The Neonate

Patrick Myers
Guest Editor

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Normal Infant or Sick Newborn: The Challenge of Evaluating an Infant in the Emergency Department

By Patrick J. Myers, MD

There are about 3,900,000 babies born in the United States every year. The vast majority of babies will require no medical care or intervention, but all of them will be thoroughly evaluated before discharge from the newborn nursery. About 10% of infants require limited help in the delivery room, with only 1% requiring significant support. Just after delivery, most infants quickly transition to routine postdelivery care which consists of being dried, warmed, and placed with the infant’s mother. The next 48 hours consists of learning to feed, bonding with his or her new family, and monitoring expected weight loss. Before discharge, infants undergo a battery of screening tests to identify those infants with an underlying abnormality. Those screening tests include total bilirubin, hearing evaluation, metabolic testing, and pulse oximeter screening for congenital heart disease. Former preterm infants will also receive a car seat test and may have had a head ultrasound. Despite the extensive postdelivery evaluation of all newborns, a small number of infants will return to the emergency department for additional workup. These recently normal but now sick infants can be hard to evaluate for a variety of reasons.

The evaluation of a young infant or neonate can be challenging because normal physical examination findings and laboratory results for an infant can be considered pathological at other times of life. In the first days of life, basic electrolytes may not herald an electrolyte abnormality but rather abnormalities in fluid balance. Vital signs are both different than those for other children but also change over the course of the first days and weeks of life. The presence of jaundice on physical examination can be normal if laboratory data support an acceptably low bilirubin level. Infants often have a combination of normal physical findings that in other settings would be alarming such as a respiratory rate in the 50s, right ventricular hypertrophy, and a liver slightly below the costal margin. Actual disease presentation can also be difficult in newborns because they present with either limited or different physical findings than older patients.

Infants also are required to make the unique transition of being almost fully supported by the
placenta to being self-sufficient in a matter of seconds. This transition causes some unique and often temporary disease states such as transient tachypnea of the newborn, pulmonary hypertension, hypoglycemia in an infant of a diabetic mother, and newborn jaundice. These will all resolve if correctly identified and if the infant is supported.

Conversely, pathological diseases and unsustain-able congenital malformations are unmasked once the support of the placenta is removed. Congenital heart disease is the prime example of this type of condition. In utero, the fetus is frequently capable of surviving with heart disease that requires immediate surgical correction. In addition, infants are born with a patent ductus arteriosus that closes in the first 24 to 72 hours of life, and once the patent ductus arteriosus closes, it can unmask significant heart disease. Metabolic syndromes are also often protected against by the placenta and only manifest once metabolites build up over the first several days of life. Being a newborn means that it is the first time that the infant will do almost anything: breathe, stool, eat, and hear. Each of these firsts is a moment that can unveil pathology: choanal atresia, tracheoesophageal fistula, imperforate anus, and a myriad of other structural abnormalities.

Even the medications and therapy we provide infants are different. The rules for medications can seem complex with the dosing amount often based on a combination of birth weight, gestation age, and day of life. Changing renal and liver functions, over the first few days of life, can require monitoring of drug levels and dosing changes. Commonly used medications can have unique physiological interactions in newborns that limit their usefulness. As an example, ceftriaxone displaces bilirubin from albumin and is not recommended for neonates. Phototherapy, high-frequency oscillators, and umbilical lines are all therapies that are unique to the practice of newborn medicine.

The contrast between the overwhelming wellness of the vast majority of infants in the first 30 days of life and the subtle presentations of disease is the most problematic aspect of treating the newborn. To combat this difficulty, developing a strong sense of normal vs abnormal is important but not enough. Because most infants will end up being normal, the temptation is to treat all infants as well. The reverse approach, imagining each infant as ill, proves to be more fruitful with a careful questioning approach for each infant. For each infant, it is valuable to ask a series of questions and then rule the infant in or out based on the answers received. What are the parents’ concerns, and are these concerns normal findings among newborns? Does the infant partake in normal infant behavior, stool enough but not too much, eat without effort, have the appropriate amount of wet diapers, spit up but not have emesis, soothe when comforted, and respond when disturbed? Are there physical findings that are unacceptable in a newborn, such as: bilious emesis, lethargy, grayness, and bulging fontanel? Are laboratory findings and imaging consistent with normal infant values?

Once these questions are asked and answered, the clinician can move onto disposition and treatment. Infants, particularly former preterm infants, tend to have less reserve and are more likely to require readmission or transfer to a higher level of hospi-
tal. If doubt remains, consultation with the appropriate specialist can aid definitive medical therapy. Finally, communication with the infant’s medical home or primary care physician can help the family and child transition to home.

The articles in this issue of Clinical Pediatric Emergency Medicine address the challenges that can surround the newborn presenting to the emergency department. This issue reviews how to address the birth and death of an infant in the emergency department, how to transport an infant, approaches to the “fussy baby,” and ways to evaluate and treat common neonatal illnesses.

REFERENCES

Management of an Unexpected Delivery in the Emergency Department

Arika G. Gupta, MD*, Mark D. Adler, MD†

For those who do not routinely care for newborn infants in the immediate postdelivery period, facing a sudden unexpected need to fill the role can be anxiety provoking, particularly if the infant requires care beyond the routine. Successful transition from the intrauterine to extraterine environment is dependent on several significant physiologic changes that must occur at the time of birth. Most (approximately 90%) infants effectively transition at delivery without requiring any special assistance. However, about 10% of infants will require some level of intervention, and 1% will require extensive resuscitative measures at birth. The focus of this article is on the preparation for and management of an unexpected delivery in the emergency department. We will highlight the unique aspects of newborn resuscitation, as well as recent changes to the Neonatal Resuscitation Program from the 2015 American Heart Association Guidelines.

Keywords: neonatal; neonatal resuscitation; newborn; unexpected delivery; precipitous delivery; delivery; emergency department

Abstract: Successful transition from the intrauterine to extraterine environment is dependent on several significant physiologic changes that must occur within minutes of birth. Most infants effectively transition at delivery without requiring any special assistance. However, about 10% of infants will require some level of intervention, and 1% will require extensive resuscitative measures at birth. The focus of this article is on the preparation for and management of an unexpected delivery in the emergency department. We will highlight the unique aspects of newborn resuscitation, as well as recent changes to the Neonatal Resuscitation Program from the 2015 American Heart Association Guidelines.

CASE PRESENTATION

A 15-year-old patient is brought to the ED by her parents for a complaint of severe abdominal pain at home. She has received care at your institution before for orthopedic issues and thus chose to come to your hospital for this issue as well. She has had episodic abdominal pain that has been recurring every 3 minutes, nausea, and a report of recent weight gain. She cannot tell you her last menstrual period. Your examination quickly reveals what the history strongly suggested: she is pregnant and in active labor. You have staff with ultrasound training on the unit and their examination reveals an intrauterine fetus. On pelvic examination, the patient is fully...
dilated, and the fetus is crowning. You immediately begin preparing for a precipitous delivery in the ED and make arrangements for the appropriate teams to be notified.

Some of the most important considerations in this situation include the following:

1. What are the key questions to ask to prepare for the impending delivery of a neonate?
2. What needs to be prepared for the resuscitation of this infant?
3. What are the principles and steps behind resuscitation of a newborn infant?

**PREPARING FOR AN UNEXPECTED DELIVERY**

First, take a deep breath and remember that 90% of babies require no assistance at birth and transition to the extrauterine environment appropriately all on their own. However, given that the remaining 10% of neonates require some intervention at birth, we must ensure we are prepared to support any newborn that requires assistance. In some instances, there may be time to discuss an impending delivery with a neonatologist; however, in other cases, the ED team must respond rapidly and rely exclusively on their own expertise.

To prepare efficiently and effectively, it is helpful to know what questions you must ask the patient before the delivery. These questions include the following:

1. Did the patient receive prenatal care? Was the pregnancy known to the patient?
2. How many babies are expected to be delivered?
3. Approximate gestational age in weeks or date of last menstrual period?
4. Any major complications during the pregnancy or labor (eg, gestational diabetes, gestational hypertension, concerns about fetal growth, maternal infection or fever, prolonged rupture of membranes)?

On the basis of this information (or lack thereof), you can begin preparing the appropriate number of team members and resuscitation supplies. If time permits, it is helpful to prebrief with the team to review the plan for resuscitation, assign roles, and delegate tasks. In addition, obtaining further history can be valuable, such as finding out if there were any known anatomic abnormalities on prenatal ultrasound. There are certain prenatal abnormalities, such as congenital diaphragmatic hernia, congenital heart disease, anterior abdominal wall defects, or lumbosacral defects, to name a few, which would affect your immediate evaluation and resuscitaion of the infant and prompt more immediate consultation with a neonatologist.

It is critical to have separate teams with predesignated roles, with one team to manage the mother and the other team for the newborn. The focus of this article is on preparing for and managing the neonate; the care of the mother, including maternal labor or delivery complications, will not be discussed. Given the low-frequency, high-stakes nature of an unexpected delivery in the ED, a standardized checklist of supplies and equipment for a newborn resuscitation is helpful to ensure that all necessary items are prepared and checked before delivery. Table 1 provides a list of recommended supplies and equipment for an impending delivery. Because of the rarity of such events, the supplies and equipment may be more difficult to find or may be missing, unlike frequently used ED equipment. It is useful to identify where these supplies are kept and have a process to ensure they are checked and stocked on a regular basis. Unplanned deliveries are sufficiently challenging without this additional distractor.

**PRINCIPLES AND STEPS OF NEWBORN RESUSCITATION**

The American Heart Association (AHA) published updated guidelines on neonatal resuscitation in November 2015. These guidelines are meant to apply to newly born infants who require assistance

<table>
<thead>
<tr>
<th>TABLE 1. List of equipment needed for neonatal resuscitation.</th>
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<tbody>
<tr>
<td>Radiant warmer</td>
</tr>
<tr>
<td>Warm blankets/towels</td>
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<tr>
<td>Hat</td>
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<tr>
<td>Plastic wrap (such as NeoWrap)</td>
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<tr>
<td>Thermal mattress</td>
</tr>
<tr>
<td>Bulb suction, suction catheter, and suction tubing</td>
</tr>
<tr>
<td>Neonatal face mask (all sizes)</td>
</tr>
<tr>
<td>Endotracheal tubes (all neonatal sizes: 2.5, 3.0, 3.5, and 4.0), stylet (optional)</td>
</tr>
<tr>
<td>Laryngoscope blades (all neonatal sizes: 00, 0, 1 Miller)</td>
</tr>
<tr>
<td>Bag valve mask device (ideally flow-inflating bag; however, self-inflating bag would be adequate)</td>
</tr>
<tr>
<td>Laryngeal mask airways (size 1 neonatal)</td>
</tr>
<tr>
<td>Oxygen source and blender</td>
</tr>
<tr>
<td>O₂ saturation probe and monitor</td>
</tr>
<tr>
<td>Cardiac leads and monitor</td>
</tr>
<tr>
<td>CO₂ detector</td>
</tr>
<tr>
<td>Tape</td>
</tr>
<tr>
<td>Umbilical line kit</td>
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<tr>
<td>Stethoscope</td>
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in making the transition to the extraterine environment but are also applicable to neonates who require resuscitation in the first few weeks after birth. Neonatal cardiac arrest is predominantly caused by asphyxia; as a result, initiation of effective ventilation remains the mainstay of initial neonatal resuscitation, whether in the delivery room or in the few weeks after birth.

A summary of these guidelines, with an emphasis on the key aspects relevant to the immediate management of an unexpected delivery in the ED, will be reviewed here. Please refer to Figure 1 for a stepwise approach to neonatal resuscitation.

The most important 3 questions to ask upon delivery of the neonate include the following:

1. Term gestation?
2. Good tone?
3. Breathing or crying?

If all 3 questions are answered “yes,” then the newborn may stay with the mother and receive routine newborn care, which entails keeping the newborn warm to avoid hypothermia. Infants that are born precipitously either at home or on the way to the hospital should be triaged similarly to the infant born in the ED by asking the same 3 questions.

If any of the above questions are answered “no,” then the infant should be immediately placed under a preheated radiant warmer for a full assessment by the infant’s resuscitation team. The steps to be performed (see Figure 1) are summarized here and then detailed further below:

1) Initial stabilization: includes warming and drying the infant, positioning the airway, clearing secretions from the airway (if needed), and stimulating the infant, including flicking the soles of the feet or rubbing the infant’s back.

2) If heart rate (HR) is below 100 beats per minute (bpm) at the first assessment (by 1 minute of life) or if the infant has inadequate respiratory effort (apnea, gasping, hypopnea), then effective ventilation and oxygen delivery must be delivered, via bag valve mask (BVM). Multiple corrective measures to ensure effective ventilations must be attempted, including endotracheal tube (ETT) or laryngeal mask airway (LMA) placement, if necessary.

3) If HR is below 60 bpm, despite adequate ventilation (including advanced airway placement), initiate chest compressions/breaths at a ratio of 3:1. Note the difference between this rate and the pediatric advanced life support algorithm with a 15:2 ratio. You may need to be explicit with your team if they are not familiar with the neonatal resuscitation recommendations.

4) If HR remains below 60 bpm, despite intubation, effective ventilation/oxygenation, and chest compressions, intravenous epinephrine (see access approaches below) should be administered, followed by consideration of other causes of hemodynamic compromise, including hypovolemia, tension pneumothorax, or cardiogenic shock.

SPECIAL CONSIDERATIONS AT THE TIME OF DELIVERY

Delayed Cord Clamping

On the basis of data from a recent systematic review,

delayed umbilical cord clamping is now recommended for both term and preterm infants who do not require resuscitation at birth, when possible. Delayed umbilical cord clamping is defined as clamping the umbilical cord approximately 30 to 60 seconds after birth. Delayed umbilical cord clamping has been associated with less all-grade intraventricular hemorrhage (IVH), higher blood pressure and blood volume, decreased need for postnatal transfusion, and lower incidence of necrotizing enterocolitis. Delayed umbilical cord clamping has not been shown to decrease mortality or incidence of severe IVH. The only adverse consequence of delayed umbilical cord clamping found was a slightly increased level of bilirubin, associated with more need for phototherapy.

Thermoregulation

Hypothermia is known to cause an increase in oxygen consumption and metabolic demand and is independently associated with increased morbidity and mortality in the neonate of all gestational ages. Low-birth weight and preterm infants, in particular, are at increased risk of hypothermia due to rapid loss of body heat due to their large body surface: body mass ratio, thinner skin, and decreased subcutaneous fat. Therefore, it is vital to take all measures to ensure appropriate thermoregulation after birth. It is recommended that the temperature of a newborn infant be maintained between 36.5°C and 37.5°C (97.7°F–99.5°F) through stabilization. There is a commensurate increase in mortality for temperatures below 36.5°C. Strategies to ensure thermoregulation

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Neonatal Resuscitation Algorithm—2015 Update

Antenatal counseling
Team briefing and equipment check

Birth

Term gestation? Good tone? Breathing or crying?

Yes

Infant stays with mother for routine care: warm and maintain normal temperature, position airway, clear secretions if needed, dry. Ongoing evaluation

No

Warm and maintain normal temperature, position airway, clear secretions if needed, dry, stimulate

Apnea or gasping? HR below 100/min?

Yes

PPV
SpO₂ monitor
Consider ECG monitor

No

Labored breathing or persistent cyanosis?

Yes

Position and clear airway SpO₂ monitor
Supplementary O₂ as needed
Consider CPAP

No

HR below 100/min?

Yes

Postresuscitation care
Team debriefing

Check chest movement
Ventilation corrective steps if needed
ETT or laryngeal mask if needed

No

HR below 60/min?

Yes

Intubate if not already done
Chest compressions
Coordinate with PPV
100% O₂
ECG monitor
Consider emergency UVC

No

IV epinephrine
If HR persistently below 60/min
Consider hypovolemia
Consider pneumothorax

Targeted Preductal SpO₂
After Birth

<table>
<thead>
<tr>
<th>Time (min)</th>
<th>SpO₂ (%)</th>
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<tbody>
<tr>
<td>1</td>
<td>60%-65%</td>
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<tr>
<td>2</td>
<td>65%-70%</td>
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<tr>
<td>3</td>
<td>70%-75%</td>
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<tr>
<td>4</td>
<td>75%-80%</td>
</tr>
<tr>
<td>5</td>
<td>80%-85%</td>
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<tr>
<td>10</td>
<td>85%-95%</td>
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</table>

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Figure 1. 2015 Neonatal Resuscitation Algorithm. Reprinted with permission from Circulation (2015;132:S543-S560). Copyright 2015, AHA.
in the delivery room, or in this case the ED, include use of a preheated radiant warmer, plastic wrap for preterm neonates (Figure 2), warm and dry linens to dry and swaddle infant, hat, increased room temperature, thermal mattresses, and use of warmed humidified resuscitation gases.

ELEMENTS OF NEONATAL RESUSCITATION

Airway

If a newborn is demonstrating signs of airway obstruction or inadequate ventilation requiring positive pressure ventilation (PPV), it is appropriate to open the mouth, if needed, adjust the airway (using chin tilt or jaw thrust), and/or suction secretions from the airway. However, it is recommended that unnecessary suctioning of the nasopharynx be avoided given the risk of inducing a vagal response and reflexive bradycardia. In addition, the most recent AHA guideline no longer recommends routine tracheal intubation in the setting of a depressed newborn with meconium-stained amniotic fluids, as had previously been recommended. The focus in these patients should be on initiating ventilation within the first minute of life if the infant remains depressed and is not breathing or ineffectively breathing.

Breathing

Despite the initial steps of drying, stimulation, and airway maneuvers, if the infant’s HR is under 100 bpm at the first assessment at about 1 minute of life, it is essential to promptly begin PPV via BVM at a rate of 40 to 60 breaths per minute. This applies to all newly born infants, regardless of the amniotic fluid appearance. If PPV is initiated, the team should consider placing a pulse oximetry probe on the right upper extremity (preaductal) to monitor oxygen saturations. Resuscitation of all infants, including preterm infants, should be initiated with low oxygen concentrations (21% ideally, but up to 30% for preterm infants is reasonable). The oxygen concentration should subsequently be titrated to achieve target preaductal oxygen saturations based on minutes of life, as seen in Figure 1. It is appropriate for a newborn with normal transition to have a preaductal saturation ranging between 60 and 70% in the first minute of life with a gradual increase until about 10 minutes of life when the preaductal saturations typically increase to 85 to 95\%. If a blender device that allows the fine titration of FiO\textsubscript{2} is not available, it is reasonable to initiate PPV with room air and increase to 100% FiO\textsubscript{2}, if resuscitation is necessary. The AHA recommendation to avoid administration of excessive oxygen is an attempt to limit the potential deleterious effects of hyperoxia.

Throughout the resuscitation, once PPV has been initiated, the team should be consistently reevaluating the infant’s HR, spontaneous respiratory effort, effectiveness of assisted ventilation, and the preaductal oxygen saturations. Ventilation corrective measures should be considered if assisted breaths are not effective or HR does not quickly begin to rise. An increase in the newborn’s HR is one of the most sensitive indicators we have during a resuscitation of the effectiveness of ventilation. If despite effective ventilations, the HR does not rise, the team should consider placement of an advanced airway, including an ETT or LMA. Laryngeal mask airways may be considered as an alternative to tracheal intubation in infants ≥34 weeks or ≥2 kg when tracheal intubation is unsuccessful or not feasible.

When administering PPV, it is critical to assess the following frequently:

- Heart rate
- Spontaneous respiratory effort
- Effectiveness of assisted breaths
- Preaductal oxygen saturations

Figure 2. Premature neonate wrapped in plastic wrap immediately after delivery to optimize thermoregulation.
If chest compressions are necessary, it is recommended that an advanced airway be placed prior, to ensure effective ventilation as well as to optimize compression performance. In addition, if compressions are being given, it is appropriate to increase the FiO₂ to 100% until the HR recovers and then titrate down the oxygen concentration once the HR improves.

If at any point in the resuscitation, the HR is greater than 100 bpm and the neonate is breathing spontaneously, but is demonstrating signs of respiratory distress (labored breathing or grunting) or has persistent cyanosis, the team should consider initiation of continuous positive airway pressure. In the setting of the ED, continuous positive airway pressure is probably most easily provided using a flow-inflating bag with the positive end-expiratory pressure set to 5 to 6 cm H₂O.

**Circulation**

As noted above, a newborn's HR is evaluated frequently throughout a resuscitation and is the most sensitive indicator of effective ventilation, whether from the infant's spontaneous respiratory effort or delivered mask ventilations. Typically, either auscultation of the HR or palpation of the pulse at the base of the umbilicus is the most effective method used to evaluate HR in the first minute of life. Previously, it was recommended to use pulse oximetry to supplement this assessment if the infant requires interventions. However, the newest recommendation is to use a three-lead electrocardiogram for monitoring HR during a resuscitation, in addition to continuous pulse oximetry for evaluation of oxygen saturation.⁴,¹⁰,¹¹

If the HR remains less than 60 bpm despite adequate ventilation, including placement of an advanced airway, chest compressions should be initiated at a ratio of 3 compressions: 1 breath for a total of 120 events per minute. Compressions and ventilations should be coordinated to avoid simultaneous delivery, allowing the chest to fully recoil between compressions and optimize lung expansion during assisted ventilation. Note that this differs from pediatric advanced life support in which compressions and breaths are not coordinated after intubation.⁵ In terms of technique, the two-thumb technique is the preferred method for compressions, as it has been shown to be more effective and is associated with less rescuer fatigue.⁶,¹³ The two-thumb technique involves encircling the newborn's torso with the thumbs placed on the lower third of the sternum and the fingers under the infant's back, supporting the spine.

**Medications and Fluids**

Given that most cases of neonatal cardiac arrest or bradycardia are secondary to inadequate ventilation, establishing adequate ventilation remains the foundation of neonatal resuscitation. However, in situations in which the HR remains below 60 bpm despite adequate ventilation with an advanced airway and 100% FiO₂ and chest compressions, it is appropriate to administer either epinephrine or a volume expander, or both. For persistent bradycardia, intravenous epinephrine at a dose of 0.01 to 0.03 mg/kg, of 1:10 000 concentration, may be considered. If venous access is not available, the resuscitation team may consider a dose of endotracheal epinephrine at a dose of 0.05 to 0.1 mg/kg, until intravenous access is established. In such circumstances, once intravenous access has been obtained, an intravenous dose of epinephrine should be given immediately, irrespective of when the endotracheal dose was given. Options for emergency intravenous access in a neonate include placement of a peripheral intravenous catheter, low-lying umbilical venous catheter (UVC), or intraosseous line (IO).⁴–¹⁷ In situations requiring emergent access, the peripheral venous system can often be difficult to catheterize due to peripheral vasoconstriction. Therefore, UVC or IO placement may be more practical until the neonate is effectively resuscitated and stabilized. The decision of which (UVC vs. IO) to place emergently should ultimately be guided by the clinician's experience and comfort level with each of the procedures. The specific procedural steps or indications and contraindications of these various lines are beyond the scope of this article.

