Emergency Care of Children With Special Health Care Needs

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As one reflects on the timeline for pediatric technical achievements since the 1980s, tremendous advancements for various subspecialties have occurred. Most importantly, these have increased survival for medically complex children. It has been explained by numerous physicians that the technical advances in the survivability for these particular patients is purely a result of the advances by NASA, other governmental agencies, the private sector, and through the collaborative effort between some of these parties and pediatric health care providers. If one trained in pediatrics during the 1980s, premature neonates or children with complex medical disorders required the greatest resources, the longest hospital stays, and the most complex medical equipment to survive day-to-day in the hospital, let alone in the community. Their survival was purely dependent upon the ingenuity of health care providers and, most importantly, adapting adult technologies to these patients. If these children did survive to discharge, their daily needs were complex and required numerous medical devices. Their emergencies were not only physiologic but also a majority of time mechanical. As these technological advances were refined for various pediatric patients, their survivability was tied to complex machines, special nutrients, higher metabolic demands, and a highly complex balance for these patients. These patients often had long and complex hospital stays with various medical and physical requirements with secondary complications associated with their increased survival.

As the advent of numerous new treatment modalities for the premature infant appeared, the rate of chronic lung disease, intraventricular hemorrhage, and bouts of necrotizing enterocolitis, as compared with that seen during the 1980s, has dramatically decreased with improved survival for these patients. Although their survival has increased, their medical needs have taken on even greater challenges, especially to emergency care providers. In these particular patients, besides the
Routine emergencies that occur in a child's life, the frequency and complexity of their emergencies are unique to their particular chronic disease state and their delicate physiologic balance between healthy or sudden morbidity. Some of these patients have multiple complex physiologic system involvement, which can pertain to one particular emergency or, in conjunction with the other physiologic systems, may have a complex emergency. This medical knowledge is constantly changing, and dissemination of this knowledge base to the emergency health care provider is slow and infrequent.

The topic for emergencies in the special needs or high-tech pediatric patient is more of evidence-based practice knowledge dissemination to the emergency medical community vs a plethora of certain prospective well-designed research studies. This knowledge base regarding both common and unusual emergencies in these patients has never really been well defined nor has a particular journal been devoted to the dissemination of an evidence-based practice knowledge base. The articles in this issue of Clinical Pediatric Emergency Medicine will be devoted to the dissemination of information pertinent to the assessment and treatment of emergencies that occur in these children. Each article will take a primary organ system involvement approach.

The articles in this issue will also approach these patients from a continuum of emergency medical services perspective. Most will offer a format that begins with a typical clinical scenario for a medically complex child, much as might be encountered in day-to-day emergency care. The articles have been written to offer insight and educate first responder emergency medical services providers, who must capably assess, treat, and then transport these children, and likewise emergency department nurses and physicians who must be ready to appropriately assess and manage emergencies in these complex children. The care of these patients during their emergencies may not always be in the tertiary or quaternary children's hospital, but the occurrence is commonly located in the community emergency department or a walk-in clinic. Necessary emergency interventions must begin where these children initially present, not at the referral pediatric center. Therefore, the role of this issue is to enlighten, alleviate fears, and improve the care that the emergency care providers can deliver to these complex children until they are transported to their designated referral center. Timely and effective implementation of stabilization procedures for these children will greatly decrease their morbidity and mortality. The authors will also present some of the challenging emergent situations associated with newer therapeutic measures and devices that are unique to those particular special needs children.

In this issue, of particular note will be an article devoted to disaster management for special needs or high-tech children. This is a new topic, one that has dramatically challenged current disaster management ideology. In the field of disaster management, these particular patients are often not considered, or they are ignored due to lack of knowledge or possibly due to the absence of pediatric experts in disaster planning. The authors for the disaster management article discuss the management of special needs children using a mass casualty incident scenario, which offers an organ system and technology support perspective in the prioritization of care and with emphasis on parental involvement, equipment issues, and overall care for these particular patients.

This issue also includes two Emergi-Quiz case presentations from the American Academy of Pediatrics Section on Emergency Medicine Scientific Session held at the October 2011 National Convention and Exhibition. These ‘clinical puzzlers’ should prove to be most enlightening.
Congenital Heart Disease: Complications Before and After Surgical Repair

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VIGNETTE

A parent finds her 18-month-old child with a history of single ventricle and recent congenital heart surgery crying and cyanotic. She is concerned because her daughter has been very irritable, with decreased activity and decreased oral intake. The child has developed a low-grade fever with coughing and episodes of vomiting. Mother is concerned because the patient's oxygen saturations while sleeping are significantly lower than normal (saturations usually reside in the high 80s). She called emergency medical services (EMS) because the patient has become difficult to arouse.

EMERGENCY MEDICAL SERVICES ASSESSMENT

Children with congenital heart defects can be a challenging group of patients to manage, especially if you are not commonly exposed to these children. Emergency medical services (EMS) personnel should focus their treatment on stabilizing airway, breathing, and circulation (the ABCs) and getting the patient to the emergency department (ED) safely.
One of the most important assessment resources an EMS provider can use is the patient's caregivers. Children with cardiac defects can often be pale and cyanotic and have increased work of breathing at their baseline. Most parents will be able to help guide assessment and treatment of these children. First, assure that the airway is patent and then assess the child's breathing pattern. Always involve the parents and caregivers in the assessment of the patient's breathing pattern, work of breathing, and overall baseline effort. Questions regarding respiratory status should always be referenced in relationship to the patient's baseline. Another key component of the cardiopulmonary system assessment in patients with corrected congenital heart disease is their baseline oxygen requirement and oxygen saturation. If the parents state the oxygen saturations have been low in comparison with the child's baseline, then apply supplemental oxygen, although start with a small amount of additional oxygen and titrate the supplemental oxygen to meet the child's baseline oxygen saturation goal. Once the baseline has been met, additional supplementation may prove to be harmful. The cardiovascular system in children with congenital heart disease may have altered pathways of blood flow; too much oxygen can cause increased blood flow to the lungs leading to pulmonary edema.

The assessment of circulation in a patient with cardiac defect includes evaluation of the child's color, capillary refill time, central and peripheral pulses, and blood pressure. If the EMS assessment reveals compromise in the circulatory system, a peripheral intravenous (IV) catheter should be placed. A fluid bolus of normal saline may be given; the dose for children with cardiac defects at 5 to 10 mL/kg. Achieving vascular access in patients with cardiac defects is often difficult. If circulatory compromise is present and IV access cannot be obtained, intraosseous access should be considered. This decision should be tempered in relationship to the patient's degree of compromise and the perceived risk-benefit ratio. Other factors that should be considered before IV placement are the skill set of each provider encountering the child. The person who is best at starting IVs should attempt IV placement on these children because crying and fussiness with prolonged access efforts may actually worsen the patient's condition. If there is minimal circulatory compromise noted on evaluation, it may be better to transport the child to a more controlled environment where the IV can be placed.

The last consideration is patient transport. If the child is stable, it may be more beneficial for this patient to come directly to a medical center that has pediatric cardiology specialists. Of course, if the child is unstable, he/she should be taken to the closest medical facility for stabilization. Input from parents and/or experienced caregivers may be helpful in determining the most appropriate receiving facility.

Perhaps the most important take-home point in the prehospital care of children with congenital heart disease is involving the parent or caregiver. Not involving or eliciting input from these resources only increases the risk for inaccurate patient assessment and deterioration due to inappropriate medical interventions. These children are unique in many aspects, and their disease state, adaptability, and response to stress and medical interventions are very patient specific and cannot be generalized. Therefore, using the parents' intuition and knowledge about their child's baseline and their experience with past illness is key toward accurate patient assessment and effective interventions by EMS personnel stabilizing and transporting these patients.

Another important aspect of prehospital care of these children is the transmission of information to the receiving ED. During a medical report, it is important to give a detailed description of the patient's underlying cardiac issues, current vital signs (with baseline vitals if the caregiver was able to provide this), and patient assessment with particular interventions that were required. The call should end with a final assessment of the needs that this particular patient may require from the ED staff. This information from EMS will greatly enhance the quality and efficiency of the care provided in the ED.

**EMERGENCY DEPARTMENT NURSING CARE**

The bedside nurse will often encounter these patients before the doctor is available to see them. Assessing a child with a congenital heart defect should begin just like the assessment of any adult or pediatric patient, ensuring that the ABCs are intact. Remember that the best resource for information regarding this child may be the parents or caregivers as they will help guide your care and be able to answer many questions. The critical thinking process for these patients with corrected congenital heart will be based on a primary assessment by the ED nurse focused on determining if the patient is in respiratory distress or failure, or has evidence of heart failure or cardiovascular compromise.
The nursing assessment for a child with a congenital cardiac defect should include the following:

- **Airway:** is the airway patent or in jeopardy—yes or no?
- **Breathing:** is the patient in respiratory distress or failure—yes or no?
- Are there retractions, nasal flaring, increased respiratory rate, or a decrease in oxygen saturation? Remember that providing supplemental oxygen unnecessarily, or too high of a flow rate or concentration, may change the flow of blood in the heart, leading the child to decompensate. Nurses should consider using a blender for oxygen delivery in these children; if this is unavailable, then starting with a small amount of oxygen and titrating up to the child’s goal saturation are acceptable.
- While listening to breath sounds, heart sounds should also be auscultated. Does the child have a murmur, or can you hear the shunt? Murmurs can be hard to auscultate, but hearing a murmur in a child with a Blalock-Taussig shunt is a normal finding. If you are reassessing a patient and the murmur has become softer, make sure that the physician is aware of this change because this could be a sign of shunt compromise.

**Circulation:** is the patient in heart failure—yes or no?

- Assessment for circulation should include the child’s color, capillary refill, central and peripheral pulses, blood pressure, and the palpation of the liver for hepatomegaly. If an infant presents to the ED and there is suspicion of a cardiac defect, blood pressure should be measured on all 4 extremities. Normally, the upper- and lower-extremity pressures are nearly the same, with the lower extremities having slightly higher systolic pressures than the upper extremities. If a difference of greater than 20 mm Hg exists between the upper and lower extremity blood pressures, then it is possible that a coarctation is present.
- Another test that can be done is measurement of a preductal and postductal oxygen saturation. This can be completed by placing a pulse oximetry probe on the right hand and the left foot at the same time. If there is a difference of 10% or greater between the two, there is a possibility of a cardiac defect.

- Another aspect that needs to be discerned is the following: does the patient have a history of underlying cardiac dysrhythmias at baseline, managed by certain medications? And if so, what interventions in the past have been used to correct dysrhythmias?

During the nursing assessment, the child should be placed on a cardiac monitor, and vascular access obtained, with laboratory studies and fluid resuscitation initiated.

- These patients should always be placed on full cardiac monitoring. The heart rhythm will often not be normal sinus rhythm. It is important to compare the rhythm to previous rhythm strips and electrocardiograms (ECGs) to determine if there are any changes in the patient’s cardiac rhythm.
- Peripheral IV placement in a child with a cardiac defect can be a challenging task; therefore, the person with the most skill at placing an IV should be used for this procedure. Multiple IV attempts can cause prolonged crying that can alter the flow of blood, causing oxygen desaturation. If IV attempts are unsuccessful, then an intraosseous needle (IO) should be used.
- Blood should be drawn with the IV start. Useful laboratory tests include a complete blood count and serum electrolytes including magnesium and phosphorus. Other laboratory test results that are commonly obtained include B-type natriuretic peptide (BNP), ionized calcium, prothrombin time, and partial thromboplastin time. Less commonly ordered studies include troponin and creatine phosphokinase MB isoenzyme. If available, a venous blood gas should be obtained to provide a quick reference to the child’s metabolic state and electrolyte balance and to determine if the lactate is increased.
- Fluid resuscitation in a child with cardiac defects requires close monitoring and reassessment of the child. Fluid boluses in these children should begin with 5 to 10 mL/kg of isotonic fluid (normal saline). One of the most important aspects of fluid administration in children with single ventricle cardiac defects is to make sure that there are no air bubbles in the tubing as air in the line can cause these children to have a stroke. If the child needs fluids, the bolus may need to run in over a half hour to an hour. Reassessment of breathing and liver size should be done before
additional boluses to make sure that the child is not being put into a fluid overload state.

