Chapter 7
Surgery

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Questions

1. Which of the following organisms are associated with sepsis occurring after abdominal surgery?
   A. *Haemophilus influenzae* type B
   B. *Neisseria gonorrhoeae*
   C. Staphylococci
   D. Enteric gram-negative rods

2. Gastroschisis differs from omphalocele in the following way(s):
   1. Gastroschisis is usually a 2–4 cm defect, often with a right paramedian location.
   2. In infants with gastroschisis, the bowel is not covered by membranes.
   3. Omphalocele is associated with a greater incidence of non-GI anomalies such as congenital heart disease, bladder exstrophy, and/or cloaca.
   4. A silo is often used in closure of larger defects.
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

3. Which is the most common type of esophageal atresia?
   A. Esophageal atresia (EA) with a posterior tracheoesophageal fistula (TEF) near the carina
   B. "H"-type EA/TEF
   C. Esophageal connection to the trachea with a second, more distal TEF
   D. Esophageal connection to the trachea without a distal TEF

Answers

1. D. Enteric gram-negative rods

The administration of prophylactic antibiotics in the OR and in the perioperative period is an important measure in reducing the incidence of infection. Overuse of antibiotics has been implicated as a cause for the development of *Clostridium difficile* toxin-associated diarrhea.

2. E. All of the above

Omphalocele is a midline defect of variable size involving the omphalos (Greek: "navel"). Omphalocele is associated with chromosomal abnormalities, other GI defects (35%), cardiac defects (20%), and also cloacal exstrophy. Only 10% of patients with omphalocele are born preterm. Omphalocele is part of the Beckwith-Wiedemann syndrome (macroglossia, hyperinsulinism, hypoglycemia, and gigantism). Omphalocele results when the intestines fail to return into the abdomen from the umbilical coelom. With omphalocele, the bowel is covered with membranes, decreasing fluid losses. In gastroschisis, a part of the small intestine herniates through the abdominal wall.

Sixty percent of patients with gastroschisis are born prematurely. A few affected patients have jejunal atresia, but other anomalies are not seen with this condition. Repair of either defect may involve a "silo" if the extruded intestines do not fit into the smaller abdominal cavity.

3. A. Esophageal atresia (EA) with a posterior tracheoesophageal fistula (TEF) near the carina

TEF is the failure of the linear division of the trachea and esophagus during embryogenesis. The most common type (85–90%) is a proximal blind pouch with a distal TEF. All patients present with aspiration at birth with respiratory distress and inability to handle oral secretions. Mortality is approximately 3% in term infants but can be much higher in preterm newborns and in those with other congenital anomalies. Forty percent of infants born with TEF have associated anomalies, with cardiovascular anomalies being seen most often. TEF is also seen as part of the VATER (vertebral anomalies, anal atresia or arterial anomalies, TEF, renal anomalies) or VACTERL (vertebral anomalies, anal atresia or arterial anomalies, TEF, renal anomalies, limb anomalies) associations.

4. The VACTERL complex includes a tracheoesophageal anomaly and:
   1. PDA, ASD, or VSD
   2. Renal defects
   3. Abnormalities of the bones of the forearm
   4. Spinal dysraphism
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

5. Regarding acute appendicitis in children:
   1. In many children younger than 2 years of age, the appendix is found perforated at operation.
   2. More males than females develop acute appendicitis.
   3. It is unusual for the child with acute appendicitis to have an appetite.
   4. Among school-aged children, the diagnosis is more often missed in girls than in boys.
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

6. Midgut volvulus:
   1. Occurs when the bowel twists upon itself
   2. Occurs when *incomplete* intestinal rotation leads to a shortened mesentery
   3. Leads to vascular compromise of the bowel
   4. Is seen in over 60% of neonates with malrotation
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

7. Duodenal atresia:
   1. Often presents with vomiting which may or may not be bilious
   2. Is often accompanied by other intestinal obstructions in both the small and large intestine
   3. Is associated with trisomy 21
   4. Generally does not present until 1–2 months of life
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

4. E. All of the above

The VATER association includes vertebral anomalies, imperforate anus or arterial anomalies, TEF, and renal anomalies. A single umbilical artery is often seen. In VACTERL, the L stands for limb anomalies. An association does not have a single known etiology. The evaluation of an infant with TEF should include a search for the anomalies in the VACTERL association.