When there is a known history of blood loss during delivery and/or signs of hypovolemic shock in the newborn, volume expansion may be considered when the infant has failed to respond to all other resuscitative measures. Appropriate volume expanders include isotonic crystalloid solution or emergency blood, at a dose of 10 mL/kg. In premature infants, it is recommended to avoid giving these volume expanders rapidly, as this may be associated with increased risk of IVH.¹⁸
POSTRESUSCITATION CARE, MONITORING, AND EVALUATION

After initial assessment and resuscitation of the newborn, it is important to ensure that the appropriate monitoring, treatment, and stabilization are provided while arrangements are made for transfer to an appropriate level of nursery.

- Thermoregulation—Hypothermia is associated with increased neonatal morbidity and mortality, including increased risk of respiratory problems, metabolic derangements, IVH, and late-onset sepsis. Likewise, hyperthermia should be avoided in neonates, as it can be associated with an increase in mortality and morbidity, including meconium aspiration syndrome, respiratory distress syndrome, neonatal seizures, and need for assisted ventilation. Therefore, the target temperature for a normal newborn, independent of gestational age, is between 36.5°C and 37.5°C (97.7°F-99.5°F).

- Therapeutic hypothermia—in special circumstances for infants ≥36 weeks gestation who have suspected moderate to severe hypoxic-ischemic encephalopathy, induced therapeutic hypothermia should be offered and implemented at a neonatal intensive care unit or other intensive care unit with the appropriate resources, monitoring, and nursing staff for this treatment protocol. If the newborn meets criteria for therapeutic hypothermia, treatment should ideally be initiated by 6 hours of life, when possible. Induced therapeutic hypothermia, when used in this special circumstance, has been associated with reduced mortality and major neurodevelopmental disability to 18 months of age.

- Glucose monitoring—Infants who require resuscitation or have other risk factors for dysregulated glucose levels (such as infants who are growth restricted, who are small for gestational age, who are large for gestational age, have exposure to gestational diabetes, have suspected sepsis, or are born prematurely), require close monitoring of their glucose levels to ensure glucose homeostasis in the first several hours after birth. Symptoms of hypoglycemia in a neonate include lethargy, poor feeding, hypotension, apnea, irritability, hypotonia, tremors, jitteriness, or seizures. In neonates, the majority of circulating glucose is used by the brain; therefore, prompt administration of intravenous dextrose, in the setting of hypoglycemia, is critical to optimize a newborn's neurodevelopment. If hypoglycemia is present, a bolus of intravenous D10% water solution is given at a volume of 2 mL/kg, followed by initiation of intravenous dextrose-containing fluids, not saline-based fluids. Typically, a D10% water solution is used as the maintenance fluid in a newly born infant to provide a dextrose infusion rate of about 4 to 6 mg/kg/min, which is equivalent to total fluids of 60 to 80 mL/kg/day. Frequent reassessment of blood glucose measurements should be taken until the newborn's glucose level has normalized, and adjustments in fluids or administration of additional boluses should be provided, as necessary. In fact, any neonate that is critically ill at birth who will require intensive care monitoring and will remain Nil per os (NPO) for a period should be initiated on D10%-containing intravenous fluids to avoid dehydration and hypoglycemia.

- Critical prenatal labs—When an infant deliveries precipitously and unexpectedly, the mother's medical and obstetrical history may not be known at the time of delivery. For all newborn infants, it is critical to know the following maternal lab results as soon as possible after delivery: hepatitis B status, human immunodeficiency virus status, syphilis testing results, and blood type and antibody testing. These laboratory tests, in addition to other routine prenatal labs, are performed in all pregnant women who receive prenatal care, as the results of these tests may affect the outcome of the pregnancy for the mother and or/the fetus. Refer to Table 2 for a list of these critical prenatal labs and immediate actions to be taken, based on maternal lab results. There are other prenatal labs that are not reviewed in Table 2, but are also important for the newborn care provider to be aware of, including group B streptococcal culture, chlamydia testing, gonorrhea testing, rubella screening, and gestational diabetes screening.

- Erythromycin eye ointment—In the United States, it is mandatory that all newborns receive prophylaxis against gonococcal eye infection by administration of ophthalmic antibiotic agents shortly after birth, ideally within the first hour of life. The current standard treatment, recommended by the American Academy of Pediatrics and Centers for Disease Control and Prevention, is 0.5% erythromycin ophthalmic ointment, with application of a 1-cm ribbon in each eye, in
the lower eyelid. The ointment should be spread by gentle massage of the eyelids, and excess medication can be wiped away after 1 minute. Evaluation of the eye, including the red reflex and pupillary response, can be difficult for several hours after administration of the ointment, so it is recommended to try and complete the eye examination before administration of this medication.

- Vitamin K administration—The American Academy of Pediatrics recommends vitamin K$_1$ be given to all newborns as a single intramuscular dose of 0.5 to 1 mg to prevent both early- and late-onset vitamin K deficient bleeding, previously referred to as hemorrhagic disease of the newborn. This intramuscular medication is typically given in the anterolateral aspect of the proximal thigh. The gluteus and deltoid muscles are important to avoid in a neonate, due to concern for nerve injury and absorption at these sites. Administration of vitamin K$_1$ can be deferred to the destination hospital, if transferring, but it should be made clear to the receiving institution whether this has been given to avoid errors.

- Apgar scoring—The Apgar scoring system was first devised by Dr. Virginia Apgar and was designed to be a quick method to assess the clinical status of the newborn infant. The Apgar score is composed of 5 components, each of which are given a score of 0, 1, or 2. Score components include: (1) color, (2) HR, (3) reflex irritability, (4) muscle tone, and (5) respiration. The patient is assessed, and the score is reported at 1 and 5 minutes after birth. However, if the 5-minute score is less than 7, then scoring
continues every 5 minutes thereafter (until 20 minutes of life) until the score is 7 or higher. This scoring system is a universally accepted and convenient standardized assessment tool of the newborn infant’s status after birth, as well as their response to resuscitative efforts. However, this score should not be used to predict future outcomes, particularly mortality or adverse neurological outcomes.32

SUMMARY

This article focuses on the preparation and management of an unexpected delivery in the ED. Although most newborns transition appropriately without intervention, about 1% of newborns require extensive resuscitation at birth. Because of the unanticipated nature of such an event, it is critical to ensure that the ED has processes in place to effectively and efficiently manage a newborn resuscitation. We provide an itemized checklist of equipment needed for such a situation, discuss principles of neonatal resuscitation, review the neonatal resuscitation algorithm, and provide a brief overview of immediate postresuscitative care. In such low-frequency, high-stakes situations, such as an unexpected delivery outside of the delivery room, simulation-based training can be a useful adjunct to clinical experience, as it can provide a forum for learning, practicing, and perfecting complex medical management.32

REFERENCES

The idea that “children are not just small adults” is pervasive in pediatrics.1,2 Neonatologists take this adage one step further, reminding trainees that “neonates are not just small children.” When a neonate presents to the emergency department (ED), it is essential that health care providers recognize presenting signs and symptoms of neonatal conditions whose accurate diagnosis requires a keen awareness of the distinction between “sick” and merely “fussy” newborns.

**APPRAOCH TO THE FUSSY INFANT**

**Clinical Presentation**

Crying up to 2 hours per day is normal in neonates. “Colic” is traditionally defined as crying more than 3 hours a day, 3 days a week for 3 weeks or longer in an otherwise well-fed, growing, and healthy infant.3 The crying might begin suddenly and for no apparent reason, during which time the infant is difficult or impossible to console. When a newborn presents to the ED with crying, it is important to exclude illness so as to provide appropriate reassurance. Ensure that the baby is being appropriately fed, changed, soothed, and not
Sources of crying not to miss in the neonatal period include the following:

- **Sepsis/meningitis**: A neonate with sepsis or meningitis may present as a fussy, irritable infant who cannot be easily consoled (see below).
- **Hair tourniquet**: Perform a full body examination of the infant to exclude a hair tourniquet, typically involving a thread of hair wrapped around the toes, fingers, or genitalia with associated pain, erythema, and edema.
- **Corneal abrasion**: An unusually fussy infant who is unwilling to open her eyes may be suffering from a corneal abrasion. Neonates may scratch the cornea with sharp fingernails, causing severe eye pain, inability to open the eye, watery eye(s), and light sensitivity.
- **Anomalous origin of the left coronary artery from the pulmonary artery**: Although rare, anomalous origin of the left coronary artery from the pulmonary artery is life-threatening and should be considered in a neonate who presents with inconsolable crying. Shortly after birth, pulmonary artery pressure and pulmonary vascular resistance (PVR) decrease, resulting in the left ventricular myocardium being perfused by relatively desaturated blood under low pressure, leading to myocardial ischemia. Initially, this myocardial ischemia is transient during periods of increased myocardial demand such as crying and feeding. Inadequate myocardial perfusion causes chest pain (angina), which may be misinterpreted as “colic.”

**APPROACH TO NEONATAL RESPIRATORY DISTRESS AND HYPOXEMIA**

“Doctor, my baby is breathing fast. Is this normal?” In contrast to older children, a normal neonatal respiratory rate ranges from 30 to 60 respirations/min (Table 1). New parents are often concerned by periodic breathing, a benign neonatal breathing pattern characterized by pauses for up to 10 seconds followed by a series of rapid, shallow breaths with spontaneous resolution. In contrast, retractions, grunting, and nasal flaring suggest the presence of parenchymal lung disease with decreased lung compliance and associated respiratory distress. Upper airway obstruction and metabolic acidosis with respiratory compensation can present as respiratory distress in neonates. Cyanosis with either comfortable tachypnea or without respiratory distress is more likely due to congenital heart disease (CHD) and warrants an echocardiogram and consultation with neonatology and pediatric cardiology.

**Evaluation for Neonatal Respiratory Failure and Cyanosis**

- **Pulse oximetry**: Preductal (right hand) and postductal (lower extremity) saturation readings by pulse oximetry can help determine the etiology of hypoxemia in a neonate. Equivalent preductal and postductal saturations in a cyanotic neonate suggest that either the ductus arteriosus is closed or the ductus is patent with subsystemic PVR. Hypoxemia in this setting may be caused by parenchymal lung disease with intrapulmonary shunting, or cyanotic CHD with ductal-dependent pulmonary blood flow. Ductal-dependent systemic blood flow lesions (eg, hypoplastic left heart syndrome, critical aortic stenosis, interrupted aortic arch, and coarctation of the aorta) can present with postductal desaturation. Anatomic pulmonary vascular disease (eg, pulmonary venous stenosis, and total or partial anomalous pulmonary venous return with obstruction) can cause suprasystemic PVR with right-to-left shunting across the patent ductus arteriosus and resulting postductal desaturation.
- **Chest radiograph**: It is important to note whether the severity of hypoxemia is out of proportion to radiographic findings. Marked hypoxemia despite provision of supplemental oxygen, when coupled with the absence of parenchymal lung disease on radiograph, is concerning for an extrapulmonary right-to-left shunt. The presence of parenchymal lung disease, pneumothorax, or a cystic mass (congenital pulmonary airway malformation) can direct management accordingly.

**TABLE 1. Normal resting vital signs in neonates.**

<table>
<thead>
<tr>
<th>Normal Resting Vital Signs in Neotones Aged &lt; 28 d</th>
</tr>
</thead>
</table>
| Heart rate (beats/min) | 100-160  
| Respiratory rate (breaths/min) | 35-55  
| Blood pressure (mm Hg) | Systolic range: 65-85  
| | Diastolic range: 45-55  
| Temperature (°C) | 36-37.9  

Sinus bradycardia with a heart rate of 80 to 100 beats/min during sleep is normal in a full-term healthy infant.
• Arterial blood gas: Evaluate gas exchange and pH (see section on inborn errors of metabolism [IEMs] regarding the presence of significant metabolic acidosis with respiratory compensation).
• Complete blood count with differential: Evaluate for signs of infection. Respiratory distress is one of the most common presenting signs of neonatal sepsis (see section on neonatal sepsis).5
• Blood pressure measurements: Obtain blood pressure measurements in the right arm and lower extremity to evaluate for aortic obstruction (interrupted aortic arch or coarctation).
• Response to respiratory support: If there is marked improvement in saturation of arterial blood with supplemental oxygen, this suggests intrapulmonary shunt or ventilation/perfusion mismatch due to lung disease.5 The response to mask continuous positive airway pressure also helps discriminate between severe lung disease and other causes of hypoxemia. If the preductal saturation of arterial blood never improves despite high inspired oxygen and mechanical ventilation, that raises the likelihood of cyanotic CHD.
• Echocardiogram and electrocardiogram: Obtaining an echo is important to rule out structural heart disease causing hypoxemia, findings which might impact the decision to transport and the need to start prostaglandin. Recall that at birth, the right ventricle (RV) is normally larger and thicker than the left ventricle, because the RV must pump blood through the relatively high-resistance pulmonary circulation in utero.5 Increased physiologic demands upon the fetal RV produce hypertrophy in neonates, including marked rightward axis, dominant R wave in V1, and T-wave inversions in V1-V3.7 Compared with adults and older children, conduction intervals in neonates will be shorter due to smaller cardiac size, heart rates will be much faster (>100 beats/min), and sinus arrhythmia may be normal.7

Clinical Pearl: Infants with severe coarctation of the aorta or aortic arch hypoplasia who escape detection in the newborn nursery can present in extremis with poor perfusion and absent femoral pulses after closure of the ductus arteriosus. This presentation warrants initiation of prostaglandin E, echocardiography, and transfer to a quaternary care facility with neonatology, pediatric cardiology, and pediatric cardiovascular surgery capabilities.

APPRAOSH TO NEONATAL SEPSIS/MENINGITIS

Early-onset sepsis remains a major cause of neonatal morbidity and mortality, although sepsis-associated mortality has declined since 2001 due to the introduction of intrapartum antibiotic prophylaxis in pregnant women during labor and delivery.8 Early-onset sepsis is defined as onset of sepsis in the first 3 days of life and is generally due to vertical transmission of bacteria from mothers to infants during the intrapartum period.8 There is no evidence that chemoprophylaxis prevents late-onset disease.9,10 Late-onset sepsis generally occurs in infants 7 days of age or older and is attributed to the horizontal transmission of pathogens acquired postnatally.9 Late-onset sepsis is associated with a higher frequency of meningitis, osteomyelitis, and septic arthritis.9 Group B streptococcus (Streptococcus agalactiae) and Escherichia coli are bacterial pathogens which together are responsible for 70% of early-onset sepsis in the neonatal period.6 Listeria monocytogenes sepsis is less common than group B streptococcus or E coli sepsis but can cause invasive disease in the newborn period.11

Clinical Presentation of Neonatal Sepsis

Clinical signs of neonatal sepsis include temperature instability, respiratory distress (tachypnea, grunting, nasal flaring, subcostal and intercostal retractions), apnea (pauses in breathing lasting 20 seconds), cyanosis, jaundice, hepatomegaly, abdominal distension, feeding abnormalities, and neurologic abnormalities.5

Diagnostic Approach to Neonates With Suspected Sepsis

All symptomatic neonates should have a complete blood count with differential, blood culture, and chest radiograph.12 The criterion standard for detection of bacteremia in newborn infants with suspected sepsis is a positive blood culture.8 Ideally, 1 to 3 mL of blood should be obtained. Catheterized urine sample should be obtained in infants older than 3 days with concern for sepsis. Cerebrospinal fluid (CSF) via lumbar puncture (LP) should be obtained for analysis, although LP should be deferred in infants with clinical instability or uncorrected bleeding disorders (see Table 2).12
White blood cell count and neutrophil indices

Neutropenia is the best predictor of neonatal sepsis, whereas the immature to total (I/T) ratio has the best sensitivity of all the neutrophil indices (see Table 3).¹³ Unlike older children, neutrophilia does not correlate well with sepsis in neonates. The absolute neutrophil count (ANC), the absolute band count of immature neutrophils, and the I/T neutrophil ratio are all more useful than total leukocyte counts in the diagnosis of neonatal sepsis. In approximately two-thirds of infants with sepsis, the ANC is abnormal.¹³

Platelet Counts

Twenty-five to 30% of infants with sepsis exhibit thrombocytopenia at the time of diagnosis (see Table 2).¹⁴ This percentage increases as duration of illness increases. Infected infants have accelerated platelet destruction and possibly depressed bone marrow production. Severely affected infants may demonstrate disseminated intravascular coagulopathy. If there is evidence of bleeding, bruising, or petechiae on exam, a coagulation panel should be obtained.

Diagnosing Bacterial Meningitis in Neonates

Common presenting symptoms for bacterial meningitis in neonates include lethargy, feeding problems, instability of temperature regulation, vomiting, respiratory distress, and apnea. A bulging fontanelle may be seen (usually a late manifestation). Seizures can be observed due to central nervous system inflammation or metabolic abnormalities (hypoglycemia, hyponatremia).

Cerebrospinal Fluid Analysis

The criterion standard for diagnosis of meningitis in neonates is CSF analysis including white blood cell (WBC) count, glucose and protein levels, Gram stain, and culture. However, between 1 and 10% of infants with proven meningitis have normal results on CSF fluid analysis.¹⁵ Of note, 30% of infants with positive CSF cultures for bacterial pathogens have negative blood cultures.¹⁵,¹⁶

### TABLE 2. Normal laboratory reference values in newborns after 72 hours.

<table>
<thead>
<tr>
<th>Selected Laboratory Value in Neonates After 72 h of Age</th>
</tr>
</thead>
<tbody>
<tr>
<td>Serum WBC count</td>
</tr>
<tr>
<td>Serum platelet count</td>
</tr>
<tr>
<td>ANC: neutropenia</td>
</tr>
<tr>
<td>I/T ratio</td>
</tr>
<tr>
<td>CSF/plasma glucose ratio</td>
</tr>
<tr>
<td>CSF protein (95th percentile)</td>
</tr>
<tr>
<td>0-14 d</td>
</tr>
<tr>
<td>15-28 d</td>
</tr>
<tr>
<td>Hypoglycemia seizure threshold</td>
</tr>
<tr>
<td>Hypocalcemia seizure threshold</td>
</tr>
<tr>
<td>Total serum calcium</td>
</tr>
<tr>
<td>Ionized calcium</td>
</tr>
</tbody>
</table>

Abbreviations: ANC, absolute neutrophil count; CSF, cerebrospinal fluid; I/T, immature to total neutrophil count.


### TABLE 3. Predictive values of laboratory markers for sepsis in neonates.

<table>
<thead>
<tr>
<th>Laboratory Value</th>
<th>Sensitivity (%)</th>
<th>Specificity (%)</th>
<th>Predictive Value (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>ANC &lt;1750 cells/mm³</td>
<td>38-96</td>
<td>61-92</td>
<td>20-77 96-99</td>
</tr>
<tr>
<td>ANC &lt;10% (5580 cells/mm³ at term)</td>
<td>48</td>
<td>73</td>
<td>4 98</td>
</tr>
<tr>
<td>I/T ratio 0.2</td>
<td>90-100</td>
<td>30-78</td>
<td>11-51 98</td>
</tr>
<tr>
<td>I/T ratio 0.3</td>
<td>35</td>
<td>89</td>
<td>7 98</td>
</tr>
<tr>
<td>CRP &gt;1 mg/dL</td>
<td>70-93</td>
<td>78-94</td>
<td>27 100</td>
</tr>
</tbody>
</table>

Abbreviations: ANC, absolute neutrophil count; CRP, C reactive protein; I/T, immature to total neutrophil count.

Adapted from Gerdes.¹⁵
**Cerebrospinal Fluid White Blood Cell Count**

During the first week of life, CSF WBC count slowly decreases in full-term infants. There is no change in CSF WBC counts or protein content with gestational age, but there is a significant decrease with postnatal age. Mild pleocytosis (6-19 cells/mm³) can be seen in symptomatic infants younger than 28 days without central nervous system involvement.

**CSF Protein and Glucose**

Cerebrospinal fluid protein values decline over the first few months of life as the infant's blood-CSF barrier matures. A 2010 cross-sectional study conducted at the Children's Hospital of Philadelphia found that in 375 infants 56 days of age or younger who had a LP performed in the pediatric ED (excluding traumatic LP and bacterial or viral meningitis), the median CSF protein value was 58 mg/dL. Average CSF protein concentration decreases by 6.8% with each 1-week increase in age.

Clinical Pearl: Herpes simplex virus should be considered in the differential diagnosis of neonatal sepsis, particularly when infants deteriorate rapidly with associated hemodynamic instability, liver failure, or coagulopathy, even in the absence of characteristic skin vesicles.

**APPROACH TO NEONATAL SEIZURES**

**Clinical Presentation**

Paroxysmal activity that interrupts normal behavior of a newborn infant and is repetitive or stereotyped should raise clinical suspicion for neonatal seizures. Examples include repetitive buccolingual movements, orbital-ocular movements, and bicycling. Periodic alterations in heart rate, blood pressure, or oxygenation in association with repetitive movements are clues that seizures are occurring. Symmetric “back and forth” movements of a tremulous or jittery infant are generally not epileptic in origin. In tremor, the flexion and extension phases are equal, whereas in seizures, a rapid flexion phase is followed by a slower extensor movement. Benign sleep myoclonus can also be mistaken for seizures and is a diagnosis of exclusion. With tremors or sleep myoclonus, gentle flexion of the affected body part should suppress the movement, as will changing position or waking up the infant. Clonic seizures will persist and cannot be suppressed by restraint. Normal motor automatisms in neonates can include rhythmic sucking, chewing, or tongue protrusions that may be provoked or intensified by stimulation. Rowing or swimming movements that can be suppressed by restraint or repositioning are typically nonepileptic. Sudden arousal with transient increased random activity of the limbs is a normal neonatal startle reflex. Normal neonatal reflexes include rooting, sucking, grasp, and Moro reflexes, which are fully developed in healthy term newborns and typically begin to disappear at 2 months of age.

