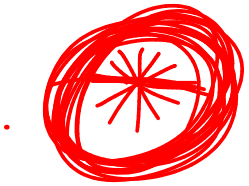
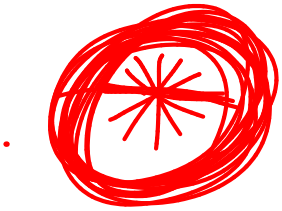
1. Polyhydramnios is frequently observed in all of the following conditions except:
A. Esophageal atresia.
B. Duodenal atresia.
C. Pyloric atresia.
D. Hirschsprung's disease.
E. Congenital diaphragmatic hernia.
Answer: D



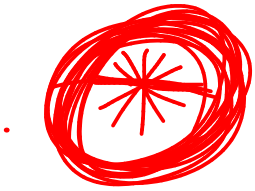
DISCUSSION: Polyhydramnios is defined as excessive amounts of fluid (>2000 ml.) in the amniotic sac during pregnancy. The amniotic pool is a dynamic pool with a relatively rapid turnover. In the fourth intrauterine month the fetus begins to swallow amniotic fluid (25% to 40% of the volume) and absorbs the fluid from the upper gastrointestinal tract. The fluid is urinated back out into the amniotic pool by the fetal kidneys and a functioning bladder. Although there are maternal causes of polyhydramnios (cardiac failure, renal failure, other causes of fluid retention) and some idiopathic cases, many instances are related to the presence of fetal anomalies. These include central nervous system problems such as anencephaly, which prevents normal swallowing, and any high alimentary tract obstruction that blocks the passage of the amniotic fluid and prevents its absorption (including esophageal atresia, pyloric atresia, and duodenal atresia). In addition, infants with congenital diaphragmatic hernia have obstructions due to herniation of the stomach and bowel into the thoracic cavity. This is a poor prognostic finding in these infants. Hirschsprung's disease is a form of low intestinal obstruction, and therefore an adequate length of proximal patent intestine is available for absorption of the swallowed amniotic fluid and polyhydramnios is usually not present.

2. Which of the following statements about Hirschsprung's disease is/are true?
A. There are no ganglion cells seen in Auerbach's plexus.
B. There is an increased incidence of Down syndrome.
C. It is more common in girls.
D. It may be associated with enterocolitis.
E. It may involve the small intestine.
Answer: ABDE



DISCUSSION: The affected segment of bowel in patients with Hirschsprung's disease has hypertrophic nerves in Auerbach's intermyenteric plexus, but no ganglion cells are present. Ganglion cells are also absent in Meissner's submucosal plexus. Some 3% to 5% of babies with Hirschsprung's disease also have Down syndrome. Hirschsprung's disease should be suspected in infants with Down syndrome that manifest evidence of abdominal distension and constipation. Hirschsprung's disease is much more common in boys (4:1). The enterocolitis of Hirschsprung's disease is a condition associated with delay in diagnosis, low bowel obstruction, severe abdominal distension, explosive diarrhea, and colonic mucosal ulceration. The course may be fulminant. This complication is associated with increased morbidity and mortality. Bacterial translocation and endotoxemia may complicate the condition. Treatment includes nasogastric suction, intravenous fluids, antibiotics, and rectal tube decompression of the obstructed rectosigmoid segment. In approximately 10% of cases aganglionosis extends into varying lengths of small bowel. In rare instances, the entire small bowel and colon may be aganglionic.

3. Which of the following statements is/are true of infants with gastroschisis?
A. It is associated with malrotation.
B. There is a high incidence of associated anomalies.
C. There is prolonged adynamic ileus following repair.
D. It is complicated by intestinal atresia in 10% to 12% of cases.
E. It is associated with chromosomal syndromes.
Answer: ACD



DISCUSSION: Because of intrauterine herniation of bowel to an extra-abdominal location, normal intestinal rotation and fixation do not occur. Most infants with gastroschisis have nonrotation. In contrast to infants with omphalocele, in which a high incidence of associated anomalies coexist, babies with gastroschisis have little else wrong. Following repair of the abdominal wall defect, infants with gastroschisis have a long delay in return of intestinal function. They usually require total parenteral nutrition to supply adequate caloric intake until gut function returns (3 to 4 weeks). Intestinal atresia is observed in 10% to 12% of neonates with gastroschisis. This is caused by bowel ischemia due to intrauterine volvulus or compression of the herniated viscera in a small, tight defect in the abdominal wall. Although infants with omphalocele frequently have chromosomal syndromes such as Beckwith syndrome or trisomy 13 to 15 or 16 to 18, babies with gastroschisis do not.

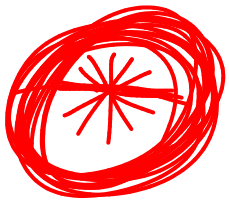
4. In neonates with congenital diaphragmatic hernia, which of the following statements is true?
A. The defect is more common on the right side.
B. Survival is significantly improved by administration of pulmonary vasodilators.
C. An oxygen index of 20 is an indication for extracorporeal membrane oxygenation (ECMO).
D. Oligohydramnios is a frequent occurrence.
E. Mortality is the result of pulmonary hypoplasia.
Answer: E

DISCUSSION: In infants with congenital diaphragmatic hernia the defect is more common on the left side (85%). Polyhydramnios is sometimes noticed and is a poor prognostic indicator of survival. Oligohydramnios is noted in fetuses with urinary tract obstruction and may be associated with pulmonary hypoplasia with an intact diaphragm. Although pulmonary vasodilators were used extensively in babies with congenital diaphragmatic hernia, they have not significantly improved survival. An oxygen index of greater than 40 is the usual indication for ECMO. Pulmonary hypoplasia is the main cause of mortality in babies with congenital diaphragmatic hernia.

5. Which of the following statements are true regarding the premature neonate?
A. A 15% to 20% right-to-left shunt occurs across the foramen ovale and patent ductus arteriosus.
B. Surfactant levels are normal after 30 weeks' gestation.
C. Fluid requirements are higher than in a full-term baby.
D. Rectal temperature is the best indicator of core body temperature.
E. They are more at risk for infection than the full-term infant.
Answer: ACE

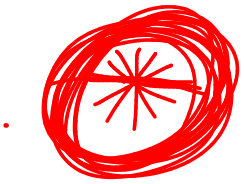
DISCUSSION: The newborn infant has a relatively elevated pulmonary artery pressure and shunts a significant amount of unoxygenated blood through the foramen ovale and patent ductus arteriosus. The normal PaO 2 below the ductus, as measured through an umbilical artery catheter, would be between 60 and 80 mm. Hg. Surfactant levels do not approach normal until after the 34th week of gestation, when enzyme levels in the surfactant pathway mature. Amniocentesis is performed to measure the lethicin-to-sphingomyelin ratio (L:S ratio) and determine whether maturation has occurred. Fluid requirements in the premature infant are between 140 and 150 ml. per kg. per day in comparison with those of the normal neonate in whom 80 ml. per kg. per day would be adequate. Increased insensible losses and the need for overhead warmers play a role in this increase. Axillary or skin probe temperature monitoring is more accurate than the rectal temperature in the neonate. The rectal temperature is not a good indicator of core body temperature until approximately 18 months of age. Premature infants lack immunoglobulin A (IgA) and have low levels of IgM, the C3b component of complement, and decreased opsonins. In addition, the leukocytes have reduced phagocytic ability, creating an increased risk of infection. Escherichia coli and beta-hemolytic streptococcus are the two most common infectious agents affecting the neonate.

6. In neonates with necrotizing enterocolitis, which of the following findings is an indication of significant bowel ischemia?
A. Increased gastric residuals.
B. Septic shock.
C. Cardiac failure due to a patent ductus arteriosus.
D. Elevated platelet count.
E. Erythema of the abdominal wall.
Answer: E



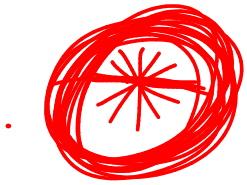
DISCUSSION: Necrotizing enterocolitis (NEC) is a condition that occurs in 2% of babies admitted to neonatal intensive care facilities. Increased gastric residuals can occur for a number of reasons and are seen as an early indicator of NEC, but they may not reflect the presence of ischemic bowel. Septic shock may be due to a wide variety of causes besides NEC. Cardiac failure due to patent ductus arteriosus may predispose to NEC but is not necessarily an indicator of ischemic bowel. Most babies with NEC have a progressive decrease in their platelet count in association with bowel ischemia. Erythema of the abdominal wall is an indication for surgical exploration and is consistent with NEC with perforation and inflammation of the peritoneum and abdominal wall.

7. The treatment of choice for neonates with uncomplicated meconium ileus is:
A. Observation.
B. Emergency laparotomy, bowel resection, and Bishop-Koop enterostomy.
C. Intravenous hydration and a gastrograffin enema.
D. Emergency laparotomy, bowel resection, and anastomosis.
E. Sweat chloride test and pancreatic enzyme therapy.
Answer: C



DISCUSSION: Meconium ileus is a form of intestinal obstruction that occurs in 10% to 15% of neonates with cystic fibrosis. The obstruction is related to intraluminal concretions of abnormal meconium. The treatment of choice is adequate hydration and evacuation with a hypertonic gastrograffin enema. The hyperosmolar contrast material causes an outpouring of fluids into the bowel lumen, which flushes out the obstructing meconium and negates the need for laparotomy. Observation alone is not a useful method of treatment. When gastrograffin evacuation fails, laparotomy, placement of a pursestring suture in the bowel wall, and intraluminal irrigation with saline and gastrograffin (administered through a catheter inserted through a small enterotomy within the pursestring) will often clear the obstructing meconium. This obviates the need for resection or enterostomy in most cases. Postoperatively, a sweat chloride test should be obtained to confirm the diagnosis of cystic fibrosis. Pancreatic enzyme should be given when diet is initiated.

