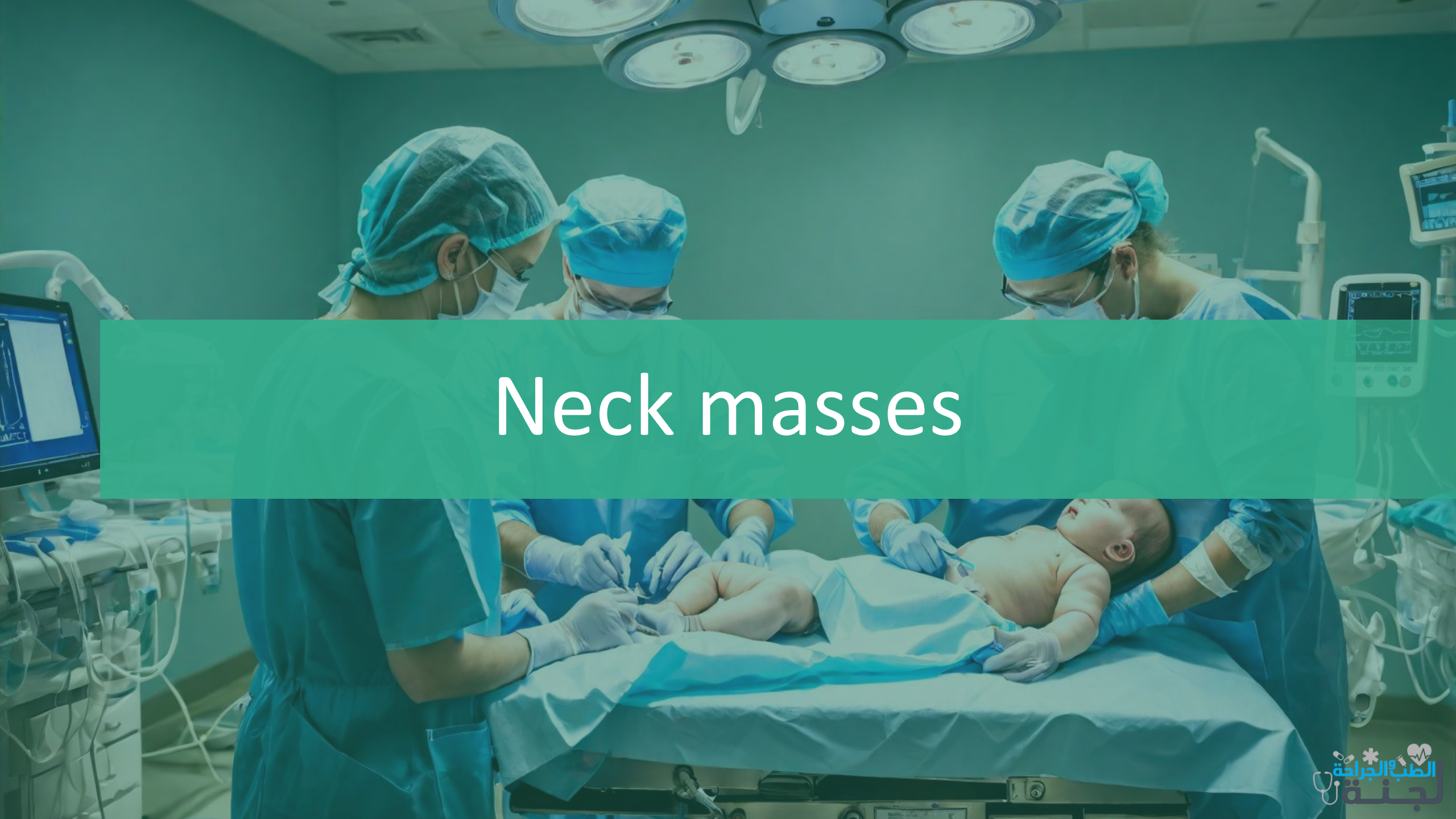
Pediatrics Surgery

2024 Edition

إعداد محمود بركات

الملاحظات

- الملف شامل لمواضيع جراحة أطفال سنة رابعة وسادسة
- مصادر الملف هي المحاضرات والسمينارات وموقع أمبوس
- الملف مقسم حسب عناوين محاضرات الدكتور
- الملف معمول بشكل عام لسادسة بمعنى فيه زيادات عن رابعة وتركيز في بعض السلايدات على خوارزميات التشخيص المطلوبة عموما من سادسة وليس رابعة
- الكلام الأخضر: معلومات إضافية
- الكلام الأزرق: ملاحظات أو إضافات مفيدة
- الكلام الأحمر: مهم



Neck masses

Congenital neck masses DDx

Congenital midline neck mass DDx

1. Thyroglossal duct cyst
2. Dermoid cyst
3. Cervical teratoma

Other midline non-congenital DDx

1. Thyroid Nodule/Goiter
2. Thymic Cyst

Congenital lateral neck mass DDx

1. Branchial cleft cyst
2. Cystic hygroma
3. Hemangioma

Other lateral non-congenital DDx

1. Lymphadenopathy (most common cause of neck masses)
2. Tumors (e.g., Salivary Gland, Neurogenic, Carotid Body)

It’s not always easy to decide if the mass is from midline structure or is lateral and thus always consider both DDx in any neck mass

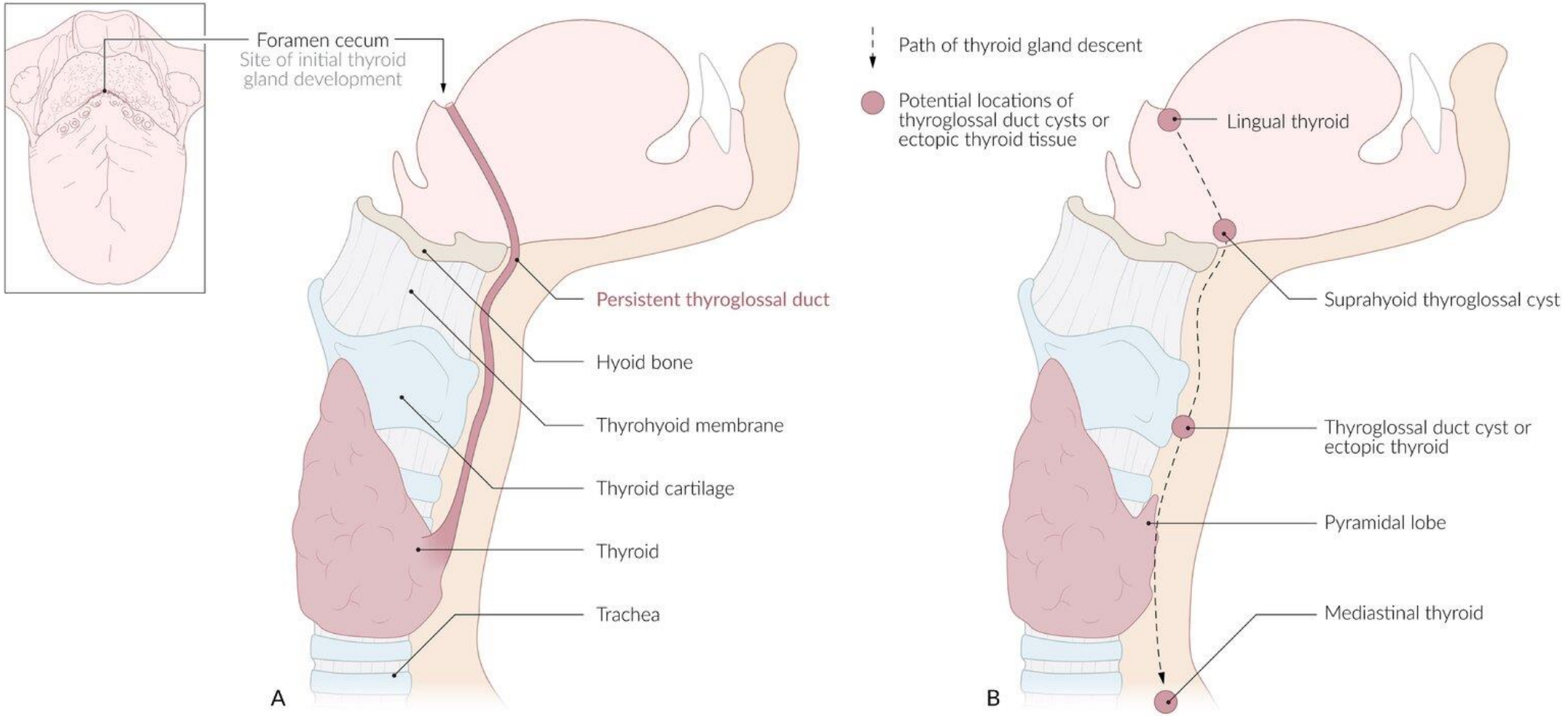
Cervical lymphadenopathy

- Most common cause of neck masses; most common in children < 5 years
- Etiology:
  - Acute unilateral cervical lymphadenopathy: most commonly caused by bacterial infections
  - Acute bilateral cervical lymphadenopathy: most commonly caused by viral infections of the upper respiratory tract
  - Rare causes: Leukemia, Lymphoma, Kawasaki disease
- Most commonly involved LN is submandibular or deep cervical nodes
- It’s a clinical diagnosis; In case of mild disease no further testing is necessary
- Imaging (if indicated): ultrasound
- Management: conservative in most cases

Thyroglossal duct cyst

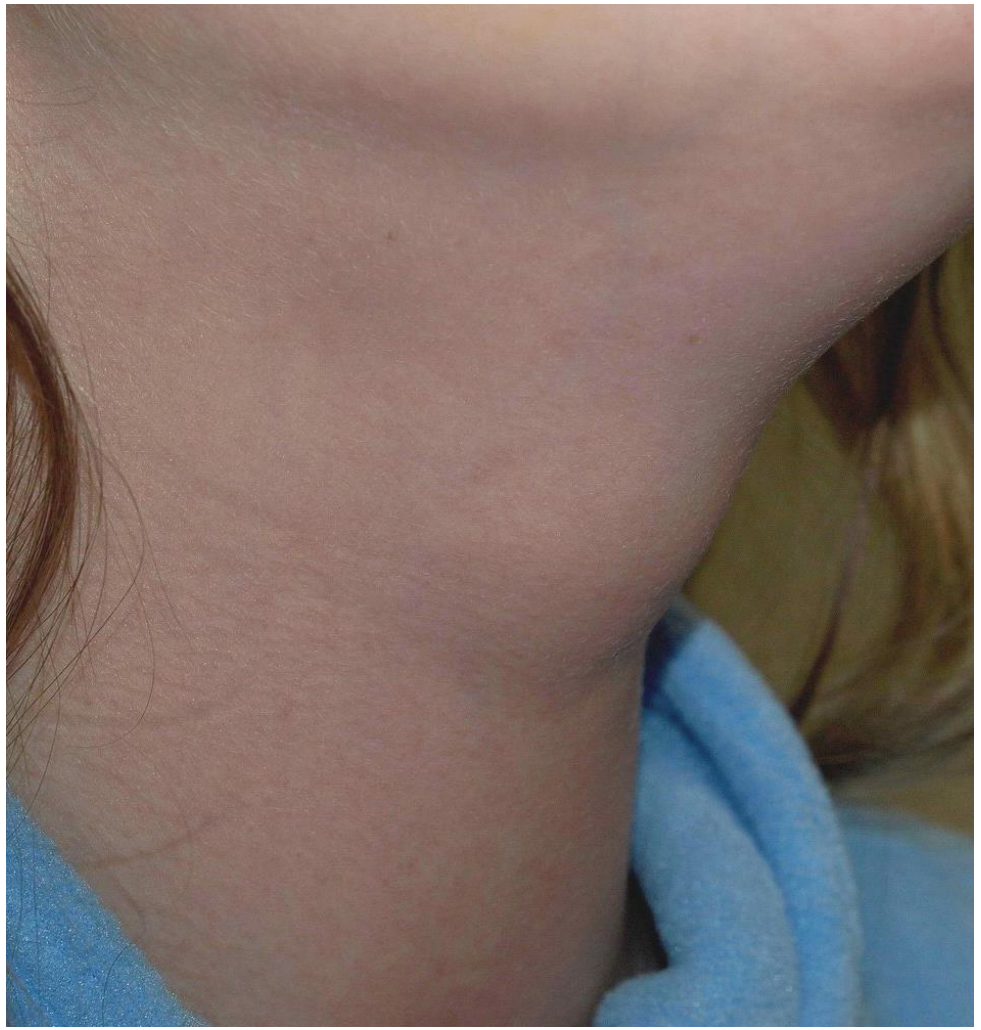
- **Definition**: a remnant of the thyroglossal duct, which forms during the embryonic development of the thyroid gland and normally regresses before birth
- Second most common neck abnormality after lymphadenopathy
- **Pathophysiology**
  - The thyroid gland originates from the foramen cecum at the base of the tongue and descends caudally into the neck, forming the thyroglossal duct.
  - Normally, the duct disappears by the time the thyroid reaches its appropriate position by 5–8 weeks of gestation
  - If the thyroglossal duct fails to obliterate, midline neck cysts or ectopic thyroid tissue can develop anywhere along its path.

Thyroglossal duct cyst



Thyroglossal duct cyst – Clinical features

- The cyst is present from birth and is usually detected during early childhood (< 5 years).
- **Description**: Painless, firm, midline neck mass that elevates with swallowing and tongue protrusion.
- Usually located near the hyoid bone.
- May cause dysphagia or neck/throat pain if the cyst enlarges.
- **Complications**
  - Infection of the cyst with possible abscess formation
  - Sinus tract formation with persistent drainage
  - Ectopic thyroid tissue
  - Malignancy arising from ectopic thyroid tissue



Thyroglossal duct cyst – Diagnostics

- Neck and thyroid examination
- **Ultrasound** of the neck to evaluate the cyst and confirm the location of the thyroid
  - Patients with thyroglossal duct cysts can have an ectopic thyroid gland.
  - In the absence of ectopy, it is important to assess the anatomical relation of the cyst to the thyroid for preoperative planning.
- Contrast-enhanced CT of the neck: preferred imaging modality
  - Thyroglossal duct cysts are demonstrated as well-defined lesions with homogenous fluid attenuation and surrounding rim enhancement, typically close to the hyoid bone.
  - Allows for assessment of anatomical location, relation, and extent of the cyst as well as its relation to normal orthotopic thyroid tissue.
- TSH levels
- If an infection is suspected, fine needle aspiration should be performed for Gram stain and culture (including AFB and mycobacterial culture).

Thyroglossal duct cyst – Treatment

- Treatment of any active infection with antibiotics before surgery
  - Thyroglossal cysts are usually infected with organisms from the oral cavity, most commonly H. influenzae, S. aureus, and S. epidermidis.
- Elective surgical excision (**Sistrunk procedure**) to prevent infection: includes removal of the cyst, a portion of the hyoid bone, and excision of tissue comprising the path of descent from the foramen cecum.
- **Complications of the Sistrunk procedure**
  - **Major complications** include recurrence, hematoma or abscess, entry into the airway (tracheotomy), hypoglossal nerve paralysis, hypothyroidism (ectopic thyroid).
  - **Minor complications** include seroma formation, wound dehiscence, local wound infection, and stitch abscesses

Dermoid cyst

- **Definition**: a cyst that forms during embryonic development when skin and skin structures become trapped in layers of connective tissue.
- In the skin, dermoid cysts usually appear as small, raised bumps or lumps. They can vary in size and may contain a mixture of hair, skin cells, oil, and other debris. Dermoid cysts are usually painless unless they become infected or inflamed.
- **How to differentiate thyroglossal duct cyst from dermoid cyst?**
  - Palpate the lesion while the child sticks out his or her tongue.
  - Owing to its attachment to the foramen cecum, the thyroglossal duct cyst usually moves cephalad when the tongue protrudes.
  - This maneuver is more reliable than asking the child to swallow

Branchial cleft cyst

- **Definition**: remnants of the embryological second branchial cleft or cervical sinus, which normally regresses before birth
- **Epidemiology**
  - Accounts for ~ 20% of pediatric neck masses
  - ~ 95% of all branchial cleft malformations are anomalies of the second branchial cleft.
- **Pathophysiology**: formed due to incomplete obliteration of branchial clefts and pouches
- **Clinical features**: usually diagnosed in late childhood or in adulthood after a previously undiagnosed cyst becomes infected
  - History of upper respiratory infection
  - Painless, firm mass
    - Located lateral to the midline, usually anterior to the sternocleidomastoid muscle
    - Does not move with swallowing
  - There may be a small draining opening if a fistula is present

Branchial arches development

First - ear, eustachian tube, **m**astoid air cells, tympanic **m**embrane, auditory canal, **m**axillary artery, **m**uscles of **m**astication, trigeminal nerve, **M**eckel cartilage (stapes, styloid process, portion of hyoid bone)

Second - facial nerve, muscle of facial expression, stapedial artery, tonsillar fossa, palatine tonsil, Reichert cartilage (stapes, styloid process, hyoid bone)

Third - inferior parathyroid gland, thymus, carotid artery, stylopharyngeus muscle, glossopharyngeal nerve, hyoid

Fourth - superior parathyroid, thyroid c-cells, cricothyroid muscle, most of the pharynx and palate, vagus and superior laryngeal nerves, thyroid and epiglottic cartilages

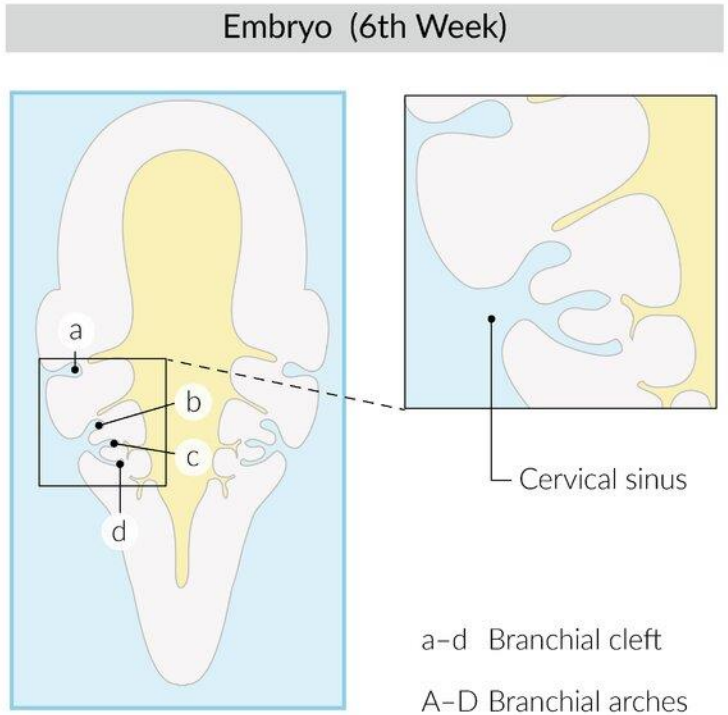
Fifth - regresses

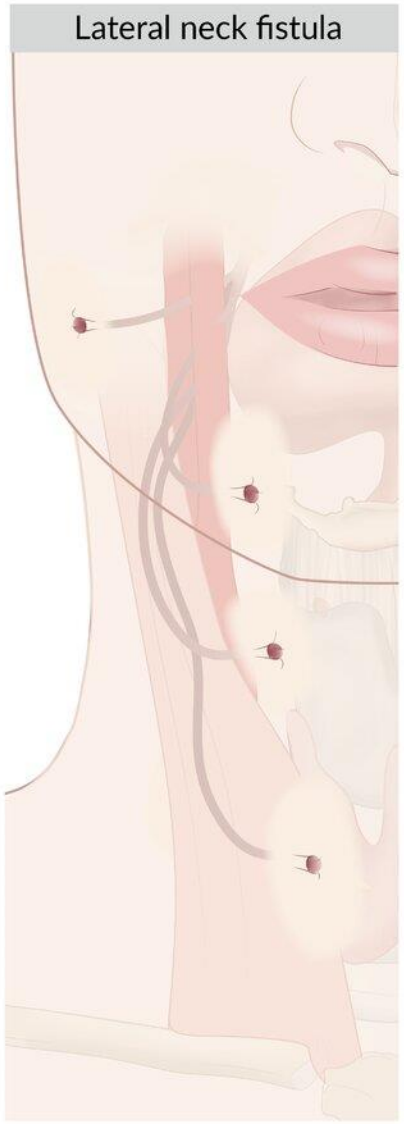
Sixth- pulmonary artery, muscle of larynx except cricopharygeus, vagus and recurrent laryngeal nerves, cricoid cartilage, arytenoid complex

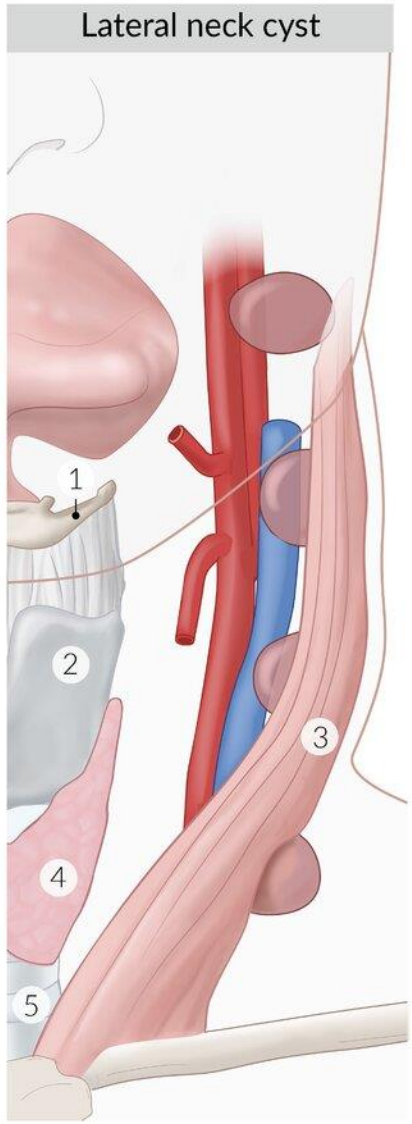
Presentation

- First
  - Duplications of the external auditory canal
  - Cyst, sinuses and fistula between the external auditory canal and angle of the mandible
- Second (most common)
  - They present as a cyst, sinus or fistula anterior to the SCM in the lower neck
- Third and fourth
  - These may present with recurrent neck infections or abscess, suppurative thyroiditis and enlarging cyst or abscess may cause airway compromise, or dysphagia

Branchial cleft cysts and fistulas



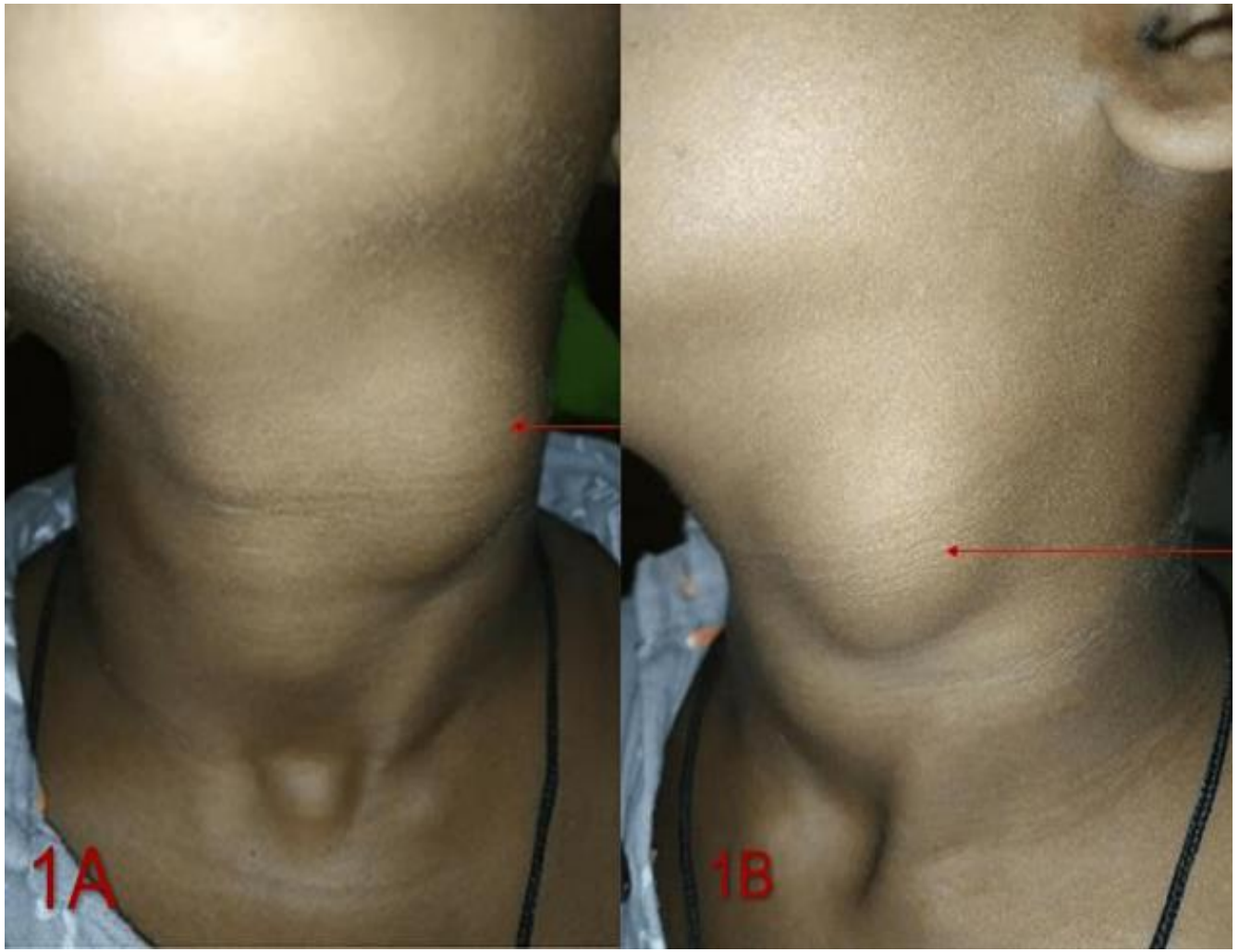




1 Hyoid bone
2 Thyroid cartilage
3 Sternocleidomastoid muscle
4 Thyroid gland
5 Trachea

Branchial cleft cyst

- **Diagnostics**
  - Neck examination
  - Ultrasound
  - CT or MRI to further assess anatomical structures for surgical planning
- **Treatment**: complete surgical excision of both the cyst and any associated tracts
- **Indications for surgery**
  - **Complications**: infection of the cyst, tract, or sinus, with possible abscess formation
  - Possible malignant transformation



Cystic hygroma

- **Definition**: a congenital lymphatic cyst (macrocystic lymphangioma) in the posterior triangle of the neck
- **Epidemiology**
  - Strongly associated with fetal aneuploidy (e.g., Turner syndrome)
- **Clinical features**
  - Present at birth as a soft, compressible, painless, posterior triangle neck mass
  - Can cause dysphagia or airway compromise
  - Positive transillumination test
- **Diagnostics**
  - Ultrasound to identify mass in infancy
- **Treatment**: Small masses may regress spontaneously, but surgical excision is usually indicated to prevent infection or airway compromise, as well as for cosmesis

Esophageal Atresia and Tracheoesophageal Fistula

Gross classification of esophageal atresia

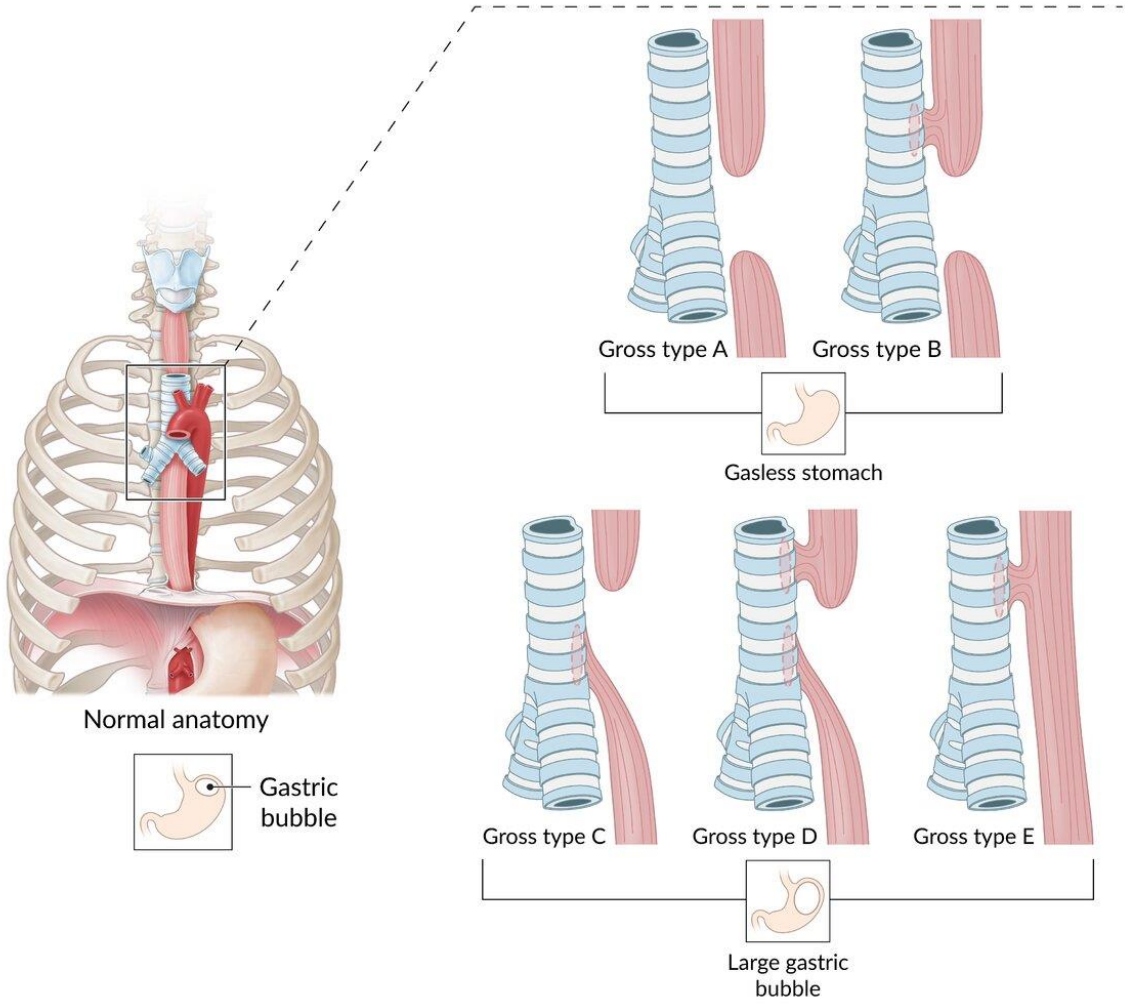
Gross type A: esophageal atresia without tracheoesophageal fistula

