The care of a critically ill neonate in the emergency department setting presents a special challenge for practitioners who do not routinely encounter compromised neonates. This review will provide guidelines for the initial stabilization of these infants as well as identify differential diagnoses that should be considered. As with any critically ill patient, achieving physiologic stability of the neonate is the first priority. However, in addition to maintaining the airway, breathing, and circulation, thermoneutrality must be achieved for a successful outcome. Obtaining the history and initiating diagnostic evaluations should occur during or shortly after the course of stabilization to promptly initiate disease-specific therapies. The mnemonic NEO SECRETS may help focus the care and promote early identification of the cause of the infant’s deterioration.

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KEYWORDS neonatal emergency care, neonatal emergencies

Rapid evaluation and stabilization of a critically ill neonate is a required skill in the emergency department (ED). Neonates can acutely decompensate from a variety of causes. However, neonates have a limited repertoire of responses to stress and the presenting signs can be nonspecific. Therefore, a logical and multisystem approach to the evaluation and management of common causes is essential to stabilizing these infants. The following review summarizes our approach to a critically ill newborn in the setting of a busy ED. Although history, physical examination, and laboratory evaluation are described in sequence, it is understood that emergent cardiopulmonary stabilization takes precedence over detailed evaluation of the neonate for underlying causes. A rapid appraisal of the newborn for some of the more common diagnoses is essential though, for institution of specific therapies.

History
Initial evaluation of the critically ill neonate may be focused on immediate needs but should be orderly so as not to overlook significant signs or symptoms. The elements of a neonatal history should begin with a maternal history, such as maternal antenatal laboratory tests (status of rubella, hepatitis B, syphilis, human immunodeficiency virus, group B streptococcus [GBS] screens, and blood type) medications, complications during pregnancy and maternal illnesses. A birth history should follow and includes the gestational age at delivery, indication for delivery (maternal pregnancy induced hypertension, fetal distress, etc), the route of delivery including any instrumentation, complications during delivery, birth weight, and resuscitation required. The immediate neonatal course should also be reviewed for difficulties in transitioning to extraterine life such as respiratory distress, hypoglycemia, feeding difficulties, jaundice, and length of hospital stay. The mother may remember the infant’s discharge weight and this should be asked.

The neonate’s medical history consists of the time from discharge from the birth hospital to the time of presentation to the ED. Details on the infant’s oral intake including the amount of water used to mix formula or any additives used should be obtained. The infant’s urine output as well as stooling habits should also be assessed. If available,
information from the first well child check should be obtained, either from the mother or the primary care provider’s office. The neonate’s examination and growth parameters from the last examination should be reviewed and compared with the weight obtained in the ED. Results of the state mandated newborn screening test results may be obtained from the primary physician’s office, hospital of birth, or directly from the state newborn screening program. These test results are important in the evaluation of certain metabolic and endocrine causes of a neonate’s acute illness as discussed later in this review.

**Physical Examination**

The physical examination of the neonate begins with inspection and an overall observation of the neonate’s condition. It is important to remember that the presenting symptoms and signs can be nonspecific and common to a variety of diseases including sepsis, heart failure, and metabolic diseases. Therefore, it is important to consider a list of common disorders that present with immediate deterioration in the neonatal period. The overall appearance of the infant is often more significant in assessing the severity of the illness. A flexed posture with spontaneous movements and pink perfusion is reassuring. A flaccid posture with decreased spontaneous movements, cyanosis, and respiratory distress signifies a gravely ill neonate in need of immediate attention. Review of vital signs on admission may also provide an indication of distress. Axillary temperatures for neonates vary widely but are generally thought to be between 36.2°C and 37.7°C [1]. Heart rates between 110 and 160 beats per minute are normal for healthy term infants and may vary during sleep or active awake states [2]. Respiratory rates between 40 and 60 breaths per minute are also normal for a neonate [3]. Normal systolic blood pressures during the first month of life range from 65 to 75 mm Hg and diastolic pressures from 40 to 50 mm Hg [4]. An easy rule of thumb is that the mean blood pressure should approximate the estimated gestational age in weeks at the time of measurement. For example, a 2 week old former 38 week gestation infant’s mean blood pressure should be around 40 mm Hg. Incorrect blood pressure readings may be obtained if the sphygmomanometer cuff is too small or large. Four extremity blood pressures should be obtained on all hemodynamically unstable neonates because discrepancies between upper and lower limb blood pressures may indicate a ductal dependent coarctation of the aorta.