8. The pentalogy of Cantrell includes all of the following except:
A. Epigastric omphalocele.
B. Sternal cleft.
C. Intracardiac defect.
D. Pericardial cyst.
E. Ectopia cordis.
Answer: D



DISCUSSION: The pentalogy of Cantrell includes an epigastric-located omphalocele, ectopia cordis, anterior pleuropericardial defect in the diaphragm, sternal cleft, intracardiac defect (most commonly a ventricular septal defect), and in approximately one third of the cases a diverticulum of the left ventricle. Pericardial cysts are not part of the pentalogy.

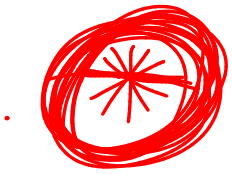
9. In infants with duodenal atresia all the following statements are true except:
A. There is an increased incidence of Down syndrome.
B. Duodenal atresia can be detected by prenatal ultrasound examination.
C. It may occur in infants with situs inversus, malrotation, annular pancreas, and anterior portal vein.
D. It is best treated by gastroenterostomy.
E. There is a high incidence of associated cardiac defects.
Answer: D

DISCUSSION: The diagnosis of duodenal atresia can be made prior to the infant's birth with a prenatal ultrasound examination. Infants with duodenal atresia are often premature and have a high incidence of associated anomalies, especially congenital heart disease. Duodenal atresia may also coexist in patients with annular pancreas, situs inversus, malrotation, and anterior portal vein. Approximately one third of the cases occur in babies with Down syndrome. The operative treatment of choice is a duodenoduodenostomy. Duodenojejunostomy is an alternative procedure. Gastrojejunostomy is not recommended.

10. The initial treatment of choice for a 2.5-kg. infant with a 20.0-cm. long proximal jejunal atresia and 8.0 cm. of distal ileum is:
A. Laparotomy, nasogastric suction, proximal dilatation to lengthen the atretic jejunum, total parenteral nutrition, and delayed anastomosis.
B. Laparotomy and proximal end-jejunostomy.
C. Laparotomy and immediate small bowel transplantation.
D. Laparotomy and double-barrel enterostomy (jejunum and ileum), with refeeding of jejunal contents into distal ileum and delayed anastomosis.
E. Laparotomy, tapering jejunoplasty, and end-to-oblique jejunoileal anastomosis.
Answer: E

DISCUSSION: The patient has short bowel syndrome with most of the bowel length involving the dilated proximal jejunal atresia. The treatment of choice is to perform a tapering jejunoplasty to preserve bowel length and construct an anastomosis. Early feedings are initiated when bowel function returns in order to stimulate bowel adaptation. Jejunal dilatation will not significantly lengthen the atretic jejunum and will not alter its abnormal motility. End-jejunostomy decompresses the obstruction but produces a high ostomy with excessive loss of succus entericus. A double-barrel enterostomy might allow refeeding of jejunal content into the distal ileum and colon, but the proximal atretic loop may have poor function. Small bowel transplantation is not a feasible alternative in the neonate at the present time.

11. A 2.8-kg. neonate with excessive salivation develops respiratory distress. Attempts to pass an orogastric catheter fail because the catheter coils in the back of the throat. A chest film is obtained and shows right upper lobe atelectasis and a gasless abdomen. The most likely diagnosis is:



A. Proximal esophageal atresia without a fistula.
B. Proximal esophageal atresia with a distal tracheoesophageal (TE) fistula.
C. “H-type” TE fistula.
D. Esophageal atresia with both proximal and distal TE fistula.
E. Congenital esophageal stricture.
Answer: A

DISCUSSION: Proximal esophageal atresia will result in excess salivation and aspiration of saliva. It is often associated with right upper lobe collapse. Infants with a distal TE fistula will have air in the stomach and intestine, as will babies with both proximal and distal fistulas, “H-type” fistula, and an esophageal stricture. Infants with proximal atresia but without a TE fistula will not have air beneath the diaphragm. Attempts at passing an orogastric catheter will be met by an obstruction and coiling of the catheter in the infant's mouth. The catheter will pass into the stomach in infants with “H-type” TE fistula and in most with esophageal stricture.

12. Neonates with NEC may demonstrate all of the following findings on abdominal films except:

A. Pneumatosis intestinalis.
B. Portal vein air.
C. Pneumoperitoneum.
D. Colovesical fistula.
E. Fixed and thickened bowel loops.
Answer: D

DISCUSSION: Infants with NEC do not develop a colovesical fistula as an initial x-ray finding. Pneumatosis, portal vein air, pneumoperitoneum, and fixed intestinal loops with thickened bowel wall are all observed with some regularity in babies with NEC.

13. The most common type of congenital diaphragmatic hernia is caused by:
A. A defect in the central tendon.
B. Eventration of the diaphragm in the fetus.
C. A defect through the space of Larrey.
D. An abnormally wide esophageal hiatus.
E. A defect through the pleuroperitoneal fold.
Answer: E

DISCUSSION: Eventration of the diaphragm is related to phrenic nerve paralysis. It is more commonly observed after a breech delivery and may be associated with torticollis and Erb's palsy. The space of Larrey is located anteriorly just off the midline. A Morgagni hernia passes through this potential space. An abnormally wide esophageal hiatus would most likely create a sliding hiatal hernia. The most common type of congenital diaphragmatic hernia in the neonate is the posterolateral Bochdalek hernia, which passes through a defect in the developing pleuroperitoneal fold.

14. The calorie-nitrogen ratio for an infant should be maintained at:
A. 75:1.

B. 100:1.
C. 50:1.
D. 150:1.
E. 25:1.
Answer: D

DISCUSSION: The calorie-nitrogen ratio should be maintained at 150:1 for most infants. Fever, major illness, sepsis, or trauma may increase the caloric requirements significantly.

15. All of the following conditions are derived from the primitive embryonic foregut except:
A. Bronchogenic cyst.
B. Cystic adenomatoid malformation.
C. Gastric duplication.
D. Mesenteric cyst.
E. Pulmonary sequestration.
Answer: D

DISCUSSION: Mesenteric cysts are derived from lymphatic anlage in the abdomen and are unassociated with foregut development. The lung buds arise from the primitive foregut and anomalies associated with tracheopulmonary development are therefore all derived from foregut. These include tracheoesophageal fistula, congenital lobar emphysema, enteric cysts (which may communicate to the normal esophagus, lung, or spinal canal; e.g., neurenteric cyst), cystic adenomatoid malformations, solitary lung cysts, intra- and extralobar sequestrations, and bronchogenic cysts. The stomach and first part of the duodenum are also of foregut origin, and thus a gastric duplication is by definition derived from foregut.

16. For a 22-kg infant, the maintenance daily fluid requirement is approximately which of the following?

a. 1100 ml
b. 1250 ml
c. 1550 ml
d. 1700 ml
e. 1850 ml
Answer: c

Maintenance water and electrolyte requirements are illustrated in the table indicated below. The volume calculation is demonstrated. The composition of the intravenous fluids is generally that of D5 1/4 normal saline or D5 1/2 normal saline with 10 mEq/l of KCL.

17. Which of the following statements regarding nutritional requirements in infants are true?

a. The total daily water requirement is estimated to be 100 ml/100 kcal ingested
b. The resting energy expenditure is approximately twice that of an adult
c. The highest rate of nitrogen retention with parenteral nutrition occurs in infants given approximately 40% of the calories as carbohydrate and the remainder as fat
d. The protein requirement for a newborn infant is approximately 2.5 g/kg/day
Answer: a, b, c, d

Taking all factors into account, total daily water requirements for a term infant are estimated to be 100 ml/100 kcal ingested, assuming an insensible loss of 50 ml/kg/day and a growth requirement of approximately of 15 ml/kg/day. The energy expenditure of normal neonates is approximately twice that of normal adults (50 kcal/kg/day versus 25 kcal/kg/day).
In most circumstances, high carbohydrate/low fat ratios in parenteral nutrition result in high rates of energy expenditure and decreased nitrogen retention, while low carbohydrate/high fat ratios result in excessive fat deposition. A balanced ratio (approximately 40% carbohydrate) provides the highest rate of nitrogen retention, and is consistent with the proportion of carbohydrate found in breast milk and with the estimates of minimal carbohydrate needs determined by isotope infusion studies. A consensus statement by the World Health Organization and the United Nations University estimates the protein

requirement at 2.5 g/kg/day for an infant and 1.25 g/kg/day for a one-year-old child. For preterm infants, the protein need ranges from 2.5 to 3.9 g/kg/day if the weight is less than 2.5 kg.

18. A term infant 48 hours of age suddenly develops hypoxemia, irritability, and glucose and temperature instability. Which of the following statements are true?

a. Empiric antibiotic coverage for b-hemolytic Streptococci and Escherichia coli should be initiated
b. An intravenous infusion of prostaglandin E1 should be initiated immediately
c. Exogenous surfactant should be given immediately
d. The mortality rate for this child is approximately 50%

Answer: a,d

This infant has the classical findings of neonatal sepsis. This is defined as a generalized bacterial infection accompanied by a positive blood culture during the first month of life. Early onset sepsis occurs during the first week of life, and is due primarily to maternal organisms, such as b-hemolytic Streptococci, Escherichia coli or Listeria monocytogenes. The mortality rate of early onset sepsis is approximately 50 percent. Late onset sepsis is due primarily to hospital acquired organisms such as Staphylococcus epidermidis, Staphylococcus aureus or Pseudomonas species, and the mortality rate for this entity is approximately 20 percent.
The signs and symptoms of neonatal sepsis are subtle and nonspecific. Early signs include lethargy, irritability, temperature instability, change in the respiratory pattern, or changes in the feeding pattern. Hematologic findings include thrombocytopenia, leukocytosis, or leukopenia. Hemodynamic manifestations occur late. Presumptive therapy should be based upon the suspected organism, but often includes Ampicillin or an anti-Staphylococcal agent plus an amino glycoside.
A prostaglandin E1 infusion is inappropriate as this relates to patients with ductal-dependent congenital heart disease. Exogenous surfactant is unlikely to be helpful in a full-term infant who has previously been well and can be expected to begin his illness with a normal complement of pulmonary surfactant.