History: as stated previously, the parents are going to be an invaluable resource for this patient and can help guide your care. The following are good questions to ask the family when assessing your patient:

- What is the child's normal oxygen saturation?
- How does the child look to them? Ask about normal activity, playfulness, whether the child cries around strangers, and how they are interacting with you.
- What is different today? Is there increased respiratory effort, or did they have to turn the oxygen up at home?
- Is there breathing difficulty while feeding, or does the child sweat or tire easily during feeding?

**CYANOTIC CONGENITAL HEART DISEASE: OVERVIEW**

Cyanotic congenital heart diseases (CHDs) encompass all heart lesions present at birth that result in cyanosis, or poor blood oxygen levels. Patients born with these lesions may have been diagnosed in utero by prenatal ultrasound or may go undiagnosed until birth or shortly thereafter. Each lesion presents at a different time based on several factors including (but not limited to) type of defect, presence of associated lesions, severity of the congenital defect(s), and gestational age at the time of delivery. In this section, we will review the most common cyanotic heart lesions (and 1 noncyanotic lesion), go through how they change as they are repaired, and discuss complications specific to the lesion and repair. For some of the lesions, specific management advice is covered within the section. At the end of the section, general management is reviewed.

It is important to remember that despite having the underlying condition, patients with may be presenting with an unrelated general pediatric illness. This illness may or may not be impacted (or have an impact on) their underlying heart disease. For example, a patient with CHD presenting with acute otitis media who is otherwise well hydrated and without respiratory issues may simply require routine antibiotic treatment. In contrast, a patient with CHD presenting with gastroenteritis or bronchiolitis—where dehydration and respiratory function may be complicated or may complicate the underlying heart disease—may need additional support in comparison with the otherwise healthy pediatric patient. It is important for the physician and other ED staff providing care to the patient to be aware of the patient’s underlying disease and how it may complicate routine pediatric illnesses. Furthermore, it is important for emergency care providers to decipher if the patient's CHD is playing a role in the patient's current illness and symptoms because the appropriate intervention may need to be adjusted for the patient with CHD.

The 5 “Ts” of cyanotic heart lesions are as follows:

1. Truncus arteriosus
2. Transposition of the great arteries (TGA)
3. Tricuspid atresia, including hypoplastic left heart syndrome (HLHS)
4. Tetralogy of Fallot (TOF)
5. Total anomalous pulmonary venous return (TAPVR)

**Truncus Arteriosus**

Truncus arteriosus is a lesion where both the aorta and the pulmonary artery are together as a single trunk arising from normally formed ventricles (Figure 1). These children, thus, have mixed blood circulating through their body by way of this common trunk. Before surgical repair, patients with truncus arteriosus may have oxygen saturations in the 70 to 80% range.

Repair is usually done around 2 weeks of age. The pulmonary arteries are taken off of the common trunk and connected to the right ventricle (RV) via a homograft or conduit (Figure 2). The associated ventricular septal defect (VSD) is also closed at this time.

Complications arising from this surgery include valvular insufficiency, arrhythmias, and outgrowth of the conduit.
1. With valvular insufficiency, the child may present with pulmonary congestion or pulmonary hypertension crises. Patients in this scenario may benefit from furosemide or other diuretics. You may appreciate crackles on lung examination or respiratory distress in these patients, and a chest x-ray may show pulmonary congestion with increased lung markings. Use caution in giving patients in this scenario oxygen because this may vasodilate the lungs further and worsen the condition.

2. Common arrhythmias seen are right bundle-branch blocks and heart blocks.

3. If the patient has outgrown the conduit, it may not be supplying enough blood to the lungs, and in this instance, patients may benefit from oxygen supplementation.

**Transposition of the Great Arteries**

This is a lesion where the pulmonary and aortic trunks are placed on the opposite ventricle, thus requiring on the right side of the heart to pump blood to the body (Figure 3). This lesion relies on some sort of mixing lesion in association with the transposition to be compatible with life (atrial septal defect [ASD], VSD, or patent ductus arteriosus [PDA]). There are 3 surgical repair approaches that these patients may have had; the most common is the arterial switch.

1. Arterial switch: in this repair, the pulmonary and aortic trunks are simply switched to the proper ventricle. Although the trunks are switched, the valve stays with its original ventricle. The coronary arteries are also disconnected and then reimplanted onto the new aortic valve (Figure 4).

2. Rastelli procedure: this is used in cases where the patient had left ventricular (LV) outflow tract obstruction and an associated VSD (Figure 5). During the repair, the VSD is closed such that the LV pushes slightly into the RV, thus alleviating the obstruction in outflow. A conduit is then created from the RV to the pulmonary artery.

3. Mustard/Senning procedure: this is rarely seen in practice; however, some cases do still arise (Figure 6). This approach is used when the patient is felt to have poor LV function and, thus, will need to rely on the RV for systemic circulation (or in cases when the coronaries are in an aberrant location). In this repair, the atria are switched (instead of the usual arterial switch). Ultimately, the LV pumps to the lungs and the RV pumps systemically.

There are several complications that may arise from the repair of TGA:

1. Myocardial ischemia: most often seen in cases where coronaries are reimplanted and may have poor/inadequate flow. These patients may benefit from IV fluids, oxygen, and general supportive care.

2. Supravalvar stenosis: seen especially with the Rastelli procedure, where the patient had LV outflow tract obstruction presurgical repair. These patients may benefit from IV fluid to overcome the stenosis. They may
also benefit from drugs that decrease afterload (nitroprusside), thus allowing the LV to overcome the obstruction with less strain.

3. Arrhythmias: these are more commonly seen with Rastelli and Mustard/Senning repairs, where the RV has been manipulated.

4. Poor LV function: more commonly seen in patients undergoing the Mustard/Senning procedure or in patients where surgical repair was performed later in life.¹⁻³

**Tricuspid Atresia and Hypoplastic Left Heart**

Although the 2 lesions differ anatomically, functionally they both are considered “single ventricles” and the surgical repairs are almost identical for both. Figure 7 depicts tricuspid atresia in which the tricuspid valve is poorly formed, thus resulting in an underdeveloped RV. Figure 8 depicts HLHS in which the mitral valve is poorly formed, resulting in an underdeveloped LV and atretic aorta.

In both cases, the patient will rely on a mixing lesion (ASD, VSD, PDA) to supply oxygenated blood to the body. Oxygen saturations will be in the 70 to 85% range before surgical correction. Repair of these lesions is done in 3 stages, as described below:

- Norwood procedure (done only in HLHS)
- Stage I: Blalock-Taussig shunt (BTS) or Sano modification (at birth)
- Stage II: Glenn shunt (6-9 months)
- Stage III: Fontan procedure (18-36 months)

**Figure 4.** Arterial switch repair for transposition of the great arteries.

**Figure 5.** Rastelli procedure repair for transposition of the great arteries.
1. Norwood + BTS or Norwood + Sano (Figure 9)

- The Norwood procedure is only done in HLHS as these children have an atretic aorta. This procedure creates a new (neo) aorta from the existing aorta and part of the pulmonary artery. The main pulmonary artery is then ligated.
- BTS is the creation of a conduit from the right subclavian artery to the right pulmonary artery, thus allowing for pulmonary blood flow.
- Sano modification is a creation of a conduit from the RV to the pulmonary artery.
- In all cases, an ASD will be created or augmented to allow mixing of blood returning to the heart. By the end of this first stage of surgical repairs, the patient will be considered a “single ventricle.”

- The decision to use a BTS or Sano modification is based on the patient and the surgeon. Both procedures are used until the second stage of repairs can be completed. Each has its advantages and disadvantages.
  - BTS does not cut into the RV; however, during diastole, the BTS will “steal” blood from the coronaries, thus resulting in poorer coronary blood flow.
  - The Sano requires cutting into the RV, which may result in arrhythmias and abnormal conduction. However, it allows for improved coronary blood flow. \(^4,5\)
  - Oxygen saturations after this procedure are generally 75 to 85%.

**Figure 6.** Mustard/Senning repair for transposition of the great arteries.

**Figure 7.** Tricuspid atresia.

**Figure 8.** Hypoplastic left heart syndrome.
2. Glenn (Hemi-Fontan) procedure (Figure 10)
   - The BTS or Sano is taken down. The superior vena cava (SVC) is then connected either to the right pulmonary artery or to the main pulmonary artery (bidirectional Glenn). This results in passive blood flow directly into the lungs from the upper body. Blood returning from the lower body continues to flow into the right atria. Saturations after this procedure are generally 75 to 85%.

3. Fontan procedure (Figure 11)
   - This is the final stage of the repair where the inferior vena cava (IVC) is connected to the pulmonary arteries, thus allowing all blood flowing back to the heart to flow directly (passively, because no valve exists) into the lungs, and thus, bypassing the right atrium (RA). This leaves the heart responsible solely for pumping oxygen-rich blood to the body. Once complete, patients usually have oxygen saturations in the normal reference range. This procedure may be accomplished in a few different fashions:
     - Extracardiac: the IVC is connected to the pulmonary artery (PA) without being connected to the heart at all.
     - Intracardiac: the IVC is connected to the heart by way of “tunneling” through the RA.
     - In either type, a fenestration may be placed at the RA serving as a “pop-off valve” in cases where pulmonary resistance is high and blood is backing up.\(^1\text{–}^3,^6\)

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**Figure 10.** Glenn procedure.

**Figure 11.** A and B, Fontan procedure.
Complications arising from this 3-stage repair are several and are associated with the manipulations being performed.

1. When a shunt is placed, the size is very important. Too small of a shunt will prevent adequate flow and lead to backup, whereas a large shunt will lead to fluid overload (in a BTS, this will result in pulmonary edema). 6

2. In connecting the SVC and IVC directly to the PA, no valve or pumping device exists in the situation where pulmonary resistance increases. Thus, it is impossible to overcome this resistance, and blood flow will back up, resulting in SVC syndrome and/or liver enlargement.

3. If a fenestration is placed for a “pop-off” in cases where pulmonary vascular resistance (PVR) is elevated so that blood flow does not back up significantly, emboli may occur. 4,5

Tetralogy of Fallot

Tetralogy of Fallot consists of 4 components (Figure 12):

1. Pulmonary infundibular stenosis
2. Overriding aorta
3. Right ventricular hypertrophy
4. Ventricular septal defect

The component with the most significant impact on the severity of the condition is the pulmonary stenosis. Thus, children with more profound pulmonary stenosis are often more symptomatic and may require earlier correction (or palliation until correction).

