Newborn Emergencies: The First 30 Days of Life

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The evaluation and appropriate management of the critically ill neonate (≤28 days of age) require an intimate knowledge of the physiologic changes and life-threatening pathologic conditions that may present during this time. The innate differences in this fragile population may provoke anxiety for the emergency department (ED) physician. For this reason, a broad systematic approach to evaluating the neonate is necessary to provide a comprehensive yet specific differential diagnosis for a presenting complaint or symptom. The efficient recognition and prompt management of illness in the neonatal period may be life saving. In recent years, it has become more important for the ED physician to be familiar with the neonate because of early discharge policies from newborn nurseries. This review provides a systematic approach to the recognition, emergency stabilization, and management of the more common newborn emergencies.

Neurologic emergencies

Recognizing a neurologic insult in neonates may be difficult. The clinical symptoms may be nonspecific. The history may reveal only a change in feeding pattern or subtle behavioral changes. A useful mnemonic to recall the broad differential diagnosis of a neonate with altered mental status, “THE MISFITS,” is outlined in Box 1. Keeping this mnemonic in mind as well as a high index of suspicion during the initial history and physical examination (non) will help guide the evaluation and management.

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Seizures occurring during the neonatal period are often difficult to recognize. The cortical development is not complete, and as a result, generalized motor activity is less common. Subtle seizures in the term neonate can include abnormal eye movements (usually horizontal, sustained eye deviation), lip smacking, abnormal tongue movements, pedaling, or apnea [1,2].

Although hypoxic-ischemic events are the more common cause of neonatal seizures (60%), the list of other causes is extensive [3]. Box 2 provides the different causes of neonatal seizures based on the age of presentation. Because intracranial infections account for 5% to 10% of neonatal seizures, a thorough sepsis evaluation should be completed on all neonates in the absence of any other immediately apparent cause.

The initial evaluation should include stabilizing the airway, breathing, and circulation (ABC) and obtaining the blood glucose level. The correction of hypoglycemia (≤40 mg/dL) should be accomplished with a 10% dextrose solution, 2 to 4 mL/kg intravenous (IV) [4]. While obtaining vascular access, additional blood tests should include serum electrolytes, a complete blood count (CBC), and blood culture. Lorazepam, 0.1 mg/kg IV, is the initial drug of choice in neonatal seizures [4]. Lorazepam is considered superior to diazepam because of the smaller volume of distribution and longer half-life [5]. If the seizure continues then phenobarbital should be the second-line choice, followed by phenytoin or fosphenytoin. Table 1 includes the pharmacologic management and doses of antiepileptic medications for status epilepticus in the neonatal period.

Other electrolyte imbalances that can result in seizures include hypocalcemia and hyponatremia. Hypocalcemia (≤7 mg/dL) should also be corrected with 100–300 mg/kg IV of 10% calcium gluconate solution. Hyponatremia (≤125 mg/dL) should be corrected with 3% saline, 5 to 10 mL/kg IV [4].

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**Box 1. Causes of altered mental status in a neonate: THE MISFITS**

T–Trauma (nonaccidental and accidental)
H–Heart disease and hypovolemia
E–Endocrine (eg, congenital adrenal hyperplasia and thyrotoxicosis)
M–Metabolic (electrolyte imbalance)
I–Inborn errors of metabolism
S–Sepsis (eg, meningitis, pneumonia, and urinary tract infection)
F–Formula mishaps (eg, under- or overdilution)
I–Intestinal catastrophes (eg, volvulus, intussusception, and necrotizing enterocolitis)
T–Toxins and poisons
S–Seizures

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Seizures

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After the seizure has stopped, a CT scan or ultrasonogram of the head should be obtained, and a sepsis evaluation should be completed. Broad-spectrum antibiotics should be administered, and the initiation of antiviral therapy (acyclovir) should be considered. The treatment for sepsis or meningitis should not be delayed if a lumbar puncture cannot be performed at the time. All neonates

Box 2. Causes of seizures in infants

First day of life

Anoxia/hypoxia
Drugs
Hypoglycemia/hyperglycemia
Infection
Intracranial hemorrhage
Pyridoxine deficiency
Trauma

