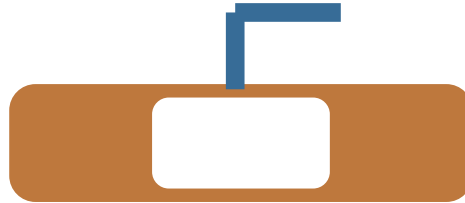


Atresia, Fistulae, and Duplications

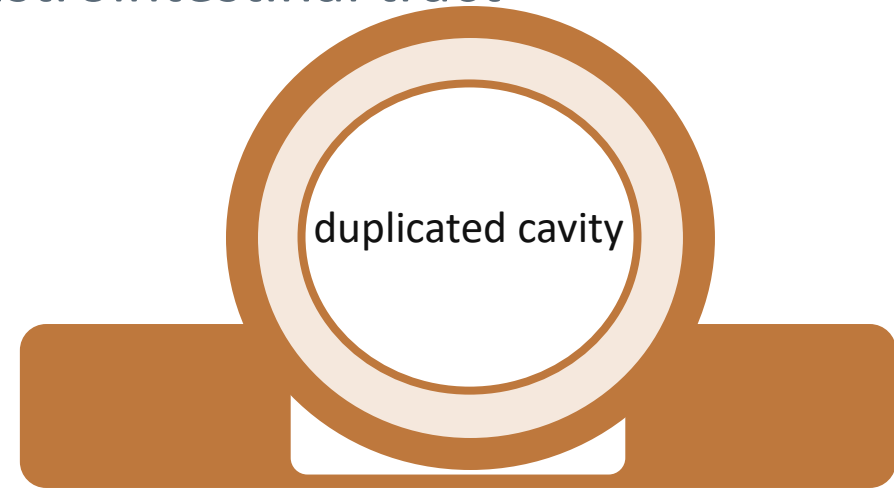
Congenital structural abnormalities of the gastrointestinal tract



Atresia



Fistula



Duplication

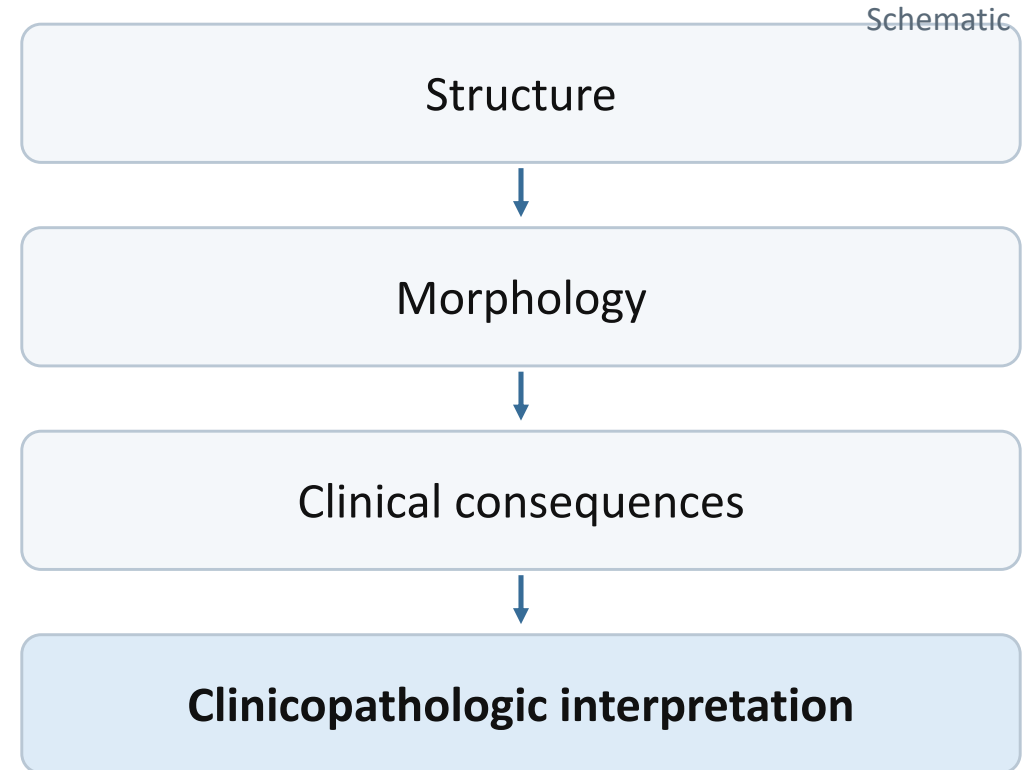
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Learning objectives

- Distinguish atresia, stenosis, fistula, and duplication anatomically.
- Explain neonatal symptoms from obstructed GI continuity or airway communication.
- Recognize major esophageal and intestinal patterns and their complications.
- Apply morphologic criteria and imaging logic to alimentary duplications.



Congenital GI defects: Three structural problems

Schematic

- Atresia: Segmental loss of a patent lumen.
- Fistula: Abnormal connection between epithelial-lined structures.
- Duplication: Extra alimentary segment, often a cyst sharing a gut wall.

ATRESIA

Discontinuity

FISTULA

Abnormal connection

DUPLICATION

Additional segment

Anatomic pattern predicts clinical consequences

Schematic

- Luminal interruption causes impaired transit and upstream distension.
- Airway communication creates a route for aspiration or gas entry.
- A duplicated segment may compress bowel or develop mucosal complications.

Lumen interrupted → retention



Airway connection → aspiration

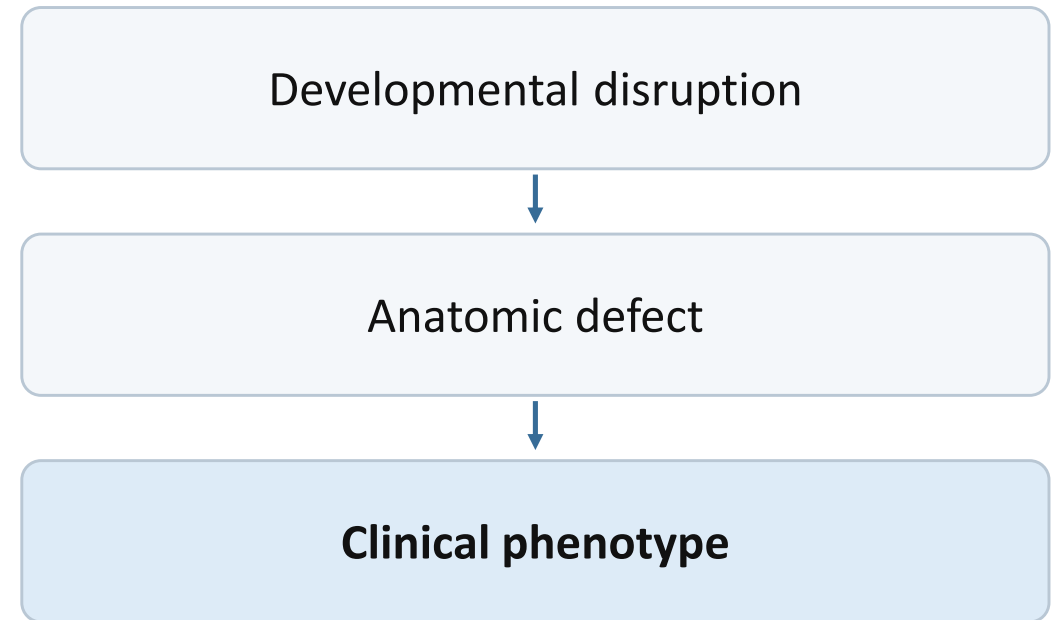


Duplicated wall → mass or ulceration

Embryogenesis: One insult can affect several organs

Schematic

- Congenital anomalies vary with developmental process and timing.
- Concurrent organ development explains associated abnormalities.
- A GI birth defect should trigger a directed extraintestinal assessment.



Atresia versus stenosis

Schematic

- Atresia is an interruption of normal lumen formation or continuity.
- Stenosis retains a narrowed passage and may obstruct partially or completely.
- Degree and location of blockage determine timing and severity.

Atresia: No patent continuity



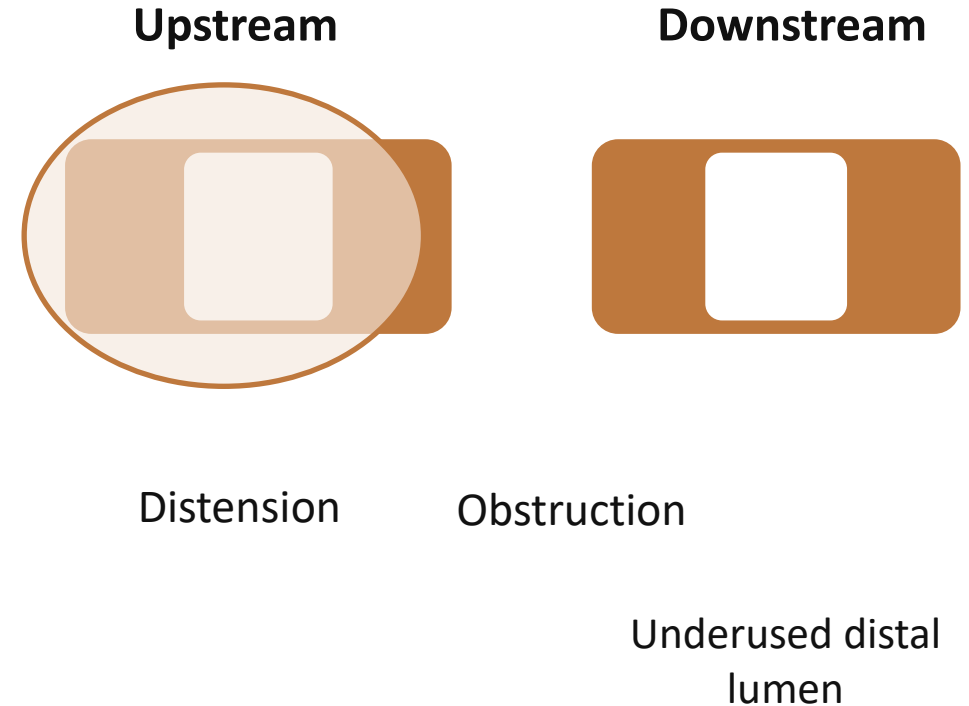
Stenosis: Narrowed lumen



Complete versus partial obstruction

Schematic

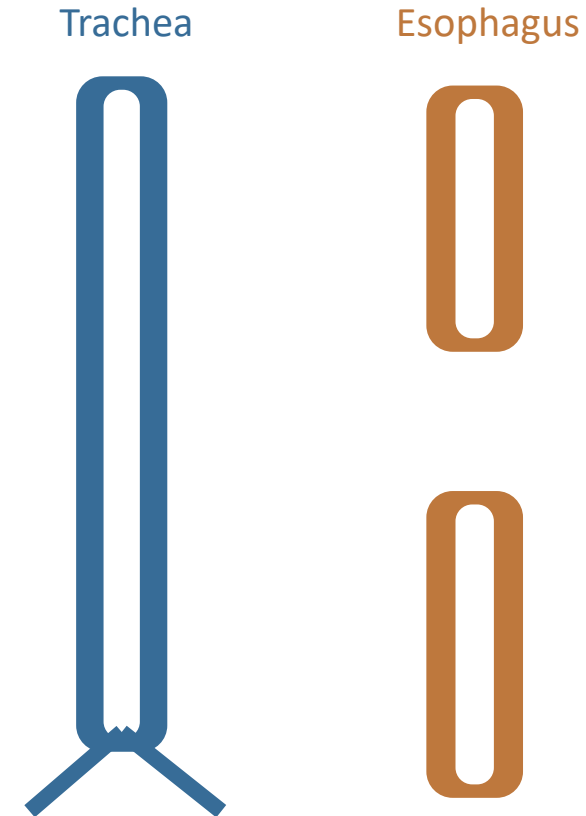
- Complete obstruction stops passage across the affected level.
- Partial obstruction permits some transit but causes retention and distension.
- Clinical expression depends on site, residual caliber, and feeding.



Esophageal atresia: Core morphology

Schematic

- A segment of the developing esophagus is absent or noncanalized.
- A blind-ending proximal pouch prevents food reaching the stomach.
- A thin noncanalized cord may connect the esophageal ends.



No fistula • discontinuous esophagus

Agnesis, atresia, and stenosis are distinct

Schematic

- Agnesis: Extreme absence of esophageal formation; Very rare.
- Atresia: Discontinuous lumen with blind pouch or pouches.
- Stenosis: Residual lumen with reduced caliber.