5. A. 1, 2, 3

The annual incidence of acute appendicitis is 4:1000. In adolescent girls, the diagnosis is more difficult because other causes of abdominal pain such as ovarian cysts, ovulatory pain, menstrual pain, and pelvic inflammatory disease mimic appendicitis. Other diagnoses in the differential include gastroenteritis, mesenteric adenitis, inflammatory bowel disease, RUL pneumonia, and urinary tract pathology.

6. E. All of the above

In 70% of patients with malrotation and volvulus, the presentation is within the neonatal period, and in half of these, the presentation is in the first 10 days of life. The balance of cases can present at any time, even into adulthood. Malrotation is twice as common in boys as girls. Presentation includes distention and bilious vomiting. X-ray studies (plain film or upper GI contrast study) confirm the diagnosis. In malrotation, the duodenum is seen in an abnormal position, with the duodenojejunal junction located to the right of the spine.

7. B. 1, 3

Duodenal atresia occurs in 1:20,000 live births. Common findings in affected infants include abdominal distention and jaundice. Maternal polyhydramnios is also often found. Associations in addition to trisomy 21 are congenital heart disease, TEF, and renal anomalies.

8. Intussusception:
   1. Has a peak incidence in infants less than 1 year of age
   2. Presents with colicky abdominal pain, bloody (currant jelly) stools, and vomiting
   3. May be reduced with a carefully performed enema
   4. Is nearly always caused by a "lead point" such as a polyp or duplication
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

9. Hirschsprung's disease, the absence of normal enteric ganglionic neurons:
   1. Has a much higher incidence in Caucasians
   2. Is limited to the rectum and sigmoid in over 50% of cases
   3. Is initially managed with stool softeners since some cases spontaneously resolve
   4. Rarely involves not only the rectum and sigmoid but the entire colon
      A. 1, 2, 3
      B. 1, 3
      C. 2, 4
      D. 4 only
      E. All of the above

10. Hirschsprung's disease can be diagnosed by:
    1. Rectal manometry
    2. Surgical rectal biopsy
    3. Suction biopsy
    4. Barium enema
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

11. Other than organomegaly, which of the following may cause abdominal distention in the neonate?
    1. Pneumoperitoneum
    2. Intestinal obstruction
    3. Ascites
    4. Pyloric stenosis
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

8. A. 1, 2, 3

Intussusception is seen in children from the age of 3–18 months. The incidence varies from 0.5 to 4/1000 live births. In almost all children under 1 year of age, no clear cause is found. The nonspecific signs of vomiting and colicky pain are nearly always part of the presentation. Later in the course of the illness, fever and lethargy are seen and finally the child may pass a currant jelly stool. A carefully done barium enema is used as a diagnostic and therapeutic tool. Excessive pressure in the performance of the enema will perforate the bowel. Up to 50% of patients with intussusception are successfully treated with hydrostatic reduction.

9. C. 2, 4

Hirschsprung's disease is a functional obstruction of the colon or rectum that results from failure of migration of ganglion cells in the developing colon. It is the cause for up to 25% of all cases of bowel obstruction in the newborn and is seen more often in males. The aganglionic segment does not permit normal colonic motility. More than 80% of cases involve only the rectum and a small part of the colon. Management is surgical since the aganglionic segment is permanently contracted. Generally, a colostomy is performed at the level of normal innervation (the so-called leveling colostomy) with a later colonic or ileal pull-thru.

10. E. All of the above

The diagnosis should be suspected in any newborn that does not pass meconium within the first day of life. A frozen section can be done with the biopsy material, which is stained for acetylcholine to identify abnormal nerve trunks. H&E stains confirm the absence of ganglion cells.

11. A. 1, 2, 3

Hepatomegaly and hepatosplenomegaly are possible causes of abdominal distention in the newborn. Pneumoperitoneum is usually a result of GI tract perforation, often in preterm newborns. Among the causes of neonatal ascites, urinary ascites is the most common, followed by cardiac and idiopathic. Pyloric stenosis is an incomplete obstruction at the gastric outlet not associated with ascites.

