Pediatric Emergency Medicine Reports

The Practical Journal of Pediatric Emergency Medicine

**Volume 16, Number 4 / April 2011** **www.ahcmedia.com**

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**Statement of Financial Disclosure**

To reveal any potential bias in this publication, and in accordance with Accreditation Council for Continuing Medical Education guidelines, we disclose that Dr. Dietrich (editor), Dr. Skrainka (CME question reviewer), Dr. Tucker (author), Dr. Cagle (author), Dr. Eyal (peer reviewer), Ms. Mark (executive editor), and Ms. Hamlin (managing editor) report no relationships with companies related to the field of study covered by this CME activity.

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# Neonatal Emergencies

*The neonatal population (birth to 1 month of age) provides a unique and difficult challenge for diagnosis and treatment in the emergency department, and a systematic approach is critical to allow for rapid diagnosis and subsequent therapy in the setting of a potentially sick neonate. The following is a review of the recognition, evaluation, and treatment of the most common and potentially life-threatening neonatal emergencies by system.* (See Table 1.)

*— Ann Dietrich, MD, FAAP, FACEP*

## Cardiovascular Emergencies

**Congenital Heart Disease.** Congenital heart disease can be differentiated into two specific categories: cyanotic and acyanotic heart disease.

To help differentiate between cardiac and noncardiac causes of cyanosis, the hyperoxia test should be performed.[1] This test involves obtaining an arterial blood gas (ABG) on room air, then obtaining a second ABG after placing the patient on 100% oxygen for 10 minutes. If the $PaO_2$ is less than 100 mmHg after placement on 100% $O_2$, then the cyanotic neonate most likely suffers from congenital heart disease. If the $PaO_2$ rises to greater than 200 mmHg, then the etiology of the cyanosis is likely respiratory.[1]

Initial evaluation of the child with suspected congenital heart disease should also include four extremity blood pressures, preductal and postductal oxygen saturations, chest radiograph, electrocardiogram, and emergent cardiology consult for a diagnostic echocardiogram.[1] When obtaining blood pressures in all four extremities, a difference between upper-extremity blood pressures and lower-extremity blood pressures of more than 20 mmHg should raise suspicion for coarctation of the aorta. If oxygen saturation in the right arm is normal (i.e., normal preductal saturation) and saturation in one of the legs is low (i.e., low postductal saturation), then a right-to-left shunting heart lesion should be considered. If the preductal saturation is low and the postductal saturation is higher, then there should be concern for transposition of the great vessels. Ductal-dependent lesions are best organized into two categories: left-sided (ductus preserves systemic blood flow) and right-sided lesions (ductus preserves pulmonary blood flow). Infants with ductal-dependent lesions may be deceivingly normal while the ductus remains open, but as the ductus begins to close, the infant can begin to show signs of hypoxemia and cardiogenic shock. If the infant has a right-sided ductal-dependent lesion, such as transposition of the great vessels, closure of the ductus will produce severe cyanosis and tachypnea, which will ultimately progress to acidosis and shock if the hypoxemia is not corrected. If the infant has a left-sided ductal-dependent lesion (such as critical coarctation of the aorta), closure of the ductus will result in profound hypotension, tachycardia, and a metabolic acidosis. The ductal-dependent lesions are categorized in Table 2.

Opening or re-opening the ductus arteriosus is achieved by administration of prostaglandin E1 (PGE1), a life-saving intervention in the neonate who

## Executive Summary

- To assist with the differentiation between cardiac and noncardiac causes of cyanosis, the hyperoxia test should be performed.
- The examination of a child with NAT to the abdomen may range from normal to multiple obvious contusions and distention.
- Only one in five children with disseminated herpes will go on to develop the classic vesicular rash.
- The diagnosis of inborn errors of metabolism is difficult and requires a high degree of suspicion in the neonatal period.

presents in cardiogenic shock. The initial dose of PGE1 is 0.05 mcg/kg/dose as a bolus, followed by 0.05-0.1 mcg/kg/minute as a continuous infusion. The only congenital heart lesion for which PGE1 could be potentially harmful is total anomalous pulmonary venous return. In this specific abnormality, re-opening the ductus can lead to increased blood flow from the systemic system directly into the anomalous pulmonary veins, shunting blood away from the systemic circulation. When administering PGE1, the physician must be aware that apnea may develop, and proper monitoring equipment as well as bag mask ventilation and advanced airway equipment should be readily available at the bedside. Other potential, though less harmful side effects, include flushing and fever.

Acyanotic heart disease usually manifests itself in the form of congestive heart failure (CHF).[2] Common causes of acyanotic heart disease are ventricular septal defect (VSD), atrial septal defect (ASD), patent ductus arteriosus (PDA), and coarctation of the aorta. Infants with CHF as a result of an acyanotic cardiac lesion generally present with worsening tachypnea, poor/slow feeding, sweating, and poor weight gain.[2] Treatments for CHF include furosemide 1 mg/kg IV and cardiotropic medications, such as dopamine (5-15 mcg/kg/min) and dobutamine (2.5-15 mcg/kg/min).[2]

### Table 1. Neonatal Emergencies by System

| | |
|---|---|
| Cardiovascular | Congenital heart disease (cyanotic, acyanotic), congestive heart failure |
| Respiratory | Apnea, hypoxia, obstruction, non-accidental chest trauma |
| CNS | Trauma (non-accidental and accidental), jaundice, seizures |
| GI | Intestinal catastrophes (volvulus, intussusception, hypertrophic pyloric stenosis), acute gastroenteritis with hypovolemia, nonaccidental abdominal trauma |
| Infectious | Serious bacterial infections (urinary tract infections, pneumonia, bacteremia, meningitis) |
| Endocrine | Inborn errors of metabolism, congenital adrenal hyperplasia |
| Electrolyte | Hyponatremia, hypoglycemia, hypocalcemia |

## Respiratory Emergencies

**Apnea/Acute Life-threatening Event.** Apnea is defined as the cessation of breathing for > 20 seconds with associated color changes or bradycardia.[3] The differential diagnosis is extensive, with gastroesophageal reflux being the most common cause; the ED physician must consider all potentially life-threatening causes, including sepsis, hypoxia, bronchiolitis, croup, pneumonia, hypothermia, hypoglycemia, anemia, nonaccidental trauma, pertussis, dysrhythmias, and seizures.[4] Initial evaluation and treatment should include securing the ABCs and placing the patient on a cardiorespiratory monitor with supplemental oxygen therapy, if indicated. Depending upon the history, one should consider a complete blood count with differential, urinalysis, body-fluid cultures (blood, urine, and CSF), an electrolyte panel, bedside glucose evaluation, fluorescent antibody testing for respiratory syncytial virus (RSV), and neuroimaging.[3] Appropriate therapy should be instituted based upon the presumptive cause of the apnea (e.g., antibiotics for sepsis and pneumonia, glucose administration for hypoglycemia, a trial of bronchodilators for bronchiolitis, ICP management or surgical intervention in the case of non-accidental head injury).

**Airway Obstruction.** Airway obstruction can be separated into infectious and noninfectious causes, as well as upper and lower airway causes. The infectious causes include viral bronchiolitis, pertussis, croup, pneumonia, and bacterial tracheitis. Noninfectious causes include choanal atresia/stenosis, airway foreign bodies, laryngotracheomalacia, vocal cord paralysis, tracheal stenosis, vascular rings, airway webs, and masses in and around the airway.[5] The infectious and noninfectious causes can be further subdivided into upper and lower airway obstruction, as illustrated in Table 3.

The initial evaluation and treatment should focus on evaluating and stabilizing the airway. Diagnostic evaluation should include neck and chest radiographs, followed by CT or MRI as indicated. The patient will

also require continuous cardiorespiratory monitoring until the underlying cause can be delineated. On X-ray, a number of signs/findings can help lead the physician toward a specific diagnosis. These findings are summarized in Table 4.

*Laryngotracheomalacia (LTM)* is a disorder of airway stiffness that typically presents with stridor early in infancy. It is usually benign and requires no intervention, and can be differentiated from other upper airway obstructions by placing the infant in the prone position; if the stridor improves, LTM is the most likely cause.[5] In severe cases with desaturations or recurrent hospitalizations, ENT consultation should be considered for possible surgical intervention.