Second day of life

Benign familial neonatal seizures
Congenital anomalies or developmental brain disorders
Drug withdrawal
Hyperphosphatemia
Hypertension
Hypocalcemia
Hypoglycemia
Hyponatremia/hypernatremia
Inborn errors of metabolism
Sepsis
Trauma

Day 4 to 6 months of age

Benign idiopathic neonatal seizures
Congenital anomalies or developmental brain disorders
Drug withdrawal
Hyperphosphatemia
Hypertension
Hypocalcemia
Hyponatremia/hypernatremia
Inborn errors of metabolism
Infection
with seizures require admission to the hospital for observation and completion of the evaluation.

**Nonaccidental head trauma: shaken baby syndrome**

The terms nonaccidental head trauma or shaken baby syndrome (SBS) are more recent terms that were adapted from Caffee’s [6] landmark 1974 article, which introduced the concept of “the whiplash shaken baby syndrome.” The diagnosis of nonaccidental trauma may be challenging, depending on the presenting signs and symptoms. In fact, one study found that 31% of infants with unrecognized abuse had been evaluated previously by a physician [7]. Nonaccidental trauma is an important diagnosis to consider in an infant with a suspicious history or nonspecific symptoms because the long-term morbidity from SBS is as high as 70%, and mortality is as high as 30% [8,9]. Although these infants may have no external signs of trauma, the presence of any scalp hematoma is associated with an increased incidence of intracranial hemorrhage.

The initial management should include ABC stabilization as well as a thorough physical examination that includes an attempt to visualize the retinas for hemorrhages. While obtaining IV access, blood may be obtained for a complete blood count, platelet count, prothrombin time, and partial thromboplastin time. Further laboratory evaluation, including cultures and bedside glucose evaluation, will be determined by the clinical presentation. Once the infant is stabilized, a CT of the head should be performed. These infants should be admitted to the hospital for further medical evaluation, management, and investigation by the appropriate authorities. Any suspicion of nonaccidental trauma should be reported from the emergency department.

**Apparent life-threatening event**

An apparent life-threatening event (ALTE) is defined as “an episode that is frightening to the observer that is characterized by some combination of apnea, color change, marked change in muscle tone, choking or gagging. In some cases the observer fears that the infant has died” [10]. Because the diagnosis is subjective and dependent on the observer’s interpretation, the ED evaluation will

<table>
<thead>
<tr>
<th>Drug</th>
<th>Dosage</th>
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<tr>
<td>Benzodiazepines</td>
<td></td>
</tr>
<tr>
<td>Lorazepam</td>
<td>0.05–0.1 mg/kg IV</td>
</tr>
<tr>
<td>Diazepam</td>
<td>0.2–0.3 mg/kg IV or 0.5 mg/kg rectal</td>
</tr>
<tr>
<td>Midazolam</td>
<td>0.1 mg/kg IV or 0.2 mg/kg intramuscular</td>
</tr>
<tr>
<td>Phenobarbital</td>
<td>20 mg/kg IV initially then repeat 10 mg/kg IV q10 min (maximum of 50–60 mg/kg)</td>
</tr>
<tr>
<td>Phenytoin/fosphenytoin</td>
<td>15–20 mg/kg IV</td>
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</table>
vary depending on the available history and physical examination of the neonate. One study found that only 2.5% of patients presenting with an ALTE had positive diagnostic tests [11]. Box 3 provides a list of the more common causes of an ALTE. The workup may include a full sepsis evaluation, electrolytes, chest radiography (CXR), ECG, and respiratory syncytial virus (RSV) and pertussis nasal swabs. Hospitalization for observation and monitoring is appropriate.

**Respiratory emergencies: bronchiolitis**

Although respiratory distress in a neonate is usually obvious, finding the underlying cause may be more challenging. Respiratory symptoms are most commonly a result of pulmonary problems but may be also caused by cardiac, central nervous system, metabolic, endocrine, or gastrointestinal (GI) emergencies. Lower airway abnormalities such as congenital pulmonary malformations, including a diaphragmatic hernia, tracheoesophageal fistula, cystic adenomatous malformation, and congenital lobar emphysema, and upper airway lesions such as laryngoe- or tracheomalacia or airway hemangioma may be the reason for the respiratory distress. Because respiratory failure is the most common cause of cardiac arrest in children, aggressive and early interventions may be life saving.