TET Spells

TET spells, seen specifically in patients with uncorrected TOF, occur when there is an increase in right to left shunting. The cause of the increase in shunting is any increase in lung pressures (including crying), which leads to higher pulmonary artery pressures and, thus, more blood flow shunting across the VSD down the path of least resistance. The result is an increased amount of oxygen-poor blood-entering circulation. Treatment for this condition is based on increasing systemic vascular resistance (SVR), thus decreasing the right to left shunting. All of the options below may be used alone or in conjunction with each other to increase SVR and decrease pulmonary pressures:

1. Place the child in a knee to chest position
2. Try and calm the child (stop crying)
3. IV fluids
4. Oxygen
5. Morphine

Repair for patients with TOF is often performed around 6 months of age. The procedure entails closing the VSD and augmentation of the pulmonary artery. In severe cases, a conduit may need to be created to bypass the pulmonary stenosis. Complications that may arise secondary to this repair include the following:

1. Residual VSD
2. RV failure—this may be secondary to significant hypertrophy before surgical correction or in cases of significant residual VSDs
3. Conduction abnormalities—this is more common in cases where a conduit was created and the RV wall was surgically manipulated
4. Valvular insufficiency—seen more often when a pulmonary conduit is placed1-3

Total Anomalous Pulmonary Venous Return

Patients with TAPVR have pulmonary veins that connect to systemic (non-pulmonary) veins, and finally, draining into the RA (Figure 13). Thus, the RA receives both oxygenated and deoxygenated blood. For survival, these patients must have some mixing lesion (usually an ASD; sometimes a VSD). There are several courses the veins may take to ultimately reach the RA, and the course is a significant component in determining the severity of the disease. The 4 main types of TAPVR are as follows:

1. Supracardiac: pulmonary veins drain into the RA via the SVC
2. Infracardiac: pulmonary veins drain into the RA via hepatic veins and the IVC
3. Cardiac: pulmonary veins drain into coronary sinus
4. Mixed: pulmonary veins split-up and drain via more than 1 of the above options

The repair of TAPVR involves detachment of the pulmonary veins from the RA and reattachment to the LA. The intracardiac mixing lesion is closed. Complications from this repair are generally related to stenosis of the anastomosis resulting in pulmonary edema. In these cases, decreasing SVR or short-term use of diuretic may be useful to prevent pulmonary overload. 1-3

Acyanotic Lesions—Coarctation of the Aorta

In this condition, there is a narrowing of the aortic arch, most commonly just beyond the aortic arch in the “juxtaductal region” because the narrowing is often near or at the site of the PDA (Figure 14). The degree of narrowing, along with any associated lesions (PDA, VSD, bicuspid aortic valve), determine the timing and severity of presentation. Symptoms may be nonspecific, including tachypnea, tachycardia, poor feeding, and vomiting. Other, more specific findings include congestive heart failure (especially if an ASD/VSD is present), lower-extremity cyanosis, poor femoral pulses, lower blood pressure, and oxygen saturations in the lower extremities. A murmur may or may not be
present. In patients who have been repaired, the most common complication is recurrence of the coarctation.\textsuperscript{1-3}

**ASSESSMENT AND MANAGEMENT OF ILL CHILDREN WITH CONGENITAL HEART DISEASE**

The assessment of the child with known or suspected CHD—whether repaired or unrepaired—should follow a systematic approach (see Figure 15). The emergency management of these children should encompass both general interventions and those specific to the suspected underlying cause.

**General Interventions**

A general approach to the child with CHD should include the following considerations:

1. ABCs
2. Consider supplemental oxygen
   a. Is this patient below their baseline oxygen saturation? If so, attempt to administer oxygen to get them back to their baseline (not to 100%—unless that is their baseline).
3. Vascular access
   a. Caution with line placement in the neck as resultant clots may prevent patient from future operations (Glenn and Fontan).
4. Prudent fluid management
   a. This can start with a 5 to 10 mL/kg bolus if unsure of fluid status.
   b. If concern exists for dehydration leading to thrombus and/or other complications, consider 10 to 20 mL/kg bolus.
5. Laboratory studies: venous blood gas, complete blood count, complete metabolic panel, and B-type natriuretic peptide
6. Chest radiograph—2 views
7. ECG
   a. Look for changes from previous ECGs, including rhythm changes and new hypertrophy/strain patterns.
8. Ultrasound to assess heart function and fluid around the heart
9. Cardiology consult
10. Echocardiogram

**Disease-Specific Interventions**

1. Suspected shunt malfunction
   a. If a shunt is too large, increased pulmonary flow and overload may occur, especially in cases of RV-PA conduits and BTS. In these patients, decreasing blood flow to the lungs is key; thus, decreasing inspired oxygen to vasoconstrict pulmonary flow may be useful along with diuretics. Look for signs of pulmonary congestion and respiratory distress.
b. If the shunt is too small, just the opposite may occur, with too little flow reaching the lungs. In these patients, oxygen administration to vasodilate the lungs may be beneficial. Look for signs of jugular venous distention and for liver enlargement without evidence of pulmonary congestion. These patients are likely to have increased cyanosis.

c. Thrombus formation within the shunt occurs more often in states of dehydration. In these patients, appropriate fluid resuscitation is key. Oxygen administration may be useful, but proceed with caution and monitor respiratory status closely. These patients will typically have had an acute decompensation and drop in oxygen saturation, usually in a state of dehydration.5

2. Fenestration
   a. Although helpful in providing a pop-off valve in states of high pressure with difficult flow, fenestrations allow for the formation of emboli. Treatment is based on where the emboli are located. Look for a patient with an acute decompensation/sudden change in neurologic status.4,6

3. RV conduit complications
   a. Arrhythmias—including bundle-branch blocks. Usually, these are noted immediately postoperatively, and proper precautions are taken to maintain adequate function (eg, via pacemaker). However, if unrecognized postoperatively, or in an event where a pacer is not working properly, patients may require medications or external pacing until a definitive solution can occur. These patients may present with episodes of syncope or be called in because of a known malfunction (ie, malfunction was noted during a recent interrogation).
   b. Over time with scarring and reorganization of the RV, function may decrease resulting in right heart failure, leading to signs of jugular venous distention and liver enlargement. Medical therapy may be necessary to help these patients through acute issues until long-term therapy can be determined

4. Complications of improved pulmonary circulation
   a. With the benefit of improved pulmonary circulation comes the risk of pulmonary overload, leading to edema, effusions, and backup. These patients will often require surgical correction, but in the acute setting, management with diuretics and careful oxygen control to prevent further overload are key.
5. Complications of medical therapy to prevent fluid overload

a. Medical therapy is integral to many patients to control fluid status and prevent fluid overload. However, when these medications work too well, or in cases where the patient becomes dehydrated secondary to acute illness, poor flow, and possibly, thrombus formation may occur. These patients need careful fluid resuscitation with 10 to 20 mL/kg bolus(es). Look for a patient with an acute change in cardiopulmonary status with an intercurrent illness.

Cardiogenic Shock

The patient assessment findings that you would see with cardiogenic shock are the same as other shock etiologies. This includes weak pulses, poor capillary refill time, decrease in urine output, change in mental status, and hypotension. Specific assessment findings for cardiogenic shock include cardiac dysrhythmias, S3 or S4 heart sounds, jugular venous distension (difficult to assess in young children), edema, and hepatomegaly. Aggressive fluid volume replacement is generally needed for the shock state. In patients with CHD, although caution should be taken with fluid resuscitation, judicious fluid boluses (20 mL/kg) with reassessment after each bolus may be indicated to overcome the patient’s hypovolemia. If during the fluid resuscitation, auscultation of the lungs reveals crackles and respiratory distress ensues or worsens, a diuretic should be considered.

If the patient continues to have poor perfusion and hypotension despite adequate fluid administration, then an inotrope and vasopressor may be needed to support perfusion and blood pressure. Dopamine (0.5-20 μg/kg/min) is frequently the first-line inotrope used for cardiogenic shock. Often, Milrinone (0.25-1 μg/kg/min) is chosen because it reduces afterload and has a positive inotropic effect on the heart. Dobutamine, epinephrine, and

<table>
<thead>
<tr>
<th>Drug</th>
<th>Mechanism</th>
<th>Inotropy</th>
<th>Chronotropy</th>
<th>Vaso</th>
<th>PVR</th>
<th>SVR</th>
<th>Dose</th>
<th>Side effects</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dopamine</td>
<td>Alpha (high) and Beta (low)</td>
<td>✗</td>
<td>✗</td>
<td>constriction</td>
<td>↑</td>
<td>↑</td>
<td>0.5-20 mcg/kg/min</td>
<td>arrhythmia</td>
</tr>
<tr>
<td>Dobutamine</td>
<td>B2&gt;B1</td>
<td></td>
<td></td>
<td>dilatation</td>
<td></td>
<td></td>
<td>0.5-10 mcg/kg/min</td>
<td>arrhythmia</td>
</tr>
<tr>
<td>Epinephrine</td>
<td>B</td>
<td></td>
<td></td>
<td>constriction</td>
<td>↑</td>
<td>↑</td>
<td>Bolus 1-20 mcg/kg; Infusion 0.05-2 mcg/kg/min</td>
<td>hypertension, tachycardia, myocardial necrosis</td>
</tr>
<tr>
<td>Norepinephrine</td>
<td>Alpha</td>
<td>✓</td>
<td>✓</td>
<td>constriction</td>
<td>↑</td>
<td>↑</td>
<td>0.05-0.1 mcg/kg/min</td>
<td>arrhythmia, hypertension</td>
</tr>
<tr>
<td>Phenylephrine</td>
<td>Alpha</td>
<td></td>
<td></td>
<td>constriction</td>
<td></td>
<td>↑</td>
<td>0.05-1 mcg/kg/min</td>
<td>arrhythmia</td>
</tr>
<tr>
<td>Isoproterenol</td>
<td></td>
<td>✗</td>
<td>✗</td>
<td></td>
<td>↑</td>
<td>↑</td>
<td>0.05-1 mcg/kg/min</td>
<td>arrhythmia</td>
</tr>
<tr>
<td>Milrinone</td>
<td>Type III PDE I</td>
<td>✗</td>
<td>✗</td>
<td>dilatation</td>
<td></td>
<td></td>
<td>Load 50 mcg/kg; Infusion 0.25-1 mcg/kg/min</td>
<td>hypotension, thrombocytopenia</td>
</tr>
<tr>
<td>Sildenafil</td>
<td>PDE I</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>↓</td>
<td>PO/NG start at 0.25 mg/kg Q6 and titrate</td>
<td>hypotension</td>
</tr>
<tr>
<td>Nitroprusside</td>
<td>Vasodilator A&gt;V (A afterload)</td>
<td></td>
<td></td>
<td>dilatation</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Vasopressin</td>
<td>AVP1 receptor</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>↑</td>
<td>0.0003-0.002units/kg/min</td>
<td>bradycardia, hypotension</td>
</tr>
<tr>
<td>Amiodarone</td>
<td>Class III antiarrhythmic</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>↓</td>
<td>load 5mg/kg; Infusion 5-10mcg/kg/min</td>
<td>slows repolarization and AV conduction; prolongs QTc</td>
</tr>
</tbody>
</table>

Figure 16. Cardiovascular drugs and drips. Abbreviations: A, arterial; B, beta; PDE I, phosphodiesterase inhibitor; PO, by mouth; NG, nasogastric; V, venous; AVP1, arginine vasopressin 1.
norepinephrine can all be used for their inotropic effects (see Figure 16 for dosing).\(^7,8\)

**Intubation, Ventilator Set-up, and Cardiopulmonary Resuscitation in Congenital Heart Disease**

In cases where the patient must be intubated, maintaining the balance of adequate ventilation and oxygenation along with pulmonary and systemic perfusion can be challenging. Positive pressure ventilation of any sort in patients with cyanotic heart lesions can lead to decreased pulmonary blood flow as the positive pressure impedes pulmonary flow. Specifically, patients with Glenn or Fontan physiology are more likely to have increased PVR, leading to decreased cardiac output. They lack an RV response to overcome this PVR because the Glenn and Fontan allow passive flow from the SVC and IVC, respectively. In patients with TOF with RV hypertrophy and RV dysfunction (which may be seen after surgical repair), diastolic filling is impaired, resulting in poor cardiac output (Figure 17).\(^9\)

Therefore, if intubation or positive pressure ventilation of any sort is necessary, special considerations must be made to achieve this balance. Blood pressure, perfusion, and respiratory status must be carefully monitored to ensure that this balance is met and maintained. Low end-expiratory pressure is essential to allow for adequate blood flow. Constant pressure via square wave ventilation is usually recommended. Settings starting with low respiratory rates, short inspiratory time, and a tidal volume of 5 to 6 mL/kg are generally ideal. In cases where a significant pulmonary process may be present, higher inspiratory pressures may be needed initially and can be adjusted as the respiratory status starts to improve.\(^8,9\)

When setting oxygen levels during ventilation, remember to maintain the patient’s baseline oxygen saturations. Remember that oxygen is a vasodilator.