*Choanal atresia/stenosis* is characterized by desaturations or apnea while the neonate is comfortable or asleep, followed by improvement of saturations to baseline when the infant cries. If the physician suspects this diagnosis, then he or she should attempt nasogastric tube placement. Inability to place the nasogastric tube successfully helps confirm the diagnosis.[5]

*Pertussis*, also known as whooping cough, is a respiratory infection caused by the bacteria *Bordetella pertussis*. In the United States, children are routinely vaccinated against this pathogen via the diphtheria/tetanus/pertussis vaccine, although immunogenicity only lasts 3-5 years post-vaccination. Small infants are more likely to have severe illness, require hospitalization, and develop complications from the infection. Symptoms occur in 3 stages: the catarrhal stage, the paroxysmal stage, and the convalescent stage. The catarrhal stage, the most infectious stage of illness, presents similarly to common upper respiratory infections. Symptoms include nasal congestion, rhinorrhea, sneezing, and low-grade fever. The paroxysmal stage is characterized by coughing episodes that can last up to several minutes, with a "whoop" sound punctuating the paroxysms of coughing. It is from this "whoop" sound that the layperson diagnosis of "whooping cough" originated. Young infants and neonates do not always exhibit the stereotypical whoop, but can present with frank apnea; they are also at great risk of exhaustion in the event that they are experiencing paroxysmal coughing spells. The convalescent stage is characterized by a chronic cough that may last several weeks. Treatment of pertussis usually occurs during the paroxysmal stage because patients are generally diagnosed during this phase. Although antibiotic therapy does not decrease the severity or duration of illness, it does decrease the bacterial load in the airway and, therefore, decreases the spread of infection.[6] First-line antibiotics are macrolides, including erythromycin and azithromycin.[7] Infants younger than one month of age should receive azithromycin because it carries less risk of development of hypertrophic pyloric stenosis when compared to erythromycin. Second-line antibiotics include trimethoprim-sulfamethoxazole for patients with macrolide allergies.[7]

*Croup*, or laryngotracheobronchitis, is a viral upper respiratory tract infection. It is most commoly caused by the parainfluenza virus.[8] It generally is a benign illness, but can become life-threatening if severe enough. Common symptoms include hoarseness, barking cough (referred to as "seal-like"), and possibly respiratory distress.[8] A patient with suspected croup should be placed on cardiorespiratory monitoring and continuous pulse oximetry to evaluate for early hypoxia and impending respiratory failure. Historically, cool mist administration was utilized as a mainstay of treatment because it was believed that mist moistens airway secretions, soothes irritated mucosal tissue, and decreases mucous viscosity. Recent studies have not shown any difference in response among

### Table 2. Ductal-dependent Lesions

| Right-sided Heart Lesions (Ductus preserves pulmonary blood flow) | Left-sided Heart Lesions (Ductus preserves systemic blood flow) |
|---|---|
| Transposition of the great vessels | Coarctation of the aorta |
| Tricuspid atresia<br>Tetralogy of Fallot | Interrupted aortic arch<br>Hypoplastic left heart syndrome |
| Pulmonary stenosis or atresia | |

### Table 3. Airway Obstructions

| Upper Airway Obstructions | Lower Airway Obstructions |
|---|---|
| Laryngomalacia** | Tracheomalacia** |
| Choanal atresia/stenosis | Bronchiolitis |
| Foreign body aspiration** | Foreign body aspiration** |
| Pertussis | Pneumonia |
| Croup | Vascular rings |
| Bacterial tracheitis | |
| Airway masses** | Airway webs and masses** |
| Vocal cord paralysis/dysfunction** | Vocal cord paralysis/dysfunction** |

** Indicates diagnoses that can have a component of both upper and lower airway obstruction.

## Table 4. Radiographic Findings

| Diagnosis | X-ray Finding | Description |
|---|---|---|
| Croup | Steeple sign | On anterioposterior (AP) neck X-ray, there is a gradual narrowing of the upper airway that resembles the top of a steeple. |
| Epiglottitis | Thumbprint sign | On lateral view of neck X-ray, the epiglottic swelling resembles a thumbprint. |
| Foreign body | Persistent airway expansion or overexpansion | Ideally, one should obtain both inspiratory and expiratory chest films, and the side with the foreign body will appear expanded/overexpanded on both views while the non-affected lung will reveal expansion on inspiration and appropriate collapse on expiration. Given that neonates do not inhale and exhale on command, decubitus films serve a similar function in that normal lungs should collapse when dependent and expand when non-dependent; in the setting of a foreign body, the affected lung would remain similarly expanded whether the lung is dependent or non-dependent. |
| Bronchiolitis | Hyperinflation | On AP CXR the lungs appear hyperinflated, with flattened diaphragms and possible areas of atelectasis. |
| Pneumonia | Focal or diffuse infiltration | On AP CXR the lungs show a focal area of consolidation or may show diffuse patchy consolidation in the case of atypical or viral pneumonias. Also obtain lateral CXR to visualize the retrocardiac area properly. |

those infants with moderate to severe croup who received moist air therapy versus those infants who did not.[9] Treatment modalities of choice today include glucocorticoids, nebulized epinephrine, and heliox therapy.[8] A single dose of dexamethasone at 0.6 mg/kg/dose IM or PO (with a maximum of 16 mg) has been shown to reduce the overall severity of croup when given in the first 4-24 hours after initial symptoms develop.[10] Racemic epinephrine nebulizer treatment is usually reserved for moderate to severe cases of croup with respiratory distress and stridor at rest. Heliox is an inert gas that is a combination of helium and oxygen. Theoretically, it works by increasing laminar flow through the airway, as it is less viscous than room air alone. Current studies dispute its efficacy in the treatment of croup, and it is usually reserved for those patients in whom respiratory distress is severe.[11]

*Bacterial tracheitis* is an uncommon illness in which the upper airway becomes infected at or around the cricoid cartilage level of the trachea. Affected patients initially have croup-like symptoms and may be misdiagnosed at this stage. The two disease entities can be differentiated, because the patient with bacterial tracheitis does not respond to standard croup therapy and is more likely to exhibit respiratory decompensation.[12] *Haemophilus influenzae* type B was historically the most common cause of bacterial tracheitis, but the HIB vaccine has made *H. influenzae* an increasingly rare pathogen. Recent studies have identified *Moraxella catarrhalis* as the most commonly identified bacteria with the most severe course, mandating intubation more often than the other pathogens. Other bacterial causes include: *Staphylococcus aureus*, *Streptococcus pyogenes*, *Streptococcus pneumoniae*, Pseudomonas species, Klebsiella species, *Mycoplasma pneumoniae*, *Mycobacterium tuberculosis*, Peptostreptococcus species, Bacteroides species, Prevotella species, and some anaerobes.[13] Treatment should begin with stabilization of the airway. If there are concerns for impending respiratory failure, immediate consultation with ENT, anesthesia, or a pediatric intensivist is warranted. Initial antibiotic coverage should be broad and include a third-generation cephalosporin and penicillinase-resistant penicillin (oxacillin or nafcillin) or vancomycin to cover for methicillin-resistant *Staphylococcus aureus*.

*Vocal cord paralysis/dysfunction* is uncommon in the neonatal population but can occur as a result of previous intubations. It may present with symptoms of both intrathoracic and extrathoracic obstruction, manifesting as both inspiratory and expiratory stridor. If the clinician is suspicious of this diagnosis, it is imperative that the airway be carefully evaluated for patency, and an otolaryngologist should be consulted for possible diagnostic laryngoscopy.

*Bronchiolitis* is an acute viral infection of the bronchioles that causes increased mucus production, inflammatory reaction and cell death, and sloughing of the cellular debris. The increased debris and inflammation of the airway lead to runny nose, cough, tachypnea, and retractions. Symptoms will worsen over the first 3-5 days, and begin to resolve by 1 week. Bronchiolitis usually occurs in infants 2 to 24 months of age,

with the most common etiology being RSV. Epidemiologically, RSV bronchiolitis occurs in the fall and winter months, and infants younger than the age of 6 months are at particularly high risk for morbidity and mortality. This age group is particularly susceptible to the development of apnea during the acute phase of infection; therefore, clinicians should consider admission for neonates with RSV infection early in the illness. Treatment is primarily supportive, including oxygen therapy for the hypoxia, aggressive nasal suctioning after administration of nasal saline drops,[13] and intravenous fluids for those babies unable to take oral fluids. Some literature suggests that nebulized albuterol (2.5 mg-5 mg jet nebulizer) and nebulized racemic epinephrine (0.25-0.5 mL jet nebulizer of 2.25% epinephrine) have been used, with reduction of symptoms in roughly 10% of cases.[14] Current recommendations include giving a trial of these medications and, if there is improvement with either therapy, then it can be continued as needed.

*Vascular rings* are abnormal configurations of the aortic arch or associated vessels that form a ring around the trachea and esophagus. Symptoms include reflux, vomiting, weight loss, stridor, cyanosis, and respiratory distress that progressively worsen as the infant ages. The evaluation of choice is an upper GI to look for any compression of the esophagus. In diagnostically difficult cases, chest CT with three-dimensional reconstruction of the aorta can demonstrate the full vascular ring ensnaring the trachea and esophagus. Treatment is surgical, so consultation with a cardiothoracic surgeon is warranted.

