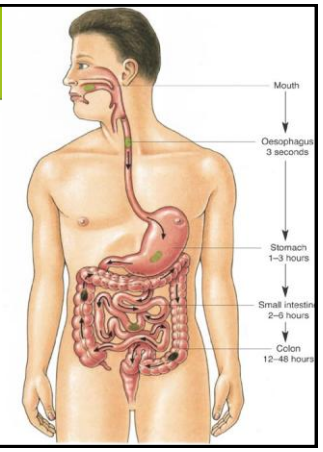
Frequency:
- 3 X per week to 3X per day!

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

Average weight:
- 100g = 3.5 oz.

Transit times

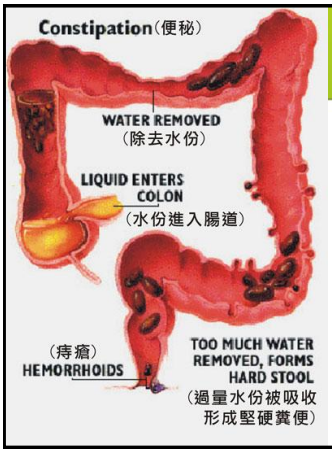
- Mouth: 1 min
- Esophagus: 4-8 secs
- Stomach: 2-4 hrs
- Small intestine: 3 to 5hrs
- Colon: 10 hrs to several days



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

Kids & adults?

- Similar prevalence so likely is present early in life & does not necessarily develop over time
- Intrinsic motility problem rather than overstretching (functional retention)

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

Table 3

Drugs Associated with Constipation[12]

Amantadine	Anticonvulsants
Antidiarrheal agents	Antihistamines
Antipsychotics (e.g., phenothiazines)	Barbiturates
Benzodiazepines	Beta-blockers
Cholestyramine	Clonidine
Calcium channel blockers (verapamil>diltiazem>dihydropyridines)	Diuretics
Lithium	$5HT_3$ antagonists (e.g., ondanestron)
Non-steroidal anti-inflammatory drugs	Opioids
Polystyrene sodium sulfonate	Pseudoephedrine
Vinca alkaloids	Tricyclic antidepressants (e.g., amitriptyline)
Cation-containing agents (aluminum, calcium, iron, bismuth, barium)	

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)
(**notify MD ASAP!)

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

Other concerns?

- Cholecystitis (4Fs!)
- Gastritis & PUD
  - *H. Pylori*
  - Meds
  - G-tube placement

Monitoring

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

Dx tests?

- Measure abdominal girth?
- Abdominal X-ray (flat plate)
- Distended abdomen & masses LLQ
- TFTs
- Pb screening if pica

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!