<table>
<thead>
<tr>
<th>Lesion</th>
<th>Repair</th>
<th>Complications</th>
<th>Oxygen Saturations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Truncus Arteriosus</td>
<td>1. Pulmonary arteries moved from the main trunk and connected to RV 2. Patch closure of VSD</td>
<td>1. Arrhythmias 2. Valve insufficiency leading to pulmonary hypertension and pulmonary edema</td>
<td>Pre-repair: mixing lesion - sats 75-85%  After repair: ~100%</td>
</tr>
<tr>
<td>Transposition of the Great Arteries</td>
<td>Arterial switch (back to normal position) or Rastelli (uses a conduit from RV to PA) or Mustard/Senning (Atrial Switch)</td>
<td>1. Myocardial ischemia 2. Arrhythmias 3. Valvular stenosis 4. Poor LV function and CO</td>
<td>Pre-repair: mixing lesion - sats 75-85%  After repair: ~100%</td>
</tr>
<tr>
<td>Tricuspid Atresia Hypoplastic Left Heart Syndrome (HLHS)</td>
<td>1. Norwood +BTS or Sano (birth) 2. Glenn (6-9 months) 3. Fontan (18-36 months)</td>
<td>1. Norwood BTS or Sano: shunt thrombosis, pulmonary edema or cyanosis if shunt size not correct 2. Glenn: SVC syndrome because of increased PVR 3. Fontan: SVC syndrome if elevated PVR; Risk of emboli if fenestration is placed</td>
<td>Pre-repair, post Norwood and Glenn: 75-85%  After Fontan: Near 100% (Unless patient is utilizing the fenestration)</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>Around 6 months of age: 1. Augmentation of PA and valve (or placement of a conduit) 2. Closure of VSD</td>
<td>1. Arrhythmias 2. Valve insufficiency leading to pulmonary hypertension and pulmonary edema</td>
<td>Pre-repair: Dependent on pulmonary stenosis  After repair: 100%</td>
</tr>
<tr>
<td>Total Anomalous Pulmonary Venous Return</td>
<td>1. Reattachment of pulmonary veins to LA 2. Rerouting (if needed) of pulmonary vessels</td>
<td>Anastomosis stenosis with resultant pulmonary edema</td>
<td></td>
</tr>
</tbody>
</table>

*Figure 17. General overview of congenital heart lesions.*
It can lead to pulmonary fluid overload in patients not accustomed to such high levels. Conversely, oxygen may be helpful for patients with pulmonary hypertension or poor pulmonary flow.

Cardiopulmonary resuscitation (CPR), likewise, offers physiologic challenges in patients with CHD. This specifically applies to patients like those discussed previously, including those with Glenn and Fontan repairs and TOF physiology. In these patients, CPR, much like mechanical ventilation, creates positive pressure that can impede pulmonary, and ultimately, systemic blood flow. However, in situations where CPR is indicated, there is no alternative because it is a potentially lifesaving measure that must be performed. In these scenarios, everything must be done to attempt to alleviate any PVR and restore systemic perfusion.

In patients with CHD (corrected or uncorrected) who require CPR, the importance of coordinated chest compressions with ventilations to assure effective ventricle filling is the same as with other patients, although with a few unique caveats. The use of sodium bicarbonate and calcium chloride/gluconate has been well documented as a beneficial adjunct in these patients during CPR and post-resuscitation stabilization. In these particular patients, significant metabolic acidosis is an underlying issue in relation to the effectiveness of the resuscitation efforts. A state of acidosis is counterproductive to the myocardial function and its responsiveness to resuscitative agents. These patients with cardiac defects are even more affected, and thus, have a less responsive myocardium to resuscitation measures in an acidic environment. Thus, correction of the underlying acidosis is imperative to improve outcomes.

**SUMMARY**

Children with congenital heart disease represent a population of patients presenting to the ED with unique medical issues. It is important for emergency care providers of all levels caring for these patients to understand their underlying heart lesion, repairs, and potential complications specific to these patients. Providers must be aware of how patients with congenital heart disease present with both complications specific to their condition as well as common general pediatric illnesses. Actively seeking baseline information and involving parents and home health care providers are essential in providing the best care to these patients. Furthermore, by remembering the basic principles of congenital heart disease, evaluation and management of these patients will be less intimidating and overwhelming.

**REFERENCES**

Difficult Airways, Difficult Physiology and Difficult Technology: Respiratory Treatment of the Special Needs Child

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VIGNETTE

Dispatch to Medic 6, we’ve got a call to a rural location for a 4 year old child in respiratory distress. You roll down the long dirt driveway to an isolated home in a rural county. The foster parents meet you at the door and bring you into a bedroom where a small child is in a hospital bed with a tracheostomy attached to a
ventilator. The little boy is much smaller than you expected for his age, appears pale throughout and bluish around the lips. His chest moves laboriously in and out with such force you can clearly see all his ribs. His parents tell you that he's had a cold for a few days and has been producing a lot of sputum through his tracheostomy tube. They have had to turn up his oxygen steadily for the past day or so. However, he started breathing like this about an hour ago and his home pulse oximeter has been reading oxygen saturation levels at around 70-75%. What's the next step?

**EMERGENCY MEDICAL SERVICES ASSESSMENT**

The emergency medical services (EMS) approach to the acute respiratory management of the child with special needs follows the same basic principles as all emergency care. However, the respiratory pathology found in this population of children will often require very timely recognition and intervention such that minutes and seconds matter. The proper approach is to calmly assess the situation, identify the problem, and act to address the most life-threatening complications first. As in every emergent patient, it is necessary to start with the ABCs, specifically airway and breathing, in the child with respiratory distress.

**AIRWAY**

The first priority in every child with respiratory distress is to ensure the presence of a patent airway and whether the child has special needs or not. However, in this population, the patient's airway may be maintained with a tracheostomy tube, which will require a different approach.\(^1\)

**Original Anatomy**

For special needs children who breathe using their original anatomy and without the use of an artificial airway, the first thing to remember is that they do not necessarily have a “normal” airway. A systematic approach to each child is recommended.

- Apply jaw thrust: many special needs children may have conditions that translate to a small or partially obstructed airway, such as a small mouth, large tongue, or laxity of the airway cartilage and soft tissue. A jaw thrust will improve the patency of the airway by moving the tongue forward and opening the oropharyngeal airway. Often, this simple maneuver is enough to alleviate the respiratory distress to a significant degree in these children.\(^1\)
- Suction mouth and nose: medically complex children often have difficulty managing their secretions. In some cases, they may be treated with an anticholinergic drug (ie, glycopyrrolate) to decrease secretions. However, this plus their tendency to "mouth breathe" may result in thicker secretions that may lead to mucus plugs in both the upper and lower airways. Saline instillation followed by suctioning of secretions from the mouth and nose will additionally improve the caliber of the airway. Remember that airway resistance is inversely proportional to airway diameter, and even small increases in airway size may significantly decrease work of breathing.\(^1\)
- Inspect for foreign body and remove if present: many special needs children are developmentally delayed, sometimes significantly. Therefore, it is necessary to consider a foreign body obstruction in much older patients than would be typical in the healthy pediatric population.
- Consider an airway adjunct: for a child that is breathing on their own, all that may be necessary is to support airway patency. For the conscious patient, a correctly sized nasopharyngeal tube will provide a semirigid opening to the oropharynx without provoking a gag reflex. In an unconscious patient with minimal or absent gag reflex, the placement of an oropharyngeal airway may assist in achieving better patency before a more secure airway method is attempted.\(^1\)

**Tracheostomy**

The presence of a tracheostomy can present a unique challenge in the child with respiratory distress. It is necessary to rule out the tracheostomy tube as the source of obstruction in every child with distress.\(^2\)

- Check for presence and placement of tube: a common etiology of respiratory distress in a child with a tracheostomy is accidental removal or displacement of the tube. If present, make sure that the tube is fully inserted up to the hub and that the tracheostomy tube ties are secured.
• Pass suction catheter through tube: this serves the dual purpose of checking the patency of the tube and simultaneously removing potential or actual obstructions in the tube.

• Try some positive pressure: attach a self-inflating bag to the tracheostomy hub and attempt to provide a few breaths with the goal of dislodging a potential mucus plug in the tube itself. If you feel significant resistance in delivering positive pressure, abandon this immediately because the tube may have migrated out of the trachea.

• Replace the tube: if the above interventions are not successful, one should assume the tube as the source of the problem, and the tube should be replaced. Locate a same-size replacement from available equipment or by asking the family; many families will have a replacement tube. When present, replace the tracheostomy tube as follows:

  1. Prepare the replacement tube by removing packaging and inserting the stylet; set this immediately beside the patient. If present, test the balloon on the new tube before insertion.
  2. Deflate the current tracheostomy's indwelling balloon and undo the tracheostomy ties.
  3. Grasp the tracheostomy hub on both sides and remove smoothly.
  4. Grasp the replacement tracheostomy tube in the same manner and insert fully to the hub, aiming both posteriorly and inferiorly.
  5. Inflate the indwelling balloon and secure around the neck using the tracheostomy ties.

If a tracheostomy tube is not available, an endotracheal tube of the same or 0.5 size smaller may be used as a temporizing measure and may be a lifesaving adjunct. Finally, if there is resistance to passage of a tracheostomy or endotracheal tube, the lumen may not be patent and a false passage may have been created. This could result in subcutaneous emphysema and compromise of the airway. Do not replace a tube against significant resistance. Rather, attempt to ventilate the patient orally with a self-inflating bag.

**BREATHING**

After achieving a patent airway, your next priority in a child with respiratory distress is adequate oxygenation and ventilation. The pulmonary pathology or compromised anatomy of a special needs child may necessitate a specialized approach.

• Bag-mask ventilation: for children receiving bag-mask ventilation, a proper technique is important to overcome the challenges of a naturally floppy airway. Provide jaw thrust by gripping the mandible and pulling the child’s face into the mask, rather than pushing the mask down. Remember the importance of maintaining the “sniffing” position. Keep fingers on the mandible and out of the submental space to avoid compressing soft tissues. It is very easy to accidentally occlude the airway in small children with excessive compression.

• Take them off the vent: the long-term ventilators that most special needs children use are designed to assist in their ventilation in a well state. They are not designed to automatically compensate for the increased needs of a sick child. Therefore, the first thing to do for a ventilated patient is to take them off their home device and provide manual positive pressure ventilation.

• Positive pressure ventilation with a tracheostomy: attaching a self-inflating bag directly to the hub of a tracheostomy tube is the equivalent of bagging an intubated patient. The same rules for adequate aeration and chest rise still apply.

• Adjust the positive end-expiratory pressure (PEEP): many children in the special needs population may be at risk for the complications of chronic lung disease (CLD), which can include stiff, noncompliant lungs, and difficulty clearing secretions. A gradual and judicious increase in PEEP can often help compensate for these conditions.

**EMERGENCY DEPARTMENT NURSING: TIPS AND TRICKS FOR CHILDREN WITH COMPLEX RESPIRATORY PROBLEMS**

Rarely is a nurse more intimidated than when presented with a chronically ill child with an acute illness. These children require a broad assessment and knowledge of the respiratory system that encompasses an understanding of both the child’s baseline anatomy and their current pathophysiology. The following is a guide to creating a beneficial experience for both the nurse and the patient while in the emergency department (ED).
First, Stay Calm

It will not be as difficult as you think. Parents and home caregivers are usually an invaluable resource for the ED team, particularly in the assessment of the child’s baseline function, and throughout the emergency care experience. The emergency nurse need not be an expert in every disorder, but knowing one’s resources, having access to the right equipment, and remembering the basics will afford the best chance of success in caring for medically complex children.

Remember the Basics

Much of the pathology occurring in infants or children involves the respiratory system, to some degree. Whether the patient presents with cerebral palsy, a tracheostomy, or Pierre Robin syndrome, basic nursing techniques should always be considered. Anticipatory care for these patients is variable depending on the diagnosis, but nurses can simply remember the ABCs and focus mainly on positioning, suction, breathing assistance, and oxygen as needed. These basic interventions alone may address many presentations of acute respiratory illness in this population.