Atresia: No patent continuity



Stenosis: Narrowed lumen



Robbins frequency estimate for esophageal atresia

Schematic

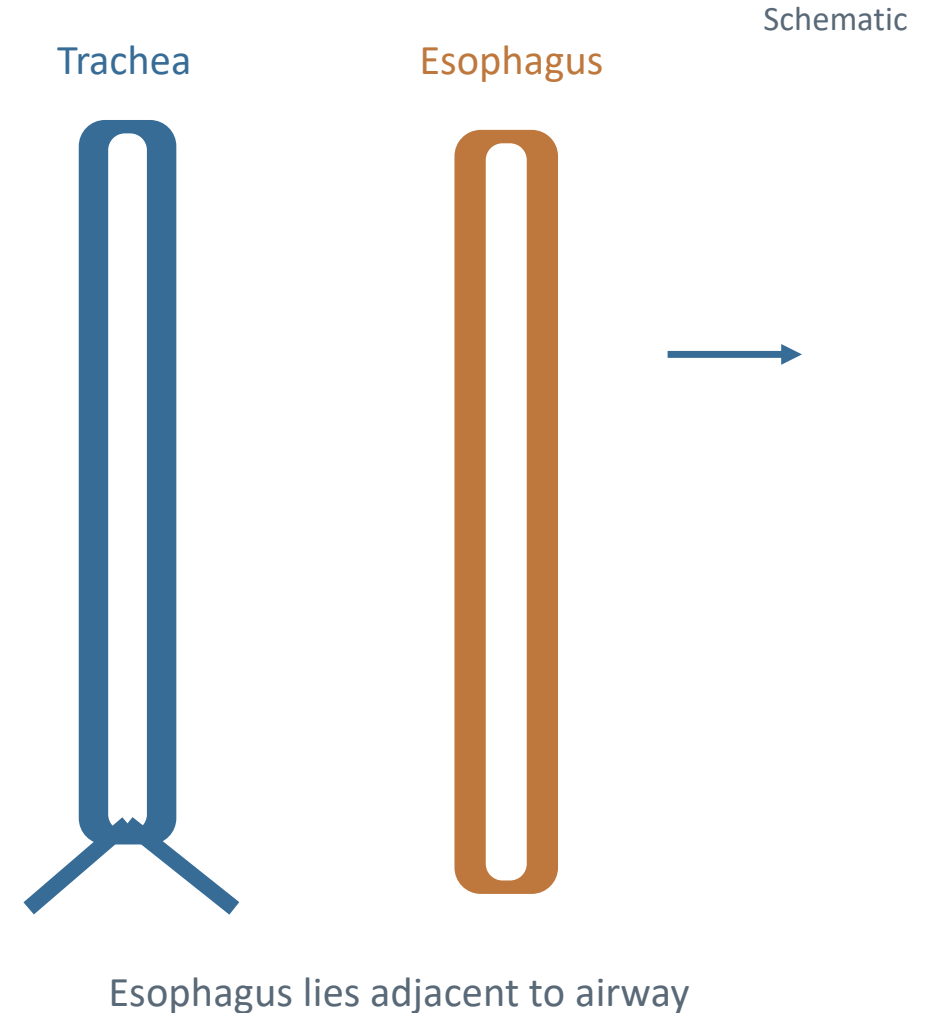
- Esophageal atresia is reported in 1–5 infants per 10,000 live births.
- This frequency refers to atresia, not all congenital GI defects.
- Anatomical subtype and associated anomalies shape clinical risk.

1–5

Per 10,000 live births

Relation to the tracheal bifurcation

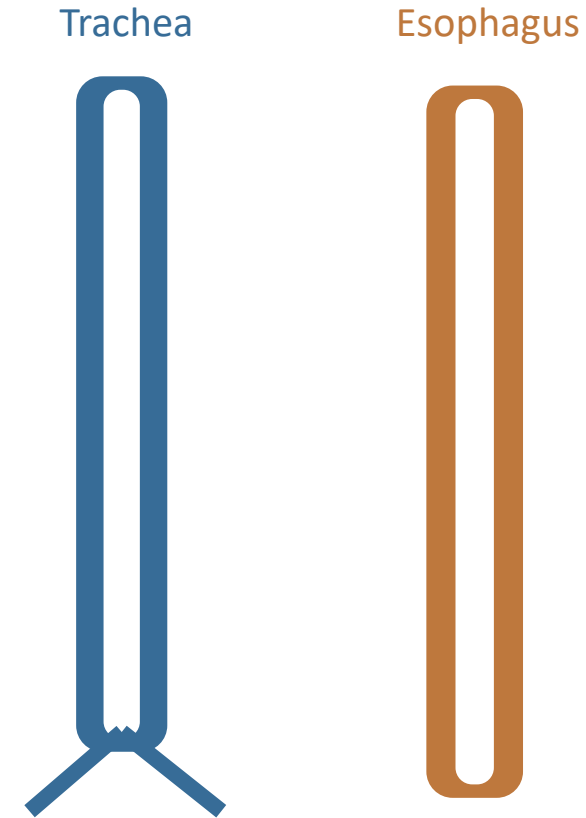
- The atretic segment most often occurs at or near the tracheal bifurcation.
- Adjacent esophageal and respiratory structures permit fistulous connections.
- The exact fistula arrangement changes both air and feeding pathways.



Normal esophagus–trachea relationship

Schematic

- Esophagus and trachea are separate adjacent conduits.
- Food should pass toward the stomach without entering the airway.
- The airway is not normally connected to the esophageal lumen.

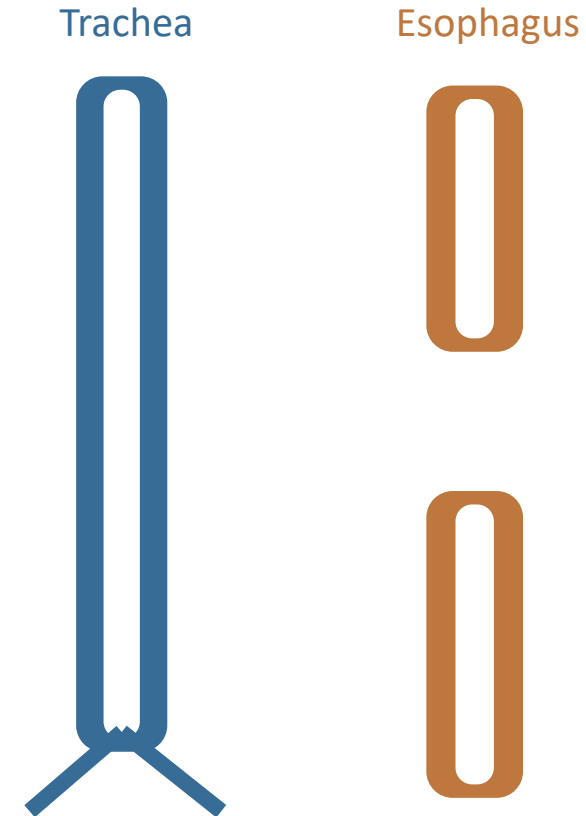


Two separate continuous conduits

Isolated atresia: No airway fistula

Schematic

- Upper and lower esophageal segments end blindly.
- No communication with the trachea is depicted.
- Obstructed saliva and feeds accumulate in the proximal pouch.

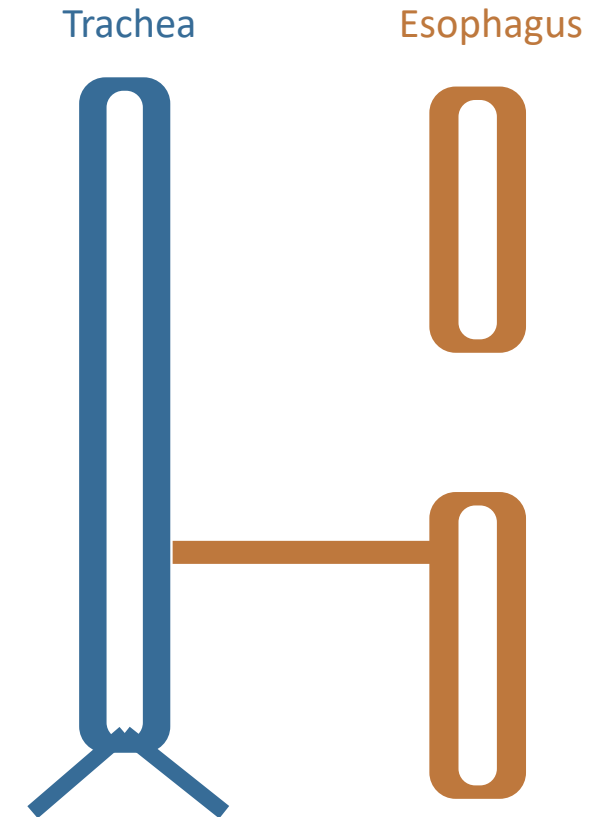


No fistula • discontinuous esophagus

Distal fistula: Atresia with airway connection

Schematic

- The proximal esophagus ends blindly.
- The distal esophageal segment communicates with the trachea.
- This is the most common Robbins-depicted configuration.

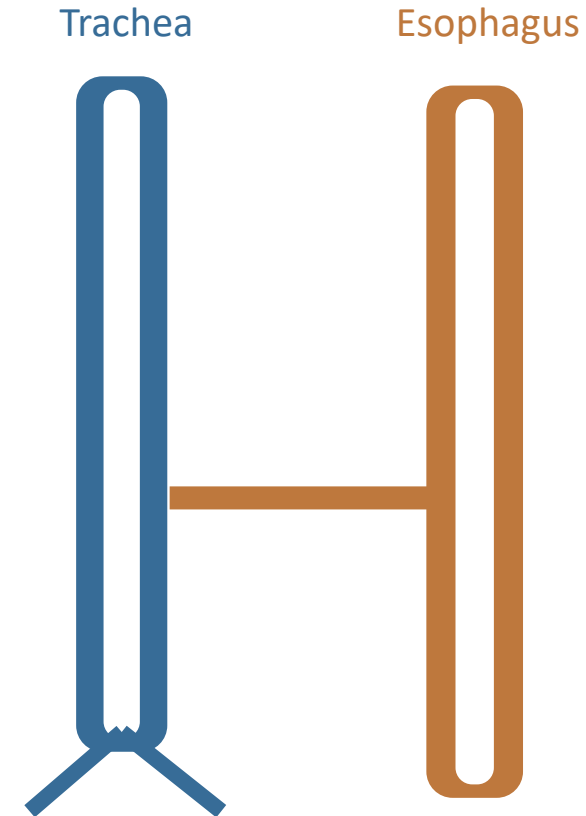


Blind proximal pouch • distal fistula

Isolated fistula: Esophagus remains continuous

Schematic

- The esophageal lumen remains continuous.
- A communication connects esophagus and trachea.
- Feeding-associated airway symptoms may occur despite luminal continuity.



Continuous esophagus • communicating fistula

Five Gross-classification anatomic types

Schematic

- Isolated atresia; Proximal fistula; Distal fistula; Double fistula.
- Isolated fistula without atresia (often termed H-type).
- Identify proximal and distal communications rather than memorizing letters alone.

A Atresia only

B Proximal fistula

C Distal fistula

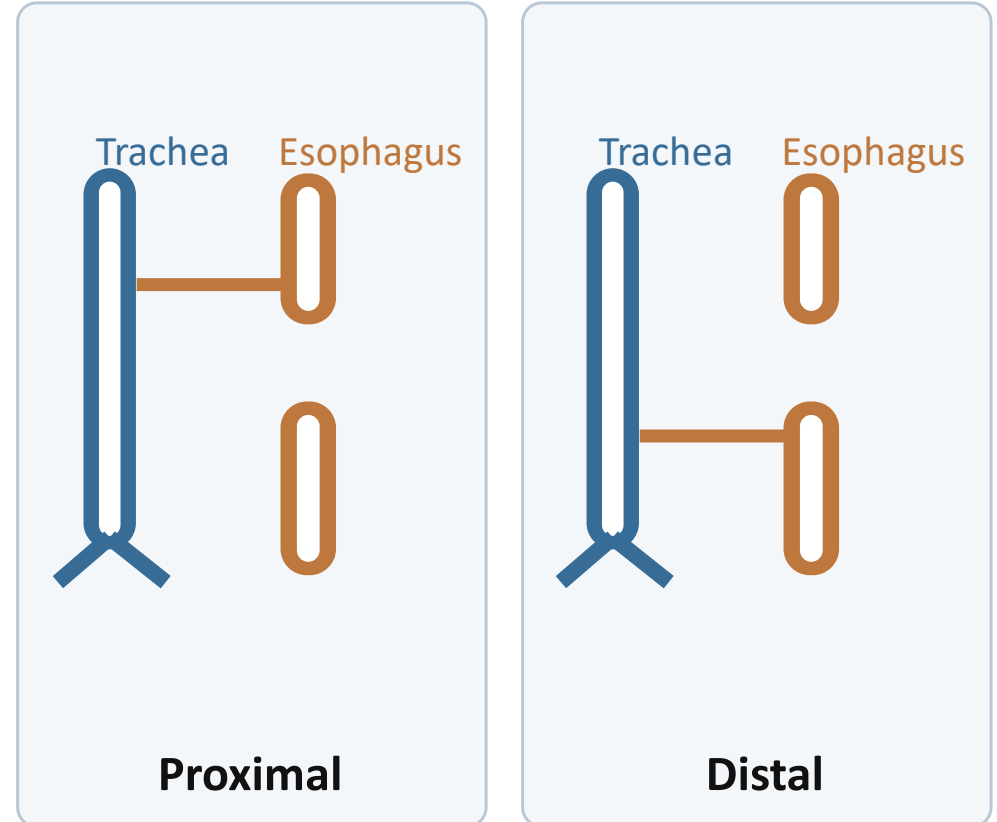
D Two fistulae

E Isolated fistula

Proximal versus distal fistula: Why it matters

Schematic

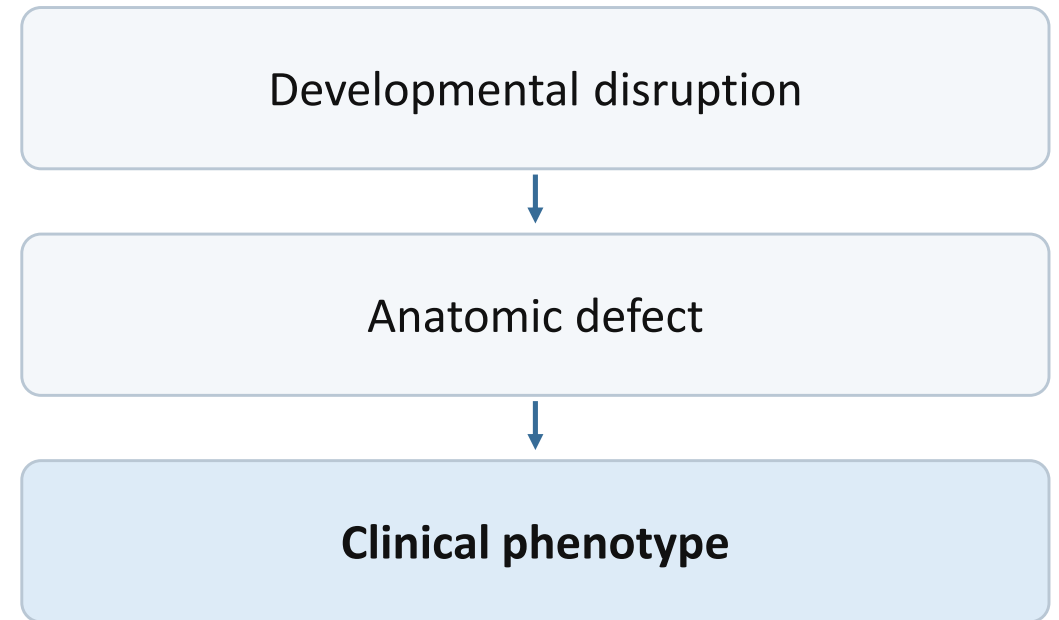
- A proximal fistula offers a direct route for pooled upper-pouch secretions.
- A distal fistula can conduct tracheal gas toward stomach and bowel.
- Both abnormal pathways create respiratory and nutritional hazards.