The remainder of the physical examination should follow and may suggest the source of illness. Inspection, palpation, and auscultation of the infant should occur from head to toe. The head should be palpated and carefully inspected for trauma in the encephalopathic neonate. The fontanelle should be auscultated to assess for a cranial bruit, especially if the neonate is hemodynamically unstable as this may be a sign of a cerebral arterio-venous malformation. Fixed and dilated pupils with a persistent oculocephalic response may indicate lidocaine toxicity, especially if there is a recent history of a circumcision or other medical procedure. The mouth, neck, shoulders, and arms should be assessed. Brachial pulses should be palpated and compared with femoral pulses to rule out a significant coarctation of the aorta. Examination of the chest may reveal a pronounced cardiac murmur, unequal breath sounds, or significant subcostal retractions. A distended, tender, and tympanic abdomen should prompt surgical consultation and a focused diagnostic search. The genitalia should be inspected. Testicular torsion or incarcerated inguinal hernias may be found. Ambiguous genitalia in a female infant or a hyperpigmented scrotum in a male infant may suggest congenital adrenal hyperplasia (CAH). Bloody stools may suggest gastrointestinal necrosis. All extremities should be palpated to ensure that bony structures are intact. A neurologic examination including evaluation of the primitive neonatal reflexes (root, suck, gag, Moro) should be done.

The history, physical examination, and initial stabilization usually occur concurrently in a deteriorating infant and may easily be interrupted. Hence, care must be taken to ensure a thorough physical examination is completed on all critically ill infants.

**Initial Stabilization**

As in all pediatric patients, stabilization prioritizes securing the airway, then establishing breathing and maintaining adequate circulation. Endotracheal intubation and fluid resuscitation are usually required in critically ill neonates, and these procedures, if indicated, should not be delayed while waiting for diagnostic evaluation. Establishing optimal ventilation and oxygenation is often sufficient to improve both respiratory and cardiac insufficiency; however, intravenous (IV) fluids and resuscitation may be required in the gravely ill neonate. An initial bolus of 10 mL/kg of isotonic fluid (0.9 NS) should be given over 10 to 15 minutes and repeated if necessary. If the umbilical stump is still present, this fluid may be given through an umbilical venous catheter [5].

If the physical examination identifies poor peripheral perfusion and cyanosis with or without a cardiac murmur, a continuous infusion of prostaglandins (PGE\textsubscript{1}, 0.01-0.05 μg/kg per minute) should be promptly initiated and pediatric cardiology consulted. If abdominal distension or discoloration is noted on physical examination, a surgical abdomen must be considered. Bilious emesis and/or abdominal distention in the neonate should be treated as a surgical emergency and should be promptly evaluated with appropriate diagnostic tests and surgical consultation. Neonates with enteric emergencies often require additional fluid resuscitation as well as decompression of the gastrointestinal tract with a large bore repogle tube. Two-view abdominal radiographs should be done to assess the
bowel gas pattern and rule out perforation of a viscus. Pediatric surgery should be notified, and transfer to an appropriate facility should be initiated if necessary.

Measures must be taken to promote thermoneutrality because this is essential for successful stabilization of a critically ill neonate. In vivo thermoregulation is an active process that requires oxygen, glucose, corticosteroids, and catecholamines [6]. A critically ill neonate who is hypoxic, hypoglycemic, or adrenally suppressed will be unable to maintain a normal body temperature and will become hypothermic. Prolonged hypothermia may result in cold stress that can affect all organ systems. The cold stressed infant may present with peripheral vasoconstriction to maintain heat, respiratory distress to increase oxygenation, metabolic acidosis due to hypoglycemia, and anaerobic metabolism if hypoxia persists, which further worsens the respiratory distress [7]. This vicious cycle of hypothermia and hypoxia will impede the response to resuscitation measures.

