Esophageal Atresia & Tracheo-Esophageal Fistula

- Esophagus dead-ends in a blind pouch.
- TE fistula is an abnormal passage between the esophagus and trachea.
- Occur together in 85% of cases.
- Results from failure of trachea and esophagus to divide at 34-36 days of gestation.
- Not always obvious on prenatal US

Esophageal Atresia & Tracheo-Esophageal Fistula

- Incidence: 1 in 4500
- Frequently, history of polyhydramnios
- Identification
  - Cannot swallow saliva with atresia
  - Choking or cyanosis with feeding
  - Gastric Tube will not pass to stomach
  - May develop gastric distension if there is a fistula, as air cannot get out of stomach

Esophageal Atresia & TEF: Management

- Elevate head of bed 30° - prone may help avoid aspiration
- NPO
- IV fluids
- Feeding tube or Replogle to low suction in pouch
- Assess for associated anomalies (30-70%)
VACTERR (or VACTERL)

V: Vertebral
A: Anorectal
C: Cardiac
T: Tracheal
E: Esophageal
R: Renal
R: Radial (or L: Limb)

TEF/Esophageal Atresia: Repair

- Primary repair often possible
- Repair is delayed if there are coexisting problems or baby is too small.
- Staged repair when the gap between distal and proximal esophagus too large. Gastrostomy placed and repair done in 6-8 weeks.
- Prognosis excellent if no other anomalies. Over 90% survival.

Gastrochisis:
Evisceration of the bowel through a defect beside the umbilicus (no sac) in an otherwise normally formed abdomen.

Gastrochisis: Incidence
- Increasing incidence reported in reports from around the U.S. and the world since early 1990's. (~ 4 in 10,000)
- Teen mothers

Gastrochisis: Features
- Defect is small, to the right of the umbilicus
- No sac covering
- Small and large intestine, rarely the liver, stomach, or bladder
- Intestine may be edematous and inflamed due to exposure to amniotic fluid
- An isolated defect - other anomalies uncommon, except malrotation and acquired atresia
Gastroschisis: Management

- Protect defect and prevent heat and fluid loss
- Place in sterile bowel bag moistened with sterile saline and cinched at the axilla
- Leave intestine visible to watch gut perfusion
- Position on side to prevent tension on defect causing vascular compromise

Gastroschisis: Management

- Keep handling to a minimum
- Sterile gloves
- NPO, NG tube or Replogle to low suction
- IV for fluids at 150 ml/Kg/day -- increased hydration needs
- Antibiotics

Gastroschisis: Management

- Surgical emergency: repair ASAP
- Primary repair
- Staged reduction: partial replacement of intestine to abdomen, placement of a silastic silo, and daily reduction until closure is possible
- Long postoperative course, requiring TPN and slow feeding

Omphalocele: Definition

- Herniation of the abdominal viscera into the base of the umbilical cord.
- Usually covered by a sac (or remnant) with umbilical arteries and vein inserted into the base of the defect.
Omphalocele: Features
- Defect can be small. Any unusually large umbilical cord should be inspected carefully prior to clamping.
- 1/5,000 - 6,000 live births
- Larger defects may include liver, stomach, spleen as well as intestines
- Associated with many anomalies; chromosomal or other structural defects in 50-70%
- Development of abdominal muscle and peritoneal layers incomplete; abdominal cavity often small and underdeveloped

Omphalocele: Management
- In delivery room and preparation for transport, same as gastroschisis
  - IV fluids
  - Cover with bowel bag or transparent drape wrap
  - NPO, gastric decompression (NG or replogle)
  - Assessment for associated anomalies

Omphalocele: Surgical Management
- Primary repair: all intestine returned to abdomen, skin closed
- Break in sac may require silo technique
- Management of choice: Dressing and epithelialization with later closure of fascia
Bowel Obstruction

- Blockage of the GI tract may be mechanical (anatomical), acquired mechanical, or functional

Bowel Obstruction: Causes

- Atresias (1 in 2,500-5,000 births)
- Hirschsprung Disease (1 in 5,000 births)
- Meconium ileus/plug (? Cystic Fibrosis)
- Hernia
- Malrotation/volvulus
- Necrotizing enterocolitis (more common in preemies, but can occur in term infants)