19. Which of the following statements about pulmonary surfactant are true?

a. Endogenous surfactant deficiency is the key physiologic problem in preterm infants with the infant respiratory distress syndrome
b. Surfactant function can be restored to normal using aerosolized phosphatidylcholine administration
c. Exogenous surfactant replacement has been shown to reduce mortality in preterm infants with the infant respiratory distress syndrome
d. Surfactant is produced by Type I alveolar epithelial cells

Answer: a, c

The pulmonary surfactant complex lowers surface tension, stabilizing the alveolus even at low lung volumes. It is a complex material secreted by Type II alveolar epithelial cells. It is composed of 80 to 90 percent phospholipid (primarily phosphatidylcholine) and unique surfactant-associated proteins (10 percent). Surfactant proteins appear to play a critical role in the organization of the phospholipid molecules, and modify the surface-active properties of the lipids. Phospholipid synthesis and the expression of surfactant proteins increase with advancing gestational age. Amniotic fluid surfactant concentrations have been used for many years to predict pulmonary maturity. Surfactant deficiency is the primary factor in the pathophysiology of the neonatal respiratory distress syndrome. The use of exogenous surfactant replacement therapy is currently under investigation for the treatment of neonatal respiratory distress and has been shown to reduce mortality in a variety of specific circumstances involving preterm infants. Several commercially available preparations are available and are undergoing clinical investigation.

20. Which of the following statements regarding premature infants are true?

a. Complications of prematurity account for approximately 85% of fetal deaths
b. Prematurity is defined by the World Health Organization as birth prior to 35 weeks gestation
c. Infants with intrauterine growth retardation have physiologic problems which are more dependent on the birth weight than the gestational age
d. Preterm infants are at increased risk for hypocalcemia and hypoglycemia when compared to term infants

Answer: a, d

Prematurity is defined by the World Health Organization as a gestational age at birth of less than 37 weeks. Complications of prematurity account for approximately 85% of fetal deaths. These deaths are commonly due to perinatal asphyxia, respiratory failure and infection. The term intrauterine growth retardation describes a pathophysiologic process that results in restriction of fetal growth. Fetal, placental or maternal abnormalities are common. These infants are a heterogeneous population and they tend to have neonatal problems related more to their gestational age than to their birth weight. These problems include asphyxia, hypoglycemia, hypothermia, hypocalcemia, pulmonary hemorrhage, necrotizing enterocolitis and other complications related to specific syndromes or congenital anomalies.

21. Other than the history and physical exam, which of the following tests is considered an essential feature of the preoperative evaluation of a patient with a suspected thyroglossal duct cyst?

a. Cervical ultrasound
b. Thyroid scan
c. Serum T3 and T4 levels
d. Needle aspiration
e. None of the above

Answer: e

A thyroglossal duct cyst is typically a midline structure connected to the foramen cecum at the base of the tongue, that is pulled proximal and superior as the tongue protrudes. It may be superior or inferior to the hyoid and is occasionally slightly off the midline. Because the thyroglossal cyst may rarely contain the patient's only thyroid tissue, some have recommended a technetium-99m radioisotope thyroid scan before excision. However, excision of the cyst is indicated regardless, because infection of the cyst is likely, and the dysgenetic thyroid tissue in the cyst has malignant potential. For these reasons, patients with suspected thyroglossal duct cysts require routine surgical excision. Preoperative ultrasound, thyroid scan, T3 and T4 levels, or needle aspiration is not necessary. For those patients who have thyroid tissue in their cysts by pathologic examination, postoperative thyroid function tests identify those who have no remaining thyroid tissue and replacement therapy can be prescribed.

22. Suppurative cervical lymphadenitis in a 3-year-old child is commonly related to which of the following organisms?

a. Staphylococcus aureus
b. Atypical mycobacterial organisms
c. Streptococcal organisms
d. Lymphoma with secondary pyogenic organisms
e. Cat scratch

Answer: a, c

Acute suppurative lymphadenitis related to bacterial pathogens is generally straightforward to diagnose. There is often accompanying infectious illness. The lymph nodes enlarge rapidly, are tender and erythema of the overlying skin is present. Fever and an elevated white blood count with a left shift are usually present. Fluctuant nodes may be aspirated. Streptococcus and Staphylococcus aureus are the most common organisms and the initial course of antibiotic therapy is directed to these organisms. If the adenopathy fails to resolve in 2 to 3 weeks, then the patient should likely undergo an excisional biopsy. Atypical Mycobacteria, cat scratch disease, and Mycobacterium tuberculosis are more uncommon than bacterial lymphadenitis and do not typically produce tenderness or systemic signs. Lymphoma with secondary pyogenic infection is similarly uncommon.

23. Branchial cleft remnants most often present with which of the following clinical problems?

a. Infection
b. Airway obstruction
c. Hemorrhage
d. Malignant degeneration
e. Pain

Answer: a

The more common second branchial cleft anomalies present typically as a pinpoint opening on the anterior border of the sternocleidomastoid muscle. Attention is drawn to the defect often by the appearance of small drops of clear fluid at the orifice or by occurrence of infection in the tract itself. Less frequently, a mass may present anterior to the upper portion of the sternocleidomastoid muscle representing a cyst derived from this tract. Infection is a more common problem in the older age group. Airway obstruction and hemorrhage are rare presentations. Pain is usually secondary to infection and malignant degeneration is reported but exceedingly rare. Treatment consists of surgical excision. If infection is present, a course of antibiotics is administered first.

24. Proximity to which of the following structures places it at risk during surgical excision of a second branchial cleft remnant?

a. Internal carotid artery
b. External carotid artery
c. Hypoglossal nerve
d. All of the above
e. None of the above

Answer: d

Operative excision of a second branchial cleft remnant begins with an elliptical transverse incision at the sinus opening and cephalad dissection of the tract to its furthest extent, generally reaching the floor of the tonsillar pillar. The dissection is kept directly on the tract to avoid injury to contiguous structures such as the internal jugular vein, the internal or external carotid arteries (between which it passes), and the hypoglossal nerve. The operation can almost always be carried out through a single cosmetic incision if the tract is kept under traction and digital pressure is exerted through the tonsillar fossa. Dissection of the sinus tract may be facilitated by passing a fine silver probe or piece of heavy nylon suture through the length of the tract.

25. Standard therapy for acute epiglottitis in a child is:

a. Tracheostomy
b. Intravenous antibiotic treatment in an ICU setting
c. Endotracheal intubation in the operating room and intravenous antibiotic therapy
d. Indirect laryngoscopy and intravenous antibiotics
e. Intravenous steroids and antibiotics

Answer: c

Acute epiglottitis is a common cause of acquired airway obstruction in the pediatric age group. Haemophilus influenzae B is nearly always the causative organism, and most children are toxic at presentation with an elevated temperature and an increased pulse and respiratory rate. Prolonged inspiratory stridor that worsens in the supine position is characteristic. The child usually sits erect, anxious and drooling and becomes increasingly exhausted with air hunger. No attempt should be made to visualize the larynx outside of the operating room for fear of sudden airway occlusion. The standard therapy is short-term endotracheal intubation performed in the operating room with general anesthesia. The inflammatory process resolves rapidly with intravenous antibiotics and intubation is seldom required beyond 3 days. In the past, tracheostomy was the standard therapy, but comparative reviews demonstrate that short-term endotracheal intubation is associated with less morbidity and fewer complications.

26. Which of the following statements regarding congenital diaphragmatic hernia are true?

a. The incidence of right and left-sided lesions is equal
b. Malrotation is to be expected
c. Left-to-right shunting via a patent ductus arteriosus is a serious but expected physiologic consequence of pulmonary hypoplasia
d. Survival rates of 75% are reported in several contemporary series
e. Congenital heart disease is present in approximately 20% of these infants

Answer: b, d, e

During organogenesis closure of the right hemidiaphragm normally precedes that of the left. This asynchronous closure and the presence of the liver on the right account for the finding that 85% to 90% of congenital diaphragmatic hernias involve the left hemidiaphragm. Malrotation is an expected finding with diaphragmatic hernia because intestinal herniation into the thorax normally precedes the fixation of the gut to the posterior body wall. Pulmonary hypertension is a major feature of congenital diaphragmatic hernia and right-to-left shunting via a patent ductus arteriosus is present in virtually all of these children. Fifteen to 25% of infants with a diaphragmatic hernia have an associated anomaly, the most important being cardiovascular abnormalities. Although ventricular septal defects and aortic coarctations are most common, virtually all cardiac and great vessel anomalies have been reported. Cardiac ECHO screening examinations are therefore routine. Survival rates as high as 75% to 90% in selected high-risk congenital diaphragmatic hernia patients have been reported in several large clinical series over the last five to ten years. This is an apparent improvement from the historic range of 50%.