Gross type B: esophageal atresia with an upper tracheoesophageal fistula

Gross type C: esophageal atresia with a lower tracheoesophageal fistula

Gross type D: esophageal atresia with upper and lower tracheoesophageal fistula

Gross type E: tracheoesophageal fistula without atresia



Overview of types of esophageal atresia

	Type A	Type B	Type C	Type D	Type E
Description	• Esophageal atresia without tracheoesophageal fistula	• Esophageal atresia with tracheoesophageal fistula connected to the proximal esophageal segment	• Esophageal atresia with tracheoesophageal fistula connected to the **distal esophageal segment**	• Esophageal atresia with tracheoesophageal fistula connected to the proximal and distal esophageal segments	• **H-type** tracheoesophageal fistula without atresia
Epidemiology [1]	• ~ 8% of cases	• ~ 1% of cases	• ~ 87% of cases	• ~ 1% of cases	• ~ 4% of cases

Clinical features	• **Polyhydramnios**
	• **Excessive secretions/foaming at the mouth**
	  ◦ Choking, drooling
	  ◦ Inability to feed
		• **Polyhydramnios**
		• **Excessive secretions/foaming at the mouth**
		  ◦ Choking, drooling
		  ◦ Inability to feed
		• **Aspiration pneumonia**
		  ◦ Coughing spells
		  ◦ Rales
		  ◦ Cyanotic attacks
					• **Aspiration pneumonia**
					  ◦ Coughing spells
					  ◦ Rales
					  ◦ Cyanotic attacks
					• Respiratory distress

X-ray findings	• Gasless abdomen • Esophageal pouch		• Large **gastric bubble** • Esophageal pouch		• Large **gastric bubble**

Pathophysiology

- A wedge of mesoderm called the tracheoesophageal septum separates the developing foregut (esophagus) from the trachea.
- Esophageal atresia and tracheoesophageal fistula are caused by a defect in mesodermal differentiation
- About 50% of cases are associated with other mesodermal defects (**VACTERL** association)
  - **V**ertebral anomaly
  - **A**nal atresia
  - **C**ardiac anomaly
  - **T**racheoesophageal fistula
  - **E**sophageal atresia
  - **R**enal anomaly
  - **L**imb malformation
- Other anomalies are most seen with type A and is least seen with type E

Clinical features – Overview

	Type A	Type B	Type C	Type D	Type E
Polyhydramnios	✓	✓	✓	✓	-
Excessive secretions	✓	✓	✓	✓	-
Aspiration pneumonia	-	✓	✓	✓	✓
Gastric distention	-	-	✓	✓	✓

Clinical features – Esophageal atresia

- **Definition**: a congenital defect in which the upper esophagus is not connected to the lower esophagus and ends blindly instead
- **Prenatal features**
  - Polyhydramnios
    - The fetus is unable to swallow amniotic fluid.
    - Occurs in approx. 57% of pregnancies
  - Associated with an increased risk of premature birth
- **Postnatal features**
  - Pooling of secretions → excessive secretions/foaming at the mouth
    - Choking, drooling
    - Inability to feed
    - Vomiting

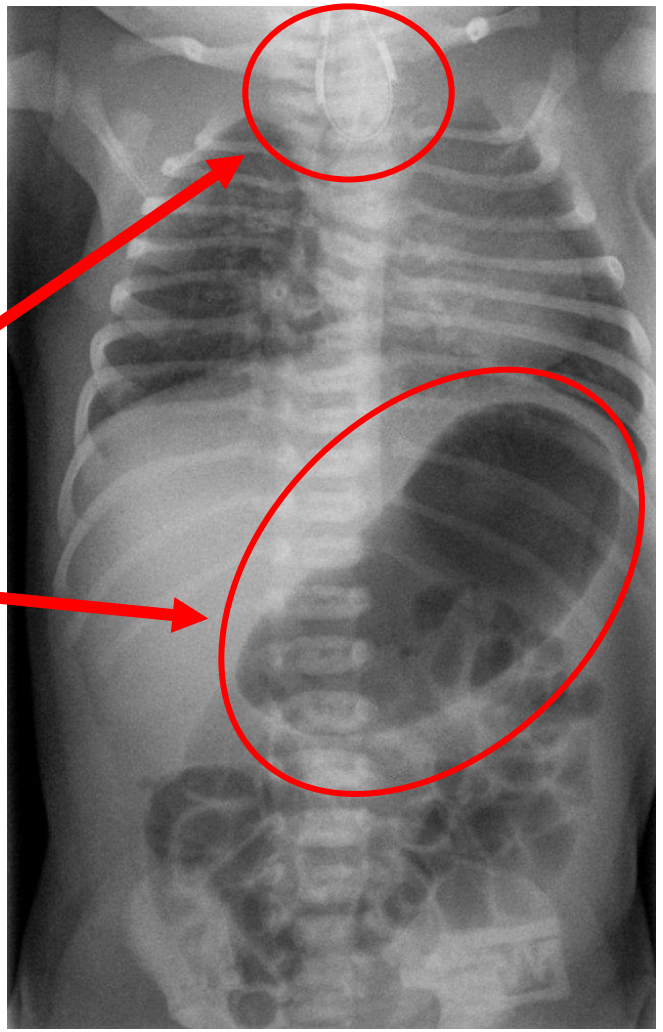
Clinical features – Tracheoesophageal fistula

- **Definition**: an abnormal connection between the trachea and esophagus that may be connected to the proximal and/or distal esophageal segment
- **Postnatal features**
  - Aspiration and subsequent aspiration pneumonia (especially if the fistula is connected to the proximal esophageal segment)
    - Coughing spells (Saliva overflows from the esophagus into the trachea)
    - Rales
    - Cyanotic attacks due to reflex laryngospasms that prevent reflux aspiration
  - If the fistula is connected to the distal esophageal segment: gastric distention

Note: Newborns usually present with symptoms directly after birth. The exception is Gross type E fistula, in which the diagnosis of a small H-type tracheoesophageal fistula may occur as late as adulthood

Diagnostics

- Placement of a feeding tube: the feeding tube cannot pass through the esophagus in the case of esophageal atresia
- X-ray of the thorax/abdomen
  - Esophageal pouch (except with an H-type fistula)
  - Large gastric bubble: air in the stomach (gross types A and B present with a gasless abdomen)
- Further diagnostics concerning VACTERL anomalies
  - Ultrasound of the abdomen
  - Echocardiography



Management

❖ Preoperative

- Placement of an oroesophageal or nasoesophageal tube for continuous suction of secretions to prevent aspiration and facilitate breathing.
- Upper body elevated, left lateral decubitus position.
- Antibiotics in case of aspiration pneumonia.
- Keep infant breathing spontaneously; routine endotracheal intubation should be avoided because of the risk for gastric perforation and worsening respiratory distress as the abdomen becomes distended from ventilation through the TEF.
- Infants who potentially have **esophageal atresia** should not be fed orally under any circumstances.

❖ Surgery

- Surgical treatment should be performed within the first 24 hours of birth
  - Timing of surgery can be quite emergent, as every inspiration may be diverted into the stomach

Management cont.

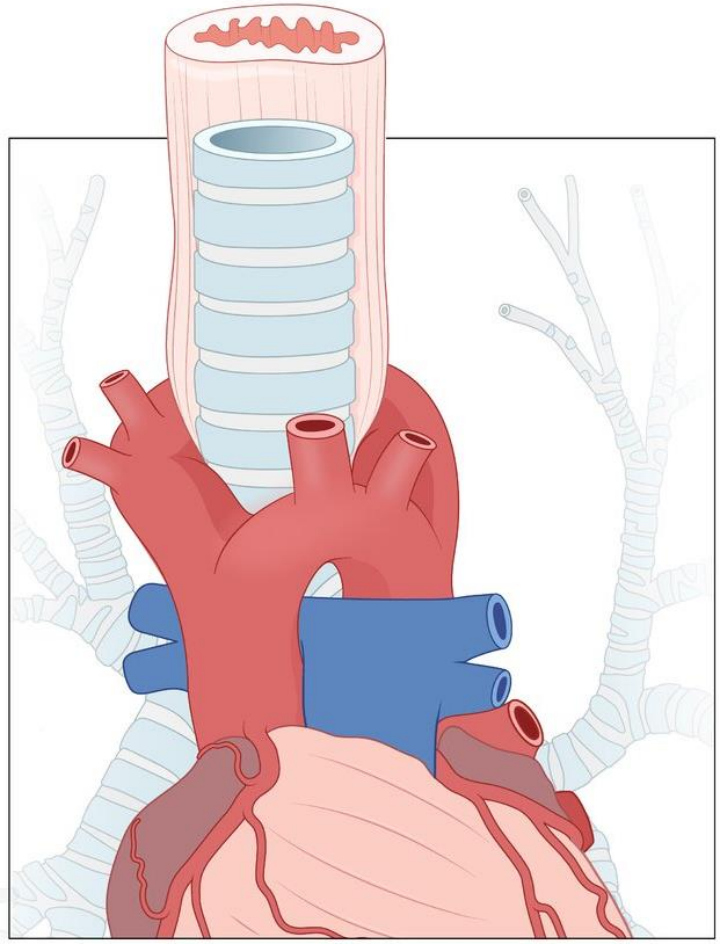
- **Surgery** cont.
  - The goal is to reconnect the upper esophageal pouch and the lower esophagus
  - A long gap between both ends of the esophagus (more than 3 vertebral bodies) may not allow primary repair.
    - In this case, a gastrostomy tube is necessary to allow enteral feeding
    - Treatment options include promoting elongation of the esophagus (via the Foker technique) and colon interposition
  - **Operation**
    - Confirmation of diagnosis via bronchoscopy, opportunity to localize fistula and possibly occlude with balloon catheter
    - **Right thoracotomy** to close/divide fistula
      - If a right-sided aortic arch is present → left-sided thoracotomy
    - Mobilization of proximal/distal esophagus and create tension free anastomosis
    - Same can be done thoracoscopically

Management cont.

- **Surgery** cont.
  - Peri-operative complications
    - Air leak at tracheal repair site
    - Anastomotic leak of esophagus
  - Long Term
    - Anastomotic stricture of esophagus
    - Gastroesophageal reflux w/wo stricture formation
    - Tracheomalacia
- **Post-operative Management**
  - Routine milestones for post op care
    - Assessment of patency of esophagus with esophagogram (post-op day 5-7)
    - Oral feeds
    - Gastroesophageal reflux prophylaxis

DDx: Double aortic arch

- **Definition**: vascular ring anomaly; with subsequent constriction of the trachea and esophagus
- **Clinical features**
  - Manifests within the first weeks of life
  - Tracheal constriction: inspiratory and expiratory stridor, dyspnea, respiratory arrest
    - Acute episodes of severe constriction and/or apnea with cyanosis may occur (can be life-threatening)
    - Hyperextension of the head to improve airflow
  - Esophageal constriction: dysphagia, choking, retching, vomiting, failure to thrive
- **Diagnostics**
  - Chest x-ray (anteroposterior and lateral): shows anterior tracheal bowing and narrowing
  - Chest CTA or MRA
- **Treatment**: surgical division of the minor arch



Double aortic arch

إضافي

Differential diagnoses of newborn swallowing disorders [2][10]	
Differential diagnosis	**Findings**
Esophageal atresia	• Excessive secretions/**foaming** at the mouth • **Coughing spells** • **Cyanotic attacks**
Status post C-section	• Excessive secretions • Reversible condition, as opposed to esophageal atresia
Choanal atresia	• Cyanotic attacks • Attacks normalize after crying or opening the mouth
Esophageal stenosis	• Delayed diagnosis (after introduction of solid food) • Dysphagia • Regurgitation
Achalasia	• Very rare during childhood • Delayed diagnosis (after introduction of solid food) • Dysphagia • Regurgitation
Defective swallowing reflex	• Central nervous system disorders • Peripheral neuromuscular disorders

Case discussion

Hx: Newborn child, normal vaginal delivery, prenatal U/S was unremarkable, now 3 hours old with difficulty of feeding

Q1: What other points of the history do you want to know?

Q2: What specifically would you look for? (Physical Exam)

Q3: What labs are needed?

Q4: What imaging is needed, findings?

Q5: What is your diagnosis?

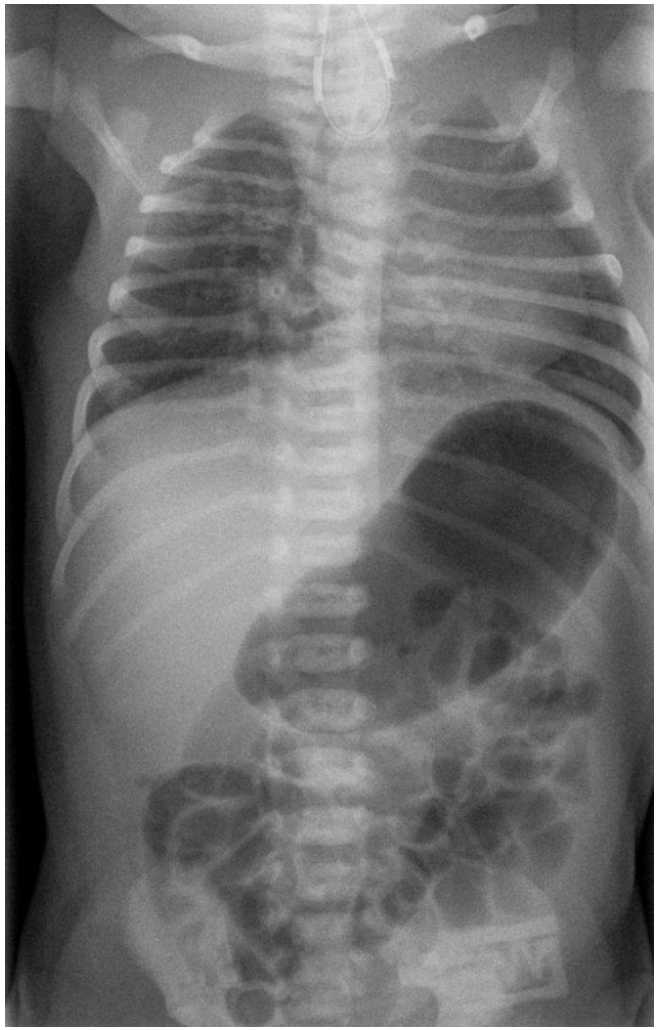
Q6: What is your plan?

Case discussion

- **What other points of the history do you want to know?**
  - **Characterization of symptoms**: spitting and coughing during attempted breastfeeding
  - **Temporal sequence**: immediate with beginning of feeding
  - **Alleviating / Exacerbating factors**: appears fine while not feeding; may have excessive secretions
  - **Associated signs/symptoms**: otherwise, normal appearing child
  - **Pertinent PMH**: vaginal delivery
  - **Perinatal**: mild polyhydramnios
  - **Meds**: none
  - **Relevant Family Hx**: none
  - **Relevant Social Hx**: none

Case discussion

- **What specifically would you look for?**
  - **Vital Signs**: HR 135bpm; RR 40/min; O_2 Sat 97%sat on RA
  - **Appearance**: Well-appearing
  - Relevant exam findings for a problem focused assessment
    - Oral secretions
    - Mild upper abdominal distension
- **What labs are needed?**
  - Standard labs (CBC, X-match)
- **What imaging is needed, findings?**
  - Chest and Abdominal Radiograph, after placement of NG tube
  - Findings: Coiled NG tube (i.e., atresia) and Air in GI tract (i.e., TEF)



Case discussion

- **What is your diagnosis?**
  - Esophageal atresia with tracheoesophageal fistula
- **What is your plan?**
  - VACTERL work-up
    - Vertebral anomaly → Sacral X-ray, spine ultrasound
    - Anal atresia → Physical exam
    - Cardiac anomaly → Echo + Position of the aortic arch
    - Renal anomaly → Renal ultrasound
    - Limb malformation → Physical exam
  - Consent: Rigid bronchoscopy and right thoracotomy, EA/TEF repair
  - Operative
    1. Bronchoscopy
    2. Ligation and division of TE Fistula
    3. Esophageal anastomosis

Congenital diaphragmatic hernias

Embryology of the diaphragm

- **The diaphragm consists of two parts**
  - The peripheral muscular part: made up of radial muscle fibers
  - A central tendinous part: a flat aponeurosis into which the peripheral muscle fibers converge and insert
- **Embryologically, the diaphragm is derived from four embryonic structures**
  - **Anteriorly**: Septum transversum
    - Forms at four weeks of gestation as an incomplete diaphragm centrally separating the pleural and peritoneal cavities and close by the eighth week of gestation
    - Develops into the central tendon of the diaphragm
  - **Dorsolaterally**: Pleuroperitoneal membranes
    - At the 5th week of gestation, pleuroperitoneal membranes form lateral to the central tendon, on either side.
    - They fuse with the septum transversum and the mesentery of the esophagus to complete the partition between the pleural and peritoneal cavities.
    - The left side fuses later, possibly explaining why most CDH are left-sided.
  - **Dorsomedially**: Dorsal mesentery of the esophagus: develops into the diaphragmatic crura
  - **Posteriorly**: Mesoderm of the body wall: forms the peripheral rim of the diaphragm

Congenital diaphragmatic hernias

❖Epidemiology

- Prevalence of CDH: 2.3–2.4 per 10,000 live births
- **More common** in male infants
- **More common** at the left side
  - The liver protects the right hemidiaphragm
  - The right-side closes before the left
- 60% of CDH cases are isolated, 40% associated with anomalies: CVS (27.5%), UG (17.7%), MSS (15.7%), and CNS (9.8%), GI system (atresia)

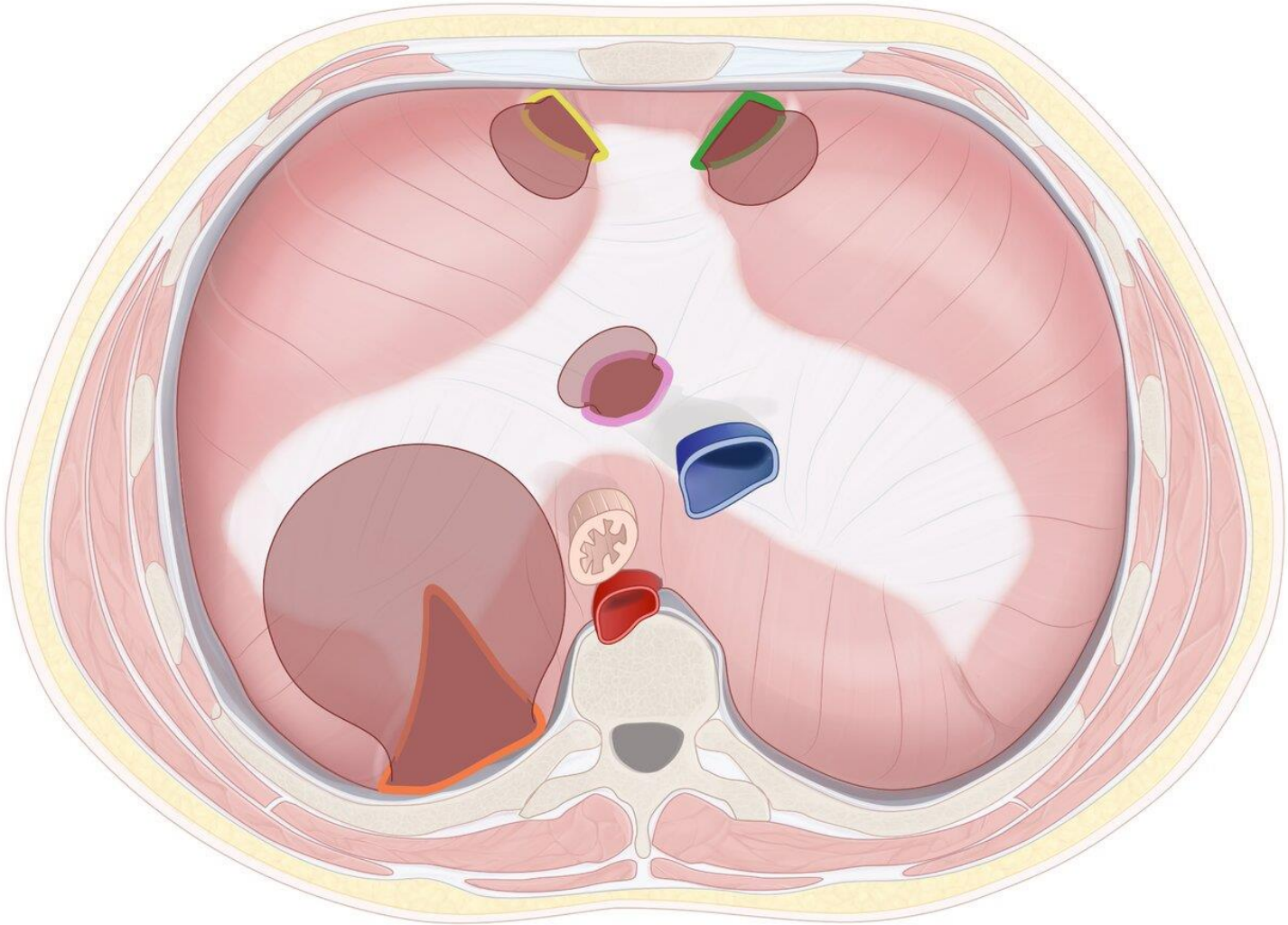
❖Pathophysiology

- Impaired development and/or fusion of embryonic structures (pleuroperitoneal membrane) → defect in the diaphragm persists during fetal development → displacement of abdominal contents into the pleural cavity → compression of lung tissue → pulmonary hypoplasia (Affects the contralateral lung as well)
- The pulmonary arteries exhibit a decrease in density as well as an increase in muscularization (CDH-associated pulmonary hypertension)

Types of congenital diaphragmatic hernias

- Failure of fusion of the septum transversum postero-laterally with the pleuroperitoneal membranes → **Bochdalek hernia**.
  - Most common CDH, accounting for 90% of cases.
  - Postero-lateral (lumbocostal) CDH (85% are left-sided)
- Failure of fusion of the septum transversum anteriorly with the sternum and ribs → **Morgagni hernia** (Morgagni-Larrey hernia)
  - Rare: < 5% of CDH
  - Anterior (sternocostal/parasternal) CDH (90% are right-sided)
  - Infants with Morgagni hernia often present late.

Types of congenital diaphragmatic hernias



Lumbocostal triangle
Bochdalek hernia

Central tendon
Central diaphragmatic hernia

Sternocostal triangle (right)
Morgagni hernia

Sternocostal triangle (left)
Larrey hernia

Clinical features

- Presentation depends on the degree of pulmonary hypoplasia and pulmonary hypertension
  - Can present immediately after birth (minutes to hours) or later (days to weeks), and can range from asymptomatic to severe respiratory distress
- Respiratory distress (e.g., nasal flaring, tachypnea, cyanosis, intercostal retractions, grunting)
  - Respiratory distress is due to severe pulmonary hypoplasia, PPHN, and poor surfactant production, which are typical characteristics of congenital diaphragmatic hernias. Postnatally, the hypoxia and resulting respiratory acidosis → reactive vasoconstriction of the preexisting muscularized pulmonary arteries → PPHN.
- Barrel-shaped chest, scaphoid abdomen (concave anterior abdominal wall), and auscultation of bowel sounds in the chest (due to abdominal contents in chest)
- Decrease in breath sounds bilaterally
- Mediastinal shift: shift of heart sounds/apex beat to the right side
- Possible syndromic dysmorphism (e.g., craniofacial, spinal dysraphism, cardiac)
  - ~ 50% of neonates have other congenital anomalies

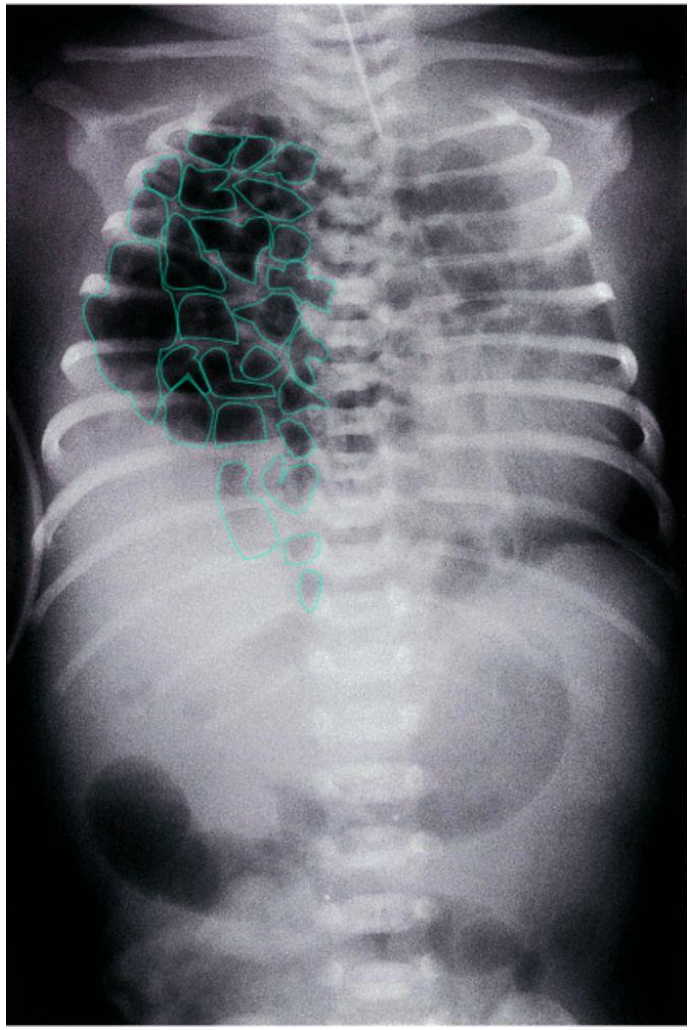
Diagnostics

- **Antenatal ultrasound**: most cases are diagnosed on routine antenatal ultrasound
  - Fluid-filled stomach/bowel seen in the thorax
  - Peristalsis may also be noted in the chest, confirming the diagnosis.
  - Esophageal compression can cause polyhydramnios
  - Hydrops fetalis may also be seen in severe cases
  - The lung-to-head ratio (LHR) is a prenatal US assessment ratio, utilizing the contralateral lung area to the head circumference, which predicts CDH severity. If the LHR is 1 or less, the prognosis is poor.
- **Chest x-ray**
  - Abdominal contents, air/fluid-filled bowel, and poorly aerated lung in the ipsilateral hemithorax
  - Mediastinal shift to the right and compression of the contralateral lung
  - In doubtful cases, a naso-gastric tube is inserted and a chest radiograph is taken: the feeding tube will be seen in the thorax.
  - In right-sided CDH: the liver appears as an intrathoracic soft tissue mass + absence of the normal intra-abdominal liver shadow

Chest x-ray

Right diaphragmatic hernia with enterothorax

- Chest x-ray (PA view) of a female newborn
- Multiple loops of bowel (green overlay) can be seen within the right hemithorax. Hypoplasia of the right lung and mediastinal shift to the left is also visible.
- These findings are characteristic of a right diaphragmatic hernia with enterothorax.



Management

- **Prenatally diagnosed CDH**: antenatal glucocorticoids; to decrease morbidity of pre-term delivery
- **Postnatal therapy**
  - Initial medical resuscitation
    - Intubation and mechanical ventilation: indicated in all infants with CDH
    - Gastric decompression (insert NG tube and continuous suction)
    - Surfactant administration: in infants born < 34 weeks of gestation and x-ray findings suggesting neonatal respiratory distress syndrome
  - **Surgical repair** (thoracotomy or laparotomy)
    - Indicated in all cases of CDH
    - **Timing**: after the infant is stabilized, often after 24–48 hours
    - **Procedure**: reduction of the hernial contents and primary closure of the defect

Poor prognosis

- Large defect size
- Major congenital heart disease
- Prematurity
- Liver herniation
- Lung-to-Head Ratio (LHR) < 1
  - The Lung-to-Head Ratio is calculated by measuring the area of the lung and comparing it to the circumference of the head. A lower LHR indicates more severe lung hypoplasia, which is associated with a poorer prognosis.
- Right more than left

Acute Abdominal Pain in Children

Extra Sources: Pediatric Surgery Library

Acute Abdominal Pain in Children

- **The most common presentation** of abdominal emergency is pain
- **The most common surgical cause** of acute abdominal pain in children is acute appendicitis. Within specific age groups other medical and surgical causes of acute abdominal pain should also be considered. (See slide 3)
- The initial presentation in patients with abdominal pain can be associated with obstruction, inflammation or both. (See slide 4)
- **Initial management**
  - Perform ABCDE survey.
  - Establish NPO status.
  - Perform a focused clinical evaluation, including pelvic, testicular, and rectal examination, if indicated.
  - Perform targeted diagnostic workup of acute abdomen.
  - Obtain urgent specialty consult as needed, e.g., general surgery, urology, OB/GYN
  - Administer supportive care for acute abdominal pain as needed.
  - Identify and treat the underlying cause.

By Age	Any age	Newborn	Infant
Common causes of pediatric abdominal pain	1. Intestinal malrotation/volvulus 2. Mechanical bowel obstruction 3. Meckel diverticulitis 4. Neutropenic enterocolitis 5. Perforated viscus	1. Neonatal obstruction, 2. Congenital lesion internal hernia/volvulus– omphalomesenteric duct remnant 3. Duplication cyst 4. Mesenteric cyst 5. Necrotizing enterocolitis 6. Incarcerated inguinal hernia	1. Intussusception 2. Incarcerated inguinal hernia 3. Nonaccidental abdominal trauma 4. Hirschsprung-associated enterocolitis 5. Abdominal / retroperitoneal neoplasm
Age group	**Toddler**	**Pre-adolescent**	**Adolescent**
Common causes of pediatric abdominal pain	1. intussusception 2. appendicitis– complicated 3. non-accidental abdominal trauma 4. Hirschsprung-associated enterocolitis 5. abdominal/retroperitoneal neoplasm 6. pneumonia	1. Appendicitis 2. Gallstone complications a. Cholecystitis b. Choledocholithiasis c. Gallstone pancreatitis 3. Epiploic fat torsion/infarction 4. Omental torsion/infarction 5. Henoch-Schonlein purpura 6. Viral gastroenteritis 7. Ovarian torsion (female)	1. Appendicitis 2. Gallstone complications a. Cholecystitis b. Choledocholithiasis c. Gallstone pancreatitis 3. GERD 4. Inflammatory bowel disease 5. Ovarian pathology (female) 6. pelvic inflammatory disease(female) 7. perforated gastric/duodenal ulcer 8. epiploic fat torsion/infarction 9. omental torsion/infarction 10. Henoch-Schonlein purpura 11. urinary tract infection 12. urolithiasis

By Pattern	Obstruction	Inflammation	Both	Other
Diagnoses	**Congenital** 1. intestinal malrotation/volvulus 2. internal hernia/volvulus 3. omphalomesenteric duct remnant/Meckel diverticulum 4. intestinal duplication cyst 5. mesenteric cyst 6. incarcerated inguinal hernia 7. intraluminal obstruction 8. distal intestinal obstruction syndrome 9. functional obstruction 10. Hirschsprung associated enterocolitis **Acquired** 1. intussusception 2. adhesive bowel obstruction 3. Crohn disease partial obstruction, stricture 4. superior mesenteric artery syndrome 5. intraluminal obstruction	1. Appendicitis 2. Gallstone complications a. Cholecystitis b. Choledocholithiasis / cholangitis 3. Pancreatitis 4. Pelvic inflammatory disease 5. Other intestinal 6. Meckel diverticulitis 7. Crohn disease – phlegmon/abscess, fistula, perforation 8. Necrotizing, neutropenic enterocolitis 9. Perforated viscus 10. Peptic ulcer disease 11. Foreign bodies 12. Nonaccidental abdominal trauma 13. Pneumonia 14. Viral mesenteric adenitis / gastroenteritis	1. intestinal obstruction with ischemic / infarcted bowel 2. complicated appendicitis 3. inflammatory bowel disease 4. Crohn disease – phlegmon/abscess, fistula, perforation 5. ulcerative colitis-megacolon 6. functional obstruction 7. Hirschsprung associated enterocolitis	1. Abdominal/retroperitoneal neoplasm 2. Ovarian pathology a. Torsion b. Ruptured cyst 3. Epiploic fat torsion / infarction 4. Omental torsion / infarction (male > female) 5. Biliary dyskinesia 6. GERD

What are the critical components of the history?