For the cold infant, heat should be provided along with initial stabilization measures. Warmth can be initiated with a radiant warmer bed or overhead heat lamps. Although other methods such as chemical heating pads have been used, radiant heat is the most common and efficient in rewarming an infant [7]. In an ED setting, the radiant warmer also allows unrestricted access to the neonate for medical procedures. However, its use in the cold infant must be done with caution. Rapid warming can result in apnea, hypotension, and shock [8]. To prevent overheating of the neonate, use the servo-control option on the radiant warmer, set the skin temperature to 36.5°C, and apply the skin probe to the anterior abdomen of the neonate. Oxygen consumption will increase as the infant’s core temperature returns to normal. If the infant remains hypoxic because of respiratory or cardiac insufficiencies, rewarming may worsen tissue hypoxia. These complications must be monitored for, and if seen, the rewarming process should be slowed by resetting the servo-control skin temperature to 1°C above the neonate’s core temperature. Once the neonate achieves that temperature, the servo skin temperature is increased by 1°C again until the neonate attains a core temperature of 36.5°C. Care should be taken to avoid hyperthermia in neonates with suspected hypoxic ischemic encephalopathy because elevated core temperature can exacerbate neurologic injury in these cases.

**Laboratory Evaluation**

Laboratory evaluation of a critically ill neonate can be directed toward a specific etiology if suspected. In absence of a clear etiology, laboratory samples are also frequently obtained to help determine diagnosis and management. Appropriate routine laboratory work in a critically ill neonate consists of a complete blood count with a differential, blood and urine cultures, urinalysis, electrolytes including blood urea nitrogen and creatinine, glucose, and an arterial blood gas. Bedside blood glucose estimation and ionized calcium measurements should also be considered. Serum calcium, magnesium, phosphorus, blood ammonia, lactate and pyruvate, and cerebrospinal fluid studies should be performed if the neonate demonstrated central nervous system signs such as seizures and encephalopathy. Empiric use of IV broad-spectrum antibiotics is usually initiated in the ill appearing neonate after the initial blood work is obtained.

**Differential Diagnoses**

Once the neonate has recovered from the initial decompensation, efforts can be directed toward determining the etiology of the infant’s condition. Working through the differential diagnosis in a critically ill neonate can be exhaustive and time-consuming. Commonly considered causes for a neonate’s catastrophic deterioration are infection, congenital heart disease, and metabolic disorders (Table 1). However, there are other etiologies for critical illness in the neonate that must be considered as well. The following mnemonic, “NEO SECRETS,” may help focus the care and promote early identification of the cause of the infant’s deterioration (Table 2).

**Inborn Errors of Metabolism**

Most neonates with inborn errors of metabolism become symptomatic after oral feeds and ingestion of the offending agent; therefore, most present between days 2 to 7 of life and may be erroneously diagnosed as having sepsis. Early suspicion of an inborn error of metabolism is essential for prompt diagnosis and the removal of the harmful metabolite.

Affected neonates commonly demonstrate poor feeding with associated poor suck. Irritability, vomiting and failure to thrive, hepatosplenomegaly, jaundice, and abnormal bowel gas pattern and rule out perforation of a viscus. Pediatric surgery should be notified, and transfer to an appropriate facility should be initiated if necessary.

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urine odors may be found. If the disease progresses unchecked, apnea, lethargy, a comatose state, and death may occur, and this deterioration can occur quickly. Metabolic acidosis, persistent hypoglycemia, or a suggestive family history should increase the suspicion for an inborn error of metabolism. Prompt consultation with a neonatologist or medical specialist in genetics or inborn errors of metabolism is imperative. In addition to the initial stabilization blood work previously mentioned, directed laboratory evaluation includes serum amino acids, ammonia, pyruvate, lactate, urine organic acids, and urinalysis for ketones (Figure 1). If a confirmed serum ammonia level is more than 200 μmol/L, immediate transfer to an intensive care unit with the ability to perform neonatal dialysis must be arranged.

Specific treatments are indicated based on the final diagnosis and with the guidance of the medical specialist in genetics or inborn errors of metabolism. Immediate general measures will include the elimination of the inciting metabolite by stopping all enteral feeds, initiation of IV fluids, and correction of associated acid-base and electrolyte abnormalities.

Electrolyte Abnormalities

Electrolyte abnormalities in the critically ill neonate are commonly the result of an underlying process rather than the initiating factor. However, delayed management may hinder any resuscitative efforts and may contribute to the neonate’s ultimate demise. Water and electrolyte metabolism are closely associated, and the management of one may affect the other.

Water balance in the neonate is primarily controlled by antidiuretic hormone whose action results in the uptake of water from the distal renal tubules as well as the cortical and medullary collecting ducts. This conservation of water is triggered by contraction of the intravascular volume and/or an increase in the serum osmolality. However, the renal response to antidiuretic hormone is diminished in neonates, thereby placing the critically ill neonate at high risk for imbalance. Water requirements increase with advancing age. Neonates require 60 to 80 mL/kg per day in the first 2 days of life, 100 to 150 mL/kg per day during days 2 through 7, and 120 to 180 mL/kg per day thereafter until 28 days of life.