27. Of the following cystic malformations of the tracheobronchial tree, which is most likely to be asymptomatic when discovered?

    a. Intralobar pulmonary sequestration
    b. Extralobar pulmonary sequestration
    c. Congenital cystic adenomatoid malformation
    d. Congenital lobar emphysema

Answer: b

Intralobar pulmonary sequestration and cystic adenomatoid malformations typically present with either neonatal respiratory distress or infection related to inadequate clearance of secretions. Given enough time, nearly all of these lesions will become infected. Congenital lobar emphysema is characterized by air-trapping within an otherwise normal lung. This typically presents with respiratory distress which ranges from mild to life-threatening. Hemodynamic instability requiring emergency thoracotomy is occasionally present.
Extralobar sequestration is typically a mass of disorganized pulmonary parenchymal tissue within its own investing pleura and outside of the normal lung parenchyma. This does not communicate with the normal tracheobronchial tree. Infection is rare and although hemorrhage, arterial venous shunting, mediastinal compression and occasional malignancy may occur, these lesions are typically asymptomatic and indeed often discovered via prenatal ultrasound. Excision is recommended for each of these lesions, typically involving a lobectomy.

28. Infants with a double aortic arch most commonly present with which of the following problems?

    a. Dysphagia
    b. High output cardiac failure related to a patent ductus arteriosus
    c. Positional hyperemia and edema of the right upper extremity
    d. Symptomatic tracheal compression

Answer: d

The double aortic arch represents the most common type of complete vascular ring. It arises from the ascending aorta and bifurcates, with one arch passing on either side of the trachea and the esophagus. The symptoms of complete vascular rings are due to compression of the trachea, the esophagus or both. The child with a double aortic arch is generally the most symptomatic and most patients have symptoms in infancy. The typical clinical picture is one of symptomatic tracheal compression and includes inspiratory wheezing, coughing, noisy breathing, shortness of breath, stridor and frequent bouts of pneumonia. Feeding problems may become apparent when solid foods are started but this is less common than tracheal compression. The most important screening test is the barium swallow, and CT or MRI are definitive. Angiography and endoscopy are not usually helpful. Any patient who is symptomatic from a vascular ring should be treated surgically.

29. Which of the following is most common after primary esophagostomy for esophageal atresia with a distal tracheoesophageal fistula?

    a. Anastomotic leak
    b. Esophageal stricture
    c. Recurrent tracheoesophageal fistula

d. Gastroesophageal reflux
e. Tracheomalacia requiring aortopexy

Answer: d

The preferred approach for esophageal atresia with a distal tracheoesophageal fistula in a patient without other problems is an extrapleural right thoracotomy with division of the tracheoesophageal fistula and primary esophagostomy. No gastrostomy is ordinarily used. The results with this approach are better than those with a staged approach. Three major complications are related to the esophageal anastomosis: leak, stricture and recurrent fistula. The incidence of leak varies from 10% to 20% depending on the type of anastomosis done and the degree of tension. A distinct advantage of an extrapleural anastomosis is the predictable resolution of these leaks if adequately drained. Similarly, the stricture rate varies between 10% and 25%, again depending on the type of anastomosis done. Many infants require one or two dilations, but few have significant long-term problems. The incidence of recurrent esophageal fistula is difficult to determine since few authors emphasize this technical problem, but it appears to be about 10% in most reports.
Gastroesophageal reflux secondary to dysmotility of the distal esophagus is a significant problem and occurs to some degree in virtually all of these patients. A significant number of these infants, 25% to 30% or more, are refractory to medical therapy and require surgical fundoplication. Tracheomalacia is a complication of the malformation, not of the repair. This appears to result from inadequate cartilagenous tracheal rings at the level of the fistula. The reported incidence is up to 25% in some recent series. Most infants with tracheomalacia improve with growth and time, but a small percentage develop severe respiratory difficulty which requires surgical aortopexy.

30. Which of the following is the most common primary lung tumor in infants and children?

a. Pulmonary blastoma
b. Squamous cell carcinoma
c. Endobronchial carcinoid
d. Leiomyoma
e. Metastatic osteogenic sarcoma

Answer: c

Bronchial adenomas are the most common primary lung tumors in childhood. Nevertheless, they are quite rare. Typically these are low-grade adenocarcinomas that include endobronchial carcinoids, cylindromas, mucoepidermoid tumors, and bronchomucous gland adenomas. Carcinoid tumors are the most common bronchial adenomas and represent over 80% of the total.
Although common in adults, bronchogenic carcinoma of the lung is extremely rare in childhood. A review of the world's literature in 1983 revealed only 47 such patients. Pulmonary blastoma is a rare malignant lung tumor composed of cells that resemble fetal lung. Its incidence is actually highest in adults, although it is reported in children as well. Benign tumors of the lung are also rare in childhood. The most common of these are pulmonary hamartomas or chondromas. Others include leiomyoma, leiomyoblastoma and mucus gland adenoma. Metastatic osteogenic sarcoma is more common than any primary lung tumor, but is by definition a secondary metastatic lesion and therefore not the correct answer to the question posed here.

31. Which of the following statements regarding congenital chest wall deformities are true?

a. Children with pectus excavatum deformities typically have physiologically insignificant limitation of exercise tolerance
b. The rate of recurrence after operative repair of a pectus excavatum deformity is between 5% and 10%
c. Pectus carinatum is the most common congenital chest wall defect
d. The most common indication for operative repair of congenital chest wall deformities is cosmesis

Answer: a, d

The most common congenital chest wall deformity is pectus excavatum, representing approximately 90% of the total. Approximately 5% to 7% of the lesions are pectus carinatum and a variety comprise the remainder. Most children with pectus excavatum are asymptomatic. There have been numerous efforts to document associated cardiac and pulmonary abnormalities. Objective data show that although there are minor demonstrable cardiopulmonary abnormalities demonstrable, these do not appear to be significantly improved by surgery and they are generally insignificant physiologically. As a result, the indications for repair of chest wall deformities are essentially cosmetic and psychological.

The importance of these however should not be minimized in a largely adolescent population. The repair is technically straightforward but involves moderate morbidity. The long-term results are excellent. Recurrence is rare (approximately 1%) in most institutions with large experiences.

32. The definitive evaluation of a child with a suspected congenital cystic abnormality of the tracheobronchial tree is best done using which of the following?

a. Rigid bronchoscopy
b. Computerized tomography or magnetic resonance imaging
c. Chest x-ray
d. Angiography
e. Barium esophagogram

Answer: b

Plain film radiography is the first imaging study performed and remains a cornerstone for the diagnosis and follow-up of this group of lesions. The use of additional imaging provides definitive diagnosis and allows planning for the surgical approach as well. Computed tomography (CT) and magnetic resonance imaging (MRI) can separate cystic from solid components in a radiopaque lung mass. These are the most definitive diagnostic studies available. The MRI has reconstructive capabilities that obviate the need for angiography. Intravenous contrast with CT scan provides similar anatomic information. Ultrasonography is less costly, more readily performed and in select cases may be as sensitive.
Angiography is not employed regularly because these alternative imaging strategies provide similar information at lower cost with less morbidity. A barium esophagogram is helpful in the diagnosis of children with dysphagia but that is a rare presentation for these lesions. Bronchoscopy is rarely helpful for these lesions and in these infants and small children carries the risk of general anesthesia and positive pressure ventilation. In children with congenital lobar emphysema and cystic adenomatoid malformation, hyperinflation following positive pressure may induce mediastinal compression and create a surgical emergency. For these reasons, CT or MRI are considered the best definitive diagnostic imaging choices after the initial chest x-ray.

33. A newborn infant develops coughing, choking and cyanosis with his first feeding. He is noted to have excessive drooling. What are the important associated anomalies that must be screened for prior to surgical intervention?

a. Right-sided aortic arch
b. Hydrocephalus
c. Genitourinary obstruction
d. Congenital heart disease

Answer: c

This child has a classical history for esophageal atresia and has a very high (90% or more) probability of a distal tracheoesophageal fistula. The simplest way to establish the diagnosis is to attempt to pass a catheter through the mouth or nose into the stomach. If the tube encounters obstruction, a plain radiograph should document the atresia.
Patients with esophageal atresia and tracheoesophageal fistula frequently have associated anomalies. This incidence is about 30% to 50% in most reports. Anomalies vary from minor skeletal deformities to uncorrectable cardiac defects. The most common associated anomalies are cardiac and gastrointestinal, especially imperforate anus (10%). Vertebral, genitourinary and limb anomalies are also seen. Importantly, approximately 5% of patients with esophageal atresia have a right-sided aortic arch. This is an important technical issue as the normal approach is via a right thoracotomy and this should be changed to a left thoracotomy in the presence of this finding. There is no association with hydrocephalus.

34. Which of the following ventilation strategies is the best initial approach for a neonate with a left congenital diaphragmatic hernia and the following post ductal arterial blood gases: PaO2 50 mm Hg, PaCO2 60 mm Hg, pH 7.35?

a. High-frequency jet ventilation
b. Permissive hypercapnia with convential pressure controlled ventilation
c. Extracorporeal membrane oxygenation (ECMO)
d. Induced respiratory alkalosis
e. Inhaled nitric oxide with conventional volume controlled ventilation

Answer: b

Contemporary congenital diaphragmatic hernia management emphasizes permissive hypercapnia using any necessary mode of respiratory support. Ordinarily, pressure controlled ventilation is the initial mode of support. The purpose is to reduce the iatrogenic lung injury associated with high-pressure mechanical ventilation. It is this latter problem, barotrauma-induced lung injury, which has reduced enthusiasm for induced respiratory alkalosis. Although alkalosis can sometimes be achieved, the price is often prohibitive mean and peak airway pressures. High-frequency jet ventilation, ECMO and inhaled nitric oxide are all evolving strategies which must be considered developmental and are reserved for use after the initial strategy is unsuccessful.