The parents of younger children can provide insight based on their observations. It is often difficult for children younger than ten years to pinpoint their symptoms and they may only describe a generalized feeling of pain, nausea and discomfort. Relevant features of abdominal pain include

- **Abdominal pain features**
  - Pattern – constant, intermittent
  - Character – dull, sharp, crampy
  - Intensity – increasing, stable/constant vs intermittent, decreasing
  - Impact on normal activities
  - Location – migratory or consistent location, distribution
- **Associated symptoms**
  - Nausea, emesis – emesis color
  - Diarrhea, constipation, obstipation; stool color/presence of blood
  - Constitutional – fever, malaise, lethargy
  - Other – cough, sore throat, dysuria
- **Duration** (time from onset) and sequence of symptoms
- **Exacerbating** and/or **relieving** factors

Cont.

- **The cardinal symptoms**
  - **Pain**; lasting 3-4h
  - **Vomiting**; persistent vomiting should raise the possibility of bowel obstruction
  - **Diarrhea**; may suggest pelvic lesion if last 24h & combined by abdominal pain
- Emesis (especially if bilious), obstipation and intermittent abdominal pain imply presence of intestinal obstruction or disordered motility such as an ileus.
- Nonspecific symptoms such as fever, malaise and constant pain in a persistent location may be caused by inflammatory conditions.
- Cough, sore throat or dysuria suggests a potential medical cause of abdominal pain.
- Also important to assess is a prior history that includes comorbidities, previous symptoms, operations, ill contacts, recent travel and any trauma.

What are the critical components of the physical exam?

- A careful physical exam is essential but potentially challenging especially in young children.
- Observation of a child’s appearance and movement while obtaining his or her history provides insight into the degree of discomfort.
- Utilizing some means of distracting the sick child may be the only way to obtain a good abdominal exam.
- Warming the hands or examining younger children and infants while still clothed or in their parents' arms may be necessary to help the child relax in order to allow the exam to proceed.
- Pertinent findings include distension and pain with palpation and/or percussion while noting the degree of tenderness and location.
- Adjunct maneuvers include assessing costovertebral angle tenderness and referred pain from costochondritis as well as the presence of Rovsing, obturator and psoas signs.
- Other physical exam components may also support a non-abdominal source for symptoms such as diminished breath sounds for pneumonia or pharyngeal erythema/tonsillar enlargement with Streptococcal pharyngitis.

Diagnostics

The diagnostic workup should be guided by the pretest probability of the diagnoses under consideration. The following list includes some commonly used diagnostic tools that can help to diagnose or rule out possible etiologies in a patient with acute abdominal pain.

- **Laboratory studies**: CBC, CRP, ESR, BMP, CMP, Amylase, Lipase, Urinalysis
- **Radiological Tests**:
  - Abdominal X-Ray (Erect/Supine)
  - Abdominal/Pelvis Ultrasound
  - UGI Contrast study/SBFT/Contrast Enema
  - Abdomen/Pelvis CT scan
  - Abdominal MRI
  - CXR when respiratory symptoms or signs are present

Acute appendicitis

- **Uncomplicated appendicitis**: appendicitis with no evidence of an appendiceal fecalith, an appendiceal tumor, or complications
- **Complicated appendicitis**: appendicitis associated with perforation, gangrene, abscess, an inflammatory mass, an appendiceal fecalith, or an appendiceal tumor
- **Peak incidence**: 10–19 years of age
- **Clinical features**
  - Initial diffuse periumbilical pain (**visceral pain**) then localizes to the RLQ within ~ 12–24 hours (**somatic peritoneal pain**)
  - **Associated nonspecific symptoms**: Anorexia, Nausea, Vomiting, Low-grade fever, and Diarrhea
  - **Clinical signs of appendicitis**: McBurney point tenderness, RLQ guarding and/or rigidity, Rebound tenderness, Rovsing sign, Psoas sign, and Obturator sign

Acute appendicitis – Management

- Keep patients NPO and initiate supportive care: e.g., IV fluids, analgesia, antiemetics
- **Low likelihood of appendicitis**
  - Associated scores: Alvarado score ≤ 2–4
  - Management: Additional testing for appendicitis may not be required.
- **Moderate likelihood of appendicitis**
  - Associated scores: Alvarado score ≤ 5–6
  - Management: confirmatory imaging required, e.g., ultrasound abdomen, CT abdomen
- **High likelihood of appendicitis**
  - Associated scores: Alvarado score ≥ 7–9
  - Management: Urgent surgical consult for admission and definitive treatment required
    - Begin empiric antibiotic therapy for acute appendicitis.
  - Next steps
    - Laparoscopic appendectomy within 24 hours for uncomplicated appendicitis
    - Emergency appendectomy for complicated appendicitis with systemic manifestations

Alvarado score (MANTRELS) [27]

Characteristics		Score
Symptoms	Migration of pain to RLQ	1
	Anorexia	1
	Nausea and/or vomiting	1
Physical examination	Tenderness in RLQ	2
	Rebound pain	1
	Elevated temperature > 37.3°C (99.1°F)	1
Laboratory parameters	Leukocytosis (> 10,000/mm^3)	2
	Shift to the left (≥ 75% neutrophils)	1

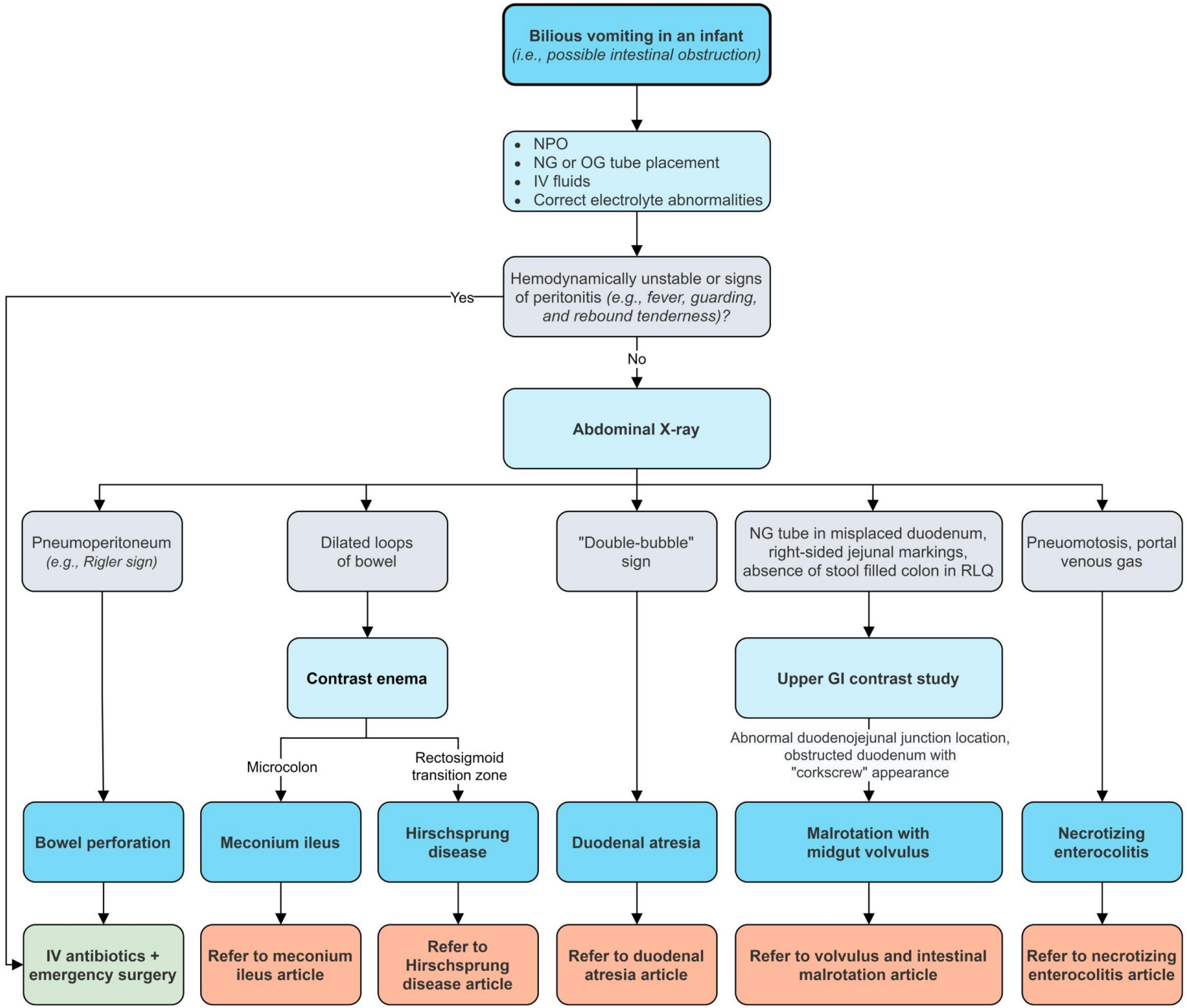
Likelihood of appendicitis

- ≤ 4: Low [16]
- 5–6: Moderate
- ≥ 7: High [16]

Acute mesenteric adenitis

- **Definition**: Mesenteric lymphadenitis refers to nonspecific self-limiting inflammation of the mesenteric lymph nodes
- **Clinical features**
  - It causes a clinical presentation that is often difficult to differentiate from acute appendicitis particularly in children
  - **Include the following**: RLQ pain, Fever (38-38.5°C), Diarrhea, Malaise, Anorexia, Concomitant or antecedent URTI, Nausea and vomiting (which generally precedes abdominal pain, as compared to the sequence in appendicitis), Voluntary guarding rather than abdominal rigidity
- **Management**
  - Patients with mild, uncomplicated presentations do not require antibiotics, and supportive care generally suffices

Neonatal Intestinal Obstruction


Bilious vomiting in an infant
(i.e., possible intestinal obstruction)
NPO
NG or OG tube placement
IV fluids
Correct electrolyte abnormalities
Hemodynamically unstable or signs of peritonitis (e.g., fever, guarding, and rebound tenderness)?
Yes
No
Abdominal X-ray
Pneumoperitoneum (e.g., Rigler sign)
Dilated loops of bowel
"Double-bubble" sign
NG tube in misplaced duodenum, right-sided jejunal markings, absence of stool filled colon in RLQ
Pneuomotosis, portal venous gas
Contrast enema
Upper GI contrast study
Microcolon
Rectosigmoid transition zone
Abnormal duodenojejunal junction location, obstructed duodenum with "corkscrew" appearance
Bowel perforation
Meconium ileus
Hirschsprung disease
Duodenal atresia
Malrotation with midgut volvulus
Necrotizing enterocolitis
IV antibiotics + emergency surgery
Refer to meconium ileus article
Refer to Hirschsprung disease article
Refer to duodenal atresia article
Refer to volvulus and intestinal malrotation article
Refer to necrotizing enterocolitis article

Hypertrophic Pyloric Stenosis (HPS)

- **Epidemiology**
  - Occurs in 1.5-4 per1000 children
  - More common in boys (~ 5:1), in white populations, and in first-born
- **Etiology**
  - **Environmental factors**: Exposure to nicotine during pregnancy & Bottle feeding
    - Bottle-fed infants drink more milk in less time, which may lead to pylorus muscle hypertrophy through overstimulation. Another hypothesis maintains that formula components make it harder to digest and gastric emptying is delayed, which may also burden the pylorus muscle.
  - **Genetic factors**: patients with affected relatives have a higher risk of HPS
  - **Macrolide antibiotics**: Erythromycin and azithromycin are associated with a higher risk of hypertrophic pyloric stenosis, especially when administered within 2 weeks after birth
- Pyloric muscle hypertrophy and hyperplasia results in gradual complete obstruction of the pyloric channel

Hypertrophic Pyloric Stenosis (HPS)

❖Clinical features

- Symptoms usually develop between the 2nd and 7th week of age (rarely after the 12th week).
- Frequent regurgitation progressing to projectile, nonbilious vomiting immediately after feeding
- An enlarged, thickened, olive-shaped, nontender pylorus (diameter of 1–2 cm) is palpable in the epigastrium in 70–90% of patients
- A peristaltic wave, moving from left to right, may be evident in the epigastrium
- "Hungry vomiter": demands re-feeding after vomiting, demonstrates a strong rooting and sucking reflex, irritable
- If left untreated: dehydration, weight loss, failure to thrive
  - These are no longer common symptoms of hypertrophic pyloric stenosis because the diagnosis is made earlier nowadays.

Hypertrophic Pyloric Stenosis (HPS)

❖Diagnostics

- **Initial imaging**: abdominal ultrasound shows an elongated and thickened pylorus
  - The diagnostic criteria for pyloric stenosis is a muscle thickness of ≥4 mm and a pyloric length of ≥16 mm.
  - If the US findings are equivocal → barium studies
- **Alternative imaging**: barium studies shows string sign: elongated, thickened pylorus
- **Laboratory tests**: Hypochloremic, hypokalemic metabolic alkalosis, a classic result, is now uncommon because infants are typically diagnosed and treated early.
  - The loss of gastric hydrochloric acid from emesis results in increased bicarbonate and decreased chloride concentrations in the blood.
  - Hypokalemia usually occurs in infants that have been vomiting for many days or even weeks.

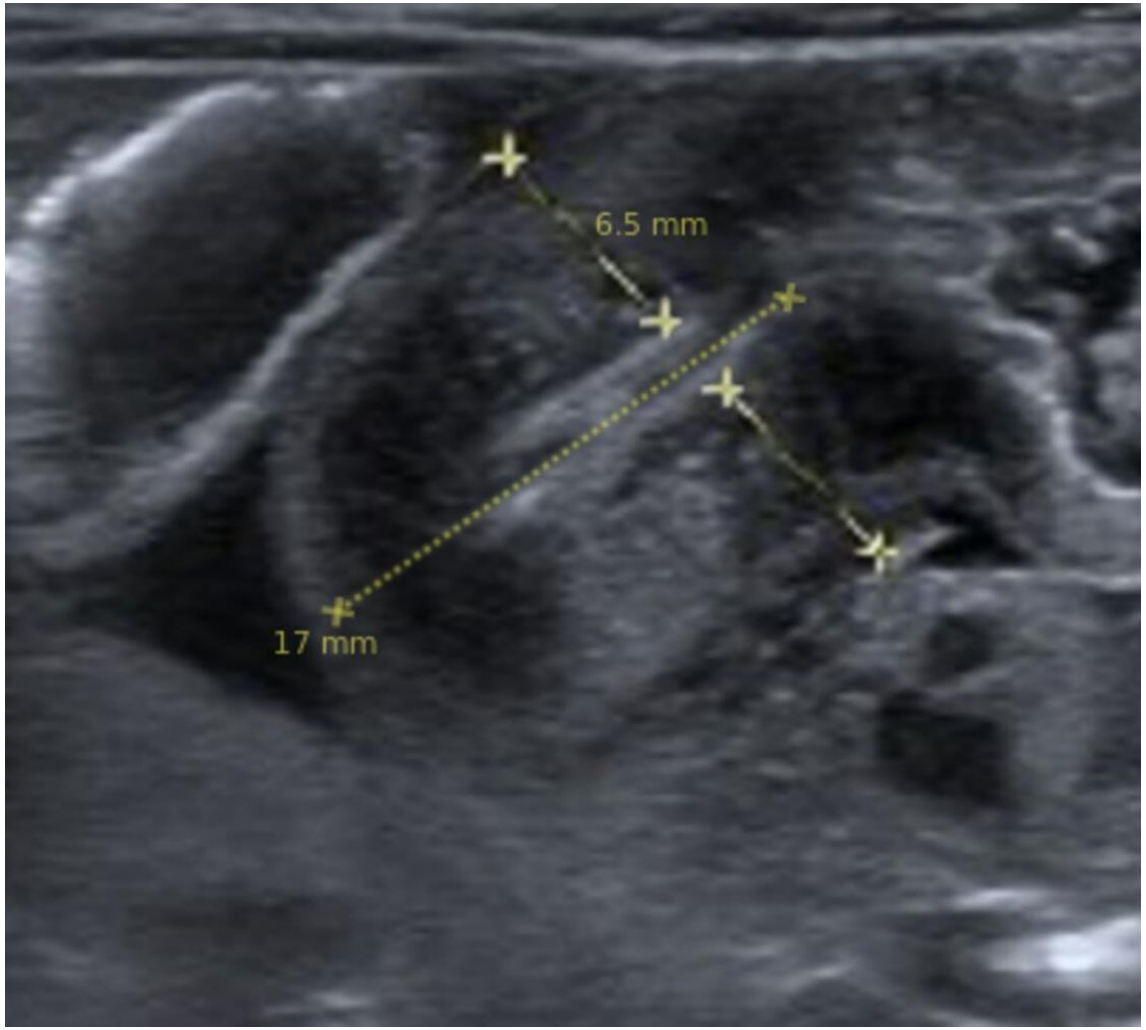
❖Management

- **Pre-op**: NPO, IV fluids, avoid NG tube, Correction of metabolic alkalosis if present
- **Surgery**: Pyloromyotomy (Open or Laparoscopic)

Hypertrophic Pyloric Stenosis (HPS)

Ultrasound abdomen (pylorus; longitudinal plane) of an 8-week-old male infant

- The single wall thickness of the muscular layer measures 6.5 mm (normal cutoff: 3 mm). The pyloric channel measures 17 mm (normal cutoff: 14 mm). No fluid was seen to pass through the pylorus into the duodenum during real-time evaluation when the infant was fed.
- The imaging findings confirmed the diagnosis of pyloric stenosis.



Duodenal atresia

- **Epidemiology**: Occurs in 1 per 5000–10,000 live births, affecting boys more commonly than girls
- **Etiology**: Associated anomalies have been reported in 45–65% of cases. Most commonly, trisomy 21 is found in almost half the cases.
- **Pathophysiology**: Due to failure of recanalization (usually between the 8th and 10th weeks of gestation)
- **Clinical features**
  - Intrauterine: polyhydramnios
  - Postpartum
    - Vomiting that is typically bilious if the stenosis is distal to the major duodenal papilla
      - Atresia or high-grade stenosis: vomiting a few hours after birth
      - Mild stenosis: vomiting after a few days
    - Distended upper abdomen and scaphoid lower abdomen
    - Delayed meconium passage

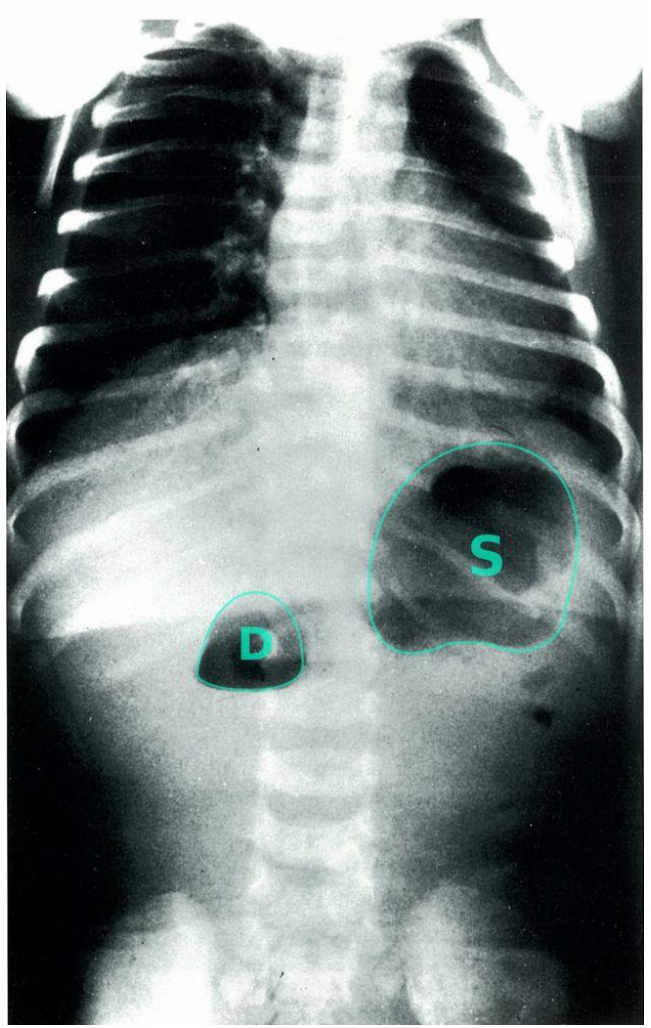
Duodenal atresia

❖Diagnosis

- **Prenatal**: ultrasound (Double bubble sign)
  - If present, test for associated anomalies (e.g., karyotyping, microarray)
- **Postnatal**: X-ray of the abdomen (gasless distal bowel & double bubble sign)

❖Treatment

- Preoperative management
  - Parenteral nutrition via a central catheter shortly after birth
  - Fluid replacement and restoration of the electrolyte balance
  - Gastric decompression
- Surgical repair is duodenoduodenostomy

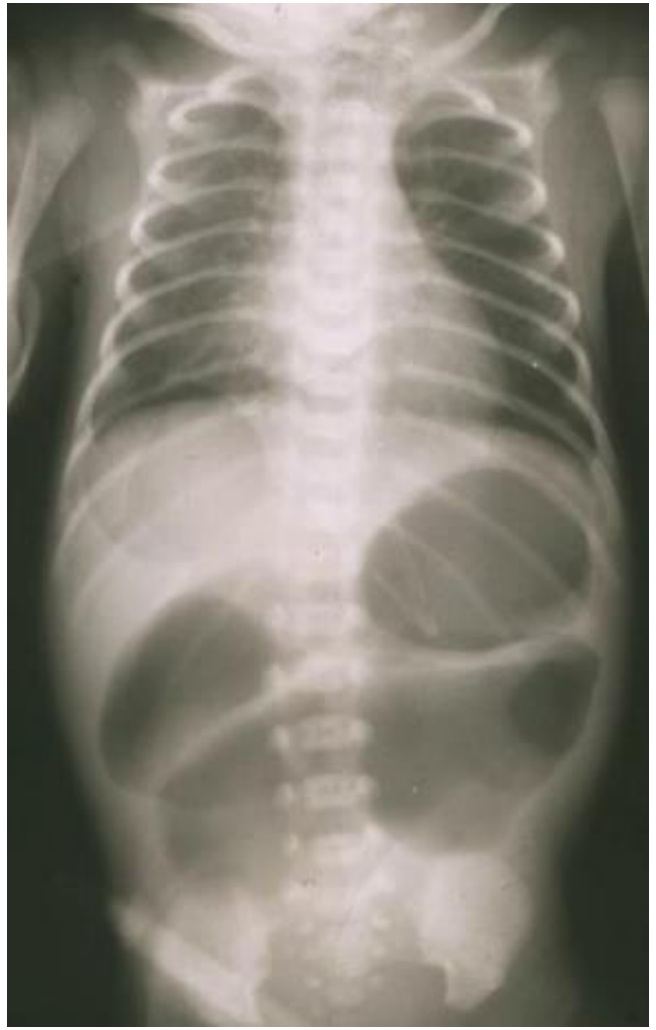


Intestinal Atresia

- **Definition**: absence of the jejunal or ileal lumen
- **Epidemiology**
  - Occurs in approximately 1 in 5000 live births. It occurs equally in males and females
  - Premature infants more common than full term
  - Jejunum more common than Ileum
  - Associated anomalies are rare, unlike DA
- **Etiology**
  - Associated with cystic fibrosis
  - **Risk factors**: maternal smoking and/or use of vasoconstrictive agents (e.g., cocaine, MDMA) during pregnancy
- **Pathophysiology**: vascular accident in utero (usually a disruption of superior mesenteric artery) → ischemic necrosis and reabsorption of the jejunum or ileum → discontinuous bowel → obstruction

Intestinal Atresia

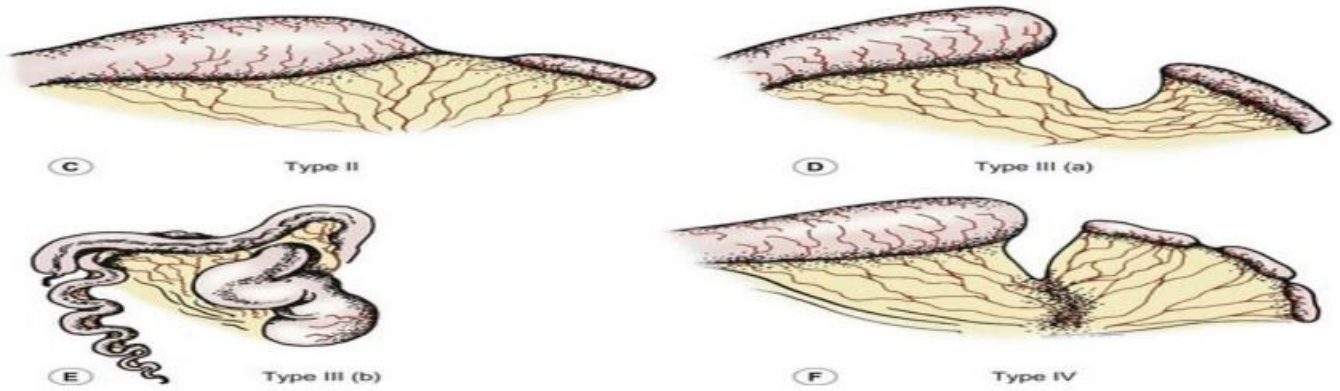
- **Clinical features**: like duodenal atresia
  - Polyhydramnios (intrauterine)
  - Bilious vomiting and upper abdominal distention (postpartum)
- **Diagnostics**
  - Abdominal x-ray shows a triple bubble sign (dilated small bowel loops and air-fluid levels; the three bubbles correlate with the distended stomach, duodenum, and jejunum proximal to the obstruction) and gasless colon.
- **Treatment**
  - Abdominal exploration with resection of proximal, dilated bowel with primary end-to-end anastomosis



Intestinal Atresia

❖Classification

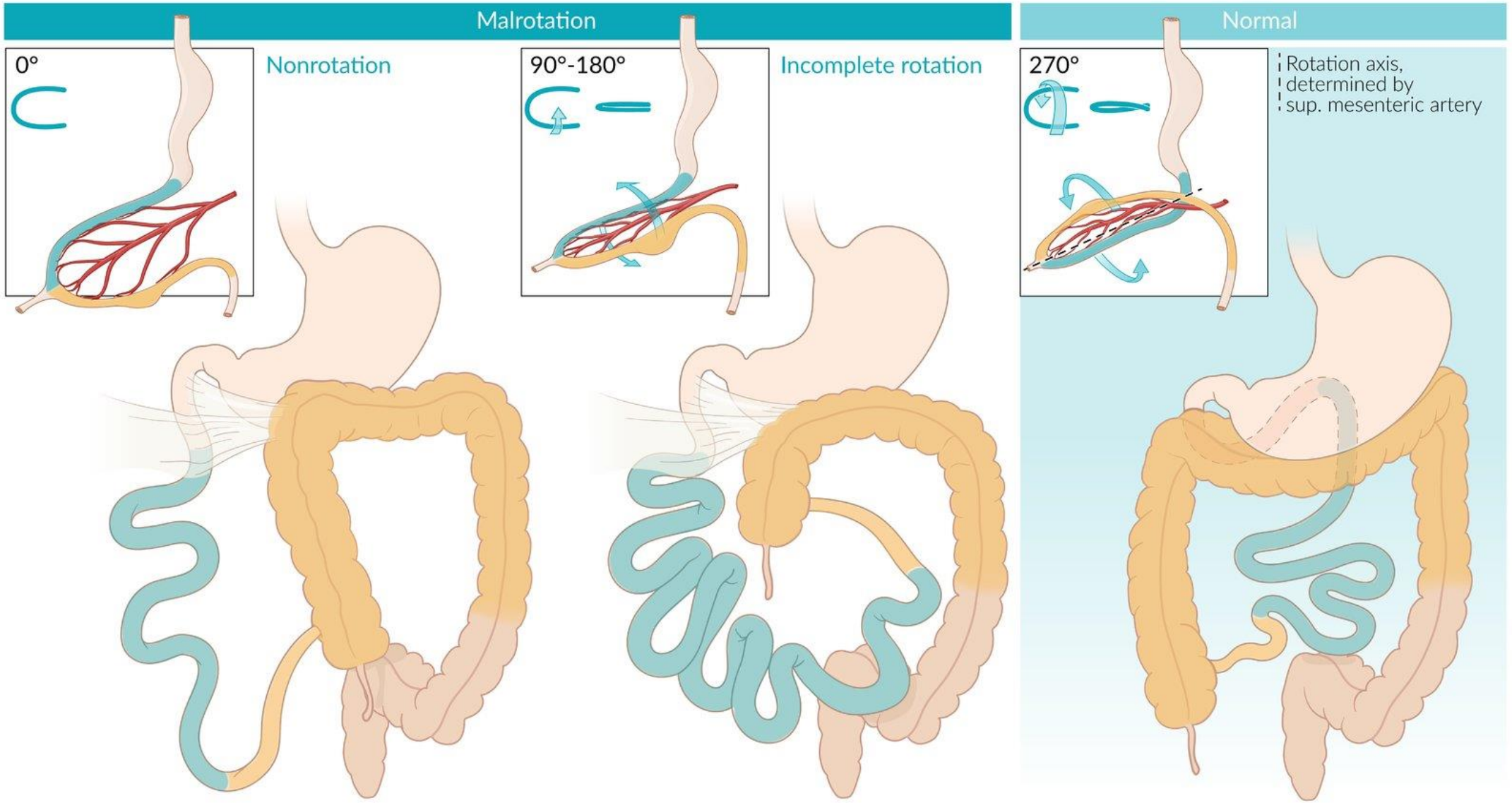
- **Type 1**: intra-luminal web, normal bowel length
- **Type 2**: fibrous cord, normal bowel length
- **Type 3A**: complete atresia with mesenteric defect
- **Type 3B**: (Apple peel or Christmas tree deformity): complete atresia with mesenteric defect and distal small bowel in helix configuration around ileocolic pedicle, significant loss of bowel length
- **Type 4**: (String of sausages deformity) multiple atresias, significant loss of bowel length



Intestinal malrotation

- **Epidemiology**:
  - 1/6000 live birth incidence
  - 90% present within first year of life (75% within 1 month of life)
- **Pathophysiology**
  - **Normal intestinal rotation**: the midgut starts to elongate in utero (4th week) → herniation of the midgut out of the umbilicus (6th week) → 90° counter-clockwise rotation of the midgut → re-entry of the midgut into the abdominal cavity(10th week) → 180° rotation inside the abdominal cavity (a total of 270°) → fixation of the duodenojejunal flexure and cecum to the posterior abdominal wall
  - **Intestinal malrotation**: arrest in the normal rotation of the gut in utero, resulting in an abnormal orientation of the bowel and mesentery within the abdominal cavity

