Chapter 17

Abdominal Wall, Peritoneum, and Diaphragm

Abdominal Wall Defects

- Gastroschisis (Greek for “belly cleft”)
  - Bowel is histologically normal, but thickened and shortened due to prolonged contact with amnionic fluid; these changes are reversible with time and after reduction into the abdominal cavity
  - Defect is almost always to the right of the umbilical cord (which is normally positioned) and separated from it by a skin bridge
  - Midgut, stomach, and gonads are the most commonly herniated organs (liver rarely herniates in gastroschisis)
  - There is no sac covering the herniated viscera
  - Malrotation is always present (in both gastroschisis and omphalocele)
  - Associated anomalies are rare (except intestinal atresia)

- Omphalocele
  - In most cases, the bowel is covered by an intact membrane, from which the umbilical cord arises (ie, herniation into the base of the umbilical cord) so gastrointestinal tract function is usually normal; in a “ruptured” omphalocele, the membrane is not intact
  - Malrotation is present
  - Often contains liver

- Incidence
  - Gastroschisis-to-omphalocele ratio: 2–3:1
  - Both are associated with prematurity

- Anomalies
  - Gastroschisis
    - Associated conditions
      - Undescended testicles (common)
      - Atresias due to vascular compression in utero
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- Hypoperistalsis
- Necrotizing enterocolitis

Omphalocele
- 60% have associated abnormalities (cardiac and chromosomal; eg, trisomy 13, 18, or 21)
- Associated with Beckwith-Wiedemann syndrome, which can result in:
  - Macroglossia, which may cause airway problems
  - Visceral hypertrophy (cardiomegaly, pancreatic β-cell hyperplasia that results in hypoglycemia)
- Associated with Meckel’s diverticulum, gastrointestinal tract duplications, and ambiguous genitalia
- Increased association of malignant tumors (Wilms, neuroblastoma, adrenal)

- Diagnosis: prenatal ultrasound (> 13 wk) is usually accurate
- Delivery: vaginal delivery is not associated with more complications than cesarean delivery, with the exception of large omphaloceles
- Treatment
  - General
    - Set orogastric tube to low suction
    - Use a “bowel bag” or plastic cling film to conserve body heat, minimize evaporative heat loss, and prevent traction or twisting of the mesenteric blood supply—the most important aspects of pretransport and preoperative preparation
  - Antibiotics
    - Ampicillin (100 mg/kg/day)
    - Gentamicin (5–7 mg/kg/day)
  - Intravenous (IV) fluids
    - 20 cc/kg Ringer’s lactate or 5% albumin
    - 150–175 cc/kg Ringer’s lactate during the first 24 hours
    - If not adequately hydrated, the child may become hypotensive on induction of anesthesia due to hypovolemia
  - Vitamin K
    - Premature infant: 0.5 mg intramuscular (IM)
    - Full-term infant: 1 mg IM
Omphalocele and gastroschisis

Steps for primary closure

- Excise the sac (except if attached to liver)
- Inspect the umbilical cord and number of umbilical arteries (presence of a single artery may be associated with an absent kidney)
- Inspect the bowel for atresias or perforations
- Manually stretch the abdominal wall
- In gastroschisis, if a larger opening is required to reduce the herniated viscera, open the fascia cephalad in the midline
- Manually extrude meconium from the colon after anal dilatation and saline irrigations (do not perform an enterotomy to evacuate meconium)
- A silicon-plastic silo or bioprosthetic may be used to retain the herniated viscera
- Do not excise a Meckel’s diverticulum
- The liver must be reduced carefully to avoid torsion of the hepatic veins or occlusion of portal vein inflow that results in hemodynamic instability and injury to capsule
- Complications of an excessively tight closure include:
  - Respiratory insufficiency (secondary to excessive pressure on the diaphragm; peak inspiratory pressure should be $< 35 \text{ cm H}_2\text{O}$ after fascial closure, abdominal compartment pressure should be $< 15 \text{ cm}$)
  - Vena cava compression, resulting in decreased venous return
  - Decreased renal vein flow, resulting in decreased glomerular filtration rate
  - Decreased mesenteric artery flow, resulting in bowel ischemia (an immediate primary repair in an infant with respiratory distress syndrome is contraindicated due to resultant high mortality)
  - If there is any question about bowel viability, place a silo and perform a second look in 12–24 hours
  - Do not attempt definitive repair until chromosomal abnormalities and possible cardiac defects have been evaluated
Delayed primary closure