35. There is an emerging consensus that the surgical repair for congenital diaphragmatic hernia is best done:

a. Emergently at the bedside, eliminating the risks of transporting an unstable neonate
b. While on extracorporeal membrane oxygenation
c. When the infant is potentially extubatable
d. Within the first 48 to 72 hours of life

Answer: c

Infants with diaphragmatic hernias have considerable variation in the degree of respiratory distress, and the degree of distress dictates timing of repair. Traditionally, infants were rushed emergently to the operating room for reduction of the herniated viscera and diaphragmatic closure. Because effective preoperative decompression of the intestine is usually possible and because it has become clear that the underlying pulmonary hypoplasia is not reversed by emergency operation, this sense of urgency is no longer accepted. Most surgeons now commit themselves to a period of preoperative stabilization which is used to confirm the diagnosis and optimize medical care. The current recommendation is that operative repair be undertaken in a stable patient who is nearing extubatable levels of ventilatory support, regardless of the means of support which have been employed. This means that repair is done at or after the end of cardiopulmonary bypass in infants where that is necessary. The results with a delayed approach appear at least equivalent and in several series better than with emergent or early repair. The concept is that the deferral of the iatrogenic operative injury to a time when the neonatal pulmonary vascular bed is less vulnerable to vasospasm is desirable.

36. Meckel's diverticulum may present with which of the following signs or symptoms?

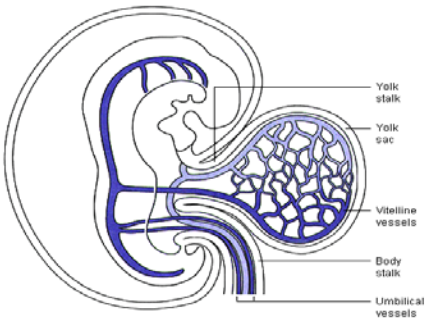
a. Hemorrhage
b. Intussusception
c. Volvulus
d. Patent omphalomesenteric duct
e. Right lower quadrant peritoneal findings

Answer: a, b, c, d, e

The most frequent congenital anomaly of the GI tract is a Meckel's diverticulum. The incidence is about 2% of the general population, many of whom remain asymptomatic throughout life. Estimations of the frequency with which symptoms develop among people with Meckel's diverticula vary from 4% to 30%, but is clear that the risk diminishes substantially with age. About half of those who become symptomatic are under the age of 2. Hemorrhage, acute diverticulitis, perforation, and small bowel obstruction or intussusception are all classic presenting scenarios for a child with a Meckel diverticulum. (Table) The approximate frequency of these presentations is shown above. Below are illustrations of the various anatomic abnormalities with their associated clinical presentations. (Figure) These presentations include Meckel's diverticulitis which is virtually indistinguishable from acute appendicitis.

SIGNS AND SYMPTOMS OF MECKEL'S DIVERTICULUM

Clinical Presentation	**Approximate Frequency (%)**
Hemorrhage	30–35
Small bowel obstruction	30–35
Diverticulitis	20–25
Umbilical fistula	10
Other	Uncommon



Normal embryologic relations of the embryonic yolk sac, yolk stalk, and gut.

37. A 3-week old infant has a barium upper gastrointestinal series to evaluate vomiting. The duodenojejunal flexure is found to be to the right of the midline as well as more caudal and anterior than a normal ligament of Treitz. The child is seen to reflux barium spontaneously to the level of the mid-thoracic esophagus. You would recommend which of the following?

a. Barium enema
b. Emergency laparotomy
c. A trial of H2, blockade and cisapride therapy
d. Upper gastrointestinal endoscopy
e. Overnight pH probe analysis

Answer: b

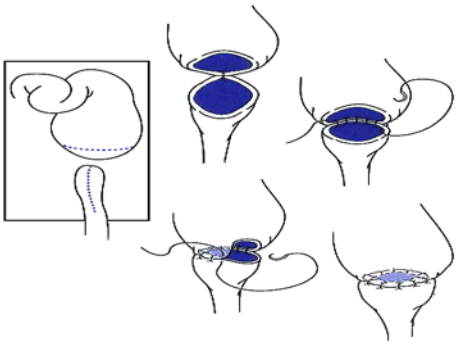
This child has malrotation and is at risk for midgut volvulus based on the imaging findings. Typically, malrotation produces an incomplete obstruction of the duodenum with a corkscrew or coiled appearance in the third or fourth portions of the duodenum. Malposition of the duodenojejunal junction is diagnostic. In particular, this includes a location to the right of the midline. Additionally, failure to achieve normal posterior and cephalad fixation is typical. The small bowel resides in the right abdomen and the colon and cecum are on the left. Attempts to radiographically differentiate malrotation with or without volvulus are unreliable and therefore hazardous. This child has gastroesophageal reflux which is likely secondary to the partial duodenal obstruction from malrotation. None of the alternatives other than emergency laparotomy are appropriate. There is no role for nonoperative management of malrotation in the neonate. Assessment, resuscitation and preoperative preparation should be conducted concurrently as the child is prepared for laparotomy. This urgency is because a delay measured in hours may represent the difference between a viable or infarcted midgut at laparotomy. Fifty to 75% of malrotations are discovered in the first month of life and about 90% occur in children less than one year of age.

38. Which of the following statements regarding duodenal atresia are true?

a. 20% to 40% of these infants have Trisomy 21
b. When associated with an annular pancreas, division of the pancreas at the site of obstruction is curative
c. Bilious vomiting is typical because the obstruction is usually distal to the ampulla of Vater
d. Reconstruction is best achieved with Roux-en-Y duodenojejunostomy

Answer: a, c

Twenty to 40% of infants with congenital duodenal obstruction have Trisomy 21. Because this syndrome is not always apparent during the physical examination, a routine karyotype should be obtained. A preoperative ECHO ultrasound examination is also appropriate to evaluate the possibility of associated congenital heart disease. Feeding intolerance and bilious vomiting in the first 24 to 48 hours of life are characteristic. The malformations are typically distal to the ampulla of Vater and thus the infants present with bilious vomiting and proximal duodenal and gastric distention. Malformations proximal to the ampulla of Vater result in nonbilious vomiting and this possibility must not be ignored.
Generally, bypass of the obstructing lesion is the best approach regardless of whether an atresia or annular pancreas is responsible. Division of an annular pancreatic band is inappropriate for two reasons: 1) The duodenum is virtually always atretic in addition. 2) Division of this pancreatic tissue necessarily divides the accessory pancreatic duct within it creating the real possibility of a pancreatic fistula. Generally, the construction of a duodenostomy that minimizes the length of defunctionalized duodenum is preferred. The procedure generally employed is a diamond-shaped duodenostomy. Simple duodenojejunostomy without a Roux-en-Y is occasionally necessary for lesions in the distal duodenum.



Diamond-shaped duodenoduodenostomy.

39. A 1500-gram, 30-week gestation neonate is fed at 2 weeks of age. He develops abdominal distention, bilious vomiting and guaiac-positive stool. A plain film of the abdomen demonstrates pneumotosis intestinalis. Which of the following related statements are true?

a. An emergency barium upper GI series should be done to rule out malrotation
b. The child should have a nasogastric tube placed, broad spectrum intravenous antibiotics begun, and sequential abdominal films obtained.
c. The likelihood of intestinal perforation is in excess of 50%
d. The expected survival rate is in the range of 70%

Answer: b, d

This clinical history is classical for neonatal necrotizing enterocolitis (NEC), an idiopathic clinical condition characterized by mucosal intestinal injury that may progress to transmural bowel necrosis. Typically it occurs in critically ill, preterm infants and is characterized by abdominal distention, bilious vomiting and either occult or gross blood in the stool. In this setting, pneumotosis intestinalis is pathognomonic. When the diagnosis is suspected on clinical grounds and the plain film, no contrast imaging should be done as this may complicate or contribute to the problems of perforation. In particular, the child presented here has an unequivocal history and an upper GI series would be inappropriate.
Half of all infants with NEC have birth weights less than 1500 g, and 80% of these infants weigh less than 2500 g at birth. The incidence is approximately 1 to 2 in 1000 live births. It is the most common surgical emergency in neonates in North America. The initial management of infants with NEC consists of nasogastric decompression; broad-spectrum antibiotic administration; and correction of hypoxemia, hypotension, acidosis, fluid or electrolyte disorders, and other reversible medical problems. Up to 90% of infants with NEC can be managed successfully nonoperatively, but this is widely variable among institutions because of differences in referral and practice patterns. Intestinal perforation is characterized by pneumoperitoneum and is an indication for operation. Although the incidence of perforation is variable, it is generally less than 20% to 40%.
The overall survival rate for neonates with NEC is about 60% to 70% for both operative and nonoperative management groups. This represents a substantial improvement from the 20% to 30% survival probability when the entity was first recognized 30 to 40 years ago.

40. The most common cause of pyogenic liver abscess in children today is which of the following?

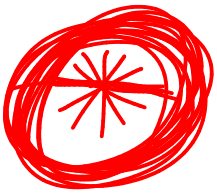
a. Perforated appendicitis
b. Blunt liver injury
c. Immunocompromised host
d. Percutaneous liver biopsy
e. Omphalitis

Answer: c

In the preantibiotic era, pyogenic hepatic abscesses occurred most frequently after perforated appendicitis. This complication is rarely seen today. Chronic granulomatous disease of childhood is a principle condition associated with hepatic abscess. This disease is the result of deficient oxidant-mediated bacterial killing by circulating granulocytes. In the pediatric age group, 40% of pyogenic liver abscesses occur in children with chronic granulomatous disease, and another

30% occur in children with other immunodeficiencies, most commonly leukemia. Other rare causes of liver abscesses in pediatric patients are umbilical vein catheter-induced infection, omphalitis and other biliary disease. Pyogenic liver abscesses following blunt liver injury or percutaneous liver biopsy are distinctly rare events.