**Differential Diagnosis for Seizures in the Neonatal Period**

Neonatal seizures can be seen with meningitis, neonatal encephalopathy, hypoglycemia, and hypocalcemia. Hemorrhagic or ischemic cerebrovascular lesions can be associated with neonatal seizures. Central nervous system malformations can lower seizure thresholds. Inborn errors of metabolism are rare causes of neonatal seizures, but warrant consideration in the presence of intractable seizures, as seen in association with elevated lactate/pyruvate levels in the blood and CSF. Newborns with maternal history of prenatal substance abuse may be at increased risk for neonatal seizures. Progressive epileptic syndromes do not usually manifest during the first month of life.

Clinical Pearl: When treating seizures in neonates, glucose and other electrolytes should be corrected with rapid infusion before antiepileptic medications are administered. Serum magnesium concentrations should be measured, as hypomagnesemia may accompany hypocalcemia.

**APPROACH TO INBORN ERRORS OF METABOLISM IN THE NEONATAL PERIOD**

**Clinical Presentation**

Inborn errors of metabolism may present in the neonatal period as an acute or more indolent illness. In IEM, a gene mutation leads to an absent or defective gene product or enzyme with resulting accumulation of the enzyme's precursor (or its byproduct) or a shortage of a downstream product of the enzymatic reaction. These inborn errors cause derangement of gene expression and biochemical function. It is beyond the scope of this article to discuss the specific IEMs. However, some of these disorders may be symptomatic in the newborn period or early infancy, and thus present to the ED with progressive lethargy, poor suck, and neurologic deterioration. Typically, these patients will be the product of normal pregnancies and deliveries and become ill after 36 hours of life, when the offending metabolites accumulate.
will be detected by expanded state newborn screens, so it is critical to call the newborn screening follow-up hotline as soon as possible when there is clinical suspicion for IEM.

Triggers to distinguish between infants with sepsis and those who might have IEMs include the following: negative results of an infectious disease work up, normal white count, hypoglycemia, unexpectedly severe metabolic acidosis (with or without increased anion gap), lactic acidosis, respiratory alkalosis, or hyperammonemia. Other concerning laboratory findings for infants with IEMs can include the following: ketosis, positive urinary reducing substances, discolored urine, and bone marrow suppression (leukopenia, anemia, thrombocytopenia). Concerning physical examination findings for infants with IEMs can include hepatomegaly, splenomegaly, coarse facial features, macroglossia, dystonia, microcephaly or macrocephaly, bone or limb deformities/contractures, persistent diarrhea, and skin abnormalities. On ophthalmologic examination, infants may have a macular “cherry red spot,” retinitis pigmentosa, optic atrophy or hypoplasia, corneal clouding or opacities, cataracts, or a dislocated lens. Further workup may reveal cardiomegaly and ventricular hypertrophy, electrocardiographic abnormalities, stippled calcifications of the patellae, dysostosis multiplex, cerebellar atrophy or hypoplasia, and agenesis of the corpus callosum.

Clinical Pearl: An acutely ill child with an IEM is an emergency and requires rapid rescue treatment. If IEM is on the differential diagnosis, supportive care focused on cardiorespiratory and hemodynamic status, with administration of fluids, and evaluation of electrolytes is the mainstay of treatment. Transfer to a tertiary care center with experience in the care of these children should be performed as quickly as possible.

APPROACH TO FEEDING DIFFICULTIES IN THE NEONATE

Clinical Presentation

Neonates frequently demonstrate functional gastrointestinal symptoms. Gastroesophageal reflux refers to retrograde movement of gastric contents into the esophagus. Gastroesophageal reflux is a normal physiologic process that occurs multiple times a day in all healthy infants. Regurgitation refers to the passage of refluxed gastric contents into the mouth or pharynx. In contrast to regurgitation, vomiting is a central nervous system reflex that uses skeletal muscles. In order to diagnose regurgitation in a healthy infant older than 3 weeks (“happy spitters”), there should be regurgitation at least twice daily for at least 3 weeks in the absence of hematemesis, aspiration, apnea, failure to thrive, difficulty in feeding or swallowing, or abnormal posture. If there is accompanying vomiting, irritability/crying, arching, coughing fits, or failure to thrive, consider gastroesophageal reflux, cow’s milk protein intolerance, or anatomical anomalies. When dealing with feeding difficulties in neonates, it is helpful to evaluate feeding frequency, technique used in nursing or bottle-feeding, and the use of pacifiers. Parental education is key to the management of physiologic regurgitation (ie, overfeeding exacerbates regurgitation, upright positioning for 30 minutes after feeds can decrease regurgitation). In infants with physiologic regurgitation, thickened antiregurgitation formula can also decrease frequency and volume of regurgitation.

Clinical Pearl: Bilious vomiting in a neonate should be equated to malrotation with midgut volvulus until proven otherwise. This is a surgical emergency requiring upper gastrointestinal series to rule out malrotation/small bowel obstruction and transfer to a tertiary care facility with availability of pediatric surgeons.

SUMMARY

In 1896, Dr Thomas Rotch, former Chair of Pediatrics at Harvard, published a treatise on the medical treatment for children, declaring: “The mere knowledge that certain diseases exist, and the usual methods of diagnosticking them, prove to be very inadequate when we are brought face to face with a sick and fretful child, or with an infant who is unable to describe its symptoms...” As health care providers to neonates, we must endeavor to learn this alphabet, to allow us to provide the highest level of care to these uniquely vulnerable patients.

REFERENCES

The number of preterm births is rising. Just under 10% of infants born in the United States will be born premature. A total of 1.4% of all births will be very low birth weight (VLBW) infants (<1500 g), and less than 1% of all births will be extremely low birth weight or less than 1000 g. Premature infants are at risk for multiple comorbidities related to their prematurity, and these comorbidities may affect every organ system. Term infants with congenital anomalies or a difficult hospital course may similarly be discharged with complex medical issues. The goal of this article is to highlight some of the common complications affecting infants who are discharged from the neonatal intensive care unit, especially issues that may arise in the first year of life and may prompt parents to seek care in the emergency department (ED) or urgent care setting.

**NUTRITION AND GROWTH**

Growth in the neonatal intensive care unit (NICU) and in the postdischarge period is critical for determining long-term outcomes in premature infants. Catch-up growth in all areas is particularly important for former preterm infants, but poor growth in head circumference after discharge is associated with poor long-term outcomes. Growth of a former preterm infant should be monitored closely and prematurity adjusted for until...
the infant reaches 24 months. It is important to use the appropriate preterm, sex-specific growth curves on former preterm infants.

Infants with a prolonged NICU course often have delayed feeding and increased energy needs, and may even develop oral aversion. These infants may be especially challenging for parents to feed in the initial postdischarge period. Typically, VLBW infants are discharged home on a transitional 22-kcal/oz formula or human milk fortified to 22 kcal/oz. Increasing energy densities, up to 30 kcal/oz, may be required for a select group of infants. Parents may have difficulty understanding how to mix the formula, or parents with low incomes may try to make the amount of formula “stretch” by diluting it down with extra water. Infants with improperly mixed formula are at risk not only for poor growth but also for electrolyte derangements. A formerly preterm infant who presents to the ED with weight loss or poor feeding should be evaluated for electrolyte abnormalities, and the physician should have a discussion with parents on how the formula was prepared. Access to formula should be assessed whenever an infant is evaluated for poor growth. Many specialty formulas are expensive and cannot be obtained without ordering them ahead of time at grocery and drug stores.

MEDICAL NEGLECT OR ABUSE

Children with complex medical needs can often present a challenge for families, especially families with limited income and resources. Failure to thrive may be a sign of an overwhelmed parent, or even medical neglect or abuse. Studies have shown that children in the birth-to-3 years age group have the highest rates of victimization, with children less than 1 year of age accounting for the highest percentage of victims (9.6%). One study showed that infants born at low birth weight had an adjusted relative risk of 2 for neglect or abuse compared with infants born at a normal birth weight. Physicians who are treating former preterm infants should be aware of this risk when these children present with failure to thrive or other signs suspicious for medical neglect or abuse.

GASTROSTOMY TUBES

Infants born with congenital anomalies or with a history of prolonged hospitalization will occasionally require partial or full nutritional support with the assistance of a gastrostomy tube (G-tubes) (Figure 1). There are multiple reasons for the use of G-tubes in premature infants, and defining the underlying etiology may help the medical team provide appropriate treatment. Premature infants with complicated courses may exhibit oral aversion and will take time to develop proper oral feeding skills. Infants with congenital anomalies of the head and neck (eg. micrognathia) may be unable to feed orally at the time of discharge. Complex genetic syndromes or neurologic syndromes may also leave infants unable to feed orally.

Gastrostomy tubes, although important for nutrition and growth, may provide challenges to the family in terms of management. Infants may dislodge the G-tubes themselves, or a parent or sibling may accidentally dislodge the tube and the family may present to the ED for assistance. Gastrostomy tubes may also need to be replaced because of cracks in the tubing or a defective balloon. It is important to know that simple G-tubes take at least 1 to 2 weeks to form a mature tract. If a fresh G-tube is dislodged, the tract may begin to narrow or even close in a matter of hours. Replacement of a G-tube into a mature tract should be completed as soon as possible. If the tract is less than 8 weeks old, the service that initially placed the tube should be contacted, and correct placement should be confirmed before feeding. In the meantime, a Foley catheter or the defective G-tube may be placed into the tract to ensure that it does not narrow further.

Two common G-tube related complaints presenting to the ED are granuloma and G-tube leaking. Granulomas are fleshy out-pouchings that are often mucosa-like in appearance (Figure 2). Most granulomas are not infected; they represent the result of local irritation by the plastic in the gastrostomy tube, which can cause bleeding or even irritability in the infant. A conservative approach is to apply a small amount of triamcinolone ointment or cream.
to the area 1 to 2 times per day for 14 days. The skin around the abdomen should be spared, and a cotton tip swab is often the most accurate way to apply the treatment.

Common illnesses such as upper respiratory tract infections and gastroenteritis may cause G-tubes to leak due to an overall increase in secretion volume. Parents often feel that they are leaking “most” of the infused formula. Typically, this type of leaking gets better as the acute illness ends. However, it is often prudent to check the balloon water fill amount to make sure that the correct amount is still in place. Gastrostomy tube feeds can be lessened in volume and increased in frequency to help minimize leaking during acute illness.

NECROTIZING ENTEROCOLITIS AND SHORT BOWEL SYNDROME

Necrotizing enterocolitis (NEC) typically presents with abdominal distention, poor feeding, and emesis. It may progress to blood in the stool, bowel perforation, intestinal necrosis, and shock. The hallmark findings of NEC on abdominal radiograph is pneumatosis intestinalis and intrahepatic portal venous gas. Infants are unlikely to present to the ED with fulminant NEC, but the long-term complications of NEC are important to understand when undertaking both acute and long-term care of former NICU graduates.

Necrotizing enterocolitis is a disease that primarily affects premature infants, although 10% of cases occur in term infants. Necrotizing enterocolitis in either age group carries a potential for high morbidity and mortality in the neonatal period. The incidence of NEC increases with decreasing gestational age, affecting about 7 to 13% of infants born at 22 to 28 weeks’ gestation. Infants with NEC can often be treated medically, but others will progress to surgical NEC, which can lead to resection of large amounts of bowel and short bowel syndrome (SBS). The mortality rate for NEC is high, between 20 and 30%, with the majority of mortality occurring in surgical NEC. All infants with NEC are at increased risk for neurodevelopmental delay compared with age-matched counterparts, in addition to increased length and cost of hospitalization.

Necrotizing enterocolitis is the most common cause of SBS in infants. In one study, the readmission rate of premature infants with SBS was 79% in the first 2 years of life. The majority of these hospitalizations are due to infection, but other complications including dumping syndrome and mechanical problems with central lines or gastrostomy tubes may also cause parents to seek care. Infection can occur from diminished mucosal barrier function leading to bacterial overgrowth and translocation or from contamination of a central line. These children will likely present to the ED, and the emergency physician should have a high suspicion for sepsis in any infant with a history of SBS, especially when the infant has a central line in place. A blood culture should be obtained for every infant with a central line and fever even when another potential source has been identified. Another common complication of SBS is “dumping syndrome,” where large amounts of watery stool are generated. Malabsorption will occur, and these infants will be at risk for dehydration, poor nutrition, and electrolyte derangements.

Former NICU graduates who present with obstructive symptoms should be evaluated for a history of NEC because NEC can lead to stricture formation for several months to years after NICU discharge. Medical or surgical NEC imparts the same risk of stricture development. Although multiple strictures can occur, the most likely location for a post-NEC stricture is the colon. A pediatric surgical consult is appropriate if history, physical examination, and imaging are consistent with NEC-related obstruction.

BRONCHOPULMONARY DYSPLASIA

Bronchopulmonary dysplasia (BPD) is a chronic lung disease affecting up to 25% of VLBW infants and is the most common severe complication of infants born prematurely (Figure 3). Although advancements such as antenatal steroids and exogenous surfactant administration have improved our ability to treat neonatal respiratory distress syndrome, the incidence of BPD over the recent years in the VLBW
Infants born at less than 32 weeks’ gestation will have an interruption in the normal development of the saccular stage of lung development, causing decreased septation and alveolar hypoplasia. Volutrauma from mechanical ventilation can then cause irreversible lung injury. A diagnosis of BPD is defined as treatment with oxygen greater than 21% for at least 28 days. Bronchopulmonary dysplasia is further classified into mild, moderate, or severe depending on the infant’s degree of oxygen requirement and mode of oxygen delivery after 28 days.

Chronically, infants with BPD will have a higher risk of neurodevelopmental delay as well as adverse complications from pulmonary vascular remodeling such as pulmonary hypertension, and higher rehospitalization rates. There is evidence that early lung injury persists into late childhood, as a study found decreased pulmonary function scores in 9 to 15-year-old patients with BPD compared with former preterm infants without BPD.12

There are a multitude of reasons why infants with BPD present to the ED. Infants with BPD may present because of their need for increased nutritional and energy requirements. Those who are discharged with or without oxygen may go home on a chronic diuretic (such as a thiazide or spironolactone) and have electrolyte abnormalities secondary to their diuretic use. Diuretics may also have been recently changed or weaned off, resulting in a slow but progressive worsening of the infant’s respiratory status. Preterm infants may also present to the ED with exacerbations of their BPD, complications from equipment malfunction at home, or viral respiratory infection, which may impair their already fragile status.

Viral illness in the face of BPD, regardless of the need for supplemental oxygen, often triggers hypoxia and wheezing. Respiratory viral panels may assist the clinician in diagnosing the primary reason for worsening respiratory status. Although bronchodilators are controversial in term infants with bronchiolitis, they are more acceptable in the early care of preterm infants with BPD especially when respiratory status is worsening. Infants in crisis may require increases in their diuretics and a burst of inhaled, oral, or intravenous steroids. Inhaled bronchodilators can be useful during exacerbations but do not improve long-term function.12,13

All ill-appearing preterm infants should be assessed for the degree of increased work of breathing as well as for hypoxia. Former preterm infants can present with “head bobbing” as a sign of respiratory distress. Blood gas and metabolic laboratory values should be assessed if the preterm infant’s condition or support is significantly above baseline. Treatment of preterm infants with a respiratory viral syndrome is mainly supportive. They may need increased FiO₂ or higher flow if on home oxygen. Increases of more than 0.5 L of flow above baseline home oxygen use are considered significant, and it is difficult to ensure safety of these patients without observation. Overnight observation can help elucidate if preterm infants with BPD need small increases in oxygen flow and supportive care only or whether they might need either noninvasive or invasive positive pressure interventions.

TRACHEOSTOMY

Infants with moderate or severe BPD may need to be discharged on home oxygen. In rare cases, about 2% of preterm infants14 will require a tracheostomy and a home ventilator (Figure 4). Infants with a tracheostomy are at high risk for readmission and death.15 When infants with tracheostomy present to the ED in distress, they should be appropriately triaged so that they receive immediate attention and receive the highest level of care.

Infants with tracheostomy are classified as either obligate or nonobligate tracheostomy users. Those that are nonobligate can breathe through their upper airway. Even those infants on a ventilator may be either obligate or nonobligate tracheostomy users.
An accidental decannulation of a tracheostomy tube should be treated seriously, and rapid attempts to reestablish an airway should be undertaken. Urgent consultation with otolaryngology can be critical to reestablishing the airway on a patient with an obligate tracheostomy. The obturator should be placed in either the old tracheostomy tube or a replacement tube, and then an attempt should be made to reinsert the tracheostomy tube. If the initial attempt, repositioning, and a 1-size-smaller tube all fail, a 10F to 12F suction catheter could be used to provide a “guide” for tube insertion and possibly as a means for oxygenation. If all attempts fail, the stoma site can be occluded and ventilation can be attempted from the upper airway.

Tracheostomy tubes can plug and may require suctioning to alleviate the associated respiratory distress. When suctioning, it is important to preoxygenate the infant, limit the number of attempts, know how deep to suction, and limit the total time suctioning. Suctioning a tracheostomy tube is not without risk, and acute complications include bleeding, hypoxia, bronchospasm, blood pressure instability, pneumothorax, perforation, increased intracranial pressure, and cardiac arrest.

Bleeding, stoma site breakdown, infection, and airway trauma are also possible complications of a tracheostomy tube. When an infant with a tracheostomy tube arrives in the ED, it is important to evaluate the patency of the tube and stoma site even if there are other etiologies for the visit.

Intraventricular hemorrhage (IVH) is another complication that increases with decreasing gestational age and can place infants at risk for seizures and adverse neurodevelopmental outcome. Bleeding is caused by immaturity of the capillaries in the subependymal germinal matrix, which rupture and bleed into the germinal matrix and ventricles, and sometimes may extend into the parenchyma. Intraventricular hemorrhage is graded on the basis of severity, with grade 1 being limited to the germinal matrix itself, grade 2 extending into the ventricles, grade 3 extending into the ventricles with associated dilation, and grade 4 extending into the ventricles with associated dilation and also involving the surrounding parenchyma (Figure 5). Grades 3 and 4 are considered severe. The overall incidence of IVH ranges from 7 to 23% of preterm deliveries.

One of the complications of severe IVH, besides developmental delay and seizures, is posthemorrhagic hydrocephalus (PHH). If PHH is left untreated, it can lead to increased intracranial pressure. The treatment for PHH is serial ventricular punctures to remove cerebrospinal fluid, and ultimately, the infant may require a ventriculoperitoneal (VP) shunt. Approximately one third of extremely low birth weight infants with IVH will develop PHH, and 15% of those infants will require a VP shunt.

Figure 5. A head ultrasound of a neonate with IVH.
Infants presenting to the ED with a history of VP shunt are at greater risk for both infection and shunt malfunction. Presenting signs and symptoms may include vomiting, poor feeding, lethargy, irritability, fever, sunsetting, or bulging fontanelle. Affected infants may also present with seizure (first time or increased frequency). These infants need emergent head imaging, and perhaps electroencephalography, and immediate consultation with neurosurgery, and perhaps neurology, as well as contact with their medical home.

**Gastroesophageal Reflux Disease**

Gastroesophageal reflux (GER) is defined as the involuntary passage of stomach contents into the esophagus and is a normal process in children, especially newborn infants. Regurgitation or spitting up is reported to occur daily in 50% of all infants. Gastroesophageal reflux disease occurs when the patient begins to have adverse symptoms from the GER, and the diagnosis is most often made clinically. Preterm infants are at especially high risk for GERD. Neurologic impairment, esophageal atresia, BPD, and chronic respiratory disorders are also independent risk factors for GERD.

Symptoms of GERD (including emesis, discomfort with feeding, respiratory issues including apnea and desaturation, sleep disturbance, and irritability) can be difficult to distinguish from cow’s milk protein allergy, or the symptoms may coexist with or complicate the GERD. Therefore, diet and other feeding modifications should be the first line of treatment before medications are introduced. Infants may respond to increased feeding frequency, with decreased volume to avoid overfeeding. Keeping the infant with her head elevated above her stomach following a feed or feeding with prone or side-lying positioning (while awake only to avoid an increased risk for sudden infant death syndrome) may improve symptoms as well. In formula-fed infants, changing to a protein hydrolysate formula may improve symptoms and help distinguish GERD from cow’s milk protein allergy. In breastfed infants, restricting the mother’s diet from cow’s milk and eggs may be trialed. Thickening feedings using up to 1 tablespoon of dry rice cereal per ounce of formula or breast milk may improve symptoms, although physicians should be aware of an association between NEC and thickened feedings in preterm infants.

Historically, medical therapy for GERD in infants includes the use of proton pump inhibitors, histamine receptor antagonists, and prokinetic agents. These medications should be reserved for those infants who are symptomatic from their reflux and who fail lifestyle modification, and should be added by the infant’s medical home provider. If the infant is not responding to the medication, therapy should be discontinued. Surgical therapies, including Nissen fundoplication and jejunal feedings, should be reserved for the most extreme cases only.

**Communication**

The ED is an important bridge to hospital-based care, emerging problems requiring additional subspecialty care, routine problems requiring follow-up, or normal newborn presentations perceived as problems. Beyond the important role that ED physicians play in diagnosis and recovery, they can also facilitate communication between parents, subspecialists, and primary care physicians that enhances the long-term prognosis for the NICU graduate. A phone call, page, or e-mail outlining the visit for the primary care physician is an important way to move a complex NICU graduate along a positive trajectory.

**Summary**

Pediatricians and emergency medicine physicians should be aware of the potential issues affecting high-risk newborns discharged from the neonatal intensive care unit. These infants are at risk of being both medically complex and medically fragile, and families may be struggling to meet their complex needs. Emergency care providers play an essential role in the management of these infants. When working with these infants and families, careful attention to each organ system is warranted. Following discharge from the ED, communication regarding the infant’s emergency care and treatment plan with the primary care provider is essential to ensure that the infant remains closely followed. As the complexity of care increases, ED personnel will likely become experts in the pathophysiology of preterm diseases, the management of acute illness, and the dependence of families on many interlocking systems that improve health.

The authors report no conflict of interest.

**References**


Respiratory Distress in the Newborn: An Approach for the Emergency Care Provider

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Timing is critical in the assessment of the newborn infant with respiratory distress. It is important to act quickly because of the potential for rapid deterioration. However, knowledge about the newborn’s historical timing including the gestational age (GA) at the time of birth, the actual age of the infant at the time of presentation, and length of time symptoms have been present is also vital. These crucial pieces of information along with the maternal, birth and recent history, and a thorough physical examination will be key in narrowing down the broad differential diagnosis of neonatal respiratory distress.