Foregut separation and abnormal communications

Schematic

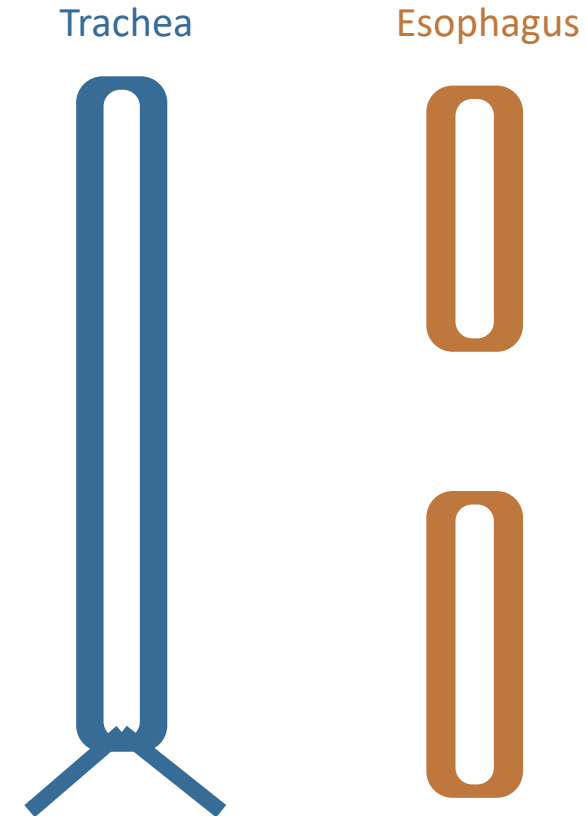
- Trachea and esophagus develop from the foregut.
- Abnormal separation or development is implicated in EA–TEF.
- The precise embryologic sequence varies across anomalies.



Why newborns regurgitate during feeding

Schematic

- An atretic proximal pouch cannot deliver swallowed material distally.
- Saliva or milk returns to the oropharynx.
- Regurgitation shortly after birth is a classic Robbins clue.

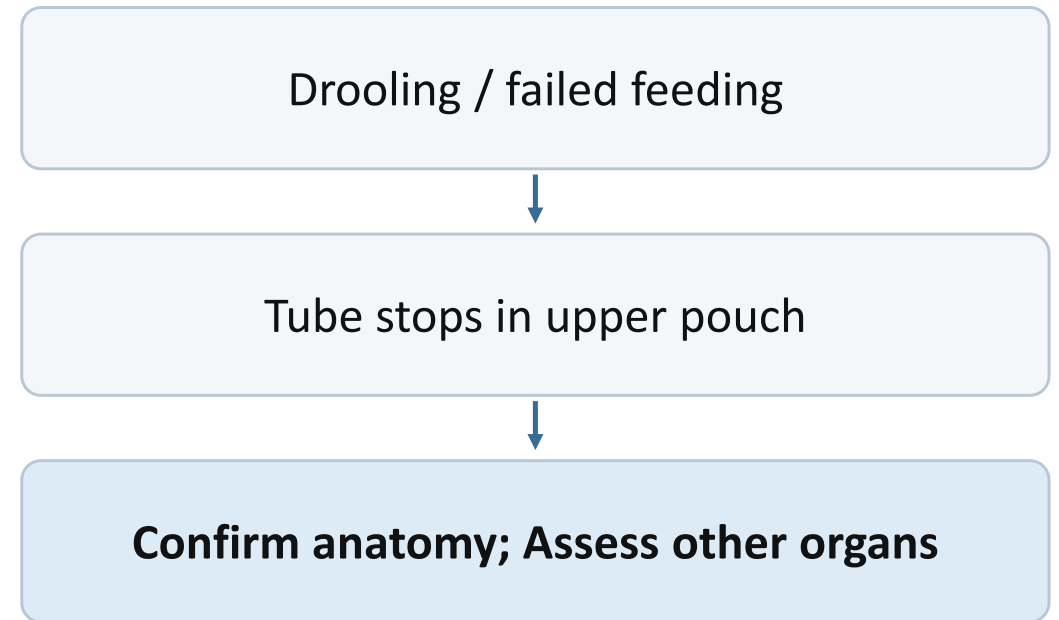


No fistula • discontinuous esophagus

Neonatal recognition: Saliva, choking, cyanosis

Schematic

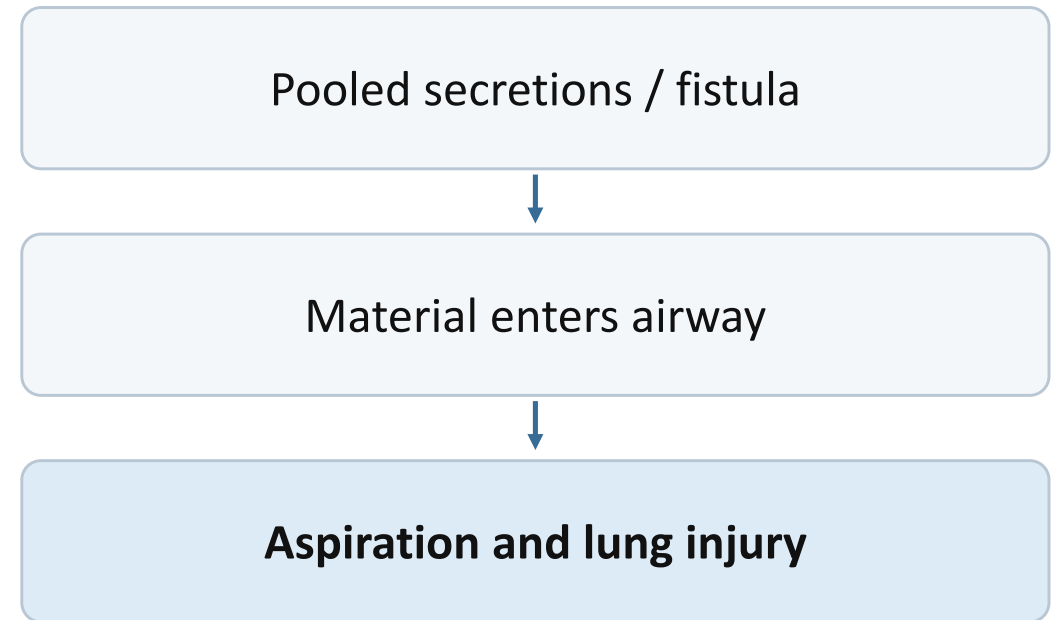
- Excessive drooling indicates pooled secretions.
- Coughing or choking during feeds suggests esophageal or airway pathology.
- Failure to pass an orogastric tube raises suspicion for atresia.



Aspiration and pneumonia: An anatomic mechanism

Schematic

- Pooled secretions may enter the upper airway.
- A tracheoesophageal connection adds a second aspiration route.
- Repeated aspiration can precipitate respiratory distress or pneumonia.



Respiratory deterioration may be rapid

- Suffocation and aspiration as major hazards.
- Milk, saliva, and gastric contents threaten the lungs through different routes.
- An apparent feeding problem may therefore be an airway emergency.

Airway risk

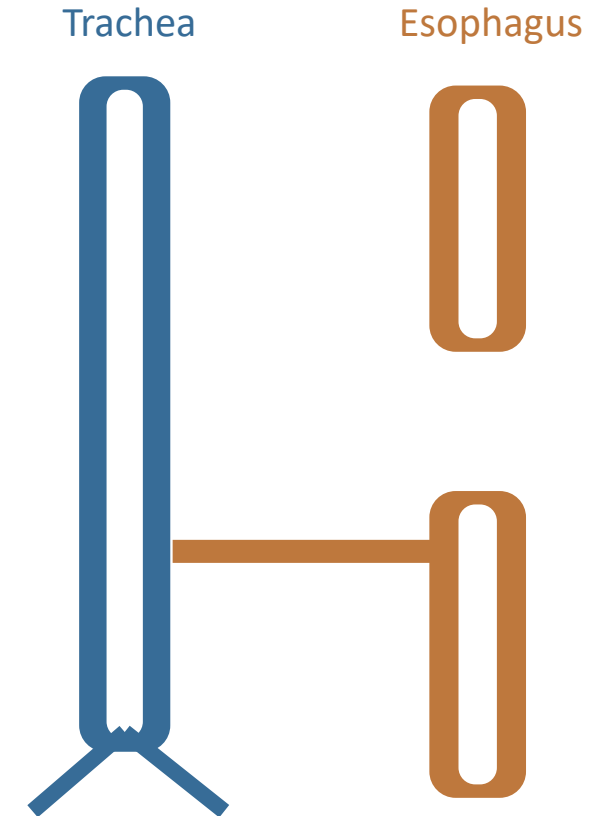
Aspiration • choking

Pneumonia • suffocation

Distal fistula and gastric air

Schematic

- Air can pass from the trachea through a distal fistula.
- Gastric and bowel gas may persist despite proximal esophageal atresia.
- This distinguishes many distal-fistula cases from isolated atresia.

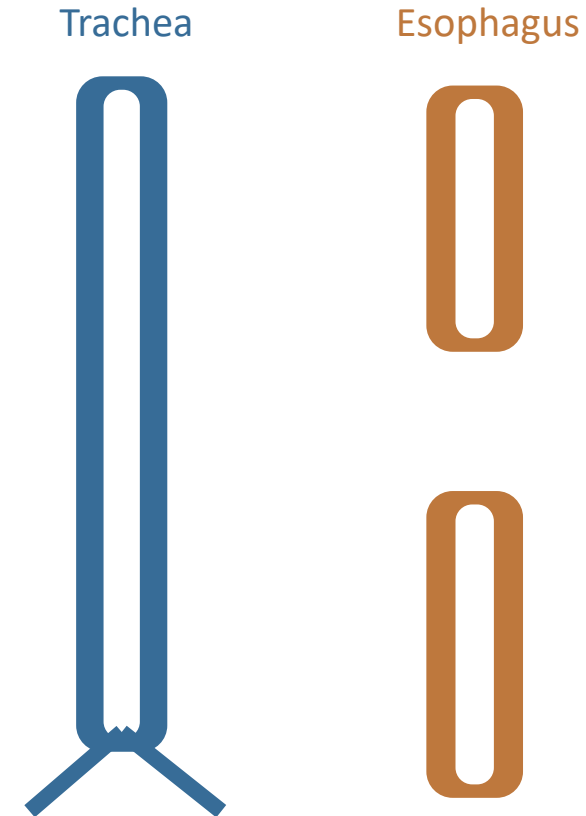


Blind proximal pouch • distal fistula

Isolated atresia and a gasless abdomen

Schematic

- Without a distal fistula, the airway cannot directly aerate the stomach.
- Absent distal abdominal gas can support isolated esophageal atresia.
- Interpret bowel gas with anatomy and clinical findings; Not alone.