41. Which of the followings statements regarding an infant with meconium ileus are true?



a. The probability is 100% that he will have cystic fibrosis
b. Nonoperative therapy resolves this problem in approximately two-thirds of patients
c. The average life expectancy is approximately 26 to 28 years for this infant at present
d. The finding illustrated below on plain film is an absolute operative indication (Figure 103-23A)

Answer: a, b, c, d

Meconium ileus refers to the characteristic obstruction of the small intestine in neonates with cystic fibrosis (CF). Some 10% to 20% of infants with CF present initially with meconium ileus. All infants with meconium ileus have cystic fibrosis. CF is characterized by a transport defect of epithelium that results in impermeability of the chloride ion and therefore water. Inspissated secretions in the pancreas and gut lead to obturator obstruction of the terminal ileum from meconium in the neonate. Approximately two-thirds of these infants have simple meconium ileus, the remainder have complications such as proximal volvulus, perforation or atresia. These latter problems may be associated with the development of a meconium cyst. In this instance, speckled calcifications on plain radiograph or ultrasound are diagnostic. The film above is that of a meconium pseudocyst consistent with intraperitoneal spillage of meconium from intestinal perforation. This finding requires surgical exploration.
Sixty to 70% of infants with simple meconium ileus can be treated by enema installation of one several irrigation solutions into the obstructed terminal ileum. Saline, hyperosmolar contrast agents, dilute N-acetylcysteine and a variety of other solutions have been used successfully. Following resolution of the obstruction, most institutions now report survival rates as high as 70% to 100%.
The average life expectancy for CF patients is now well into the third decade of life. It is primarily determined by the course of the pulmonary disease rather than GI problems. A number of important medical advances, including the realistic prospect of gene therapy are foreseeable for these infants.

42. You are asked to recommend therapy for an asymptomatic 2 year old who swallowed a small alkaline watch battery 4 hours ago. A plain film shows the intact battery in the intestine beyond the stomach. The best course of therapy is?

a. Immediate laparotomy, enterotomy and removal of the battery
b. Enteroscopy with extraction
c. Laparoscopy with ultrasound localization and extraction
d. Cathartics and a follow-up plain film in 48 hours if the child remains asymptomatic

Answer: d

Passage of an ingested foreign body beyond the gastroesophageal junction is associated with a 95% probability of uneventful distal transit. The character of the foreign body is largely irrelevant but batteries, particularly alkaline disk batteries, present a potentially serious hazard to children and may require an aggressive approach. A number of reports have documented the unique risk of intestinal perforation resulting from disruption of the battery casing and associated spillage of the toxic contents. Although some have advocated routine immediate removal, this does not appear necessary. If the battery is endoscopically accessible in the esophagus, it should be removed when recognized. Cathartics and enemas may help expedite passage of batteries discovered when already in the small or large bowel. Delay for more than a few days, casing rupture on plain radiograph, or symptoms of any kind require extraction. Despite the risks, most batteries pass without these sequelae.

43. A jaundiced 6 week old infant has biliary atresia. Which of the following statements are true?

a. Portoenterostomy is the initial procedure of choice
b. Primary liver transplantation using either a reduced sized cadaveric graft or a living related graft is now the procedure of choice

c. Approximately two-thirds of patients managed with portoenterostomy will develop chronic liver disease sufficient to indicate liver transplantation
d. Because biliary atresia has pathogenic components of acute and chronic inflammation, antiinflammatory therapy is known to delay onset of liver failure

Answer: a, c

Biliary atresia is an idiopathic process in which the extrahepatic biliary ducts are replaced in whole or in part with dense fibrous tissue containing evidence of both acute and chronic inflammation. There is an intrahepatic component as well. Although antiinflammatory therapy is of some theoretical interest, there are no data to suggest that antiinflammatory pharmocologic therapy will influence the natural history of liver disease associated with biliary atresia.
The approach for the usual infant in whom biliary atresia is discovered within the first 90 days of life is to confirm the suspected diagnosis by operative cholangiogram at laparotomy and then proceed with portoenterostomy. In general terms, one-third of these infants do well on a long-term basis, one-third have prompt failure, and the remainder have chronic liver disease that becomes problematic more slowly. Therefore, approximately two-thirds of these patients develop chronic liver disease for which liver transplantation is a reasonable alternative. Hepatic transplantation is best considered a necessary and complementary approach to portoenterostomy for infants with biliary atresia. Data support its use in infants with failed portoenterostomy or in older infants with established cirrhosis at the time of presentation. Growth failure, hepatic synthetic failure and sequelae of portal hypertension are indications to proceed with transplantation.

44. Of the following, which is the most likely cause of hemodynamically significant lower gastrointestinal bleeding in a 6 month old male child?

a. Meckel diverticulum
b. Henoch-Schonlein purpura
c. Intussusception
d. Crohn's colitis
e. Hemolytic uremic syndrome

Answer: a, c

All of the choices are possible causes of lower GI bleeding in a 6 month old; intussusception and a bleeding Meckel diverticulum are the most common. Upper GI hemorrhage with passage of blood distal should also be considered, but is not discussed further here as sampling nasogastric aspirate is a relatively easy and reliable means of differentiating these problems. The magnitude of blood loss associated with intussusception is usually minor, but the associated vomiting and bowel obstruction may lead to significant volume depletion with hemoconcentration. The magnitude of the hemorrhage is usually more significant with a Meckel diverticulum.
Infectious diarrheas also occur in this age group. Typically, signs and symptoms include fever and ileus with bloody diarrhea. The diagnosis is confirmed with stool examination for leukocytes and cultures for specific pathogens.

45. Which of the followings statement regarding Hirschprung's disease are true?

a. Suction rectal biopsy is virtually always diagnostic if the specimen includes submucosa
b. Hirschprung's disease is the result of a sex linked dominant gene
c. The endorectal pullthrough is demonstrably superior to other forms of surgical construction
d. Ninety percent or more of patients have an excellent or good functional result following reconstructive surgery
e. The important cause of mortality in contemporary practice is enterocolitis

Answer: a, d, e

The incidence of Hirschprung's disease is about 1 per 5000 live births, with no racial predilection, but with a marked male-to-female (4:1) preponderance. Most cases are sporadic, but long-segment or total colonic aganglionosis and female gender are strongly associated with familial disease. Recent data suggest an association with the RET protooncogene. The genetic basis of Hirschprung's disease is under active investigation and it appears that several genes including those located on chromosomes 10, 13, 22 and possibly others are involved. It is neither sex linked nor dominant. There is a rare association with the MEN syndromes, particularly medullary carcinoma of the thyroid.
The accuracy of suction rectal biopsy is 100% with a correctly done biopsy that includes submucosa and experienced pediatric pathology in several large series. This requires both a search for ganglion cells and evaluation of the axons of the myenteric neurons using either conventional staining techniques or histochemical staining for acetylcholinesterase.

Definitive operations for congenital aganglionosis all depend on resection or bypass of the distal aganglionic rectum with a low rectal anastomosis to normally innervated pulled-through proximal intestine. Selection among the several described operations depends more on a surgeon's individual training and preference rather than upon demonstrable differences in outcome. Although the endorectal pull-through is one of the widely practiced and popular procedures, it is not demonstrably superior to the procedures described by Duhamel or Swenson (see text). Eighty to 90% or more of patients have excellent or normal bowel function following reconstructive surgery for Hirschprung's disease when evaluated after 5 years, regardless of the procedure employed.
The primary remaining cause of mortality directly attributable to Hirschprung's disease itself is enterocolitis. When it occurs, this is typically found in infants or neonates for whom the diagnosis has been delayed. Postoperative enterocolitis does occur, but it tends to be substantially less virulent. Undiagnosed neonatal Hirschprung's enterocolitis can lead to death in 12 to 24 hours from overwhelming sepsis.

46. The operative procedure of choice for managing the most common type of choledochal cyst is which of the following?

a. Cyst gastrostomy
b. Cyst jejunostomy
c. Excision with Roux-en-Y hepaticojejunostomy
d. Transduodenal marsupialization
e. Endoscopic sphincterotomy

Answer: c

The most common choledochal cyst is a type I cyst; 80% to 90% of the total in most reports. These are characterized by fusiform dilation of the choledochus itself. These cysts typically involve the entire common bile duct with only mild dilation of the common hepatic duct and a normal intrahepatic system. The treatment of this lesion is always surgical. Internal drainage procedures without cyst resection (e.g., cyst duodenostomy, cyst gastrostomy and cyst jejunostomy) were routinely performed for type I choledochal cysts until the 1970s. The rate of failure (e.g., stricture, recurrent cholangitis, stone formation, pancreatitis) ranged from 30% to 75%, depending on the length of follow-up and the type of procedure. As these late complication rates became apparent, the risk of bile duct adenocarcinoma in the residual cyst also became widely recognized. Therefore, the preferred operative treatment of a type I choledochal cyst is total transmural excision with Roux-en-Y hepaticojejunostomy. Occasionally, adults with severe inflammation and fibrosis may require intramural cyst dissection, leaving the posteriomedial (outer) wall of the cyst in situ to protect the adjacent portal vein and hepatic artery.