12. Abdominal masses in the newborn:
    1. Are generally not malignant
    2. Are retroperitoneal in approximately 66% of cases
    3. If retroperitoneal, are most often renal in origin
    4. If due to a tumor, are most likely abdominal teratomas
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

13. Which of the following have been implicated as having a role in the development of necrotizing enterocolitis (NEC)?
    1. Intestinal ischemia
    2. Bacterial colonization of the bowel
    3. Feeding
    4. Multiple births
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

14. Which of the following are associated with intestinal ischemia?
    1. Polycythemia
    2. Umbilical vessel catheterization
    3. Congestive heart failure
    4. An open patent ductus arteriosus
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

15. Patients with extrahepatic biliary atresia (incidence = 1:10–15,000 live births):
    1. Will be cured for life if a Kasai operation is done within the first 3 weeks of life
    2. Are generally well initially and then develop jaundice at 3–6 weeks of age
    3. Should be started on phenobarbital to induce liver enzymes in the remaining normal liver
    4. Will likely die within the first year of life without surgical intervention
       A. 1, 2, 3
       B. 1, 3
       C. 2, 4
       D. 4 only
       E. All of the above

12. E. All of the above

Approximately 10–15% of abdominal masses in the newborn are due to malignant tumors. Types of renal masses seen in newborns include hydronephrosis, polycystic disease, renal vein thrombosis, Wilms' tumor, and mesoblastic nephroma. Other retroperitoneal masses seen in this age group include neuroblastoma, ganglioneuroma, and sacrococcygeal teratoma.

13. A. 1, 2, 3

NEC is the most common gastrointestinal emergency in the infant. The incidence ranges from 1% to 4% of all NICU admissions, with a higher incidence in infants with birth weights <1000 g. The most commonly implicated etiologic factors are an ischemic insult to the gut and the presence of intraluminal bacteria or viruses and substrate (formula or milk). Approximately 93% of infants who develop NEC have been fed enterally. Because of the role of bacteria in NEC, antibiotics are often part of the therapy.

14. E. All of the above

Mesenteric blood flow in the newborn is affected by a variety of factors in addition to those mentioned. During hypoxia, the so-called diving reflex shunts blood from the mesenteric, renal, and peripheral vascular systems to the brain and heart. Polycythemia and also exchange transfusions have been implicated in intestinal ischemia. Other possible etiologies for NEC are RDS, hypotension, hypothermia, and birth asphyxia. The presentation of NEC may include abdominal distention, vomiting and gastric residual, lethargy, hypotension, apnea, and temperature instability.

Lab findings include pneumatosis intestinalis on X-ray, thrombocytopenia, blood and/or reducing substances in the stool, and metabolic acidosis.

15. C. 2, 4

This condition is defined as atresia or hypoplasia of any part of the extrahepatic biliary system.

The most common form includes atresia up to the porta hepatis and even intrahepatic ducts. Approximately 15% of affected children have other defects. Clinical presentation includes jaundice in the second to third week of life; acholic stools; enlarged, hardened liver; and splenomegaly. Conjugated bilirubin is elevated along with alkaline phosphatase, gamma-glutamyl transferase, and transaminases.

16. In pyloric stenosis:
    1. The child develops metabolic alkalosis due to continued vomiting of gastric contents.
    2. There is non-bilious vomiting which may contain “coffee ground” material.
    3. The child may develop hypochloremia.
    4. The diagnosis is best made with a barium swallow.
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

17. Pyloric stenosis, the most frequently occurring cause of gastric obstruction in the newborn:
    1. Is seen in approximately 1:2500 live births
    2. Is equally common in all races and ethnic groups
    3. Is seen with the same frequency in males and females
    4. Is seen in infants as young as 2 weeks and as old as 12 weeks
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

18. Regarding children in MVAs who have a history or physical exam indications of lap belt injury:
    1. These injuries are most common in children under the age of 1 year.
    2. There may also be damage to the spinal cord in these children.
    3. Diagnostic peritoneal lavage is always indicated.
    4. Hollow viscus injury may not be apparent for 12–24 h.
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

16. A. 1, 2, 3

Pyloric stenosis is much more common in males. It is seen in 1:150 males and 1:750 females. The presentation includes progressive, relentless vomiting generally starting between the second and fourth week of life with a range of 2–12 weeks. Typical electrolyte abnormalities seen in infants with pyloric stenosis include hypochloremia, hypokalemia, and hyponatremia along with azotemia. The severity of the child's condition is graded by the degree of hypochloremia. An infant with more severe dehydration will have, in addition to the metabolic acidosis, a paradoxical aciduria. The kidney, in an effort to maintain intravascular volume, absorbs as much sodium as possible, but the only anion available to absorb is bicarbonate. Thus, the urine is acidic despite the systemic alkalosis. Prior to surgical repair, the child should be adequately hydrated and the electrolyte abnormalities corrected. Given the nature of the pathology, gastric outlet obstruction leading to intractable vomiting, administration of contrast is a poor way to make the diagnosis. Ultrasound or air contrast plain X-rays are more current diagnostic tools.