**Intrathoracic Nonaccidental Trauma (NAT).** The most common nonaccidental intrathoracic emergency is from multiple rib fractures. Anterior and posterior rib fractures are more specific for NAT than lateral rib fractures. In fact, a recent systematic review by Kemp et al showed that neonatal anterior or posterior rib fractures are 71% specific for NAT.[15]

## CNS Emergencies

**Trauma (Non-accidental and Accidental).** Evaluating a neonate with a suspected head injury is challenging. The history may be nonspecific and, in the case of nonaccidental trauma, may be vague and change frequently with repeat interviews. Furthermore, neonates with head trauma may exhibit subtle physical exam abnormalities or may appear completely normal. Common symptoms include lethargy, irritability, seizures, increased or decreased tone, impaired consciousness, vomiting, poor feeding, breathing abnormalities, and apnea.[16] In fact, seizures are reported in 40-70% of all shaken babies.[17] Approximately half of all patients with the shaken baby syndrome have severe impairment, are unresponsive, have opisthotonos, or are moribund.[16]

The most common types of head injuries include skull fractures, subdural hematomas, and epidural hematomas. Skull fractures result directly from the trauma and may be associated with intracranial hemorrhages. Subdural hematomas are seen below the inner dural layer, external to the brain and arachnoid membrane. They can be classified as acute (rapidly occurring after the injury), subacute (3-7 days after injury), or chronic (2-3 weeks after initial injury). They occur when the intracranial bridging veins become sheared, either from direct insult from the trauma or from indirect (contra-coup) insult as the brain is shaken inside the skull. As the hematoma expands, it places direct pressure on the brain and leads to an increase in the intracranial pressure (ICP). Because the bleeding is venous in nature, the patient usually has a slower neurologic decline when compared to those patients with epidural hematomas.

Epidural hematomas usually arise from a direct blow to the head and are generally associated with an overlying skull fracture. The bleeding usually results from damage to the middle meningeal artery or its dural branches. This brisk arterial bleeding leads to rapid hematoma accumulation, causing abrupt increases in ICP and rapid decline in neurologic status. Early diagnosis and subsequent aggressive treatment can improve long-term outcomes;[18] therefore, the workup of a neonate with suspected abuse or accidental head trauma, after initial stabilization, should always start with imaging. Computed tomography (CT) remains the best initial screening tool,[19] although cranial ultrasound and magnetic resonance imaging (MRI) are possible alternatives to detect intracranial bleeding. Cranial ultrasound can only be used for infants with an open fontanelle, and a negative ultrasound does not exclude intracranial pathology.[19] MRI is more sensitive and can provide details about anatomy, vascular structures, and myelination; however, it is much more expensive and more difficult to obtain in emergency situations.[20]

Emergency department management of head injury should include stabilization of airway, breathing, and circulation; determination of a bedside glucose; and regulation of temperature. If an intracranial bleed is suspected, then complete blood count, platelet count, prothrombin time, and partial thromboplastin time should be obtained.[21] After initial stabilization, neuroimaging should be performed emergently, and neurosurgical consultation obtained as warranted.[21]

It is imperative to recognize quickly the signs of increased ICP and impending hernation. These signs include hypertension, bradycardia, and respiratory abnormalities (either bradypnea or tachypnea), the trio known as Cushing's triad. Other signs include either unilateral or bilateral mydriasis that is unreactive to light. A blown pupil indicates compression or distortion of cranial nerve III, which occurs in head trauma as the brain herniates through the foramen magnum. Patients with severe head injury should be intubated immediately, and medical therapy for increased intracranial pressure should be

## Table 5. Medical Therapy for Increased Intracranial Pressure

| Treatment | Dose | Mechanism of Action |
|---|---|---|
| Head elevation and midline position | 30 degrees | Improves CSF and venous drainage[47] |
| Hyperventilation | Goal end-tidal $CO_2$ of 35. | Current recommendations are short, intermediate episodes of hyperventilation only in times of impending herniation.[12] It causes an immediate effect of decreasing ICP, but the effect is transient. Also it is unknown if there is any effect on long-term outcome. |
| Mannitol | 0.5-1 gm/kg/dose IV | Works by two mechanisms. First, mannitol causes a decrease in blood viscosity, leading to reflex vasoconstriction intracranially, thereby decreasing intracranial pressure. Second, it induces hyperosmolarity and, therefore, an extracellular fluid shift. Mannitol may accumulate in areas of cerebral vascular interruption, potentially leading to reverse osmosis with increased intracranial pressure in these areas. |
| Hypertonic saline (multiple concentrations, most common is 3%) | Bolus 3% saline 2-4 mL/kg/dose followed by infusion of 0.1-1 mL/kg/hr to keep ICP < 20 | Hyperosmolar agent that works by causing an extracellular shift of fluid, therefore decreasing intracellular edema. It has been demonstrated that it also decreases overall fluid requirement, decreases therapies in the first 3 days, decreases the duration of mechanical ventilation, and decreases the length of hospitalization.[31-33] |
| Barbiturates (drug-induced comas) | Generally, pentobarbital 3-10 mg/kg loading dose followed by 1-2 mg/kg/hr infusion | It has been shown to decrease excitotoxicity, decrease ICP, and increase brain oxygenation.[34-36] |

instituted as outlined in Table 5.

A new potential therapy that is currently under experimental trials is induction of hypothermia. The goal is to keep core temperatures between 32-34°C. One study showed a 20% improvement in ICP in the hypothermic group as compared to the control group.[22] This therapy is still very controversial and needs more supporting evidence before universal implementation.

**Hyperbilirubinemia.** Jaundice may be a normal part of the neonatal period or a pathologic finding, and it is the ED physician's responsibility to distinguish between physiologic bilirubin elevation and a potentially fatal bilirubin elevation. The ultimate goal is the prevention of kernicterus, which is defined as abnormal accumulation of unconjugated bilirubin in the brain. Symptoms of kernicterus include lethargy, high-pitched crying, and decreased muscle tone with intermittent periods of increased muscle tone. As the condition progresses, the newborn may exhibit fever and opisthotonos.

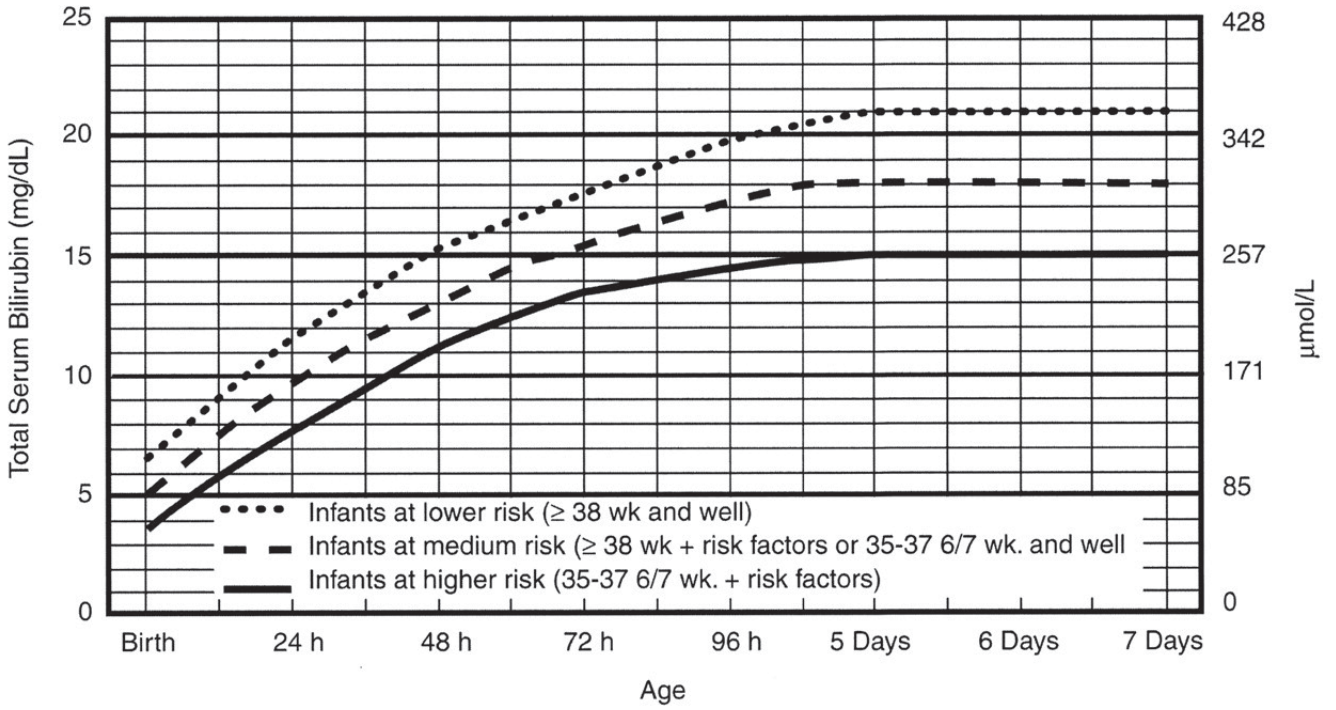
## Table 6. Conjugated vs. Unconjugated Hyperbilirubinemia

| Conjugated Hyperbilirubinemia | Unconjugated Hyperbilirubinemia |
|---|---|
| Biliary atresia | Breastfeeding vs. breast-milk jaundice |
| Alpha-1-antitrypsin deficiency | ABO incompatibility |
| Hepatitis | Sepsis |
| Biliary cholestasis | Glucose-6-phosphate dehydrogenase deficiency |
| | Hereditary spherocytosis |
| | Gilbert's and Crigler-Najjar syndrome |