47. Which of the following is the most common liver tumor of childhood?

a. Hemangioma and hemangioendothelioma
b. Hepatoblastoma
c. Hepatocellular carcinoma
d. Mesenchymal hamaratoma

Answer: b

Primary liver tumors are uncommon in children. Of these, about one-third are benign and two-thirds are malignant. The most common presenting feature for a liver tumor is an asymptomatic abdominal mass. The diagnostic evaluation is generally an ultrasound initially followed by either computed tomography or magnetic resonance imaging, for definitive diagnosis. The relative incidence of liver tumors is illustrated in the table below.
Hepatoblastoma is the most common liver tumor of childhood. Most hepatoblastomas are discovered within the first two years of life. Although these are large and may require primary chemotherapy, a 65%–75% survival is achievable with resection that yields histologically clear resection margins.

48. The risk of biliary tract adenocarcinoma developing in a patient with a choledochal cyst left in situ is approximately which of the following?

a. Less than 1%
b. 3% to 5%
c. 10% to 15%

d. Greater than 25%

Answer: b

Adenocarcinoma of the biliary tract develops in 3% to 5% of patients who have choledochal cysts. Although this represent a small number of patients, the total number reported exceeds 50 and the incidence is about 1000 times that of the normal population. In addition, the carcinoma may develop as early as the adolescent years, a marked contrast to the normal population in which a fifth or sixth decade of life presentation is typical. Neoplastic transformation of the dysplastic biliary cyst epithelium may result from chronic inflammation. Consideration of this potential problem has contributed significantly to the current emphasis on surgical excision of these cysts.

49. The most common cause of acute pancreatitis in childhood is which of the following?

a. Pancreas divisum
b. Cholelithiasis
c. Trauma
d. Valproic acid
e. Annular pancreas

Answer: c

Although adult pancreatitis is usually related to cholelithiasis or alcohol ingestion, pediatric causes are considerably more varied in etiology. Fifty to 70% of childhood acute pancreatitis is either idiopathic or posttraumatic in origin. Trauma is the single most common cause of acute pancreatitis in childhood. Cholelithiasis is an important etiology in the adolescent population and in children with hemotologic disorders. Annular pancreas normally is associated with duodenal atresia and produces neonatal duodenal obstruction, but not acute pancreatitis. Pancreas divisum is an anatomic variation present in 10% to 15% of normal children that is occasionally the cause of acute pancreatitis. Valproic acid is an important anti-convulsant used in pediatric neurology. One of its possible complications is the development of acute pancreatitis or necrotizing pancreatitis. Fortunately, this is rare as it is often lethal.

50. Which of the following statements regarding gastroschisis are true?

a. Primary fascial closure can be achieved in only about 25% of these infants
b. These infants have an incidence of approximately 40% to 50% of associated anomalies
c. Overall survival is approximately 80% to 90%
d. When the diagnosis is known prenatally, planned cesarean section is the safest method of delivery

Answer: c

Gastroschisis is a full-thickness defect of the abdominal wall with herniation of a variable amount of uncovered intestine. Prenatal diagnosis has enabled the diagnosis of gastroschisis to be made prior to delivery. Prospective evaluation comparing vaginal delivery with elective cesarean section has demonstrated no difference in outcome. Therefore, careful vaginal delivery generally remains the birthing method of choice. Unlike omphalocele which is commonly associated (50%) with other anomalies, other structural anomalies are associated with gastroschisis in approximately 10% of patients.
Primary fascial closure after reduction of the herniated viscera is the best surgical option and this is possible in 60% to 70% of infants. Care must be taken not to generate excessive intraabdominal pressure when performing a primary abdominal wall closure. Generally, an intraabdominal pressure less than 20 cm H2O is well tolerated. If the herniated viscera cannot be reduced primarily, a silastic pouch constructed to temporarily contain the extra abdominal bowel and a series of partial reductions are begun. This approach combined with adequate nutritional support by total parenteral nutrition yields survival rates of at least 80% to 90% in most contemporary series of gastroschisis.

51. Which of the following are typical causes of neonatal intestinal obstruction?

a. Intussusception
b. Meconium ileus
c. Hirschprung's disease
d. Meckel's diverticulum
e. Incarcerated hernia

Answer: b, c, e

Neonatal bowel obstruction is defined as intestinal obstruction developing in the first 30 days of life. The cardinal manifestation is bilious vomiting often in conjunction with abdominal distention. Meconium ileus and Hirschprung's disease are classical causes of neonatal intestinal obstruction outlined in the table. Incarcerated inguinal hernias are the most common cause of neonatal intestinal obstruction. Hernias usually do not present a diagnostic dilemma, as simple inspection of the groin yields an obvious diagnosis. Intussusception, while a cause of distal bowel obstruction in infants, does not usually become a consideration until at least 3 to 6 months of age. Intestinal obstruction related to Meckel's diverticulum is generally related to either intussusception or volvulus associated with a band from the Meckel's diverticulum to the abdominal wall. Both of these events tend to occur later than the neonatal period.

52. A 6-week-old child presents with generalized seizures, a serum glucose of 30 mg/dL and concurrent hyperinsulinemia. This child's first priority is which of the following?

a. Permanent central venous access and glucose infusion
b. Administration of cortisone and adrenocorticotropic hormone
c. Computerized tomographic scan of the abdomen to look for an islet cell adenoma
d. Urgent pancreatic resection

Answer: a

Most cases of hyperinsulinemia in the first 2 years of life are due to nesidioblastosis, a condition associated with excessive and diffuse formation of neoislets from primitive pancreatic ductal cells. Hyperinsulinemia secondary to islet cell adenoma or carcinoma or islet cell hyperplasia is more common in the older child. Infants with nesidioblastosis, such as the one described here, typically present with symptomatic hypoglycemia, seizures and hyperinsulinemia. An insulin-to-glucose ratio (insulin in IU/ml divided by glucose in mg/dL) that is greater than 0.5 with fasting is highly suggestive.
Infants with nesidioblastosis are managed initially medically with maintenance of blood glucose levels above 40 mg/dL. This is best carried out by the infusion of hypertonic glucose solutions through a permanent central venous catheter. In addition, diazoxide, cortisone and adrenocorticotropic hormone and streptozocin have been used to treat the hypoglycemia.
Definitive management of nesidioblastosis may require pancreatic resection. This requires a pancreatectomy usually estimated at approximately 95% to 99% with splenic and duodenal preservation. Following 90% to 95% pancreatectomy, over 90% of infants with nesidioblastosis are rendered permanently euglycemic. A CT scan of the abdomen to search for an adenoma is an appropriate diagnostic maneuver, but it is not the first priority.

53. At what age is surgical orchiopexy recommended for a child with a unilateral undescended testis?

a. Promptly upon discovery, regardless of age
b. 1 year
c. 5 to 6 years
d. Any time prior to puberty

Answer: b

Undescended testis occurs in 30% of premature boys, 3.4% of full-term boys, 0.8% of 1-year olds, and 0.8% of adults. The implication is that if spontaneous testicular descent has not occurred by 1 year of age, it is unlikely to occur subsequently. Therefore, orchiopexy is deferred until the patient reaches 1 year of age. Testes that remain undescended during childhood have a significant reduction in the number of germ cells and have little function after puberty. Demonstrable histologic changes are present by the age of 2 years. For this reason, delay beyond 1 year is not recommended.

54. An infant is noted to have a left flank mass shortly after birth and an ultrasound examination demonstrates left hydronephrosis. The most common cause of this finding is which of the following?

a. Neonatal Wilm's tumor
b. Congenital ureteropelvic junction obstruction
c. Multicystic dysplastic kidney
d. Vesicoureteral reflux

Answer: b

The most common cause of neonatal hydronephrosis is congenital ureteropelvic junction obstruction. It is important to rule out bilateral ureteropelvic junction obstruction when this is found as this can present in 20% to 30% of neonatal cases. The management requires establishment of dependent drainage of the renal pelvis, and this is usually accomplished by a dismembered pyeloplasty with resection of the obstructing segment. A successful outcome is expected in over 90% of patients.
Neonatal Wilm's tumor is exceedingly rare and does not ordinarily present with obstruction of a functioning kidney, but rather as a solid mass. Multicystic dysplastic kidney occurs in about 1 in 4000 births and is usually unilateral. It is the most common form of renal cystic disease in infants, but these cysts do not communicate with a functional renal pelvis and there is little or no functional renal cortex. Vesicoureteral reflux, while common, does not present in the newborn period with hydronephrosis and a palpable kidney.

55. The medical indications for circumcision include which of the following?

a. Infants with a history of urinary tract infection
b. Hypospadias
c. Phimosis
d. Enuresis
e. Vesicoureteral reflux

Answer: a, c, e

The American Academy of Pediatrics has published guidelines on the indications for circumcision. Circumcision should be encouraged in infants with a history of urinary tract infection or vesicoureteral reflux to decrease the chances of ascending infection. On the contrary, in an infant with hypospadias, chordee, significant penoscrotal webbing, or other congenital problems, circumcision should be discouraged to preserve the foreskin for use in later reconstruction. In normal infants, circumcision is a matter of family choice and not an important issue for medical debate. Phimosis is a fibrotic contraction of the preputial aperture so that retraction is impossible. Circumcision or dorsal slit are the most effective solutions to phimosis. Enuresis is unrelated to the preputial skin and has no bearing on the decision for circumcision.