17. D. 4

The incidence of pyloric stenosis is 1:2500 live births. It is seen predominately in whites and is most common in firstborn males. The average age of onset is 3–4 weeks with a range of 2–12 weeks. Interestingly, this anomaly is not seen at birth, and in cases where it is managed medically, the hypertrophic pylorus eventually (after 4–6 weeks) returns to normal and the child stops vomiting, all without surgical intervention. Pyloric stenosis is a common reason for surgery in the neonatal period.

18. C. 2, 4

Injuries to the pancreas can result from either lap belt injury or bicycle handlebars. Injury to the pancreas is difficult to diagnose. Elevations in amylase and lipase may not be seen until 1–2 days after the injury. Rapid deceleration while in a lap belt may also damage the intestines with perforation or even transection possible. If there is intestinal damage, in addition to a bruise over the abdomen in the area of the lap belt, back pain may be a symptom.

19. In the evaluation of the child who has suffered blunt abdominal trauma, organ damage may be indicated by:
    1. Left shoulder pain
    2. Hematuria
    3. Flank ecchymosis
    4. Bilious emesis
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

20. Gastroesophageal reflux (GER) in infants and young children can present with:
    1. Recurrent pneumonia
    2. Irritability
    3. Wheezing, stridor, or hoarseness
    4. Apnea
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

21. Treatment(s) for GER include(s):
    1. Thickened, small, and frequent feeds
    2. Magnesium sulfate to increase gastric pH
    3. H-2 receptor blockade
    4. Avoidance of high-carbohydrate meals
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

22. Esophageal foreign bodies in children:
    1. Are best diagnosed with a barium swallow
    2. Often lodge at or just below the cricopharyngeus muscle
    3. Always require removal if they pass into the stomach
    4. May cause stridor in infants
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

19. E. All of the above

The spleen is the abdominal organ most often damaged following blunt abdominal trauma in children. Kehr's sign, left shoulder pain resulting from pressure on the left upper quadrant, is suggestive of splenic injury. The severity of splenic injury is graded by CT from 1 to 5. Grade 1 is a tear in the capsule, while grade 5 indicates a completely ruptured spleen. Renal damage following blunt abdominal trauma is relatively common in children. Abdominal CT with renal contrast will make the diagnosis in most cases. Flank pain and bruising and urinalysis with blood and protein should raise the suspicion of renal damage.

20. E. All of the above

GER must not be confused with other causes of regurgitation in the newborn/infant such as pyloric stenosis, duodenal stenosis, annular pancreas, malrotation, or any of a host of metabolic diseases. Contrast studies and/or 12–24-h pH probe studies may confirm the diagnosis. Pneumonia and wheezing develop with aspiration of refluxed gastric contents, irritability is due to the pain of reflux esophagitis, and apnea is a possible reaction to the presence of aspirated gastric contents in the trachea or larynx.

21. B. 1, 3

Medical treatment of GER is directed to lowering the pH of the gastric contents and decreasing the amount of reflux. In the well, thriving child with a small amount of post-feeding reflux, observation and reassurance are all that is needed. In more severe cases, placing the child at an angle (30° head up) after feeding may limit the reflux as will the institution of frequent small feedings instead of larger ones.

22. C. 2, 4

A barium study of the esophagus may be useful in the evaluation of upper airway obstruction, often demonstrating posterior esophageal compression from a vascular ring. The cricopharyngeus muscle, located high in the esophagus, often stops a swallowed esophageal foreign body from progressing further. A foreign body which passes into the stomach will likely be passed through the entire GI tract, so retrieval is often not undertaken unless it is indicated by the specific nature of the foreign body (such as an open safety pin).