Bronchiolitis is the most common cause of respiratory disease in children less than 2 years of age [12]. The respiratory syncytial virus is responsible 80% of the time, but other viral causes include the adenovirus, influenza, and parainfluenza
viruses [12]. Epidemics occur usually in the winter and spring months. Presenting symptoms include nasal congestion, tachypnea, wheezing, retractions, and apnea. Apnea may be an early symptom, appearing before any respiratory symptoms have developed.

ED management includes ABC stabilization and an initial CXR. Other diagnostic interventions will be directed by the clinical presentation. Pharmacologic treatment may include a trial of nebulized epinephrine or a β-agonist. Epinephrine may be beneficial in initially improving respiratory distress but has not been shown to significantly shorten the length of stay compared with albuterol [13]. Controversy exists over the use of steroids in children with bronchiolitis. However, although corticosteroids have not been demonstrated to have significant benefit in the management of either mild or severe bronchiolitis [14], there may be a subset of patients who do respond to steroid use. Patients with recurrent episodes of wheezing, a strong family history of atopy, or severe disease on presentation may be candidates for therapy. The overall mortality of previously healthy infants hospitalized with RSV is ≤1%, but those infants who are at high risk (eg, have an underlying disease such as coronary heart disease, chronic lung disease, or prematurity) have an overall mortality of 3.5% [15].

Neonates with bronchiolitis who demonstrate any respiratory distress or apnea should be admitted to a monitored bed for supportive care. In addition, special consideration should be given to infants with comorbid conditions.

Infectious emergencies

As with other neonatal emergencies, the presenting signs and symptoms may range from minor complaints to shock. Fever (≥100.4°F, rectally) should always prompt a full evaluation for sepsis, but other symptoms, including hypothermia and irritability, may be just as concerning. An undeveloped immune system and recent exposure to bacteria present in the birth canal put the neonate at high risk for developing a severe bacterial illness. These infections include sepsis, meningitis, skin infections, pneumonia, and osteomyelitis. A pertinent birth history should include maternal group B streptococcal status, the presence of a sexually transmitted disease, prolonged rupture of membranes, mode of delivery, and any invasive monitoring (eg, a scalp monitor) should increase suspicion for a severe bacterial illness.

Sepsis

A full sepsis evaluation should be initiated in any neonate with a fever or other nonspecific symptoms that do not have an obvious explanation. In fact, finding a possible source (including RSV) should not limit the evaluation. Each sepsis evaluation should include a CBC, blood culture, urinalysis, urine culture (catheterized or suprapubic specimen), CXR, and lumbar puncture for the analysis of cerebrospinal fluid (CSF). In addition, broad-spectrum antibiotics (Table 2)
should be administered, and the neonate should be admitted to the hospital, regardless of normal laboratory and radiographic findings.

**Neonatal herpes**

Neonatal herpes is an uncommon diagnosis but has an increasing incidence, with a rate of 2000 neonatal infections per year in the United States [16]. The symptoms may be subtle, without skin findings, but should not be excluded because 17% to 39% of cases never have skin lesions [17]. A high index of suspicion is important because early detection and treatment with acyclovir, 20 mg/kg IV, has been shown to decrease the mortality from 90% to 31% [18]. Despite this improvement, the long-term morbidity remains high. Treatment should be considered in any patient with fever, irritability, abnormal CSF findings, and especially seizures. Laboratory evaluation should include an analysis of a CSF biopsy for herpes simplex virus by polymerase chain reaction, viral culture, and liver function tests. A CXR may demonstrate pneumonitis. Management should include supportive care, broad-spectrum antibiotics for sepsis, and administration of acyclovir. Critically ill neonates or those with a high suspicion for neonatal herpes should be admitted to a monitored bed and receive critical care support.