56. Which of the following statements regarding neuroblastoma are true?

a. Neuroblastoma is the most common abdominal malignancy of childhood
b. Approximately 80% of neuroblastoma patients are diagnosed prior to age 4 years
c. N-myc oncogene copy number in neuroblastoma tissue is inversely related to survival probability
d. Trk proto-oncogene expression in neuroblastoma tissue is inversely related to survival probability
e. All of the above

Answer: a, b, c

Neuroblastoma is the most common extracranial solid tumor and the most common abdominal malignancy of childhood. The incidence is approximately 8 to 10 per million children under the age of 15 years. The incidence is uniform throughout the world. This results in approximately 500 new cases reported each year in the United States. The median age at diagnosis is about 2 years and 80% of children are less than four years of age at diagnosis. The N-myc oncogene, whose function and mechanism of action remain the subject of investigation, was empirically shown to be a useful predictor of survival and risk. It was found that patients with increased copies of the N-myc gene had a much worsened prognosis. Currently, most authorities consider a copy number of more than ten to be significant. The trk proto-oncogene is a component of the high-affinity nerve growth factor receptor and is expressed in human neuroblastoma tissue. Trk-A expression is inversely correlated with N-myc amplification and is associated with lower stage at diagnosis and improved prognosis.

57. Which of the following are considered low risk features for neuroblastoma patients?

a. Age less than one year
b. Stage 2A and 2B disease (International Staging Criteria)
c. Stage 4S disease (International Staging Criteria)
d. Neuron specific enolase plasma level less than 100 ng/ml
e. None of the above

Answer: a, b, c, d

Risk status is determined in neuroblastoma patients by a number of characteristics which have been elaborated over the last twenty years.

58. A one month old female infant is brought to you for evaluation of afriable polypoid mass prolapsing through the vaginal introitus. Your presumptive diagnosis is which of the following?

a. Ectopic ureterocele
b. Rectal prolapse
c. Congenital adrenal hyperplasia with ambiguous genitalia
d. Embryonal rhabdomyosarcoma

Answer: d

Vaginal and cervical primary rhabdomyosarcomas often prolapse through the vaginal orifice as a friable polypoid mass and may hemorrhage. Botyroid tumors are really of the embryonal subtype but grow into a hollow space such as the vagina or bladder so they undertake a characteristic “grape-like” appearance. Other patterns of rhabdomyosarcoma include alveolar and pleomorphic morphology. The incidence of rhabdomysarcoma is biphasic with one peak in infancy followed by the second in the adolescent years. The nature of presentation is site dependent. This patient has a classical presentation for a vaginal botyroid rhabdomyosarcoma. The emphasis in evaluation is to perform a thorough pretreatment workup that defines completely local tumor extent and evaluates the regional and distant sites of metastases.

59. Which of the following approaches is considered standard care for most Wilms’ tumor patients in the United States today?

a. Adriamycin and vincristine therapy followed by surgical resection
b. Needle biopsy followed by either chemotherapy or resection depending upon the histology
c. Primary surgical resection followed by chemotherapy
d. Radiation therapy if judged unresectable on CT or MRI imaging

Answer: c

The standard of care for Wilms’ tumor patients in the United States is initial surgical resection. Exceptions to this rule include extensive intracaval tumors which require cardiopulmonary bypass for extraction, obviously unresectable tumors with documented invasion of contiguous structures, and possibly bilateral tumors, especially if it is unclear which side is most heavily involved. All resectable Wilms’ tumor patients receive postoperative chemotherapy with the possible exception of Stage 1 favorable histology patients who are younger than 24 months of age at diagnosis and have tumors less than 250 grams in weight at resection. Chemotherapy followed by surgical resection is practiced in Europe with roughly equal outcomes to those in the United States but this approach has the disadvantage of changing the surgical and pathologic staging which are the basis for the National Wilms’ Tumor Studies and the cornerstone of treatment in the United States. Needle biopsy has a very limited role for unusual presentations of Wilms’ tumor as the diagnosis is generally apparent with modern imaging techniques. Radiation therapy is not a primary mode of therapy for Wilms’ tumor under contemporary National Wilms’ Tumor treatment protocols.

60. Which of the following statements regarding rhabdomyosarcoma are true?

a. Surgical resection of the primary tumor results in cure of approximately 80 to 90% of all patients
b. Currently recommended therapy includes complete resection of primary tumors prior to chemotherapy for small noninvasive lesions, or after documented response with more formidable primary tumors
c. Alveolar histology is a favorable prognostic finding
d. Overall survival of all patients is now approximately 50%

Answer: b, d

Surgical resection of the primary tumor was the mainstay of treatment 30 years ago for rhabdomyosarcoma but only resulted in overall survival rates in the range of 20%. This improved to approximately 50% with the addition of chemotherapy. Survival is stage dependent and if all cases (both high and low risk) are included, the overall survival from

rhabdomyosarcoma is now approximately 50%. Presently it is recommended that complete resection of primary tumors should be undertaken either before chemotherapy for small noninvasive lesions or after documented response with more formidable primary tumors. In certain situations where chemotherapy results in a complete or very good tumor regression, external beam radiation may be employed as a primary means of local control. Debilitating or disfiguring surgery is only performed if residual tumor is present after both chemotherapy and therapeutic irradiation. Alveolar histology is associated with a particularly poor prognosis for rhabdomyosarcoma.

61. Patients with Wilms' tumors most frequently present with which of the following?

a. Bilateral metachronous lesions
b. Bilateral synchronous lesions
c. An extrarenal primary
d. A multicentric primary lesion
e. A unifocal, unilateral lesion

Answer: e

It is currently hypothesized that Wilms' tumor arises from primitive metanephric blastema and individual tumors often contain not only primitive metanephric cells but also cartilage, skeletal muscle and squamous epithelium. Most tumors arise unifocally within the kidney but approximately seven percent of unilateral Wilms' tumors are multicentric. The proportion of synchronous bilateral tumors among all nephroblastoma patients ranges from 4.4 to 7.0% while that of metachronous tumors is 1.0–1.9%. Wilms' tumors are equally distributed with regard to the left and right side and may occur with no apparent connection to the kidneys. Usually, extrarenal Wilms' tumor occurs in the retroperitoneal area but other reported sites include pelvis, scrotum and inguinal region.

62. Hepatoblastomas are childhood liver tumors characterized by which of the following features?

a. Multicentricity
b. Cirrhosis in the uninvolved liver
c. Unresectable tumors subjected to cytoreductive chemotherapy may be resected with long-term survival
d. Jaundice

Answer: c

Children with hepatoblastoma most commonly present with an abdominal mass or diffuse abdominal swelling. The child is typically in good health and the lesion may be observed by an observant parent or clinician on routine examination. Weight loss and other symptoms are unusual. Liver function tests are usually normal or nonspecifically altered. Jaundice is uncommon. The most useful tumor marker is the serum (a-fetoprotein (AFP)) level which is elevated in approximately 90% of the cases.
Hepatoblastoma usually presents as a single, pseudo-encapsulated lesion often reaching large proportions before becoming apparent. The umbilical fissure is generally not breached. Multicentricity occurs in less than 20% and cirrhosis of the surrounding liver is unusual. Multicentricity and associated cirrhosis are typical of hepatocellular carcinoma. Complete surgical resection remains the major objective of therapy for hepatoblastoma. At presentation approximately 60% of patients with hepatoblastoma have resectable tumors. Chemotherapy is the major treatment option available for unresectable tumors. Over the last decade it has become apparent that some of these patients may be rendered resectable by preoperative chemotherapy.

63. Common sites of neuroblastoma metastasis are which of the following?

a. Lung
b. Regional lymph nodes
c. Bone marrow
d. Cortical bone
Liver

Answer: b, c, d

Neuroblastoma metastasizes to both regional lymph nodes and distant sites, most frequently bone marrow and/or cortical bone. The liver and lungs are rarely the site of metastatic spread. Cortical bone involvement as manifested by a positive bone scan is a particularly poor prognostic indicator. The majority of patients present with locally advanced disease at the time of diagnosis.

64. Which of the following statements regarding renal tumors of childhood and adolescence are true?

a. Clear cell sarcoma is presently considered a variant of Wilms' tumor with a poor prognosis
b. Clear cell sarcoma of the kidney has a high rate of metastasis to bone
c. Rhabdoid tumors may arise in the kidney, mediastinum or brain
d. Childhood rhabdoid tumors of the kidney carry an excellent prognosis

Answer: b, c

Clear cell sarcoma of the kidney is presently considered a distinct histopathologic and clinical entity from Wilms' tumor. It has a similar age distribution as that observed in Wilms' tumor, but a markedly worsened prognosis. It is characterized by a proclivity to metastasize to bones and indeed has been called the bone metastasizing renal tumor of childhood. Relapse and death occur in 75% of patients with over half dying within one year of diagnosis. Aggressive systemic chemotherapy is recommended for all stages of the disease. Likewise, postoperative radiation to the tumor bed is recommended regardless of stage.
Rhabdoid tumors are rare malignancies that most commonly involve the kidney in childhood but may also occur primarily in the mediastinum or brain. Outcome is particularly poor and there is no proven chemotherapy regimen. Rhabdoid tumors of the kidney occur in infancy with a median age at presentation of 13 months. Survival rates are almost zero and even Stage I patients fare poorly. Aggressive therapy is warranted including surgical resection, local radiation therapy and systemic chemotherapy.

65. Which of the following syndromes are associated with the development of Wilms' tumor?

a. Beckwith-Wiedemann Syndrome (hemi-hypertrophy, macroglossia, aniridia)
b. Neurofibromatosis
c. Denys-Drash syndrome (pseudohermaphroditism, glomerulopathy)
d. Gonadal dysgenesis
e. Hemolytic uremic syndrome

Answer: a, b, c, d

A number of syndromes are associated with the development of Wilms' tumors and when present, require routine periodic screening. There is no known association of the hemolytic uremic syndrome with the development of Wilms' tumor.