Intestinal malrotation



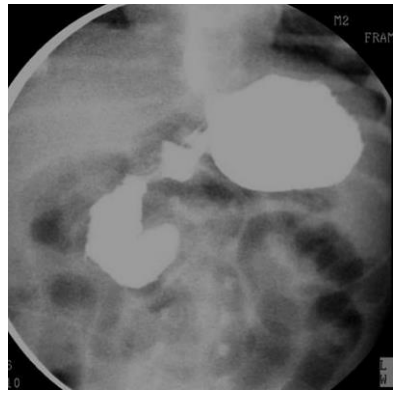
Intestinal malrotation

❖Clinical features

- Presentation varies but most severe type is with midgut volvulus which is due to twisting of the abnormally fixated small bowel around a narrow-based mesentery and SMA
- Midgut volvulus
  - Bilious vomiting with abdominal distension in a neonate/infant
  - Signs of bowel ischemia: hematochezia, hematemesis, hypotension, and tachycardia
- Bilious emesis in newborn is malrotation until proven otherwise

❖Diagnostics

- **Upper Gastrointestinal series (UGI)** is diagnostic test of choice
  - DJ junction lies right to spine
  - Malpositioning of the cecum (left-sided cecum)



Intestinal malrotation

- **Management**
  - **Midgut volvulus with/without peritonitis**
    - **Initial resuscitation**: NPO; nasogastric tube insertion; IV fluids; correction of electrolyte imbalance; broad-spectrum IV antibiotics
    - **Emergency surgery (Ladd procedure)**
      - The volvulus is reduced/untwisted, and the Ladd bands removed.
      - Gangrenous/necrotic bowel, if present, is resected and either anastomosed or created into a stoma.
      - Appendix is removed (to prevent future diagnostic/operative difficulties)
      - Development of postoperative adhesions decreases the chance of recurrent volvulus.
  - **Incidentally detected/asymptomatic intestinal malrotation**: elective surgery (Ladd procedure)

Open Ladd's Procedure

- RUQ transverse incision or midline laparotomy
- Eviscerate intestinal contents
- Detorse counter-clockwise if volvulus is present
  - Resect grossly necrotic bowel
- Release Ladd's cecal bands
- Broaden the small intestine mesentery
- Incidental appendectomy
- Place small bowel on right and colon on left

Meconium ileus

- **Definition**: failure to pass the first stool in neonates (meconium usually passes within the first 24–48 hours after birth)
- **Etiology**: Cystic fibrosis is the cause in > 90% of cases
- **Clinical findings**: signs of a distal small bowel obstruction
  - **Simple**: bilious vomiting, abdominal distention, no passing of meconium or stool
  - **Complicated**: perforation, peritonitis, ascites
- **Diagnostics**
  - X-ray abdomen (with contrast agent): Neuhauser sign (soap bubble appearance): a mottled or bubble-like appearance in the distal ileum and/or cecum as a result of meconium mixing with swallowed air
- **Treatment**: Contrast enema if simple, resection if complicated, treat underlying CF disease if present

Hirschsprung Disease (HD)

❖ Epidemiology

- 1 in 5000 live births
- 4:1 male-to-female ratio

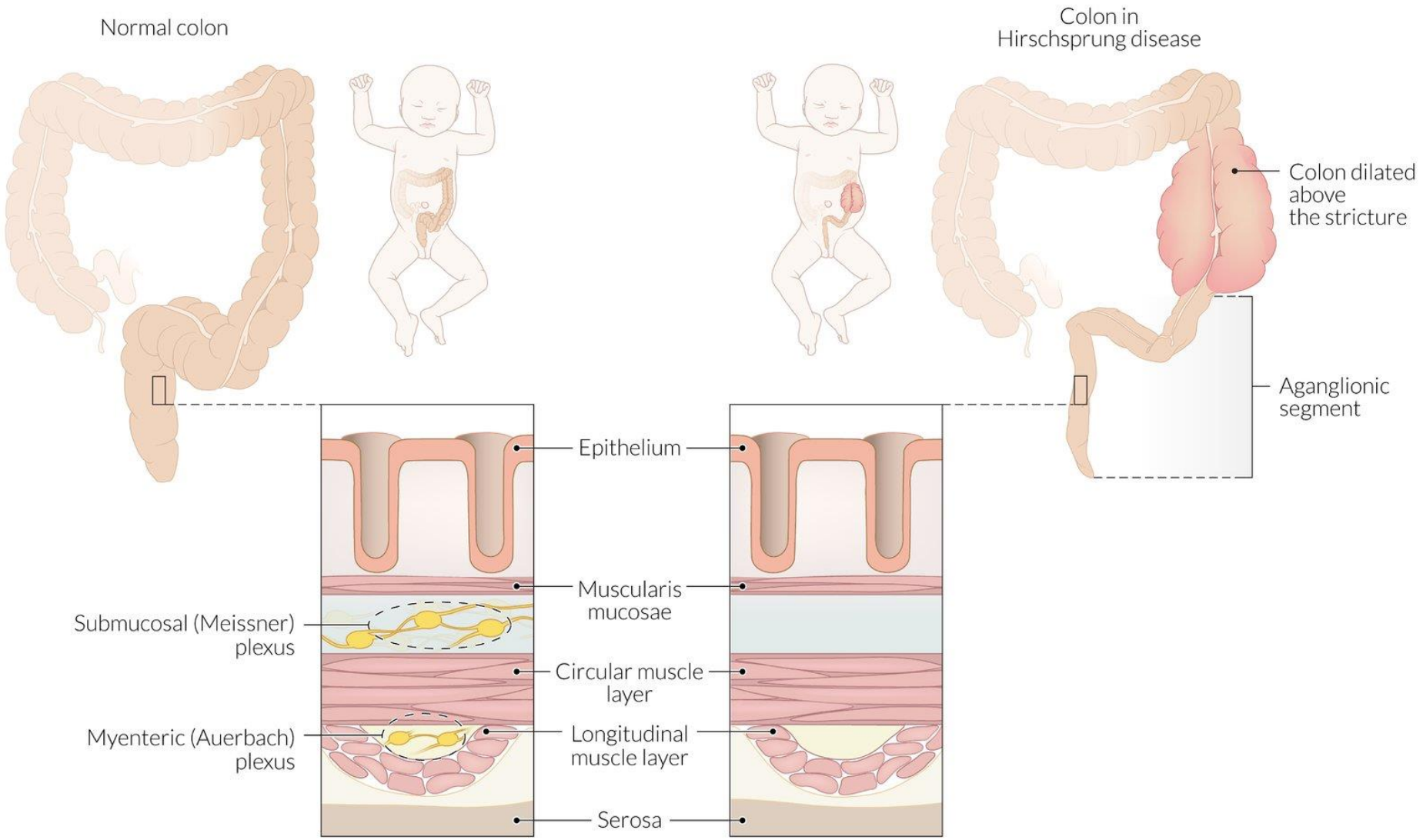
❖ Etiology

- **Genetic causes**: RET gene mutations associated with multiple endocrine neoplasia type 2 (MEN2) and familial Hirschsprung disease
- **Associated conditions**: Trisomy 21, Multiple endocrine neoplasia type 2

❖ Pathophysiology

- The absence of parasympathetic ganglion cells in the myenteric and submucosal plexuses of the intestine due to failure to migrate to distal bowel (craino-caudal migration) → distal bowel fails to relax → functional, distal bowel obstruction

Hirschsprung Disease (HD)



Hirschsprung Disease (HD)

❖Distribution of Disease (subtypes)

- All patients with Hirschsprung Disease have absence of ganglia in rectum
- **Proximal distribution of aganglionosis varies**
  - Short-segment (rectum only)
  - Rectosigmoid (~75% of all patients)
  - Long-segment (distal to mid transverse colon)
  - Total colonic HD (mostly associated with trisomy 21)
  - All of colon and some or all of small bowel (rare)

❖Clinical presentation

- **Early presentation**: Most patients present during the neonatal period with delayed passage of meconium (> 48 hours) and neonatal distal intestinal obstruction (abdominal distention and bilious vomiting)
- **Late presentation**: Some patients may present in toddler years or older with failure to thrive, chronic constipation, and/or enterocolitis

Hirschsprung Disease (HD)

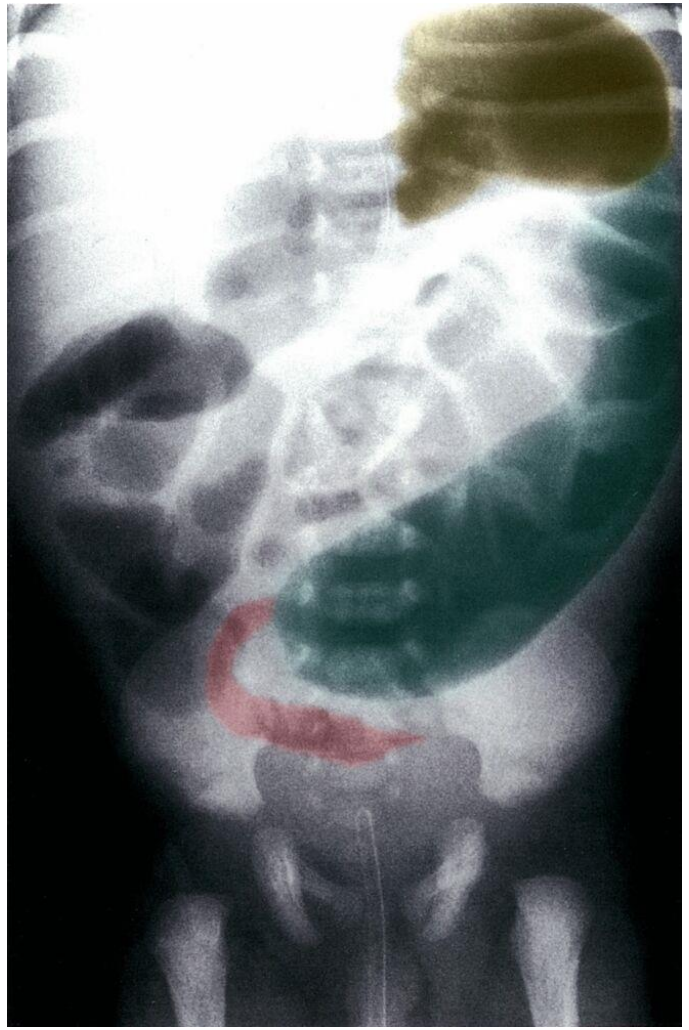
❖ Diagnostics

1. **Abdominal x-ray**
   - **Indication**: usually performed initially in newborns with abdominal distention and delayed passage of meconium
   - **Findings**: decreased or absent air in rectum and dilated colon segment immediately proximal to aganglionic region
2. **Barium enema**
   - **Indication**: usually performed in addition to x-ray, to localize and determine the length of the aganglionic segment prior to surgery
   - **Findings**: Change in caliber along the affected intestinal segment (transition zone)
3. **Anorectal manometry**
   - **Indication**: screening in atypical presentations or in older children
   - **Findings**: Absent relaxation reflex of the internal sphincter after stretching of the rectum

Hirschsprung Disease – Abdominal X-ray

X-ray AP view of a 5-day-old newborn

- A massively dilated colon (green overlay) is seen proximal to a narrowed distal segment (red overlay).
- The appearance and location of the narrowing are consistent with short-segment Hirschsprung disease.
- Yellow overlay: gastric bubble
- Reversal of recto-sigmoid ratio (rectum smaller than sigmoid)
- Transition zone (Funnel shape)



Hirschsprung Disease (HD)

❖Diagnostics cont.

4. Rectal biopsy
   - **Indication**: confirmatory test
   - **Procedure**
     - Full-thickness biopsy under general anesthesia
     - Suction rectal biopsy
   - **Findings**
     - Absence of ganglion cells in an adequate tissue sample
     - Elevated acetylcholinesterase activity
     - Hyperplasia of the parasympathetic nerve fibers
     - Absent calretinin immunostained fibers
     - Stained forAcetylcholinesterase historically (positive in hypertrophic nerves) and more commonly, Calretinin (negative in aganglionosis)

Hirschsprung Disease (HD)

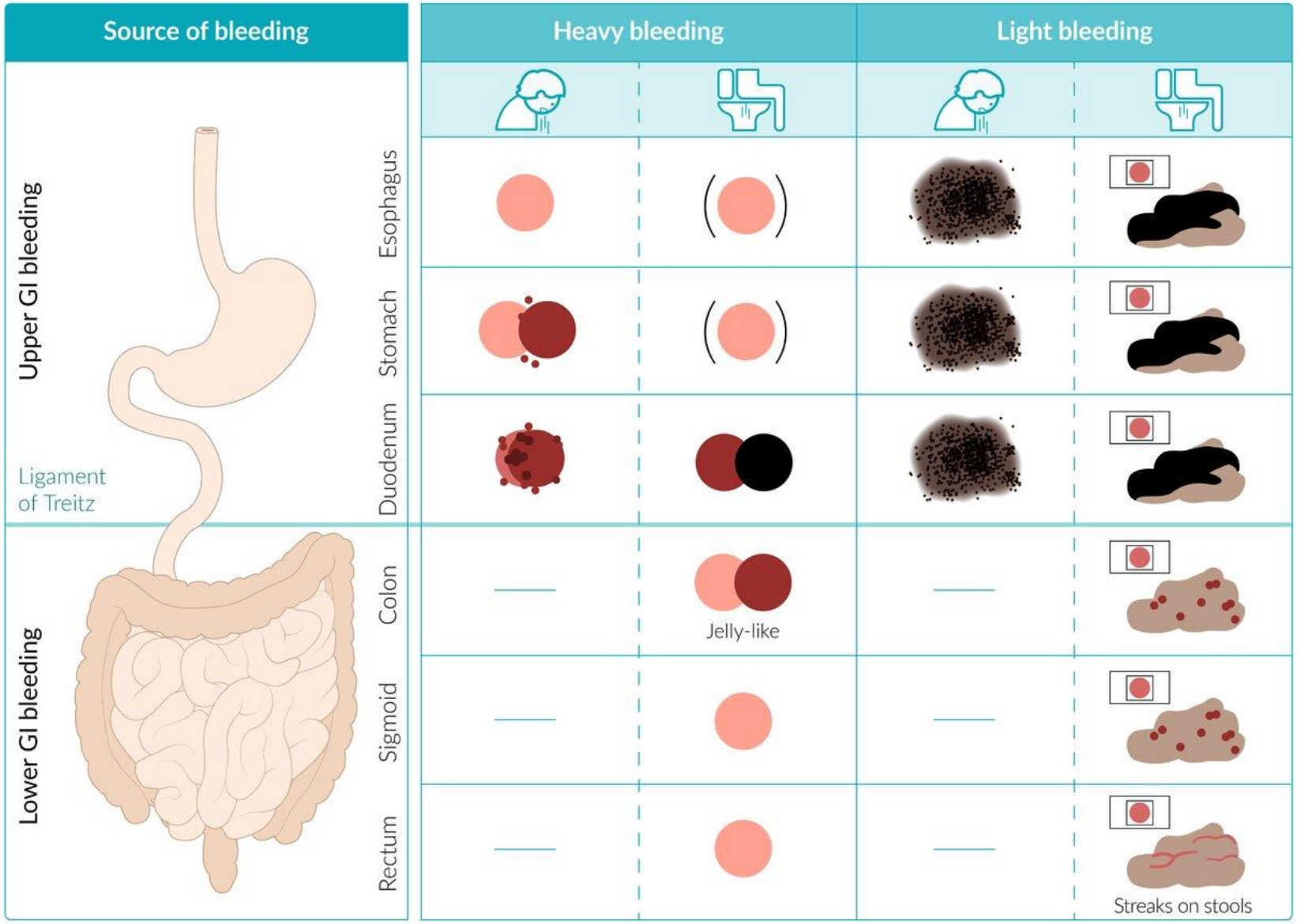
Initial management	○ IV fluid resuscitation ○ Nasogastric decompression ○ IV antibiotics with anaerobic coverage, if indicated ○ Colonic irrigation may be required.
Surgical correction Procedures	○ Total trans-anal endorectal pull-through: preferred method that can be done in one stage, usually after age of 3 months. ○ Abdominoperineal pull-through (Soave procedure): traditionally performed in two stages: • **First stage**: diverting colostomy to relieve the dilated bowel. • **Second stage**: Resection of the aganglionic segment ○ If rectal irrigation is not working at any time, or diagnosis was made at old age, leveling colostomy is the better choice before pull through procedure (at level of ganglionic cells)
Postoperative management	○ Routine colonic irrigation ○ Prophylactic antibiotic therapy

GI bleeding in pediatrics

Gastrointestinal bleeding

Upper gastrointestinal bleeding	Lower gastrointestinal bleeding
~ 70–80% of GI hemorrhages	~ 20–30% of all GI hemorrhages
The source of the bleeding is proximal to the ligament of Treitz (suspensory muscle of the duodenum)	The source of the bleeding is distal to the ligament of Treitz, usually in the colon
Presents with hematemesis (vomiting of red blood or coffee ground-like material) and/or melena (black, tarry stools)	Presents with hematochezia (bright red or maroon-colored blood or fresh clots per rectum)

Gastrointestinal bleeding

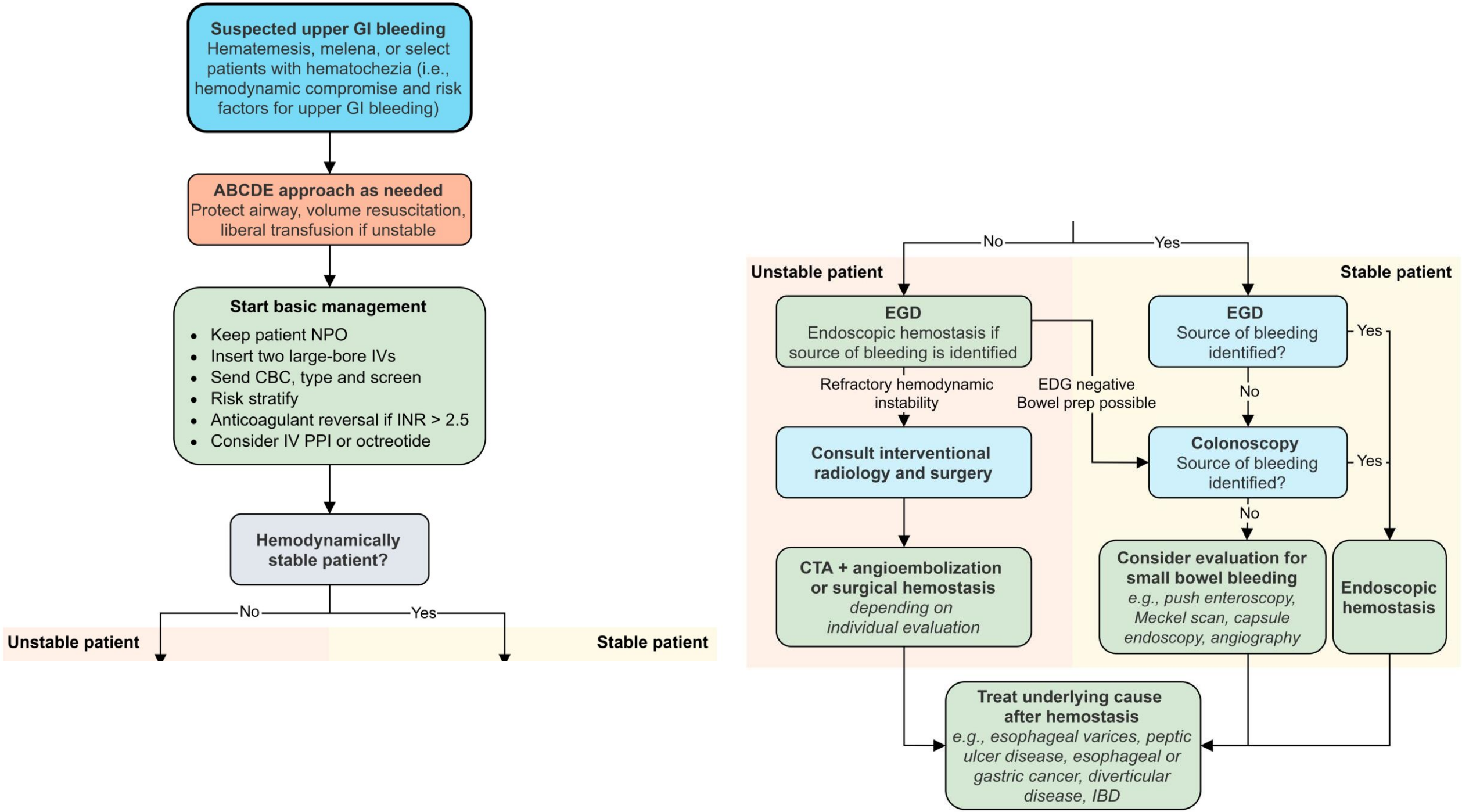


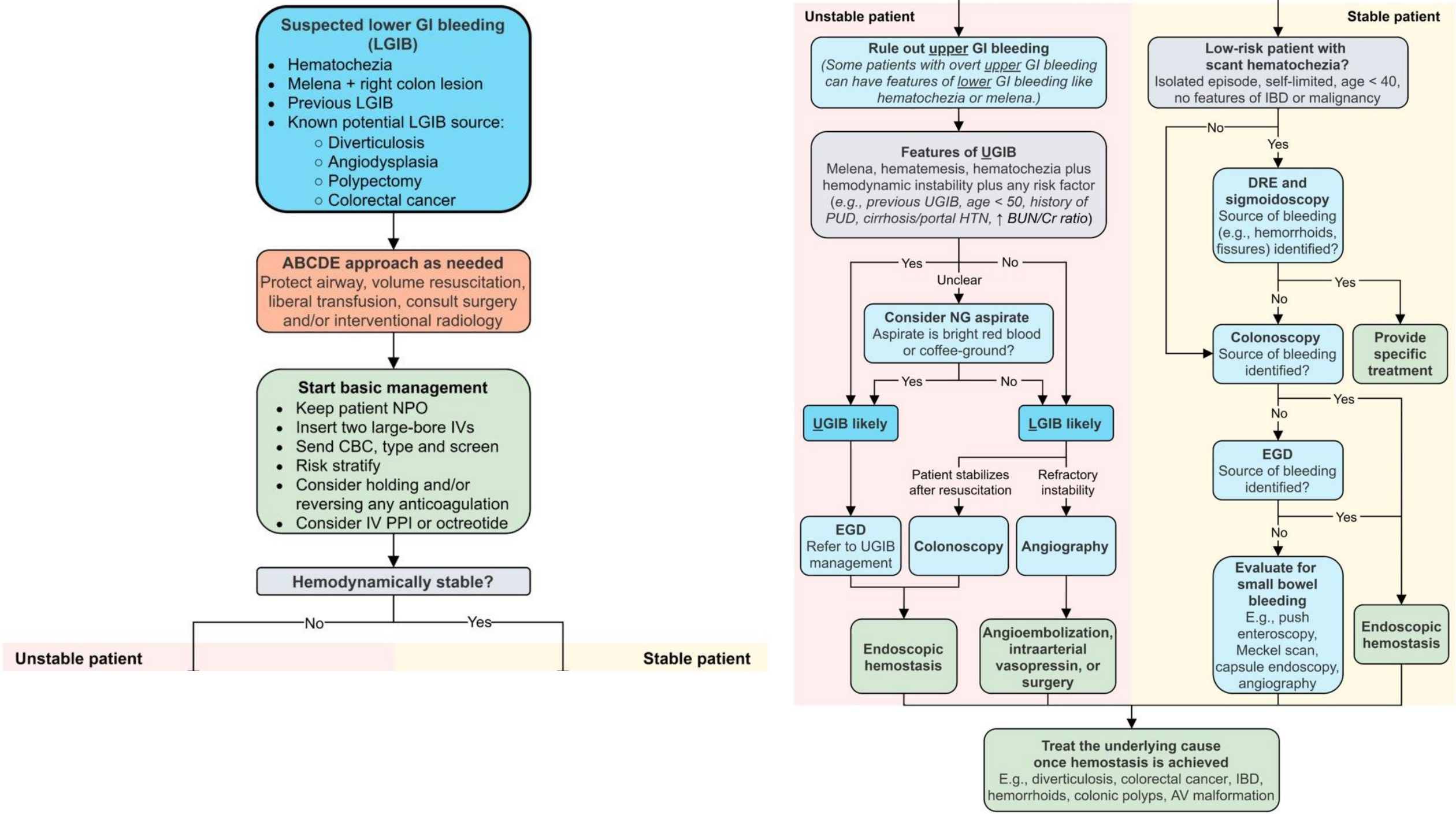
The blood volume is approximately 80 ml/kg

TABLE 3-1 SIGNS AND SYMPTOMS OF HEMORRHAGE BY CLASS

PARAMETER	CLASS I	CLASS II (MILD)	CLASS III (MODERATE)	CLASS IV (SEVERE)
Approximate blood loss	<15%	15–30%	31–40%	>40%
Heart rate	↔	↔/↑	↑	↑/↑↑
Blood pressure	↔	↔	↔/↓	↓
Pulse pressure	↔	↓	↓	↓
Respiratory rate	↔	↔	↔/↑	↑
Urine output	↔	↔	↓	↓↓
Glasgow Coma Scale score	↔	↔	↓	↓
Base deficit[a]	0 to −2 mEq/L	−2 to −6 mEq/L	−6 to −10 mEq/L	−10 mEq/L or less
Need for blood products	Monitor	Possible	Yes	Massive Transfusion Protocol

[a] Base excess is the quantity of base (HCO_3^-, in mEq/L) that is above or below the normal range in the body. A negative number is called a base deficit and indicates metabolic acidosis.


Suspected upper GI bleeding
Hematemesis, melena, or select patients with hematochezia (i.e., hemodynamic compromise and risk factors for upper GI bleeding)
ABCDE approach as needed
Protect airway, volume resuscitation, liberal transfusion if unstable
Start basic management
Keep patient NPO
Insert two large-bore IVs
Send CBC, type and screen
Risk stratify
Anticoagulant reversal if INR > 2.5
Consider IV PPI or octreotide
Hemodynamically stable patient?
No
Yes
Unstable patient
Stable patient
No
Yes
Unstable patient
Stable patient
EGD
Endoscopic hemostasis if source of bleeding is identified
EGD
Source of bleeding identified?
Yes
Refractory hemodynamic instability
EDG negative
Bowel prep possible
No
Consult interventional radiology and surgery
Colonoscopy
Source of bleeding identified?
Yes
No
CTA + angioembolization or surgical hemostasis
depending on individual evaluation
Consider evaluation for small bowel bleeding
e.g., push enteroscopy, Meckel scan, capsule endoscopy, angiography
Endoscopic hemostasis
Treat underlying cause after hemostasis
e.g., esophageal varices, peptic ulcer disease, esophageal or gastric cancer, diverticular disease, IBD


Suspected lower GI bleeding (LGIB)
Hematochezia
Melena + right colon lesion
Previous LGIB
Known potential LGIB source:
Diverticulosis
Angiodysplasia
Polypectomy
Colorectal cancer
ABCDE approach as needed
Protect airway, volume resuscitation, liberal transfusion, consult surgery and/or interventional radiology
Start basic management
Keep patient NPO
Insert two large-bore IVs
Send CBC, type and screen
Risk stratify
Consider holding and/or reversing any anticoagulation
Consider IV PPI or octreotide
Hemodynamically stable?
No
Yes
Unstable patient
Stable patient
Unstable patient
Stable patient
Rule out upper GI bleeding
(Some patients with overt upper GI bleeding can have features of lower GI bleeding like hematochezia or melena.)
Features of UGIB
Melena, hematemesis, hematochezia plus hemodynamic instability plus any risk factor (e.g., previous UGIB, age < 50, history of PUD, cirrhosis/portal HTN, ↑ BUN/Cr ratio)
Yes
No
Unclear
Consider NG aspirate
Aspirate is bright red blood or coffee-ground?
Yes
No
UGIB likely
LGIB likely
Patient stabilizes after resuscitation
Refractory instability
EGD
Refer to UGIB management
Colonoscopy
Angiography
Endoscopic hemostasis
Angioembolization, intraarterial vasopressin, or surgery
Low-risk patient with scant hematochezia?
Isolated episode, self-limited, age < 40, no features of IBD or malignancy
No
Yes
DRE and sigmoidoscopy
Source of bleeding (e.g., hemorrhoids, fissures) identified?
Yes
No
Provide specific treatment
Colonoscopy
Source of bleeding identified?
Yes
No
EGD
Source of bleeding identified?
Yes
No
Evaluate for small bowel bleeding
E.g., push enteroscopy, Meckel scan, capsule endoscopy, angiography
Endoscopic hemostasis
Treat the underlying cause once hemostasis is achieved
E.g., diverticulosis, colorectal cancer, IBD, hemorrhoids, colonic polyps, AV malformation

Etiology Classification

	Newborn	1m – 1 year	1 – 2 years	> 2 years
UGIB	1. Hemorrhagic disease 2. Swallowed maternal blood	1. Esophagitis 2. Gastritis	1. PUD	1. Varices
LGIB	1. Anal fissure 2. NEC	1. Anal fissure 2. Intussusception	1. Polyps 2. Meckel Diverticulum	1. Polyps 2. IBD 3. Intussusception

Necrotizing enterocolitis

❖Epidemiology

- NEC is the most common cause of acute abdomen in premature infants.
- Peak incidence: 2nd–4th week after birth

❖Pathophysiology

- Idiopathic NEC: reduced bowel perfusion combined with oral feeding causes bacterial translocation into the bowel wall

❖Risk factors

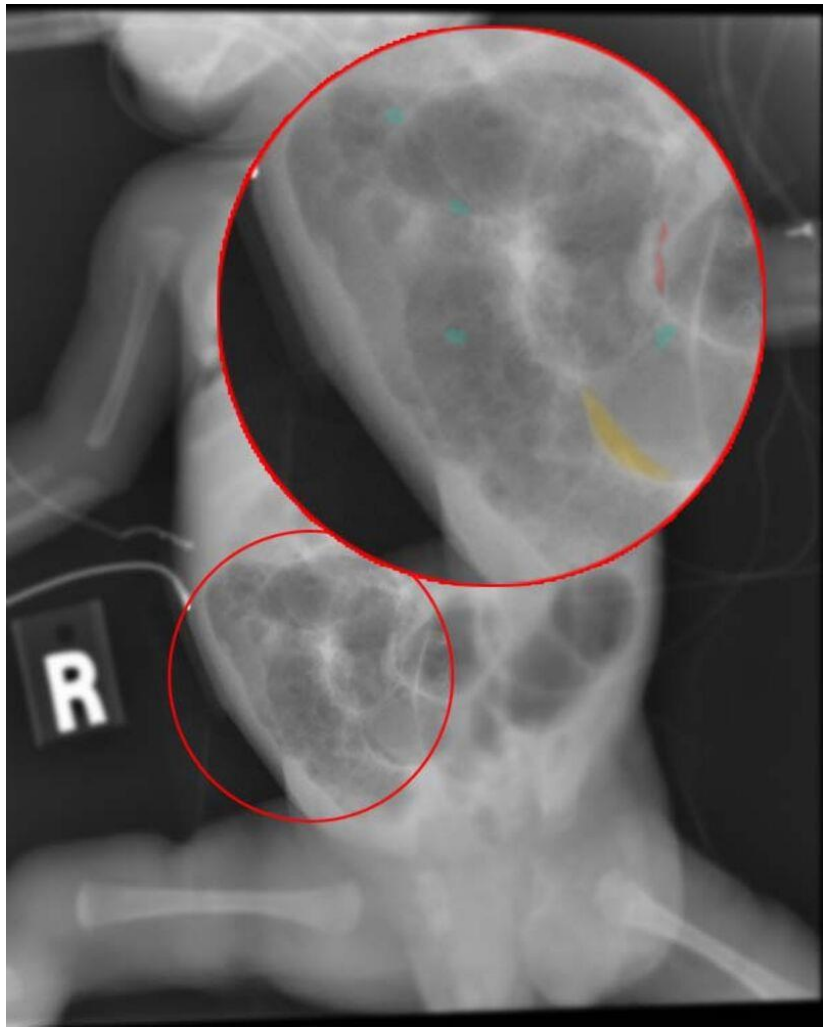
- Prematurity
- Low birth weight
- Congenital heart anomalies and perinatal asphyxia (Difficult delivery)

❖Clinical features (Bell staging criteria)

- **Stage I**: **Suspected NEC**: Lethargy, distended and shiny abdomen, gastric retention, vomiting, diarrhea, rectal bleeding
- **Stage II**: **Proven NEC**: Stage I symptoms + abdominal tenderness, visible intestinal loops lacking peristalsis
- **Stage III**: **Advanced NEC**: Intestinal perforation, symptoms of sepsis, flank redness and if left untreated: rapid progression to disseminated intravascular coagulation (DIC) and shock

Necrotizing enterocolitis

- **Diagnostics**
  - **Laboratory tests**
    - Check for signs of DIC (Platelets, PT, PTT, Fibrinogen)
    - Arterial blood gas analysis: Metabolic acidosis is associated with advanced NEC
  - **Imaging (Abdominal radiography)**
    - Pneumatosis intestinalis: bubbles of gas within the wall of the intestine
    - Portal venous gas
    - Increased intestinal wall thickness



Necrotizing enterocolitis

Treatment should be initiated promptly when NEC is suspected to prevent complications such as perforation, peritonitis, and sepsis.