**Dermatologic infections**

Neonatal skin infections deserve special mention because a full sepsis evaluation is warranted. The antibiotic coverage should be expanded to include an anti-staphylococcal agent. Nafcillin, 50 mg/kg IV, may be appropriate, but the use of this drug should be determined by the resistance profile in the community.

Omphalitis, an umbilical and periumbilical infection, is considered a surgical emergency if it extends to the peritoneum, and it requires ABC stabilization, fluid resuscitation, IV antibiotics (ampicillin, gentamicin, and flagyl), and immediate pediatric surgical consultation. Any erythema that surrounds the umbilicus and extends to the abdominal wall is suspicious for omphalitis, regardless of the presence of fever, and the infant should undergo a septic evaluation, the initiation of antibiotics, and hospital admission.
Gastrointestinal emergencies

Although acute viral gastroenteritis may present in the neonatal period and should be evaluated with specific concern for hydration status, the life-threatening diseases should always be excluded first. Symptoms of GI emergencies may be subtle, including irritability or feeding intolerance, or they may be more apparent, with vomiting (bilious or nonbilious), abdominal distention, and shock. GI emergencies are discussed more extensively elsewhere in this issue.

Malrotation with midgut volvulus

Any history of bilious emesis should be considered midgut volvulus until it is proven otherwise. Malrotation occurs in 1 in 5000 live births, and approximately 80% of cases present with volvulus in the first month of life [19]. The initial evaluation should include ABC stabilization, fluid resuscitation, nasogastric tube placement, and abdominal radiographs. The most common radiographic finding in the case of malrotation with volvulus is a normal gas pattern [20]. With severe duodenal obstruction, a “double bubble sign” may be present. An upper GI is the study of choice for definitive diagnosis. Immediate pediatric surgical consultation is imperative.

Necrotizing enterocolitis

Necrotizing enterocolitis (NEC) is, classically, a disease of premature neonates but may also present in the term infant. Symptoms may include feeding difficulties, irritability, abdominal distention, and hematochezia. Management includes ABC stabilization, fluid resuscitation, and the administration of antibiotics. Laboratory evaluation should include a CBC, blood culture, and serum electrolytes. The classic radiographic finding is pneumatosis intestinalis or portal air. Free air may be present if the bowel has already perforated. Management should include surgical consultation and admission to a critical care service.

Toxic megacolon

Hirschsprung’s disease occurs in 1 in 5000 live births, with a male:female ratio of 4:1 [21]. This condition results from the failure of neural crest cells to migrate in the colon, resulting in an aganglionic section of bowel. This diagnosis should be considered in the presence of constipation or failure to pass meconium in the first 24 hours of life.

Enterocolitis (toxic megacolon) may develop in these patients and may present similarly to NEC. The management and stabilization are also similar. A radiograph may demonstrate an enlarged or dilated section of colon. These patients also warrant a surgical consultation and should receive critical care management.
Hyperbilirubinemia

Neonatal hyperbilirubinemia (jaundice) is a common newborn finding that, although usually is benign, may represent a more serious underlying diagnosis. The level of concern will be directed by the age of presentation, associated clinical symptoms, and most importantly, if the bilirubin is primarily unconjugated (indirect) or there is an elevated conjugated (direct) portion. As a result, the initial evaluation should include a total and direct bilirubin test, hematocrit level, reticulocyte count, and Coombs’ test. The causes of unconjugated hyperbilirubinemia are usually physiologic or related to breast feeding but may also include ABO or other minor blood group incompatibilities, spherocytosis, glucose-6-phosphate dehydrogenase deficiency, sepsis, acidosis, lethargy, temperature instability, or albumin < 3.0 g/dL. Guidelines are for phototherapy use, which is indicated when total serum bilirubin values exceed those values within the table.