The differential diagnosis of jaundice should be separated into diseases that cause conjugated (direct) hyperbilirubinemia and those that cause unconjugated (indirect) hyperbilirubinemia. Conjugated hyperbilirubinemia is defined as > 2 mg/dL direct bilirubin or > 20% of the total bilirubin.[23] (*See Table 6 for details.*)

No clear total bilirubin level defines when kernicterus will occur, but according to the U.S. Kernicterus Registry, the total bilirubins of affected individuals ranged from 20.5 to 59.9. The U.S. Kernicterus Registry defines two phases of symptoms: acute and chronic. Acute kernicterus is characterized by irritability, hypertonia with early opisthotonos, drowsiness, poor feeding, high-pitched cry, alternating tone, or failed hearing screens. Chronic kernicterus sequelae include extrapyramidal movement disorders, gaze abnormalities, auditory disturbances, intellectual deficits, and enamel dysplasia of deciduous teeth. In 2004 the AAP released a practice guideline that provided guidance for initiation

## Figure 1. AAP Nomogram for Initiating Phototherapy




- Use total bilirubin. Do not subtract direct reacting or conjugated bilirubin.
- Risk factors = isoimmune hemolytic disease, G6PD deficiency, asphyxia, significant lethargy, temperature instability, sepsis, acidosis, or albumin < 3.0g/dL (if measured)
- For well infants 35-37 6/7 wk can adjust TSB levels for intervention around the medium risk line. It is an option to intervene at lower TSB levels for infants closer to 35 wks and at higher TSB levels for those closer to 37 6/7 wk.
- It is an option to provide conventional phototherapy in hospital or at home at TSB levels 2-3 mg/dL (35-50mmol/L) below those shown but home phototherapy should not be used in any infant with risk factors.

**Reprinted with permission from: Subcommittee on Hyperbilirubinemia. Management of hyperbilirubinemia in the newborn infant 35 or more weeks of gestation. *Pediatrics* 2004;114:297-316. American Academy of Pediatrics**

of phototherapy based on risk factors and gestational age.[24] See Figure 1 for the nomogram as provided by AAP.

In the practice guideline, the AAP recommends that for infants greater than 35 weeks gestation, exchange transfusions should be strongly considered for total bilirubins of 20-25 mg/dL, based on the post conceptual age and the bilirubin to albumin ratios.[24] The diagnostic evaluation of hyperbilirubinemia should include blood type with Rh factor, Coomb's testing, total and direct bilirubin, reticulocyte count, and a complete blood count with differential. The provider should also strongly consider obtaining blood, urine, and spinal fluid for culture, as sepsis can be a cause of hyperbilirubinemia.

**Seizures.** Neonatal seizures are often very challenging to diagnose. The history is usually difficult to elicit, and parents may report vague symptoms.[25] Since the cortical development of the neonate is incomplete, generalized tonic-clonic seizures may not be present.[26] Particularly concerning historical features include lip smacking, abnormal tongue and eye movements, and apnea. The emergency physician should focus on the potentially life-threatening etiologies, including hypoxia, trauma, intracranial bleeds, inborn errors of metabolism, tumors, sepsis (bacterial and viral), hypoglycemia, hyponatremia, hypernatremia, and hypocalcemia.[27]

Initial stabilization should include placing the patient on a cardiorespiratory monitor, securing the airway, breathing, and circulation, and obtaining electrolytes (including glucose, sodium, and calcium); treatment should be aimed at the underlying cause. Hyponatremic seizures should be treated with 3% saline (Amount of 3% NaCl [mL] = (125 mEql/L – actual serum Na) X (Body weight [kg] X 0.6 L/kg over 60 min). For hypoglycemic seizures, raising the glucose back into the normal range may be sufficient to stop the seizures,[28] and the dose administered should be 5 mL/kg of D10. In nonhypoglycemic seizures, further evaluation should include complete blood count with differential and complete sepsis work-up (blood, urine, and cerebrospinal fluid [CSF] cultures).[25] The first-line treatment for seizures is lorazepam 0.1 mg/kg/dose intravenously. This dose may be repeated twice before moving on to the next agent. The use of phenobarbital versus phenytoin as second-line therapy in neonatal seizures is actively debated,[29] but the

success rate of either agent alone is less than 50%.[30] The dose of phenobarbital is 20 mg/kg IV, then 10 mg/kg every 10 minutes up to a maximum of 50-60 mg/kg; the dose of phenytoin/fosphenytoin is 15-20 mg/kg IV. For refractory seizures, pyridoxine 50-100 mg IV may be given, but the patient should undergo EEG monitoring during administration to determine its effect.[25] Furthermore, broad-spectrum antibiotics and acyclovir should be considered for treatment of potential infectious causes, and all electrolyte abnormalities should be corrected (see electrolyte emergencies section).

## GI Emergencies

**Abdominal Emergencies.** It is often difficult to distinguish between common viral gastroenteritis and intestinal catastrophes because they share vomiting and feeding intolerance as presenting symptoms. One feature that can distinguish gastroenteritis from an intestinal catastrophe is the presence or absence of diarrhea. When faced with an infant with vomiting and no diarrhea, the clinician must consider a catastrophic intestinal event. Some of the more common intestinal emergencies are outlined below.

*Malrotation With Midgut Volvulus.* Malrotation results from incomplete rotation and fixation of the bowel during development, more specifically incomplete rotation around the superior mesenteric artery. Malrotation occurs in 1 out of every 500 infants and is commonly associated with other congenital abdominal malformations, including diaphragmatic hernias, omphalocele, and gastroschisis. Symptoms usually occur within the first year of life, when the malrotation may lead to a volvulus. Volvulus is a twisting of the intestines that results in obstruction of the bowel lumen as well as obstruction of the arterial blood flow to the associated small bowel. Symptoms include abdominal distention, abdominal pain, and acute onset of bilious emesis. If the volvulus persists, then it can lead to hematemesis or hematochezia. Physical exam will show abdominal guarding, peritoneal signs, and abdominal distention. Ultimately, if the volvulus persists, the infant will develop decompensated shock.[31] Initial management should include securing ABCs, making the infant NPO, starting broad-spectrum antibiotics, placing a nasogastric tube to low intermittent suction, initiating fluid resuscitation and vasopressor medication if needed, and requesting a surgical consultation. The first-line vasopressor should be dopamine at 3-5 mcg/kg/min, as this medication increases the splanchnic artery blood flow. Workup should include a complete blood count with differential, arterial blood gas, electrolytes, type and screen, clotting studies, and abdominal X-ray. The diagnostic study of choice is an upper GI series. In a normal study, the contrast will travel down the esophagus and into the stomach. From here it will cross the abdominal midline as it enters the first part of the duodenum. It should then turn down into the middle duodenum, make a right turn, and recross the midline as it passes through the distal duodenum and into the proximal jejunum. This pattern forms the normal duodenal "C" loop. In a case of malrotation, the contrast will exit the stomach normally and cross the midline into the proximal duodenum. It may then make the downturn but will not recross the midline and, therefore, will not complete the full duodenal "C." Finally, if the malrotation has an associated volvulus, the contrast will abruptly end at the level of the volvulus and will not continue to pass into the distal small bowel. Ultimate treatment is a surgical repair called a Ladd's procedure.[32] This procedure is characterized by reduction of the volvulus with mobilization of the small bowel to the right side of the abdomen and the large bowel to the left side. Surgeons also perform an appendectomy.

*Intussusception.* This condition occurs when one portion of the intestine telescopes into a distal portion. The segment that prolapses distally is referred to as the intussusceptum, and the part that receives the intussusceptum is the intussuscipiens.[33] This telescoping obstructs the normal blood flow, leading to acute abdominal pain; the bowel will become necrotic if the obstruction does not resolve spontaneously or is not reduced. Intussusception most commonly occurs at the ileocecal junction, generally affects children 3 months to 6 years of age, and may occur in 1 in 1000 live births.[33] It is more likely to occur in infants after a recent viral illness, and in infants with Meckel's diverticulum, cystic fibrosis, Henoch-Schönlein purpura, intestinal polyps, or any other condition that can lead to development of a lead point. The normal intestinal peristalsis is disrupted in the affected areas, and the adjacent intestines are allowed to telescope into the mass or swollen intestinal section. These areas of disrupted peristalsis are referred to as lead points. In neonates, intussusception is rare but may be seen in the setting of a pre-existing lead point.