ADAPTATIONS FROM FETAL LIFE TO NEWBORN

Full-term infants are born within the GA range of 39 weeks 0 days to 41 weeks 6 days; preterm infants, prior to 37 weeks; early term, 37 weeks 0 days to 38 weeks 6 days; late term infants, 41 weeks 0 days to 41 weeks 6 days; and postterm infants, after 42 weeks GA.¹ As the neonate transitions from intrauterine to extrauterine life, several physiologic changes have to occur which depend on the GA and actual age. For the newborn to appropriately ventilate and oxygenate independently after birth, the lungs must have sufficient development and have started to...
produce pulmonary surfactant, a lubricant-like layer that lowers the surface tension and reduces atelectasis. Both the degree of lung development and amount of surfactant production are dependent on the GA.  

At birth, newborns transition from fetal circulation to neonatal circulation in which the placenta is replaced by the lungs for oxygenation. Fetal shunts such as the ductus arteriosus that allowed blood to mostly bypass the lungs in utero close in the first hours to weeks of life. In addition, the elevated pulmonary vascular resistance (PVR) necessary in utero must decrease after birth to allow for pulmonary blood flow and adequate gas exchange to take place. Clamping of the umbilical cord removes the low resistance system of the placenta and increases systemic vascular resistance. With the newborn's first breath, lung volume increases and PVR decreases. The ductus arteriosus starts to close in response to the changes in pulmonary and arterial vascular resistance, removal of placentaletly derived prostaglandins, and increased arterial oxygen content. Initial closure occurs at 10 to 15 hours of life, with final closure at 2 to 3 weeks of life in term infants. Respiratory distress can be caused by maladaptation to neonatal physiology but can also develop after normal transitioning in those infants who are reliant on the persistence of fetal circulation, like some congenital heart diseases.

NORMAL NEWBORN PARAMETERS

Important vital signs to assess in newborns include heart rate (HR), respiratory rate (RR), preductal and postductal oxygen saturations, 4 extremity blood pressures, and unclothed weight. The range for normal HR at term is 110 to 160 beats/min, but this can decrease to 85 beats/min during sleep. Normal RR is 40 to 60 breaths/min with tachypnea defined as sustained RR > 60 breaths/min. The mean blood pressure in neonates 1 to 3 days old is 64/41 (mean arterial pressure [MAP] 50 mm Hg), which increases to 95/58 (MAP 72 mm Hg) for children 1 month to 2 years of age. Normal oxygen saturations immediately after birth are close to 65% but by 10 minutes of life and thereafter, they should remain greater than 90%. Weight gain is an excellent indicator of newborn health, and lack of expected growth can be an indication of an underlying problem. Newborns typically lose up to 10% of birth weight over the first several days of life and then regain birth weight by roughly 2 weeks of life. Weight should be obtained and plotted on an infant sex and GA-specific growth chart.

PERTINENT HISTORY AND EXAMINATION FINDINGS

A thorough maternal and birth history can be invaluable in directing the initial work up and narrowing the differential diagnosis. A template that can be used to obtain and communicate this information follows:

The infant is an _ (AGA/SGA/LGA) _ (number of days) _ old _ (gender) _ born at _ (gestational age in weeks) _ via _ (mode of delivery) _ under _ (type of anesthesia) _ to a _ (age of mother) _ (gravida/parity status) _ complicated by _ (any fetal, labor, or maternal history prior to delivery including medications/drugs) _ with _ (appropriate/inappropriate) _ weight gain.

With any newborn in respiratory distress, a full examination may be deferred until the infant is stabilized. Once stable, the newborn requires a head-to-toe examination to help rule in or out diagnoses on your differential. The general examination should note vitals, degree of distress, dysmorphic features, or cyanosis. Head and neck examination should assess the palate, nose, and oropharynx for patency or masses as well as masses along the neck. Observe the chest for asymmetry or barrel shape. The respiratory examination should note symmetry and quality of breath sounds, abnormal sounds (eg, stridor and wheezing - including which phase[s] of respiration these sounds are heard), retractions, grunting, nasal flaring, and head bobbing. The cardiac examination should include heart sounds, murmurs, central and peripheral perfusion, and brachial and femoral pulses. The abdomen should be assessed for scaphoid
appearance, bowel sounds, organomegaly or tenderness. Assess extremities for abnormalities in the limbs or digits and skin for color, rashes, or lesions. Finally, the neurologic examination should assess level of alertness, tone, and reflexes including suck.  

### STABILIZATION AND ASSESSMENT

All newborns with respiratory distress should be placed on continuous cardiorespiratory monitoring with pulse oximetry. Addressing inadequate ventilation or airway obstruction is essential in preventing further decompensation. Inadequate ventilation is often due to atelectasis and requires effective pressure, particularly positive end-expiratory pressure to re-recruit lung volume. Required respiratory support varies from nasal cannula, nasal or mask continuous positive airway pressure (CPAP), positive pressure ventilation via mask or endotracheal tube, or laryngeal mask airway placement. Blow-by oxygen or non-rebreather masks are not appropriate modes of respiratory support in the newborn. In cases of upper airway obstruction, oral airways, nasal trumpets, or prone position may be needed to stabilize the airway. If intubation is necessary, a tertiary care neonatal intensive care unit (NICU) or pediatric intensive care unit should be contacted and can assist in mode of ventilation and further management. In addition, newborns with respiratory distress should have vascular access obtained, be nil per os and started on maintenance intravenous fluids containing dextrose, with a nasogastric tube placed to decompress the stomach. A chest radiograph (CXR) and preferably arterial blood gas should be performed. Oxygen should be provided as needed to achieve normal oxygen saturations.

### ETIOLOGIES OF RESPIRATORY DISTRESS

**Transient Tachypnea of the Newborn**

Transient tachypnea of the newborn (TTN) is a period of self-limited tachypnea in term or near-term infants. The onset is typically within the first 6 hours of life and most symptoms resolve by days of life 2 to 5. Transient tachypnea of the newborn, also called “wet lung,” is due to a delay in fetal lung fluid resorption and subsequent decreased lung compliance. Fluid resorption is hormonally mediated by activation of sodium channels. Increased risk for TTN is seen in cesarean delivery without labor, diabetic mothers, male sex, and earlier gestation. Physical examination reveals tachypnea, grunting, and nasal flaring. Initial CXR often shows fluid in the pleural fissures, streaky infiltrates, and increased perihilar markings (Figure 1). Blood gas may be normal or show respiratory acidosis with hypoxemia. Treatment is supportive with either nasal cannula or CPAP, and intubation is rarely needed. Because TTN presents similarly to other causes of neonatal respiratory distress, it is often considered a diagnosis of exclusion. It is appropriate to evaluate and initiate therapy for infectious or other etiologies until more information is known and clinical course becomes more apparent.

**Respiratory Distress Syndrome**

Respiratory distress syndrome (RDS) is a disorder of surfactant deficiency seen in preterm infants, although term infants with diseases that affect surfactant production or function can also be
affected. Symptoms begin shortly after birth and worsen over the first few hours of life. The incidence of RDS is indirectly related to GA. As mentioned, because surfactant lowers the surface tension in the lung, a deficiency can lead to increased atelectasis and impaired ventilation and oxygenation. On examination, infants will have tachypnea, retractions, and nasal flaring, in addition to grunting which functions as an auto-positive end-expiratory pressure mechanism to increase intra-airway pressure and prevent alveolar collapse. Chest radiograph demonstrates diffuse granular or “ground glass” infiltrates (Figure 2). Treatment involves providing appropriate respiratory support which can range from CPAP to intubation depending on work of breathing, degree of respiratory acidosis, and fraction of inspired oxygen (FiO\textsubscript{2}) requirement. Once admitted to an intensive care unit, physicians may consider administering endotracheal surfactant for FiO\textsubscript{2} > 30%-40%. Patients with suspected RDS should be evaluated for infection with blood culture and complete blood count. Empiric antibiotics may be appropriate given newborn and maternal history. Because RDS may require prolonged respiratory support, goals of support are to prevent barotrauma and unnecessary FiO\textsubscript{2} exposure.

Pneumonia and Other Infections

Pneumonia or other infections including bacteremia, urinary tract infection, and meningitis can all present with respiratory distress. Symptoms present depending on timing of infection but can develop within the first few hours of life. On examination, the infant may have respiratory distress and/or other nonspecific symptoms such as poor feeding, somnolence, temperature instability, and hypoglycemia. Chest radiograph findings for neonatal pneumonia can be quite variable and difficult to diagnose radiographically but most commonly have bilateral alveolar densities with air bronchograms (Figure 3). However, CXR findings might also appear like that of TTN or RDS or have pneumothorax or pleural effusion. A complete blood count with differential and blood culture should be sent on all patients with signs or symptoms concerning for infection. Depending on the infant’s history, associated symptoms, and examination, urine, cerebrospinal fluid, and tracheal cultures should be considered, as well as respiratory viral panel or other viral testing, such as herpes simplex virus. Empiric antibiotics (eg, ampicillin and gentamicin) that treat the common neonatal pathogens should be started in all newborns with suspected infection. Infection can present similarly to TTN but the symptoms will worsen or persist for a longer period, especially without treatment.

Meconium Aspiration Syndrome

Meconium aspiration syndrome (MAS) is a condition of the term or postterm infant involving the aspiration of meconium into the lungs which can result in mechanical obstruction, chemical pneumonia, surfactant inactivation, and persistent pulmonary hypertension. Meconium is fetal stool which is typically passed after birth but can be passed in utero during times of fetal stress or hypoxia. The frequency of meconium-stained amniotic fluid occurs in 10 to 15% of deliveries. Infants born through meconium-stained amniotic fluid can be asymptomatic, but about 7% will have neonatal respiratory distress. As of 2015, routine intubation and tracheal suctioning is no longer necessary.
recommended for nonvigorous infants born through meconium-stained fluid. Infants will present with respiratory distress of variable severity but can progress to respiratory failure. They will often have meconium-stained skin or umbilical cord. Chest radiograph can show diffuse patchy infiltrates with areas of overinflation related to air trapping and atelectasis (Figure 4). Arterial blood gases should be obtained to determine the degree of hypoxemia. Management should focus on respiratory support to improve ventilation and oxygenation. Infants should be monitored for signs of air trapping or pneumothorax and worsening hypoxemia or persistent pulmonary hypertension. Administration of surfactant should be considered as well as the need for extracorporeal membrane oxygenation (ECMO). Consultation with neonatologist is recommended.

### Pneumothorax
Pneumothorax is a leakage of air into the pleural cavity that can cause collapse of the lung. It usually develops secondary to an underlying disease process but can occur spontaneously in 1% of newborns, although only about 10% of these are symptomatic. The cause can be spontaneous from increased intrathoracic pressure that occurs in the first few minutes of life during transitioning, or it can result from resuscitation efforts or an underlying disease process like RDS, MAS, pneumonia, or any congenital malformation of the lung. Symptoms vary in severity, but pneumothorax should be considered in any infant with an abrupt worsening of respiratory status. Typically, infants will have tachypnea, grunting, flaring, retractions, and diagnostically, asymmetric breath sounds. Diagnosis may be emergently made by trans-illumination of the thorax or clinical examination, but CXR is preferred (Figure 5). Interventions are based on clinical significance with mild respiratory distress treated with supportive care and monitoring. Severely affected newborns, especially those with tension physiology including severe hypoxia, distress, or cardiac instability, may require immediate needle decompression and/or chest tube placement. Consultation should be made with a neonatologist and/or pediatric surgeon (Figure 5.)

### Congenital Diaphragmatic Hernia
Congenital diaphragmatic hernia (CDH) occurs when a portion of the diaphragm is absent and the contents from the abdominal cavity herniate through the defect into the thorax. Up to 95% of diaphragmatic hernias are posterolateral (Bochdalek) and are usually seen on the left. Other locations for CDH include the anterior diaphragm (Morgagni) or at the central tendon location. These defects are not always diagnosed in utero for a variety of reasons including the timing of herniation and possible fluidity of the herniated contents, as well as the quality of the study and presence of other associated anomalies. In addition, the fetal timing of herniation and amount of abdominal contents present in the thorax during critical points in development can severely affect the pulmonary tissue and vasculature leading to both ipsilateral and contralateral pulmonary hypoplasia and pulmonary hypertension. Although most infants present with respiratory distress soon after birth, 5 to 25% can present later in life. Isolated CDH can have a survival rate of up to 80%, but with associated anomalies, survival can be as
low as 10%. On examination, infants can have tachypnea and other signs of respiratory distress, asymmetric breath sounds, displaced heart sounds, scaphoid abdomen, and auscultation of bowel sounds in thoracic cavity. Chest radiograph will show loops of bowel in the thorax, often with displacement of the cardiac silhouette (Figure 6). In the acute neonatal period, immediate intubation and gastric decompression followed by gentle mechanical ventilation with goal of minimizing barotrauma are recommended. These infants should be transferred as soon as possible to a tertiary care NICU where pediatric surgery and extracorporeal membrane oxygenation (ECMO) are available. In addition to respiratory support, the patient may need treatment of pulmonary hypertension including the use of inhaled nitric oxide and inotropic support for hemodynamic instability.

For infants with late presentation, they may have intermittent or chronic respiratory symptoms and/or may present with gastrointestinal symptoms like abdominal pain or vomiting. They also may present with acute respiratory distress in the setting of superimposed infection. Chest radiograph findings may mimic pneumonia or may be nonspecific necessitating a chest computed tomography for diagnosis. Prompt referral to a pediatric surgery center is recommended.

**Persistent Pulmonary Hypertension**

Persistent pulmonary hypertension is a disease in which the newborn’s PVR does not appropriately decrease after birth and remains abnormally elevated resulting in impaired pulmonary blood flow and gas exchange. This can be due to an abnormality in vasculature constriction due to parenchymal disease (eg, pneumonia and MAS), vasculature remodeling (eg, idiopathic), or vasculature development (eg, CDH). Persistent pulmonary hypertension can present within the first few hours of life or later as a sequela to another lung disease. Infants affected by persistent pulmonary hypertension present with tachypnea and cyanosis, due to right-to-left shunting across the ductus. Chest radiograph may show variable findings depending on etiology. Echocardiogram will reveal elevated pulmonary artery pressures and right-to-left shunts. Because the goal of therapy is to decrease PVR, treatment should be aimed at promoting pulmonary vasodilation including maintaining appropriate lung volumes, oxygenation, and ventilation as well as maintenance of normal blood pressure, volume status, hemoglobin levels, and electrolytes. Mechanical ventilation is often needed to improve acidosis and oxygenation.

The use of inhaled nitric oxide should be considered as it has been shown to decrease the need for ECMO. Patients may need inotropic support and those with respiratory failure and severe hypoxemia may progress to ECMO. Urgent referral to a tertiary care facility with ECMO capability is recommended.

**Congenital Heart Disease**

Infants with congenital heart disease, in particular ductal-dependent heart lesions like hypoplastic left heart syndrome or coarctation of the aorta, can present with respiratory distress. As mentioned, the ductus arteriosus has a functional and permanent closure. In those infants reliant on the ductus for systemic perfusion, they may develop respiratory distress progressing to cardiogenic shock as this shunt closes. On examination, infants with congenital heart disease may have respiratory distress, cyanosis, decreased systemic perfusion or abnormal pulses, heart murmur, abnormal rhythm, or organomegaly. In addition, given the association with genetic syndromes, the infant may have dysmorphic features to help guide the diagnosis. Primary evaluation should include pulse oximetry, CXR, and arterial blood gas along with treatment to support ventilation and oxygenation. Chest radiograph may show pulmonary edema and cardiomegaly (Figure 7). These infants can be difficult to differentiate from other presentations of neonatal respiratory distress or shock so treatment should include management for neonatal sepsis/pneumonia. In differentiating cardiac from noncardiac causes, an oxygen saturation unresponsive to 100% FiO₂ suggests a congenital heart defect. Diagnosis is confirmed by echocardiogram. In any infant with concern for a
ductal-dependent heart lesion, intravenous prostaglandin E1 should be started to prevent permanent ductal closure. Urgent consultation with pediatric cardiologist and/or neonatologist should be obtained.

Esophageal Atresia/Tracheoesophageal Fistula
Esophageal atresia (EA) and tracheoesophageal fistula (TEF) are congenital abnormalities of the esophagus and/or trachea that usually involve a connection or fistula between the 2 structures. The types of EA and TEF vary by presence or absence of EA and location of the TEF. The most common TEF is type "C," which is characterized by proximal EA with distal TEF. In infants with TEF, about 50% have associated anomalies like VACTERL association (Vertebral anomalies, Anal atresia, Cardiac defects, TEF, Renal abnormalities, Limb anomalies) or chromosomal abnormalities. Infants with EA typically present in the immediate neonatal period but TEF without EA may have a delayed presentation. Pregnancy complications may include polyhydramnios. Infants with EA/TEF have difficulty handling their secretions due to the blind esophageal pouch and are prone to aspiration. They can present with drooling, cough, respiratory distress, or dehydration. Diagnosis is made by inability to pass a nasogastric tube. Chest radiograph will show the nasogastric tube coiled in the proximal esophageal pouch (Figure 8). Treatment should include placement of a decompression tube in the esophageal pouch to clear secretions, respiratory support as needed, and placement of peripheral intravenous for administration of fluids. These infants should be transferred to a NICU with pediatric surgery capability.

Figure 8. Esophageal atresia with tracheoesophageal fistula. Note looped gastric tube in the esophageal pouch and presence of gastric air.

Upper Airway Disorders
Upper airway disorders are a broad category referring to abnormalities in nasal, oral, laryngeal, or tracheal portions of the airway. The presentation varies in timing and severity depending on specific etiology. Because newborns are obligate nasal breathers, an obstruction in the nasal passages like choanal atresia or intranasal tumor can present soon after birth. Oral lesions like lymphatic malformations or cysts (eg, dermoid) can cause airway obstruction due to mass effect. Laryngeal and tracheal airway anomalies such as laryngomalacia, laryngeal web, or hemangiomas can present closer to 4 to 6 weeks of life. Laryngomalacia is the most common cause of stridor in the newborn. Findings may include cyclic cyanosis (cyanosis when mouth is closed), difficulty feeding, tachypnea or apnea, retractions, inspiratory or biphasic stridor, abnormal cry, masses, or improvement when prone. Further evaluation can include attempting to pass a 6F catheter through both nares to assess for patency of choanae. Chest radiograph can assist in diagnosis of vasculature malformations or to rule out other etiologies. Consultation with an otolaryngologist is recommended for further airway management and definitive diagnosis that may require endoscopy, advanced imaging, and corrective procedures.

Interstitial Lung Diseases
Childhood interstitial lung disease (chILD) is a broad classification for disorders that result in impaired gas exchange due to abnormalities in the pulmonary interstitium or airway. Onset of respiratory symptoms can be variable even within the same type of chILD. Consider this diagnosis if there is a family history of lung disease or in infants with persistent or worsening symptoms despite ruling out the more common causes of respiratory distress. Common types of chILD which present in the newborn period include genetic disorders of surfactant proteins (eg, surfactant protein C deficiency), diffuse developmental disorders (eg, alveolar capillary dysplasia), growth abnormalities (eg, pulmonary hypoplasia), or undefined (eg, neuroendocrine cell hyperplasia of infancy or pulmonary interstitial glycogenosis). Newborns with chILD are most likely to present with tachypnea, but hypoxemia, abnormal lung examination results including crackles or wheezing, cough, and failure to thrive can be seen. Workup for this disorder should include a CXR and echocardiogram to rule out diseases that present similarly to chILD. Further evaluation can include lung computed tomography, lung biopsy, or genetic testing for surfactant
dysfunction mutations. Consultation with a neonatologist or pediatric pulmonologist with expertise in chILD is recommended.

**SUMMARY**

Newborns with respiratory distress can present with a wide range of symptoms that can overlap with those seen in a variety of diseases. It is critical to first stabilize the infant and then initiate further evaluation including a thorough history and examination, blood work, and CXR, which will help direct your differential diagnoses. Important diagnoses to consider include transient tachypnea, RDS, infection, MAS, pneumothorax, CDH, persistent pulmonary hypertension, congenital heart disease, EA/TEF, upper airway disorder, and interstitial lung disease. Prompt stabilization, diagnosis, and treatment are key to a successful outcome, as is early consultation with neonatologists and other pediatric specialists when appropriate.

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**REFERENCES**

Abstract:
The symptom of emesis in the neonate is common and caused by a myriad of clinical states, some pathologic and some benign. There are many clinical data points that steer the astute clinician toward certain diagnoses and away from others. The focus of this article is to provide a framework for evaluating a neonate that presents to an emergency department with emesis. After reading this article, the emergency department clinician will have a better understanding of the clinical presentation and evaluation of surgical and nonsurgical etiologies of emesis in the neonate.

Keywords:
neonate; emesis; emergency department; newborn; pyloric stenosis; gastrointestinal; gastroesophageal reflux; gastroesophageal reflux disease; malrotation; volvulus; intestinal atresia; Hirschsprung disease; incarcerated hernia; necrotizing enterocolitis

Emesis in the Neonate: Recommendations for Initial Management

Ann G. Downey, MD

Emesis in the neonate is a common presenting complaint in the emergency department (ED). Studies have noted a chief complaint of emesis in 11 to 36% of neonatal ED visits. The severity of vomiting is broad and ranges from simple, uncomplicated gastroesophageal reflux (GER) to life-threatening etiologies. It can be challenging to determine the severity of overall illness and emesis in an infant amid what is often a high level of parental concern. The focus of this article is to provide a framework for evaluating a neonate that presents to the ED with emesis. By the end of this article, the clinician will know the common causes of neonatal emesis and have a solid grounding in what tests to order, when to admit patients, and when to consult pediatric surgery.

HISTORY AND PHYSICAL EXAMINATION

When evaluating the presenting complaint of neonatal emesis, strong skills in history taking and physical examination are paramount. There are several striking features in the history and the physical examination that can narrow the focus for the clinician, help eliminate tests, and minimize delay in diagnosis of potentially life-threatening etiologies of neonatal emesis.

Key features in the history include a birth history focusing on gestational age, delivery complications, need for admission to the neonatal intensive care unit, and birth weight. Elements notable in the prenatal history include maternal laboratory values, prenatal ultrasound scans, and the levels of amniotic fluid, as polyhydramnios may be one of the first indications of intestinal obstruction. Finally, a
focused family history includes details such as milk-protein allergy in siblings and if an infant is a first-born male (at higher risk for pyloric stenosis). The presence of fever and a history of accidental or nonaccidental trauma are also critical elements of the history.