Table 3
Management of hyperbilirubinemia in newborns

<table>
<thead>
<tr>
<th>Risk category</th>
<th>Total serum bilirubin (mg/dL)</th>
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<tbody>
<tr>
<td></td>
<td>Postnatal time</td>
</tr>
<tr>
<td></td>
<td>24 h</td>
</tr>
<tr>
<td>High risk (35–37 wks gestational age + risk factors)</td>
<td>8</td>
</tr>
<tr>
<td>Medium risk (≥38 wks gestational age + risk factors, or 35–37 wks and well)</td>
<td>10</td>
</tr>
</tbody>
</table>

For photo-therapy in infants ≥ 35 weeks’ gestation
Total bilirubin value is used
Risk factors include: isoimmune hemolytic disease, asphyxia, G6PD deficiency, sepsis, acidosis, lethargy, temperature instability, or albumin < 3.0 g/dL

Table 4
American Academy of Pediatrics recommendations for phototherapy and exchange transfusion in the healthy term (≥ 38 wk) neonate

<table>
<thead>
<tr>
<th>Age (h)</th>
<th>Phototherapy (g/dL)</th>
<th>Exchange (g/dL)</th>
</tr>
</thead>
<tbody>
<tr>
<td>24</td>
<td>12</td>
<td>19</td>
</tr>
<tr>
<td>48</td>
<td>15</td>
<td>22</td>
</tr>
<tr>
<td>72</td>
<td>18</td>
<td>24</td>
</tr>
<tr>
<td>≥ 96</td>
<td>20</td>
<td>25</td>
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Presence of ill-appearing infant, unstable vital signs, lethargy, apnea, tachypnea, fever, poor feeding, or behavior change

Are there other signs of sepsis, like fever or omphalitis, infected circumcision, etc

Are there signs of GI obstruction, e.g. bilious or persistent vomiting, abdominal series showing air-fluid levels, smooth bowel walls?

Complete sepsis workup indicated ** Sepsis workup probably indicated in ALL neonates with above symptoms regardless of final etiology

Consider hypothyroidism, congenital adrenal hyperplasia, or inborn errors of metabolism ** can present with vomiting

Is there a family history or ethnic origin associated with jaundice or hemolytic anemia??

Order G6PD screening, RBC smear, reticulocyte count; consider G6PD, hereditary spherocytosis or elliptocytosis

Does Mom have diabetes?

Is total bilirubin > 12 mg/dl, increasing faster than 0.5 mg/dl/hr or lasting greater than one week??

Cause is physiologic neonatal hyperbilirubinemia:
If bilirubin > 20 mg/dl, admit for phototherapy.
If lower, stop breast-feeding temporarily, expose infant to sunlight if possible, and recheck in 24 hours

Cause is probably breast-feeding jaundice or breast milk jaundice in addition to physiologic neonatal hyperbilirubinemia; Stop breast feeding temporarily; Also must consider other causes such as Crigler-Najjar syndrome or Lucey-Driscoll syndrome

Is the Coomb's Test positive?

Then consider Maternal Fetal Blood Incompatibility, e.g. Rh, blood type or maternal immune antibodies

Is the infant anemic?

Is there a cephalohematoma, large ecchymosis, intracranial bleed, or polycythemia?

Cause due to hemolysis or polycythemia

Cause due to Maternal diabetes-related jaundice

Workup for volvulus, duodenal atresia, pyloric stenosis, Hirschsprung’s or other GI obstructions

Order G6PD screening, RBC smear, reticulocyte count; consider G6PD, hereditary spherocytosis or elliptocytosis

Does Mom have diabetes?

Is total bilirubin > 12 mg/dl, increasing faster than 0.5 mg/dl/hr or lasting greater than one week??

Cause is physiologic neonatal hyperbilirubinemia:
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Cause is probably breast-feeding jaundice or breast milk jaundice in addition to physiologic neonatal hyperbilirubinemia; Stop breast feeding temporarily; Also must consider other causes such as Crigler-Najjar syndrome or Lucey-Driscoll syndrome
of concern, and the differential diagnosis includes biliary atresia or obstruction, hepatitis, and α₁-antitrypsin deficiency. The current management criteria for unconjugated hyperbilirubinemia are outlined in Tables 3 and 4. Fig. 1 shows an algorithm for the evaluation of these neonates. Hospitalization and further evaluation of infants with conjugated hyperbilirubinemia typically is warranted.