- Sew silicon-plastic or Gore-Tex (WL Gore & Associates, Inc, Newark, Del) sheets to the edges of the fascia; a silicon-plastic bag (with an integrated spring to hold the base open) can also be used, and does not require suturing
- Gradually reduce the silo over several days
- Remove the silo and perform primary fascial closure after about 7 days
- Allow the sac (which must be intact) to thicken and epithelialization to occur before applying escharotic or desiccating agents
  - Use Acticoat (Smith & Nephew, London, England) dressing and change every 4–5 days
  - Povidone-iodine may be associated with iodine absorption and suppression of thyroid-stimulating hormone
  - 0.5% silver nitrate may be associated with hyponatremia
  - Apply silver sulfadiazine twice a day for 2 weeks, and perform primary skin closure in about 2 months
- Indications for applying escharotic agents include suspected chromosomal syndromes (eg, trisomy 13 or 18); severe, unstable cardiac defects (eg, hypoplastic left heart, hypoplastic aortic arch); and, in premature infants, hyaline membrane disease, primary pulmonary hypertension, and sepsis
- Intestinal atresia
  - Conserve bowel when feasible
  - Perform ileostomy

Postoperative considerations
- Respiratory: ventilator support for 48–72 hours, pharmacological paralysis (using pancuronium bromide)
- Provide total parenteral nutrition (TPN) until full enteral feeds can be resumed
  - Normal intestinal absorption may be delayed for weeks
  - Dysmotility problems may persist for weeks to months
If gastrointestinal contrast studies show no evidence of obstruction, do not operate; operating will only create new adhesions
- Place a gastronomy tube, continue TPN, administer physical therapy for sucking, give erythromycin to increase motility, and be patient
  - Administer 5% albumin infusions as needed
  - Give antibiotics until the silicon-plastic silo has been removed
  - Patient may experience increased gastroesophageal reflux due to increased intraabdominal pressure; this rarely requires fundoplication
  - After successful skin closure of a giant omphalocele, a ventral hernia repair may be associated with hemodynamic instability due to hepatic venous anatomy

- Long-term complications
  - Stricture may result in a small-bowel obstruction
  - Ventral hernia
  - Undescended testicles

- Survival depends on prematurity, size of the defect, and severity of associated anomalies

**Disorders of the Umbilicus**

- Umbilical drainage
  - Omphalitis
    - **Etiology**
      - Poor hygiene
      - Most commonly caused by *Staphylococcus aureus*
      - *Clostridium perfringens* (purple wound) is rare, but is associated with high mortality
    - May result in portal vein thrombosis, leading to portal hypertension and, eventually, to upper gastrointestinal bleeding (treated nonoperatively)
  - Granuloma
    - Small: treat with silver nitrate
    - Large: treat by surgical excision (electrocautery)
  - Polyp
    - Physical appearance: glistening, cherry-red nodule
Pathology: vitelline duct remnant consisting of a small piece of intestinal mucosa

Treatment: surgical excision with central core of umbilicus (silver nitrate does not work!)

- Persistence of vitelline duct structures: omphalomesenteric sinus, fistula, Meckel’s diverticulum, cysts, bands

Urachal sinus

- Definition: cord-like, mucosal-lined structure extending from the dome of the bladder to the lower border of the umbilical ring
- Symptoms: urinary fistula
- Associated conditions
  - Bladder outlet obstruction (eg. posterior urethral valves)
  - Recurrent cystitis
- Diagnosis: voiding cystourethrogram
- Treatment: excision of entire tract down to the bladder via an infraumbilical incision