Intestinal Malrotation with Midgut Volvulus

Bowel Obstruction: Features

- History of polyhydramnios
- Abdominal distension with visible bowel loops, absence or hyperactivity of bowel tones
- Vomiting, especially bile-tinged
- Greater than 20-30 cc stomach aspirate at birth
- No meconium
- Depending on cause, timing of onset varies - may be present at birth or develop soon after
Bowel Obstruction: Management

- NPO with NG or Replogle to low suction, or aspirate every 20-30 minutes
- IV for fluid, glucose, electrolyte management
- Measure and track abdominal girth
- Abdominal films may help diagnose if air is absent from the distal bowel.
- Good surgical outcomes

Myelomeningocele (Spina Bifida)

- Neural tube defects result from failure of the neural tube to close normally at 3-4 weeks gestation
- Range of defects from spina bifida occulta (covered with skin) to myeloschisis, in which the whole spinal cord is without dermal or vertebral covering

Myelomeningocele

Myelomeningocele (Spina Bifida) Spina Bifida

- Prenatal detection by quad screen (blood), ultrasound, and amnio
- 80% in lumbar region
- 0.7 in 1,000 births in U.S.
- Incidence of mental retardation less than 20%, but most develop hydrocephalus and need ventriculoperitoneal shunt.

Spina Bifida

- Prenatal detection by quad screen (blood), ultrasound, and amnio
- 80% in lumbar region
- 0.7 in 1,000 births in U.S.
- Incidence of mental retardation less than 20%, but most develop hydrocephalus and need ventriculoperitoneal shunt.

Myelomeningocele: Management

- Ventilate or resuscitate from side if necessary.
- Keep on side or abdomen.
- Sterile, NON-LATEX, powder-free gloves for handling
- Measure and examine defect, then dress to keep sterile, moist, and protected. Prevent HEAT LOSS.
- NPO, IV, antibiotics if ruptured (high infection risk).
Secure feeding tube to back of Telfa dressing and attach syringe of sterile saline.

Protect with sterile “donut” to cushion spinal cord lesion from injury

Cover the dressing with a piece of steri-drape.

Secure the donut and dressing with a 6” piece of Bandnet or Kerlix around the baby’s middle.

For a final touch, the mudflap...

Diaphragmatic Hernia

- Herniation of abdominal organs into the thoracic cavity through a defect in the diaphragm due to early failure of the closure of the diaphragm. This usually results in hypoplasia of the lung.

Diaphragmatic Hernia: Features

- Incidence: 1 in 4000; 90% on left
- Detectable on prenatal ultrasound
- 50% associated with other anomalies: neural tube defects, heart defects, intestinal malrotation
- May be mild and asymptomatic or severe and life-threatening

Diaphragmatic Hernia: Features

- Respiratory distress at birth or soon after
- Cyanosis, decreased breath sounds on one side of chest
- Muffled or displaced heart sounds on (usually) right side of chest
- Bowel tones in chest
- Diagnosis confirmed by X-Ray

May or may not have barrel chest, scaphoid abdomen:
Diaphragmatic Hernia: Management

- Avoid bag-mask ventilation! Early intubation recommended.
- Decompress stomach with OG or Replogle.
- Elevate head of bed
- NPO, IV for fluid and electrolyte management
- Antibiotics if risk of sepsis

Surgical repair usually deferred until pulmonary hypertension is controlled.

May have lung hypoplasia and need high frequency ventilation, nitric oxide or ECMO

Primary closure usually possible

Patch or muscle flap may be used to close defect.

Survival depends on preoperative status. 40-60% survival if severe symptoms appear within the first 6 - 8 hours of life.

Depends on severity of defect and any other associated anomalies.

Imperforate Anus

- Failure of differentiation of urogenital sinus and cloaca during embryological development. May be high or low in colon.

Imperforate Anus: Features

- 1 in 5,000 live births
- High imperforate anus associated with lack of innervation
- Low imperforate anus may have dimple or appear normal in rectum.
- 50% or more have associated anomalies, most frequently genitourinary
Imperforate Anus: Management

- NPO, IV for fluids, glucose, electrolytes
- Gastric decompression by NG (Replogle) to low intermittent suction
- Diagnosis by physical exam, X-Ray, and ultrasound
- Surgical emergency: Procedure dependent on level of anorectal pouch. Low pouch usually can be a primary repair.

References