Positioning

Initially, the positioning of the patient should be corrected to optimize airway patency. Several medical conditions result in anatomical changes in the airways of children, and therefore, may require external manipulation to obtain adequate patency. For example, a patient with Down syndrome may have an extremely large tongue that can block the airway and occlude respiration. The aforementioned baby with Pierre Robin also has challenging airway anatomy because of a small mandible and decreased mouth opening. Positioning the patient for optimal airway patency should therefore be tailored to the individual anatomy and circumstances of the patient. It is beneficial to recognize the position in which the patient is least stressed. For example, some patients with severe cerebral palsy do much better lying on a particular side and seem to increase their heart rate, respiratory rate, and work of breathing when positioned otherwise. As mentioned previously, parents and caregivers are an invaluable resource regarding what the child prefers and should be valued as partners in care. Other examples of positioning techniques for special needs children that may be considered include elevating shoulders slightly to minimize tongue occlusion or head manipulation in contracted patients whose tendency is to drop their chin or favor one side with head placement.

Clearing the Airway

Secretions can also be a challenge for the special needs child and patients with a tracheostomy tend to have increased difficulty managing their secretions. In the setting of respiratory illnesses, secretions may become thicker and more copious. Suctioning is needed more often, and diligence is necessary to keep the airway clear and the patient comfortable. Humidification through a tracheostomy collar can be very helpful in promoting secretion mobilization and comfort. Most tracheostomy care parents will normally have everything needed for the care of their child when they present for emergency care. Parents may be supplied, for example, with an extra tracheostomy tube and one that is a size smaller in case of emergent displacement. They will normally have a clean set of tracheostomy ties as well. Families are taught to keep these tubes, along with the appropriate-sized suction catheters, with the patient at all times. If there is an inability to gain access to the appropriate pediatric tracheostomy supplies, parents may be able to provide what is needed.

When suctioning is required, an appropriately sized suction catheter should be inserted into the tracheostomy. Take care not to extend the catheter beyond the length of the tube itself. Again, parents are an extremely good resource for these details, such as suction catheter depth. One recent study has indicated the instillation of saline into the tracheostomy before suctioning offers highly questionable benefit because the mucous and saline never mix, defeating the purpose of “loosening” the mucus for easier withdrawal. However, the temptation to do this is acknowledged and could be considered if simply trying to elicit a cough.

Patients with chronic respiratory disease who do not have a tracheostomy may also present with an abundance of thick secretions. Patients who are nonambulatory are particularly at risk for ineffective airway clearance due to a weak cough, immobility, and decreased muscle tone. These factors combined with their compromised lung and airway anatomy predispose to complications such as pneumonia. Pediatric Yankauer suction attachments are effective at clearing oral secretions, but deep suction with the appropriate-sized suction catheter may be necessary for the patient who has difficulty clearing pharyngeal secretions. Chest physiotherapy may be a helpful adjunct for the
immobile patient that cannot expel secretions. Mist and frequent repositioning of the patient are helpful with secretion clearance.

**Equipment Pearls**

Home ventilators are a commonality with special needs patients and are usually best managed by respiratory therapists upon arrival to the ED. Home ventilated patients almost always have a tracheostomy in place. Patients on home ventilators may present to the ED in respiratory distress. In this scenario, it is best to disconnect the ventilator from the patient and manually assist respirations with a self-inflating or anesthesia bag until a tracheostomy assessment can be performed, much as one would do for an intubated patient under the same circumstances.

Complex patients may present to the ED with gastrostomy tube feedings infusing. A good rule of thumb by which to practice is to turn off infusing feeds if there are any respiratory concerns. These fragile children have a high risk of aspiration pneumonia and respiratory illness and the risk is even greater when paired with these feedings. Be sure to address the child's hydration status with the care team if the feeds are discontinued. In the presence of respiratory distress, vent the feeding tube to decrease intra-abdominal pressure, which can contribute to increased work of breathing and decreased ventilation.

**Monitoring**

Pulse oximetry parameters in the special needs child rarely differ from that of the typical patient, with the exception of those with a history of cardiac anomalies. Oxygen saturations should remain above 90%, with a target range of 95% to 100%. A prolonged expiratory phase with a corresponding drop in oxygen saturations may be seen in children with restrictive lung disease, such as those with cerebral palsy. This can sometimes be harder to recognize clinically in chronically ill patients because they are typically very pale in complexion and may have irregular breathing patterns at baseline. A good plethysmography waveform is imperative for accurately interpreting the oxygen saturation as measured by pulse oximetry readings. Pulse oximetry should remain in place throughout the ED visit and requires evaluation and intervention for drops below 90%. Placement of the oximetry probe can sometimes be tricky and may require multiple attempts if poor perfusion is present. If extremity perfusion is inadequate, consider placement on the ear lobe.

Patients with bronchospasm who require bronchodilators such as albuterol may be at risk for hypokalemia and should be placed on cardiac monitoring for rhythm and rate recognition. Intravenous access and basic laboratory work can typically be anticipated in these children. Blood gases may be ordered to evaluate the effectiveness of ventilation. It would be a benefit to the patient if acquiring blood for these laboratory tests is considered while vascular access is being established. The child's caregivers will be a good resource for venipuncture as well, often times guiding the nurse to successful veins for access.

These are just a few guiding principles in the ED care of the special needs child. They are indeed a challenge to care for even with appropriate resources at the ready. Keeping the ED assessment and care systematic and concentrating on the basics will guide the ED visit to an optimal outcome. Remember that parents and home health and regular caregivers are always ready and willing to assist, so be ready and willing to accept their guidance.

Finally, advocate for the basics in pediatric emergency care equipment in your department so that a basic need does not become a major obstacle when the time comes. Published recommendations for pediatric readiness offer an excellent guide as to what equipment and medications are needed.

**RESPIRATORY THERAPY: MEDICATIONS, EQUIPMENT, AND TECHNOLOGY**

A description of commonly used equipment for the respiratory care of medically complex children follows.

**Nasal Cannula**

This low-flow oxygen delivery device fits into the patient's nares. This delivery source takes advantage of the anatomical dead space of the nasopharynx to concentrate an oxygen reservoir for inspiration. Operation requirements include the following: oxygen source of 50 psi, flow meter, prefilled humidifier, blender (optional), and pulse oximeter.

- Flow rate range: 0.25 mL/min to 5 L/min
- Expected fraction of inspired oxygen (FIO₂): 21% to 40%

Hazards include the following:
- Lack of humidification leads to drying of secretions and nasal bleeding with resultant increased airway resistance and/or obstruction
• Nasal obstruction
• Bacterial colonization

**Simple Mask**
This face mask provides oxygen for patients with spontaneous breathing who may require higher O₂ concentrations or cannot tolerate or cooperate with a nasal cannula. One disadvantage is that the mask is an open system in which FIO₂ cannot be tightly regulated and high concentrations cannot be assured.

- **Typical flow ranges:** 6 to 10 L/min

**Venturi Mask**
This face mask provides oxygen to patients with spontaneous breathing who require higher oxygen concentrations. The advantage of the venturi mask over the simple mask is that O₂ concentrations can be controlled with reasonable accuracy and ease of use. The Ventimask has interchangeable adapters that control the amount of air, and oxygen blended together.

- **Typical flow ranges:** 6 to 10 L/min
- **FIO₂ color-coded adapters:** 24% to 50%
  - Blue (24%), yellow (28%), white (31%),
  - green (35%), pink (40%), orange (50%)

Disadvantages are as follows:
- It cannot reliably deliver an FIO₂ greater than 50%.
- Frequent adjustments may make it less feasible for an acute setting.

**Non–Rebreathing Mask**
This face mask provides high-flow oxygen to patients with spontaneous breathing who require 100% O₂ concentrations. The non–rebreathing mask is a partially closed system that consists of 2 exhalation valves and an attached reservoir bag ensuring that the patient will receive a FIO₂ as close to 100% as possible without an advanced airway.

- **Typical flow rate:** 15 L/min

**Metered Dose Inhaler With Spacer**
This inhaler is appropriate for any patient older than 6 months with a normal breathing pattern who is spontaneously capable of taking a deep breath with a breath-hold maneuver. Metered dose inhalers have been demonstrated to be as effective as nebulizer devices in the acute care setting and may offer relative efficiency in staff time owing to their shorter administration time.

- **Albuterol:** 2 to 4 puffs every 4 hours as needed for wheezing or increased work of breathing; designed for use in the acute setting as a rescue inhaler
- **Fluticasone:** 2 puffs every 12 hours daily as a maintenance medication to decrease and prevent inflammation of airways

**Handheld Nebulization**
Handheld nebulization is used to deliver aerosolized medications, typically with a mouthpiece or mask. Set the flow meter between 5 and 7 L/min, and monitor vital signs and heart rhythm for any acute changes. Common drugs that can be delivered via handheld nebulization include the following:

- **Albuterol**
- **Ipratropium**
- **3% hypertonic saline**
- **2 to 3 mg of 1:1000 epinephrine**
- **Racemic epinephrine**
- **Budesonide**

**Continuous Nebulizer**
Continuous nebulizer is used to deliver continuous aerosol treatments over a 1- to 4-hour time frame. It is typically used with a mask that is attached to corrugated tubing delivering the aerosol from a large-bore nebulizer attached to a blender (flow meter set at 11 L/min). Three of the most common drugs used include the following:

- **20-mL bottle of 5 mg/mL albuterol sulfate mixed with 1.5 mg ipratropium and 91 mL of saline**
- **3 to 6 mg of 1:1000 epinephrine with 24 to 27 mL of saline**
- **3 mg of 1:1000 epinephrine with 15 mL of 3% hypertonic saline**

**Mini-Heart Nebulizer**
Mini-Heart nebulizer is designed to deliver a continuous aerosol over 1 to 4 hours inline in the setting of positive pressure ventilation, typically in concert with either bilevel positive airway pressure (BPAP) or a mechanical ventilator. The nebulizer receives flow from extension tubing attached to a blender set at 2 L/min with FIO₂ greater than 30%. Three of the most common drugs used include the following:

- **10 mL of 5 mg/mL albuterol sulfate mixed with 1.5 mg ipratropium and 6 mL saline**
- **3 mg of 1:1000 epinephrine with 2 mL saline**
Noninvasive Ventilation

**Vapotherm**

This device is used to provide humidified oxygen via specialized cannula at a set flow rate, typically from 4 to 20 L/min. The benefit of Vapotherm is that the provider can give a controlled amount of humidified oxygen and flow. Vapotherm must be set up to deliver either “low flow” or “high flow” rates, which require different types of equipment.

- Low flow: 1 to 8 L/min
- High flow: 5 to 40 L/min

**Nasal Continuous Positive Airway Pressure**

This is a system used to provide continuous positive airway pressure (CPAP) to an infant via nasal prongs or nasal mask. The manufacturer produces a mask/prong size card that can be matched with the patient to determine proper sizing. The prong circles on the card should equal the size of the patient’s nares. Once the prongs, mask, and head gear are secured, the nasal CPAP will provide a measured PEEP based on the O₂ flow rate. To increase the PEEP, one will need to increase the flow rate. The FIO₂ can also be independently adjusted to the desired concentration. When assessing the patient, listen to the baby's chest for a “CPAP roar” to confirm prong placement and CPAP delivery.

**Bilevel Positive Airway Pressure**

As a type of noninvasive ventilation, BiBPAP helps keep the upper airways of the lungs open by providing positive pressure through a sealed face mask. It has been found to be especially useful for pediatric patients with asthma and other chronic pulmonary conditions that may be predisposed to hypercarbia due to neuromuscular disease or compromised lung parenchyma. The name “Bi” meaning 2 and “PAP” meaning positive airway pressure represents how this device works. Positive pressure is set at 2 levels:

1. Inspiratory positive airway pressure (IPAP)
2. Expiratory positive airway pressure (EPAP)

A higher pressure is used to aid inspiration (IPAP), and a lower pressure is used when exhaling (PEEP). Other BiPAP settings include FiO₂, inspiratory time, and a backup rate, which is typically set between 4 and 8 breaths/min.

