Specific Clinical Risk Factors:
GI Problems in People with Developmental Disabilities

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Dysphagia, esophageal disorders & GE reflux

OPENING COMMENTS:

Don't confuse "usual" with "normal"
Don't ignore the signs that a problem exists ("I told ya and I told ya")
It's important to be proactive (anticipate)
Kitchen to bathroom

DYSPHAGIA

- difficulty swallowing (difficulty in passage of food, solid or liquid, from the mouth to the stomach)
- inability to handle oral secretions
- inability to safely take medications orally

ESOPHAGEAL DISORDERS:

- anatomical problems (hiatal hernia, esophageal stricture, esophageal web, esophageal diverticulum, esophageal ring, tumors)
- inflammation (esophagitis) – due to GE reflux, medications (e.g., ASA, NSAIDs, KCl, iron, vit C, TCN), chemicals (lye or acid)
- infections
- esophageal dysmotility - difficulty with movement of food, solid or liquid, through the esophagus due to decreased or ineffectual peristalsis (e.g., presbyesophagus), diffuse spasm, achalasia
- may involve retrograde movement of material from the esophagus to the pharynx and result in aspiration

GE REFLUX

- retrograde movement of gastric contents from the stomach into the esophagus and higher, the latter possibly resulting in aspiration of contents into the trachea and lungs
- natural occurrence
- symptomatic vs. asymptomatic
- degree of esophageal damage varies (most severe – Barrett's)
- effects on pharynx, larynx, and tracheobronchial system
- antireflux barrier:
- lower esophageal sphincter (LES)
- esophageal clearance (gravity, peristalsis, salivation, anchoring of distal esophagus in abdomen)
- gastric reservoir (dilatation, increased intragastric pressure, delayed gastric emptying, increased acid secretion)

Those at special risk:

- Individuals with cerebral palsy, Down Syndrome (especially as they age)
- Individuals with facial malformations (e.g., cleft palate)
- Individuals who have had strokes or problems resulting in paralysis of muscles involved in swallowing
- Individuals with Bell's palsy
- Individuals who have difficult to control seizure disorders
- Individuals with Parkinson's Disease, neuromuscular disorders
- Elderly
- Individuals with skeletal deformities such as severe (kypho)scoliosis
- Individuals with a collagen disease affecting the esophagus (scleroderma, polymyositis)
- Premature infants
- Individuals who are marginally compromised and are put on new medications that have adverse side effects (e.g., psychotropic drugs, anticholinergics, anticonvulsant medications, medications for spasticity, any medication causing lethargy, calcium channel blockers, theophylline)
- Individuals who have (or have a history of) esophageal lesions or cancers
- Recumbent positioning
- Increased abdominal tone
- Constipation
- Individuals who steal food
- Individuals who eat too fast

Clinical Implications:

Morbidity (illness)

Recurrent respiratory infections, changes in pulmonary status
Inadequate hydration, leading to problems with blood electrolytes, lethargy, worsening constipation
Inadequate nutrition (malnutrition) leading to compromised health status
Inability to take medications properly (e.g., seizure control)
Esophageal changes (esophageal stricture, Barrett's esophagus, esophageal cancer)

Mortality (death)

What triggers the need for an investigation?

- It's helpful when the individual can communicate verbally or otherwise.
- Sometimes the desire to please or fear can interfere.
- Sometimes there is a delay in recognition of the problem.

Signs & Symptoms:

- Coughing, choking, cyanosis when eating or drinking
- Crying, tearing, irritability while eating or drinking
- Rales, stridor, wheezing, or congestion ("gurgling") during or after eating or drinking
- Obvious difficulty chewing or swallowing
- Obvious discomfort, pain, fear, or distress while eating or drinking (e.g., feeling of food getting stuck)
- Abnormal head/body positioning (especially backward arching at head/neck)
- Food/meal refusal (sometimes related to unfamiliar staff)
- Food spillage
- Fatigue with eating
- Recurrent emesis (may be behavioral but may be a symptom of GI discomfort or of constipation)
- Emesis during or after meals (including self-induced vomiting)
- Vomiting of blood or "coffee-ground" material
- Nasal reflux or regurgitation
- Excessive salivation or mucus production, difficulty handling secretions
- Rumination
- Recurrent respiratory infections/aspiration pneumonias
- Weight loss, chronic underweight status, or inadequate weight gain
- Persistent or recurrent dehydration
- Low grade fevers or spiking fevers of unknown cause
- Unexplained anemia (iron deficiency anemia when there has been sufficient blood loss or inadequate iron intake)
- Chronic pharyngitis, laryngitis
- Behavior problems around mealtime
- Evidence of interstitial fibrosis on chest x-ray
- Decreased serum protein, albumin, prealbumin levels

Evaluation:

- History and Physical exam
- Lab – CBC, chemistries, stool for blood, emesis for blood, x-rays
- Occupational or swallowing therapy assessment via history, exam, mealtime evaluation, videofluoroscopy
- History – previous x-rays, pulmonary pathology)
- Exam of oral structures, facial symmetry, muscle tone, dentition, tongue movements, lip and jaw closure, method of processing food, drooling)
- Videofluoroscopic assessment of:
    - pharyngeal structure, symmetry, delay, seepage, residue, timing and swallow, aspiration or penetration)

  - esophageal structure/abnormalities, motility/peristalsis (primary/secondary/tertiary waves), lower esophageal esophageal sphincter relaxation/patency, GE reflux, hiatal hernia
  - gastric structure, motility, emptying
- Consultation with gastroenterologist who may elect to do:
  - esophageal manometry (pressure measurements)
  - esophagogastroduodenoscopy (looking into the esophagus, stomach, and duodenum, taking biopsies, looking for H. pylori)
  - esophageal pH probe

Treatment:

General Treatment: Avoid constipation. If it is a problem, treat it (adequate hydration, fiber, other dietary measures, medications, avoid medications that cause it or worsen it)

Treatment for Dysphagia & Esophageal Dysmotility:

- Diet texture changes, thickening of liquids
- Feeding techniques
- Thermal stimulation
- Physical management – positioning
- NG tube (short term when cause is self-limited or responsive to other treatment)
- G-tube

G-tubes

- short-term or long-term
- for supplemental use or total nutrition/hydration
- types – open surgical, PEG
- indications: > 2months with an NG tube, documented aspiration or aspiration pneumonia, protracted feeding times, failure of more conservative treatment, esophageal obstruction or dysfunction (Note that GERD is not on this list)
- risks post-op: wound infection, hemorrhage, malposition of tube, granulation tissue, pressure necrosis of the abdominal or gastric wall, diarrhea, pneumoperitoneum, gastrocolic or gastroenteric fistula, migration of the tube, enlargement of the stoma, aspiration pneumonia
- contraindications: gastric outlet obstruction, severe intractable gastroparesis, noncompliance
- benefits: convenient, easy to maintain and use, natural use of GI tract, improved nutritional and hydration status, medication administration

G-tubes do not solve other GI problems such as GERD, aspiration of oral secretions, gastroparesis.

Treatment for GERD:

Positioning measures: Elevate head of bed
Remain upright after meals/snacks

Avoiding predisposing factors: overeating, bedtime snacks, high fat foods, smoking, alcohol, medications that make it worse, foods that make it worse (high-fat, peppermint, chocolate, high acid content, caffeine), tight clothing over the abdomen, posture that increase intraabdominal pressure

Medications:

- Antacids
- H2 blockers (H2 receptor antagonists)–Tagamet, Zantac, Pepcid, Axid
  - side effects – rare – headache, lethargy, confusion, depression, hallucinations, hepatitis, hematological toxicity
  - medication interaction – primarily with Tagamet – theophylline, Coumadin, Dilantin, Lidocaine
- Proton pump inhibitors – Prilosec, Prevacid, Aciphex, Nexium, Protonix
  - side effects – gynecomastia, myopathy, rashes, interstitial nephritis, concern about bacterial overgrowth and gastric tumor with long-term use
  - medication interaction – Valium, Coumadin, Dilantin, Digoxin, Prednisone
- Prokinetic agents - Reglan
- Ulcer adherents - Carafate

Esophageal dilatation for stricture

Surgical Intervention:

- Suggested criteria: persistence or recurrence of symptoms or complications after 8-12 weeks of intensive acid suppression therapy, increased esophageal exposure to gastric acid evident on 24-hour pH monitoring, documentation of a mechanically defective LES on manometry
- Factors to consider: strength of propulsive movements, anatomic shortening of esophagus (e.g., hiatal hernia), symptoms suggestive of duodenogastric reflux, hypersecretion of gastric acid, delayed gastric emptying
- Most common procedure: Nissen fundoplication (open vs. laparoscopic)
  - Contraindications to laparoscopic repair: remedial repair, need for other procedure that cannot be done by laparoscopy, incisional abdominal hernia that also needs repair
  - Relative contraindications: obesity, large hiatal hernia

Important factors to remember when dealing with GI issues in the DD population:

- Be proactive – early detection, evaluation, and intervention
- Monitor the weight record, intervening early
- Provide education/inservicing of staff, especially about the physical/nutritional management plan (PNMP)
- Advise direct care staff:

  - Don't leave individuals at risk unattended during meal preparation or at mealtimes.
  - Be consistent in following the individual's PNMP
  - Don't change diet texture without permission.
  - Maintain proper positioning at all times, in all situations
  - Report any concerns to nurse or supervisory person.
- Monitor the PNMP for consistency, effectiveness, need for modification, user-friendliness (easy to put into place, keep clean), comfort, enhancement of feelings of security.
- Periodically review the medication regimen to see if any may be exacerbating the problem.
- Watch for trends.