For the dehydrated neonate, fluid resuscitation requires the replacement of the fluid deficit in addition to maintenance water requirements. The percentage of dehydration is simply calculated as follows: fluid deficit (L) = pre-illness weight (kg) − illness weight (kg). For the neonate, the birth weight, hospital discharge weight, or the 2-week office visit weight should be available for comparison with the weight in the ED.

Figure 1 Approach to the neonate with a suspected inborn error of metabolism. FAO, fatty acid oxidation; RTA, renal tubular acidosis.

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<tr>
<th>N</th>
<th>Inborn errors of metabolism</th>
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<tr>
<td>E</td>
<td>Electrolyte abnormalities</td>
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<tr>
<td>O</td>
<td>Overdose (toxin, poison)</td>
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<td>S</td>
<td>Seizures</td>
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<td>E</td>
<td>Enteric emergencies</td>
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<td>C</td>
<td>Cardiac abnormalities</td>
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<td>R</td>
<td>Recipe (formula, herbs, additives)</td>
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<td>E</td>
<td>Endocrine crisis</td>
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<th>Hypoglycemia</th>
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<th>Metabolic Acidosis</th>
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<td>Ketosis</td>
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Sodium balance in the neonate is primarily determined by the nephron. The fractional excretion of sodium is inversely proportional to gestational age. Although most healthy term infants have a mature basal sodium balance with a fractional excretion of sodium of less than 1%, certain conditions, including hypoxia and respiratory distress, can result in higher urinary losses of sodium and sodium imbalance [12]. Sodium supplementation in the first 24 hours of life is not necessary because hyponatremia at this time is due to excess free water and not sodium deficiency. During the remainder of the first week, neonates require 2 to 4 mEq/kg per day of sodium; this increases to 3 to 5 mEq/kg per day during the remainder of the first month of life. Similarly, potassium supplementation should be started on the second day of life. Potassium requirements in the first week of life are 1 to 2 mEq/kg per day and 2 to 3 mEq/kg per day for the remaining month of life.

Hyponatremia, hyperkalemia, and metabolic acidosis may also be caused by congenital renal anomalies that are associated with progressive renal failure. The common causes include autosomal recessive polycystic kidney disease, renal dysplasias, and renal tubular acidosis. These are usually accompanied by significant alterations in blood urea nitrogen and creatinine.

The clinical signs and diagnostic evaluation of sodium and potassium abnormalities are shown in Table 3. The management of these derangements is the same: calculate the deficit and replace the losses while avoiding rapid correction. If the neonate is seizing because of hyponatremia, 4 to 6 mL/kg of 3% NaCl IV may be given rapidly without increased risk of central pontine myelinosis [13]. Hyperkalemia with electrocardiogram changes also requires additional measures: calcium gluconate 10% 100 mg/kg IV over 3 to 5 minutes, NaHCO₃ (1-2 mEq/kg IV over 5-10 minutes), and insulin (0.1 U/kg/hr) IV infusion.

### Overdose

Neonatal toxicology is very different from pediatric toxicology. Accidental ingestion is common in toddlers but rare in neonates. Their immature pharmacokinetics