A feeding history including what an infant consumes, total volume of feeds, frequency, feeding behavior, and an elimination history for both urine and stool should be obtained. Calculating the percentage an infant is below birth weight is important. Healthy neonates regain their birth weight by 2 weeks of age at the latest and should not be greater than 10% below birth weight at any given time following birth. In addition, infants should gain 20 to 30 g/d after the below birth weight at any given time following birth. In the case of necrotizing enterocolitis or intussusception. In addition, establishing the presence of a patent anus is a critical element of the examination.

Hallmark features of concern in the history and physical examination include bilious emesis, weight loss, lethargy, an obtunded infant, shock, dehydration, and an acute abdomen.

**DIAGNOSTIC EVALUATION**

The evaluation of a neonate presenting with emesis offers the clinician a wide array of tests to rule in or out specific etiologies (Table 1). However, many tests are nonspecific and do not provide clarity of diagnosis. In addition, for many well-appearing, hydrated neonates with nonbilious emesis, a complete history and physical examination should suffice as long as there is close follow-up from the primary care physician. A metered blood glucose should be done for any lethargic, obtunded, or dehydrated infant. Electrolytes may yield information about a neonate’s hydration status with hypernatremia or a low bicarbonate level indicate dehydration in an infant. The presence of hypokalemic, hypochloremic metabolic alkalosis is

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**TABLE 2. Imaging mode of choice for common etiologies of emesis.**

<table>
<thead>
<tr>
<th>Etiology</th>
<th>Imaging Mode of Choice</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypertrophic pyloric stenosis</td>
<td>Pyloric ultrasound scan</td>
</tr>
<tr>
<td>Intestinal malrotation ±</td>
<td>Upper gastrointestinal (GI) series</td>
</tr>
<tr>
<td>midgut volvulus</td>
<td></td>
</tr>
<tr>
<td>Intestinal atresia or stenosis</td>
<td>Abdominal plain film ± upper or lower GI</td>
</tr>
<tr>
<td>Meconium syndrome</td>
<td>series</td>
</tr>
<tr>
<td>Hirschsprung disease</td>
<td>Abdominal plain film followed by lower GI</td>
</tr>
<tr>
<td>Necrotizing enterocolitis</td>
<td>2-View abdominal plain film</td>
</tr>
<tr>
<td>Increased intracranial pressure</td>
<td>MRI if available and time allows. CT if</td>
</tr>
<tr>
<td></td>
<td>MRI not available or need is emergent.</td>
</tr>
</tbody>
</table>

Abbreviations: CT, computed tomography; MRI, magnetic resonance imaging.
seen with hypertrophic pyloric stenosis while the triad of hyponatremia, hyperkalemia and hypoglycemia may be seen in congenital adrenal hyperplasia. A blood gas to ascertain a neonate’s acid base status may be helpful especially in neonates that are ill appearing. Infants with a metabolic acidosis may be presenting with an inborn error of metabolism, sepsis, an ingestion, or shock. (See Table 2.)

Traditional radiographic studies include abdominal plain films, abdominal ultrasound, upper gastrointestinal (GI) series, and lower GI contrast series and are chosen based on which disease process is most likely. The use of multichannel intraluminal impedance studies or endoscopy for GER is generally reserved for the outpatient or inpatient settings. The use of abdominal computed tomography (CT) or magnetic resonance imaging (MRI) for neonates with emesis is unusual and most often used when neonatologists and pediatric surgeons care for infants in the neonatal intensive care unit. If raised intracranial pressure is suspected, imaging of the CNS may be necessary.

GENERAL MANAGEMENT OF THE SICK NEONATE WITH EMESIS

Infants with emesis that are ill or toxic appearing should have stabilization of their airway, breathing, and circulation. Intravenous access, most commonly with a peripheral venous catheter, should then be established in sick neonates with emesis. If the neonate is dehydrated, a 10 mL/kg bolus of isotonic sodium chloride solution should be administered and repeated at intervals depending on clinical status. A blood glucose should be measured, and swift correction of hypoglycemia with a 2 mL/kg bolus of D10W should be done. Repeated glucose measurements should be taken with repeated boluses of D10W until hypoglycemia has been corrected. Electrolyte abnormalities should also be recognized and corrected. Gastric decompression should be performed in the case of an obstructive etiology and infants should be made nil per os.

MANAGEMENT OF SPECIFIC SURGICAL ETIOLOGIES

Hypertrophic Pyloric Stenosis

Hypertrophic pyloric stenosis (HPS) is a condition that develops in an infant when the circumferential muscle of the pyloric sphincter becomes thickened and creates a narrowing of the gastric outlet. The incidence is 0.1 to 1% of the population, and it classically presents in term, male infants 3 to 6 weeks of age. The history is notable for progressive, nonbilious, projectile emesis with every feeding and an infant that appears hungry and interested in feeding following the episodes of emesis. At the point that many patients present to the ED, infants may have progressed and have symptoms of dehydration. A family history of HPS may also be present.

Physical examination of the infant may reveal an infant that is dehydrated or jaundiced, and may also have the classic physical finding of a small, round, mobile mass in the epigastric region termed an olive. When identified, the presence of this olive has a specificity greater than 97%.

Evaluation of the infant with concern for HPS includes an electrolyte panel which may reveal hypokalemia, hypochloremia, and a contraction metabolic alkalosis. The imaging mode of choice is an ultrasound exam of the pylorus which measures the diameter, wall thickness, and pyloric channel length. When findings are equivocal, an upper GI study can be diagnostic when it shows an elongated and narrowed pyloric channel with characteristic shoulders of the hypertrophied pylorus protruding into the gastric lumen.

Surgery for HPS is not emergent. Time may be taken to ensure that a patient with HPS is hydrated, euglycemic, and without acid-base derangement before surgery. The treatment for HPS is a pyloromyotomy which may be performed open or laparoscopically. Most infants are discharged from the hospital 24 to 48 hours following a pyloromyotomy.

Malrotation With Midgut Volvulus

Malrotation presents in the first month of life in more than half of patients with about 75% of patients presenting in the first year. Bilious emesis in the child younger than 1 year should be presumed to be malrotation until proven otherwise. It occurs when the bowel fails to make two 270° counterclockwise rotations in the 6th and 12th weeks of gestation and to fix itself appropriately within the abdomen. When this does not occur, the bowel is at risk for obstruction or volvulus. Although the incidence is estimated to be 1% of the population, it only leads to clinical discovery in 1 in 6000 infants. Although the presentation may be variable in older patients, neonates generally present with bilious emesis and progress to a toxic-appearing infant with shock and the passing of bloody stools.

Physical examination of the infant may reveal an unstable, toxic-appearing infant prompting immediate clinical concern. However, if the malrotation is intermittent, this ill appearance may wax and wane.

The standard for diagnosing intestinal malrotation is an emergent upper GI series which should
not be delayed to obtain blood work. The upper GI series reveals the position of the duodenojejunal junction anterior, low and either midline or to the right of the midline. It is normally positioned to the left of midline. If volvulus is present, the small intestine may develop the appearance of a bird’s beak sign or a corkscrew appearance if the intestine is partially obstructed. The upper GI series must be performed by an experienced radiologist.

The most critical element in the management of an infant with malrotation with midgut volvulus is to consult with a pediatric surgeon emergently. This condition, left untreated, can lead to necrosis of the bowel, short gut syndrome, and death. Although multiple steps of management should be taken simultaneously to care for an infant with this condition, surgery should not be delayed. If time allows, fluid resuscitation, placement of nasogastric tube, blood for type and cross-match, and administration of broad spectrum antibiotics should be done.

The surgery performed for this condition is called the Ladd procedure which can be performed laparoscopically or by opening the abdomen. It involves detorsion of the volvulus, widening the mesenteric base, division of the mesenteric or “Ladd” bands, and placing the small intestine on the right and colon on the left in a nonrotation fashion. An incidental appendectomy is also done to avoid possible confusion for the clinician in the future, as the appendix would otherwise be returned to the left upper quadrant.

Recovery times following the surgery vary.

Intestinal Atresia

Intestinal atresia and stenosis may occur anywhere along the GI tract. Depending upon the level of atresia or stenosis, in relation to the ampulla of vater, bilious emesis may be present. An infant may be able to pass stool, often liquid, if an intestinal stenosis is present. Prenatal history may be notable for polyhydramnios. Most infants presenting to the ED with an intestinal atresia or stenosis will have poor weight gain and potentially evidence of dehydration.

Evaluation of the infant includes plain films of the abdomen, which may reveal the double-bubble sign in the case of duodenal atresia (Figure 1) or the triple-bubble sign in the case of jejunal atresia. Distended air-filled loops of bowel with air fluid levels are usually observed, with a greater number of these seen when the obstruction is present in the distal bowel. When intestinal obstruction or stenosis is suspected but not confirmed by an abdominal plain film, an upper GI series or lower GI series should be considered based on the suspected level of the lesion.

Consultation with a pediatric surgeon is essential for correction of an intestinal atresia or stenosis. Management is focused on prompt placement of a nasogastric tube for gastric decompression and intravenous access for fluid resuscitation. Serum studies of electrolytes and complete blood counts should be obtained before operative management. Surgical management depends entirely upon the level of the atretic or stenotic region of bowel and involves identifying the level of the lesion, excising the lesion, and performing an anastomosis on the remaining bowel. Complications of intestinal atresia and stenosis include the development of anastomotic stricture, anastomotic leakage, and sepsis.

Meconium Syndromes

Meconium syndromes often present with intestinal obstruction due to the presence of thick and inspissated meconium that develops in utero. There are multiple meconium syndromes, but for the purposes of the ED physician, meconium ileus is the most relevant. Although infants should not be discharged from birth hospitals until they have passed meconium, for infants born at home or in centers without this requirement for discharge, clinical signs of intestinal obstruction often develop in the first few days of life with failure to pass stool and bilious emesis.

Abdominal films often reveal classic features of intestinal obstruction. The tiny air bubbles in meconium will present as granular, soap-like bubbles on plain film. A contrast enema usually demonstrates a microcolon. Consultation with a pediatric surgeon is paramount because this condition can progress to volvulus, necrosis of the bowel, and perforation of the bowel if left untreated.
Meconium ileus is associated with cystic fibrosis about 80% of the time.  

Hirschsprung Disease
The vast majority of patients with Hirschsprung disease present in the neonatal period. They present with symptoms of obstruction, abdominal distention, emesis, and the failure to pass meconium. Although most infants fail to pass meconium, roughly 5% of patients do pass some stool in the first 24 hours of life.  

An abdominal plain film will likely reveal evidence of obstruction. A contrast enema may reveal a rectum that is smaller in diameter than the colon, a transition zone, and a normal caliber colon proximal to the transition zone. Contrast is often not evacuated completely until 24 hours after the contrast study is performed. Consultation with a pediatric surgeon is necessary, and the definitive diagnosis is made by rectal biopsy when aganglionosis and hypertrophy of the nerve trunks are identified.

There are several approaches to treating Hirschsprung disease, but the principle for all approaches is to resect the aganglionic segment of bowel and unite the remaining bowel. This may be done in a single operation or may be performed in a staged approach.  

Incarcerated Inguinal Hernias
An incarcerated inguinal hernia is a diagnosis usually made by history and physical examination. Former preterm infants are particularly vulnerable to inguinal hernias. Parents are often aware of the hernia or at least of the changed appearance of the groin or scrotum. Clinical examination of the diaper area will confirm the diagnosis. If not detected by history or physical examination, the abnormality can be seen on abdominal plain film with bowel appearing in the inguinal region or scrotum.

Manual reduction in the ED should be attempted by the emergency physician alone or in consultation with a pediatric surgeon. One study found that manual reduction was successful in 90% of patients with incarcerated inguinal hernias. Regardless of success, the patient should have a herniorrhaphy either emergently or electively following manual reduction.  

MANAGEMENT OF
SPECIFIC NONSURGICAL ETIOLOGIES
Gastroesophageal Reflux and Gastroesophageal Reflux Disease
Gastroesophageal reflux refers to the passage of gastric contents into the esophagus. This can result in infants spitting up or having emesis. It is a normal physiologic process that occurs in about 50% of infants in the first 3 months of life. Infants that are gaining appropriate weight; are well appearing; and have nonbilious, episodic regurgitation are considered to be part of this 50% of normal infants. Reassurance and possibly thickened formula for babies not breastfed are the only interventions needed.

Gastroesophageal reflux disease (GERD) refers to the complications of GER. It is generally considered a diagnosis of exclusion because there are no symptoms that can reliably diagnose GERD or predict treatment response. To accurately diagnose whether GER symptoms are associated with the presence of both gastric contents and acid exposure in the esophagus, a combined multiple esophageal impedance-pH study is needed, although this study is not often performed, and instead, staged treatment is guided by primary care providers or pediatric gastroenterologists prescribing specific interventions over time.

Some initial therapies that can be suggested to help parents are holding infants upright after a feeding for a period of about 30 minutes; avoiding placing infants in car seats directly after feeding to avoid an increase intraabdominal pressure; avoiding excessive feeding volumes; feeding breast milk alone; or, if formula feeding, conducting a trial of reflux specific formula. A trial of an elemental formula for infants with possible cow’s milk-protein allergy can be considered but generally should be initiated by a primary care physician. Reflux medications such as histamine-2 antagonists and proton pump inhibitors have been linked in recent years to gastroenteritis and pneumonia in pediatric populations and have not been shown to be effective in multiple studies for treating symptoms associated with GERD. Black box warnings for these agents have led the American Academy of Pediatrics to recommend the cautious and limited use of acid suppression agents in infants.

Given these concerns, trials of acid suppression medications, promotility agents, and gastric-emptying agents should not be prescribed in EDs for infants. The American Academy of Pediatrics recommends consultation with a pediatric GI specialist before starting medications for infants with GERD.  

Sepsis and Infection
Sepsis and infection in the neonate can be accompanied by emesis. Key elements of the history include fever, lethargy, and sick contacts. The differential diagnosis in the neonate with emesis and fever includes pneumonia, sepsis, urinary tract
infection, meningitis, gastroenteritis, viral syndromes, and rarer causes such as osteomyelitis. According to a policy statement by the American College of Emergency Physicians, for infants younger than 28 days with fever of at least 38°C, a full sepsis evaluation with blood culture, catheterized urine culture, and cerebrospinal cultures should be obtained before starting broad-spectrum intravenous antibiotic therapy and admitting the infant to the hospital.17

Additional data that are helpful in the evaluation of emesis with fever in an infant with suspected infection include a complete blood count and an abdominal plain film, which might reveal evidence of a septic ileus. A more detailed review of sepsis in the newborn is addressed in other articles in this issue.

Inborn Errors of Metabolism
Metabolic disease, often detected with state newborn screening programs, may present to the ED before the results of the screening test becoming available. Infants are usually ill appearing with electrolyte disturbances and, often, metabolic acidosis noted on blood gas measurements. Fluid resuscitation, elimination of protein and fat intake until further information is obtained, and referral to a hospital with pediatric metabolic specialists are warranted.

Increased Intracranial Pressure
Increased intracranial pressure can lead to emesis in the neonate from a variety of etiologies including hemorrhage from trauma (accidental or not), stroke, tumor, meningitis, hydrocephalus, and CNS malformation, among others. History may reveal clues to the etiology, and physical examination may reveal an infant with a bulging anterior fontanelle; splitting of the cranial sutures; and, in more advanced cases, sundowning of the eyes, obtundation, and apnea with bradycardia.

Emergent imaging of the central nervous system with CT or MRI followed by consultation with a pediatric neurosurgeon or pediatric trauma service may be necessary.

Necrotizing Enterocolitis
Although necrotizing enterocolitis occurs most frequently in preterm infants, it can present in late-preterm and term infants with abdominal tenderness and distention, bloody stools, and emesis (Figure 2). Characteristic abdominal plain film findings include pneumatosis intestinalis, with portal venous air, and sometimes free air. Infants with necrotizing enterocolitis should be made nil per os and have blood cultures drawn, broad-spectrum intravenous antibiotic therapy should be started, and they should be admitted to a hospital where pediatric surgery is available for consultation.

Feeding Intolerance
Emesis in the neonate is occasionally caused by intolerance of an infant’s feeding regimen. Cow’s milk-protein allergy can present in both formula-fed and breast-fed infants, although it is much less common in breast-fed infants. Altering an infant’s formula or dairy elimination in a mother’s diet may be all that are needed to treat this condition, which is best managed in the outpatient setting unless it is severe.

SUMMARY
Ultimately, there are many etiologies that lead to emesis as a primary symptom in the case of intestinal obstruction or as an associated symptom in the case of sepsis. It can be challenging for any physician to determine an etiology and an initial management strategy. Some principles to remember include that bilious emesis should be considered to be malrotation until proven otherwise. Ill, toxic, or dehydrated infants always need further evaluation and usually hospital admission. Infants that are failing to thrive require hospital admission when they present to the ED. Pediatric surgery should be consulted when there is concern for hypertrophic pyloric stenosis and intestinal obstruction. For patients that are well appearing, hydrated, not failing to thrive, and without features in their history that prompt further
evaluation, reassurance and follow-up with their primary care physicians are indicated.

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CONFLICT OF INTEREST STATEMENT

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REFERENCES

Abstract:
Sepsis is a significant cause of morbidity and mortality for neonates and infants. Neonates are at increased risk for sepsis due to their immature immune system. Bacterial, viral, and fungal organisms may cause sepsis in the young patient. Identifying septic neonates upon presentation to their primary care physician or the emergency department remains a challenge given the nonspecific manifestations of illness. Suspicion for sepsis should prompt evaluation to identify a source to tailor treatment appropriately. Timely diagnosis and management of neonatal sepsis, especially for those in septic shock, will lead to improved outcomes. The following article presents an overview of the most common organisms causing disease, clinical presentation, evaluation, and management for the neonate or infant presenting with suspected sepsis.

Keywords:
neonatal sepsis; bacterial infection; viral infection; fungal infection; emergency department

Epidemiology and Pathogenesis

All neonates are at increased risk for infection due to their immature immune defenses. Studies have shown an increased rate of neonatal infection associated with lower gestational age and lower birth weight. In one study, the incidence of early-onset neonatal infection in infants born at term was 0.53 per 1000 live births. In the preterm population, the incidence of EOS was 3.71 per 1000 live births and 10.96 per 1000 live births in VLBW infants. There is also an increased incidence of LOS among VLBW neonates, especially those who require hospitalization, compared with term infants.

Risk factors for EOS include preterm birth, maternal colonization with group B Streptococcus (GBS), prolonged rupture of membranes longer than 18 hours, or maternal signs or symptoms of intraamniotic infection. GBS remains the leading cause of EOS in term neonates, although the overall national incidence has
decreased by 87% with the implementation of intrapartum antibiotic (IPA) for the prevention of early-onset GBS sepsis.\textsuperscript{13} IPA does not change the risk of getting LOS. Since the initiation of IPA for GBS, \textit{Escherichia coli} has emerged as the leading cause of EOS in preterm neonates.\textsuperscript{2,14}

In addition to low birth weight and gestational age, other risk factors for sepsis include immunodeficiency, indwelling catheters, and some inborn errors of metabolism, such as galactosemia, which may present as \textit{E coli} sepsis or urosepsis.\textsuperscript{15} After GBS, the other prevalent gram-positive organisms causing neonatal sepsis include \textit{Staphylococcus aureus}, coagulase-negative \textit{Staphylococcus}, \textit{Enterococcus}, and \textit{Listeria monocytogenes}. Other than \textit{E coli}, the most common gram-negative organisms are \textit{Klebsiella}, \textit{Enterobacter}, \textit{Citrobacter}, and \textit{Pseudomonas}.\textsuperscript{2,5,14}

Finally, viral and fungal organisms can also cause illness in neonates. Some key viral infections include herpes simplex virus (HSV), human immunodeficiency virus, enterovirus, respiratory syncytial virus, influenza, adenovirus, and rotavirus. Neonatal HSV generally presents in the first 6 weeks of life. Approximately 1500 cases of neonatal HSV infection occur every year in the United States.\textsuperscript{16} Neonatal HSV can present as skin, eye, and mouth disease; central nervous system (CNS) disease; or disseminated disease. EOS and LOS can also be caused by fungal infections, most often \textit{Candida albicans} or \textit{C parapsilosis}, found primarily in preterm VLBW infants.\textsuperscript{17}

### INITIAL MANAGEMENT AND DIAGNOSTIC TESTING

The septic infant or neonate may be “well-appearing” with few symptoms or may present in septic shock. Signs and symptoms of neonatal sepsis are more often nonspecific. Upon obtaining a complete history, any reported deviations in activity or feeding pattern should be taken seriously. An infant may have temperature instability, irritability, lethargy, respiratory distress, apnea, poor feeding, abdominal distension, jaundice, and/or tachycardia. Poor perfusion and hypotension are generally late findings but are sensitive indicators of sepsis.\textsuperscript{10}

In an infant presenting in septic shock, initial management should focus on stabilizing the patient. This may include alleviating airway compromise, providing respiratory support, and obtaining intravenous access for restoration of circulation and perfusion.\textsuperscript{18} According to the 2007 updated American College of Critical Care Medicine guidelines for management of pediatric and neonatal septic shock, fluid resuscitation with isotonic or colloid boluses should be given starting with 20 mL/kg up to a maximum of 60 mL/kg, with reassessment of liver size and rales indicating fluid overload. In a neonate in septic shock, correcting hypocalcemia and hypoglycemia is important. If fluid resuscitation does not restore perfusion, central access should be considered and inotropes started. Of note, until central access is available, inotropes may be started peripherally. Hydrocortisone should be administered to infants with suspected adrenal insufficiency, and until ductal-dependent congenital heart disease is ruled out, a prostaglandin infusion should be considered.