Urachal cyst

- May occur anywhere along urachal tract, from the umbilicus to the bladder
- Symptoms: enlarging suprapubic or infraumbilical mass, which may become infected, resulting in an urachal abscess
- Diagnosis: ultrasound, voiding cystourethrogram
- Treatment: drain abscess, excise cyst (infraumbilical incision) after infection is resolved

Patent omphalomesenteric sinus

- Etiology: persistent vitelline duct, usually at the site of a Meckel’s diverticulum (located in the ileum)
- Symptoms
  - Contents of small bowel draining through the umbilicus
  - Obliterated cord may become fixed to the umbilicus and may be associated with a closed-loop intestinal obstruction
- Diagnosis: ultrasound, sinogram
- Treatment: surgical excision via an infraumbilical excision

Umbilical hernias

- Most occur through the umbilical vein (cephalad) portion of the ring, where the cicatricial scar is less dense
- Most important factors in assessing likelihood of closure
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include:

- Age of the patient
- Size of the fascial defect (fascial defects > 1.5 cm are unlikely to close after 4–5 years of age)

Indications for operation

- Absolute: incarceration (rare in infants), strangulation, evisceration
- Relative: fascial defect persisting after age 5, a herniation that is proboscis-like (repair at 2 years)

Repair technique

- Infraumbilical incision
- Excision of the sac
- Transverse fascial closure
- Attachment of fascial skin to underlying fascia
- Pressure dressing for 72 hours to prevent a hematoma

Epigastric hernia

- Location: linea alba above the umbilicus
- Etiology: congenital fascial defect
- Pathology: protrusion of preperitoneal fat
- Symptoms: pain, tenderness
- Physical appearance: small, palpable, irreducible mass in the midline (usually supraumbilical, often multiple)
- Treatment: operative repair via transverse incision

Spigelian hernia

- Location: between semilunar line and lateral border of the rectus sheath
- Physical appearance: prominent when patient is crying and straining
- Sac may contain omentum or bowel
- Diagnosis: computed tomography (CT) scan, ultrasound
- Treatment: surgical repair

Inguinal Disorders

- Anatomy
  - Processus vaginalis
    - Defined as peritoneal diverticulum that extends through the internal inguinal ring
    - In an indirect hernia, hernia sac is anterior and medial to the cord structures, lateral to the epigastric vessels
Communicating hydrocele
- Defined as intraperitoneal fluid that has tracked down the patent processus vaginalis into the tunica vaginalis
- In females, a hydrocele may occur along the round ligament (canal of Nuck), presenting as a bulge or mass in the labia majora

Incidence of inguinal hernias
- ~ 3% overall
- Male:Female ratio = 10:1
- Right = 50%; left = 25%; bilateral = 15%, due to later descent of the right testis and delayed obliteration of the processus vaginalis
- Increased in premature infants (~ 20% incidence)
- Increased-risk patients include those with cystic fibrosis, chronic lung disease, connective tissue disease, and those on peritoneal dialysis or with a ventriculoperitoneal shunt
- Treatment: routine bilateral explorations are recommended
- Direct and femoral hernias are rare in children

Hydrocele
- Noncommunicating hydroceles usually resolve by 12 months of age; if not, they should be considered hernias and repaired
- Diagnosis
  - Usually absent upon waking, and most prominent once the child is ambulatory due to movement of intraperitoneal fluid through the processus (resulting from gravity)
  - Transillumination or ultrasound
- Treatment (if hydrocele is present after 12 months of age)
  - Ligate patent processus vaginalis, excise hydrocele
  - Aspiration is not recommended
    - If the processus is patent, the hydrocele will recur
    - If the hydrocele is encysted, it will resolve spontaneously

Symptoms of inguinal disorders
- Indirect hernia (most common)
  - Groin bulge extending toward the top of the scrotum
  - A palpable mass in a female in the labia majora usually represents an ovary (if the mass is bilateral, suspect tes-
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- **Particular feminization syndrome; strangulation is rare**
  - Visibility is better during periods of increased intraabdominal pressure (eg, crying, stooling)
  - Cord thickens as it crosses pubic tubercle (“silk glove” sign)
  - Hernia sac is anteromedial to cord structures and lateral to the epigastric artery and vein