<table>
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<th>Table 3</th>
<th>Clinical presentation and diagnostic workup guidelines for common electrolyte abnormalities in a critically ill newborn.</th>
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<tr>
<td><strong>Signs and Symptoms</strong></td>
<td><strong>Diagnostic Workup</strong></td>
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</table>
| Hyponatremia (Na⁺ <130 mEq/L) | Edema  
Vomiting  
Muscle weakness  
Seizures and/or coma | If ↑ K and shock, r/o CAH  
If ↓ UOP and ↓ serum osmolality, r/o SIADH or renal defect  
If ↑ UOP and dehydration, r/o improper mixing of formula |
| Hypernatremia (Na⁺ >150 mEq/L) | Lethargy or irritability  
Seizures, decreased tone | If ↑ UOP and ↓ urine specific gravity, r/o renal concentration defects  
If ↑ UOP and ↑ specific gravity, r/o breastfeeding failure, improper formula mixing |
| Hypokalemia (K⁺ <3 mEq/L) | Weakness  
Ileus  
Hypertension  
ECG changes: ↑ P wave amplitude, inverted T waves | If ↑ urine K, r/o renal losses  
If hypertensive, r/o excess mineralocorticoids or renin  
If normotensive, r/o RTA, diuretics, GI losses or malnutrition |
| Hyperkalemia (K⁺ >6 mEq/L) | Weakness  
Tetany  
ECG changes: peaked T waves, wide QRS, ST segment depression, V-fib | If ↓ urine K⁺ and ↑ 17-OHP, r/o CAH  
If ↓ UOP, r/o renal failure  
If ↓ chloride and metabolic acidosis, r/o type IV RTA |
| Hypocalcemia (total serum Ca²⁺<7 mg/dL, ionized Ca²⁺<1.1 mmol/L) | Clonic seizures, jerking or tetany  
Laryngospasm, stridor, weak cry  
Prolonged QT interval | If ↓ Mg⁺ correct Mg first  
If ↓ serum Phos, r/o high Phos formula or cow’s milk ingestion, ↓ PTH  
If ↓ serum Phos, ↓ urine Ca, r/o renal failure  
If ↓ PTH, ↓ Phos, ↑ AP, r/o Vit D abnormality  
If ↓ PTH, ↓ Phos, ↓ AP, r/o rickets |
| Hypoglycemia (serum glucose <40 mg/dL) | Lethargy  
Seizures | If no thymus on CXR, r/o DiGeorge syndrome  
Often symptom of other illness (sepsis, asphyxia, shock, metabolic disorder)  
If ↓ insulin ↓ insulin-like growth factor 1, r/o hyperinsulinemia |

*r/o* indicates rule out; UOP, urine output; SIADH, syndrome of inappropriate antidiuretic hormone; DTRs, deep tendon reflexes, RTA, renal tubular acidosis; ECG, electrocardiogram; V-fib, ventricular fibrillation; GI, gastrointestinal; 17-OHP, 17-hydroxyprogesterone; Ca, calcium; Mg, magnesium; Phos, phosphorus; PTH, parathyroid hormone; AP, alkaline phosphatase; Vit D, vitamin D; K, potassium; CAH, congenital adrenal hyperplasia; Na, sodium; CXR, chest x-ray.
makes them more susceptible to toxins. Unique neonatal features include increased skin surface area-to-body weight ratio, immature hepatic and renal function resulting in impaired drug metabolism and excretion, and decreased plasma protein binding. All these factors allow drug toxicity at lower doses [14,15]. The routes of neonatal poisoning also differ from those of toddlers and include dermal exposure to toxins in addition to oral ingestion of contaminated breast milk. Dermal routes of exposure can occur if rubbing alcohol (isopropyl alcohol or ethanol) is used for antiseptic cord care or in baths for fever reduction. Systemic absorption of large amounts of topical diphenhydramine may also occur. Oral overdoses of diphenhydramine have been reported in cases of caregivers giving the antihistamine to sedate a fussy infant. Homeopathic colic remedies such as Gripe Water may contain sodium bicarbonate and ethanol and can cause toxicity if given in large quantities [16].

Ingestion of contaminated breast milk is another route for toxin exposure for the neonate. Morphine, methyl mercury, and pesticides have all been implicated in cases of acute neonatal deterioration and a complete maternal history including medications as well as complementary alternative medicines should be obtained for breast-fed infants.

Seizures

Neonatal seizures commonly manifest as subtle motor automatisms such as lip smacking, bicycling movements of the legs, tongue thrusting, apnea, and staring spells. The classic tonic-clonic movements are not typical in neonates, which may lure caretakers to overlook this diagnosis. The underlying etiology and treatment of the clinical seizures will dictate the outcome and ultimate neurologic development. Causes for neonatal seizures may include subarachnoid or subdural hemorrhages, intracranial infections, electrolyte abnormalities, and drug withdrawal. Less common causes are pyridoxine deficiency and local anesthetic toxicity.

The diagnosis and management of seizures related to hyponatremia, hypocalcemia, and hypoglycemia are discussed in greater detail in the “Electrolytes” and “Endocrine” sections of this review. After electrolyte abnormalities are corrected and seizures continue, phenobarbital is the drug of choice, and 20 mg/kg IV should be given to halt seizure activity.