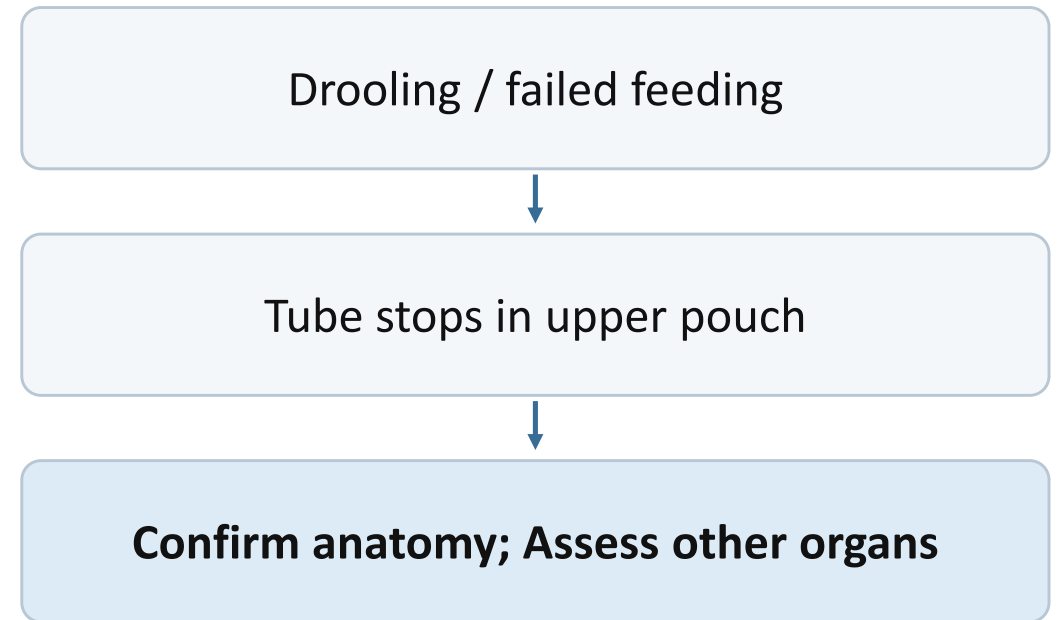


No fistula • discontinuous esophagus

Orogastric tube: What the image means

Schematic

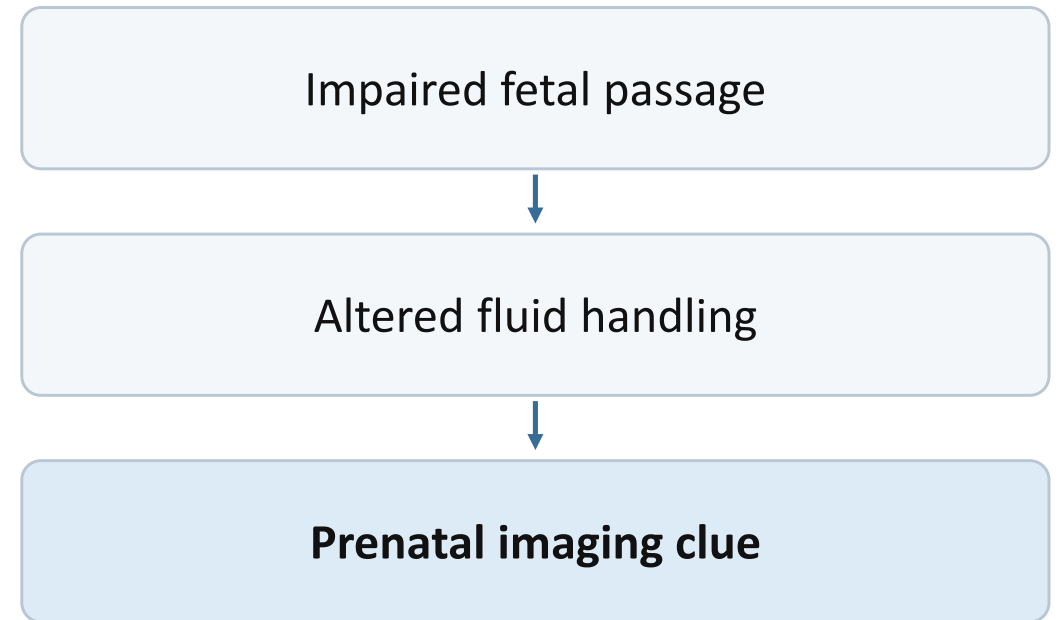
- The tube fails to reach the stomach when the upper pouch ends blindly.
- Radiography can show the tube coiled in the upper pouch.
- Distal intestinal gas provides an additional anatomical clue.



Prenatal clue: Polyhydramnios

Schematic

- Obstructed fetal swallowing may reduce normal fluid absorption.
- Excess amniotic fluid is a prenatal clue, not a specific diagnosis.
- Postnatal anatomic evaluation remains necessary.



VACTERL: Evaluate beyond the esophagus

Schematic

- Vertebral, Anal, Cardiac, Tracheoesophageal, Renal, Limb defects.
- Esophageal anomalies with this multisystem pattern.
- Associated structural disease may alter immediate surgical planning.

Vertebral

Anal

Cardiac

Tracheoesophageal

Renal

Limb

TACRD: Another associated malformation pattern

Schematic

- Tracheal agenesis/atresia and complex cardiac abnormalities.
- Radial ray defects and duodenal atresia complete the acronym.
- This association reinforces the need to survey multiple organs.

Vertebral

Anal

Cardiac

Tracheoesophageal

Renal

Limb

Additional anomalies

Schematic

- Heart, lungs, and great vessels may be affected.
- Genitourinary structures and anus may also be abnormal.
- An isolated-looking foregut lesion does not exclude major coexisting defects.

Heart

Lungs

Great vessels

Kidneys

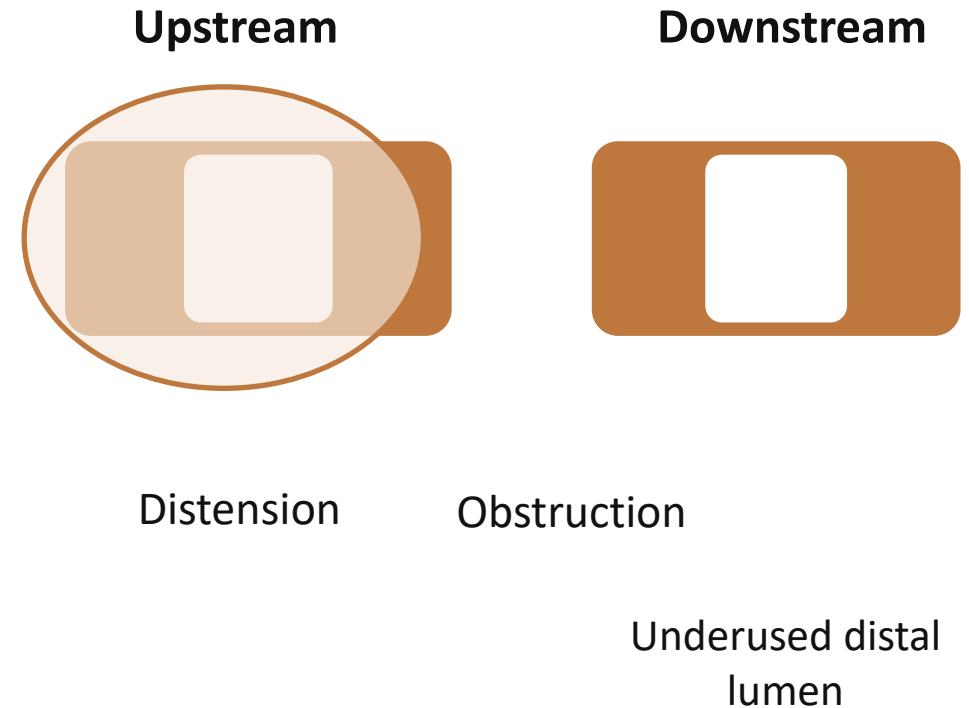
Anus

Limbs

Imperforate anus: Related developmental context

Schematic

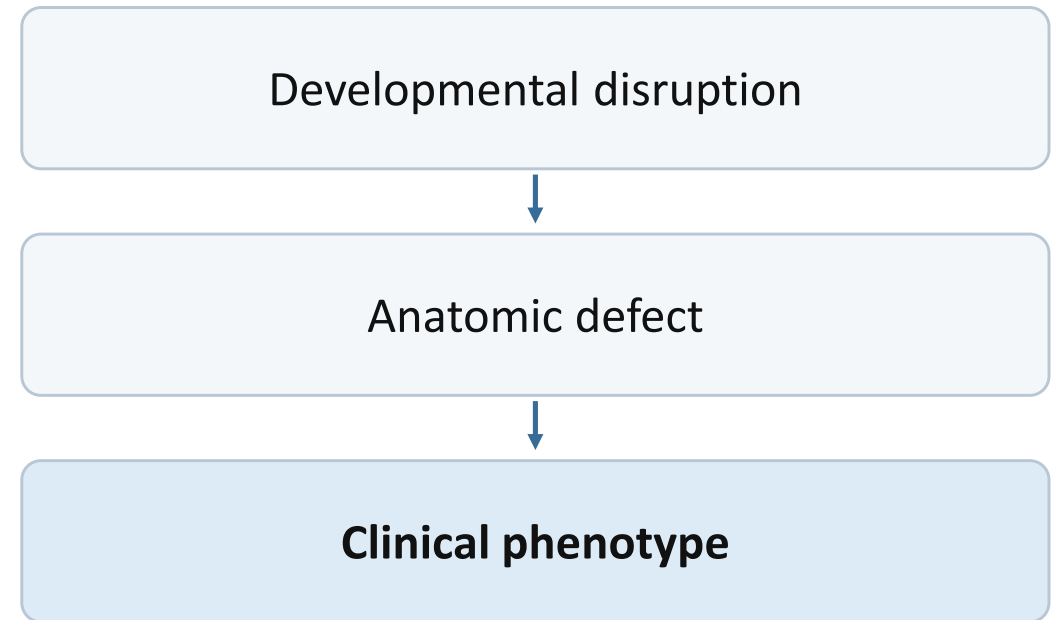
- Robbins relates imperforate anus to failure of the cloacal diaphragm to involute.
- Anorectal outlet obstruction is anatomically separate from esophageal atresia.
- Perineal inspection forms part of congenital anomaly assessment.



Fluid and electrolyte disturbances

Schematic

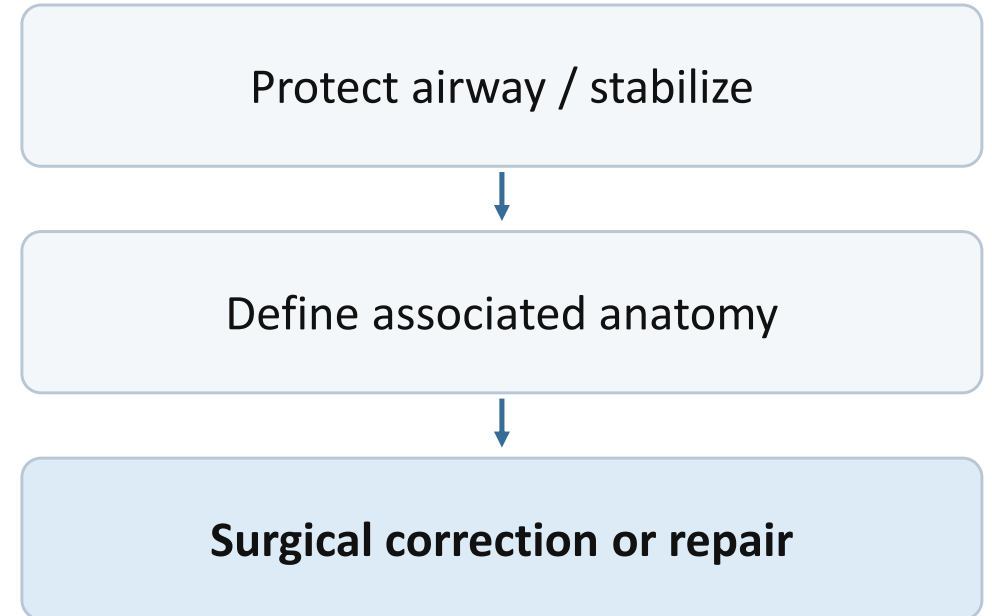
- Ineffective feeding threatens intake and hydration.
- Regurgitation and vomiting increase gastrointestinal losses.
- Severe fluid and electrolyte imbalance as a risk.



Immediate care logic: Protect the airway

Schematic

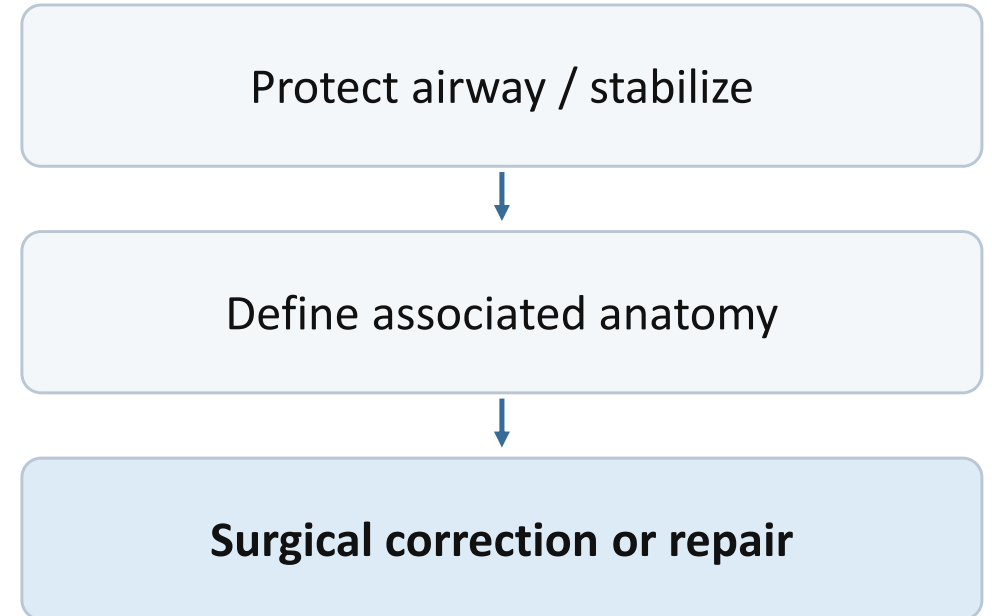
- Suspected EA–TEF requires urgent neonatal surgical assessment.
- Avoid oral feeding while airway and anatomy are evaluated.
- Protect the lungs and stabilize hydration before definitive repair.