RESPIRATORY PATHOLOGY IN THE SPECIAL NEEDS CHILD

Children with special health care needs (CSHCN) are a growing segment of the population. They possess a unique set of anatomical and physiologic factors that demand particular attention upon presentation in the ED. These children, many of whom are already on home oxygen or a long-term ventilator, often have preexisting respiratory compromise and lack pulmonary reserve. These conditions make them particularly vulnerable to the typical respiratory infections of childhood. At the same time, they are also at risk for additional respiratory complications that are unique to their underlying condition. Three broad categories of conditions that may contribute to respiratory disease among CSHCN are neurologic and neuromuscular compromise, airway obstruction (upper and lower), and anatomical abnormalities. Unlike many other children in the ED, CSHCN in respiratory distress will often have several concurrent problems that contribute to their ongoing disease process.

**Neurologic and Neuromuscular**

Neurologic conditions such as cerebral palsy, spinal myopathies, muscular dystrophies, and seizure disorders are often associated with low tone, decreased ability to protect the airway, and poor respiratory reserve. A common childhood respiratory infection such as viral pneumonia may lead to greater respiratory compromise than is expected in the general pediatric population. The congenital or acquired neurologic disorders present in these children alter their ability to protect and support their respiratory system. Their impaired neuromuscular condition may also predispose them to decreased muscle tone, inability to cough productively, gastroesophageal reflux, poor gag reflex, increased secretions, and decreased ability to ambulate. In CSHCN, these basic physiologic functions and protective mechanisms are more easily disrupted, and small insults to a child's respiratory homeostasis may quickly lead to respiratory distress and failure.

**Upper Airway Obstruction**

Partial upper airway obstruction is a fairly common reason for ED visits in children owing to their smaller airways and this plays an even greater role among CSHCN. The most obvious sign of upper airway obstruction may be inspiratory stridor, although obstruction may present nonspecifically with...
increased work of breathing, grunting, retractions,
and hypoxia. In general, children will most commonly
present with upper airway obstruction due to an
infectious process such as croup, peritonsillar or
retropharyngeal abscesses, or tracheitis. However,
CSHCN are particularly at risk for foreign body
aspiration because of underlying developmental
delay and diminished protective airway reflexes.
Children with tracheostomies have an inherent risk
for obstruction at their tracheostomy stoma or due to
partial or complete obstruction or dislodgement of the
tube. As detailed previously, timely intervention in
these cases is often a matter of life and death.

Former premature infants, those with a history of
neuromuscular disorders, those who are frequently
intubated, and those who are chronically ventilated
often have a degree of laryngobronchomalacia and/
or subglottic stenosis. For these children, it is
important to establish a baseline because many
will have some degree of clinically evident airway
“floppiness” and partial upper airway obstruction,
even during periods of wellness. Airways that lack
normal cartilaginous and muscular support are far
more predisposed to collapse during times of
respiratory distress. Often, a concurrent viral
illness, sedative medications, or simple fatigue
from prolonged illness can cause the obstruction
to worsen. Another anatomical condition in young
children and infants with a history of prolonged
intubation is subglottic stenosis. This is a complica-
tion arising from contact between the endotracheal
tube and the trachea. Necrosis, granulation tissue,
and eventually fibrosis can develop and further
decrease the compliance and circumference of the
upper airway.

**Lower Airway Obstruction**

Lower airway obstruction commonly presents as
a reactive airway disease such as asthma in children
and must be approached with caution among
CSHCN. The old maxim “all that wheezes is not
asthma” is particularly true among CSHCN. Advances
in neonatal intensive care have led to a large
cohort of former premature infants with CLD,
formerly known as bronchopulmonary dysplasia. Baro-
trauma from chronic ventilation and subsequent
altered lung development leads to prolonged oxygen
dependence in the neonatal period. As these
children age, clinical manifestations such as air
trapping, bronchospasm, wheezing, and inflamma-
tion may resemble those of asthma. Owing to their
impaired pulmonary clearance, children with CLD
are predisposed to more severe infections, particu-
larly bronchiolitis, in the first 2 years of life. This
common lower respiratory infection can manifest
with signs of increased work of breathing and
hypoxia owing to the lack of pulmonary reserve in
this population.

Cystic fibrosis (CF) is a congenital disorder of
childhood that results in progressive lower respira-
tory tract airway obstruction due to impaired
secretory function of the pulmonary epithelium. In
CF, the lungs produce an abnormally thick mucus
layer that plugs airways, compromises effective
clearance, and leads to colonization with pathogenic
bacteria. Over time, the architecture of large- and
medium-sized airways becomes disrupted, making
these children prone to air trapping, bronchiectasis,
diminished functional lung volume, and frequent
bacterial pneumonias. Among children with CLD or
CF, these subsequent infections further compro-
mise their pulmonary reserve and ability to adapt to
disruptions in their already diminished baseline
respiratory function.

Another form of lower respiratory tract obstruc-
tion that CSHCN are at risk for developing is
aspiration pneumonia or aspiration pneumonitis.
Children with impaired protective airway reflexes
and those with anatomical abnormalities of the
gastrointestinal tract leading to reflux are at risk for
aspirating gastric contents during periods of acute
illness or altered mental status. The acidic pH
destroys the surfactant lining of the airways and
causes diffuse inflammation leading to hypoxia,
tachypnea, and fever. Although an initial chest
x-ray may be normal immediately after an event,
radiologic signs of pneumonitis typically appear
within several hours of the aspiration event. In some
cases, aspiration pneumonitis may progress to
aspiration pneumonia if aspirated bacteria are
ineffectively cleared from the respiratory tract.

**Anatomical Considerations**

Although airway obstruction is a common cause
of respiratory distress among all children, special
considerations must be taken with CSHCN. Many of
these children have unique congenital or acquired
anatomical abnormalities that affect airway func-
tion, particularly during times of stress or illness.
Micrognathia is a known association with syn-
dromes such as Pierre Robin or velocardial facial
syndrome and may exacerbate upper airway ob-
struction by causing the tongue to be displaced
posteriorly into the oropharynx. Respiratory dis-
tress in these children may be successfully treated
by simply placing the child prone or by pulling the
tongue forward with a finger or hemostat. Children
with Down syndrome often have poor muscle tone
and a large tongue that can similarly lead to upper airway obstruction. Certain types of cerebral palsy and other neuromuscular disorders may be associated with severe scoliosis that inhibits full chest expansion and/or excursion of the diaphragm, reducing baseline functional lung volume.

Patients with a history of congenital diaphragmatic hernia and subsequent pulmonary hypoplasia are specifically at great risk due to a unique form of restrictive lung disease. These children typically have markedly reduced pulmonary reserve due to congenital underdevelopment of the lungs, and even small functional compromises or minor illnesses can lead to respiratory failure. If these patients require intubation, it is important to monitor airway pressures and to remember that the conventional methods of volume control ventilation do not apply. ⑥

Pulmonary Hypertension

Pulmonary hypertension is broadly defined as being present when pulmonary artery pressure is greater than half of the systemic pressure. It is a general term that encompasses the etiologies of pulmonary arterial hypertension, pulmonary venous hypertension, and pulmonary hypertension due to respiratory causes. This condition can be caused by either congenital or acquired disorders and is most commonly seen in the outpatient setting as a complication of prematurity or cardiac disease. These patients are typically managed with pulmonary vasodilators, which may be delivered via oral therapy, subcutaneous injection, or continuous infusion. A common treatment modality is the continuous delivery of the prostacyclin agonist epoprostenol through a central line. An important consideration for patients receiving an epoprostenol infusion is that the half-life of this medication is less than 6 minutes, making any loss of vascular access a true emergency. ⑧,⑨ In the scenario where an indwelling central line has failed, causing interruption of this medication, it may be given through a peripheral line temporarily until central access can be achieved, although the dose may need to be titrated. ⑨

Patients with underlying pulmonary hypertension presenting with an acute decompensation may demonstrate hypoxia, cyanosis, or right heart failure. The main treatment goals in the acute setting are to dilate the pulmonary bed vasculature and to unload the right heart. Oxygen is a simple and universally accessible method to accomplish this goal, along with the use of inhaled nitric oxide if available. ⑤ For any patient presenting with an exacerbation of pulmonary hypertension, transfer to a center capable of pediatric critical care including extracorporeal membrane oxygenation is indicated as soon as possible.

Hypercoagulopathy

Limited mobility and indwelling catheters represent a “double hit” for CSHCN in terms of increased risk for thrombosis and subsequent pulmonary embolism. The emergency care provider must maintain a high clinical suspicion for this complication because it typically presents with nonspecific symptoms including increased respiratory rate, chest pain, and dyspnea. A history of clot formation and knowledge of the child’s anticoagulation status are helpful when forming the appropriate differential diagnosis.

SUMMARY

Children with special health care needs are an ever-increasing segment of children treated in the ED. Careful attention to their underlying disease state and respiratory baseline is required because they are at increased risk for complications from common childhood illnesses. They have anatomical and developmental conditions that limit their ability to compensate for even slight decreases in their respiratory baseline. Care of these children is best accomplished by taking into consideration how their underlying disease state impacts the course of their acute illness. Family members and home care providers should be viewed as resources and partners in care because they typically possess great knowledge about these children and their diseases. Timely consultation with these patient’s pediatric specialty care providers will further inform patient assessment and treatment.

REFERENCES


The Hematology and Oncology Pediatric Patient: A Review of Fever and Neutropenia, Blood Transfusions, and Other Complex Problems

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VIGNETTE

A home health nurse arrives at the home of a 3 year old boy with pre-B cell acute lymphocytic leukemia (ALL) who is receiving induction chemotherapy. On arrival, his mother states he has not been feeling well with decreased oral intake.
since receiving his chemotherapy 2 days ago. He has not urinated today and has been vomiting despite treatment with anti-emetics. The home health nurse notes that the child is lethargic and feels warm to touch. He has rigors and his capillary refill is delayed. The home health nurse calls 911.

FEVER AND NEUTROPENIA

Neutropenia is a condition in which a patient has an abnormally low number of neutrophils. These cells play an important role in the body’s immune response. Neutropenia is a common side effect of chemotherapy because chemotherapeutic agents target rapidly dividing cells such as neutrophils. Patients who are neutropenic are at risk for serious bacterial infections (SBIs), such as bacteremia, pneumonia, and meningitis. Oncology patients receiving chemotherapy are at even higher risk for SBI if they have a central venous line. Because fever may be the first and only sign of an SBI in an oncology patient, families are discouraged from giving antipyretics to the patient for any indication as not to mask a fever.

Neutropenia is defined by calculation of an absolute neutrophil count (ANC). The ANC is calculated by taking the total number of white blood cells and multiplying it by the percentage of neutrophils plus the number of band forms seen on a complete blood count with differential. For example, a patient with a white blood cell count of 3000 with 2% bands and 8% neutrophils has an ANC of 300. For the oncology patient receiving chemotherapy, neutropenia is defined as an ANC less than 500.

VIGNETTE, CONTINUED

Emergency Medical Services (EMS) arrives several minutes later and finds the child listless on the couch in the living room. He is pale, febrile and tachycardic with thready pulses. He is weak but opens his eyes spontaneously and follows simple commands. He is loaded on the stretcher for transport to the local hospital. In the ambulance, you place him on a monitor and obtain a set of vital signs that show a heart rate of 197 with sinus tachycardia on the monitor, blood pressure of 70/palp, respiratory rate of 30 and oxygen saturation of 96% on room air. He is tachypneic with clear breath sounds bilaterally and normal work of breathing. He has weak pulses and mottled skin with a capillary refill of greater than 5 seconds.

EMERGENCY MEDICAL SERVICE ASSESSMENT

An oncology patient receiving chemotherapy is at risk for sepsis or other SBIs. Regardless of the presenting complaint, it is important to evaluate and stabilize the patient’s “airway, breathing, and circulation” (ABCs) systematically.