- **Supportive care**:
  - Stop enteral feeding → parenteral feeding and substitution of fluids
  - Gut decompression via nasogastric tube
- **IV broad-spectrum antibiotics**: e.g., ampicillin, gentamicin, and metronidazole for anaerobic coverage
- **Radiographic monitoring**: plain supine abdominal radiographs every 6–12 hours in the initial phase of the disease
- **Surgery**: primary peritoneal drainage and/or laparotomy with necrotic bowel excision
  - **Indications**: perforation, peritonitis, clinical worsening despite medical therapy and/or abdominal mass (Fixed-loop sign) with erythema of the abdomen wall

Meckel diverticulum

❖The rule of two's

- Meckel diverticulum occurs in 2% of the population, 2% are symptomatic, mostly in children < 2 years, affects males twice as often as females, is located 2 feet proximal to the ileocecal valve, is ≤ 2 inches long, and can have 2 types of mucosal lining.

❖Pathophysiology

- Incomplete obliteration of the omphalomesenteric duct → persistence of the proximal (intestinal) segment of the duct → Meckel diverticulum
- Other anomalies arise from failed regression of the omphalomesenteric duct Umbilical polyp, omphalomesenteric fistula, umbilical sinus, umbilical cyst, and persistent fibrous band.

❖Anatomy

- It is a true diverticulum that occurs in the ileum
- There may be 2 types of mucosal lining: native ileal mucosa and ectopic mucosa

Meckel diverticulum – Clinical features

- Asymptomatic (most common manifestation; detected incidentally)
- Symptomatic
  - Lower gastrointestinal bleeding (most common feature)
    - Presence of ectopic gastric mucosa or pancreatic tissue → acid or enzyme secretion within the diverticulum → ileal ulceration → bleeding
    - Can manifest as:
      - **Hematochezia**: Indicates a brisk hemorrhage
      - **Tarry stools**: Indicate a slow hemorrhage
      - **Currant jelly stools**: Indicate intussusception with bowel ischemia
  - Abdominal pain (typically in the right lower quadrant)
    - Due to volvulus or intussusception → Episodic, crampy pain, bilious vomiting, currant jelly stools
    - Diverticulitis +/- perforation → Like appendicitis

Meckel diverticulum

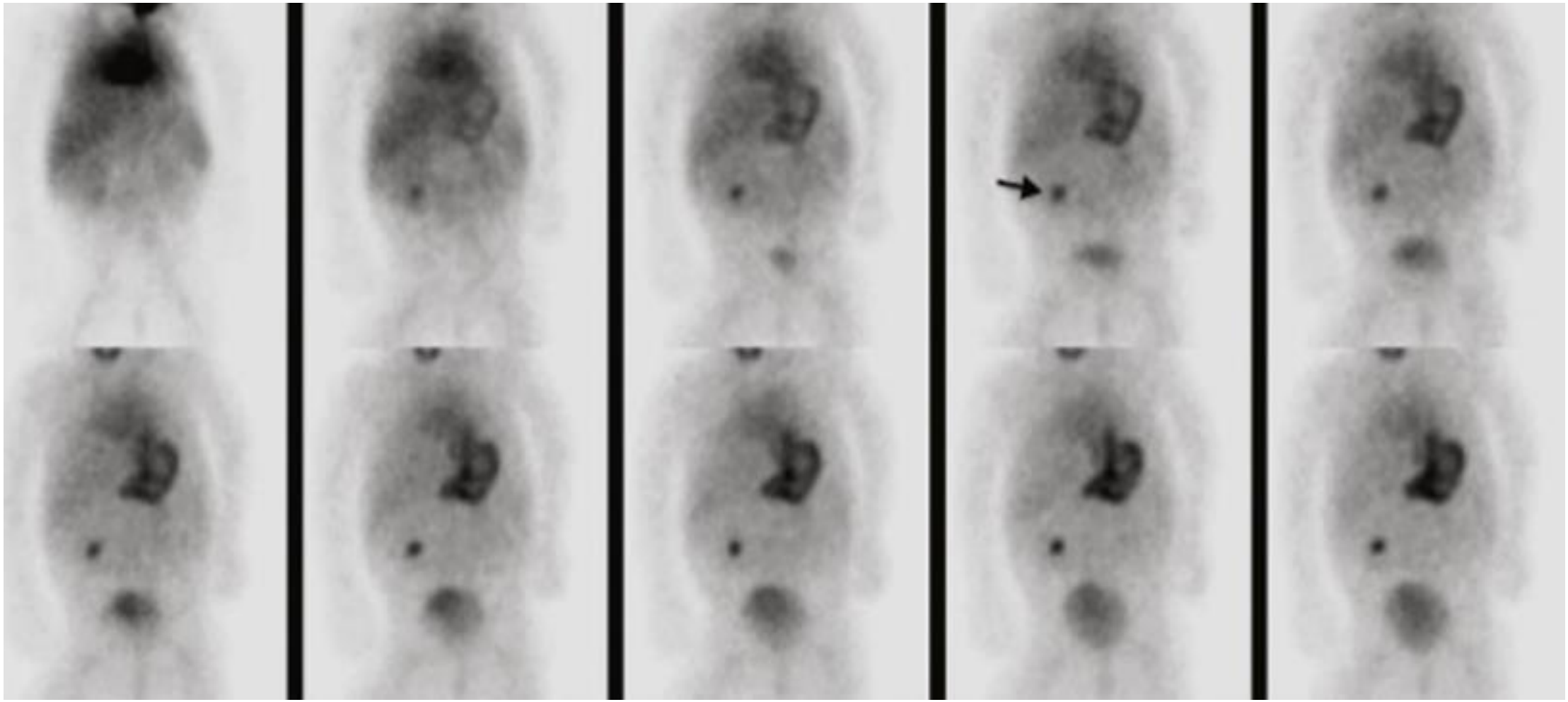
❖ Diagnostics

- The initial work-up follows the same protocol as that for lower gastrointestinal bleeding and/or acute abdomen.
- Only the imaging tests specific to Meckel diverticulum are mentioned here.
  - Meckel scintigraphy scan (Meckel scan): a noninvasive nuclear medicine imaging technique using radiolabeled technetium (99mTc), which is preferentially absorbed by the gastric mucosa and can identify ectopic gastric mucosa (sensitivity of 85% and a specificity of 95%)
  - Ultrasound for intussusception
  - CT Abd/Pelvis for obstruction

❖ Pre-op management

- Hydration/transfusion
- NG decompression
- Antibiotics

Meckel scintigraphy scan (Meckel scan)



Meckel diverticulum – Surgery

- **Asymptomatic Meckel diverticulum**
  - Incidentally detected on imaging studies: no treatment necessary
  - Incidentally detected on laparotomy/laparoscopy
    - Children or young adults: surgical resection of all incidentally detected Meckel diverticula
    - Adults < 50 years: surgical resection only for Meckel diverticula that have a high risk of developing complications
    - Adults > 50 years: no treatment necessary
- **Symptomatic or complicated Meckel diverticulum**
  - Initial stabilization of the patient
  - Surgical resection of all symptomatic/complicated Meckel diverticula
- **Surgical procedures**
  - **Segmental resection**: indicated for a Meckel diverticulum that is bleeding, has a broad base, or a palpable abnormality
  - **Diverticulectomy**: Meckel diverticulum is resected at the base

Intussusception

- **Definition**: Full-thickness invagination (telescoping) of the proximal bowel into the distal contiguous intestine
- **Epidemiology**
  - 80-90% of intussusceptions occur in children between 3 months and 2 years of age
  - The most common intussusception is ileocolic
  - Ileoileal or colocolic intussusceptions are less frequent and associated more often with a pathologic lead point (e.g., Meckel's, LN, Tumor, lymphoma, Polyp)
- **Primary intussusception**
  - Intussusception frequently follows viral illnesses like gastroenteritis or respiratory infections. This is because of hyperplasia of Peyer’s patches. These patches expand around the distal ileum, narrowing the lumen and promoting intussusception.
  - No identifiable pathologic lead point
  - Adenoviruses, rotaviruses, and rotavirus immunization have been implicated

Intussusception

❖Secondary intussusception

- Identifiable pathologic lead point
- The incidence varies from 1.5–12%
- The **most common lead point** is a Meckel diverticulum followed by polyps and duplications. Other benign lead points include the appendix, hemangiomas, carcinoid tumors, foreign bodies
- Outside the typical age for intussusception
- Usually ileo-ileal or colocolic

❖Clinical features

- Acute cyclical colicky abdominal pain (sudden screaming or crying spells), often with legs drawn up, with asymptomatic intervals
- Less than 15% of patients with intussusception present with the **classic triad of** abdominal pain, a palpable sausage-shaped abdominal mass, and blood per rectum
- lethargy, dehydration, and abdominal distention are also frequent findings

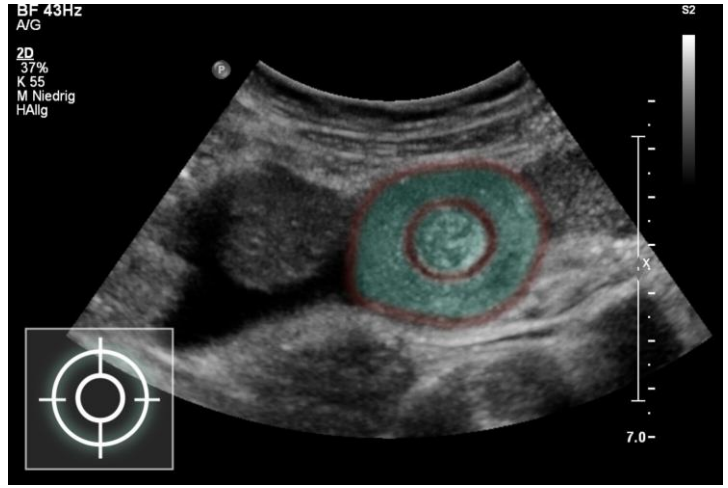
Intussusception

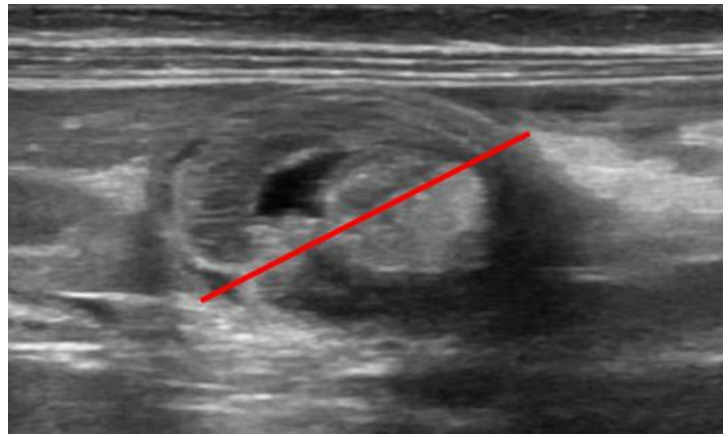
❖Diagnostics

- Abdominal ultrasound (best initial test): often sufficient to confirm diagnosis:
  - Target sign
  - Pseudokidney sign
- Contrast or pneumatic enema using ultrasound or fluoroscopy (best confirmatory test):
  - Pneumatic insufflation
  - Coiled-spring sign

❖Management

- Initial steps: nasogastric decompression and fluid resuscitation
- Nonsurgical management (performed under continuous ultrasound or fluoroscopic guidance)
  - Air enema: treatment of choice
  - Hydrostatic reduction





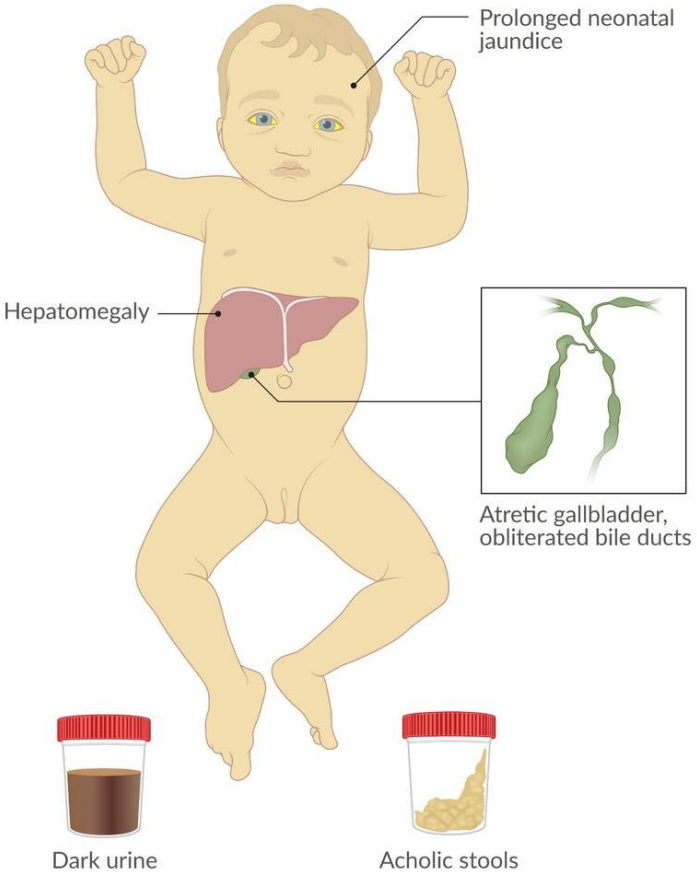
Intussusception

- **Operative treatment**
  - **Indications**
    - Irreducibility by pneumatic or hydrostatic means
    - Peritonitis (perforation), shock, and hemodynamic instability
    - Age > 6 years
    - Duration of symptoms > 24 hours
  - **Procedure**
    - Gentle manipulation by pushing the intussusceptum out of the intussuscipiens (rather than by pulling with traction)
    - If attempts at reduction cause undue injury to the bowel wall, if bowel necrosis or perforation is present, or if a pathologic lead point is identified or suspected, resection and primary anastomosis are indicated

Neonatal surgical jaundice

Biliary atresia

- **Definition**: obliteration or discontinuity of the extrahepatic biliary system, most commonly of the common bile duct
- **Pathophysiology**: discontinuity of the biliary system due to obliteration or fibrosis → obstruction of bile flow (cholestasis) → secondary biliary cirrhosis and portal hypertension
- **Clinical features**
  - Jaundice
    - Prolonged neonatal jaundice (> 2 weeks)
    - Onset 2 weeks after birth is possible.
  - Acholic stools, dark urine
  - Hepatomegaly

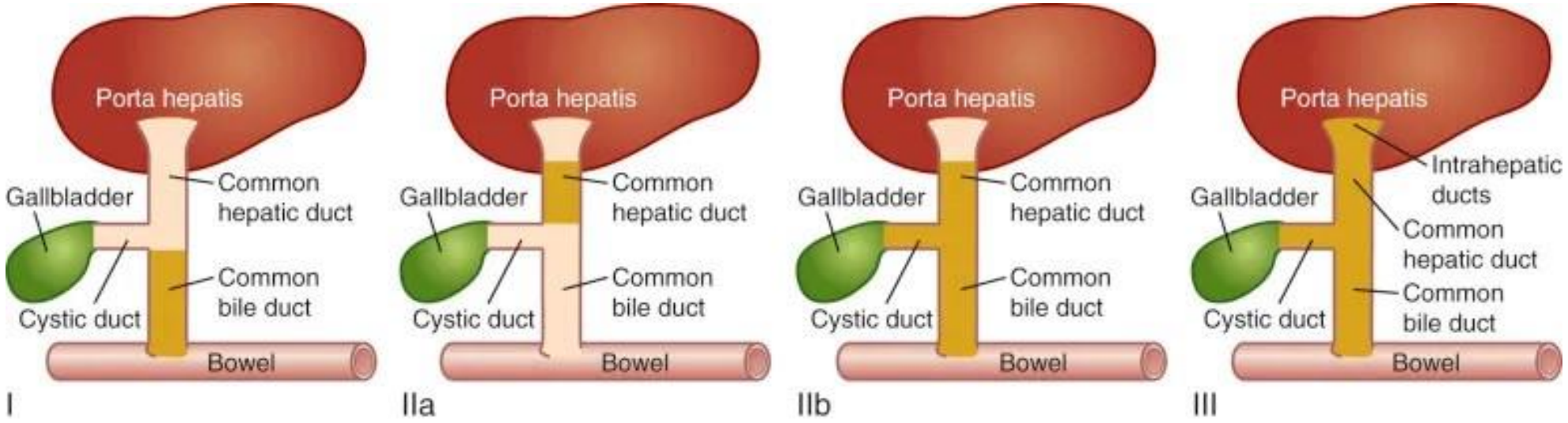


Biliary atresia

- Infants with BA can be grouped into three categories:
  - **BA without any other anomalies or malformations**: This pattern is sometimes referred to as perinatal BA and occurs in 70-85% of infants with BA. Typically, these children are born without visible jaundice, but within the first two months of life, jaundice develops, and stools become progressively acholic.
  - **BA associated with laterality malformations**: This pattern is also known as biliary atresia splenic malformation (BASM) or "embryonal" biliary atresia and occurs in 10-15% of infants with BA. The laterality malformations include situs inversus, asplenia or polysplenia, malrotation, interrupted inferior vena cava, and cardiac anomalies.
  - **BA associated with other congenital malformations**: This occurs in the remaining 5-10% of BA cases; associated congenital malformations include intestinal atresia, imperforate anus, kidney anomalies, and/or cardiac malformations.

Kasai classification of biliary atresia

Type I	Atresia at the level of the common bile duct (patent cystic and CHD)
Type IIa	Atresia is at the level of the common hepatic duct (patent cystic and CBD)
Type IIb	Atresia in the cystic duct, CBD and CHD
Type III	Is the most frequent type, atresia occurs at the porta hepatis



Biliary atresia – Diagnostics (stepwise)

Laboratory analysis	• Conjugated hyperbilirubinemia • ↑ AST, ALT, ALP and GGT
Ultrasound	**Findings**: Absence of the gallbladder and no dilatation of the biliary tree **Note**: The absence of a gallbladder is highly suggestive of the diagnosis of BA, but the presence of a gallbladder does not exclude the diagnosis
Hepatobiliary scintiscanning	If there is a good hepatic uptake with no evidence of excretion into the bowel after 24h with excretion by alternate route like the kidney and bladder then it is biliary atresia.
Percutaneous Liver biopsy	Active inflammation with bile duct degeneration and fibrosis, Bile duct proliferation, and Portal stromal edema
Intraoperative cholangiography (Gold standard)	• Performed if liver biopsy findings suggest an obstructive etiology • If the intraoperative cholangiogram demonstrates biliary obstruction (i.e., if the contrast does not fill the biliary tree or reach the intestine), the surgeon should perform a Kasai HPE at that time

Biliary atresia

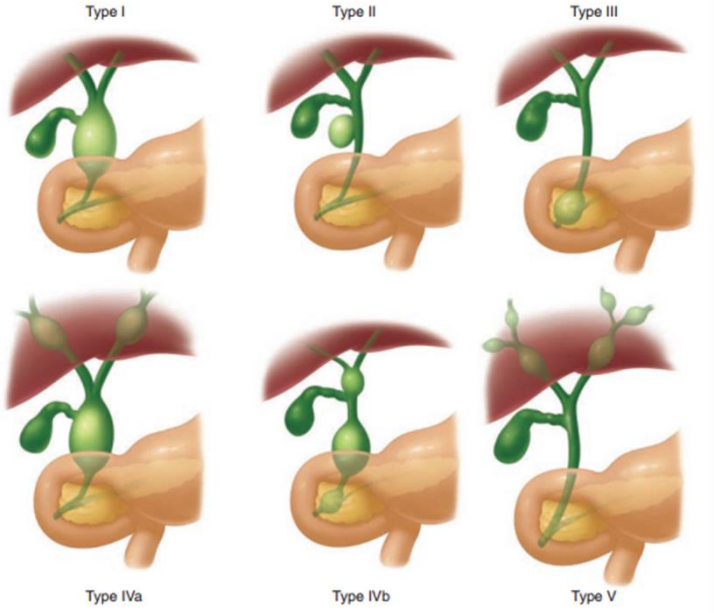
Treatment	○ Infants with BA who are jaundiced should be treated with supplements of fat-soluble vitamins and monitored for fat-soluble vitamin deficiencies. In addition, they should be given high-calorie formulas or other nutritional supplements as required to sustain normal rates of growth ○ **Kasai procedure (hepatoportoenterostomy)**: a connection is created between the liver and the small intestine to allow for bile drainage. ○ **In cases of liver cirrhosis**: liver transplantation (primary reason for liver transplantation in children)
Complications	○ If undetected → Early biliary liver cirrhosis (at approx. 9 weeks of age) ○ Younger age at the time of the Kasai HPE is associated with better outcomes
Prognosis	○ 1/3: Completely treated by KASAI ○ 1/3: Present late with liver cirrhosis → liver transplantation ○ 1/3: Failed KASAI (cholangitis; jaundice >3m) → liver transplantation

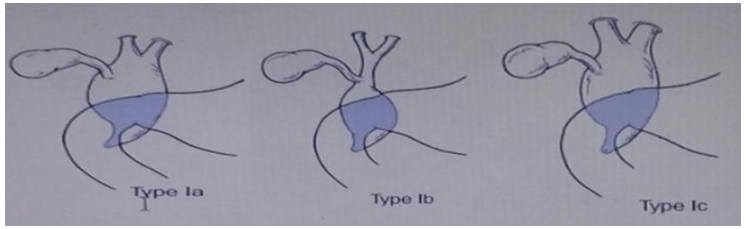
Choledochal cysts

- **Definition**: premalignant condition characterized by extrahepatic and/or intrahepatic cystic dilation of the biliary tree
- **Epidemiology**
  - More in females (4:1)
  - More common in Asians
  - Most commonly diagnosed during childhood
  - Increase risk for malignancy and cholangitis
- **Clinical features**: symptoms typically develop before the age of 10
  - The **classic triad** of abdominal pain, palpable abdominal mass, and jaundice (The entire triad is present in fewer than 30% of patients)
  - **Clinical presentation varies with age**:
    - **Prenatal**: Cystic mass in the abdomen
    - **Children ≤ 12 months**: obstructive jaundice and abdominal mass
    - **Children > 12 months and adults**: right upper quadrant pain (most common), fever
    - **In adults**: recurrent ascending cholangitis, obstructive jaundice or recurrent pancreatitis

Choledochal cysts – Anatomical classification

Type 1	• It is the most common (>50% of cases) • Cystic/saccular or fusiform dilation of the CBD (extrahepatic) • Type 1a consists of cystic dilation of the entire common bile duct • Type 1b is cystic dilation of a segment of the common bile duct • Type 1c is fusiform dilation of the common bile duct
Type 2	• It is a true, single, extrahepatic diverticulum
Type 3	• Also referred to as a choledochocele , usually has a normal CBD and main pancreatic duct with cystic dilation of the distal CBD that is either intraduodenal or intrapancreatic in location • The choledochocele is usually stenotic at their openings due to chronic inflammation
Type 4	• Type 4A: Is composed of multiple cysts located in both intrahepatic and extrahepatic bile ducts • Type 4B: Is composed of multiple cysts located extrahepatic bile duct only
Type 5	• Intrahepatic single or multiple cysts only without extrahepatic dilation • Type V cysts in conjunction with hepatic fibrosis are commonly referred to as **Caroli disease**

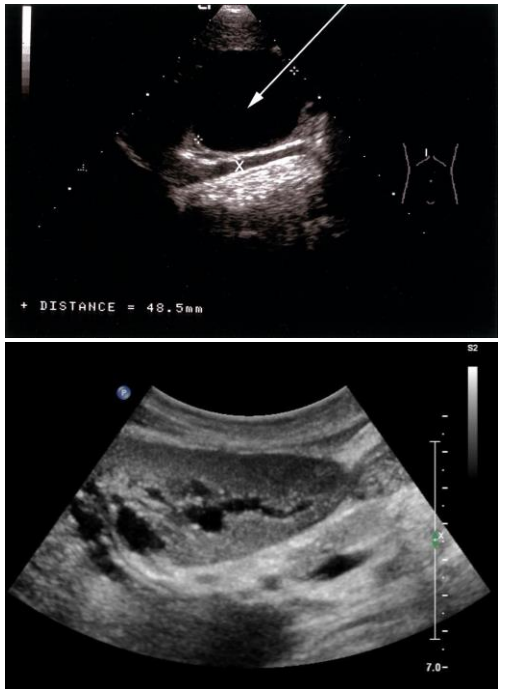




Choledochal cysts – Diagnostics

❖Laboratory studies

- ↑ direct bilirubin
- AST, ALT, GGT, ALP → assist liver and biliary damage
- Coagulation profiles
- CBC → look for anemia



❖Imaging

- Ultrasound (Initial imaging modality of choice)
- CT of the abdomen (first study in adults)
- HIDA scan: if an anomaly was detected prenatally
- Cholangiography: essential for diagnosis confirmation, identification of associated ductal pathology (e.g., choledocholithiasis, cholangiocarcinoma), and mapping of the biliary tree for surgical planning
- MRCP: usually the method of choice for preoperative imaging of the biliary tree

Choledochal cysts

❖ Treatment

- **Treatment according to types**
  - Type 1: removal biliary tree and GB + hepaticojejunostomy
  - Type 2: diverticulum excision
  - Type 3: sphincterotomy
  - Type 4: hepaticojejunostomy
  - Type 5: liver transplantation
- The treatment of choledochal cysts is typically complete surgical excision at the time of presentation, with the exception of type V (multiple intrahepatic cysts) which may benefit from conservative therapy including percutaneous drainage and medical management

❖ Complications

- Ascending Cholangitis, Pancreatitis, Biliary stone formation, Anastomotic stricture, Bowel obstruction, Malignancy: arise from the distal CBD

Abdominal Masses of Childhood

Neuroblastoma

❖Definition

- A malignant, embryonal neuroendocrine neoplasm of the sympathetic nervous system that originates from neural crest cells, potentially secretes catecholamines, and is usually found in the adrenal glands or sympathetic ganglia

❖Epidemiology

- Most common malignancy of the adrenal medulla in infants and third most common childhood cancer overall following leukemia and brain tumors
- Mean age at diagnosis: 1–2 years
- The majority of children have progressed to advanced-stage disease by the time of diagnosis.

❖Etiology

- Cause: unclear
- Genetic associations: Amplification and overexpression of N-myc oncogene

Neuroblastoma – Clinical features

❖General symptoms

- Failure to thrive or weight loss
- Fever
- Nausea, vomiting, loss of appetite

❖Paraneoplastic syndromes

- Chronic diarrhea → electrolyte imbalances
- Opsoclonus-myoclonus-ataxia: a paraneoplastic syndrome of unclear etiology characterized by rapid and multi-directional eye movements, rhythmic jerks of the limbs, and ataxia
- Possibly hypertension, tachycardia, palpitations, sweating, flushing

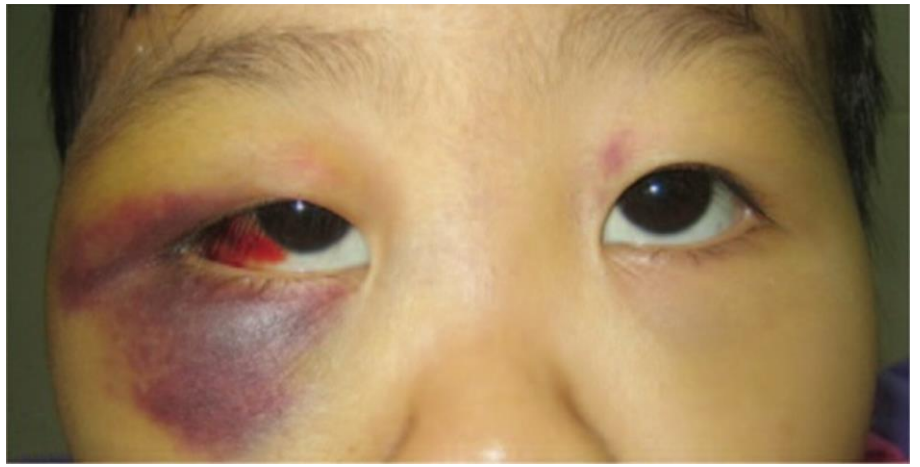
❖Localized symptoms

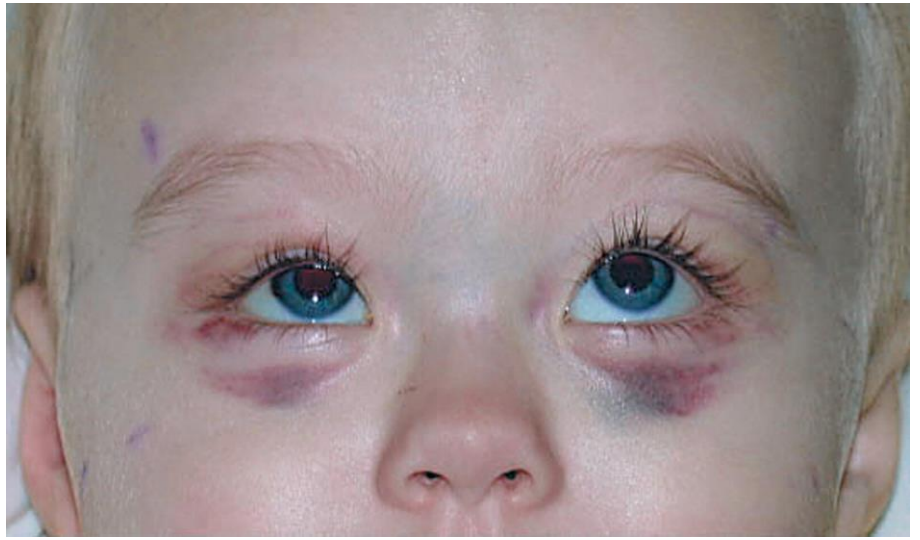
Location of primary tumors	Associated signs and symptoms
Abdomen (in > 60% of cases)	• Palpable, firm, irregular abdominal mass that may **cross the midline** (in contrast to nephroblastoma, which is smooth and usually does not cross the midline) • Abdominal distension and pain • Hepatomegaly • Constipation
Chest (in ~ 20% of cases): particularly paravertebral ganglia	• Spinal cord compression → back pain, weakness, numbness, ataxia, loss of bowel or bladder control • Scoliosis • Dyspnea, cough • Inspiratory stridor
Neck	• Horner syndrome • Symptoms due to spinal cord compressions
Location of metastases	**Associated signs and symptoms**
Orbit of the eye	• Periorbital ecchymoses (**"raccoon eyes"**) • Proptosis
Bones	• Bone pain • Anemia (bone marrow suppression)
Skin	• Subcutaneous nodules

Neuroblastoma – Clinical features

Periorbital ecchymoses

- 3-year-old girl with orbital neuroblastoma
- The periorbital region of the right eye appears swollen and shows a purplish-bluish hematoma. Proptosis of the eye and subconjunctival hemorrhage (hyposphagma) can also be seen.
- These findings are typical of a periorbital ecchymoses (raccoon eye) and are here associated with the spread of neuroblastoma.