Surgical correction: The anatomic objectives

Schematic

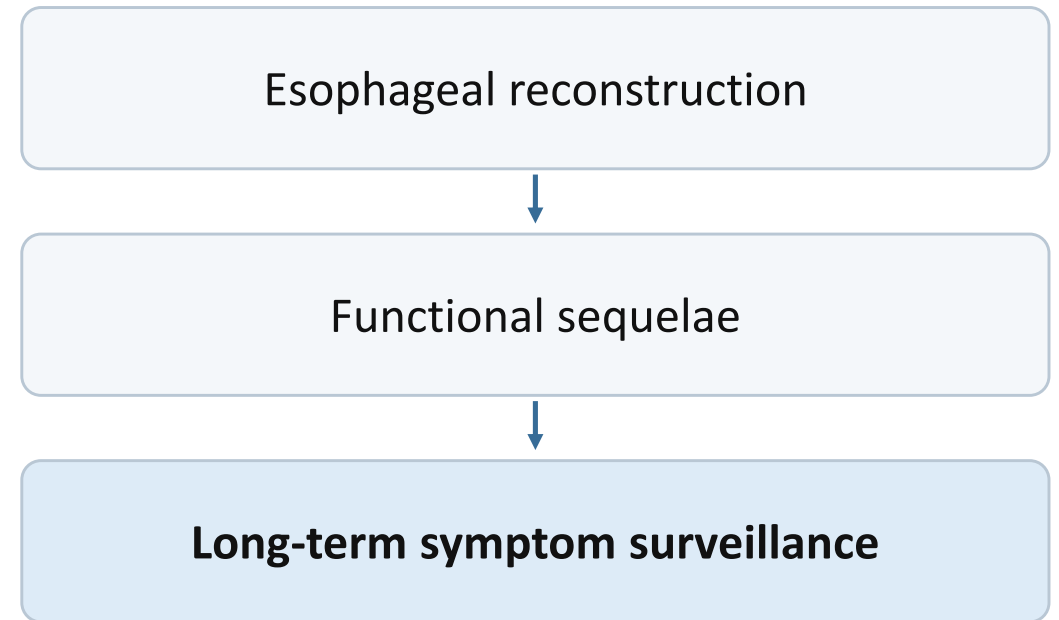
- Re-establish a usable esophageal passage when feasible.
- Divide and close the abnormal communication with the airway.
- Identify and address concurrent major malformations.



Repair does not end gastrointestinal surveillance

Schematic

- Esophageal dysmotility and feeding difficulties may persist.
- Gastroesophageal reflux and anastomotic narrowing are recognized sequelae.
- Long-term follow-up is relevant beyond the neonatal period.



Postrepair stricture: Clinicopathologic relevance

Schematic

- Anastomotic narrowing may produce dysphagia or food retention.
- Long-gap repair, leaks, and reflux can contribute.
- Distinguish postoperative stricture from congenital stenosis.

Atresia: No patent continuity



Stenosis: Narrowed lumen



Congenital stenosis: Wall narrowing

Schematic

- A markedly reduced lumen with fibrous wall thickening.
- Luminal compromise can be incomplete or severe.
- Unlike atresia, some passage may remain.

Atresia: No patent continuity



Stenosis: Narrowed lumen



Acquired stenosis: Four etiologic pathways

Schematic

- Chronic gastroesophageal reflux can produce inflammatory scarring.
- Irradiation and caustic injury can lead to fibrosis.
- Systemic sclerosis is another described setting.

Chronic reflux

Caustic injury

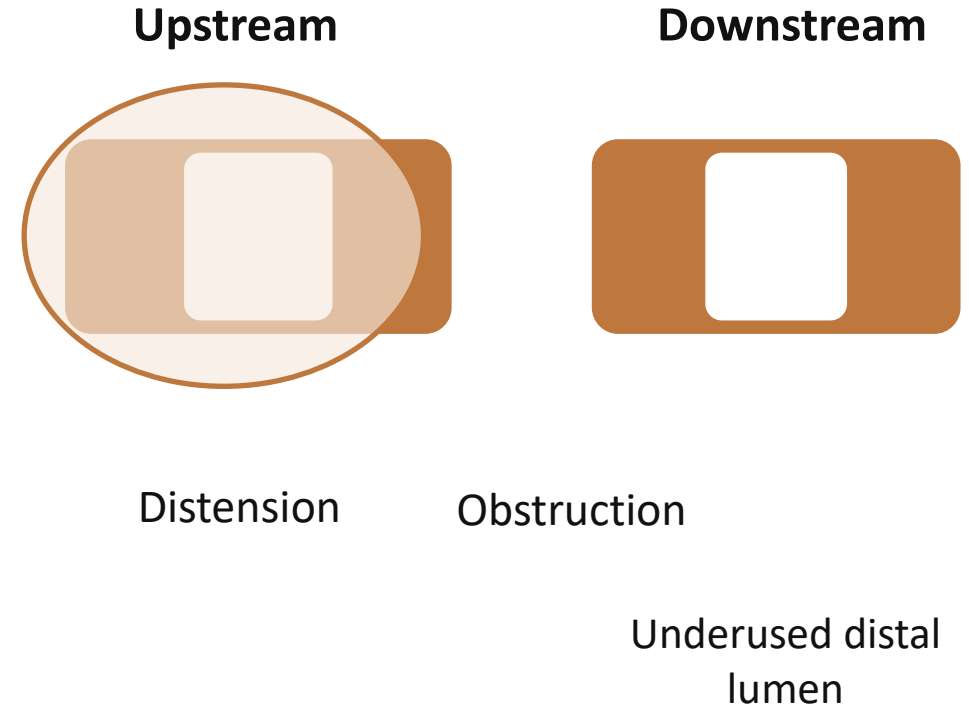
Irradiation

Systemic sclerosis

Why esophagus and small intestine are vulnerable

Schematic

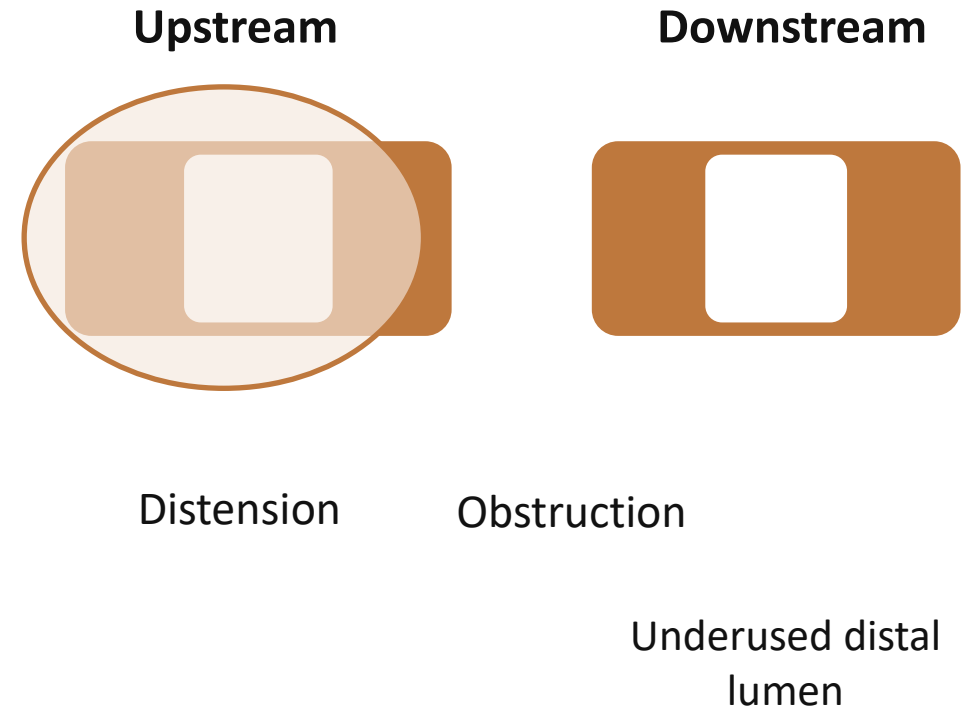
- Stenosis may occur anywhere in the GI tract.
- Clinically significant narrowing in narrower lumens.
- Even modest wall thickening may compromise transit.



Intestinal atresia: A second obstructive phenotype

Schematic

- Congenital intestinal lumen is interrupted at an intestinal level.
- Symptoms usually relate to retained content proximal to the block.
- Duodenal and jejunoileal atresias have different common developmental explanations.



Duodenal atresia: Failure of canalization

Schematic

- Normal embryonic duodenal lumen formation requires recanalization.
- Failure can result in a complete luminal block.
- The obstruction often presents early with vomiting.

Proximal duodenum



Distal duodenum

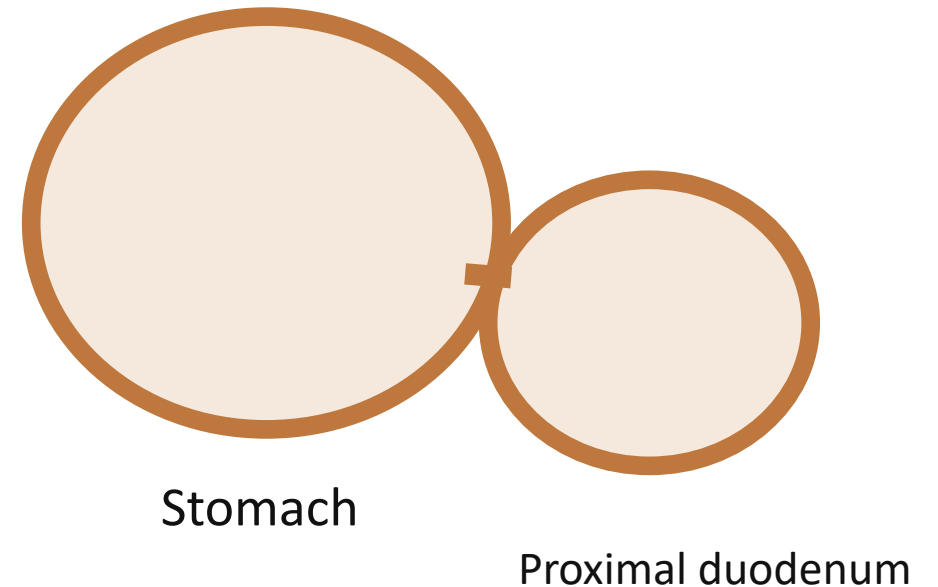


Lumen interrupted in the duodenal segment

Duodenal obstruction: The double-bubble pattern

Schematic

- Two dilated gas-filled compartments represent stomach and proximal duodenum.
- Little or no distal bowel gas favors complete obstruction.
- The sign describes anatomy, not a pathognomonic cause.



Two gas-filled proximal compartments

Double bubble: Important differential diagnoses

Schematic

- Duodenal atresia and duodenal web or stenosis.
- Annular pancreas or extrinsic bands with duodenal obstruction.
- Malrotation-associated obstruction must remain in the differential.

Duodenal atresia

Duodenal web

Annular pancreas

Malrotation / bands

Duodenal atresia and other congenital anomalies

Schematic

- Down syndrome is a recognized association.
- Cardiac and additional intestinal or rotational defects may coexist.
- An associated anomaly survey is clinically relevant.