Primary assessment: ABC

- Airway: Make sure that the patient has an intact protected airway.
- Breathing: Check for irregular breathing patterns or apnea, place on pulse oximetry, and apply oxygen as indicated by oximetry, increased respiratory rate or effort, diminished circulation, or altered mental status.
- Circulation: Place the patient on a monitor for transport and document if the child is tachycardic, has a widened pulse pressure (diastolic less than half the systolic pressure), and/or is hypotensive; do not hesitate to attempt to obtain peripheral access via either intravenous or intraosseous routes to give fluids (many oncology patients will have a central line or port-a-catheter; however, you should only access these if you are trained to do so).

It cannot be stressed enough that early recognition and treatment of shock are necessary to improve this patient’s outcome. Furthermore, never administer rectal medications or check rectal temperatures in a child at risk for neutropenia because this may result in disruption of mucosa and introduction of bacteria into the blood stream. It is always good to review reference ranges of vital signs in children to quickly recognize any vital sign abnormalities.

Applying the ABCs to the patient in the vignette, the child is maintaining his airway and is tachypneic with an otherwise acceptable respiratory examination. It would be okay to provide some oxygen for comfort. His most pressing issue is his cardiovascular status. He is tachycardic and hypotensive. Unless you are trained to properly access his port-a-catheter, peripheral access should be obtained (place an IV; consider an IO if IV access cannot be obtained quickly) followed by immediate infusion of normal saline (or lactated Ringers if saline is not available). He should be transported on a monitor with frequent assessments of heart rate and blood pressure. You should immediately notify the receiving hospital emergency department (ED) staff that you are transporting this critically ill patient.
VIGNETTE, CONTINUED

The charge nurse at the community hospital is notified that EMS is en route with a 3 year old boy with ALL. The child was reportedly sluggish, warm to touch, hypotensive and tachycardic. EMS was able to obtain one site of peripheral IV access and is administering normal saline. What items should the nursing staff have ready for when this patient arrives? Is it appropriate to triage this patient in the waiting room?

On arrival of the child to the ED, the nurse immediately places him in a room under neutropenic precautions. He is placed on a monitor and a complete set of vital signs are obtained: temperature 39.0°C, heart rate 147, respiratory rate 20, blood pressure 90/60. He weighs 15 kilograms. EMS reports that they have given a total of 500 mL of normal saline en route with some improvement in perfusion. Now that the child is in the ED, what things should the nursing staff be prepared to do quickly to provide the best care for the patient?

NURSING ASSESSMENT

Treating a pediatric oncology patient in the ED may be an overwhelming task for a nurse who is not accustomed to caring for such a complex patient. This section outlines key nursing points to keep in mind, and Table 1 summarizes the top 10 clinical considerations for emergency nurses when caring for the pediatric oncology patient. To ease this stress and prevent further exposure to other sick patients in the waiting room, it is important to immediately place the patient into a treatment, resuscitation, or isolation room. These patients should be assigned a high-acuity triage level.

In the ED, one should assume that all patients receiving chemotherapy are neutropenic until proven otherwise. Neutropenic precautions (gown, gloves, and mask) should be implemented to prevent the spread of infection to the patient. Good hand washing, preferably where the family can observe care providers doing so, is highly recommended; this may be the single, most effective intervention toward the reduction of nosocomial infections. Whenever possible, the patient should have their own stethoscope and thermometer, one that will not be used to examine other patients. Finally, never administer rectal medications or check rectal temperatures in a child at risk for neutropenia because this may result in disruption of the mucosa and introduction of bacteria into the blood stream.²

Upon immediate arrival to the treatment or resuscitation room, all patients should be placed on a monitor and have vital signs, including a blood pressure with an appropriately sized pediatric cuff, monitored closely. A manual blood pressure measurement is preferred to automated because a manual will provide the most accurate reading. A physician should be notified immediately if the child demonstrates any alterations in mental status, tachypnea, tachycardia, altered perfusion, or hypotension. Furthermore, in addition to hypotension, watch for a widened pulse pressure (diasystolic pressure that is less than half of the systolic) or a lower mean arterial pressure. Finally, observe for any trending in vital signs away from normal because this may indicate that there is not much time before the child decompensates clinically. As stated above, recognition and treatment of shock are necessary to maximize the patient's outcome.

Pediatric oncology patients will usually present with some type of intravenous access readily available. Central venous catheters are common, along with implanted ports for both blood draws and fluid and medication administration. Many families are usually very reliable in applying a topical anesthetic before leaving home, thus making initial access in the ED somewhat less traumatic for the

<table>
<thead>
<tr>
<th>TABLE 1. Top 10 clinical priorities for oncology patient nursing care.</th>
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</thead>
<tbody>
<tr>
<td>1. Assign a high-acuity triage category and isolate or place in a treatment room as soon as possible.</td>
</tr>
<tr>
<td>2. Initiate protective isolation (neutropenic precautions).</td>
</tr>
<tr>
<td>3. Wash hands thoroughly. Stock supplies that can remain in the room to prevent contamination (eg, stethoscope or thermometer).</td>
</tr>
<tr>
<td>4. Place on full cardiopulmonary monitoring. Place on oxygen if needed for comfort. Check vital signs often. Remember, a hypotensive patient may be septic.</td>
</tr>
<tr>
<td>5. Avoid invasive procedures. This includes avoiding rectal thermometers/medications, urine catheterization, intramuscular injection, and tampons for adolescent females.</td>
</tr>
<tr>
<td>6. Access the child’s central line, or obtain peripheral vascular access immediately.</td>
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<tr>
<td>7. Central line management: use only 10-mL syringes to flush central lines.</td>
</tr>
<tr>
<td>8. If febrile or concern for sepsis, draw laboratory studies in this order: blood culture, BMP, CBC, type and screen, and PT/PTT.</td>
</tr>
<tr>
<td>9. If febrile, initiate antibiotics within 60 minutes of room arrival. Always ask about allergies to any medications because these patients may have reactions that are different.</td>
</tr>
<tr>
<td>10. If febrile (temperature of &gt;38.0°C or &gt;100.5°F), administer acetaminophen; dosing is 15 mg/kg every 6 h. Never give ibuprofen because of risk for platelet dysfunction.</td>
</tr>
</tbody>
</table>

CBC, complete blood cell count; BMP, basic metabolic profile; PT, prothrombin time; PTT, partial thromboplastin time.
patient. Although topical anesthetics do an adequate job at numbing the access site for pain control, the analgesia may not be fully appreciated until 30 to 60 minutes after application, depending on the type of anesthetic used. For example, EMLA (eutectic mixture of local anesthetics) typically takes about 40 to 60 minutes to achieve satisfactory anesthesia, whereas ELA-MAX (4% liposomal lidocaine) is effective in 30 minutes. Therefore, if it is a topical analgesic has not been applied before arrival, do not waste time with application in the ED if it will delay treatment. An alternative to consider is ethyl chloride spray, which will provide some instantaneous numbing effects. Eliminating pain with needle sticks is a preferred approach for stable patients; life-saving treatment should not be delayed in a truly emergent patient. Finally, if available staff do not possess a level of comfort in accessing central lines or mediports, peripheral intravenous access should be pursued as an alternative. For a patient with a concern for sepsis or shock, optimal vascular access should be large bore (in children, this equates to the largest gauge IV that can be reasonably placed).

Laboratory evaluation and administration of prophylactic antibiotics should not be delayed. A good target for blood sample collection and initiation of empiric antibiotics is within 60 minutes of patient arrival to the ED. It is important that you draw a blood culture from each lumen of a central line if present. Obtain the culture from the first blood drawn (your waste) and place it into separate blood culture bottles if there are 2 or more lumens. This will help to distinguish which lumen might be the source of the infection; therefore, accurate labeling is crucial. If no central line is present, then a blood culture may be collected via peripheral puncture. Another important laboratory test result to obtain is a complete blood count with differential to assess for neutropenia. Other studies that may be considered once vascular access is established include metabolic panel, coagulation studies, and type and screen. The treating physician may also request viral and/or fungal cultures, urine analysis and culture, or radiographic studies. Keep in mind that, in addition to avoiding rectal medications, other invasive procedures such as urine catheterization and administration of intramuscular injection should be avoided when collecting laboratory test results and administering medications.

**Family Considerations for Nurses and Health Care Providers**

Taking the family into consideration as well as the patient is a vital component of the care of the pediatric oncology patient in the ED. Many of these patients and their families feel as if they have lost control in their lives. The ED may be a very stressful and unfamiliar place for them and remind them of their initial diagnosis. Often, oncology families show their uneasiness with the ED environment by frequently questioning care decisions and methods that are used. This is often simply because methods used, and not necessarily the care itself, sometimes differ from what they are accustomed to from the oncology team. This situation can often be de-escalated or avoided altogether by including family in the plan of care. Understanding what the family expects and what they are used to and communicating what will actually occur and why will help ease the stress associated with a variation in the way care is delivered in the ED. Parents and patients are usually very well educated about the child's disease. Be respectful of their knowledge of the disease, treatment, and the parent's knowledge of their child. Ask them how they usually have things done or what makes the patient more comfortable. They may offer useful information regarding optimal approaches (eg, best peripheral access site) or care issues unique to the patient (eg, reactions to medications). Be upfront and update them on their child's care. Talk to the family openly about what you are doing and why the interventions are being performed.

**VIGNETTE, CONTINUED**

The ED physician is notified by the charge nurse that EMS has just brought a 3 year old boy with ALL, who is ill-appearing. On arrival to the room, the physician washes her hands and dons her neutropenic precautions attire prior to entering the room. She reviews the vital signs and learns that the child has received just over 20 mL/kg of normal saline from EMS. She makes note that the child is tachycardic and febrile. The nurse is preparing to access the child's central line, while the physician performs a complete physical exam. The exam is remarkable only for a child with alopecia and pallor. He is alert and in no distress. The central line site does not have any erythema or exudates. His capillary refill is 2-3 seconds. In addition to another fluid bolus, what other interventions or medications should the physician order? What other interventions or medications should the physician consider to administer? What other information does the physician need to complete the work-up? What is the disposition of this patient?

**PHYSICIAN ASSESSMENT**

If not already done, the nursing staff should be instructed to place the patient on a cardiovrespiratory monitor. It is important to obtain and scrutinize
a complete set of vital signs. The physician must assess the following questions quickly: (1) Is the patient stable? (2) Is the child truly febrile? (3) How is the respiratory status? and (4) Is there evidence of hypovolemic or septic shock?

- An easy, quick way to calculate systolic hypotension in a child is to compare the measured pressure with \[(\text{age in years} \times 2) + 70\]. A blood pressure less than this likely represents hypotension and should be addressed immediately.

Any abnormal values should be addressed systematically. Fluid resuscitation (20 mL/kg) should begin immediately and rapidly (IV push or under pressure) to treat hypotension and shock. Rapid administration of isotonic fluid is associated with improved patient outcomes in septic shock. Patients with blood pressure changes may require multiple boluses to achieve a satisfactory response. If the child's blood pressure does not improve after 2 or 3 fluid boluses, administration of inotropic infusions should be considered, and consultation with a pediatric tertiary care center, if not already achieved, should be pursued.

After assessing the vital signs and addressing any abnormal values, perform a complete physical examination, looking for a focus of infection. It is especially important to make note of the child's mental status. Pay close attention to the central line, examining for any signs of inflammation or infection. It is important to keep in mind, however, that neutropenic patients may not have an adequate immune system to exhibit outward signs of inflammation such as erythema, warmth, and purulent drainage. Look in the patient's throat and ears and assess for upper respiratory infection symptoms. Remember, common things being common, oncology patients are at risk for upper respiratory infections, otitis media, and pharyngitis just as any other child. Finally, evaluate for any evidence of mucositis secondary to chemotherapy, bearing in mind that mucositis may be present in genital and perirectal regions and not just the oral mucosa.