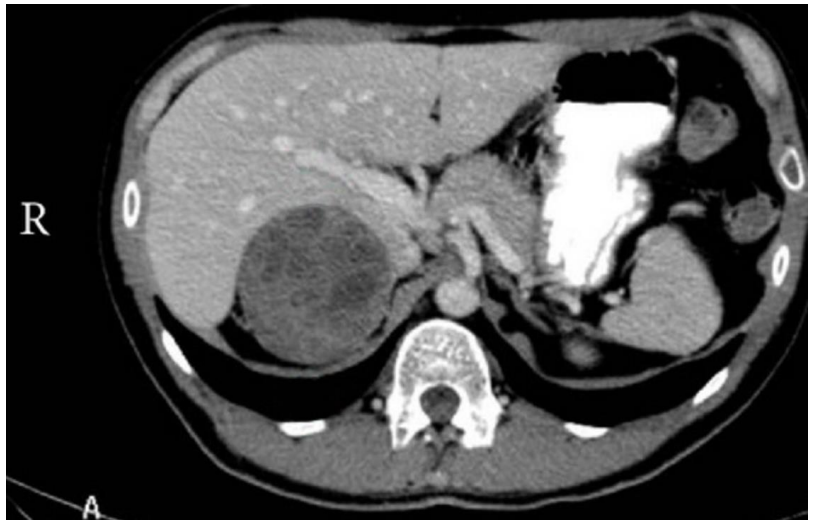
International Neuroblastoma Staging System (INSS) [13]	
Stage	**Definition**
1	• Localized tumor • Complete gross excision with or without microscopic residuals • Negative ipsilateral lymph nodes
2A	• Localized tumor • Incomplete gross excision • Negative ipsilateral lymph nodes
2B	• Localized tumor • Complete or incomplete gross excision • Positive ipsilateral lymph nodes
3	• Unresectable unilateral tumor that crosses the midline with or without lymph node involvement • Any tumor with positive contralateral lymph nodes • Midline tumor with bilateral tumor or lymph node involvement
4	• Any tumor with dissemination to distant lymph nodes or other organs (e.g., bone, liver, skin), with the exception of Stage 4S disease
4S	• Localized primary tumor with dissemination to skin, liver, or bone marrow, occurring in infants < 12 months

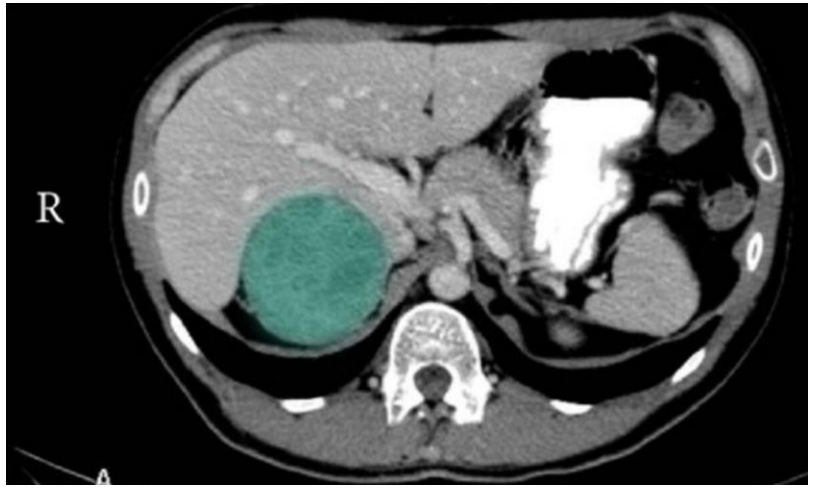
Neuroblastoma – Diagnostics

Laboratory tests	**Urine**	○ ↑ **Catecholamine metabolites**: homovanillic acid (HVA) and vanillylmandelic acid (VMA) in 24-hour urine ○ **Urinalysis**
	Blood	○ ↑ **Catecholamine metabolites** (HVA/VMA) ○ ↑ Lactate dehydrogenase (LDH), ferritin, neuron-specific enolase (NSE) ○ CBC with differential ○ Serum chemistry profile ○ Liver and kidney function tests
Other	**Imaging**	○ To identify the primary site ○ Abdominal ultrasound ○ CT or MRI (depending on the presumable site of the lesion)
	Scintigraphy	○ MIBG scan: Uptake scan of MIBG combined with a radioactive iodine tracer. ○ In MIBG non-avid tumors: technetium bone scan and plain radiographs
	Biopsy	○ Definitive diagnosis of neuroblastoma requires tissue biopsy

Neuroblastoma of the adrenal gland

- CT abdomen (with oral and IV contrast; axial plane)
- A large heterogeneous mass (green overlay) with little to no vascular enhancement is present in the area of the right adrenal gland.
- This is a common CT appearance of adrenal neuroblastoma. Although not present on this CT, accompanying calcifications are common.





Neuroblastoma – Treatment

Neuroblastoma patients are treated based on their risk category (low, intermediate, or high), which is based on the stage of their neuroblastoma (extent of disease), age at diagnosis, and the presence/absence of MYCN amplification.

Low risk	Generally, children with early-stage disease (Stages 1–2) and no MYCN amplification
Intermediate risk	Generally, children with intermediate and late-stage disease (Stages 3–4) and no MYCN amplification
High risk	Generally, children with late-stage disease and/or MYCN amplification
Stage 4S (an exception)	○ Better prognosis than other stage 4 neuroblastoma and spontaneous regression is very common ○ Children with Stage 4S belong to the low-risk or intermediate-risk groups unless they have a MYCN amplification, in which case they are high-risk patients

Neuroblastoma – Treatment

Treatment of neuroblastoma			
Treatment	Low-risk neuroblastoma	Intermediate-risk neuroblastoma	High-risk neuroblastoma
Observation only	X		
Preoperative chemotherapy (e.g., doxorubicin, cyclophosphamide, etoposide, and a platinum drug)	X	X	X
Surgery	X	X	X
Postoperative chemotherapy		X	X
Radiation		X	X
GD2 antibody, dinutuximab, granulocyte macrophage colony-stimulating factor (GM-CSF), interleukin 2 (IL-2), and cis-retinoic acid			X
Other postoperative therapies (e.g., MIBG therapy)			X

Nephroblastoma (Wilms Tumor)

❖ Epidemiology

- The second most common malignant abdominal tumor in children after neuroblastoma (5% of all childhood malignancies are nephroblastomas)
- Peak incidence: between 2 and 5 years (95% of cases are in < 10 years of age)
- Most common malignant neoplasm of the kidney in children

❖ Etiology

- The exact etiology of nephroblastoma remains unknown, but it is associated with several genetic mutations and syndromes.
- Genetic predisposition
  - Associated with loss of function mutations of tumor suppressor genes on chromosome 11
    - The WT1 (Wilms tumor 1) gene is the most important nephroblastoma tumor gene (mutated in ~ 10–20% of cases)
    - WT2 (Wilms tumor 2) gene

Nephroblastoma – Associated syndromes

- **WAGR syndrome**: Deletion of the 11p13 band leads to the deletion of the WT1 gene and other genes, such as PAX6.
  - **W**ilms Tumor
  - **A**niridia
  - **G**enitourinary anomalies
    - Pseudohermaphroditism, undescended testes in males (due to gonadal dysgenesis)
    - Early-onset nephrotic syndrome
  - **R**ange of intellectual disability
- **Denys-Drash syndrome**: point mutation in WT1 gene, which encodes a zinc finger transcription factor
  - Denys-Drash syndrome is a mild form of WAGR without aniridia or intellectual disability
  - Nephroblastoma
  - Pseudohermaphroditism, undescended testes in males (due to gonadal dysgenesis)
  - Early-onset nephrotic syndrome caused by diffuse mesangial sclerosis
- **Beckwith-Wiedemann syndrome**: mutations of the WT2 gene

Beckwith-Wiedemann syndrome

Beckwith-Wiedemann syndrome

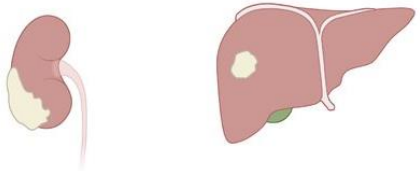
Definition
Congenital disorder of growth with a predisposition to tumor development

Etiology
WT2 gene mutation on chromosome 11 → defect in genetic imprinting

Diagnostics
Clinical diagnosis and/or genetic testing

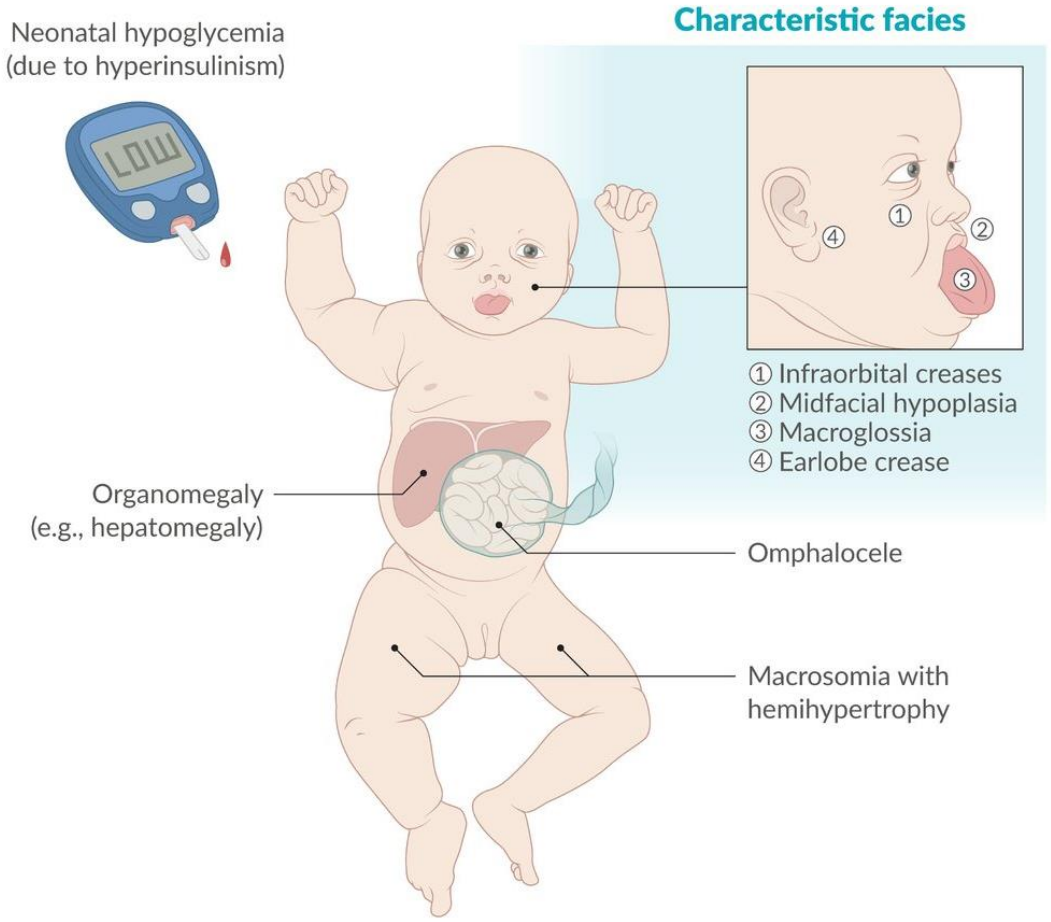
Treatment
- Treatment of manifestations (e.g., omphalocele repair)
- Regular screening for tumors and urinary tract abnormalities (e.g., abdominal ultrasound, serum AFP levels)

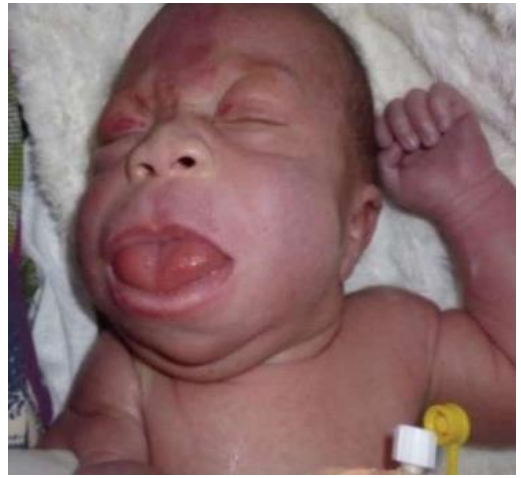
Most common tumors



Nephroblastoma

Hepatoblastoma





Macroglossia, hemihyperplasia, omphalocele, neonatal hypoglycemia, macrosomia, embryonal tumors (e.g., Wilms tumor, hepatoblastoma, neuroblastoma, and rhabdomyosarcoma), visceromegaly

Nephroblastoma – Clinical features

❖ **Abdominal symptoms**
- Palpable abdominal mass (often found incidentally)
  - Non-tender
  - Unilateral and large but not crossing midline
  - Smooth and firm
- Abdominal pain (~ 40% of cases)

❖ **Other signs and symptoms**
- Hematuria (~ 25% of cases)
- Hypertension (~ 25% of cases)
- Left sided varicocele: tumor has extended into left renal vein obstructing left testicular vein
- In cases of subcapsular hemorrhage → anemia and possibly fever
- Symptoms caused by metastatic spread (e.g., pulmonary symptoms)

- **Note**: Careless palpation of a nephroblastoma can result in rupture of the renal capsule and tumor spillage!

National Wilms Tumor Study (NWTS) system [8]

Stage	Tumor location	Tumor spread	Surgery
I	• Unilateral	• Limited to the kidney • No renal sinus vessel involvement	• Complete resection with no tumor beyond the excision margins

II	• Unilateral	• Extends beyond kidney
		  ◦ Renal capsule
		  ◦ Soft tissue of the renal sinus
		  ◦ Blood vessels beyond the renal parenchyma	

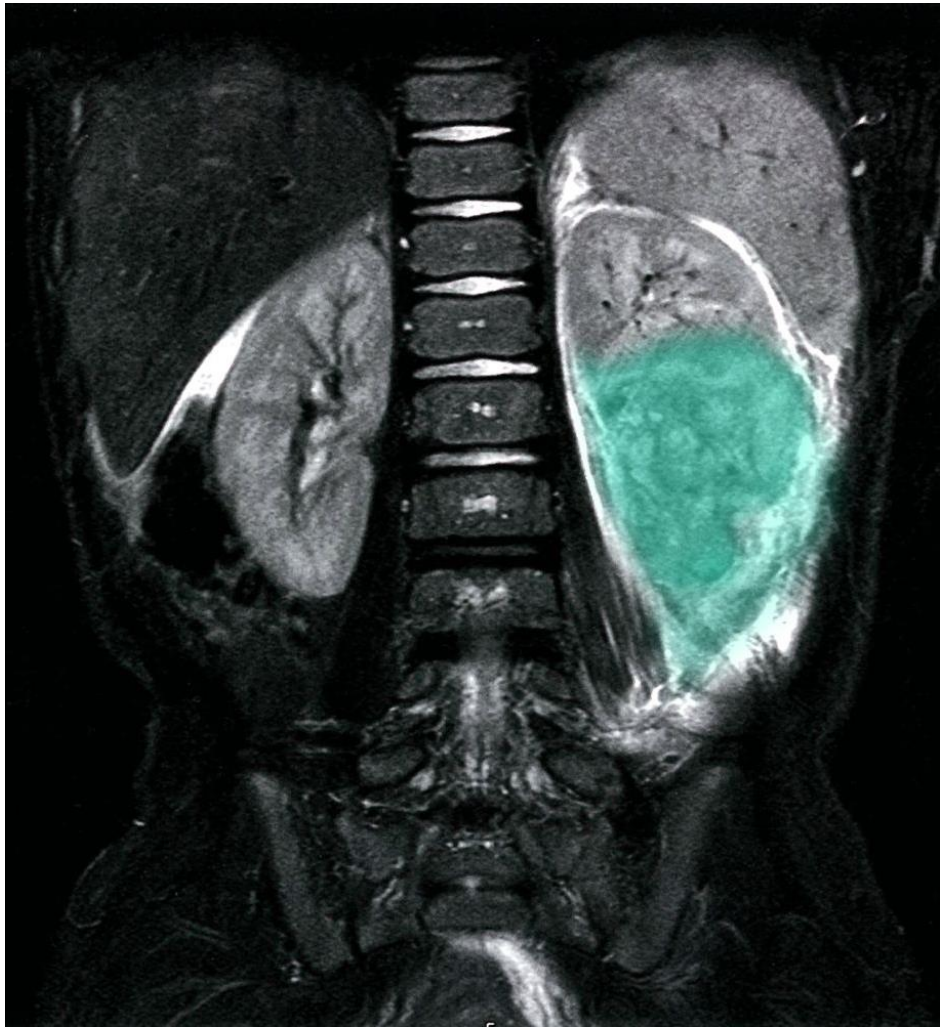
III	• Unilateral	• Confined to the abdomen • Dissemination in abdomen (e.g., regional lymph nodes, peritoneal involvement)	• Incomplete resection with residual tumor remaining postoperatively (e.g., not resectable due to infiltration into vital structures)
IV	• Unilateral	• Hematogenous metastasis (e.g., lung, liver, bone, brain) • Distant lymph nodes involvement (outside of abdomen)	
V	• Bilateral	• Both kidneys are affected	

Nephroblastoma – Diagnostics

- **Urinalysis**: hematuria may be present
- **Imaging**
  - Ultrasound (best initial test)
    - Hyper vascular tumor
    - Mostly uniform echogenicity with hypoechoic areas of necrosis
  - Abdominal CT/MRI (Next step)
    - To assess extent of involvement
    - Surgical planning
  - CT thorax/CXR (Pre-op assessment)
    - To detect metastases
    - Staging
- Biopsy is usually reserved for assessing nodules that are suspected metastases, as tumor capsule rupture and spillage results in more advanced staging and intensive treatment.

Nephroblastoma – Diagnostics

- MRI abdomen (T2-weighted with fat saturation; coronal plane) of a 3-year-old child
- A large heterogeneous but relatively well-circumscribed solid mass is centered within the lower pole of the left kidney. Normal renal tissue is seen cranial to the mass. The hypointense areas of the mass suggest soft tissue, whereas the areas of hyperintensity suggest hemorrhage or necrosis.
- Biopsy confirmed the diagnosis of nephroblastoma (Wilms tumor), the most common type of pediatric renal cancer, with a peak incidence at 3–4 years of age.



Nephroblastoma – Treatment

Treatment of nephroblastoma according to stage

Treatment		Stages I and II	Stages III and IV	Stage V (bilateral)
Surgery	Renal parenchymal-sparing resection	• No		• Yes
	Nephrectomy	• Yes		• No
Chemotherapy	Preoperative chemotherapy	• No		• Yes
	Doxorubicin	• No	• Yes	
	Dactinomycin and vincristine	• Yes		
Radiation		• No	• Possible	• Yes

- **Good prognosis**: survival rates are > 90%

Lecture – Management

- Preop
  - CT scan of chest → rule out lung metastasis
    - Most common metastatic sites are lung and liver
  - Laboratory studies
    - CBC, PT/INR, PTT, Type and Screen
- Operative
  - Radical vs partial nephrectomy
- Operation – Goals – Radical Nephrectomy
  - Safely resect entire tumor
  - Avoid upstaging tumor by complications
    - Capsular tears, gross tumor spillage, biopsy of tumor
  - Adequately stage tumor
    - Evaluate for metastasis, removal/biopsy of appropriate lymph nodes (most common operative error), evaluating vascular invasion
  - Adequately documenting pre-operative vs intraoperative tumor rupture
    - Changes post operative management with radiation

Lecture – Management

❖Operation – Contraindications

- Extension of tumor thrombus above the level of the hepatic veins
- Tumor involves surrounding structures, requiring removal of those structures to remove complete tumor
- Tumor involves bilateral kidneys
- Tumor involves a solitary kidney
- Pulmonary metastasis leading to respiratory compromise
- **All indications for neoadjuvant chemotherapy**

❖Operation – Complications

- Tumor Spill (9.7%); Break in tumor capsule
  - Includes pre-operative or intraoperative needle or core biopsy (COG protocol)
  - Transection of ureter or renal vein where tumor exists
- Bleeding (2%)
- Vascular or bowel injuries

Lecture – Management

❖Peri-operative management

- Monitor for ileus – particularly in patients with extensive retroperitoneal dissection
- Monitor for complications:
  - Wound infection
  - Bowel obstruction
  - Intussusception
- Prepare for radiation and chemotherapy

❖Final Discussion/Review

- Wilms' tumor is the most common pediatric renal mass and 2nd most common pediatric abdominal mass
- Most common presentation: asymptomatic abdominal mass
- Treatment for unilateral tumors: Radical nephrectomy followed by chemotherapy +/- radiation
- Histology and stage of tumor main prognostic indicators for Wilms tumor

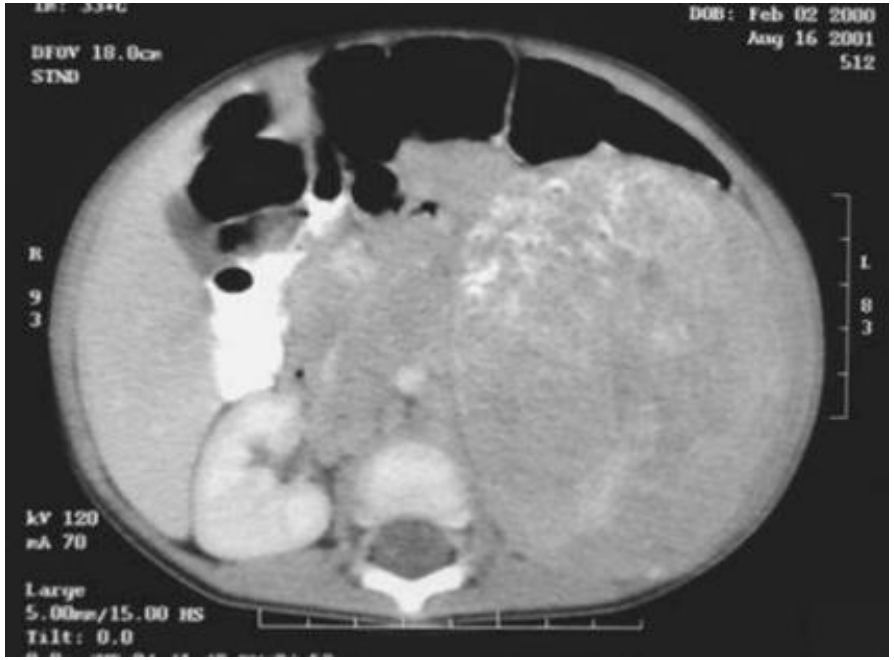
Abdominal mass DDx

Abdominal mass	Neuroblastoma	Nephroblastoma
Primary origin	Extrarenal (Adrenal gland or paravertebral sympathetic ganglia)	Intrarenal
Pattern of spread	Encasement of vessels and aortic elevation, Neural extension	Direct expansion with displacement of adjacent structures
Constitutional symptoms	More constitutional symptoms	Less constitutional symptoms
Appearance	Very sick appearance (Irritable child)	More healthy appearance
Physical examination	• Non-mobile mass • More likely to cross the midline • Painful	• Displacing mass, confined to the flank and unlikely to cross the midline • Painless
Hypertension	Less likely; is due to catecholamine release	More likely; is due to angiotensin
Other	• Externally displaces kidney • Often calcified • Racoon eyes	• Intrinsically displaces urinary collection system • Macroglossia • Hematuria

Abdominal mass DDx

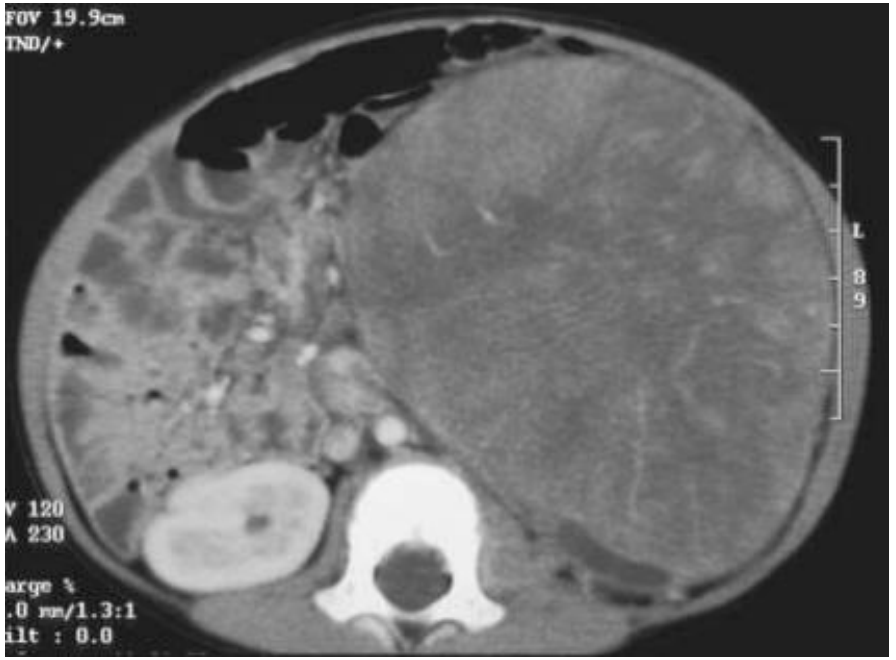
Neuroblastoma

- Calcifications more likely seen in neuroblastoma



Nephroblastoma

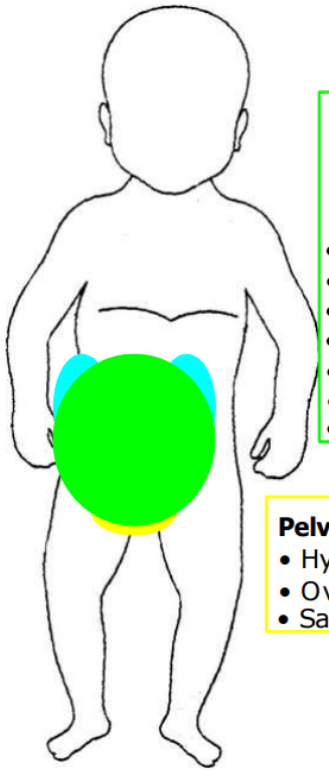
- "Claw sign: Sharp angles on either sides of the mass": Wilms mass arising from kidney



Abdominal mass DDx

INFANTS

Flank - 65%
- **Renal - 55%**
- Hydronephrosis
- Polycystic kidney
- Mesoblastic nephroma
- Renal ectopic
- Renal vein thrombosis
- Nephroblastomatosis
- Wilms tumor
- **Nonrenal - 10%**
- Adrenal hemorrhage (Waterhouse Friderichsen syndrome)
- Neuroblastoma



Intraperitoneal - 20%
- **GI Masses - 15%**
- Duplication
- Meconium ileus
- Mesenteric-omental cyst
- **Hepatobiliary - 5%**
- Hemangioendotheloma
- Hepatoblastoma
- Hepatic cyst
- Choledochal cyst
- Hydrops of gallbladder

Pelvic - 15%
- Hydrometrocolpos
- Ovarian cyst
- Sacrococcygeal teratoma

CHILDREN AND ADOLESCENTS

Flank - 78%
- **Renal – 55%**
- Wilms tumor
- Hydronephrosis
- Cystic disease
- **Nonrenal – 23%**
- Neuroblastoma
- Teratoma
- Other neoplasms

Intraperitoneal – 18%
- **GI Masses - 12%**
- Appendiceal abscess
- Other neoplasms
- **Hepatobiliary – 6%**
- Hepatoblastoma
- Hepatocellular ca
- Choledochal cyst

Pelvic – 4%
- Ovarian cyst
- Hydrometrocolpos

Case discussion

Hx: A child is seen by the PCP. The mother has noticed the child's abdomen was different upon bathing

Q1: What other points of the history do you want to know?

Q2: What specifically would you look for? (Physical Exam)

Q3: What labs are needed?

Q4: What imaging is needed, findings?

Q5: What is your diagnosis?

Q6: What is your plan?