- **Direct hernia**
  - Rare in children
  - May result from an injury to the floor of the inguinal canal during a previous hernia operation
  - Occurs medial to epigastric vessels

- **Hydrocele**
  - Communicating: intermittent scrotal swelling
  - Noncommunicating: remains the same size

- **General principles of treatment**
  - Patients needing treatment include:
    - Those with a hernia on physical examination, or with a convincing history consistent with a hernia
    - Patients who present with incarcerated hernias that can be reduced; these should be admitted and have semielective repair before discharge (waiting 24 hours from time of reduction to operation permits edema to subside)
    - Hernias in premature infants (usually indirect) should be repaired prior to the child’s discharge
    - If a patient has an undescended testicle at the time of hernia repair, perform an orchiopexy at the same time
    - Infants < 50 weeks old (corrected gestational age) who were premature should be admitted for overnight observation postoperatively to monitor for residual effects of anesthesia (apnea and bradycardia)
    - Excise an appendix testis to prevent torsion

- **Complications**
  - Incarceration: sac contents (usually bowel) cannot be reduced nonoperatively
    - Most common in infants < 1 year old
  - **Symptoms**
    - Severe irritability
    - Cramping abdominal pain
Vomiting

- Physical manifestation: firm, nontender mass in groin
- Pathology
  - Decreased venous/lymph drainage increases edema and pressure
  - This leads to decreased arterial perfusion and eventual gangrene and necrosis, which presents as scrotal redness, edema, or a mass (strangulation)

- Treatment
  - Attempt reduction
    - Sedate (using midazolam 0.05–0.1 mg/kg, or chloral hydrate)
    - Elevate the lower body
    - Apply ice to the hernia sac
  - If nonoperative reduction is successful, admit the patient for repair in 24 hours
  - If nonoperative reduction is unsuccessful, perform immediate operative reduction
  - If patient’s bowel is infarcted, perform a resection through the groin incision if possible; if not, perform a laparotomy

- Complications of incarceration
  - Gonadal infarction
  - Intestinal obstruction
  - Gangrenous bowel
    - Physical appearance: erythematous scrotum
    - Attempts at reduction are usually unsuccessful

- Recurrence
  - A direct hernia may occur at the site of repair of a previous indirect hernia if there has been an intraoperative injury to the floor of the inguinal canal
  - Excessively tight closure of the internal inguinal ring can result in recurrence
    - Symptoms: tender, swollen testis
    - Treatment: reexploration and inspection of the testis

Disorders of the Peritoneum and Peritoneal Cavity
- Abdominal compartment syndrome
  - Etiology: increased intraabdominal pressure due to an
inflammatory process (eg, perforated appendix) or any space-occupying condition (eg, bleeding, edema) that increases the volume of the abdomen

- Symptoms
  - Sepsis
  - Respiratory distress due to pressure on the diaphragm
  - Oliguria due to renal vein compression
  - Hypotension due to compression of vena cava and decreased venous return
  - Vasomotor changes
  - Acidosis due to hypoperfusion

- Physical appearance: abdominal distension

- Diagnosis: intraabdominal pressure > 20 cm H₂O, as determined by measuring bladder pressures

- Treatment
  - Assisted ventilation
  - IV fluids
  - Treatment of underlying cause
  - Abdominal decompression using a silo or wound vacuum device