Heart

Lungs

Great vessels

Kidneys

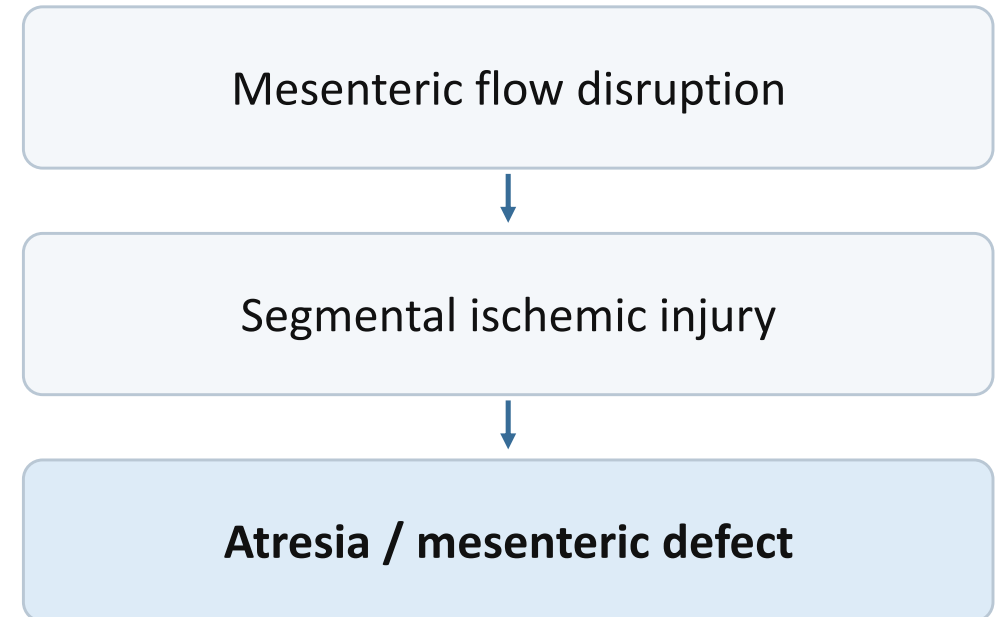
Anus

Limbs

Jejunioileal atresia: Vascular disruption model

Schematic

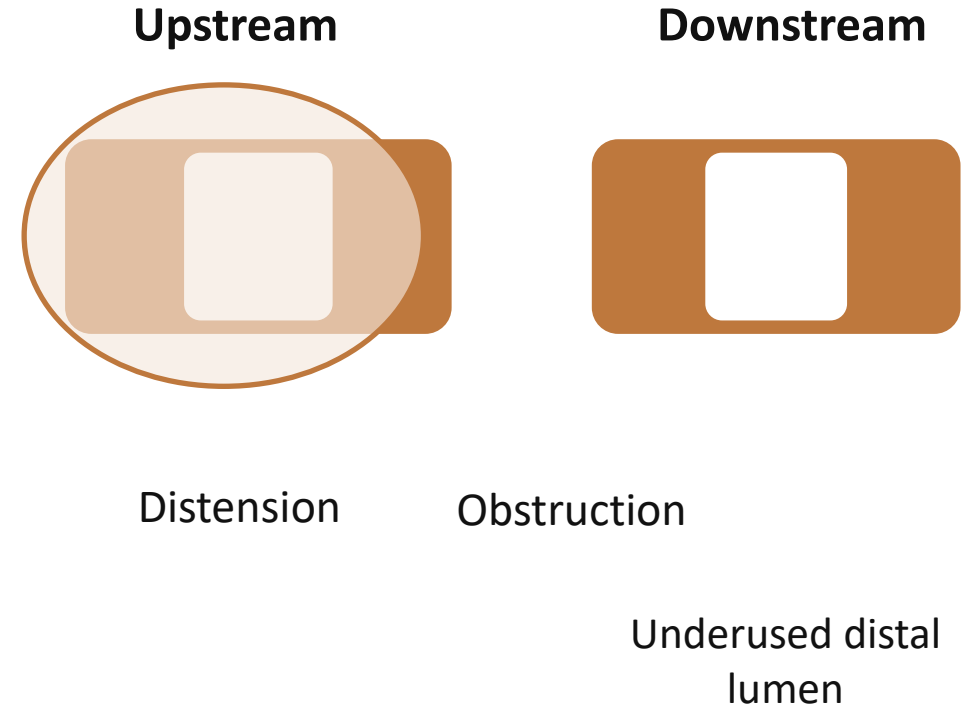
- Intrauterine mesenteric vascular compromise is a major explanation.
- Ischemic loss may interrupt a previously formed intestinal segment.
- The model does not necessarily explain every familial or complex case.



Proximal and distal bowel changes

Schematic

- Proximal bowel becomes distended by retained contents.
- Distal bowel may remain narrow or underused.
- Difference in caliber matters for surgical reconstruction.



Jejunioileal atresia Type I

Schematic

- An internal mucosal or membranous obstruction blocks the lumen.
- External intestinal serosal continuity is maintained.
- There is no associated mesenteric defect in the classical description.



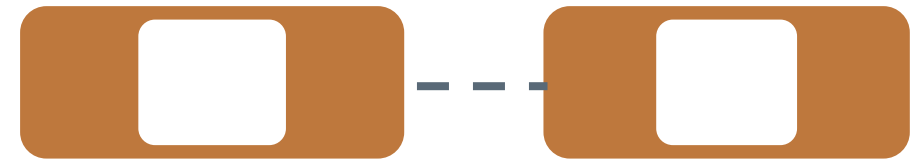
Intact outer bowel wall

Obstructing intraluminal membrane

Types II and IIIa: Interruptions and mesentery

Schematic

- Type II: Blind ends joined by a fibrous cord.
- Type IIIa: Discontinuity with a V-shaped mesenteric defect.
- The mesenteric anatomy helps distinguish these forms.



Type II: Fibrous cord

Type IIIa adds a mesenteric gap

Types IIIb and IV: Complex small-bowel atresia

Schematic

- Type IIIb: “apple-peel” configuration with an abnormal remaining mesentery.
- Type IV: Multiple atretic segments.
- Bowel-length implications are important in these forms.

IIIb: Abnormal coiled residual bowel

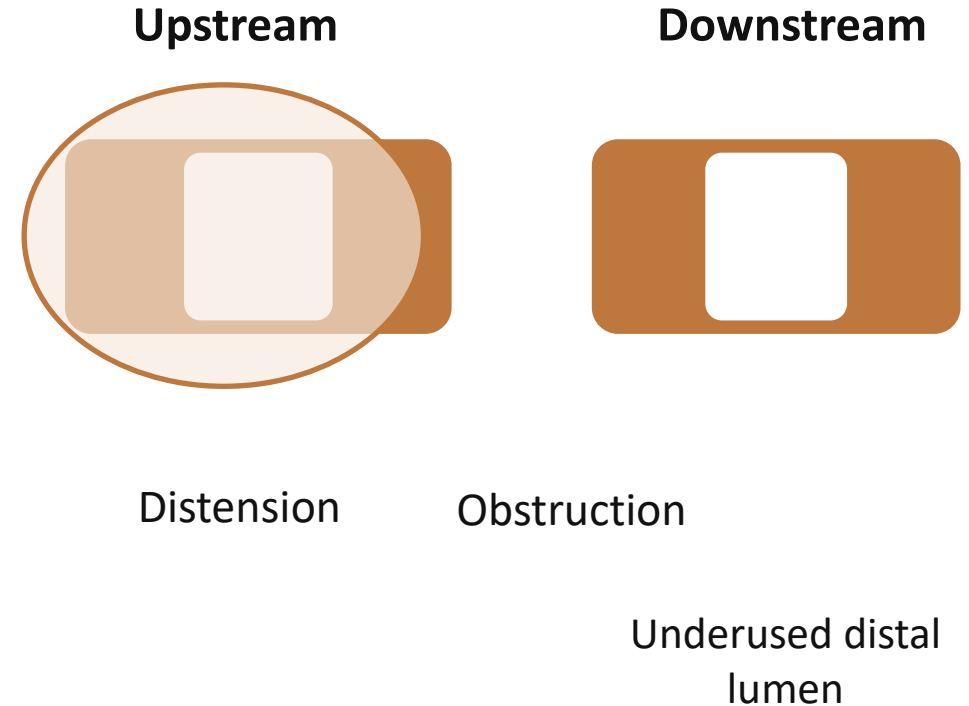


IV: Multiple interrupted segments

Neonatal intestinal obstruction: Clinical clues

Schematic

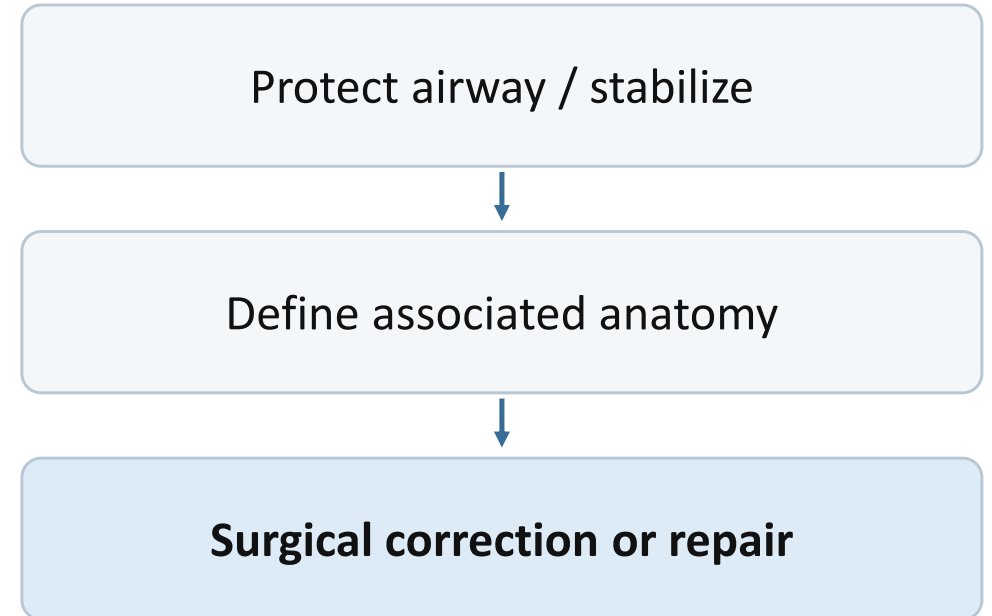
- Bilious vomiting may signal obstruction beyond entry of bile into duodenum.
- Abdominal distension and delayed passage of stool may be present.
- The anatomical level influences clinical timing and distension.



Initial evaluation of suspected intestinal atresia

Schematic

- Stabilize the infant and limit enteral accumulation.
- Use imaging to identify level and likely mechanism of obstruction.
- Pediatric surgery evaluates definitive anatomic correction.



Other causes of neonatal obstruction

Schematic

- Malrotation with volvulus may mimic a congenital obstructive lesion.
- Meconium ileus and meconium plug syndrome have distinct mechanisms.
- Colonic atresia and anal atresia are more distal possibilities.

Atresia

Malrotation / volvulus

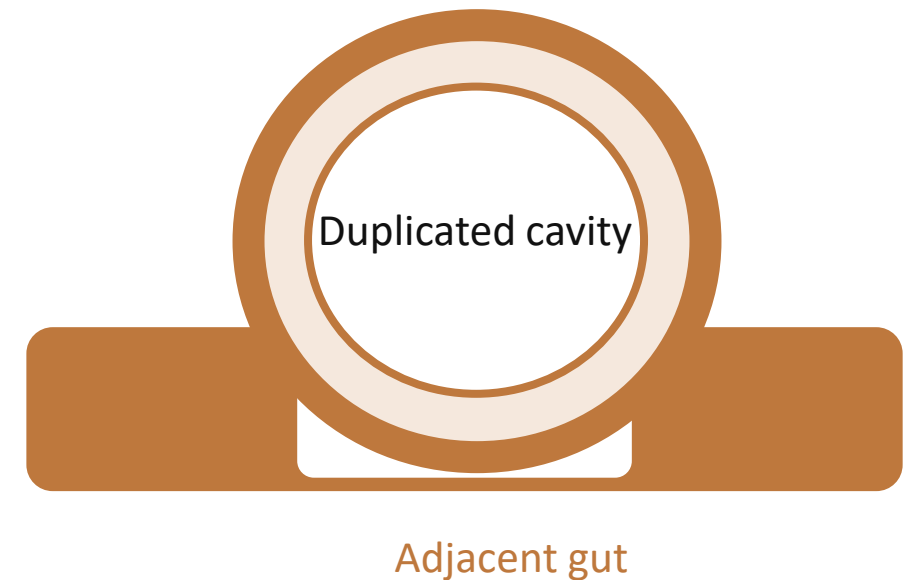
Meconium obstruction

Anal outlet defect

Alimentary duplication: Definition

Schematic

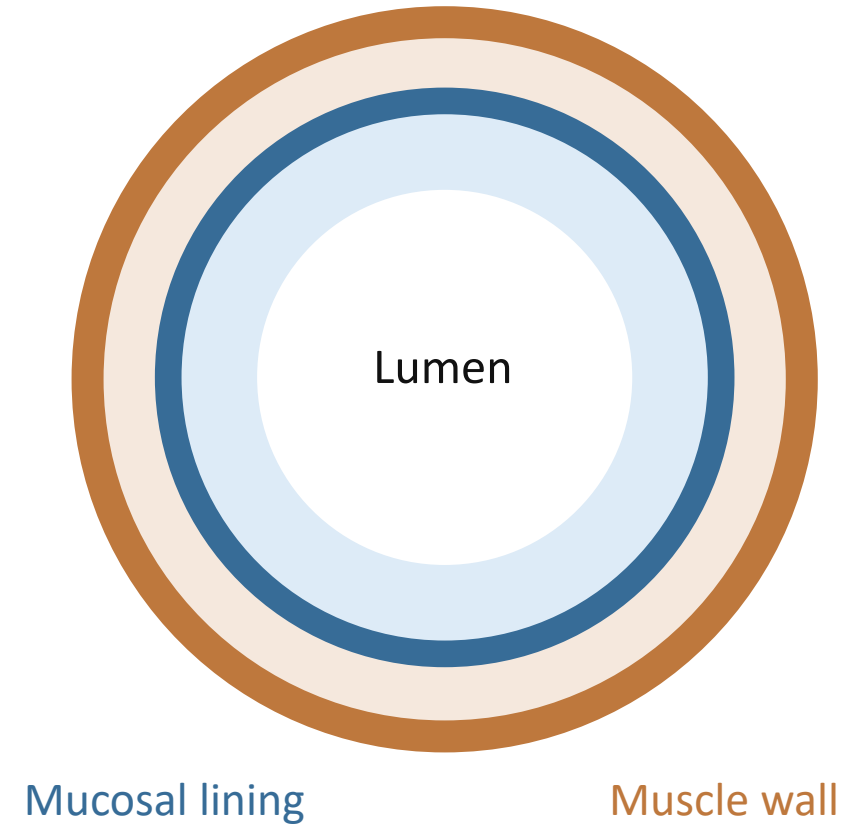
- A congenital extra segment lies in close association with gut.
- It commonly shares a smooth-muscle wall with the native segment.
- The lesion may be cystic or tubular.