The most common symptom is severe, acute, intermittent abdominal pain,[33] often associated with drawing up of the legs. Between these episodes of pain, the infant may behave completely normally. A percentage of children will exhibit intermittent periods of lethargy as their only presenting symptom of intussusception. The pathognomonic "currant jelly stool" represents bowel ischemia but is rarely seen; however, 75% of affected patients will have stools that are occult positive for blood. If an affected infant is febrile, then the clinician should be concerned about the possibility of secondary sepsis. Physical exam may reveal a "sausage-shaped" mass in the abdomen, but the exam is most commonly normal. The diagnostic evaluation should include an abdominal X-ray (may show a paucity of cecal air), immediate surgical consultation, and barium or air contrast enema.[34] The enema is diagnostic and generally therapeutic.[34] If the intussusception cannot be relieved with the enema, then immediate surgical reduction is warranted.

## Table 7. Signs and Symptoms of Dehydration

| Clinical Sign | Mild Dehydration | Moderate Dehydration | Severe Dehydration |
|---|---|---|---|
| Mental status | Crying | Sleepy but arousable | Difficult to arouse to unarousable |
| Mucous membranes | Moist | Dry and tacky | Cracked, dry, bleeding |
| Fontanelle | Flat | Flat to mildly depressed | Extremely sunken |
| Capillary refill | < 2 seconds | 2-4 seconds | > 4-5 seconds and cold |
| Heart rate | Mildly elevated | Tachycardic | Extreme tachycardia |
| Blood pressure | Normal | Normal | Hypotensive |
| Skin turgor | Normal | Normal to slow rebound | Tenting noted |
| Respiratory rate | Normal | Mild elevation | Notable tachypnea |
| Tears | Normal | Decreased but present | No tears |
| Urine output | Normal | Decreased | Anuric |

*Hypertrophic Pyloric Stenosis.* Pyloric stenosis results from progressive hypertrophy of the pyloric sphincter. As the sphincter muscle enlarges, it mechanically obstructs the opening between the stomach and the duodenum.[35] Pyloric stenosis is characterized by progressive nonbilious vomiting that worsens as the infant ages, and, in its early stages, is almost impossible to differentiate from gastroesophageal reflux. As it progresses, however, the vomiting becomes more prominent and projectile in nature.[35] After vomiting, the infant with pyloric stenosis is usually fussy secondary to hunger and, as the process continues, the infant can become significantly dehydrated. The physical exam may reveal a dehydrated-appearing infant with sunken fontanelle, dry mucosal surfaces, and tachycardia. Abdominal exam may reveal an "olive," or a small round mass in the mid to right upper quadrant (near the medial edge of the rectus abdominus muscle), representing the hypertrophied pylorus.[35] Initial work-up should include an electrolyte panel, which may reveal a hypochloremic, hypokalemic metabolic alkalosis if the infant is severely dehydrated. The diagnosis of pyloric stenosis is confirmed with an abdominal or pyloric ultrasound. An ultrasound is considered positive if the serosa to mucosal thickness is greater than 3 mm, if the pyloric canal is greater than 17 mm in length, or if the total thickness of the pylorus is 15 mm or greater. Initial treatment should include correction of electrolyte disturbances, fluid resuscitation, and an immediate surgical consult. The surgical treatment is a pyloromyotomy.[35] Although more common in the second and third months of life, pyloric stenosis can be seen in infants as young as 2-3 weeks of age.

**Acute Gastroenteritis With Hypovolemia.** Acute gastroenteritis is a common complaint in the emergency room, with parents generally reporting vomiting and diarrhea of varying frequencies. The infant may also have decreased urine output by history, and decreased skin turgor, sunken fontanelle, and dry mucosal membranes on physical exam. The degree of dehydration should be estimated to guide the clinician in goals of fluid resuscitation.[36] See Table 7 for signs and symptoms of dehydration.

In the neonatal population, dehydration status is estimated using the 5%, 10%, 15% rule for mild, moderate, and severe dehydration, respectively. Proper rehydration should be approached in two phases[36]: initial volume resuscitation followed by complete replacement of the infant's fluid losses and correction of electrolyte derangements. If the infant is only mildly dehydrated, an oral challenge should be attempted, as enteral rehydration is superior to intravenous rehydration. Generally, 5 mL, or roughly one teaspoon, every minute is tolerated. If the infant is severely dehydrated or fails enteral rehydration, then intravenous rehydration (IV) should be initiated.[36] Initial fluid resuscitation should include a bolus of isotonic fluid (normal saline or lactated Ringer's) at 20 mL/kg, which may be repeated as needed. The goal of initial volume resuscitation is stabilization or improvement in the infant's vital signs, urine output, mental status, and physical exam findings listed in Table 7. During the second phase of rehydration, an isotonic dextrose-containing IV fluid should be utilized, and potassium should also be added if the infant is making adequate urine.[36] The clinician should calculate the estimated total volume loss using the 5%, 10%, 15% rule above. The fluid administered during the initial volume resuscitation should be subtracted from the total deficit, and the deficit should be corrected over 24 hours (half over the first 8 hours and the remaining half over the subsequent 16 hours). It is important to remember that maintenance fluid requirements should be added to deficit replacement during the 24-hour replacement. The dosage of maintenance fluids is 100 mL/kg/day up to 10 kg of weight, then 50 mL/kg/day for the next 10 kg, and then 20 mL/kg/day for the remaining weight (100/50/20 rule).

### Table 8. LEG Mnemonic

| |
|---|
| *L isteria monocytogenes* |
| *E scherichia coli* |
| G roup B Streptococcus |

Another quick estimate is the 4/2/1 rule for the amount of fluid per hour. In this rule, you give 4 mL/kg/hr for the first 10 kg, then 2 mL/kg/hr for the next 10 kg, and 1 mL/kg/hr for the remaining weight.[36] Once the volume deficit has been adequately replaced, the infant should be transitioned to maintenance fluids until he or she is able to tolerate enteral fluids.

**Intra-abdominal Nonaccidental Trauma (NAT).** Visceral organ injury occurs in roughly 2-4% of NAT.[37] The most common organs affected are the duodenum and proximal jejunum because they are rich in vascular supply and are fixed in the retroperitoneum.[37] Other organs that may be affected are the liver, spleen, kidneys, and pancreas. The examination of a child with nonaccidental intra-abdominal trauma can range from a seemingly normal abdomen to an abdomen with multiple obvious contusions and associated distention.[37] If abdominal trauma is suspected, the clinician should obtain CBC, blood type and screen, coagulation studies, and a CT of the abdomen. The patient should also be made NPO while he or she awaits prompt surgical consultation.

## Infectious Emergencies

**Serious Bacterial Infections (SBI), Including Urinary Tract Infections, Meningitis, Bacteremia, and Pneumonia.**[38] It is well documented that any infant less than one month of age with a temperature greater than 100.4°F must undergo a complete sepsis evaluation because of the inability to differentiate common viral illnesses from SBIs.[38] Evaluation of fever in children from 1 to 3 months is clinician-dependent at many institutions — the only full sepsis evaluation mandated in the literature is for those infants younger than 1 month of age. The physician should start with a thorough history, including birth history and perinatal course. Specifically in the birth history, it is important to know the infant's gestational age, maternal group B streptococcus status with treatment history, and any maternal sexually transmitted diseases (HIV, any maternal HSV, gonorrhea, chlamydia, or syphilis[38]). Both viral illnesses and SBIs may present with any or all of the following: fussiness, lethargy, hypothermia, hyperthermia, jaundice, seizures, irritability, apnea, poor feeding, tachypnea, and tachycardia. In this population, a complete sepsis evaluation includes: CBC with manual differential and blood culture, urinalysis and urine culture, cerebrospinal fluid cell count and CSF culture, and a chest X-ray (as clinically indicated). Using the manual differential and "I:T Ratio" may be useful in guiding the physician in the differentiation between viral illness and SBI.[38] This ratio is calculated as below:

I/T Ratio = (immature cells [such as band cells]) / (immature cells + total neutrophils). If the I/T ratio is greater than 0.2, then a physician should be more suspicious of a serious bacterial infection.[38]

Broad-spectrum antibiotics must be started as soon as possible and should be aimed at covering the most common organisms that lead to sepsis in this age group.[7] These organisms can easily be remembered by the mnemonic LEG as shown in Table 8.