Laboratory testing should be performed on neonates with possible sepsis. Although available diagnostic testing is not always helpful in deciding who should receive antibiotics, it can assist in the decision regarding the appropriate time to discontinue treatment. According to the American Academy of Pediatrics guidelines on the management of early-onset neonatal sepsis,\textsuperscript{19} a blood culture is required for all neonates with suspected sepsis. When a single pediatric blood culture bottle is used, a minimum of 1.0 mL of blood is needed. Cultures from tracheal aspirates should be obtained if intubated.\textsuperscript{19} In addition, a complete blood count with differential and platelet count should also be obtained. If the patient presents in respiratory distress, a chest radiograph is recommended. A lumbar puncture should be performed if the infant has a positive blood culture, has suggestive laboratory data, is clinically symptomatic, or does not improve with initiation of antibiotics.\textsuperscript{19}

When obtaining a lumbar puncture, cerebrospinal fluid (CSF) should be sent for culture, cell count, and protein and glucose concentration. Acceptable values for CSF differ between term and preterm neonates. Several studies have examined the various indices. The mean number of white blood cells in uninfected preterm and term infants was less than 10 cells/mm\textsuperscript{3}.\textsuperscript{3,20,21} In infants with meningitis, the median number of white blood cells in those born less than 34 weeks was 110/mm\textsuperscript{3}, and in those greater than 34 weeks, it was 477/mm\textsuperscript{3}.\textsuperscript{3,22,23} Protein concentrations in preterm infants are inversely proportional to gestational age.\textsuperscript{19} In a term newborn, the protein concentration should be less than 100 mg/dL.\textsuperscript{20,21} Normal CSF glucose concentrations are similar to those in children. It is important to remember that some infants with meningitis have normal CSF values.\textsuperscript{20,21}

In infants 1 week of age or older, sterile urine culture and urinalysis are needed during the evaluation of LOS.\textsuperscript{19} Infants with EOS generally do
not have an ascending urinary tract infection; rather, seeding of the kidney is due to bacteremia. Additional laboratory testing should be based on each specific case taking into consideration prevalent organisms at the time of presentation. For example, nasopharyngeal secretions for antigen can be sent when respiratory syncytial virus, influenza, or adenovirus is suspected. When evaluating for enterovirus, CSF or blood should be sent for polymerase chain reaction studies or cultured from the throat, rectum, stool, blood, or urine.

Herpes simplex virus may present with nonspecific symptoms similar to bacterial sepsis; therefore, clinicians need to maintain a high index of suspicion for evaluating and treating the disease. Infants who develop HSV are frequently born to women who acquire genital HSV infection during pregnancy but either asymptomatic or have nonspecific symptoms. Typically, HSV presents in the second and third week of life, but HSV-infected infants can become symptomatic shortly after birth. Isolation of HSV by viral cell culture is the definitive laboratory method for identifying a neonate with active disease. Cultures should be obtained from any suspicious lesions, the mouth, nasopharynx, conjunctiva, and rectum. Blood and CSF can be sent for HSV polymerase chain reaction testing and are useful to determine if the patient has CNS involvement or disseminated disease.

Other laboratory tests to consider in the neonate undergoing evaluation for sepsis include bilirubin level, electrolytes, blood glucose level, blood gas, and coagulation studies. A neonate presenting with sepsis early on may also have hyperbilirubinemia requiring phototherapy treatment. Electrolytes will help identify metabolic derangements including those seen with dehydration and guide appropriate fluid resuscitation. Infants with sepsis may also have disseminated intravascular coagulation and may require transfusion for thrombocytopenia, prolonged prothrombin and activated partial thromboplastin times, and low fibrinogen levels. As in the patient with septic shock, hypoglycemia and hypocalcemia should be corrected, and respiratory support should be initiated as necessary.

**TREATMENT**

Infants presenting with suspected sepsis should be started on antibiotic coverage to target the most common pathogens with consideration for local resistance patterns. Ampicillin and an aminoglycoside, usually gentamicin, are commonly initiated when EOS is suspected. This drug combination covers *E. coli* and has synergistic activity against GBS and *Listeria monocytogenes*. If the infant has either renal or hearing issues, an alternative to aminoglycosides should be considered. A third-generation cephalosporin, commonly cefotaxime, is often considered as an alternative to gentamicin; however, development of resistance and increased risk of developing invasive candidiasis are reported. Excellent CSF penetration makes cefotaxime plus an aminoglycoside a good choice for the initial treatment of gram-negative meningitis.

Ceftriaxone should not be used in the first week of life because of potential displacement of bilirubin from albumin, increasing the risk of kernicterus. The Food and Drug Administration advises that the use of ceftriaxone and calcium-containing products should not occur within 48 hours of each other. Case reports have demonstrated that, when given together, calcium-ceftriaxone precipitate may form in newborn lungs and kidneys.

Vancomycin is often used in LOS because coagulase-negative *Staphylococcus* is a common causative organism. The Centers for Disease Control and Prevention have recommended against using vancomycin empirically because of emergence of vancomycin-resistant strains. Given concern for impaired neurodevelopmental outcomes in infants with coagulase-negative *Staphylococcus*, use of vancomycin should be used judiciously, targeting those with indwelling devices. In 1 study, most organisms causing LOS were susceptible to gentamicin with either flucloxacinil or amoxicillin. Oxacillin or nafcillin plus gentamicin is a commonly used regimen. As in EOS, a third-generation cephalosporin should only be used if meningitis is suspected.

In both EOS and LOS, when an organism is identified, the antibiotic regimen should be tailored appropriately. Duration of therapy remains controversial; however, discontinuation of empirical antibiotic therapy should be considered after 2 to 3 days of negative cultures if clinical status remains stable. Just as there are risks with not starting antibiotics in a timely fashion, unnecessary antimicrobial therapy can be harmful. Studies have shown an association with prolonged antibiotic administration in infants with EOS and necrotizing enterocolitis and death. Given high cost and emerging antimicrobial resistance patterns resulting from inappropriate antimicrobial antibiotic use, it is important to consider antimicrobial stewardship.

Intravenous acyclovir should be administered if HSV is suspected. The treatment has been shown to decrease mortality and morbidity in those with CNS disease and disseminated disease. In skin, eye, and
mouth disease, acyclovir may also prevent progression to CNS or disseminated disease.

The most common antifungal therapy includes amphotericin B and fluconazole. Newer therapy is currently being studied for safety and efficacy in neonates.

SUMMARY

Neonatal sepsis remains a significant cause of morbidity and mortality not only during initial hospitalization but also after discharge. Identifying septic neonates who present to their primary care physician or the emergency department often remains a challenge given the nonspecific manifestations of illness. Suspicion for sepsis should prompt evaluation to identify a source to tailor treatment appropriately. Timely diagnosis and management of neonatal sepsis, especially for those in septic shock, will lead to improved outcomes for the patients.

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REFERENCES


The
Decompensated Neonate in the First Week of Life

Silena C. Chapman, MD

Many critical and potentially fatal illnesses can arise in the first week of life, making rapid and efficient evaluation and stabilization paramount in this population. Neonates often present with nonspecific findings and may decompensate quickly. It is vital that caregivers are able to recognize at-risk neonates, maintain a broad differential (Table 1), and approach evaluation and management simultaneously and methodically. Diagnostic priority should be placed on ruling out immediately life-threatening etiologies such as sepsis, critical congenital heart disease (CHD), inborn errors of metabolism, endocrine crises, and hematologic emergencies.

RECOGNIZING THE CRITICALLY ILL NEONATE

The evaluation and identification of at-risk neonates often occur in triage before a physician sees the patient. The ability to identify abnormal initial history, appearance, and vital signs is key to appropriate triage and timely care. A change of behavior, noted by the family, is often the initial clue to impending decompensation.

A typical neonate sleeps approximately 15 to 18 hours per day, awakening spontaneously every 2 to 4 hours for feeding. Parents should report 8 to 12 feedings of either formula or breast milk, with the infant taking at least 2 oz or nursing for 10 to 15 minutes at the breast. The infant should also have a minimum of 6 to 8 wet diapers in a 24-hour period. Normal stooling patterns are variable; however, soft, seedy, yellow-to-green stools should be accepted as normal, and any stools with blood or mucus or that are clay colored must be considered abnormal. Frequent spitting up is a common complaint in this age group, as two thirds of normal, healthy infants have gastroesophageal reflux. Gastroesophageal reflux can be associated with vomiting of gastric contents, typically

Abstract:
The first week of life can be a critical period in which previously subclinical disorders may manifest, causing significant illness. Newborn infants also have different baseline vitals and laboratory parameters, which complicate the initial evaluation of infants. Rapid identification of sick neonates, stabilization, and directed evaluation are keys to minimizing long-term morbidity and mortality. This review targets 5 conditions that present in the first week of life: neonatal sepsis, critical congenital heart disease, inborn errors of metabolism, congenital adrenal hyperplasia, and hemorrhagic disease of the newborn. The primary focus is on key findings, initial evaluation, and immediate treatment in the emergency department where these infants often present after discharge from the newborn nursery.

Keywords:
neonate; shock; early-onset sepsis; late-onset sepsis; congenital heart disease; cyanosis; inborn errors of metabolism; congenital adrenal hyperplasia; hemorrhagic disease of the newborn; vitamin K–deficient bleeding

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described as partially digested breast milk or formula; however, reports of green-colored (bilious) or forceful (projectile) emesis require further evaluation.

Visual inspection of the infant can be quick and telling regarding the status of the neonate. A healthy neonate should appear comfortable, with a normal respiratory effort. Infants with significant respiratory distress will sometimes develop head bobbing. The infant should have flexed extremities with spontaneous movements. The infant's color should be pink to indicate adequate perfusion, although some infants may become mottled when unwrapped in cold environments. Mucous membranes, as well as palms and soles, can be evaluated for color in darkly pigmented infants. The infant may not be vigorous if sleeping but when presented with stressful stimuli, such as blood pressure measurement, should show signs of arousal.

Abnormal vital signs may be the key to identifying a baby in shock. Over the first days and weeks of life, infants' vital signs change dramatically and need to be understood based on referenced norms (Table 2). A rectal temperature is the preferred method of recording and should be between 36.5°C and 38°C, or 97.7°F and 100.4°F. The normal respiratory rate of an infant is between 30 and 60 breaths per minute. Heart rate can vary depending on the neonate's sleep-wake status and activity level. Typically, a heart rate of 85 to 205 beats per minute when awake and 80 to 160 beats per minute when sleeping is considered appropriate. Blood pressure measurement may not be standard in initial emergency department triage of neonates but is important in this population to determine hemodynamic stability. A systolic blood pressure of 65 to 85 mm Hg and a diastolic blood pressure of 45 to 55 mm Hg are considered adequate for age. A mean blood pressure that approximates the corrected gestational age of the infant can be used as a quick assessment of adequacy. After the initial transition period at birth, the normal oxygen saturation regardless of probe placement should be greater than 94%. Values outside the parameters listed in Table 2 should alert the caregiver that the infant requires immediate attention.

Initial laboratory tests and imaging can be obtained to support the evaluation of the sick neonate. Preliminary laboratory tests can include a complete blood count (CBC), blood culture, blood gas, lactate, and complete metabolic panel. Chest and abdominal plain radiographs can be obtained for infants exhibiting signs concerning for respiratory or gastrointestinal disorders. Seizing infants or those with suspected intracranial processes should undergo neuroimaging including head computed tomography or magnetic resonance imaging.

### NEONATAL SEPSIS

Infection can be a devastating and unfortunately common disorder in the first week of life, occurring in 1 to 5 neonates per 1000 live births.

### TABLE 1. Differential diagnosis for decompensation in the first week of life

<table>
<thead>
<tr>
<th>Infection</th>
<th>Cardiac</th>
<th>Metabolic</th>
<th>Endocrine</th>
<th>Hematologic</th>
<th>Miscellaneous</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bacterial/Viral Sepsis, Neonatal Pneumonia, Meningitis</td>
<td>Ductal-dependent Systemic Blood Flow (shock): Coarctation of the Aorta, Interrupted Aortic Arch, Pulmonary Valve Stenosis</td>
<td>Amino Acidopathies, Urea Cycle Enzyme Defects, Organic Acidurias</td>
<td>Congenital Adrenal Hyperplasia: 21-hydroxylase Deficiency, 3B-hydroxylase Deficiency</td>
<td>Hemorrhagic Disease of the Newborn</td>
<td>Seizures, Gastrointestinal Emergencies, Non-accidental Trauma, Toxins, Formula Mixing Errors</td>
</tr>
<tr>
<td>Neonatal Herpes: Skin, Eye, Mouth (SEM) Disease, CNS Disease, Disseminated Disease</td>
<td>Valvular Aortic Stenosis, Hypoplastic Left Heart Syndrome</td>
<td>Disorders of Carbohydrate Metabolism, Disorders of Bilirubin Metabolism, etc.</td>
<td>Deficiency, 11B-hydroxylase Deficiency, Neonatal Thyrotoxicosis</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

### TABLE 2. Normal newborn vital signs.

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Temperature</td>
<td>36.5-38°C</td>
</tr>
<tr>
<td>Respiratory rate</td>
<td>30-60 breaths per minute</td>
</tr>
<tr>
<td>Heart rate</td>
<td>85-205 beats per minute (awake)</td>
</tr>
<tr>
<td></td>
<td>80-160 beats per minute (asleep)</td>
</tr>
<tr>
<td>Blood pressure</td>
<td>Systolic 65-85 mm Hg</td>
</tr>
<tr>
<td></td>
<td>Diastolic 45-55 mm Hg</td>
</tr>
<tr>
<td>Oxygen saturation</td>
<td>&gt;94%</td>
</tr>
</tbody>
</table>
Sepsis is divided into early onset, occurring before 72 hours of life, and late onset, occurring after 72 hours of life. Although the common bacterial, viral, and fungal pathogens vary depending on timing of presentation, group B Streptococcus (GBS), Escherichia coli, and herpes simplex virus (HSV) can be causative in both early- and late-onset sepsis syndromes. Particular attention to risk factors for infection present at birth may place infection higher or lower on the differential diagnosis. Risk factors include maternal GBS colonization, history of chorioamnionitis during labor, prolonged rupture of membranes (>18 hours), history of premature rupture of membranes, or maternal history of HSV.\textsuperscript{10,11}

Presenting history and symptoms may be quite variable. Common presenting complaints include difficulty waking (lethargy), inconsolability, fever, hypothermia, poor feeding, poor tone, and signs of respiratory distress. Infants have the potential to progress from symptomatic to decompensated septic shock quickly, so the workup should progress rapidly. When sepsis is suspected, a CBC and blood culture must be obtained, and empiric treatment should be initiated without delay. Important indices on the CBC include the total white blood cell count, the mature (or segmented) neutrophil count, the immature (or band) neutrophil count, and the platelet count. Values outside the reference range for age for any of these indices may indicate the presence of an infection. In addition, an immature-to-total neutrophil (I/T) ratio greater than 0.20 can be a sensitive measure of infection, with a negative predictive value of 99%.\textsuperscript{10} In addition to blood, urine should be obtained for analysis and culture. The remainder of the sepsis evaluation can be directed by history, and the decision to perform a lumbar puncture can be based on clinical stability, as well as historical and clinical likelihood of meningitis.

If viral sepsis from HSV is suspected, the infant should have serum and cerebrospinal fluid samples sent for polymerase chain reaction assays. Many infants who acquire HSV at birth are born to women who acquire genital HSV infection during pregnancy but either are asymptomatic or have nonspecific symptoms.\textsuperscript{12} In addition, HSV viral surface cultures from the conjunctiva, mouth, nasopharynx, and rectum should be obtained.

Timely empiric treatment with broad-spectrum antibiotics is top priority in infants with suspected sepsis (Table 3). Routinely, ampicillin in combination with gentamicin or a third-generation cephalosporin, such as cefotaxime, is used for their synergy against the common pathogens in neonatal sepsis. Any infant suspected of HSV infection and who undergoes viral culturing should also receive empiric acyclovir.

### Critical Congenital Heart Disease

CHD occurs in approximately 1% of live births.\textsuperscript{9} Although many lesions will present in the immediate postnatal period before discharge from the newborn nursery, there is a subset of lesions that remain subclinical due to compensation from an open ductus arteriosus. In these cardiac defects, systemic or pulmonary blood flow is dependent on the open ductus arteriosus, resulting in infants who present with shock or cyanosis, respectively, with ductal closure.

The criterion standard for diagnosis of CHD is the echocardiogram; however, this may not be readily available in the emergency department, and several other quick diagnostic evaluations can and should be performed during initial assessment. Pulse oximetry can be used as an assessment of oxygenation. A thorough evaluation includes both preductal (right upper extremity) and postductal (either lower extremity) pulse oximetry measurements, which can be nearly 100% sensitive and 100% specific for detecting cyanotic heart disease.\textsuperscript{14} Four-extremity blood pressures should be obtained to look for differences between the upper and lower extremities, as well as overall adequacy of the blood pressure. An arterial blood gas is key to determining the presence of acidosis (pH, HCO\textsubscript{3}, base deficit) as well as inadequate oxygenation (pO\textsubscript{2}). A serum lactate will also provide information about tissue perfusion and oxygenation and can usually be obtained with the blood gas. Chest radiography can be performed to look for the presence of cardiomegaly or abnormal pulmonary vascular markings which may also be suggestive of CHD. An electrocardiogram (ECG) can be obtained but can be difficult to interpret in the first week of life because of characteristic ECG findings that are usually abnormal in other patient populations. As previously discussed, the normal heart rate of an

### TABLE 3. Empiric antibiotics and antivirals for suspected sepsis.\textsuperscript{13}

<table>
<thead>
<tr>
<th>Antibiotic</th>
<th>Dosage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amoxicillin</td>
<td>100 mg/kg per dose IV or IM every 12 h</td>
</tr>
<tr>
<td>Gentamicin</td>
<td>4 mg/kg per dose IV or IM every 24 h</td>
</tr>
<tr>
<td>Cefotaxime</td>
<td>50 mg/kg per dose IV or IM every 8 h</td>
</tr>
<tr>
<td>Acyclovir</td>
<td>20 mg/kg per dose IV only every 8 h</td>
</tr>
</tbody>
</table>

IM indicates intramuscularly.
The decompensated neonate in the first week of life

**INBORN ERRORS OF METABOLISM**

Metabolic disorders should be considered whenever a critical neonate is being evaluated in the emergency department, as incidence can be as high as greater than 1 in 500 live births. Although hospital-born infants are universally screened before discharge, results may not be available for weeks. During this time, unidentified neonates may begin to manifest symptoms of their disordered metabolic processes as a result of exposure to a particular catabolic insult. Infants that were normal in the regular newborn nursery and then suffered an acute change in status after feeding at home should be carefully evaluated for an inborn error of metabolism.

Details of each metabolic disorder and its sequelae fall outside the scope of this discussion; however, disorders can be roughly categorized as amino acid disorders, urea cycle disorders, organic acidemias, fatty acid oxidation defects, disorders of carbohydrate metabolism, and miscellaneous. Manifestations are characteristic of the disordered process; however, certain findings such as strange odor, feeding difficulties, vomiting, hepatomegaly, profound lethargy, and seizures should prompt the evaluation for an inborn error of metabolism.

The evaluation for inborn errors of metabolism is primarily laboratory driven. A blood gas should be obtained to look for the presence of metabolic acidosis, respiratory alkalosis, and an increased anion gap. Blood should be sent to evaluate ammonia, lactate, and pyruvate and for quantification of amino acids. A complete metabolic panel should be ordered to assess electrolytes, glucose, as well as renal and liver function. Ketones, reducing substances, and organic acids can be evaluated in the urine.

The goals of treatment in the evaluation of inborn errors of metabolism should focus on hydration, correction of acid-base imbalance, nutrition, and elimination of offending metabolites. Infants are allowed nothing by mouth and placed on age-appropriate parenteral fluids containing dextrose and electrolytes to prevent a catabolic state. Parenteral nutrition fluids containing protein are held until a specific disorder is identified. Comorbid issues such as sepsis or seizures should also be attended to. Many infants will require ongoing supportive care until definitive treatment can be provided with the identification of their specific inborn error of metabolism.

**CONGENITAL ADRENAL HYPERPLASIA**

The most common endocrine disorder that manifests in the first week of life is congenital adrenal hyperplasia (CAH). CAH results from 1 of 5 enzyme defects that lead to impaired cortisol production and increased adrenocorticotropic hormone production. Manifestations are characteristic to the enzyme defect; however, adrenal crisis secondary to salt wasting in 21-hydroxylase and 3β-hydroxylase deficiencies and severe hypertension secondary to salt-retention in 11β-hydroxylase deficiency are of particular interest, as approximately half of infants with untreated salt-losing CAH present in crisis between 6 and 14 days of age. Female infants will often have abnormal genitalia with some amount of virilization, whereas male infants often have normal genitalia. Infants often present with dehydration, vomiting, and shock-like symptoms.

CAH may be detected on the newborn screen; however, results may not be available within the first week of life. Serum quantification of 17-hydroxyprogesterone, 11-deoxycortisol, 17-hydroxyprogrenenolone, and cortisol may be helpful in diagnosing these disorders. These results may not be available immediately in the emergency department but should be sent as the patient is being stabilized. Electrolytes should also be obtained to identify abnormalities, with the most common type of CAH presenting with hyponatremia, hyperkalemia, and metabolic acidosis. Management should focus on correction of hypotension, dehydration, and electrolyte abnormalities. Identified cortisol deficiency should be corrected as well with

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**TABLE 4. Prostaglandin E1 dosing for ductal patency.**

<table>
<thead>
<tr>
<th>Dosage</th>
<th>Initial</th>
<th>Maintenance</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>0.05-0.1 μg/kg/min continuous IV infusion</td>
<td>0.01 μg/kg/min continuous IV infusion</td>
</tr>
</tbody>
</table>
stress-dose hydrocortisone as shown in Table 5. Inpatient management is encouraged until the specific disorder is diagnosed and ongoing medical management is established.

HEMORRHAGIC DISEASE OF THE NEWBORN

Hemorrhagic disease of the newborn or vitamin K–deficient bleeding, as it is also known, typically occurs in 3 distinct periods. Early disease, characterized by serious and catastrophic bleeding, occurs within the first 24 hours of life. Classic disease, which may be associated with cutaneous, gastrointestinal, circumsencephal, or intracranial bleeding, occurs up to 1 week of life in 1.7% of infants without vitamin K prophylaxis. Late disease can occur any time after the first week of life and may be associated with similar bleeding seen in classic disease but with a notably higher rate of intracranial hemorrhages. A recent increase in late-onset disease has been noted in the United States due to the increasing refusal of vitamin K at birth. 17,18

Active or serious bleeding should be managed promptly. A hemoglobin, hematocrit, and platelet count should be obtained to evaluate the need for blood product replacement. Coagulopathy laboratory tests including prothrombin time, partial thromboplastin time, and fibrinogen should be obtained as well. Hemorrhagic disease of the newborn is characterized by a prolonged prothrombin time, normal fibrinogen level, and normal platelet count. Unless another bleeding disorder is suspected, specific factor or coagulation assays are not necessary. Treatment with an appropriate dose of vitamin K, approximately 1 to 10 mg IV via slow push, should resolve coagulation concerns. 13 Unless associated with inadequate absorption of fat-soluble vitamins due to chronic disease, ongoing treatment should not be necessary.