23. Extraintestinal manifestations of inflammatory bowel disease (IBD) include:
    1. Growth retardation
    2. Peripheral arthritis
    3. Anemia
    4. Reactive airway disease
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

24. Common findings in patients with IBD (ulcerative colitis or Crohn's disease) include:
    1. Iron deficiency anemia
    2. First-degree heart block
    3. Hypoalbuminemia
    4. Stool cultures positive for various enteric pathogens
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

25. Cleft lip:
    1. May be unilateral or bilateral
    2. Occurs with and without cleft palate
    3. Occurs in 1:600–1000 live births
    4. Varies from being a small notch in the vermilion border to a complete separation
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

26. Problems seen in children with cleft palate ± cleft lip include:
    1. Otitis media
    2. Feeding difficulties
    3. Malpositioned teeth and dental decay
    4. Problems with phonation
        A. 1, 2, 3
        B. 1, 3
        C. 2, 4
        D. 4 only
        E. All of the above

23. A. 1, 2, 3

Inflammatory bowel disease (IBD) is a term given to both ulcerative colitis and Crohn's disease. Ulcerative colitis is a chronic inflammatory illness limited to the mucosa and submucosa of the colon and rectum. The risk for cancer is estimated to be 10–20% per decade after the first 10 years of disease. Crohn's disease is a transmural inflammation involving any and all portions of the GI tract. Management of IBD centers on nutritional support, immunosuppression, and surgery. Extraintestinal manifestations of these illnesses differ, but growth retardation and anemia, due to poor nutrition and GI blood loss, are regularly seen. Arthritis, arthralgias, and various skin manifestations, such as erythema nodosum, are also seen in both diseases.

24. B. 1, 3

The hypochromic, microcytic anemia and hypoalbuminemia seen in IBD are due to both iron losses through subtle GI bleeding and to poor nutrition. Infection of the GI tract is generally not part of the problem in IBD. Except in cases of dehydration, electrolytes are generally within normal limits.

25. E. All of the above

Recurrence patterns of this problem do not suggest a simple pattern of inheritance. Isolated cleft palate appears to be a separate entity from cleft lip with or without cleft palate. Isolated cleft palate has an incidence of 1:2500 live births. Cleft lip with cleft palate is more common that either is seen in isolation. The frequency is higher than 1:1000 in Native Americans, Japanese, and Chinese people and lower in African-Americans. Other anomalies are seen in up to 25% of all patients with cleft lip, palate, or both and more often in children with bilateral cleft lip. The Robin malformation sequence includes cleft palate as well as micrognathia and glossoptosis. Up to 20% of patients with the Robin sequence have cardiac anomalies such as ASD, VSD, or PDA.

26. E. All of the above

Cleft lip and palate often occur as isolated anomalies. Various techniques and equipment are available for feeding these infants, and no single solution is suitable for all. The management team for these infants should include a maxillofacial surgeon, audiologist, speech pathologist, otolaryngologist, exodontist, and geneticist.

27. Chiari type II malformations, also called Arnold-Chiari malformations:
    1. Are seen in nearly all patients with meningomyelocele
    2. May be asymptomatic
    3. Can be a cause of headache, particularly with coughing or straining
    4. Can be associated with vocal cord paralysis
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

28. Tests used to confirm the diagnosis of Chiari II malformation include:
    1. Sleep studies
    2. PET scan
    3. EEG
    4. MRI
    A. 1, 2, 3
    B. 1, 3
    C. 2, 4
    D. 4 only
    E. All of the above

27. E. All of the above

A Chiari II malformation is a bony defect that includes caudal displacement of the cerebellar vermis, fourth ventricle, and lower brainstem below the plane of the foramen magnum. Chiari II malformations are often asymptomatic. Presentation is often a headache, in particular after cough or with flexion/extension of the neck, lower cranial nerve signs, and, if the Chiari malformation has led to development of a syrinx, long tract signs such as lower extremity weakness.

28. D. 4

Chiari malformations are graded by the distance the CNS structures (cerebellar tonsils) extend below the foramen magnum on MRI, i.e., >6 mm in children <10 years old and >5 mm in older children. In an MRI study of adults, the prevalence of Chiari II using these diagnostic criteria was 0.5–1.0% and >70% of these patients were asymptomatic. A syrinx was noted in 30% of these subjects.