The antibiotics recommended are a penicillin, plus either an aminoglycoside or a third-generation cephalosporin. Ceftriaxone is not generally used in patients younger than the age of 1 month due to the risk of biliary sludging and secondary development of hyperbilirubinemia. Antibiotic dosing is outlined in Table 9.[7]

### Table 9. Antibiotic Dosing

- Ampicillin 100 mg/kg/dose, interval dependent upon gestational age, weight, and day of life (refer to reference)
- Gentamicin dose and interval based upon gestational age and day of life (refer to reference)
- Cefotaxime/claforan 50 mg/kg/dose Q 8 hrs from day of life 0-7 or Q 8 hrs/day of life >7
- Ceftriaxone/rocephin 50 mg/kg/dose Q 12 hrs*

* Not generally used in neonates < 30 days of age

The clinician should also keep in mind the possibility of a neonatal herpetic infection, which may present in one of three manners: skin, eye, mucous membrane (SEM) presentation, disseminated disease, and encephalitis.[39] The evaluation for all neonatal herpetic illnesses should include scraping of suspicious lesions and obtaining blood and cerebrospinal fluid for HSV polymerase chain reaction (PCR). Of note, CSF in HSV infection may appear grossly bloody given the hemorrhagic nature of the infection. Diagnostic evaluation should also include a complete blood count with manual differential and a chemistry panel including liver function tests, as herpetic infection may result in elevated transaminases and bilirubin levels.[39]

Infection with herpes simplex virus limited to the skin, eye, or to mucous membranes (SEM presentation) accounts for about 45% of all herpetic illnesses in infants. Lesions to these areas typically appear around day of life 10-12. Eye infection causes conjunctival injection and watery discharge. Vesicular lesions may be present in the oropharynx, but they occur uncommonly. If not treated immediately, SEM disease is associated with a high risk of progression to serious disease.[39]

Disseminated disease accounts

for about 25% of HSV infections in newborns. The development of antiviral therapy (acyclovir) has greatly decreased the incidence of this disease entity. In the typical course, most affected neonates appear well at birth but begin to develop symptoms over the first 10-12 days of life. As the viremia progresses during this time, the HSV begins to affect multiple organs. Disseminated HSV symptoms are a function of the organ system affected (for example, CNS, renal, GI tract, and pulmonary system), and are difficult to distinguish from common viral illnesses and serious bacterial infections. These symptoms include irritability, poor feeding, respiratory distress, jaundice, seizure activity, and evidence of bleeding (GI hemorrhage, purpura). Roughly 60-75% of infants with disseminated HSV will have CNS involvement. Only 1 in 5 infants with disseminated disease go on to develop the classic vesicular rash.[39]

Encephalitis is the sole manifestation of HSV infection in 30% of neonatal HSV cases. Patients usually present with symptoms or signs of illness at 16-19 days of life, but can present as late as 4-6 weeks after birth. Initial manifestations of encephalitis include lethargy, irritability, poor feeding, seizures (which can be focal or generalized), fever or hypothermia, and bulging fontanelle. Patients may have a history of SEM disease that was not recognized as herpetic in nature.[39]

Intravenous acyclovir at 20 mg/kg (per dose) every 8 hours should be started immediately when one is concerned about the possibility of HSV infection.[7] The clinician should also consider obtaining an electroencephalogram, since HSV encephalitis is associated with temporal-lobe seizures, and head CT or MRI may show temporal-lobe abnormalities. The duration of treatment with acyclovir depends upon which presentation the clinician is treating, as shown in Table 10.

**Table 10. Acyclovir Duration of Treatment**

| Disease Presentation | Duration of Treatment |
|---|---|
| SEM disease | 14 days of IV acyclovir |
| Disseminated disease | 21 days of IV acyclovir and repeat lumbar for HSV PCR; if positive, treat for 28 days total. If repeat is negative, then 21 days is sufficient. |
| HSV encephalitis | 21 days of IV acyclovir and repeat lumbar puncture for HSV PCR; if positive, treat for 28 days total. If repeat is negative, then 21 days is sufficient. |

## Endocrine Emergencies

**Inborn Errors of Metabolism.** Diagnosis is often difficult and delayed because inborn errors of metabolism are rare and present with nonspecific symptoms. State newborn metabolic screens do a good job of helping diagnose the most common causes, but there are more than 400 known types of inborn errors of metabolism. Prompt diagnosis requires a high level of suspicion. Nonspecific symptoms include vomiting, irritability, poor feeding, failure to thrive, or tachypnea. More specific symptoms include seizures, apnea, hypoglycemia, temperature instability, and acidosis.[40] Initial workup and stabilization should include securing of ABCs and a bedside glucose. Next, the clinician should obtain a complete blood count, serum electrolytes, liver function tests, lactate, arterial blood gas, ammonia level, and a urinalysis for ketones and urine-reducing substances. Further evaluation should include urine organic acids and plasma amino acids.[40] Treatment requires correction of severe acidosis with sodium bicarbonate at 1 mEq/kg/dose and severe dehydration with isotonic fluid at 20 mL/kg. Fluids containing 10% dextrose should also be commenced expeditiously at a rate of 1 ½ to 2 times normal maintenance IV fluids. Treatment with intravenous dextrose will decrease further catabolism and subsequent acidosis.[40] Whenever an inborn error of metabolism is suspected, an immediate pediatric genetics consultation is warranted. Some of the different inborn errors of metabolism are outlined in Table 11.

**Congenital Adrenal Hyperplasia (CAH).** The most common cause of congenital adrenal hyperplasia (CAH) is a deficiency in the 21-hydroxylase enzyme.[41] This deficiency is usually discovered on prenatal screening, but it can be missed secondary to lab error, inadequate blood sample, or low infant birth weight (< 2500 grams). If the defect is not detected early, the neonate will present with vomiting, hypotension, or possibly shock. Electrolyte imbalances are common and usually include the combination of hyponatremia, hyperkalemia, and hypoglycemia. Any infant with hypotension that does not respond to aggressive fluid resuscitation or inotropic medications should alert the treating physician to the possibility of CAH.[41] Initial management of CAH includes securing ABCs and obtaining emergent electrolytes with a bedside glucose level. The physician should also consider an electrocardiogram (ECG) if hyperkalemia is a concern. ECG changes in acute hyperkalemia begin with peaked T waves. As the hyperkalemia worsens, one will begin to see a prolongation of the PR interval, followed by loss of the P wave with widening of the QRS complex. Worsening hyperkalemia ultimately leads to ventricular tachycardia or ventricular fibrillation with resultant cardiac arrest.

## Table 11. Inborn Errors of Metabolism

| Disease | Enzyme Affected | Signs/Symptoms |
|---|---|---|
| *Urea Cycle Defects* | | |
| Citrullinemia | Argininosuccinate synthetase deficiency | Lethargy, coma, seizures, developmental delay, hepatomegaly, vomiting, poor feeding, hyperammonemia |
| Argininosuccinic aciduria | Argininosuccinic acid lyase | Lethargy, coma, seizures, developmental delay, hepatomegaly, vomiting, poor feeding, hyperammonemia, brittle hair, and skin lesions |
| Arginase deficiency | Arginase | Spasticity, developmental delay, seizures, hyperammonemia, ataxia, vomiting, poor feeding, lethargy |
| *Glycogen Storage Diseases* | | |
| Type I (von Gierke disease) | Glucose-6-phosphatase | Hypoglycemia, hepatomegaly, hyperlipidemia, growth failure, lactic acidosis, and hyperuricemia |
| Type II (Pompe disease) | Acid maltase | Hepatomegaly and progressive muscle weakness, leading to cardiac failure and death by 2 years old. |
| Type III (Cori disease) | Glycogen debrancher | Hypoglycemia, hepatomegaly, hyperlipidemia, and myopathy |
| Type V (McArdle disease) | Muscle glycogen phosphorylase | Exercise-induced cramping and recurrent rhabdomyolysis leading to myoglobinuria and renal failure |
| *Amino Acid Metabolism* | | |
| Maple syrup urine disease | Branched-chain alpha-keto acid dehydrogenase complex | Poor feeding, vomiting, dehydration, lethargy, hypotonia, seizures, ketoacidosis, sweet smelling urine, and progressive neurological decline if not diet controlled |
| Propionic acidemia | propionyl-CoA carboxylase deficiency | Progressive encephalopathy, increased infection risks, seizures, hyperammonemia, basal ganglia strokes, and cardiomyopathy |
| Homocystinuria | Cystathione beta-synthase deficiency | Connective tissue, cardiovascular, CNS, and muscle disease, increased homocystine in serum and urine |
| Phenylketonuria | Phenylalanine hydroxylase | If not well controlled, then progressive developmental delay, seizures, and cognitive impairment |

Treatment of adrenal crisis associated with CAH includes administration of 1-2 mg/kg of hydrocortisone (both glucocorticoid and mineralocorticoid effect) and correction of associated electrolyte imbalances. For symptomatic hyperkalemia or hyperkalemia with ECG changes, see Table 12.

## Electrolyte Emergencies

**Hyponatremia.** Hyponatremia is defined as a serum sodium level less than 130 mEq/L, although it is not usually symptomatic until the serum sodium has dropped to 125 mEq/L or below.[42] Hyponatremia may present with seizures and requires quick recognition and immediate correction with 5-10 mL/kg of 3% sodium chloride.[43] One of the more common culprits of hyponatremia in a neonate is improper mixing of formula.[42] Usually a parent or guardian is trying "to stretch out the formula" and may over-dilute the powdered formula with too much water. Another cause of hyponatremia includes parents supplementing infants with free water.