### Metabolic emergencies: inborn errors of metabolism

Inborn errors of metabolism (IEM) are uncommon but remain a source of anxiety in considering the diagnosis. Each state screens for specific defects of

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**Box 4. Presenting signs and symptoms of an inborn error of metabolism**

**Subtle**

- Abnormal Tone
- Irritability
- Poor feeding or feeding refusal
- Poor weight gain
- Somnolence
- Tachycardia
- Tachypnea
- Vomiting

**Overt**

- Acidosis
- Altered thermoregulation
- Apnea
- Arrhythmia
- Cardiomyopathy
- Dehydration
- Lethargy or Coma
- Persistent hypoglycemia
- Poor perfusion or hypotension
- Seizures
- Sudden unexplained death

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Fig. 1. Inborn errors of metabolism. Exceptions have been noted to the pathway as shown. (Courtesy of Ken Kwon, MD, University of California, Irvine, CA).
metabolism, and the only consistency from state to state is screening for phenylketonuria, congenital hypothyroidism, and galactosemia [22]. A high index of suspicion is important because the symptoms may be subtle, and early recognition and interventions may significantly affect long-term morbidity. Box 4 summarizes the subtle and overt symptoms that may suggest an IEM.

The initial ED management should include ABC stabilization and a bedside blood glucose test. Laboratory evaluation should include serum electrolytes, pH level, lactate, ammonia, CBC, liver function tests and urinalysis for reducing substances, and ketones. The presence of ketones in the neonate’s urine should increase suspicion for an IEM because the errors are typically inefficient producers of ketones even in the presence of hypoglycemia [23]. Other diagnostic tests should include blood and urine for amino and organic acids. The presence or absence of hyperammonemia and pH level are helpful diagnostically in differentiating between several defects. Fig. 2 provides a diagnostic pathway for normal and elevated serum ammonia. The correction of electrolytes (specifically providing parenteral glucose to prevent catabolism), fluid resuscitation, admission to the hospital for further evaluation, discontinuation of all feedings, and consultation with a pediatric geneticist are all important steps in the initial management of an IEM.

Note: exceptions to above pathway exist.
Courtesy of Ken Kwon, MD. University of California, Irvine
Endocrine emergencies

Congenital adrenal hyperplasia

The most common cause of congenital adrenal hyperplasia (CAH) is the result of a deficiency in the 21-hydroxylase enzyme [24]. There is no routine screening in all states for this defect, and male neonates may not have any physical genitalia abnormalities that are noticed in the newborn nursery. As a result, these infants with an unrecognized diagnosis of CAH may present to the ED with shock during the first 2 weeks of life. An initial evaluation that reveals hyponatremia, hyperkalemia, and hypotension unresponsive to fluids or inotropes should immediately prompt the consideration of CAH. Hypoglycemia is also a common finding. Management other than ABC stabilization should include replacement of steroids with hydrocortisone, 25 to 50 mg/m$^2$ IV [4]. The infant with CAH has a deficiency of steroids, which disables a normal stress response. These infants should be admitted to an appropriate facility that provides pediatric subspecialty and critical care support.

Thyrotoxicosis

Infants born to mothers with Graves’ disease may develop thyrotoxicosis and present with delayed symptoms to the ED. Symptoms at presentation may include poor feeding, irritability, tachycardia, respiratory distress, hyperthermia, or congestive heart failure. In the presence of these symptoms, laboratory evaluation should include thyroid functions tests. Treatment may require propranolol, 0.25 mg/kg IV, to control tachycardia. In addition, propylthiouracil, 1.25 mg/kg IV, followed by Lugol’s solution (1–5 drops orally) should be administered to help control the hypermetabolic state and hormone release [24]. These patients, once stabilized, should be admitted to a pediatric subspecialty hospital.