TABLE 5. Stress-dose hydrocortisone dosing. 13

<table>
<thead>
<tr>
<th>Weight based (kg)</th>
<th>1 mg/kg per dose IV every 8 h</th>
</tr>
</thead>
<tbody>
<tr>
<td>BSA (m²)</td>
<td>20-30 mg/m² IV every 8 to 12 h</td>
</tr>
<tr>
<td>BSA calculation</td>
<td>( \frac{(0.05 \times \text{kg}) + 0.05}{\text{BSA}} )</td>
</tr>
</tbody>
</table>

BSA indicates body surface area.

The critically ill neonate presents a challenge to caregivers and should be managed rapidly and methodically. Although this review discusses the diagnosis and initial management of these life-threatening neonatal conditions, primary focus should be placed on ensuring the neonate’s stability. Ensuring a patent airway, adequate ventilation and oxygenation, and sufficient circulatory perfusion must take priority over diagnostic measures. Once stabilized and initial evaluation and treatment have been completed, these infants should be admitted for ongoing inpatient management.

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REFERENCES

Interhospital Transport of the Neonatal Patient

Kenny D. Kronforst, MD, MPH

The regionalization of perinatal care, a strategy involving the coordination of obstetric and neonatal services within a designated network of perinatal centers, has led to major improvements in maternal and neonatal outcomes. Since the 1960s, there have been substantial declines in neonatal morbidity and mortality, largely due to effective antepartum risk assessments leading to increases in the number of in utero transfers to tertiary centers for delivery and prompt access to the appropriate level of intensive care. This continues to be the practice model today, which, coupled to rapid advances in neonatal care and technology, has allowed for the effective management of critically ill newborns.

The reality is that despite increases in neonatal survival resulting from antenatal transfer and delivery at tertiary care centers, not all high-risk fetuses have the benefit of timely identification. This results in a number of infants who are delivered at primary or secondary community settings and require transfer for higher level of care. In this setting, reported outcomes are not as positive. For example, when looking at antenatal versus postnatal transfers to tertiary care centers, study found a significantly increased need for neonatal intensive care services and total length of stay for newborns transported postnatally. They also reported significantly higher incidence of bronchopulmonary dysplasia, intraventricular hemorrhage, higher staged intraventricular hemorrhage, patent ductus arteriosus, and mortality among those infants. This significance held true when comparing the postnatally transferred group to a similar cohort of inborn babies at tertiary centers. In fact, postnatally transferred infants were twice as likely as inborn admissions to have death or major disability, particularly in the very low birth weight category.

There are several reasons why transferred infants tend to do worse with regard to neonatal outcomes. Studies have shown that negative outcomes from transfers originating at referring...
institutions likely result from equipment failure, unavailability of monitoring devices, iatrogenic trauma, and temperature instability. The degree to which the clinical staff is prepared for high-impact, low-frequency events also influences these outcomes as deterioration of skills and knowledge gaps exist even with Neonatal Resuscitation Program–trained personnel. Because of these potential deficiencies, the transfer process of high-risk infants has to include specialized support consisting of appropriately trained teams with the capacity to provide goal-directed therapies, critical care interventions, and continuous monitoring.

The American Academy of Pediatrics' Section of Transport Medicine, in keeping with efforts to provide the best care to transported patients, has made several recommendations for conducting safe and effective transport of infants to tertiary hospitals for advanced neonatal care. In particular, they support the mobilization of specialized pediatric teams over nonspecialized teams due to their dedicated expertise and ability to treat infants in case of clinical deterioration. In other words, these teams can jointly manage patients with providers at referring institutions as needed, ensuring optimal patient safety even while stabilizing for transport. In fact, studies have consistently shown that specialized teams, irrespective of composition, have overall improved outcomes. One study found a significant difference with regard to morbidity, mortality, and hospital length of stay, and specifically noted decreased hypothermia, hypotension, and acidosis in premature infants when transported by trained staff. Another study found that unplanned events during transport, such as unintended extubations, and 28-day mortality were also significantly improved when the transport team had specialized training. Thus, the participation of skilled pediatric personnel is a key element of quality care during transport, with immediate and long-term impact.

Transport of newborns, even with an ideal team, is still subject to substantial practice variation in both transport operations and clinical care. This not only influences neonatal outcomes but also curtails any type of quality initiatives to improve transport medicine. In an effort to establish best practices, Schwartz et al. endeavored to develop quality metrics for neonatal transport, and through a Delphi process, came up with 12 core measures from which to benchmark performance and drive change. Most measures focused on patient safety, particularly as it pertained to issues of airway stabilization and management of respiratory conditions, adverse events including medical errors and equipment failures, unintended hypothermia, and patient handoffs. Details regarding each measure can be accessed elsewhere. The remainder of this manuscript aims to inform on key concepts of neonatal pretransfer stabilization and transport care with the intent to standardize care and improve clinical outcomes for infants born in the community. It focuses on patient symptoms and clinical practice guidelines (where applicable) for management of sick infants until arrival at a receiving tertiary institution.

RESPIRATORY CONDITIONS AND THEIR MANAGEMENT DURING TRANSPORT

Respiratory distress ranks among the top reasons for neonatal transport requests and requires advanced knowledge of airway maintenance and management. As such, the transport team must be prepared to play a critical role in patient stabilization, particularly as an infant's condition deteriorates beyond the referring hospital's resource capabilities. This joint management requires that transport personnel perform a rapid assessment of an infant's condition upon arrival, review response to already attempted therapies, and intervene with either noninvasive techniques or invasive ventilatory support as needed. Most importantly, it requires a clear understanding of the severity of illness to be able to anticipate the patient's clinical trajectory and needs once the infant arrives in the neonatal intensive care unit (NICU). To that end, close attention must be paid to rapidly changing infant parameters such as vital signs, work of breathing, inspired oxygen requirements, pulse oximeter readings, and acid-base status.

There are 3 common respiratory conditions that are typically encountered in transport. Prompt identification and targeted therapy can result in decreased infant morbidity and mortality, even before receiving specialized NICU care. A short review of respiratory distress syndrome (RDS), persistent pulmonary hypertension of the newborn (PPHN), and pneumothorax will be presented to elucidate the extent of transport capabilities needed in patient management.

Respiratory distress syndrome is a condition affecting mostly premature babies secondary to surfactant deficiency. Surfactant is a complex protein that provides an air/water interface within an alveolus, or breathing unit, leading to reduced surface tension and effective expansion of the air space during a breathing cycle. Deficiency of this protein results in a complex disease that is...
characterized by hypoxia and impaired gas exchange. It affects about 1% of newborn infants and is the leading cause of death for babies less than 34 weeks’ gestation. This translates to about 10% of affected premature infants in the United States per year.

The incidence of surfactant deficiency is inversely proportional to gestational age and birth weight. Although the rates of RDS remain high due to this relationship, there is now decreased infant mortality driven by routine surfactant replacement therapy. This has important implications for preterm infants born at nontertiary centers as delay in receiving surfactant has been linked to worse outcomes and bronchopulmonary dysplasia. Although there is currently no national consensus on surfactant administration before interfacility transport, the combination of an acceptable safety profile and low incidence of pneumothorax makes it a reasonable therapy to provide pretransport. Interestingly, the participation of a highly skilled transport team in both the intubation and administration process increases the likelihood that an infant will receive surfactant in a community setting. This, together with the judicious use of oxygen and ventilatory support, remains the staple of RDS therapy in transport.

A second commonly encountered respiratory condition in the nontertiary setting is PPHN. Persistent pulmonary hypertension of the newborn results from failure of normal cardiopulmonary transitions where blood flow is diverted systemically secondary to increased pulmonary vascular resistance. Infants that are affected tend to be born at term and present in hypoxic respiratory failure. This condition affects approximately 2 per 1000 live born infants and may complicate as many as 10% of all neonates in florid respiratory distress. Approximately 25% of infants with moderate to severe PPHN go on to have considerable neurodevelopmental delays by 24 months of age. This significant morbidity is matched only by a mortality rate that, even with prompt and appropriate therapy, remains between 5 and 10%.

The etiology of PPHN can be classified into 3 main categories, including normal vasculature/abnormal lung parenchyma, abnormal vasculature/normal lung parenchyma, or hypoplastic vasculature from abnormally developed lungs. These categories represent several disease states that often require transport from the community to tertiary care facilities for advanced medical management. A frequently encountered clinical scenario is meconium aspiration syndrome, a condition where increased pulmonary pressures result from parenchymal lung disease and airway obstruction. This typically presents with extrapulmonary shunting of blood leading to severely compromised oxygenation and hemodynamics, and usually requires prompt evaluation for extracorporeal membrane oxygenation (ECMO), among other stabilizing measures.

The therapeutic approach to PPHN, which can begin in a nontertiary hospital and continue during transport, is aimed at maintaining adequate cardiac output through the use of inotropic support. This ensures adequate oxygen delivery to underperfused tissues and helps correct metabolic acidosis. Additional mainstays of medical management include correction of electrolyte/glucose abnormalities and maintenance of normal body temperature. Endotracheal intubation and use of appropriate ventilator strategies to maintain functional residual capacity must also be considered. Inhaled nitric oxide, now the standard of care for PPHN, is particularly desirable because it promotes selective pulmonary vasodilation with minimal systemic effects. Although it frequently precludes the need for ECMO, it can prove ineffective in 30 to 40% of sick newborns. For those infants, rapid transfer by a transport team properly outfitted to provide inhaled nitric oxide therapy without interruptions, and possibly high-frequency jet ventilation, has been associated with improved outcomes.

The final respiratory condition to be discussed in terms of its potential for transport and need for specialized management is pneumothorax. A pneumothorax is a condition that occurs when pulmonary air leaks into a potential space between the parietal and the visceral pleuras. This can happen spontaneously or can be secondary to other respiratory diseases such as RDS, pneumonia, or meconium aspiration syndrome. It can manifest subtly without clinical compromise or can result in severe respiratory failure with hemodynamic instability if it occurs under tension. Risk factors for pneumothorax include prematurity and need for mechanical ventilation. In addition, the use of positive pressure ventilation during a neonatal resuscitation has been associated with an increase risk.

The incidence of pneumothorax in infants is variable and depends largely on their gestational age, birth weight, and associated risk factors. For example, preterm infants that are mechanically ventilated have estimated rates anywhere between 6 and 33%. This range has narrowed slightly since the implementation of surfactant therapy and switch from pressure to volume-mode ventilation. However, there is still uncertainty as to the actual number affecting all infants. A more useful estimate of incidence, then, comes from a 1930s study that
reviewed chest films of 700 consecutive newborns and determined the rate to be between 1 and 2% of all live births.\textsuperscript{24} All the same, this translates into significant morbidity and mortality, particularly at the lower gestational ages.

Infants presenting with asymptomatic pneumothorax can be monitored without specific treatment and are good candidates to stay in their community nurseries. Those with respiratory distress often require needle decompression or thoracostomy tube placement for effective evacuation of air, procedures that are typically managed in the NICU and require transport. Air will usually be evacuated before transport occurs by either the local pediatric team or the transport team to observe improvement in respiratory distress and monitor for hemodynamic stability. The transport team will then continue to observe the infant for status changes until arrival in the NICU.

In addition to providing support and co-management for respiratory conditions, transport teams must be prepared to retrieve critically ill infants with possible cyanotic heart disease from community settings. This situation should be suspected if an infant remains hypoxic despite maximal oxygen therapy and assisted ventilation.\textsuperscript{25} These infants can severely decompensate without appropriate and timely interventions. As such, they require emergent stabilization and rapid transport to facilities with cardiac and neonatal intensive care units for further diagnostic and operative consideration.

Management for a suspected cardiac lesion in the community setting should be limited to correction of treatable contributing factors because delay in transport can significantly impact an infant’s outcomes.\textsuperscript{25,26} Priorities for pretransport stabilization should include airway management, vascular access, oxygen therapy, volume repletion, electrolyte correction, and inotropic support. Administration of prostaglandin E\textsubscript{1} should also be considered to maintain the patency of the ductus arteriosus and enable mixing of blood for systemic circulation. This, however, should only be done after appropriate consultation with a specialist for dose and effect.

**ADVERSE EVENTS DURING TRANSPORT**

Limited resources and equipment, staff with variable knowledge base, critically ill patients, involvement of multiple care teams, and pressure for time-critical interventions can result in opportunities for unintended harm during transport.\textsuperscript{27,28} In fact, a study by Barry et al\textsuperscript{27} found that up to 75% of children transferred had adverse events attributable to an unreliable process, though these values reflected transport by nonspecialized teams. Nevertheless, compromise to patient safety can occur throughout the continuum of patient care, but in transported infants tends to cluster around specific time points, particularly during pretransport stabilization, during transport itself, and at patient handoff.\textsuperscript{29} The most common errors include lack of appropriate cardiorespiratory support and equipment malfunction, potentially resulting in life-threatening events such as hypotension and respiratory failure. Other frequently reported events include medication errors, procedure errors, and hypothermia.\textsuperscript{28}

According to Karlsen et al,\textsuperscript{30} more than 65,000 infants per year are transported from their birth hospital to a NICU for specialty care. Despite the potential for adverse events during each of these transports, most are typically achieved in a safe and timely manner. Nevertheless, it is well established that when errors occur, most are thought to be secondary to avoidable human factors. Interestingly, Lim et al\textsuperscript{29} described that about one third of the adverse events reported in their study occurred during the preparation and stabilization of patients at the referring institution, before the arrival of any specialized transport team. Thus, the transport team provides a unique service for sick children and families in the community and bridges the care offered on-site to that provided in the receiving care units. Through this support, the transport team is able to bring education, resources, and quality initiatives to partnered referral centers to assist in the overall reduction of adverse events.\textsuperscript{29,31}

Another way to bring about improvement in the quality of the transport process is through the use of specialized assessment tools. In particular, the transport risk index of physiologic stability (TRIPS) is a risk-weighted transport score that is highly predictive of infant morbidity and mortality in the NICU when measured at a single time point.\textsuperscript{32,33} The transport risk index of physiologic stability is a composite score derived of several physiologic variables including temperature, respiratory status, blood pressure, and response to noxious stimuli. The score, when obtained at 2 time points, can also be used to elucidate a change in patient status and the potential reasons for that change. With regard to the transport process, a high pretransport TRIPS score will reflect deficiencies during initial patient stabilization, whereas a high postransport TRIPS score will reveal information regarding transport care itself. Review of these 2 scores can aid in identifying opportunities to optimize care through resuscitation training, improved communication, better equipment/personnel allocation, and overall
support. This scoring system, however, is currently not universally used and requires ongoing performance analysis to be the designated as a worthwhile transport tool.

An undisputed critical tool of care that is sparsely described in the literature is the handoff of patient information during the interhospital transfer of critically ill neonates. From the adult literature, it is known that patients who are transported into tertiary care facilities, when compared to those admitted directly through the emergency department, seem to have a significantly higher inpatient mortality. Furthermore, these patients also share a higher incidence of avoidable morbidities including longer average length of stay, higher proportion of intensive care, higher hospital costs, and lower rates of discharge home. Interestingly, poor communication during transitions of care has been identified as the key reason for this difference.

To address the issue of poor handoff communication, the Joint Commission set a National Safety Patient Goal that requires all health care providers to implement a standardized approach to patient handoff. Although there are several programs available to template these verbal exchanges, it is up to each institution or program to adopt and develop a system that works for their patient population. To date, there is no single handoff protocol that is used reliably in neonatal transports. Nevertheless, there is consensus that effective information sharing needs to be integrated into practice for adverse outcomes to be meaningfully reduced. Work is in progress to look at process mapping, checklists, and technology integration to facilitate meeting this Joint Commission goal.

**TEMPERATURE REGULATION IN TRANSPORT**

The importance of maintaining a thermoneutral environment for newborns is well established. In fact, temperature regulation is a highly prioritized element of newborn care, and currently, a recognized quality marker for measuring infant stability. This is most relevant as smaller and more preterm infants survive through NICU admission and have a limited capacity for achieving temperature ranges between 36.5°C and 37.4°C. It is also a recognized challenge for transported infants, whose most frequent diagnoses include prematurity and respiratory distress, and are at high risk for developing hypothermia.

Measures to counter heat loss during initial infant stabilization are critical to avoid injury from cold stress. Perhaps the greatest impact in NICU care has come from the use of heated humidified air during infant resuscitation and transport. In fact, this practice alone has significantly decreased the chance of hypothermia at NICU admission, a condition frequently associated with early neonatal death. Other interventions with similar influence include maintaining ambient temperatures greater than 25°C in the delivery room, covering high-risk newborns in polythene wraps or plastic bags immediately after birth, and the consistent use of radiant heat and/or servo-controlled isolettes.

Hyperthermia in newborns and transported infants is less frequently encountered, but has equal negative impact if not properly recognized and treated. This most often occurs in term or late preterm infants and can be associated with infection (less frequently) or iatrogenic oversight. Transport incubators, even in servo-control modes, will require frequent temperature checks, reduction of ambient temperature, and adjustment to infant bundling to maintain core temperature within the acceptable range. Thus, a standard approach to infant incubator monitoring and management is necessary to prevent iatrogenic harm and provide optimal care.

A condition that requires strict temperature regulation is that of hypoxic ischemic encephalopathy. This is a pathologic state that typically affects term and late preterm infants who present with a history of profound perinatal distress. A significant number of affected infants are born in community settings, and overall, represent about 1 in 1000 newborns. These infants generally require prompt evaluation and are often candidates for therapeutic hypothermia after transport to specialized tertiary centers. Timing of therapy is critical for optimal neuroprotection, and delays beyond 6 hours of life have been associated with serious injury and secondary cerebral energy failure leading to profound neurodevelopmental delays. Thus, pretransport and transport care must be well coordinated to address this critical window of time and lessen the neurologic burden of untreated moderate to severe encephalopathy. As therapeutic hypothermia becomes the standard of care for infants with encephalopathy, nurseries and transport teams will have to develop protocols for optimal management and resolve the issue of passive versus active cooling before arrival to a designated hypothermia center. Of note, consistent cooling to target temperatures between 33°C and 35°C has yet to be documented in these situations.

Furthermore, the extent of injury from overcooling remains to be explored.
FAMILY INVOLVEMENT, BACK TRANSFERS, AND ONGOING COMMUNITY RELATIONS

Current clinical practice models attempt to integrate family-centered care for all hospitalized pediatric patients. This is certainly the case for inpatient newborns and should extend to infants who require transport to tertiary centers for specialty management. An impending separation from an infant, particularly one who is sick, is a highly stressful and often unpredictable event. In these instances, the practice of family-centered care may contribute to decreased parental anxiety. However, this practice has not been standardized for infants undergoing transport, largely due to the constraints of time imposed by the process itself. Nevertheless, it is well established that parental partnership initiated by the transport team is a key driver for ongoing engagement in future infant care and contributes to improved infant outcomes. As such, it should be incorporated into the transport process and prioritized as a primary goal of transport care.

Arrival and patient handoff to the receiving unit does not finalize the involvement of the transport team in patient care. Often, they are charged with providing clinical updates to the referring institutions and, as such, remain involved in the management of their transported patients. In addition, teams may be involved in the back transfer of infants who are convalescing and no longer require tertiary care.

The concept for back transfers is grounded on the principles of cost-effectiveness, bed utilization, and parental satisfaction. In a regionalized system of care, this should be the expected outcome for all infants who still require some degree of clinical support but are expected to have a stable clinical course until discharge. In fact, it is economically justified if the projected remaining length of stay at the lower level nursery covers the expense of transport back to the community hospital. This not only improves community relations by allowing referring hospitals to offer their own services but also gives families proximity and increased flexibility in caring for their hospitalized infants.

SUMMARY

Overall, the transport of critically ill infants to centers that can offer appropriate intensive care has resulted in improved neonatal outcomes. The fact that this can happen within a regionalized system of care has further facilitated the movement of patients and supported community relationships. Standardization of care continues to be a priority with new quality initiatives aimed at improving patient safety. Training and education for improved pretransport stabilization as part of a greater transport process have also been prioritized. Development of practice guidelines regarding the management of common diseases has also been undertaken, though this also represents a small step toward standardization of care. Finally, family engagement through family-centered care and timely retrotransfer of appropriate infants to home hospitals remain important goals of transport care.

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Abstract:
The death of a neonate is devastating for all involved. Each year, critically ill neonates present to emergency departments across the United States. These infants require acute medical interventions with a goal of stabilization. Despite these efforts, hundreds of infants die every year in emergency departments across the United States. Emergency care providers, unaccustomed to providing neonatal end-of-life care, may feel unsure about how to best care for families during resuscitative measures and after neonates die. There is literature to suggest that increased knowledge and advance preparation can calm fears of providers caring for patients in such tragic situations. We aim to provide in this article a broad overview of a variety of topics related to neonatal death and bereavement care.

Keywords:
neonatal death; neonatal bereavement; palliative care; emergency department

Neonatal Death in the Emergency Department: When End-of-Life Care Is Needed at the Beginning of Life

Jessica T. Fry, MD, Natalia Henner, MD

The death of a neonate, no matter in what setting or circumstance, is devastating for all involved. The birth of a child is rightly expected to be a joyous event. Neonates and their families who instead require emergency treatment demand immediate expert care from medical teams in several forms: quick diagnostic assessment, procedural skills, clear communication, and intensive psychosocial support. Balancing these tasks is difficult but critically important for involved patients, families, and staff.

For the purpose of this article, we will focus on emergency care providers’ experiences immediately after the death of a neonate. We will present definitions of perinatal death, describe death of a neonate after resuscitation, discuss the role of palliative care, and explore the needs of families after neonatal death. Much has been written about providers’ experiences after the death of a child in the emergency department (ED). Many of these concepts apply to the neonatal patient and will be reviewed. However, neonatal loss can be viewed by providers and families as different from the loss of an older child. We will try to address differences in what families may need from providers and what providers may feel.
We hope that, by highlighting unique aspects of neonatal end-of-life care, we can improve awareness, competence, and skill of emergency providers in this area.

**DEMOGRAPHICS OF NEONATAL DEATH**

Dying patients present to EDs during various stages of the perinatal period—from a fetus with evidence of in utero distress to a neonate decompensating at home. Infant mortality, including perinatal mortality, remains a significant measure of health in modern society. It is important to understand the terminology of these deaths to accurately report them and gather information on vital statistics. Table 1 lists commonly used definitions related to birth and death as adapted from the American Academy of Pediatrics Committee on Fetus and Newborn.