**Hypoglycemia.** Hypoglycemia is the most common metabolic problem in neonates. It is defined as a blood glucose value of less than 40 mg/dL.[44] Infants with hypoglycemia are usually asymptomatic but can present with increased sleepiness, lethargy, seizures, vomiting, and unresponsiveness. Hyperinsulinism is the most common cause of hypoglycemia in the first 3 months of life and is well-recognized in infants of mothers with diabetes.[44] Other causes are sepsis and any acute illness in which there is decreased oral intake. Treatment of hypoglycemia is 2-4 mL/kg of 10% dextrose in water as an intravenous bolus. This bolus should be followed by maintenance dextrose administration with a glucose infusion rate (GIR) of 5-8 mg/kg/min.[44]

**Hypocalcemia.** Although hypocalcemia is relatively rare in neonates, it usually develops after the first week of life and is associated with conditions that have high serum phosphate levels (vitamin D deficiency, hyperparathyroidism, and maternal anticonvulsant use). Vitamin D deficiency usually resolves with reduction of the renal phosphate load or with vitamin D

## Table 12. Symptomatic Hyperkalemia

| Medication | Dosing | Method of Action |
|---|---|---|
| Calcium gluconate 10% | 100 mg/kg IV (via central line) over 3-5 mins | Cardiac stabilizer that increases threshold potential. This medication restores normal gradient between threshold potential and resting membrane potential. (Does not lower serum potassium.) Not compatible with sodium bicarbonate. |
| Sodium bicarbonate | 0.5 mEq/kg IV over 5-10 minutes. May repeat after 10 minutes. | Bicarbonate ion neutralizes hydrogen ions and raises the pH, thus driving potassium intracellularly. Onset of action is within minutes, and duration of effects is 15-30 minutes. |
| Insulin and glucose | 0.1 U/kg insulin with 2 mL/kg 25% dextrose | Stimulates cellular uptake of potassium. Effects are seen immediately, but are temporary. Dextrose is given to prevent further hypoglycemia. |
| Furosemide (Lasix) | 1-2 mg/kg/dose IV | Increase renal excretion of potassium. Effects are inconsistent and may take up to an hour before any effects are seen. |
| Sodium polystyrene sulfonate (Kayexelate) | 1 gm/kg/dose PO/PR | Binding resin that exchanges sodium for potassium in the large intestine, decreasing total body potassium. Onset of action is 2-12 hours and is longer if given PR. Lowers potassium over 1-2 hours with 4-6 hour duration of action. Multiple doses usually necessary. |

supplementation.[45] The treatment for hypocalcemia is 100-300 mg/kg of calcium gluconate administered via peripheral or central IV or calcium chloride 10-20 mg/kg/dose via central IV catheter.[46] The best way to determine the cause of hypocalcemia is to classify the causes into the following categories.[45]

*Too Little Vitamin D.* Vitamin D helps maintain serum calcium levels by aiding in absorption of calcium through the intestines. Low levels of vitamin D can result from a number of different causes. The most common example of low vitamin D is an exclusively breast-fed baby around 6 months of age with no supplementation. There are also certain medications (taken by the baby or the breast-feeding mother) associated with decreased vitamin D, including anticonvulsants (phenytoin), isoniazid, and rifampin. Finally, liver disease can affect vitamin D levels by affecting the metabolism of vitamin D to its active form (25-hydroxylation of vitamin D).[45]

*Too Little Parathyroid Hormone.* Certain congenital diseases of the midline structures, such as DiGeorge syndrome, VATER association, and CHARGE association, lead to parathyroid aplasia; fetal exposure to retinoic acid may also lead to parathyroid aplasia. In these situations, little or no parathyroid hormone is made, resulting in no/little bone reabsorption and decreased calcium reabsorption from the kidneys and gut.[45]

*Too Much Phosphate.* This situation is usually due to an increased oral intake of cow's milk, which contains much more phosphate than human milk. Renal failure also leads to decreased excretion of phosphate and increased excretion of calcium.[45]

*Spring Tetany or "Hungry Bone Syndrome."* This condition occurs when a child has had vitamin D deficient rickets to the point that the bones are depleted of calcium. In the initial stages of treatment (or sunlight exposure), vitamin D repletion leads to a massive influx of calcium back into the bones. If the infant is not receiving calcium supplementation, there will be an acute drop in the serum calcium levels, possibly leading to seizures (hence the name spring tetany).[45]

## Conclusion

When presented with a sick neonate, a pediatric emergency medicine physician must remember that ill babies can exhibit a wide variety of symptoms. An extensive history and physical exam should provide clues to the source of an infant's illness, and the priority in management is securing the ABCs followed by obtaining labs (including blood, urine, and cerebrospinal fluid cultures as indicated) that can focus treatment.

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## Physician CME Questions

33. Which of the following statements regarding head trauma in the neonate is true?
    A. Magnetic resonance imaging (MRI) is the modality of choice for an initial screening tool.
    B. Cushing's triad consists of hypotension, tachycardia, and respiratory alterations.
    C. Hyperventilation is the first-line therapy for elevated intracranial pressure.
    D. Mannitol induces hyperosmolarity and, therefore, an extracellular fluid shift.
    E. Hypertonic saline causes an intracellular fluid shift, thus increasing intracellular edema.

34. Which of the following conditions causes a conjugated hyperbilirubinemia?
    A. hereditary spherocytosis
    B. ABO incompatibility
    C. biliary atresia
    D. breastmilk jaundice
    E. Crigler-Najjar syndrome

35. Which of the following is the first-line therapy for neonatal seizures?
    A. phenobarbital
    B. phenytoin
    C. lorazepam 0.01 mg/kg/dose
    D. lorazepam 0.1 mg/kg/dose
    E. lorazepam 1 mg/kg/dose

36. Which one of the following is *not* a cyanotic heart lesion?
    A. transposition of the great vessels
    B. hypoplastic left heart
    C. pulmonary atresia
    D. tricuspid atresia
    E. total anomalous pulmonary venous return

37. A male infant presents with acute onset of wheezing that developed while he was crawling around on the kitchen floor. His mother denies any history of asthma in the patient or the family. On exam, the patient is afebrile with 96% oxygen saturations, and his wheezing is localized to the right side. What might you see on the chest radiograph?
    A. steeple sign
    B. focal infiltration
    C. thumbprint sign
    D. double bubble
    E. persistent right-sided inflation in the right lateral decubitus position

38. Which of the following is an example of lower airway obstruction?
    A. pertussis
    B. bacterial tracheitis
    C. bronchiolitis
    D. choanal atresia
    E. croup

39. An infant presents with a 2-day history of fever and cough. The parents describe the cough as "barking," and they think that he is hoarse. In the ED, he is sitting in his mother's lap and has audible stridor at rest; he also has mild to moderate suprasternal and intercostal retractions. Which of the following therapies would be indicated at this time?
    A. albuterol nebulizer treatment
    B. heliox therapy
    C. cool mist administration
    D. racemic epinephrine nebulizer treatment
    E. atrovent nebulizer therapy

40. Which of the following is true regarding volvulus?
    A. Patients are generally asymptomatic.
    B. Symptoms include nonbilious emesis, diarrhea, and fever.
    C. Symptoms include abdominal distention, abdominal pain, and acute onset of bilious emesis.
    D. Air contrast enema is the modality of choice for diagnosing and treating volvulus.
    E. Antibiotics are not considered to be part of the treatment plan.

41. A 9-day-old with an uncomplicated perinatal history presents with a history of decreased oral intake and increased sleeping. His vital signs are notable for a 38.4° C rectal temperature, and his physical exam is completely normal. After the sepsis work-up, which of the following would be appropriate antibiotic therapy?
    A. ceftriaxone
    B. ampicillin plus ceftriaxone
    C. ampicillin plus cefotaxime
    D. gentamicin plus ceftriaxone
    E. cefotaxime

42. Which of the following is true regarding congenital adrenal hyperplasia (CAH)?
    A. The most common enzyme deficiency is 21-hydroxylase.
    B. The usual electrolyte abnormalities include hypernatremia and hypokalemia.
    C. Hypotension associated with CAH should respond to fluids alone.
    D. Infants typically present with hypertension and diarrhea.
    E. Patients are generally hyperglycemic.

**Answers:** 33. D; 34. C; 35. D; 36 B; 37. E; 38. C; 39. D; 40. C; 41. C; 42. A

## Pediatric Emergency Medicine Reports

### *CME Objectives*

Upon completion of this educational activity, participants should be able to:

1. recognize specific conditions in pediatric patients presenting to the emergency department;
2. describe the epidemiology, etiology, pathophysiology, historical and examination findings associated with conditions in pediatric patients presenting to the emergency department;
3. formulate a differential diagnosis and perform necessary diagnostic tests;
4. apply up-to-date therapeutic techniques to address conditions discussed in the publication;
5. discuss any discharge or follow-up instructions with patients.

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To clarify confusion surrounding any questions answered incorrectly, please consult the source material. After completing this activity, you must complete the evaluation form that will be provided at the end of the semester and return it in the reply envelope provided to receive a credit letter. When your evaluation is received, a credit letter will be mailed to you.