Cardiac disease

Acyanotic heart disease

Clinical decompensation in acyanotic heart diseases may be a result of closure of the ductus arteriosus (DA). The onset of symptoms typically is gradual, with the onset of congestive heart failure. Different degrees of obstruction to the left ventricular outflow tract are present that result in an increase in pulmonary blood flow and a gradual development of heart failure. The classic triad of symptoms for pediatric congestive heart failure is tachypnea, tachycardia, and hepatomegaly. In addition, there may be a history of poor feeding, sweating or color change with feedings, and poor weight gain. Lower extremity edema and jugular venous distention are unlikely findings at this age. The more common causes of acyanotic heart disease are listed in Box 5 [25].
The evaluation should include a CXR, ECG, and serum electrolytes. The initial management may also be administration of prostaglandin E (PGE)\(_1\), but success is less likely because the development of heart failure is gradual and the DA may already have been closed for several days to weeks. The first-line choice in pharmacologic management of congestive heart failure is furosemide, 1 mg/kg IV; however, other adjuvants include dopamine, dobutamine, and digoxin. Pediatric cardiology consultation should be obtained, and once stabilized, the patient should be transported to the appropriate facility.

Cyanotic heart disease

Cyanotic congenital heart defects that are not detected in the newborn nursery will present during the first 2 to 3 weeks of life when the DA closes. With the

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**Box 5. Causes of acyanotic heart disease that present with congestive heart failure**

- Anemia
- Aortic atresia
- Aortic stenosis
- Arteriovenous malformation
- Coarctation of the Aorta
- Complete arteriovenous canal
- Cor pulmonale caused by bronchopulmonary dysplasia
- Endocardial cushion defect
- Hypoplastic left heart
- Interrupted aortic arch
- Mitral valve atresia
- Patent ductus arteriosus
- Truncus arteriosus
- Ventricular septal defect

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**Box 6. Causes of cyanotic heart disease: the five “terrible Ts” (and one “S”)**

1. Transposition of the great vessels
2. Total anomalous pulmonary venous return
3. Tetralogy of Fallot
4. Truncus arteriosus
5. Tricuspid atresia
6. Severe pulmonic stenosis
neonate’s first breath, oxygen and other mediators stimulate the closure of the DA. Functional closure of the ductus occurs in the first 10 to 14 hours of life, but anatomic closure may not occur until 2 to 3 weeks of age because of prematurity, acidosis, and hypoxia [26]. The congenital heart defects that classically present with cyanosis, commonly referred to as the “terrible Ts,” are listed in Box 6.

The immediate goal in evaluating a cyanotic neonate is to differentiate between cardiac and noncardiac causes. The classic hyperoxia test is carried out by obtaining an arterial blood gas (ABG), then placing the patient on 100% oxygen for 10 minutes and then by repeating the ABG. If the cause of cyanosis is pulmonary, the PaO₂ should increase by 30 mm Hg, but if the cause is cardiac, there should be minimal improvement in the PaO₂. The initial ABG should be obtained with co-oximetry because methemoglobinemia may also cause cyanosis in the neonatal period. A simpler method for completing the hyperoxia test is to provide 100% oxygen and observe the oxygen saturation on pulse oximetry for an increase of 10% in pulmonary causes and minimal change with cyanotic heart disease. A CXR and ECG should also be obtained, but these modalities usually are not specific for the diagnosis of congenital heart disease. Although it is not routinely available in the ED, an echocardiogram is diagnostic.

If the neonate’s oxygen saturation or PaO₂ fail to improve and cyanotic heart disease is suspected, then PGE₁ should be administered as a bolus of 0.05 µg/kg IV followed by an infusion of 0.05 to 0.1 µg/kg /min IV [4]. Age-appropriate airway equipment should be immediately available before starting PGE₁ because a non-dose-dependent side effect of PGE₁ is apnea, which requires intubation and mechanical ventilation. It may take 10 to 15 minutes for a response to PGE₁, and its effect can be recognized by an increase in oxygen saturation. The patient should be weaned from supplemental oxygen as soon as possible after the oxygen saturation has improved. After stabilization, the patient should be transported to an appropriate facility for pediatric cardiology and pediatric cardiovascular surgery consultation.

Summary

The differential diagnosis for each subtle or nonspecific symptom in the neonatal evaluation is extensive. This review categorizes the more common life-threatening illnesses by system, in an attempt to provide a guide to the initial recognition, evaluation, and emergency department management. It is important to keep a high index of suspicion when evaluating the neonate because some initial interventions may be life saving.

References