Endocrine Crisis

Endocrine emergencies such as hypoglycemia and hypocalcemia are relatively common in the neonatal period. Other disorders such as CAH and thyrotoxicosis are not routinely encountered in the ED setting but must be considered in critically ill neonates. Calcium metabolism is tightly regulated and involves the concerted effects of parathyroid hormone, vitamin D, and calcitonin on the targeted organs: intestines, kidneys, and bone. Glucose metabolism is also regulated by several different hormones including glucagon and insulin. Surges of epinephrine and norepinephrine after birth contribute to the mobilization of hepatic glycogen [17], gluconeogenesis, and maintenance of plasma glucose concentration. Hepatic glucose production depends on the amount of glycogen stores, gluconeogenic precursors, gluconeogenic and glycogenolytic systems, and an intact endocrine system to coordinate these processes. Abnormalities in glucose metabolism reflects either exposure to irregular maternal glucose metabolism or intrinsic metabolic problems in the neonate [18]. Large for gestational age, which may be associated with diabetes mellitus in the mother, and small for gestational age infants are at increased risk of hypoglycemia.

The clinical presentation and causes of hypocalcemia and hypoglycemia are detailed in Table 3. Similar to electrolyte abnormalities, treatment of hypocalcemia and hypoglycemia include supplementation of the deficient agent. Symptomatic hypocalcemia, which may present as seizures, requires an IV bolus of 100 to 200 mg/kg of 10% calcium gluconate followed by repeat boluses every 6 hours and/or a continuous calcium infusion. After the acute period, some neonates may need vitamin D supplementation and a low-phosphorus formula such as Similac PM 60/40 (Mead Johnson, Evansville, Indiana). Likewise, symptomatic hypoglycemia, which may also present as seizures, requires a 2 mL/kg IV mini-bolus of D10W to rapidly return the blood glucose level to normal without overshooting the desired glucose concentration [19]. This is then followed by a continuous glucose infusion. Determination of the glucose infusion rate (GIR) is determined by the following equation: GIR (mg/kg per minute) = [IV rate (mL/h) × dextrose concentration (g/dL) × 0.167] ÷ weight (kg). Most neonates will be euglycemic with a GIR of 5 to 8 mg/kg per minute. Infants requiring a higher GIR may have hyperinsulinemia. D25W boluses should be avoided.

Congenital adrenal hyperplasia describes a group of disorders with an inherited defect in an enzyme required for the adrenocortical synthesis of cortisol from cholesterol [20]. The lack of glucocorticoids, and occasionally mineralocorticoids, in conjunction with the accumulation of premetabolites (commonly testosterone) result in the clinical manifestations of the affected neonate. The most common is 21-hydroxylase deficiency resulting in the inability to convert progesterone to aldosterone or cortisol and causing an accumulation of testosterone. Female infants classically present with ambiguous genitalia with varying degrees of virilization. However, a male infant may only have a hydropigmented scrotum and no other physical abnormalities and can only be recognized by appropriate laboratory evaluation and results of the newborn screen if available. Affected infants classically have severe salt-wasting and hyperkalemia requiring aggressive fluid resuscitation and exogenous glucocorticoids and or mineralocorticoids. Some infants may be
hypertensive. If CAH is suspected, early consultation with a pediatric endocrinologist is recommended.

Another cause of neonatal endocrine emergencies is thyrotoxicosis. Although rare, affected neonates can present from birth to 6 weeks of age with tachycardia, tremors, sweating, and irritability [21]. The most common cause of neonatal thyrotoxicosis is maternal Graves disease, which results in transplacental passage of thyroid-stimulating hormone receptor stimulating antibodies [22]. A complete history will also reveal the classic complaint of failure to thrive despite hyperphagia [20]. If neonatal thyrotoxicosis is suspected, early consultation with a pediatric endocrinologist is recommended.

**Cardiac Abnormalities**

A decompensating neonate with a yet undiagnosed congenital cardiac defect is perhaps the most daunting patient for many emergency physicians. Most physicians remember the 5 classic cyanotic congenital heart defects: truncus arteriosus, transposition of the great arteries, tricuspid atresia, tetralogy of Fallot, and total anomalous pulmonary venous return. However, most of these patients become symptomatic during the first days of life and are most often diagnosed within the immediate newborn period.