- Meconium peritonitis
  - Frequently associated with cystic fibrosis as the etiology of obstruction
  - Types
    - Pseudocyst: meconium is contained by necrotic bowel and omentum; cyst wall is lined with calcium
    - Plastic: free perforation causes marked generalized inflammatory reaction and adhesions
    - Generalized: prenatal perforation with continuing leak produces meconium ascites
  - Etiology
    - Intrauterine volvulus of meconium-filled loop of bowel, leading to intestinal vascular compromise
    - This results in bowel ischemia then atresia, which leads to obstruction and, finally, perforation
  - Symptoms: polyhydramnios, abdominal distension, and bilious vomiting
  - Diagnosis
    - Prenatal ultrasound shows polyhydramnios, dilated
bowel loops, or calcifications
  ▶ Abdominal radiograph shows dilated loops of intestine and intraabdominal calcifications (linear calcifications may line the processus vaginalis and scrotum)
  ◦ Treatment
    ▶ For obstruction or pneumoperitoneum, perform laparotomy with conservative resection
    ▶ In asymptomatic cases, observe patient closely
• Omental and mesenteric cysts
  ◦ Pathology: usually lymphangiomas
  ◦ Symptoms: pain, vomiting, and abdominal mass
  ◦ Diagnosis: CT scan or ultrasound
  ◦ Treatment: excision
• Ascites
  ◦ Etiology
    ▶ Urinary tract malformation (eg, ureteropelvic junction obstruction, posterior urethral valves) resulting in obstruction (most common etiology in the neonate)
    ▶ Immune hydrops (Rh incompatibility, cardiac anomalies)
    ▶ Pancreatitis
    ▶ Ovarian cyst or tumor
    ▶ Chyle due to lymphatic abnormality
  ◦ Treat underlying abnormality
• Peritoneal adhesions
  ◦ Immediate postoperative bowel obstruction is usually due to small bowel intussusception
  ◦ Diagnosis: upper gastrointestinal study with small bowel follow through
  ◦ Treatment (initial): nasogastric tube, IV fluids, antibiotics, laparotomy with adhesion lysis for complete obstruction

Congenital Diaphragmatic Hernia
• General
  ◦ Normal openings in the diaphragm occur at the aorta, vena cava, and esophagus
  ◦ Late-onset diaphragmatic hernias
    ▶ Diagnosis: chest radiograph in an asymptomatic patient
    ▶ Symptoms
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- Gastrointestinal obstruction
- Respiratory distress (severe hypercarbia, hypoxia)
  - Treatment: repair
  - Differential diagnosis: congenital cystic adenomatoid malformation, sequestration

- Defects
  - Esophageal hiatus (Morgagni): stomach prolapses into the mediastinum
  - Congenital posterolateral defect (Bochdalek)
  - Anomalous attachment of diaphragm to sternum and ribs
  - Epigastric omphalocele and retrosternal defect in the diaphragm and pericardium (Pentalogy of Cantrell), producing herniation within the pericardium
  - Attenuation of the tendinous or muscular portion of the diaphragm produces eventration (phrenic nerve injury)

- Incidence
  - Associated with malrotation
  - Occurs more frequently in females than males
  - Occurs most often on the left (90%)

- Diagnosis
  - Prenatal diagnosis
    - Polyhydramnios (75%)
    - Associated with major central nervous system, cardiac, and chromosomal abnormalities (eg, trisomy 13 and 18); many are stillborn
  - Postnatal diagnosis
    - View prenatal ultrasound
    - Chest radiograph will show tip of the nasogastric tube to be above diaphragm, indicating herniation of the viscera into the chest and a mediastinum shift
    - Rule out associated abnormalities (occur in 15%–25% of cases) in the following:
      - Central nervous system: head, spine (meningo-myelocele)
      - Heart (most common): patent ductus arteriosus, ventricular septal defect
      - Kidneys
      - Lung: sequestration (occurring most commonly in left lower lobe)
Rule out chromosomal (trisomy 13) and metabolic abnormalities

Pathophysiology
- Herniated viscus becomes distended with air, displacing the mediastinum
- Increased pulmonary artery pressure and pulmonary vascular resistance resulting from decreased pulmonary artery branches and thickened muscularis of bronchioles
- A right-to-left shunt through a patent ductus arteriosus and the foramen ovale results in hypoxia, hypercarbia, and acidosis; these lead to increased pulmonary vasoconstriction
- Acidosis and hypercarbia lead to pulmonary vasodilation
- Alkalosis and hypocarbia lead to pulmonary vasoconstriction

Physical appearance
- Respiratory distress is evident by grunting, flaring, retracting, and cyanosis
- Scaphoid abdomen
- Shifted heart sounds, decreased bowel sounds
- Tracheal deviation
- Bilateral pulmonary hypoplasia (more severe on the side of herniation)

