

1. C. Bilious emesis should always be considered an emergency and in this newborn the most likely diagnosis is malrotation with midgut volvulus. During the 5th to 8th embryonic week, the intestine projects out of the abdominal cavity and then rotates 270 degrees before returning into the abdomen. If the rotation does not occur correctly, the intestine will not be anchored at the mesentery and is considered malrotated and at risk for volvulus. The incidence of malrotation is one per 500 live-births, with a 2:1 male to female ratio in the newborn period. Volvulus is the twisting of a loop of bowel about the mesenteric attachment. The incidence of volvulus is however much less frequent with up to seventy-five percent of cases occur within the first month of life. After initial stabilization, an NG tube should be placed, antibiotics initiated if perforation or sepsis is suspected and an immediate surgical consultation should be obtained.
2. B. Patients with congenital adrenal hyperplasia present with hypoglycemia, hyperkalemia and hyponatremia. Congenital adrenal hyperplasia is due to a deficiency in one of 5 enzymes involved in the production of cortisol. This results in decreased conversion of 17-OH progesterone to 11-desoxycortisol and a subsequent drop in cortisol levels which can lead to cardiovascular collapse. The most common form of congenital adrenal hyperplasia is due to 21-Hydroxylase deficiency, which accounts for 90-95% of cases. Most of the enzyme deficiencies lead to decreased aldosterone production and subsequent hyperkalemia and hyponatremia.. If the child has evidence of congenital adrenal hyperplasia, resuscitate with boluses of normal saline if the child has evidence of shock or volume depletion. Consultation with a pediatric endocrinologist may be helpful in deciding upon the maintenance fluids. *Administer hydrocortisone 25 mg IV, then 25-50 mg/m²/day (body surface area is equal to the square root of the: (height in centimeters multiplied by the weight in kilograms) divided by 3600)) divided every 6 to 8 hours (approximately 25 mg every 6 to 8 hrs in full-term newborns). Hyperkalemia typically responds to fluid replacement alone, however severe hyperkalemia, especially if dysrhythmias are present, should be treated with 10% calcium gluconate (100mg/kg), sodium bicarbonate 1 meq/kg, insulin 0.1 unit/kg with D10 3-10 cc/kg, and kayexalate. Monitor serum glucose closely as these patients are frequently hypoglycemic. Treatment includes hydrocortisone (remember to draw extra red top tubes for analysis prior to administration. The blood is typically tested for 17-hydroxyprogesterone, dehydroepiandrosterone, androstenedione and testosterone)*
3. The drug of choice for ductal dependent cyanotic heart disease is prostaglandin E1- with a starting infusion of -0.05-1ug/kg/min. There is a risk of apnea associated with its use so be prepared to intubate, other complications include seizures and fever. Patients with congenital heart disease present with poor feeding, sweating with feeds tachypnea, sudden onset of cyanosis or pallor that may worsen with crying, lethargy, or failure to thrive. Patients with cyanotic congenital heart disease are hypoxic but typically have a minimal response to oxygen therapy; whereas patients with a pulmonary process causing hypoxia will have an increase in their oxygen saturation when oxygen is administered. Indomethacin is used to close a patent ductus.

4. C. Newborns with hypoglycemia should be treated with D₁₀W solution with a range of 2-10cc/kg. Higher concentrations should not be used as they can cause vein sclerosis and intracranial hemorrhage. Infants and young children should be treated with D₂₅- 2-4cc/kg.
5. A. Suspect Hirschsprung Disease in a newborn who has not yet passed a meconium stool. Other possibilities include an imperforate anus or meconium plugging. Older children present with a history of chronic constipation. Hirschsprung disease is the absence of intramural ganglion cells in the rectum which extends to the sigmoid colon in 77% of patients and involves the entire colon in 15% of patients. The incidence is 1/5,000 live-births, with a male to female ratio of 4:1. The diagnosis should be suspected if the patient presents with lack of meconium stool within the first 24 to 48 hours of life. Vomiting and abdominal distension may also be present.
6. C. Excessive free water intake can result in hyponatremic seizures. Infants less than 6 months of age are particularly susceptible to these types of seizures and commonly have intractable seizures requiring intubation and hypothermia. Immediate treatment includes the administration of 3% saline 4cc/kg.
7. D This patient has Omphalitis and should undergo a full septic evaluation, administration of antibiotics and hospital admission. Surgical debridement may be required for severe cases. Omphalitis is inflammation and infection surrounding the umbilicus that can spread to the liver or peritoneum. Patients can present with symptoms ranging from mild erythema to necrosing lesions surrounding the umbilicus on the abdominal wall. Fever may be present.
8. B. This patient is at risk for herpes encephalitis and should undergo a complete septic workup and IV acyclovir should be initiated in the ED. Begin acyclovir (20mg/kg every 8 hours IV) if there is a positive maternal history of herpes, a vesicular rash, focal neurologic findings, CSF pleocytosis or elevated CSF protein without organisms on gram stain.
9. PALS defines SVT in infants as a heart rate of greater than 220 BPM. In older children the heart rate for SVT is greater than 180 BPM. The ECG demonstrates a narrow complex tachycardia without discernible p waves or beat-to-beat variability. In the stable patient, vagal maneuvers are the first treatment of choice (ice to the face, or blowing through an occluded straw in older children). Adenosine given as centrally as possible is the first drug of choice (0.1mg/kg up to 6mg for the first dose and then 0.2mg/kg for the second dose up to 12 mg) If this is not successful, then amiodarone-5mg/kg given over 20-60 minutes or procainamide 15mg/kg given over 30-60 minutes are the next drugs of choice. Unstable patients should undergo cardioversion with 0.5-1J/kg followed by 2J/kg. If an IV is accessible, a dose of adenosine can be given while setting up for the cardioversion.
10. B. Vaginal bleeding in the first few weeks of life is often due to withdrawal of maternal hormones and parental reassurance is all that is necessary.
11. A. Suspect an inborn error of metabolism in patients who have an altered level of consciousness. These patients may or may not be acidotic depending on the type of inborn error that is present. Patients with a urea cycle defect typically have a normal blood gas

but an elevated ammonia level. Patients with organic acidemias will be acidotic but may or may not have an elevated ammonia level. Patients with galactosemia will have a normal blood gas and ammonia level but will have reducing substances in the urine.

12. E. An easy way to remember the differential diagnosis of the critically ill newborn is the mnemonic “THE MISFITS”:

T - Trauma/ NAT (non-accidental trauma)

H- Heart disease- congenital/hypovolemia/hypoxia (respiratory complaints)

E- Endocrine (Congenital adrenal hyperplasia, thyrotoxicosis)

M-Metabolic disturbances (hypoglycemia, hyponatremia)

I-Inborn errors of metabolism

S-Sepsis

F-Formula dilution or over concentration

I-Intestinal catastrophes

T-Toxins (home remedies)

S- Seizures