The emergency physician must be armed with a differential diagnosis of cardiac lesions that present after the immediate newborn period to provide appropriate therapy for an affected neonate. Ductal dependent lesions usually present in the first 2 weeks of life with severe hemodynamic compromise. The ductus arteriosus is usually patent in these infants in the immediate newborn period, and these infants can be asymptomatic at the time of discharge from the birth hospital. They remain well-appearing until they develop a dramatic hemodynamic collapse as the ductus arteriosus begins to close. Hence, neonates presenting with sudden appearance of shock require an immediate evaluation for ductal dependent congenital cardiac defects and should receive aggressive IV fluid resuscitation and prostaglandins to maintain the patency of the ductus arteriosus while awaiting evaluation from the cardiac specialists. The ductal dependent lesions are usually of 2 types: left-sided and right-sided obstructive lesions. Left-sided obstructive lesions (aortic stenosis, coarctation of the aorta, interrupted aortic arch, hypoplastic left heart syndrome) present with signs of severe systemic hypoperfusion with pallor, mottling, decreased or absent pulses, severe metabolic acidosis, and cardiomegaly with pulmonary congestion on chest radiograph (CXR). Right-sided obstructive lesions (pulmonary atresia, severe pulmonary stenosis, tetralogy of Fallot, tricuspid atresia) present with severe cyanosis, metabolic acidosis, and decreased perfusion of the lung fields on CXR. Prostaglandin infusion (PGE₂ at a rate of 0.01-0.05 μg/kg per minute) should be immediately started when neonates present with these signs, with or without a cardiac murmur, and an immediate evaluation by a pediatric cardiologist should be requested. Lesions with pulmonary overcirculation (atrial or ventricular septal defects, atrioventricular canal, truncus arteriosus, and partial anomalous pulmonary venous return) can present with evidence of congestive heart failure and respiratory distress; however, the deterioration in these infants is not as dramatic as with ductal dependent lesions. Congestive heart failure may also be due to supraventricular tachycardia and is suspected when the infant's heart rate is more than 220 bpm. A pediatric cardiologist should be consulted as soon as congenital heart disease is suspected.

**Recipe**

Incorrect mixing of formula may contribute to electrolyte imbalances in the neonate ranging from severe hypernatremia (not enough water added) to severe hyponatremia and water intoxication (not enough formula added) (see “Electrolyte Abnormalities”). A thorough history of the mixing of formula is essential.

The recent trend of complementary and alternative medicine has promoted the use of “natural” foods including organic foods as well as the use of supplemental vitamins and homeopathic dietary additives. Nestle has released the first commercial infant formula with added probiotics (bifidobacteria) called “Good Start Supreme® with Natural Cultures” to “help support baby's healthy immune system.” Although studies have yet to show a significant adverse effect of the commercially added probiotics (Lactobacillus, Bifidobacterium) [23-25], concern is raised regarding unregulated addition of probiotics to expressed breast milk or formula by well-meaning care takers. Because these additives are considered dietary supplements, there is no requirement to demonstrate safety, purity, or potency before marketing probiotics [26].

**Enteric Emergencies**

Gastroesophageal reflux disease, omphalocele, tracheo-esophageal fistula, imperforate anus, meconium ileus, bowel atresias, and significant congenital diaphragmatic hernias are often diagnosed and addressed shortly after birth. Hence, they will not be included in the following discussion. Common enteric emergencies in neonates who were otherwise asymptomatic at home are discussed below.

The suspicion of a surgical abdomen can be raised even with a limited history and physical exam. A history of feeding intolerance or bilious emesis with physical findings of a tender, distended abdomen with discoloration of the overlying skin should prompt immediate consultation with a surgeon skilled in neonatal care.

A true surgical emergency is the neonate with malrotation with or without midgut volvulus. A contrast study, usually an upper GI, is required to rule out malrotation. Visualization of a dilated duodenum, abnormal positioned duodenojejunal junction with right-sided jejunal loops, and a malpositioned cecum
on a contrast study confirms malrotation of the intestines [27]. If a “corkscrew” appearance of the jejunum is seen, midgut volvulus is present. Time is of the essence because intestinal necrosis can rapidly occur if there is a delay in making this diagnosis. Definitive surgical treatment consists of an exploratory laparotomy and Ladd’s band procedure.

If an neonate is previously premature, is an infant of a cocaine-using mother, or has a history of surgical closure of an abdominal wall defect, necrotizing enterocolitis must be considered. Abdominal radiographs may reveal intestinal dilatation and thickened bowel walls. The presence of pneumatosis intestinalis or portal venous gas is pathognomonic for necrotizing enterocolitis. If there is no free air or evidence of peritonitis, medical management should consist of IV antibiotics and bowel rest. If free air or peritonitis is found, surgical intervention is indicated.