Treatment
- Support spontaneous respiration; provide sedation with paralysis only if necessary
- Endotracheal intubation (avoid bag-mask ventilation to prevent insufflation of air into the stomach and small bowel)
- Insert repogle nasogastric tube
- Ventilatory settings
  - Peak inspiratory pressure < 25 cm H₂O
  - Positive end expiratory pressure < 6 cm H₂O (to minimize barotrauma)
  - 100% oxygen
  - Adjust rate and inspiratory–expiratory ratio to maximize partial pressure of oxygen (PaO₂), decrease partial pressure of carbon dioxide (PaCO₂)
  - Maintain pH > 7.20 (permissive hypercapnea)
- Place umbilical venous catheter, umbilical arterial catheter (postductal), or right radial A line (preductal)
When placing an umbilical arterial catheter, ideal arterial blood gas levels should be as follows: $\text{PaO}_2 > 40$ mmHg, $\text{PaCO}_2 < 30$, pH > 7.5
- Administer sodium bicarbonate or trimethamine drip
- Provide volume replacement (Ringer's lactate or 5% albumin)
- Maintain oxygen content between aorta and right atrium < 5%

Oxygen content ($\text{CaO}_2$) = ($\text{SaO}_2 \times \text{Hgb} \times 1.34) + .003(\text{PaO}_2)$

$\text{SaO}_2$: arterial oxygen saturation
$\text{PaO}_2$: mixed venous oxygen saturation

- Maintain mixed venous (from right atrium) saturation > 65%
- Administer dopamine/dobutamine as needed
- Tolazoline enhances histamine release, but can result in hypotension and peptic ulcers
- Nitric oxide (an endothelium-derived relaxing factor), which decreases pulmonary vasoconstriction, may be helpful
- Perform surgical repair after pulmonary hypertension has resolved
- Appropriate pulmonary vasodilators to administer include prostaglandin E1 and E2; appropriate pulmonary vasoconstrictors include prostaglandin F, thromboxane A1 and B2, and leukotrienes

**Surgical repair**
- Perform when patient is physiologically stable, not as an emergency measure
- A diaphragmatic hernia is not an indication for fetal surgery
- Administer general anesthesia (eg, pancuronium)
- Make a subcostal abdominal incision for right-sided or left-sided defects or thoracoscopy
- Reduce herniated viscera by excising sac; a hernia sac is present in 10%–20% of patients and may be easily missed
- Primary repair
  - Small defect: use pledgets
  - Large defect: use bioprosthetic or synthetic prosthesis
- Place a thoracostomy tube to underwater seal
- A purple-brown mass near or in the left lower lobe may represent a sequestration
- Blood is usually supplied from a branch of the abdomi-
nal aorta below the diaphragm

- Treatment consists of excision with repair of the diaphragm

- Postoperative care
  - Administer IV fluids (dopamine or dobutamine may be added)
  - Anticipate pneumothorax

- Results
  - Overall mortality ~ 50% (unchanged for past 20 y)
  - Prognosis is determined by the degree of hypoplasia
  - Lung volumes approach equality on a ventilation/perfusion scan
  - Residual volume and functional residual capacity are increased
  - Vital capacity, forced expiratory volume, and minute ventilation volume are normal or slightly decreased

**Eventration of the Diaphragm**

- Most common cause is injury to the phrenic nerve from stretching of the neck at the time of birth or from direct operative trauma

- Symptoms
  - Inspiration leads to negative intrathoracic pressure, which causes herniation of viscera into the ipsilateral chest
  - This leads to mediastinum shift to the contralateral side, which results in respiratory distress and/or pneumonia
  - Occurs more frequently in the left diaphragm than the right

- Diagnosis
  - Chest radiograph will show elevated hemidiaphragm
  - Fluoroscopy of the diaphragm or ultrasound shows elevated diaphragm with paradoxical motion
  - Differential diagnosis: congenital diaphragmatic hernia with hernia sac

- Treatment
  - Intubation with assisted ventilation initially
  - Perform transthoracic (seventh intercostal space) plication **only** if patient is symptomatic (ie, ventilator support required for > 2 wk)