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**Vice President/Group Publisher:** Donald R. Johnston
**Executive Editor:** Shelly Morrow Mark
**Managing Editor:** Leslie Hamlin

**GST Registration No.:** R128870672
Periodicals Postage Paid at Atlanta, GA 30304 and at additional mailing offices.

**POSTMASTER**: Send address changes to ***Pediatric Emergency Medicine Reports***, P.O. Box 105109, Atlanta, GA 30348.





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# Neonatal Emergencies

## Radiographic Findings

| Diagnosis | X-ray Finding | Description |
|---|---|---|
| Croup | Steeple sign | On anterioposterior (AP) neck X-ray, there is a gradual narrowing of the upper airway that resembles the top of a steeple. |
| Epiglottitis | Thumbprint sign | On lateral view of neck X-ray, the epiglottic swelling resembles a thumbprint. |
| Foreign body | Persistent airway expansion or overexpansion | Ideally, one should obtain both inspiratory and expiratory chest films, and the side with the foreign body will appear expanded/overexpanded on both views while the non-affected lung will reveal expansion on inspiration and appropriate collapse on expiration. Given that neonates do not inhale and exhale on command, decubitus films serve a similar function in that normal lungs should collapse when dependent and expand when non-dependent; in the setting of a foreign body, the affected lung would remain similarly expanded whether the lung is dependent or non-dependent. |
| Bronchiolitis | Hyperinflation | On AP CXR the lungs appear hyperinflated, with flattened diaphragms and possible areas of atelectasis. |
| Pneumonia | Focal or diffuse infiltration | On AP CXR the lungs show a focal area of consolidation or may show diffuse patchy consolidation in the case of atypical or viral pneumonias. Also obtain lateral CXR to visualize the retrocardiac area properly. |

## Medical Therapy for Increased Intracranial Pressure

| Treatment | Dose | Mechanism of Action |
|---|---|---|
| Head elevation and midline position | 30 degrees | Improves CSF and venous drainage |
| Hyperventilation | Goal end-tidal $CO_2$ of 35. | Current recommendations are short, intermediate episodes of hyperventilation only in times of impending herniation. It causes an immediate effect of decreasing ICP, but the effect is transient. Also it is unknown if there is any effect on long-term outcome. |
| Mannitol | 0.5-1 gm/kg/dose IV | Works by two mechanisms. First, mannitol causes a decrease in blood viscosity, leading to reflex vasoconstriction intracranially, thereby decreasing intracranial pressure. Second, it induces hyperosmolarity and, therefore, an extracellular fluid shift. Mannitol may accumulate in areas of cerebral vascular interruption, potentially leading to reverse osmosis with increased intracranial pressure in these areas. |
| Hypertonic saline (multiple concentrations, most common is 3%) | Bolus 3% saline 2-4 mL/kg/dose followed by infusion of 0.1-1 mL/kg/hr to keep ICP < 20 | Hyperosmolar agent that works by causing an extracellular shift of fluid, therefore decreasing intracellular edema. It has been demonstrated that it also decreases overall fluid requirement, decreases therapies in the first 3 days, decreases the duration of mechanical ventilation, and decreases the length of hospitalization. |
| Barbiturates (drug-induced comas) | Generally, pentobarbital 3-10 mg/kg loading dose followed by 1-2 mg/kg/hr infusion | It has been shown to decrease excitotoxicity, decrease ICP, and increase brain oxygenation. |

## Signs and Symptoms of Dehydration

| Clinical Sign | Mild Dehydration | Moderate Dehydration | Severe Dehydration |
|---|---|---|---|
| Mental status | Crying | Sleepy but arousable | Difficult to arouse to unarousable |
| Mucous membranes | Moist | Dry and tacky | Cracked, dry, bleeding |
| Fontanelle | Flat | Flat to mildly depressed | Extremely sunken |
| Capillary refill | < 2 seconds | 2-4 seconds | > 4-5 seconds and cold |
| Heart rate | Mildly elevated | Tachycardic | Extreme tachycardia |
| Blood pressure | Normal | Normal | Hypotensive |
| Skin turgor | Normal | Normal to slow rebound | Tenting noted |
| Respiratory rate | Normal | Mild elevation | Notable tachypnea |
| Tears | Normal | Decreased but present | No tears |
| Urine output | Normal | Decreased | Anuric |

## Airway Obstructions

| Upper Airway Obstructions | Lower Airway Obstructions |
|---|---|
| Laryngomalacia** | Tracheomalacia** |
| Choanal atresia/stenosis | Bronchiolitis |
| Foreign body aspiration** | Foreign body aspiration** |
| Pertussis | Pneumonia |
| Croup | Vascular rings |
| Bacterial tracheitis | |
| Airway masses** | Airway webs and masses** |
| Vocal cord paralysis/dysfunction** | Vocal cord paralysis/dysfunction** |

** Indicates diagnoses that can have a component of both upper and lower airway obstruction.

## Inborn Errors of Metabolism

| Disease | Enzyme Affected | Signs/Symptoms |
|---|---|---|
| *Urea Cycle Defects* | | |
| Citrullinemia | Argininosuccinate synthetase deficiency | Lethargy, coma, seizures, developmental delay, hepatomegaly, vomiting, poor feeding, hyperammonemia |
| Argininosuccinic aciduria | Argininosuccinic acid lyase | Lethargy, coma, seizures, developmental delay, hepatomegaly, vomiting, poor feeding, hyperammonemia, brittle hair, and skin lesions |
| Arginase deficiency | Arginase | Spasticity, developmental delay, seizures, hyperammonemia, ataxia, vomiting, poor feeding, lethargy |
| *Glycogen Storage Diseases* | | |
| Type I (von Gierke disease) | Glucose-6-phosphatase | Hypoglycemia, hepatomegaly, hyperlipidemia, growth failure, lactic acidosis, and hyperuricemia |
| Type II (Pompe disease) | Acid maltase | Hepatomegaly and progressive muscle weakness, leading to cardiac failure and death by 2 years old. |
| Type III (Cori disease) | Glycogen debrancher | Hypoglycemia, hepatomegaly, hyperlipidemia, and myopathy |
| Type V (McArdle disease) | Muscle glycogen phosphorylase | Exercise-induced cramping and recurrent rhabdomyolysis leading to myoglobinuria and renal failure |
| *Amino Acid Metabolism* | | |
| Maple syrup urine disease | Branched-chain alpha-keto acid dehydrogenase complex | Poor feeding, vomiting, dehydration, lethargy, hypotonia, seizures, ketoacidosis, sweet smelling urine, and progressive neurological decline if not diet controlled |
| Propionic acidemia | propionyl-CoA carboxylase deficiency | Progressive encephalopathy, increased infection risks, seizures, hyperammonemia, basal ganglia strokes, and cardiomyopathy |
| Homocystinuria | Cystathione beta-synthase deficiency | Connective tissue, cardiovascular, CNS, and muscle disease, increased homocystine in serum and urine |
| Phenylketonuria | Phenylalanine hydroxylase | If not well controlled, then progressive developmental delay, seizures, and cognitive impairment |

## Symptomatic Hyperkalemia

| Medication | Dosing | Method of Action |
|---|---|---|
| Calcium gluconate 10% | 100 mg/kg IV (via central line) over 3-5 mins | Cardiac stabilizer that increases threshold potential. This medication restores normal gradient between threshold potential and resting membrane potential. (Does not lower serum potassium.) Not compatible with sodium bicarbonate. |
| Sodium bicarbonate | 0.5 mEq/kg IV over 5-10 minutes. May repeat after 10 minutes. | Bicarbonate ion neutralizes hydrogen ions and raises the pH, thus driving potassium intracellularly. Onset of action is within minutes, and duration of effects is 15-30 minutes. |
| Insulin and glucose | 0.1 U/kg insulin with 2 mL/kg 25% dextrose | Stimulates cellular uptake of potassium. Effects are seen immediately, but are temporary. Dextrose is given to prevent further hypoglycemia. |
| Furosemide (Lasix) | 1-2 mg/kg/dose IV | Increase renal excretion of potassium. Effects are inconsistent and may take up to an hour before any effects are seen. |
| Sodium polystyrene sulfonate (Kayexelate) | 1 gm/kg/dose PO/PR | Binding resin that exchanges sodium for potassium in the large intestine, decreasing total body potassium. Onset of action is 2-12 hours and is longer if given PR. Lowers potassium over 1-2 hours with 4-6 hour duration of action. Multiple doses usually necessary. |

## Acyclovir Duration of Treatment

| Disease Presentation | Duration of Treatment |
|---|---|
| SEM disease | 14 days of IV acyclovir |
| Disseminated disease | 21 days of IV acyclovir and repeat lumbar for HSV PCR; if positive, treat for 28 days total. If repeat is negative, then 21 days is sufficient. |
| HSV encephalitis | 21 days of IV acyclovir and repeat lumbar puncture for HSV PCR; if positive, treat for 28 days total. If repeat is negative, then 21 days is sufficient. |

## LEG Mnemonic

*L isteria monocytogenes*
*E scherichia coli*
G roup B Streptococcus

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