Hirschsprung disease should be considered in the neonate with a history of failure to pass meconium or infrequent stools and abdominal distension. A rectal biopsy is diagnostic and demonstrates the absence of ganglion cells. If there is rapid progression of abdominal distension and profuse vomiting, toxic megacolon must be assumed and emergent surgical treatment with a diverting colostomy may be necessary.

Trauma (After the Perinatal Period)

Neonates are not immune from accidental injuries such as motor vehicle accidents, falls, and the antics of overzealous older siblings. Unfortunately, they may also be the victims of adult anger, frustration, and anxiety. An obtunded, encephalopathic neonate may have IV a metabolic disorder or sepsis, but trauma must also be considered. Intracranial pathology includes hemorrhage, edema, contusional tears, and diffuse axonal injury [28].

Other organs can be involved in trauma. The musculoskeletal system can also be affected by fractures of varying degrees, but bony injury alone is rarely a cause of acute clinical deterioration of a neonate. However, if associated with a pneumothorax or liver laceration, emergent resuscitation may be required. Blunt abdominal trauma can result in adrenal hemorrhage, which can lead to hypovolemia and shock. Hence, radiographic imaging and ultrasonography play a crucial role in the evaluation of a critically ill neonate who has been the victim of a trauma. Pediatric neurosurgeons and pediatric general surgeons should be consulted early in the management of an injured neonate to help provide prompt surgical intervention if needed and collaborate with the medical team in maximizing the infant’s outcome.

Sepsis

Sepsis is intentionally discussed last so as to give all other potential diagnoses careful consideration before attributing a neonate’s deterioration to sepsis. Nonetheless, neonatal sepsis is a well-known cause of neonatal collapse. Common microbes of concern are GBS, Listeria monocytogenes, Escherichia coli, and Staphylococcus aureus. Maternal history of GBS status and penicillin prophylaxis at the time of delivery may be helpful but late-onset GBS is commonly community acquired. It is important to note that intrapartum and early neonatal antibiotic treatment does not prevent late-onset GBS meningitis, pneumonia, or sepsis. Maternal history of a cervical cerclage during pregnancy may increase the risk for gram-negative sepsis in the neonate, whereas a history of brown-colored amniotic fluid (which may be mistaken as meconium-stained fluid) may suggest Listeria.

Prompt initiation of broad-spectrum IV antibiotics such as ampicillin and gentamycin or ampicillin and cefotaxime is essential. Ampicillin is effective against GBS, Listeria, and S aureus, whereas gentamycin is effective against most gram-negative bacilli. If ampicillin and cefotaxime are used, remember that several strains of β-lactamase–producing E coli have become resistant to these drugs [29] and the threshold to add or change cefotaxime to gentamycin in the neonate with suspected gram-negative sepsis must be low.

Viral infections include herpes simplex virus and enterovirus and can also compromise a neonate. Disseminated neonatal herpes simplex virus infection is clinically indistinguishable from sepsis and should be considered in the differential of sepsis. A history of maternal genital lesions or respiratory symptoms may be helpful but are often negative. If sepsis is highly suspected, consider a viral infection as well. In addition to blood cultures, send surface cultures and blood or cerebrospinal fluid for polymerase chain reaction assays for the suspected virus. Acyclovir should be started in septic-appearing neonates with signs of encephalopathy and/or disseminated intravascular coagulopathy. Because pleconaril is no longer available for the treatment of neonates with enteroviral infections, supportive treatment must be maximized. A detailed review of neonatal sepsis is presented later in the article on “Evaluation and management of a newborn with sepsis.”

Summary

Neonatal symptoms of illness are limited and nonspecific, requiring the treating physician to be astute and complete in the care and management of these patients. Regardless of the diagnosis, the management of the critically ill neonate is still dependent on a rapid assessment and support of airway, breathing, and circulation. Although the history and physical examination may help narrow the differential diagnosis, beginning resuscitation and ensuring thermoneutrality are essential for the successful management of any critically ill neonate. Laboratory evaluation can be directed toward a specific diagnosis or can be broad enough to allow for the identification of a diagnosis or tracking changes in the status of the neonate. Although the
differential diagnosis is extensive, a systematic approach to initial resuscitation, followed by application of the “NEO SECRETS” mnemonic will allow the treating physician to be focused and able to implement time-efficient interventions that will lead to the best possible outcome for the critically ill neonate.

References