Most neonatal deaths happen in inpatient units, such as neonatal or pediatric intensive care units. Between 1999 and 2013, 268,225 neonatal deaths were reported in the United States. A total of 249,871 (93%) of these occurred in inpatient medical facilities. Of the remaining, 9842 (3.7%) were recorded as either occurring in an outpatient medical facility/ED or presenting dead on arrival to a facility. These statistics indicate that emergency care providers irregularly attend to dying neonates and their families. Because the vast majority of neonates die in inpatient units, it is likely that inpatient staff have more experience in this area. Emergency providers may appreciate this difference and feel ill prepared to care for dying neonates. Advance preparation, including acquiring knowledge of various issues surrounding neonatal death, may help to alleviate discomfort and improve care.

**HOW AND WHY NEONATES DIE**

There are 3 mechanisms through which infants who will die arrive in EDs. First, these infants may be born in EDs. This could include pregnancies affected by stillbirth as well as infants who are born alive with conditions causing physiologic instability. Examples of such conditions include prematurity, congenital anomalies, and birth asphyxia. Next, infants may be born outside of hospitals and present to EDs for acute care. This accounts for precipitous births outside of the hospital setting as well as planned out-of-hospital deliveries. It merits mention that out-of-hospital birth has been identified as an independent risk factor for neonatal mortality. Finally, neonates who were born in hospitals and discharged home may subsequently become acutely ill or injured and present for emergency care.

The causes of neonatal death are many and have changed over time with changing care practices. We offer that differential diagnoses for a particular neonate can be thought of as related to age after birth. Numerous studies have described how neonates die in the setting of neonatal intensive care. A parallel can be drawn between how neonatal intensive care unit infants die shortly after birth (immediately to hours) and conditions emergency care providers might encounter in an infant’s first moments of life. Although causes of early death are numerous, they generally belong to 1 or more of the following categories: complications of prematurity, congenital anomalies (genetic and/or structural), respiratory failure, asphyxia, shock/anemia, infection, or malignancy.

Causes of death for neonates who were discharged home after birth may be different than those described above. It is likely that causes of acute neonatal decompensation at home resemble the etiologies of postdischarge death for low–birth weight infants and the etiologies of sudden unexpected death in infancy. There may be some overlap, but understanding history and timing of neonatal decompensation can focus the set of possible diagnoses (Table 2).

One cause of both early and late neonatal mortality which deserves separate discussion is

**TABLE 1. Commonly used definitions of birth and death in the perinatal period.**

<table>
<thead>
<tr>
<th>Definition</th>
<th>Description</th>
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</thead>
<tbody>
<tr>
<td><strong>Live birth</strong></td>
<td>Complete expulsion or extraction from the mother of a product of human conception, irrespective of the duration of pregnancy, which, after such expulsion or extraction, breathes or shows any other signs of life such as beating of the heart, pulsation of the umbilical cord, or definite movement of voluntary muscles, regardless of whether umbilical cord has been cut or placenta is attached.</td>
</tr>
<tr>
<td><strong>Fetal death</strong></td>
<td>Death before complete expulsion or extraction from the mother of a product of human conception, which is not an induced termination of pregnancy. The death is indicated by the fact that, after such expulsion or extraction, the fetus does not breathe or show any other sign of life such as beating of the heart, pulsation of the umbilical cord, or definite movement of voluntary muscles.</td>
</tr>
<tr>
<td><strong>Infant death</strong></td>
<td>Live birth that results in death within the first year.</td>
</tr>
<tr>
<td><strong>Neonatal death</strong></td>
<td>Live birth that results in death within the first 28 d.</td>
</tr>
</tbody>
</table>

Data from Ahrens and Hart.
injury due to abuse or neglect. Situations where abuse is suspected are inevitably challenging and emotionally charged. The primary focus must remain on the critically ill neonate. However, emergency providers should follow every policy and procedure established to investigate concerns after resuscitative measures are concluded (including involvement of specialist child abuse teams, child protective services, and law enforcement as applicable). Providers should note that a number of conditions related to congenital disorders or birth trauma may have features that appear consistent with abuse.

TABLE 2. Possible etiologies of acute neonatal decompensation.

<table>
<thead>
<tr>
<th>Condition</th>
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<tbody>
<tr>
<td>Asphyxia/Suffocation</td>
</tr>
<tr>
<td>Choking/foreign body</td>
</tr>
<tr>
<td>Smothering</td>
</tr>
<tr>
<td>Wedging/entrapment</td>
</tr>
<tr>
<td>Cardiac disease</td>
</tr>
<tr>
<td>An rhythms</td>
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<tr>
<td>Cardiac tumors</td>
</tr>
<tr>
<td>Cardiomyopathy</td>
</tr>
<tr>
<td>Congenital heart disease</td>
</tr>
<tr>
<td>Coronary aneuysms</td>
</tr>
<tr>
<td>Dehydration</td>
</tr>
<tr>
<td>Drowning</td>
</tr>
<tr>
<td>Electrocuion</td>
</tr>
<tr>
<td>Hypothermia/hyperthermia</td>
</tr>
<tr>
<td>Infection</td>
</tr>
<tr>
<td>Encephalitis/meningitis</td>
</tr>
<tr>
<td>Necrotizing entercolitis</td>
</tr>
<tr>
<td>Pneumonia</td>
</tr>
<tr>
<td>Sepsis</td>
</tr>
<tr>
<td>Inborn disorders of metabolism</td>
</tr>
<tr>
<td>Intracranial hemorrhage</td>
</tr>
<tr>
<td>Arnold-Chiari malformation</td>
</tr>
<tr>
<td>Malrotation with midgut volvulus</td>
</tr>
<tr>
<td>Poisoning, accidental or intentional</td>
</tr>
<tr>
<td>Carbon monoxide poisoning</td>
</tr>
<tr>
<td>Water intoxication with hyponatremia</td>
</tr>
<tr>
<td>Respiratory failure</td>
</tr>
<tr>
<td>Undiagnosed congenital abnormalities of the respiratory system</td>
</tr>
<tr>
<td>Bronchopulmonary dysplasia</td>
</tr>
<tr>
<td>Seizures</td>
</tr>
<tr>
<td>Trauma, incidental or intentional</td>
</tr>
</tbody>
</table>

This includes causes identified as significant in post-discharge mortality of low-birth weight infants as well as in sudden unexpected death in infancy. Data from Kugelman et al and Carroll and Wood.

Regardless of the timing or underlying etiology, emergency providers who do not regularly work with dying neonates may struggle with these tragedies. For the remainder of this article, we seek to provide practical considerations surrounding the care of dying neonates, their bereaved families, and medical teams. For reference, we consolidated these recommendations into Table 3. We wish to highlight that these are some of many possible suggestions. Neonatal bereavement is a field in which there are little evidence-based data to support or refute the effectiveness of interventions in all families. Practices which some families find healing may be hurtful to others. Care should be uniquely tailored based on local practices, resources, and needs of individual families.

THE END OF A NEONATAL CODE

Once a critically ill neonate presents for emergency care, there are 2 ways through which death can occur. First, infants may die in the context of an active code. Death despite active resuscitation is due to physiologic decompensation regardless of advanced supportive measures, including intubation/ventilation and cardiopulmonary resuscitation. Emergency providers who have limited experience running a code using Neonatal Resuscitation Program and/or Pediatric Advanced Life Support algorithms may feel unsure about stopping treatment for neonates without clinical response. The patient’s clinical history and the causes of death mentioned above should be kept in mind when choosing between potential interventions. Consultation with specialized neonatal providers, as available, should be strongly considered if there is uncertainty whether therapies could change outcome.

There has been ongoing development in the area of patient and family-centered care in pediatrics, including pediatric emergency medicine. As a result, it is standard of care for many families to be present throughout active resuscitation of children. A large body of literature has ascribed many positive benefits to this practice, even in the case of dying children. These include improved parental ability to cope with loss and decreased incidence of posttraumatic stress disorder among bereaved families. Parental presence during resuscitation has also been found to be beneficial in early neonatal resuscitation. Having parents at the bedside during death may seem daunting but can be crucial...
TABLE 3. Suggested considerations for neonatal end-of-life care.

During resuscitative efforts
- Offer the family the option of being present during resuscitative efforts
- Clearly communicate assessment of the neonate’s condition and expected prognosis
- Consider designation of one member of the team (possibly social work or chaplain) to be the “point person” for family communication throughout resuscitation and after death
- If the neonate’s mother is absent from the bedside due to her own medical needs, ensure that her needs for communication can be met
- If the providers are not fluent in the parents’ primary language, interpreter services should be used

When a neonate has an advanced directive
- Follow, as applicable, orders provided by a POLST form
- Understand that families may change their minds regarding advanced directives and respect changes when they occur
- Contact home hospice agency to help understand neonate’s expected symptoms and prognosis

Communication with families after death
- Speak concisely about death; avoid euphemism
- Expect strong grief reactions in families, even manifesting as physical symptoms
- Respect silence in families; pace information as appropriate for the cues of the family
- Understand that here may be differences in how mothers and fathers hear, process, and react to information
- Avoid communication biases in cases of suspected nonaccidental trauma or regarding unconventional parenting choices
- Provide validation of grief, regardless of infant’s age or condition
- Provide clear information regarding autopsy, postmortem testing, and plans for later follow-up

Communicating with other providers after death
- Offer to contact other medical providers who have cared for the neonate and his or family (may include obstetricians, pediatricians, mental health providers, home hospice agencies, and other subspecialists)
- Contact providers in a timely manner to ensure that the family’s “medical home” is aware of their loss and can provide additional support
- Follow procedure of local OPO, and ensure that family is informed of any OPO follow-up that may occur

Bereavement care for families
- Facilitate, however possible, the process of parents parenting their neonate, including time and space to say goodbye.
  ° Clean/remove items which may be visually disturbing to families
  ° Clean the neonate’s body; remove extraneous medical equipment as allowed by medical examiner’s policies
  ° Consider use of hats and blankets
- Allow families time to be with their child’s body after death as desired
- Consider whether the family has particular religious or personal beliefs which they may wish accommodated after death; allow flexibility in routine to honor these as possible
- Reach out to Labor and Delivery or a NICU which more commonly experiences stillbirth/neonatal death to access bereavement resources. These may include:
  ° Handprint/footprint cards
  ° Molds of hands and/or feet
  ° Means to hold keepsakes (memory boxes), such as a hat, blanket, or lock of the neonate’s hair
  ° Bereavement photography, whether with a hospital camera or by a professional
  ° Services of a Child Life specialist to support bereaved siblings
  ° Access to community resources for ongoing bereavement support and/or counseling
  ° Ensuring that mother has access to health resources for her own medical and mental health follow-up
  ° Resources for lactating mothers regarding cessation of lactation and/or breast milk donation
- Provide families with contact information of medical team after they leave the hospital (written materials preferred)

Bereavement care for staff
- Appreciate challenges of caring for a dying neonate; debrief as a team and individually if necessary
- Seek assistance with debriefing from palliative care or ethics teams in especially challenging cases
- Encourage staff to seek bereavement support

NIC indicates neonatal intensive care unit. OPO, organ procurement organizations; POLST, practitioner orders for life-sustaining treatment.

for parents at the moment and beyond. Not all families will wish to remain present. Some parents feel overwhelmed, and some mothers immediately postpartum require urgent medical care themselves. To ensure ongoing family support, one staff member should be designated as family liaison during resuscitation. In addition to medical care providers, this may be a role for a social worker or chaplain. In this way, family needs for communication and decision making (at the bedside or from a distance) can be continually assessed and honored.

NEONATAL PALLIATIVE CARE

A second mechanism through which neonatal death occurs is through withholding or withdrawing of medical therapies. There has been increasing recognition of the application of palliative care principles to neonates with life-limiting conditions. Perinatal palliative care providers assist parents of babies with life-limiting conditions in various ways: delineating goals of care during perinatal and neonatal periods, ensuring collaborative approaches between providers, supporting families after delivery, assisting with neonatal...
symptom management, and aiding transition to home hospice. Common diagnoses for perinatal palliative care referral are summarized in Table 4. Many of these conditions were previously termed lethal, but there is growing awareness that some affected neonates survive beyond minutes or hours, with or without invasive interventions.26–28

Many families view their neonate’s surviving longer than predicted as a gift29–31 and rely on medical care to ensure comfort in life and death. Families may choose from a range of medical options to balance extending meaningful time in the lives of their children with ensuring comfort.28,30,32 Interventions which may improve quality of life despite seeming invasive include delivery of oxygen via nasal cannula, provision of nutrition via nasogastric or gastrostomy tube, and drainage of hydrocephalus via ventriculoperitoneal shunt. The presence or absence of medical technology should not imply a certain treatment plan. Providers should explore families’ goals and understanding of disease trajectory when discussing ongoing therapies.

Families caring for dying neonates at home face unimaginably difficult tasks. Pediatric palliative care and hospice agencies provide broad supportive services, including nursing visits, crisis care, social work assistance, and spiritual care. For most families, home hospice nurses become primary providers. Nurses communicate closely with hospice medical directors, referring physicians, and consultants (if applicable). Although there are a growing number of pediatric hospice agencies in the United States, there remain geographic gaps in care provision.33 Pediatricians or adult hospice agencies may also provide primary care for neonates at the end of life.

Although many families working with hospice develop treatment plans for home care, some will seek medical attention in the ED. There are various reasons for these visits, all potentially challenging for staff. Infants may experience distressing symptoms that are difficult to control at home such as pain, respiratory distress, or seizures. Parents may panic when death is imminent and wish to move to the hospital. Families may change plans regarding end-of-life treatment and seek alternate paths. Regardless of reason, all parents hope for reassurance and compassion during these dark moments. Emergency department staff can partner with families by gaining insight into their understanding of disease trajectories and treatment goals. It may be helpful to coordinate with primary care providers and consider consultation with experienced neonatal teams to treat symptoms and plan for ongoing care.

Most families complete a Practitioner Orders for Life-Sustaining Treatment (POLST) form when enrolling with palliative care/hospice agencies. The POLST forms serve as medical orders, similar to inpatient Do Not Attempt Resuscitation/Allow Natural Death order sets, which enable families to express preferences for life-sustaining treatments across different care locations. These forms are available online in English and Spanish, and most states have adopted POLST programs.34 If a family seeking emergency neonatal care has a completed POLST form, the orders should be followed and used to facilitate conversation about resuscitation status and medical therapies. Of note, parents do not have to disclose POLST forms and/or can ask for POLST orders to be altered. In this case, a comprehensive conversation about treatment expectations should occur.

### Table 4. Conditions with which infants may qualify for perinatal palliative care.

| 1. Newborns at the threshold of viability |
| 2. Newborns or fetuses with complex or multiple congenital anomalies incompatible with prolonged survival |
| a. Trisomy 18 and 13, thanatophoric dwarfism, or lethal forms of osteogenesis imperfecta |
| b. Anencephaly, holoprosencephaly, congenital severe hydrocephalus, neurodegenerative diseases requiring ventilation (eg, spinal muscular atrophy) |
| c. Potter syndrome/renal agenesis, some cases of polycystic kidney disease |
| d. Acardia, some cases of inoperable heart anomalies |
| e. Some cases of giant omphalocele, severe congenital diaphragmatic hernia with hypoplastic lungs, inoperable conjoined twins |
| 3. Newborns with overwhelming illness not responding to intensive medical interventions |

Data from Leuthner22 and Catlin and Carter.28

A Diagnoses provided serve as examples; institutional considerations and individual discretion should apply.

**COMMUNICATION WITH FAMILIES AFTER DEATH**

The news of a neonate’s death elicits strong emotional reactions from families. Providers should recognize and acknowledge these emotions. Relay information concisely, and avoid euphemisms (eg, say “died” instead of “passed away”). Some families may be in shock and fall silent. It is important to honor silence and remain present until communication continues. Some parents have physical reactions to the news. Birth mothers may have
different reactions from fathers or partners, including feelings of guilt. It is important to acknowledge differences in grief and to tailor communication styles. There are several things to be discussed with families after a neonate dies, although the majority can be paced to suit family needs. Disclosure of neonatal death can be an emotional experience for providers as well. Many families appreciate sincere expressions of sadness from medical personnel.35

At this emotionally charged time, special attention must be paid to admitting biases and ensuring compassionate communication. It is difficult to express empathy and maintain connection when nonaccidental death is suspected. It may also be tough to avoid judgment when parents follow unconventional health practices such as vaccination avoidance, refusal of vitamin K injections, or home births. Many of these families feel disconnected from the medical system at baseline and feel even more disenfranchised after their neonate’s death.

There is a common misconception that neonatal death is somehow “less painful” than death of an older child. Evidence suggests that parental grief following the death of a neonate is similar in duration and intensity.36,37 Bereaved perinatal parents commonly experience deep social isolation. This is likely multifactorial and related to the unique moral status of neonates,38 biases about the value of their lives,39 and the relatively small “ripple effect” of their death on communities. It is important to acknowledge the tragedy parents experience regardless of infant diagnosis or life expectancy. Avoid misguided expressions of empathy such as “you can have another child,” as parents do not find such statements comforting.40 Even when death was anticipated, avoid the assumption that it brings relief.

After the death of a neonate, there are a number of ways to demonstrate commitment to family well-being.41 Staff can offer to call family members or fulfill transportation needs. It could be helpful to explain postmortem nursing care and to invite family participation. Special attention should be paid to validating parents’ need to parent even after their child’s death.

**COMMUNICATION WITH OTHER PROVIDERS**

Neonatal patients often have primary providers who will want to extend support to their families. Staff should notify obstetricians, pediatricians, and subspecialists in a timely manner.35 A clear follow-up plan should be established, especially if autopsy information will be reviewed. It is particularly important to inform the mother’s obstetrician before postpartum visits. If not previously contacted, staff should also discuss a neonate’s death with the involved perinatal hospice team.

**POSTMORTEM EXAMINATION**

In the case of sudden neonatal death, parents often express a need to know why death occurred. Sometimes, this is evident upon reviewing history, physical examination, laboratory workup, or imaging. At other times, diagnosis may remain elusive even after extensive postmortem studies. Providers should explain the process of diagnostic evaluation, including placental pathology and autopsy. Many cases of sudden death will require referral to a coroner/medical examiner’s office. Even if autopsy is not mandated, it should be offered to all families. Data suggest that parents who choose against autopsy later express regret.42 Sensitive discussion of autopsy empowers parents to choose a postmortem examination if it meets their needs. Some families observe religious rituals surrounding the body after death. This can be difficult with autopsy but should be respected whenever possible. It is crucial to establish concrete plans for follow-up with an emergency or primary care provider and to ensure that the family knows how they will receive information on cause of death.40

**THE QUESTION OF ORGAN DONATION**

Some families of dying neonates inquire about organ donation. Individual states have unique organ procurement organizations (OPOs). Each OPO establishes their own criteria for acceptability of deceased organ donors. Emergency care providers should review OPO policies to determine criteria for organ donation as well as timing of mandatory notification surrounding neonatal death. Most neonates will not qualify for vital organ donation in the setting of sudden death, as they will not meet the strict criteria for donation after brain death or circulatory determination of death.43 However, donation of heart valves, corneas, and other tissue may be possible. OPOs, once notified, may contact parents if a neonate qualifies for donation. Providing anticipatory guidance regarding organ donation can reduce secondary trauma that families may feel if a goal of organ donation cannot be achieved or if they receive unexpected contact from an OPO.

**BEREAVEMENT CARE FOR FAMILIES**

Immediate and long-term bereavement care for families is extremely important following neonatal
death; this can start in the ED. Staff should appreciate the heightened state of parents and pay attention to verbal and nonverbal cues. Many parents share vivid memories of their child’s death and recall interactions with providers in great detail.

Most families want to bid farewell after death and appreciate support in this process. It is important not to assume parents would not want to see their baby, regardless of appearance. For some parents, spending time with their dead newborn is their only opportunity for bonding. Many parents share regret of not being offered the chance to say goodbye or declining to do so when offered. Rooms should be rid of visually disturbing items (invasive equipment, blood). Parents can be invited to participate in cleaning the body. Hats and blankets can be provided if desired. Parents should have ample time to say goodbye without rushing. Some parents may have requests regarding how bodies are transported to the morgue. Flexibility in hospital proceedings at this time can be helpful.

Emergency care providers can identify available bereavement resources with labor and delivery or nursery/neonatal units. Such resources include memory boxes for keepsakes (lock of hair, hat, toy, pacifier) and plaster molds for hands and feet (Figure 1). Bereavement photography services can be available through hospital contracts or national organizations. Families may desire photography even for extremely premature neonates or neonates with significant dysmorphology. If such services are unavailable, staff can use hospital cameras and arrange to deliver photographs to parents. Child Life resources can be invaluable for siblings at the time of death and beyond; some are available online. Social workers can help families find community bereavement counselors or share online resources. Perinatal nurses (obstetric and nursery) may provide strategies for cessation of lactation or breast milk donation.

Some families see their emergency providers as "last links" to their neonate’s life, especially if that life was brief. Emergency staff should recognize the significance of that bond. It is appropriate to make arrangements to meet with parents if requested. It is also fitting to consider phone calls or cards. Providers can seek support in bereavement outreach from social work. Attending memorial services at family invitation can further validate the relationship and may be helpful for both family and staff.

**DOCUMENTATION**

After the intensity of caring for a dying neonate, it is challenging to relive emotions through documentation of the encounter. However, entering accurate information into the medical record is crucial to the process. Studies show that medical documentation in both neonatal and emergency providers can be inconsistent, omitting important details. Although trials of documentation interventions have not demonstrated consistent results, use of standardized templates may help providers organize thoughts on the multiple elements of medical and psychosocial care.

**BEREAVEMENT CARE FOR STAFF**

Caring for dying neonates poses unique challenges for every staff member. It is important to recognize that some team members may be particularly hesitant to participate in end-of-life care (eg, pregnant women, recently bereaved providers). Team debriefings following death should focus on challenging and gratifying aspects of care. Debriefings may be especially important after death from nonaccidental trauma, and participation of a child protection team is encouraged. In cases of neonatal death due to withholding therapy for a "treatable" condition (extreme prematurity, complex heart lesions, or others), palliative care or ethics teams can be beneficial. Staff should be strongly encouraged to seek bereavement support for themselves as an important means of self-care.
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