The histologic triad of duplication

Schematic

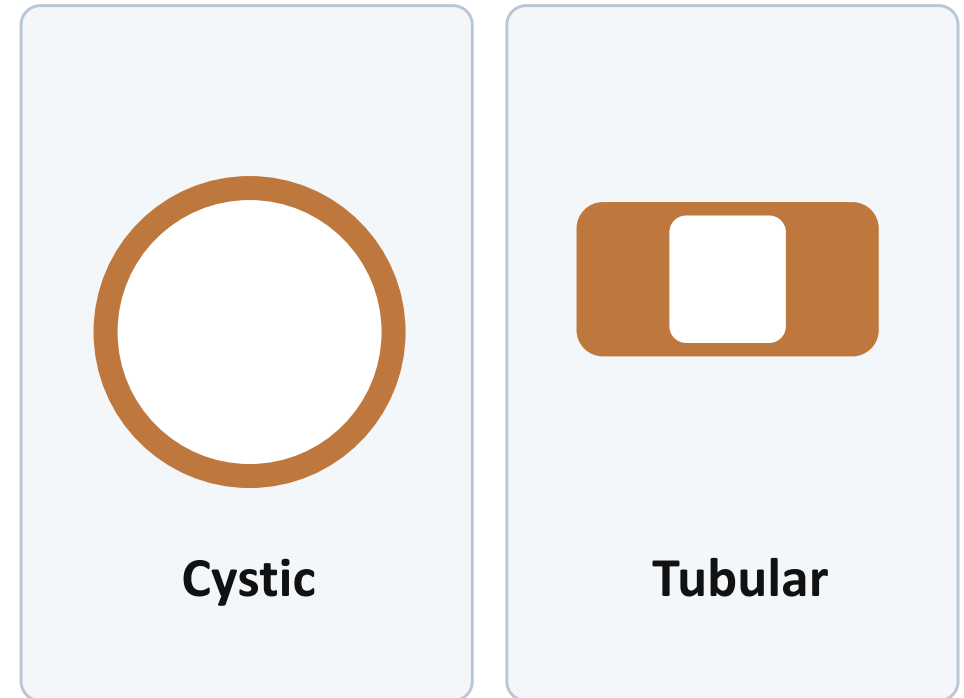
- Epithelial lining of alimentary-type mucosa.
- A defined smooth-muscle component in its wall.
- Intimate relation to adjacent gastrointestinal tract.



Cystic and tubular duplications

Schematic

- Cystic lesions commonly do not communicate with bowel lumen.
- Tubular duplications may connect to the parent bowel.
- Communication alters filling, symptoms, and surgical anatomy.

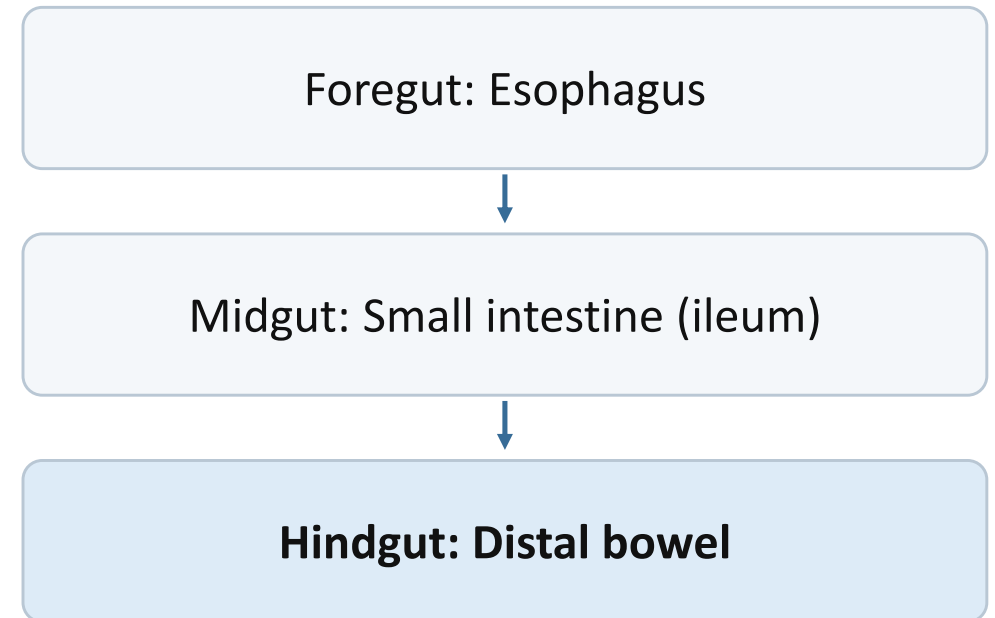


Configuration governs luminal communication

Where duplications arise

Schematic

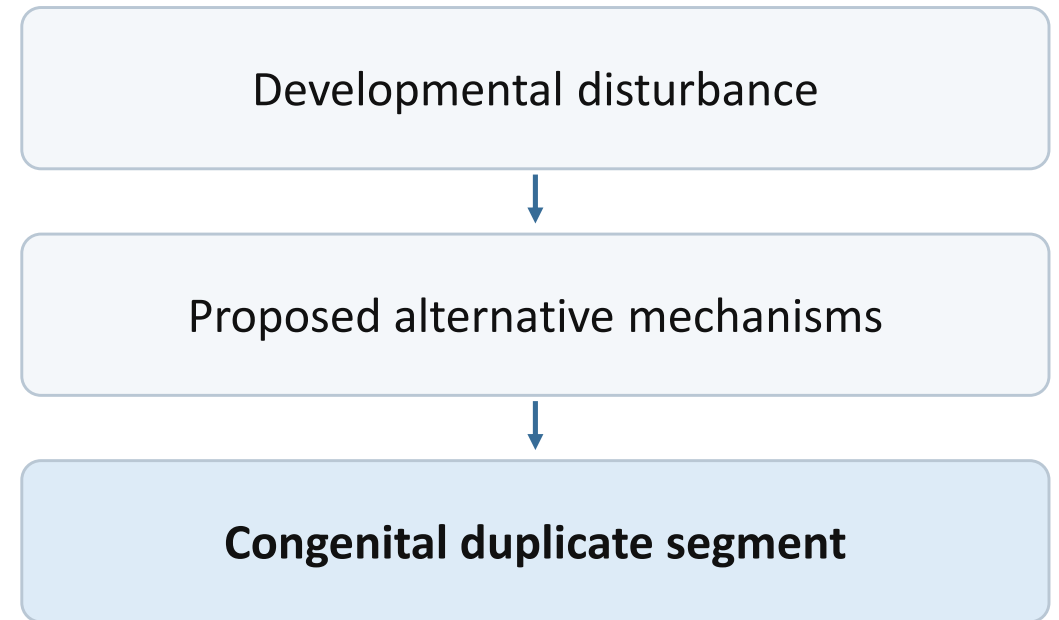
- Alimentary duplication can occur along the gastrointestinal tract.
- The ileum and esophagus are commonly described sites.
- Symptoms depend more on location and relationships than on name alone.



Embryogenesis of duplication: Multiple theories

Schematic

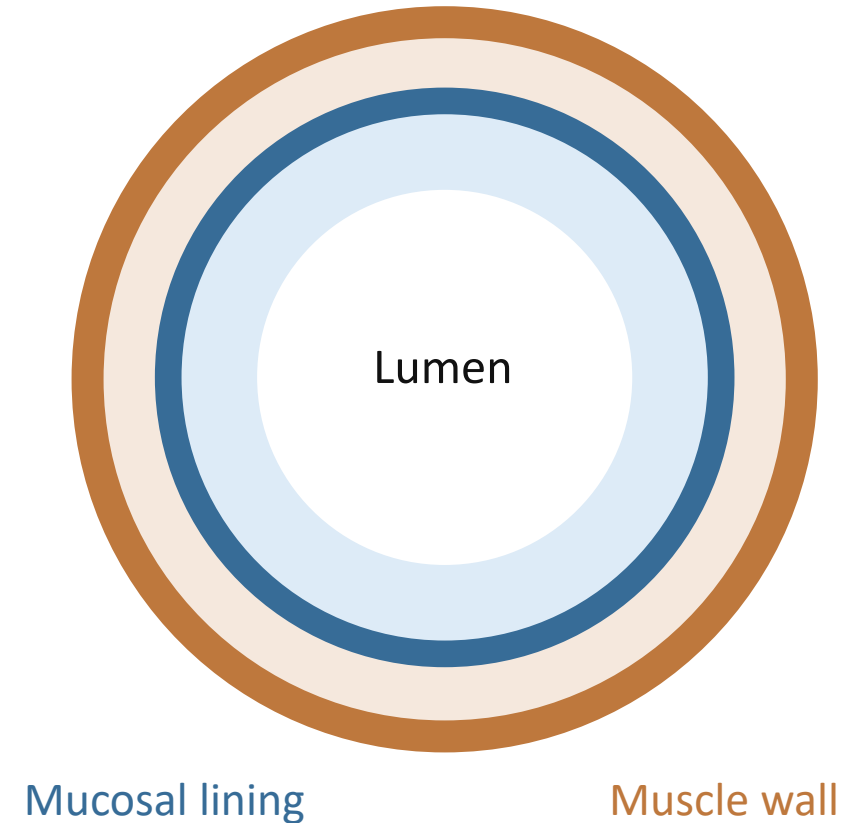
- Proposed mechanisms include abnormal recanalization and abortive twinning.
- Persistent embryonic diverticula and other theories have been described.
- No single hypothesis explains every duplication phenotype.



Duplication histology: Layers have meaning

Schematic

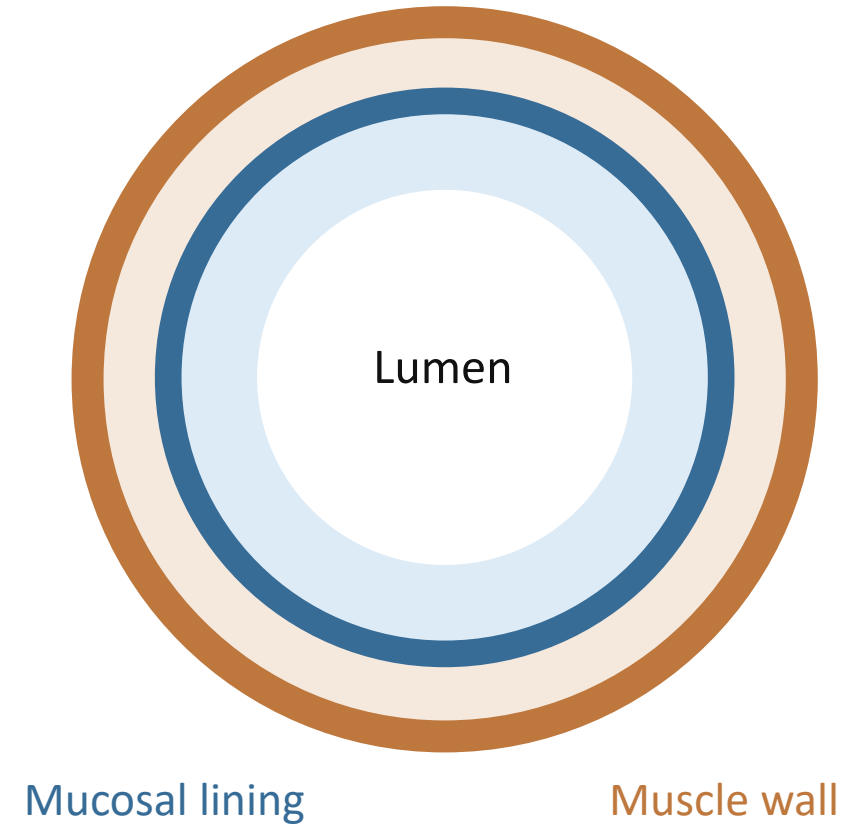
- Enteric-type epithelium lines the cyst or duplicated lumen.
- Smooth-muscle layers support an alimentary duplication diagnosis.
- Mucosal type may differ from the adjacent bowel segment.



Ectopic gastric mucosa: Why bleeding occurs

Schematic

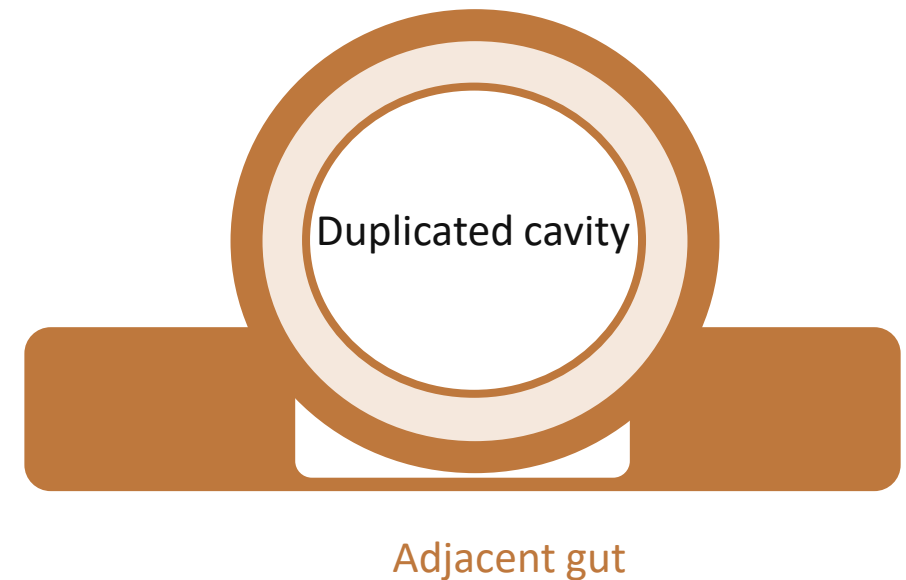
- Gastric-type mucosa may occur within an intestinal duplication.
- Acid secretion can ulcerate susceptible nearby mucosa.
- Hemorrhage is a consequence of mucosal biology, not simply mass effect.



