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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 gastrografin 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 gastrografin 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 gastrografin evacuation fails, laparotomy, placement of a pursestring suture in the bowel wall, and intraluminal irrigation with saline and gastrografin (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.