Case discussion

- **What other points of the history do you want to know?**
  - **Age**: a crucial factor that may adjust the differential diagnosis
  - **Mass**: duration, associated pain, changes in eating and elimination patterns, history of trauma
  - **Birth Hx**: prematurity, difficult birth, prenatal care
  - **Medical Hx**: associated medical illnesses. Previous concerns for hematuria or hypertension
  - **Family Hx**: syndromes (Beckwith-Wiedemann, WAGR, Gardner)
  - **Review of system**: night sweats, malaise, bleeding or bruising, skin changes

Case discussion

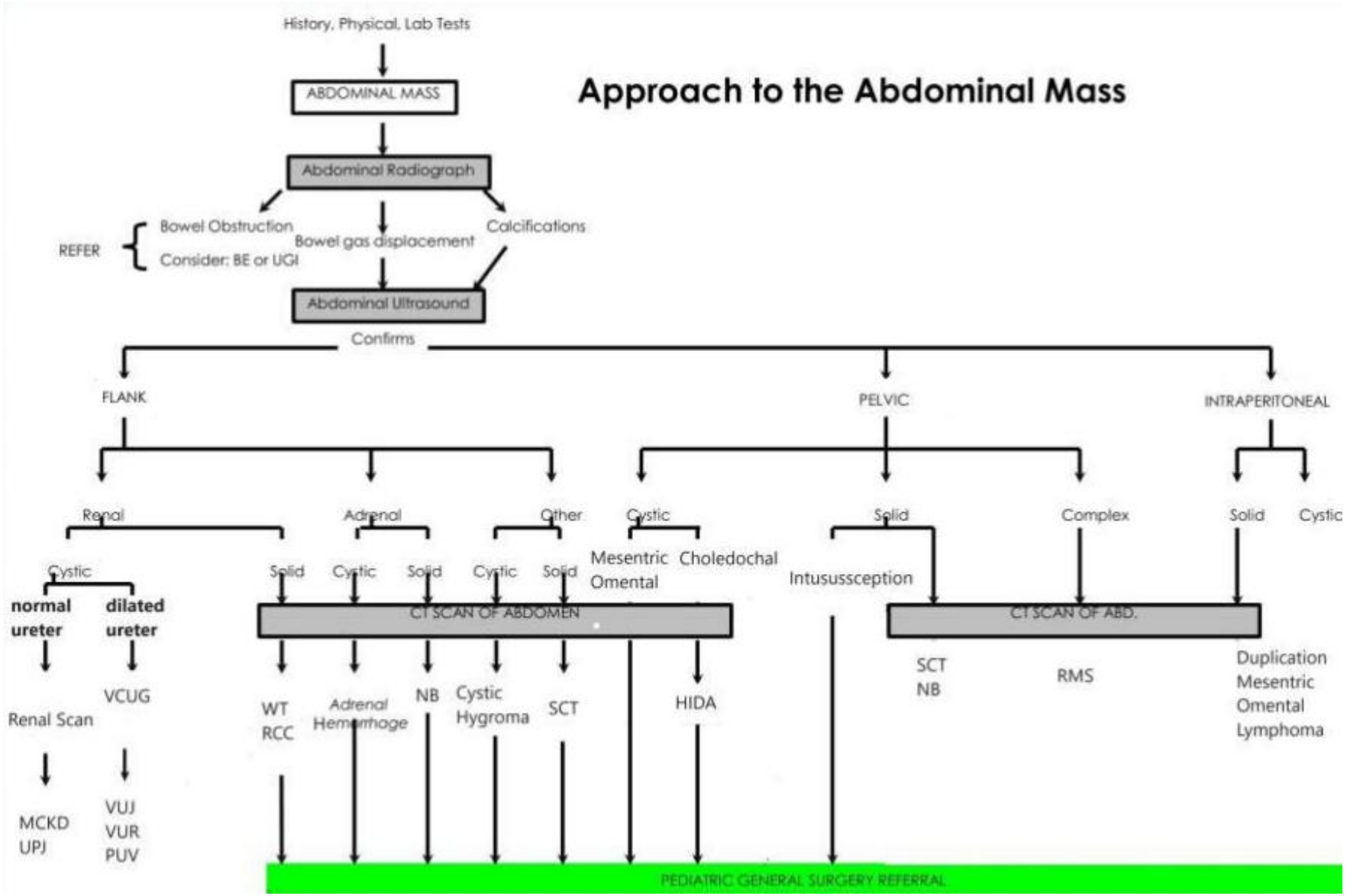
❖**What specifically would you look for?**

- **Vital Signs**: some tumors can cause elevated HR, BP; some masses may push up on diaphragm and limit breathing
- **NB**: This is a neoplasm of neural crest origin, arising in the adrenal medulla and along the sympathetic ganglion chain from the neck to the pelvis
- **Appearance**: healthy or ill appearance, Look for overgrowth (Beckwith-Wiedemann syndrome)
- **Head & Neck**: aniridia, raccoon eyes, proptosis, Horner's syndrome
- **Chest**: Rapid and shallow breathing
- **Cardiac**: congestive HF (SCT= Vascular Steal Syndrome)
- Lymphadenopathy
- **Abdomen**
  - Omphalocele, hepatosplenomegaly
  - Mass: location, configuration, size, consistency, mobility, tenderness
- **GU**: ambiguous genitalia, hypospadias, cryptorchidism

Case discussion

- **What labs are needed?**
  - CBC and differential
  - Electrolytes, BUN, Cr
  - Liver function tests
  - Amylase, lipase
  - Urine: Urinalysis, Vanillylmandelic acid (VMA), Homovanillic acid (HVA)
  - Tumor markers: alpha-fetoprotein (AFP), β-HCG
- **What imaging is needed, findings?**
  - **Ultrasound**: good first test
  - **CT scan**: good to help plan surgery and for staging
    - Wilms Tumor: “Claw sign
    - Neuroblastoma: Calcifications
  - **MRI**: limited application
  - **Nuclear scans**: selective use based on diagnosis

Case discussion – What is your Dx and plan?



Operation

❖Goals of surgery

- Staging (Invasion and LNs)
- Obtain tissue for diagnosis; only if not resectable
- Complete resection (avoid disruption of the margins to avoid tumor rupture)
- Assistance with radiotherapy, or assistance with chemotherapy (Debulking)

❖Complications

- Peri-operative
  - Ileus is common after any abdominal surgery
  - Post-op intussusception is well reported, particularly after retroperitoneal dissection
- Long Term
  - Is dependent on the tumor type and whether rupture has occurred
  - Potential for adhesive bowel obstruction

Lecture MCQs 1 (Answers at the end)

- Where do most abdominal masses arise?
  a. Flank
  b. Intraperitoneal
  c. Pelvic
  d. None of the above
- Which is the most useful first test to order to help determine the type of abdominal mass?
  a. X-ray
  b. Ultrasound
  c. CT scan
  d. MRI

Lecture MCQs 2 (Answers at the end)

- With regards to abdominal masses, the goal of surgery may include?
  a. Staging
  b. Obtain tissue for diagnosis
  c. Resection of mass
  d. Help adjuvant therapy
  e. All the above
- With regards to abdominal masses, avoiding tumor rupture is critical to?
  a. Avoiding pathologic misinterpretation
  b. Intraoperative blood loss
  c. Not upstaging the patient
  d. Decreasing likelihood of postoperative intussusception
  e. Spuriously increasing tumor markersafter surgery

Lecture MCQs 3 (Answers at the end)

- The most common presenting abnormality in patient's with a Wilms' tumor is:
  a. Hypertension
  b. Hematuria
  c. Asymptomatic abdominal mass
  d. Abdominal pain
- Which of the following does NOT increase recurrence of tumor?
  a. Pre-operative core needle biopsy
  b. Resection lymph nodes at renal hilum, along iliac vessel and para-aortic regions
  c. Transecting ureter containing tumor
  d. Sustaining minor renal capsular tear during dissection from surrounding structures

Lecture MCQs Answers

Q1: A. Flank

Q2: B. Ultrasound

Q3: E. All the above

Q4: C. Not upstaging the patient

Q5: C. Asymptomatic abdominal mass

Q6: B. Resection lymph nodes at renal hilum, along iliac vessel and para-aortic regions

Ventral Wall Defects

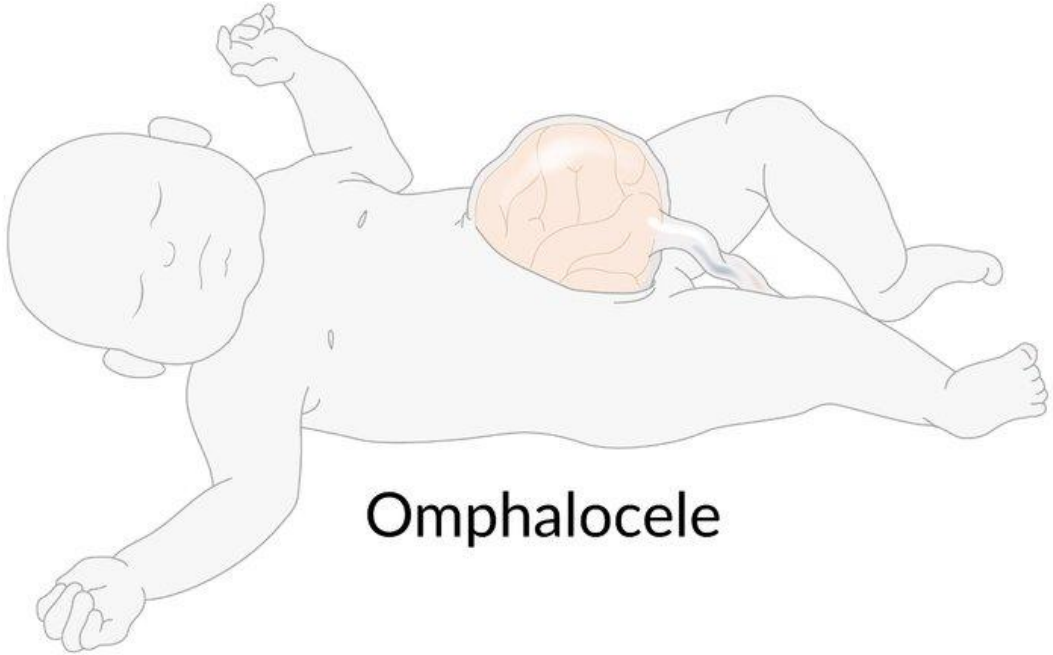
Ventral Wall Defects

❖Embryology

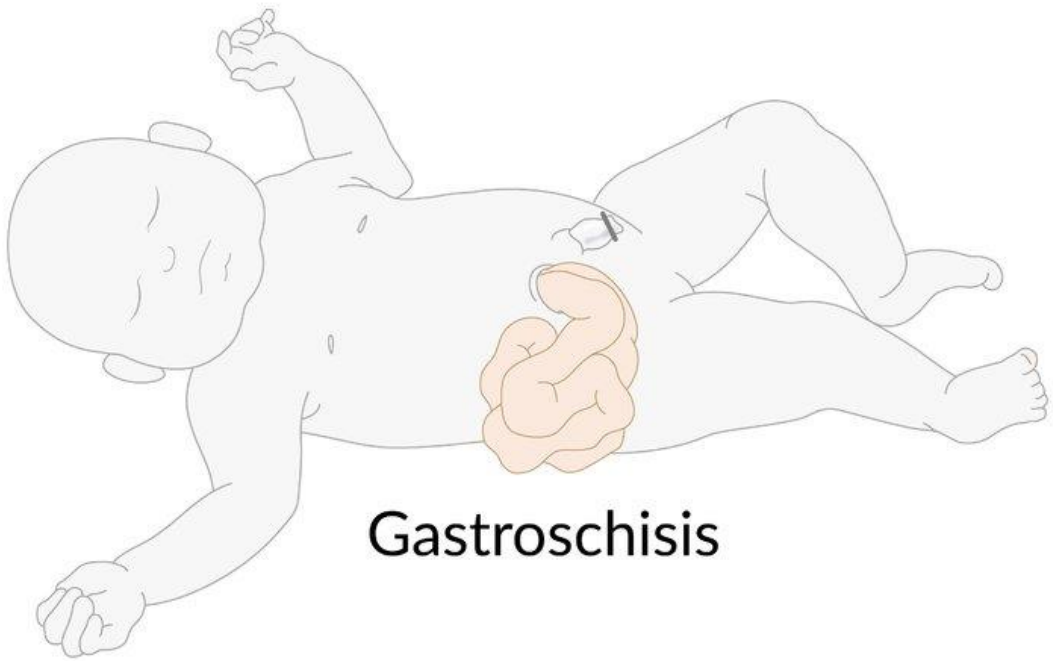
- The abdominal wall forms during the fourth week of gestation.
- During the sixth week, rapid intestinal and liver growth leads to herniation of the midgut into the umbilical cord. Elongation and rotation of the midgut occurs over the ensuing 4 weeks.
- By week 10, the midgut returns to the abdominal cavity, where the first, second, and third portions of the duodenum and the ascending and descending colon assume their fixed, retroperitoneal positions.
- An abdominal wall defect involves an interruption of these embryologic processes.

❖Possible causative factors

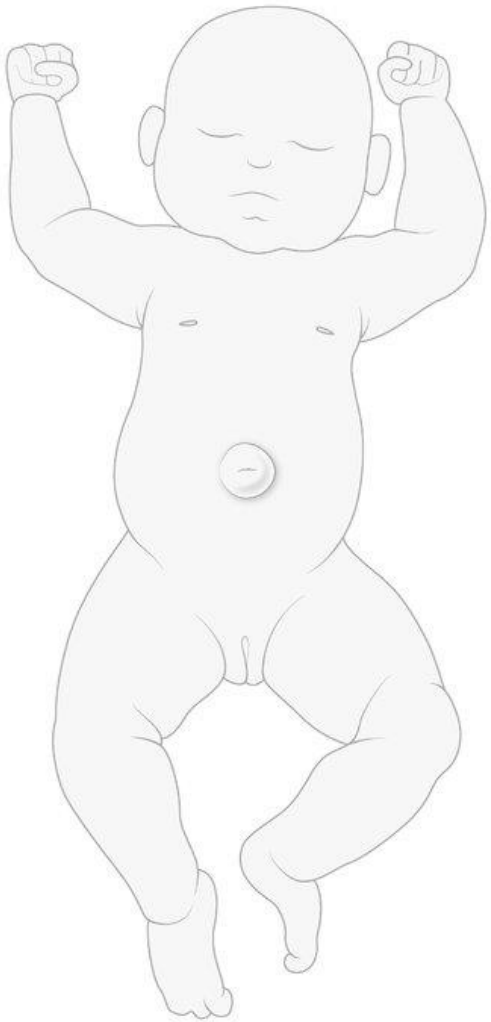
- Tobacco
- Environmental exposures (nitrosamines)
- Cyclooxygenase inhibitor use (aspirin and ibuprofen)
- Decongestants (pseudoephedrine and phenylpropanolamine)
- Low maternal age: younger than 21 years
- Prematurity



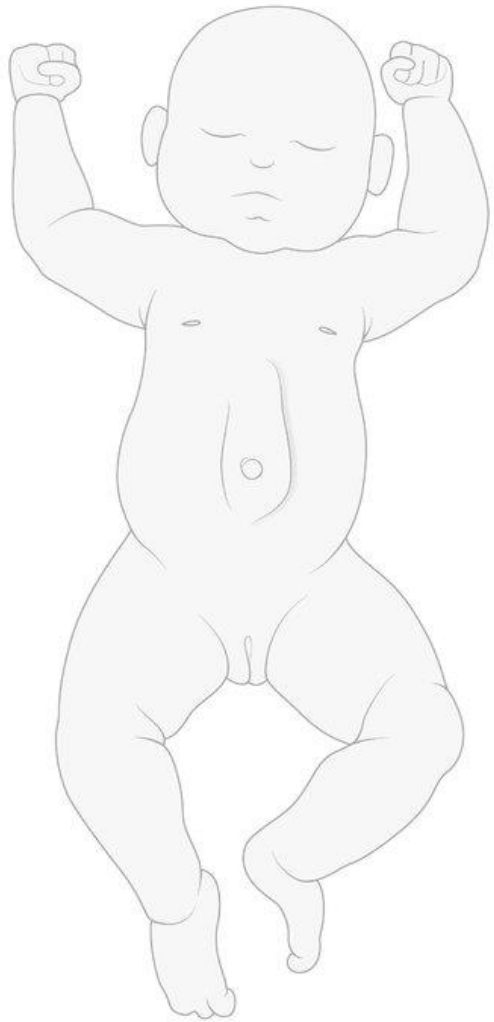
Omphalocele



Gastroschisis



Umbilical hernia



Diastasis recti

	Omphalocele	Gastroschisis	Umbilical hernia
Description	A ventral wall defect that results in congenital herniation of abdominal viscera through the abdominal wall at the umbilicus	A ventral wall defect that results in paraumbilical herniation of the intestine through the abdominal wall, without formation of a hernia sac	A ventral wall defect with a protruding sac (possibly containing intestines) at the umbilicus
Pathophysiology	Persistent herniation of the midgut derivatives due to impaired closure of the lateral umbilical folds	Failed formation of a sufficiently large peritoneal cavity → growing bowel → rupture of the anterior abdominal wall at its weakest point → herniation of bowel sections	Failed spontaneous closure of the umbilical ring following physiological herniation of the midgut → patent umbilical orifice
Anatomy	• Viscera (may also contain liver and gall bladder) herniate into the umbilical cord. • The hernia sac is covered by amniotic membrane and peritoneum.	• Protrusion of intestinal content only, usually on the right side of the umbilicus • The exposed viscera may be covered with a think inflammatory peel. Bowel loops may be densely adherent making it impossible to distinguish one loop of bowel from another.	• The mass protrudes through the umbilicus. • The hernia is covered by skin.
Defect location	Umbilicus	Right of umbilicus	Umbilicus
Associated conditions	• Additional malformations in ~ 50% • Cardiac defects are observed in 30-50 % • Karyotype abnormalities occur in 30 % of cases with trisomy 13, 18 and 21 being most common • Beckwith-Wiedemann syndrome • Pentalogy of Cantrell	• Additional anomalies rarely present • Intestinal atresia is the most common associated anomaly • Malrotation occurs in nearly every patient with gastroschisis, but midgut volvulus is not commonly seen	Incidence is more common in children with other congenital anomalies (e.g., trisomy 21, trisomy 18, congenital hypothyroidism).

	Omphalocele	Gastroschisis	Umbilical hernia
Diagnostics	Clinical diagnosis		
	Can be detected prenatally • Ultrasonography: polyhydramnios in utero • ↑ MSAFP		Usually, no further diagnostics necessary
Delivery	• Cesarean delivery • Vaginal delivery for small omphalocele without liver involvement	Usually, vaginal delivery	Vaginal delivery
Management	• Wrapping of the hernia sac/free intestines with sterile saline dressings covered with plastic wrap • Nasogastric suction • IV fluids		Conservative: closes spontaneously by the age of 5 years
Prognosis	Depend on associated anomalies	Depend on condition of bowel	Good
Surgery	Within the first 24 hours of life	Emergency surgery	Rarely necessary
Closure approaches	• Primary closure if small • Giant omphaloceles, however, are managed by allowing epithelialization of the sac with topical application of silver sulfadiazine, serial reductions and elective repair at 6-24 months (as a ventral hernia) • Ruptured omphaloceles have very poor prognosis and require silo placement with delayed primary closure.	• Placement of Silastic® silo with staged reduction of the viscera over several days followed by delayed facial closure with suture • Complete reduction of the viscera shortly after birth and primary facial closure with suture • Complete reduction of the viscera shortly after birth and covering of the defect with an umbilical cord flap (Sutureless)	-

Gastroschisis Notes

❖ Classification

- Simple gastroschisis (i.e. viable bowel, no atresias)
- Complicated gastroschisis (i.e. Perforated bowel, atresia)

❖ Complications

- Abdominal compartment syndrome (ACS) can occur within the first few days following reduction and closure of a gastroschisis defect. ACS may manifest as increasing pulmonary pressures in ventilated patients, low urine output, differential cyanosis (bluish appearing legs caused by impaired venous return) and hypotension.
- 10-15% of patients with gastroschisis develop necrotizing enterocolitis (NEC)
- Intestinal atresia

Other abdominal wall hernias

- **Umbilical Hernia** (notes not mentioned in the table)
  - The hernia sac is peritoneum
  - The extent of skin protrusion is not always indicative of the size of the fascia defect
  - Umbilical hernias are present in 15–25% of newborns
  - Premature and low birth weight infants have a higher incidence
- **Epigastric Hernia**
  - Hernias of the abdominal wall through the midline linea alba, with a location between the umbilicus and xiphoid process
  - Incidence up to 5%
  - These hernias present as either a small painless mass, which becomes painful with activity, or a small painful incarcerated mass
  - Typical contents are preperitoneal fat
  - Epigastric hernias do not resolve and should be repaired

Other abdominal wall hernias cont.

- **Spigelian Hernia: (Spigelian Triangle)**
  - Quite rare in children and can be difficult to detect and diagnose.
  - The actual defect occurs at the intersection of the linea semicircularis, linea semilunaris, and the lateral border of the rectus abdominis muscle
  - More frequently in girls
  - A tension-free closure is important to prevent recurrence
- **Lumbar Hernia**
  - Bulge in the area bordered by the 12th rib, sacrospinalis muscle, and internal oblique muscle
  - The bulge is usually due to herniated preperitoneal fat
  - Physical findings include a soft mass that is easily reducible
  - Repair is advisable because the defect never resolves spontaneously and incarceration is possible

Anorectal malformation

Anorectal malformation

Definition	A normal anal opening is nonexistent, and the colon empties anteriorly either onto the perineum or towards the vagina in a female or into the urinary system in a male. There are also instances of atresia where the rectum is blind ending with no connection or external opening
Epidemiology	○ ARM occur in one in every 4000 to 5000 births ○ They are slightly more common in males (1.2:1) ○ The risk of having a second child with an ARM is approximately 1%
Associations	○ Mesodermal defects (e.g., **VACTERL association**): The distance away from the normal anal opening correlates with the degree of associated abnormalities ○ Known association with trisomy 21
Pathophysiology	Failure of complete hindgut separation into ventral urogenital portion and the dorsal anorectal portion

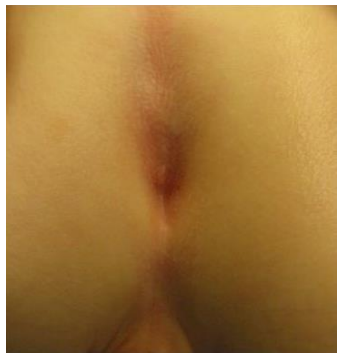
Clinical features

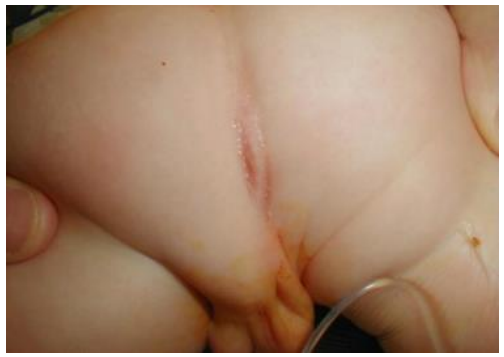
- Absence of anal opening
- Thin anal membrane in place of anal opening through which meconium is visible
- Obstipation, ileus
- Fistulas that complicate defecation (e.g., rectovestibular, rectourethral, rectovaginal)
- Small or missing anal dimple (The anal dimple is the area of the perineum that has the largest density of sphincter fibers)
- In some cases, bucket-handle malformation (A prominent skin tag located at the anal dimple under which an instrument can be passed)
- Flat bottom (absent or poorly developed midline groove between buttocks)

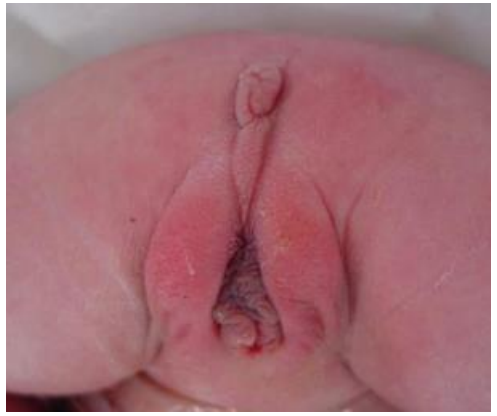
Classification

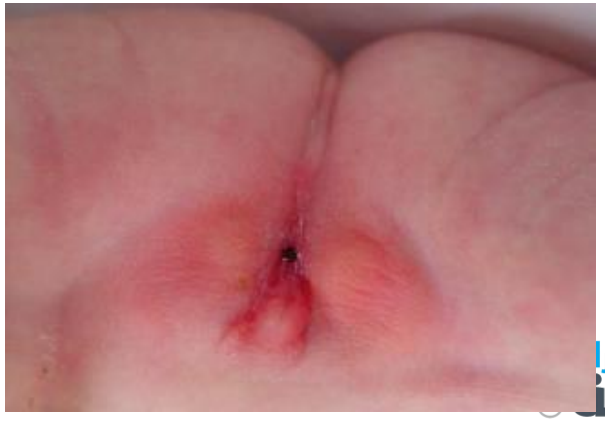
Male ARM Classification by Location	
Lower	
Anal stenosis	1%
H-type fistula	4%
ARMs with no fistula	5%
Perineal (cutaneous) fistula	10%
Recto-bulbar urethra fistula	37%
Higher	
Recto-prostatic urethra fistula	35%
Recto-vesical fistula	8%

Female ARM Classification by Location	
Lower	
Recto-vaginal fistula	<1%
Anal stenosis	1%
ARMs with no fistula	5%
Perineal (cutaneous) fistula	18%
Recto-vestibular fistula	70%
Complex	
Cloaca (Single Opening)	5%





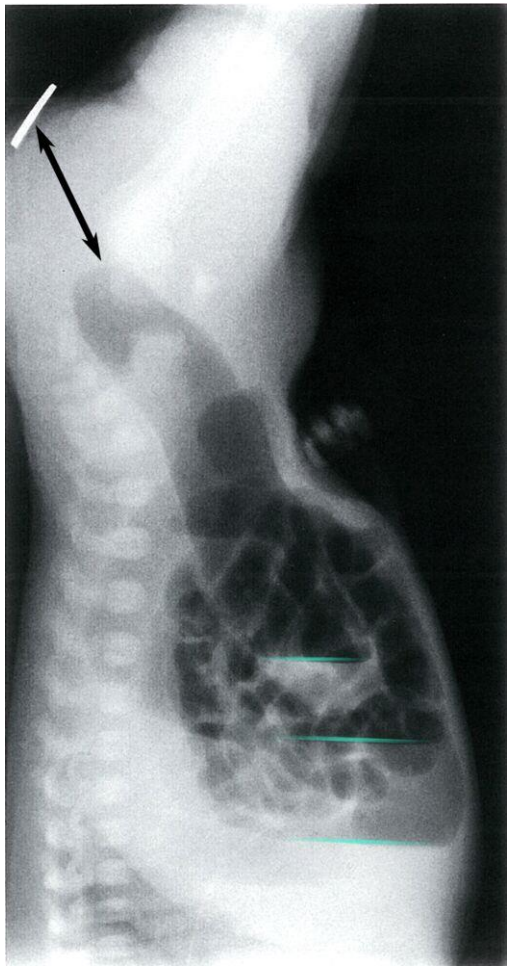




Diagnosis and investigations

- ❖It’s a clinical diagnosis
- ❖After the diagnosis number of tests are done before surgery

X-ray	Lateral pelvic radiography or invertogram to determine position of the rectal pouch
VACTERL Work up	• Echocardiogram • Renal U/S (GU anomalies are the most common in ARM) • Spine Ultrasound and X-Ray (Has prognostic factor) • Radius X-Ray if abnormality is noted on physical exam
Fistula screening	• Exam the genital area and the urethra (e.g., abdominal U/S) • Urinalysis: meconium detectable in case of a rectourinary fistula • Augmented-pressure distal colostography to assess for fistulas
Planning Staged Procedure	• VCUG • Antegrade colostogram via distal limb • Cystoscopy, Vaginoscopy



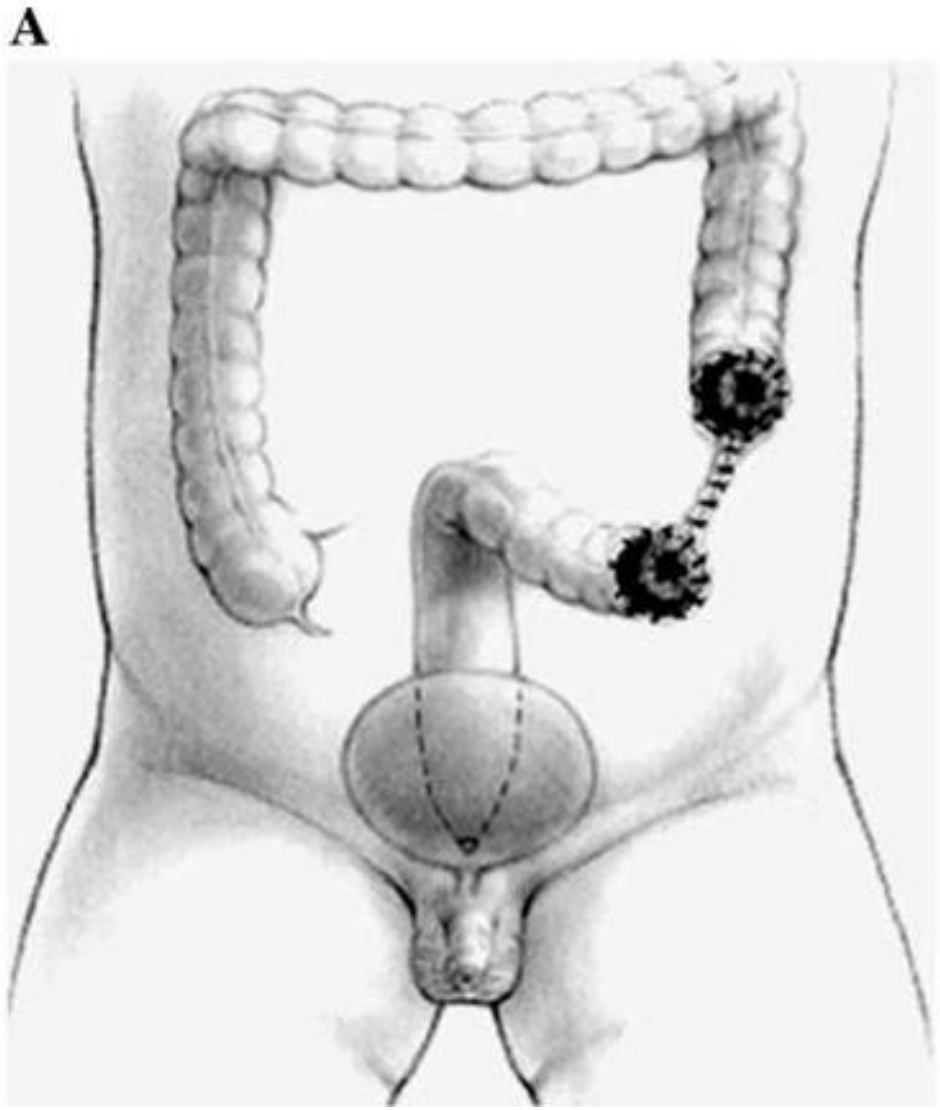
Management

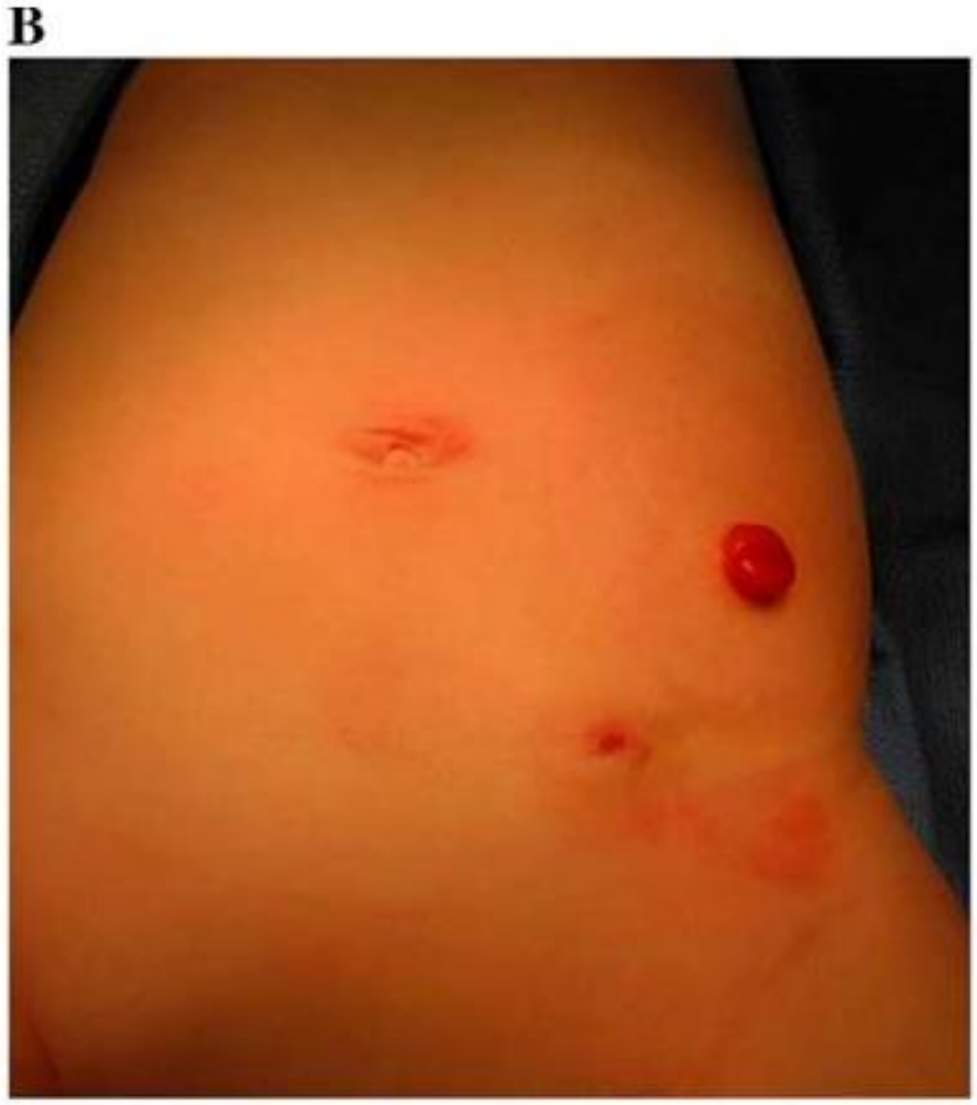
- **Include**: NPO, IV hydration and surgery
- **Surgeries**:

Anoplasty	**Indications**: Perineal Fistula
PSARP (Posterior Sagittal Anorectoplasty); with or without colostomy	**Indications**: Rectal gas below coccyx, No associated defects, Vestibula Fistula **Procedure**: • Combined perineal and abdominal approach required for higher types • Colonic diversion reversed once perineum healed (6-8 weeks after PSARP)
Diverting colostomy	**Indications**: Rectal gas above coccyx, Associated defects, Cloaca

- **Surgery complications**
  - **Peri-operative**: Infection, Bleeding, Wound Dehiscence, UTI
  - **Long Term**: Anal stenosis, Fecal incontinence, Chronic constipation

Diverting colostomy (Divided Colostomy)





Inguinoscrotal Swelling

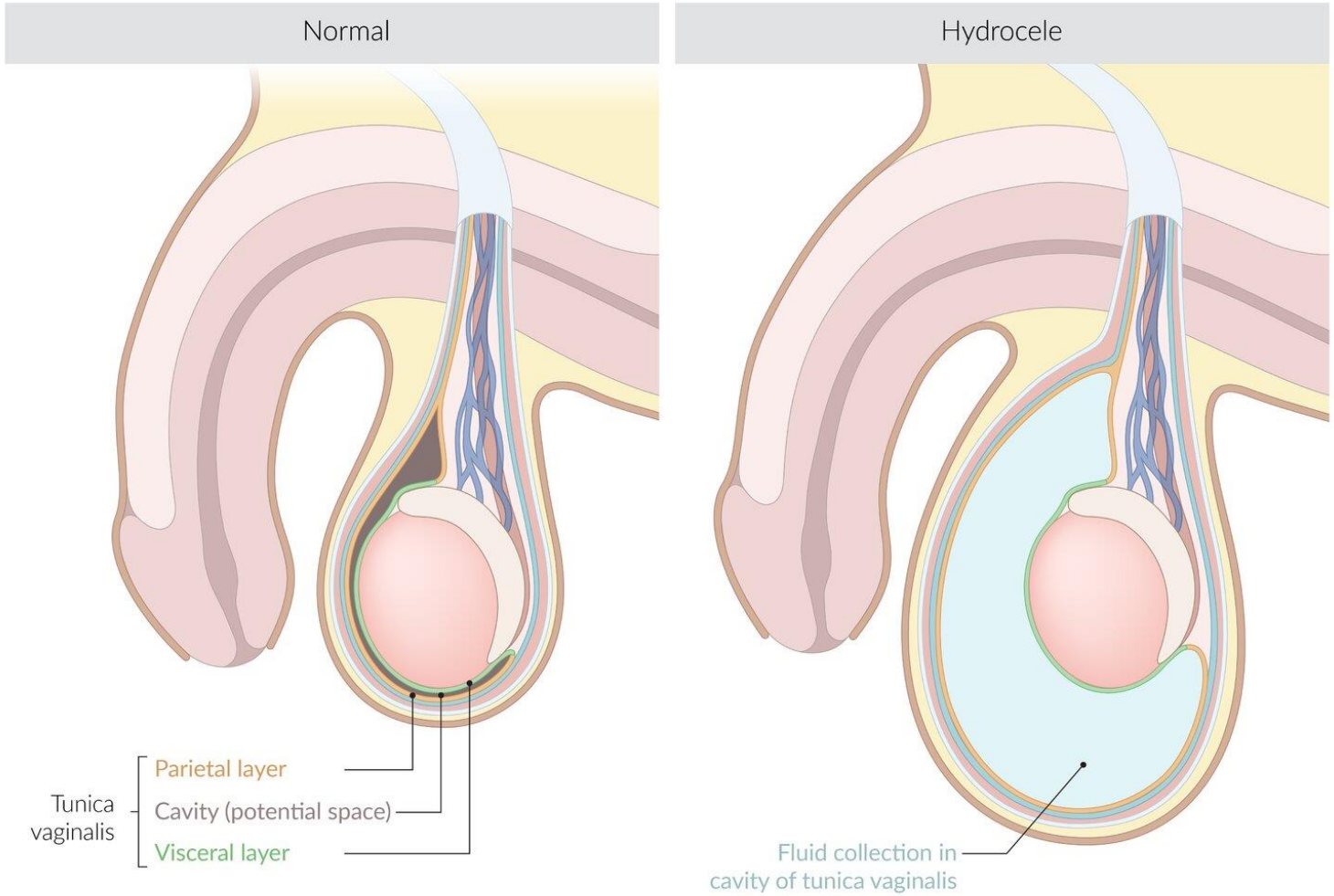
Inguinoscrotal Swelling in Children

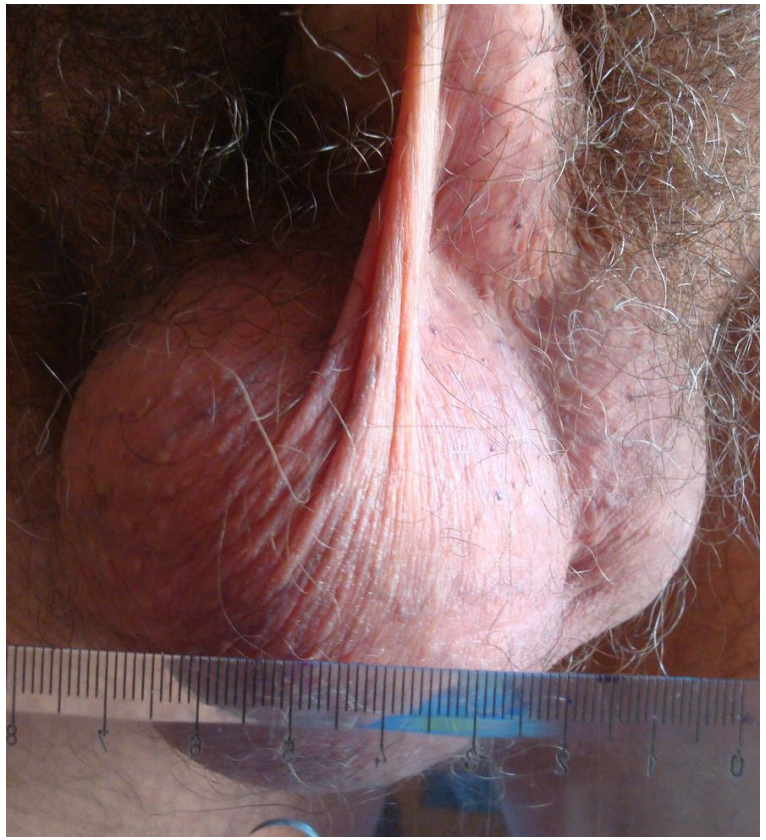
- Processus vaginalis is a peritoneal diverticulum that extends through the internal ring at approximately 3 months gestation. As the testis descends between the seventh and ninth months of gestation, a portion of the processus vaginalis attaches to the testes and is dragged into the scrotum with the testes.
- The portion of the processus vaginalis surrounding the testis becomes the tunica vaginalis. The remainder of the processus vaginalis obliterates, thereby eliminating the communication between the peritoneal cavity and the scrotum.
- Failure of obliteration or incomplete obliteration of the processus vaginalis is the underlying pathophysiology of the development of hernias and hydroceles
- A **hydrocele** occurs with incomplete obliteration of the processus vaginalis so that fluid accumulates around the testicle or the cord structures. This fluid may or may not communicate with the peritoneal cavity.
- A **hernia** occurs as a result of distal obliteration of the processus vaginalis with proximal patency or complete failure of obliteration so that both fluid and bowel may be present in the sac.

Hydrocele

Definition	Painless accumulation of fluid in a sac around one or both testicles which derives from the tunica vaginalis, a tissue covering the testes
Etiology	○ Idiopathic (most common) ○ Congenital hydrocele ○ Acquired hydrocele: 2ry to pathology (e.g., trauma, tumor, torsion, infection)
Clinical features	○ Fluctuant, painless swelling of affected scrotum ○ Positive transillumination
Treatment	Congenital hydrocele usually resolves spontaneously within 6 months of birth. **Indications for surgery** • Non communicating • Communicating with failure to resolve by 1–2 years of age • Unable to rule out hernia (bowel) component • If infertility is a concern

Hydrocele


Normal
Hydrocele
Tunica vaginalis
Parietal layer
Cavity (potential space)
Visceral layer
Fluid collection in cavity of tunica vaginalis

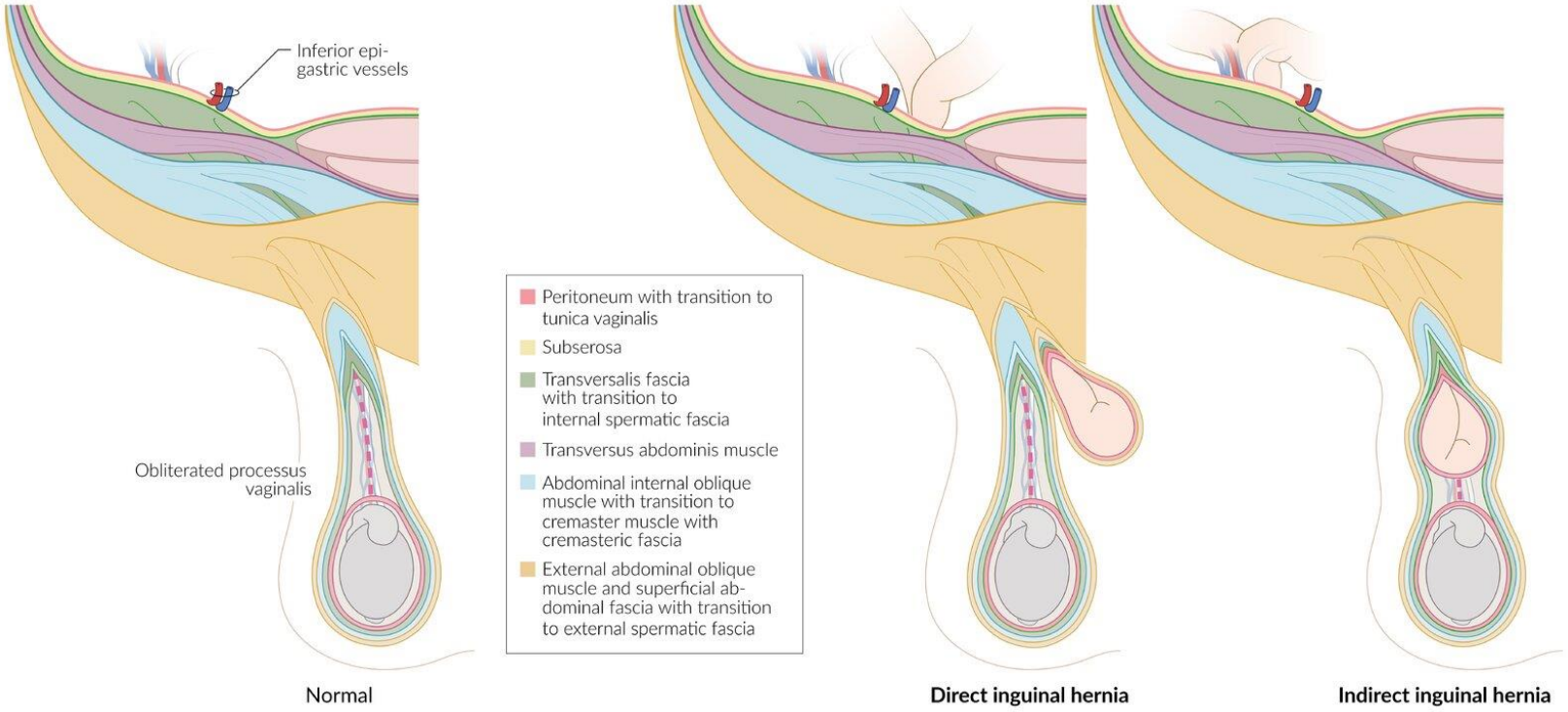


Inguinoscrotal hernia – Epidemiology

- Almost all inguinal hernias in infants and children are indirect.
- The incidence of groin hernias in infants and children is 1-5%.
- Boys outnumber girls 6-10:1
- Right-sided hernias are more common because the right testis descends later than the left testis; therefore, the right processus vaginalis obliterates at a later date than the left. (R 60%, L 30%, Bilateral 10%)
- Consequently, patients who present with a left-sided hernia have a higher incidence of an occult right sided hernia.
- The risk of incarceration exceeds 60% during the first 6 months of life.
- Most hernias are found either by parents or during a well-baby or preschool check.
- Most patients have a history of an intermittent bulge in the groin or scrotum, especially with crying or straining. Most are asymptomatic.

Pathophysiology of indirect inguinal hernia

- Most commonly results from incomplete obliteration of processus vaginalis; during fetal development (but can also be acquired).
- Lateral to the inferior epigastric blood vessels (outside Hesselbach triangle)
- Runs from the deep inguinal ring through the inguinal canal to the superficial (external) inguinal ring (in men, along with the spermatic cord)



Uncomplicated inguinal hernia – Clinical features

- **Typically manifests as** an ill-defined mass in the inguinal region
- **With the following features**
  - Increases in size when coughing or straining
  - Inguinal pain; increases with physical activity
- **Physical examination**
  - Ask the patient to perform the Valsalva maneuver and observe for an expansile cough impulse in the inguinal region.
  - Palpate the inguinal canal.
    - Invaginate the scrotal skin toward the superficial inguinal ring
    - Ask the patient to perform the Valsalva maneuver.

Complicated inguinal hernia – Clinical features

- **Definitions**
  - **Incarcerated hernia**: the hernia is irreducible.
  - **Obstructed hernia**: symptoms of mechanical bowel obstruction (sudden onset of pain)
  - **Strangulated hernia**: sudden, severe groin pain caused by constriction and ischemia (or necrosis) of hernial contents; blood may be seen per rectum
- **Symptoms and signs of an incarcerated inguinal hernia**
  - The patient may be irritable and complain of pain in the groin.
  - Signs of intestinal obstruction, including abdominal distention, vomiting, and obstipation, may follow.
  - Physical examination reveals a tender, sometimes erythematous, irreducible mass in the groin.
  - With strangulation of the bowel, blood may be seen per rectum.

Complicated inguinal hernia – Clinical features

- Most commonly the bowel become incarcerated in an inguinal hernia sac.
- In females, the ovary or fallopian tubes frequently become incarcerated.
- **Amyand's hernia**: presence of **Appendix** in an inguinal hernia sac.
- **Littre's hernia**: protrusion of a **Meckel's diverticulum** into the hernial sac.
- Transillumination is not reliable sign

Inguinoscrotal hernia cont.

DDx	Hydrocele, Cryptorchidism, Testicular torsion, Torsion of the appendix testis, Inguinal lymphadenopathy
Treatment	High ligation of the sac (herniotomy)
Surgery Timing	○ In premature infants, groin hernias should be repaired just before discharge from the NICU ○ Other infants should be scheduled electively within about 1 month ○ Incarcerated hernias that can be reduced in the emergency department should be repaired within 24-48 hours. ○ Incarcerated hernias that cannot be reduced in the emergency department should be repaired emergently.
Complications of groin hernia repair	○ Bleeding, ○ Recurrence (0.5-1%), ○ Infection (< 1%), ○ Iatrogenic cryptorchidism, ○ Injury to cord structures (< 2%)

Cryptorchidism

- **Definition**: failure of one or both testicles to descend to their natural position in the scrotum
- **Epidemiology**
  - 80% found in inguinal canal. (The testicle is located between the external and internal inguinal ring, preventing adequate mobilization)
  - Unilateral (90%) or bilateral (10%); 70% of unilateral cases occur on the right side.
  - In full-term infants, the incidence is 2.7-5.9% at birth but decreases to 1.2-1.8% by 1 year of age.
  - The incidence is 10x in premature infants. (i.e., Prematurity is a risk factor)
- **Undescended testes at risk for**
  - Injury due to direct trauma
  - Epididymo-orchitis from repeated trauma
  - Sterility – 10% risk for unilateral and 38% for bilateral undescended testes
  - Malignancy

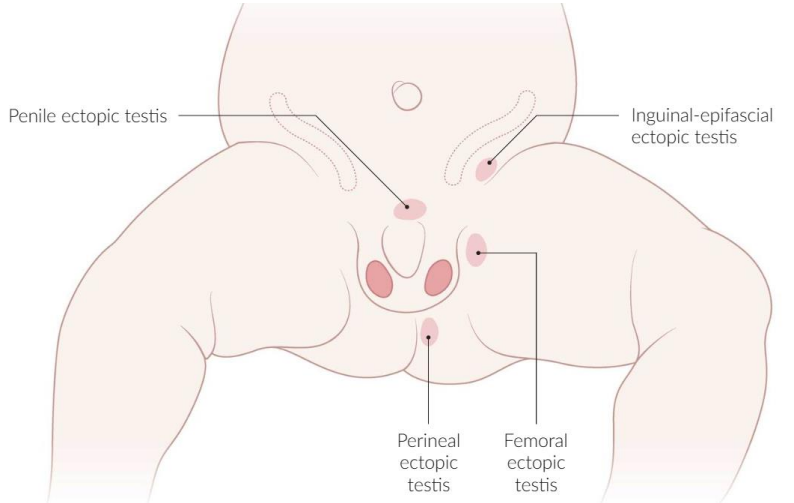
Other scrotal abnormalities

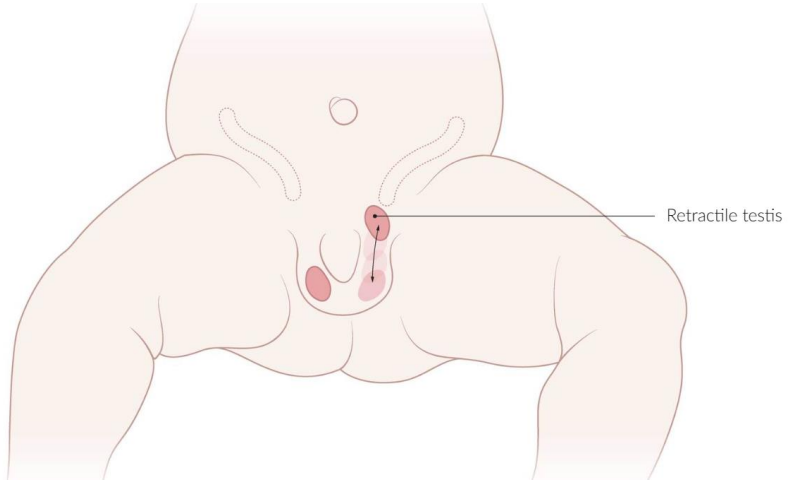
❖ Ectopic testes

- Testes found outside of the line of normal descent
- Can be found in femoral canal, perineum, contralateral scrotum, above the pubic bone

❖ Retractile testes

- Retraction of testicles in response to cold temperatures and protection from trauma
- Later in childhood
- Secondary to abnormal response to cremasteric muscle contraction
- Can be brought down into the scrotum during physical examination
- No treatment is necessary





Scrotal abnormalities

Diagnosis	○ Clinical ○ Ultrasound ○ MRI ○ Diagnostic laparoscopy
Management	○ First 6 months of age → monitor for spontaneous descent ○ Not descended → Orchiopexy ○ Hormonal therapy not recommended by the American Urological Association guidelines due to lack of response and long-term efficacy • **Testosterone** – no longer used due to causing precocious puberty • **hCG** – 6% success rate • **LHRH** – 19% success rate

Testicular Torsion

Definition	Twists of the spermatic cord, causing venous congestion, edema, and eventual arterial obstruction, which, if not treated, lead to gonadal necrosis.
Peak incidence	• Neonatal period (first 30 days of life) • During puberty (10–14 years)
Pathophysiology	○ **Intravaginal torsion**: hypotheses suggest this occurs because of a congenital abnormality in which the tunica vaginalis attaches to the superior pole of the testis (bell-clapper deformity) → increased mobility of testis within tunica vaginalis, with possible abnormal transverse lie of testis → torsion of the testis (along the spermatic cord). Most commonly 12-18-year-olds. ○ **Extravaginal torsion**: lack of fixation of the tunica vaginalis to the gubernaculum → concomitant torsion of the testis and tunica vaginalis (along the spermatic cord). Seen in neonatal torsion.

Testicular Torsion – Clinical features

- Abrupt onset of severe testicular pain and/or pain in the lower abdomen
- Typically, swollen and tender testis and/or lower abdominal tenderness
- Nausea and vomiting
- Abnormal position of the testis
  - Scrotal elevation (high-riding testis)
  - Abnormal transverse position
  - Possible undescended testes (predisposes to testicular torsion)
- Absent cremasteric reflex
- Negative Prehn sign
- In neonates
  - Possible absent testis
  - Firm, painless scrotal mass
  - Possible acute inflammation: swollen, erythematous (or blue discolored in venous engorgement), and tender hemiscrotum

Testicular Torsion – Diagnostics

- **Mainly clinical**
  - Testicular torsion is typically a clinical diagnosis. Do not delay definitive treatment for diagnostic workup if clinical suspicion is high.
- **Doppler sonography**; Characteristic findings
  - Twisting of spermatic cord (whirlpool sign)
  - Reduced or absent blood flow to/from the affected testis
  - Heterogeneous appearance of testicular parenchyma indicates testicular necrosis
- **Nuclear scintigraphy**
  - **Indications**: Inconclusive clinical findings and to evaluate for epididymitis
  - **Characteristic findings**
    - **Testicular torsion**: Areas that do not absorb radionuclide as a result of decreased blood flow to the affected testis (“Cold spots”)
    - **Epididymitis**: areas where there is increased radionuclide absorption as a result of increased blood flow in inflammation (“Hot spots”)

Differential diagnosis of scrotal pain

Disorder	History	Examination	Laboratory studies
Testicular torsion	• Sudden onset • Unilateral painful testis/lower abdomen • Nausea or vomiting	• Swollen, edematous, tender testis • Abnormal position of testis • Negative Prehn sign • Absent cremasteric reflex	• Normal inflammatory markers • Normal urinalysis
Epididymitis	• Gradual onset • Possible history of urethral discharge	• Very tender • Positive Prehn sign • Positive cremasteric reflex • Tender spermatic cord	• Increase in generalized inflammatory markers • Possible pyuria during urinalysis
Testicular tumor	• Slow progression (e.g., weeks to months) • Usually painless mass	• Easy palpation of solid mass • Possible manifestations of metastatic disease	• Possible increase in serum tumor markers (e.g., alpha-fetoprotein)
Torsion of testicular appendage	• Insidious onset of unilateral scrotal pain • Usually seen in boys 7–14 years of age	• Tender testis • May manifest with blue dot sign	• Typically, normal inflammatory markers • Typically, normal urinalysis

Testicular Torsion – Treatment

- **Manual testicular detorsion**
  - **Indication**: may be attempted prior to surgery for immediate pain relief or if surgery is not immediately available
- **Exploratory surgery**
  - **Indication**: suspected testicular torsion
  - **Timing**: ideally, within 6 hours of symptom onset
  - **Procedure**
    - Immediate surgical exploration of the scrotum with reduction (untwisting) and orchiopexy of the affected testis
    - Orchiopexy of the contralateral testis is recommended because the risk of testicular torsion on the contralateral side increases with previous or current testicular torsion.
    - Orchiectomy if the testis is grossly necrotic or nonviable

Circumcision

6th year seminar

Penile foreskin

- The foreskin is a piece of skin that covers the round tip of the penis.
- It is a double-layered fold of smooth muscle tissue, blood vessels, neurons, skin, and mucous membrane that covers and protects the glans penis and the urinary meatus. It is also described as the prepuce.
- The foreskin is mobile, fairly stretchable, and acts as a natural lubricant, and is lined up by an external keratinized layer and an internal mucosal layer.
- When a baby is born, the foreskin is adherent to the glans penis. These adhesions separate spontaneously with time, allowing the foreskin to become retractile. At 1 year of age, about 90% of boys have a non-retractile foreskin. By 16 years this declines to just 1%.
- Sometimes, the foreskin doesn’t separate when it should and remains tight, a condition called phimosis. Usually, phimosis requires further intervention or circumcision to correct it.

Indications

❖Medical indications

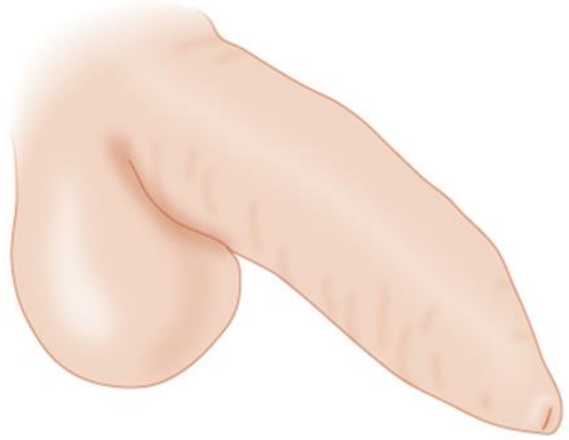
- Recurrent and/or refractory balanoposthitis
- Recurrent UTIs
- Pathological phimosis and paraphimosis (Tx: dorsal slit)
- Prevention of HIV transmission in countries with high infection rates
- Queyrat erythroplasia

❖Elective

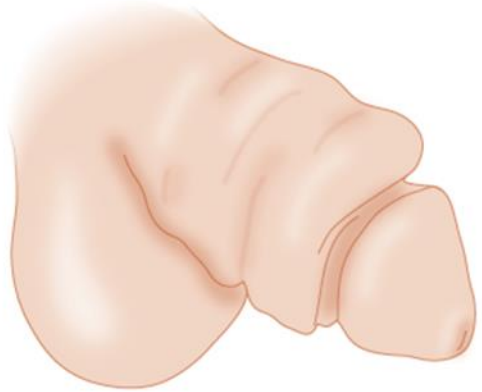
- Customarily performed during the neonatal period because of social norms and/or religious laws (e.g., in Judaism and Islam)
- Cosmetic reasons

❖Benefits

- Decreased risk of UTIs during infancy
- Decreased risk of acquiring certain STIs, such as HIV and HPV
- Decreased risk of penile cancer
- Decreased risk of some penile inflammatory disorders (e.g., balanitis)



Phimosis



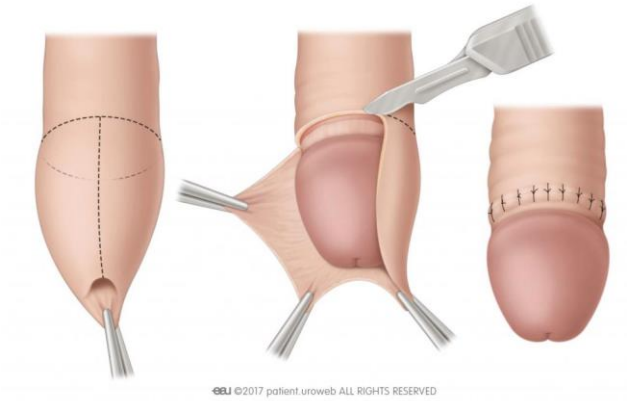
Paraphimosis

Contraindications

- Bleeding diathesis (e.g., hemophilia)
- Prematurity
- Failure to thrive
- Ill baby
- Certain genital structure abnormalities
  - Hypospadias (urethral meatus on the ventral aspect of the penis)
  - Epispadias (rare malformation in which the urethra ends in an opening on the upper aspect of the penis)
  - Chordee (head of the penis curves upward or downward)
  - Webbed penis (scrotal skin extends onto the ventral penile shaft)
  - Ambiguous genitalia, because the foreskin may be needed for reconstructive surgery
  - Buried penis (normal size but hidden under the skin of the abdomen, thigh, or scrotum)
  - Penis torsion
  - Micropenis

Management

- **What happens during a circumcision?**
  - Separates the foreskin from the head of the penis.
  - Uses a scalpel to remove the foreskin.
- **Freehand technique**
  - Physician removes the foreskin with a scalpel. Performed in operating room and requires general anesthesia. Less complications.
- **Clamp techniques**
  - A special clamp is placed to compress the edges of the foreskin before removing the 'extra' skin. Generally utilized for neonatal circumcision. Most appropriate when infant is under 4-6 weeks of age and under 12 pounds.
- **Types of clamps**
  - Gomco clamp (higher bleeding risk)
  - Plastibell clamp (higher site injuries and serious complication)
  - Mogen clamp (higher injury rates to the tip of the penis injuries)



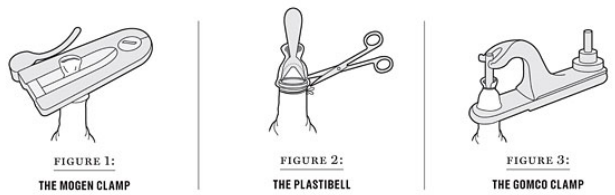


FIGURE 1: THE MOGEN CLAMP

FIGURE 2: THE PLASTIBELL

FIGURE 3: THE GOMCO CLAMP

Complications

- **Bleeding**: due to injury of the cut dermal edges or frenular artery (end arteries)
- **Infection** (rare): Wound infection, Bacteremia
- **Surgical injury**
  - Inadequate skin removal: can lead to paraphimosis which is an emergency
    - Inadequate skin removal is worse than excessive
  - Removal of excessive skin: In many cases, conservative therapy consisting of wet to dry or antibiotic ointment dressings results in adequate healing by secondary intention
  - Glans injury: If the glans is amputated, the tissue should be wrapped in saline-soaked gauze and placed indirectly on ice for transport. The patient should be transferred immediately to a referral center, as successful reattachment is possible if performed within eight hours of injury
  - Epidermal inclusion cyst occurs when an island of skin is left to heal underneath the skin of the penile shaft. Cysts usually are diagnosed by physical examination. Treatment consists removal of the entire cyst
  - Abnormal scarring, resulting in adhesions or penile curvature
  - Urethral injuries: Urethrocutaneous fistulas (correction requires a second operation that is performed six months after the initial procedure)