Clinical consequences of duplication

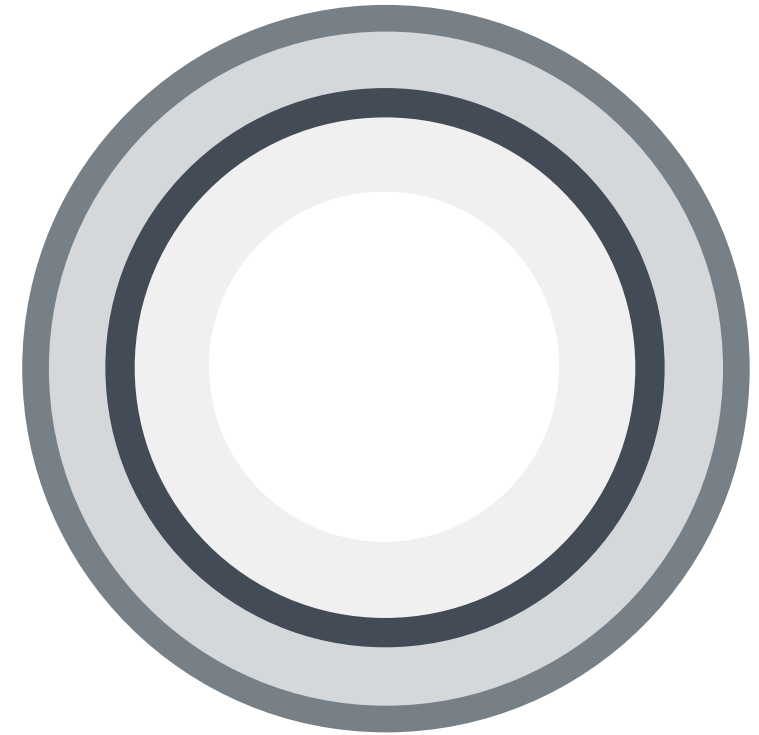
Schematic

- Mass effect can narrow or obstruct adjacent bowel.
- Intussusception, volvulus, and bleeding are recognized complications.
- Presentation ranges from infancy to incidental later detection.



Imaging clue: The gut-signature wall

- Ultrasonography may show inner echogenic mucosa and outer hypoechoic muscle.
- Layered architecture is suggestive of a duplication cyst.
- The sign has mimics; Pathology and anatomic context remain important.



Echogenic inner layer

Hypoechoic outer layer

Simplified ultrasound wall schematic

Duplication differential: Do not overcall cysts

Schematic

- Mesenteric or other abdominal cysts can resemble a duplication.
- Meckel diverticulum is a true bowel-wall diverticulum, not a closed duplication cyst.
- Location, communication, and wall histology separate diagnoses.

Duplication cyst

Meckel diverticulum

Mesenteric cyst

Other cystic lesions

Comparative pathology: Three structural phenotypes

Schematic

- Atresia primarily obstructs luminal continuity.
- Fistula creates abnormal inter-compartment communication.
- Duplication adds a second tissue-lined structure near native bowel.

ATRESIA

Discontinuity

FISTULA

Abnormal connection

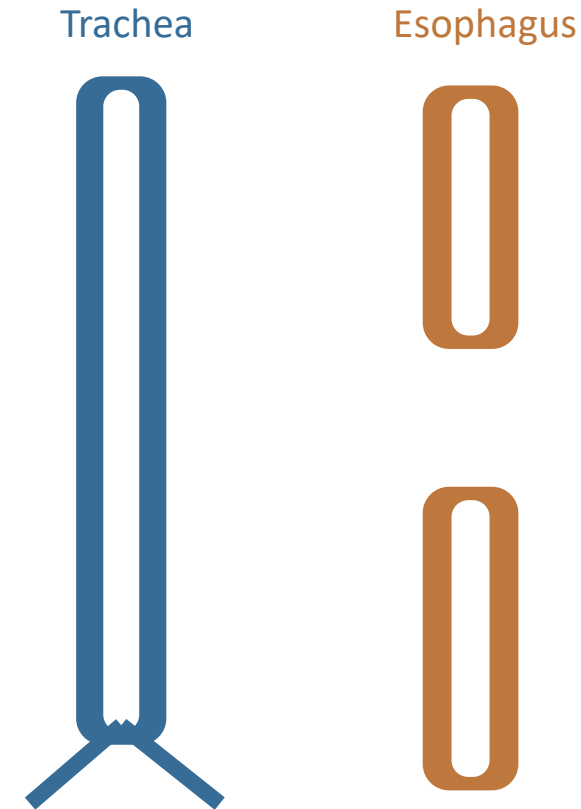
DUPLICATION

Additional segment

Case 1: Newborn with repeated choking during feeds

Schematic

- Excessive saliva and inability to advance an orogastric tube.
- A radiograph depicts the tube coiled above the stomach.
- Question: Which congenital lesion explains the findings?

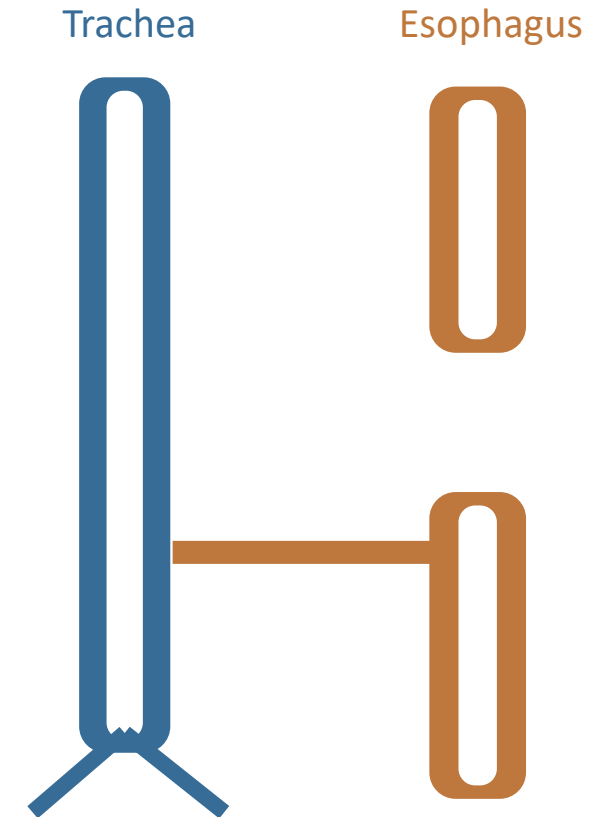


Where would a feeding tube stop?

Case 1: Anatomic answer

Schematic

- Esophageal atresia explains the blind-ending pouch.
- Inspect abdominal gas for a possible distal airway communication.
- Assess cardiac, vertebral, renal, anorectal, and limb anatomy.

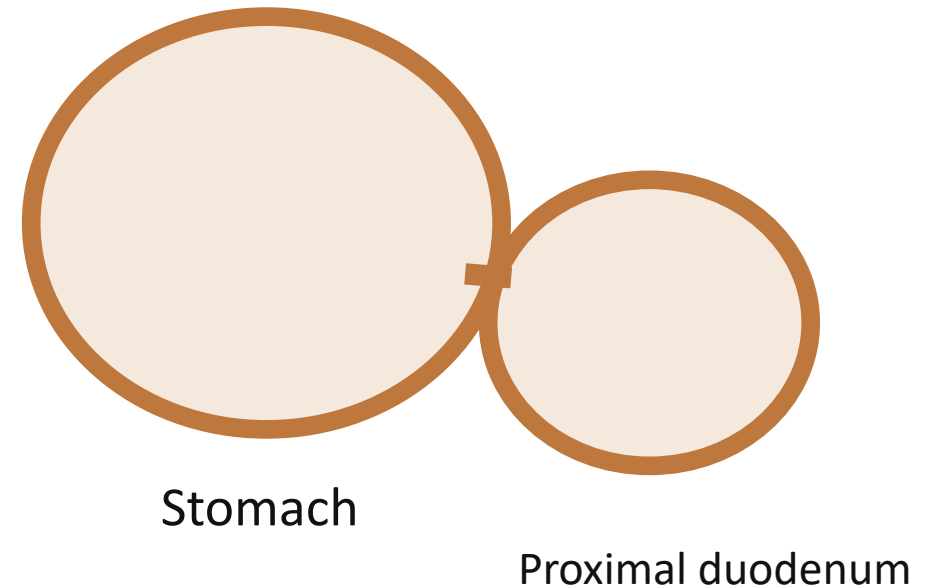


Blind proximal pouch • distal fistula

Case 2: Neonate with bilious emesis

Schematic

- Radiography shows a dilated stomach and proximal duodenum.
- Little or no distal intestinal gas is identified.
- Question: What does the two-compartment pattern localize?



Two gas-filled proximal compartments

Case 2: Level first, cause second

Schematic

- The double bubble localizes proximal duodenal obstruction.
- Duodenal atresia is important but not the only possible cause.
- Correlate anatomy and evaluate associated anomalies.

Proximal duodenum



Distal duodenum

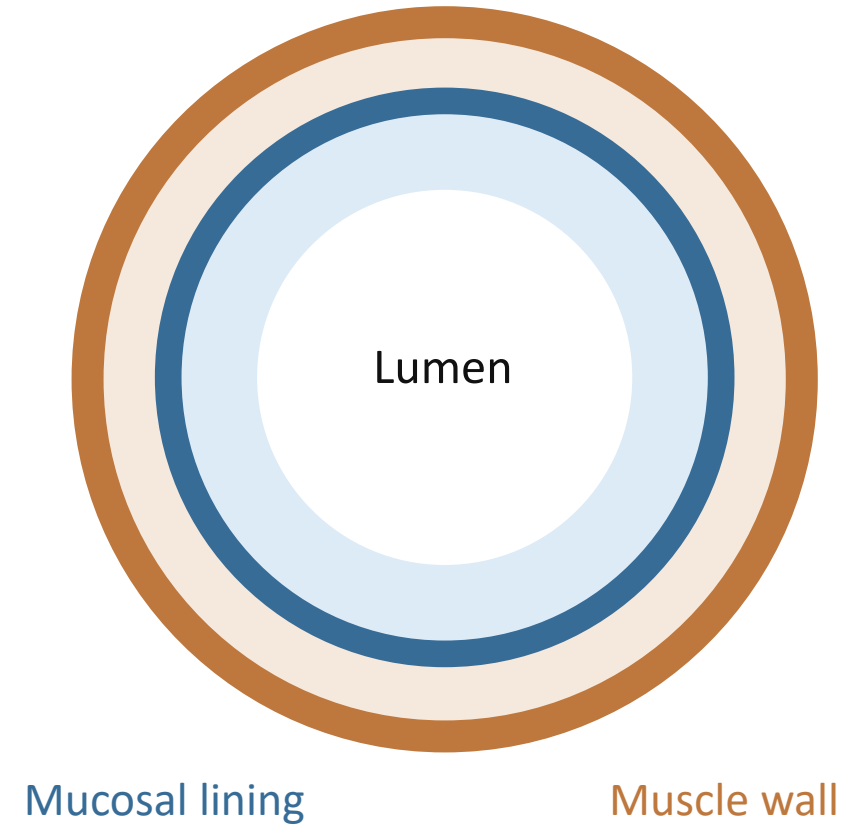


Lumen interrupted in the duodenal segment

Case 3: Enteric-lined cyst in a child

Schematic

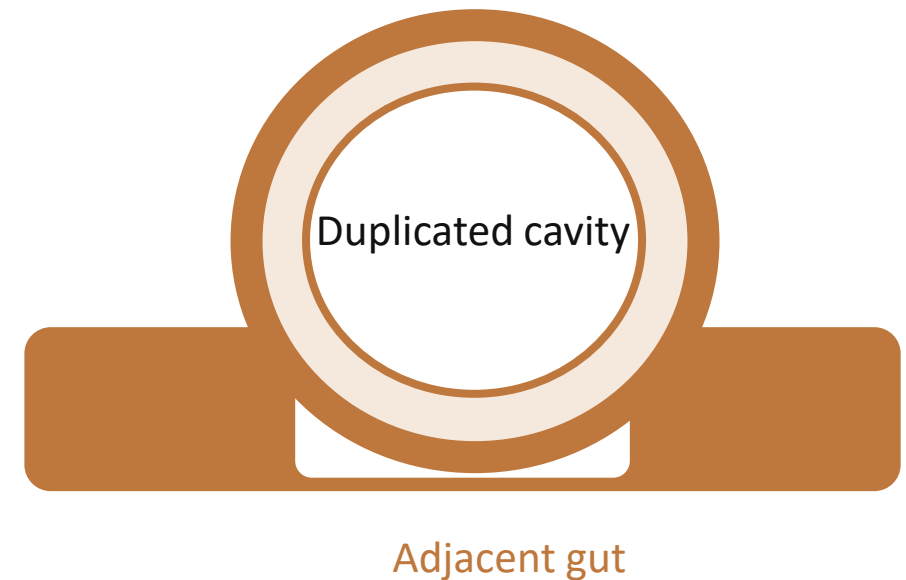
- A cystic lesion is closely attached to the small-intestinal wall.
- Histology shows mucosa and a distinct smooth-muscle coat.
- Question: Which developmental lesion is most likely?



Case 3: Integrated answer

Schematic

- An alimentary duplication cyst explains the anatomical and histologic triad.
- Check for heterotopic mucosa and ulceration.
- Clinical concern includes obstruction, bleeding, and other complications.



Synthesis: Structure predicts symptoms

Schematic

- Esophageal obstruction: Drooling, regurgitation, failed tube passage.
- Airway fistula: Aspiration risk and possible gastric aeration.
- Intestinal obstruction: Vomiting and characteristic bowel-gas patterns.
- Duplication: Cystic mass effect or heterotopic mucosal disease.

Lumen interrupted → retention



Airway connection → aspiration



Duplicated wall → mass or ulceration

References — primary text and clinical context

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