Laboratory evaluation should include a complete blood count with differential and a blood culture with sampling from all lumens of the central line, if present. Additional tests may be obtained at the discretion of the physician based on physical examination findings and in consultation with the child's specialty care provider. In addition, a venous blood gas and a serum lactate may be useful in evaluating the patient's metabolic state and severity of septic shock. Remember that shock is a clinical diagnosis, one that requires urgent intervention. No laboratory test should be relied upon to make the diagnosis and drive the initiation of therapy. It is important to bear in mind that chest radiographs may be falsely reassuring if the child is unable to mount an adequate immune response to show infiltrates. In this instance, the physician must rely on clinical suspicion based on physical examination findings to make the clinical diagnosis of pneumonia.

Antibiotics should be started without delay (Table 2). It cannot be stressed enough that antibiotics should not be withheld while awaiting laboratory results when septicemia is suspected! Ideally, antibiotics should be given within 1 hour or less of patient arrival to the ED. Early initiation of broad-spectrum antibiotics significantly reduces infection-related mortality in patients with chemotherapy-induced fever and neutropenia. Common organisms implicated in septicemia in patients receiving chemotherapy include gram-negative organisms such as Escherichia coli, Klebsiella pneumonia, and Pseudomonas aeruginosa and gram-positive organisms such as Staphylococcus aureus and streptococci. Patients with central lines are at especially high risk for infections with S aureus, coagulase-negative staphylococci, and viridians streptococci. Monotherapy with cefepime (50 mg/kg every 8 hours) or meropenem (20 mg/kg every 8 hours in children >3 months old) has proven to be good coverage for children with chemotherapy-induced neutropenia and fever. If these choices are not readily available, a \(\beta\)-lactam drug plus an aminoglycoside or a cephalosporin plus an aminoglycoside is a good alternative starting point. To avoid drug resistance, studies have shown that vancomycin (15 mg/kg) should not be administered empirically unless there are signs of hypotension, sepsis, or shock on evaluation, the

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**TABLE 2. Empiric antibiotic recommendations for fever and neutropenia in children.**

<table>
<thead>
<tr>
<th>First-line agents</th>
<th>Cefepime 50 mg/kg every 8 h, or Meropenem 20 mg/kg every 8 h (if &gt;3 mo old)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Alternative choices</td>
<td>(\beta)-Lactum plus aminoglycoside, or Cephalosporin plus aminoglycoside</td>
</tr>
<tr>
<td>Indications for vancomycin</td>
<td>Hypotension, sepsis, or shock Inflamed central venous access entry site Recent administration of high-dose AraC</td>
</tr>
</tbody>
</table>

Ara-C, cytarabine.
central line is inflamed, or the child has received high-dose arabinosylcytosine (AraC). It is acceptable and encouraged to discuss antibiotic choices with the child’s oncologist because regional variations in drug resistances and subtleties in patient history may influence the type of antimicrobial(s) chosen.

After the child’s vital signs have been stabilized and antibiotics have been administered, the child’s oncologist should be notified. The oncologist can often provide recommendations on continuing care and disposition, including admission to the local hospital or transfer to the tertiary care center for further evaluation. In the case of the child in the vignette, safe transfer to the tertiary care center is likely indicated.

**VIGNETTE, CONTINUED**

The child is given a total of 50 mL/kg of normal saline boluses with improvement in mental status, perfusion and vital signs. Laboratory evaluation is significant for an ANC of 310. He is given a dose of cefepime after a blood culture is obtained. The physician speaks with the oncologist on call at the tertiary care center who accepts the patient for transfer. The physician determines that the patient may be transported safely by a ground EMS team and updates the family.

**OTHER ONCOLOGY EMERGENCY CARE CONSIDERATIONS**

In addition to fever and neutropenia and associated risk of sepsis, other complications of chemotherapy that may be seen include severe anemia, bleeding, tumor lysis syndrome, disseminated intravascular coagulopathy, tumor compression, and other cardiopulmonary emergencies. Chemotherapy targets rapidly dividing cells, and therefore, depletes all blood cell lines, causing anemia and thrombocytopenia. Chemotherapy patients may require short-term transfusion of blood products. Peripheral neuropathy also is common in these children. Affected children may lack sensation, range of motion, and strength as a result. Chemotherapy-induced nausea and vomiting are perhaps one of the most common side effects of chemotherapy and arguably one of the most uncomfortable for the patient. Table 3 provides a quick reference of common side effects of specific chemotherapeutic agents.

<table>
<thead>
<tr>
<th>Chemotherapeutic agent</th>
<th>Side effect</th>
</tr>
</thead>
<tbody>
<tr>
<td>AraC</td>
<td>Cardiac failure, red urine, Pancreatitis</td>
</tr>
<tr>
<td>Asparaginase</td>
<td>Pulmonary fibrosis, fever</td>
</tr>
<tr>
<td>Bleomycin</td>
<td>Nephrotoxicity, ototoxicity, anaphylaxis</td>
</tr>
<tr>
<td>Carboplatinum</td>
<td>Nephrotoxicity, ototoxicity</td>
</tr>
<tr>
<td>Cisplatinum</td>
<td>Hemorrhagic cystitis, SIADH, infertility</td>
</tr>
<tr>
<td>Cyclophosphamide</td>
<td>Cardiac toxicity, red urine</td>
</tr>
<tr>
<td>Doxorubicin/daunomycin</td>
<td>Dermatologic changes, neurotoxicity</td>
</tr>
<tr>
<td>5-Fluorouracil</td>
<td>Hemorrhagic cystitis, SIADH, interstitial pneumonia, infertility</td>
</tr>
<tr>
<td>Ifosfamide</td>
<td>Allergic reaction, hepatotoxicity, diabetes, pancreatitis</td>
</tr>
<tr>
<td>L-asparaginase</td>
<td>Oral and gastrointestinal ulcers, hepatotoxicity, nephrotoxicity</td>
</tr>
<tr>
<td>Methotrexate</td>
<td>Hepatotoxicity, dermatologic changes</td>
</tr>
<tr>
<td>6-Mercaptopurine</td>
<td>Central nervous system effects</td>
</tr>
<tr>
<td>Procarbazine</td>
<td>Neurotoxicity, SIADH</td>
</tr>
</tbody>
</table>
| Vincristine/vinblastine| SIADH indicates syndrome of inappropriate secretion of antidiuretic hormone.

Finally, in addition to fever with neutropenia and complications of chemotherapy, the oncology patient may present to the ED with the complaint of abdominal pain. The oncology patient with abdominal pain may be receiving induction or maintenance proliferating, and treatment-responsive tumors. Tumor lysis syndrome occurs when large numbers of neoplastic cells are killed with the initiation of chemotherapy, leading to release of intracellular ions and metabolic byproducts into the systemic circulation. Clinically, the syndrome is characterized by rapid development of hyperkalemia, hyperuricemia, hypercalcemia, hyperphosphatemiania, and acute renal failure. The main principles of tumor lysis syndrome management are: (1) identification of high-risk patients with initiation of preventive therapy (new solid tumor diagnosis), and (2) early recognition of metabolic and renal complications with prompt supportive care. Aggressive intravenous hydration not only helps correct electrolyte disturbances by diluting extracellular fluid but also increases intravascular volume. Increased intravascular volume enhances renal blood flow, glomerular filtration rate, and urine volume, which aids the elimination of these cellular toxins.
Chemotherapy. They may also be in remission. The ED provider should begin the patient's assessment as they might for any child with a complaint of abdominal pain. This would include a complete history, thorough physical examination, and laboratory and/or radiographic studies (such as ultrasound or computed tomographic scan of the abdomen), as appropriate. In addition to evaluation for routine considerations, such as gastroenteritis, appendicitis, and trauma, the practitioner should also consider problems unique to this population such as an abdominal mass or leukemic infiltrate in the bowel. In other words, it is important not to minimize the complaint of abdominal pain in the oncology patient because it could indicate spread or recurrence of their disease.

A Few Words About Transfusions and Administration of Blood Products

In addition to oncology patients receiving chemotherapy, children with hematological disorders such as sickle cell disease, aplastic anemia, bleeding disorders, and platelet dysfunction may have depleted cell lines and require transfusion of red blood cells or platelets. Indications for transfusion of red blood cells include an acute blood loss of greater than or equal to 10% to 25% of the patient's blood volume, symptomatic anemia, oxygen requirement, asymptomatic anemia with bone marrow failure, and a hematocrit less than or equal to 18% to 20%. If time allows and the situation is not emergent, ensure that a type and cross-match is collected from the patient. When in doubt, it is a good idea to consult with the patient's hematologist or the blood bank before it can be transfused. For example, oncology patients or those who are immunocompromised require leukofiltered and irradiated blood, whereas patients with sickle cell anemia require matching of other specific blood antigens (such as Kell, C, and E). If packed red blood cells are needed emergently, as in a trauma patient or severe gastrointestinal bleed, it is best to give type-specific or O-negative blood, preferably after a type and cross-match is collected from the patient. When in doubt, it is a good idea to consult with the patient's hematologist or the blood bank before transfusing blood products.

As with adult care, pediatric blood product administration has several “golden rules” that physicians and nurses must follow. The first and foremost is education of a patient and family and informed consent. Families who have a child who has required blood products in the past are usually quite familiar with their child's disease and often have done research to understand the disease and therapeutic interventions. This being said, you can expect them to request laboratory results including blood counts and plan of care. Involving them upfront with all results will help them feel like a partner in their child's care. Parents may also offer insight on prior transfusions, any associated reactions, and need for pretreatment. Newly diagnosed patients and families will need a more thorough explanation of reasoning behind blood product administration before consent is signed.

When ordering packed red blood cells in pediatric patients, order the appropriate dose based on the patient's size. A standard unit of blood contains 250 to 300 mL. In general, a pediatric patient should be transfused 10 to 15 mL/kg of packed red blood cells. Platelets should be given to any patient who has a low platelet count and is actively bleeding or requires an invasive procedure. When transfusing platelets, 1 random donor pack raises the platelet count by 40,000 per 10 kg of weight. Platelets are also available in leukofiltered and irradiated options for immunocompromised patients; inquire in advance regarding the availability of these products in your institution. In general, blood product can remain unrefrigerated for no more than 4 hours and are typically ordered to be administered over 2 hours in children. This rate may be further reduced if necessary if there is a history of minor reactions to blood products or if a reaction is noted (see below). An exception to this is the actively bleeding patient (ie, trauma patient) who may require rapid infusion of blood products. Platelets, plasma, and cryoprecipitate typically infuse over 30 minutes. Blood products should be connected directly to the venous access catheter and never be “piggybacked.”

Monitoring blood product administration is most important over the first 15 minutes because this is when most reactions will occur. Obtain vital signs including temperature, heart rate, and blood pressure before the start of the infusion to establish the patient's baseline and watch for trends suggestive of a reaction. Reactions to blood products vary but generally begin with an alteration in temperature and are best initially detected in these patients by visual cues. Flushing, pruritus, dyspnea, chills, or rash may alert ED staff to stop the infusion and investigate further. Vitals signs may reflect fever, tachycardia, hypertension, or hypotension. Pain in the chest or back may also be present. When such a
reaction occurs, the infusion should be stopped immediately, and the blood tubing should be replaced with normal saline to keep the vein open. The ED physician must be notified immediately, and staff should follow hospital policy for infusion reactions. If a minor reaction occurs, a good rule of thumb is to slow the infusion to a quarter rate at first then back to full rate at 15 minutes if no additional reaction is noted. Flush the line with saline once the infusion is complete.

Immunoglobulin therapy initiation in patients with idiopathic thrombocytopenic purpura (ITP) can also be considered a blood product but is treated differently, as intravenous immunoglobulin (IVIG) is generally a more time-consuming therapy, one that usually involves inpatient admission. Factors 8 and 9 infusions are also in a different category because these factor replacements are achieved with quick push infusions and are often done at home by trained parents. Often, these patients will either go home immediately post infusion in the ED or not need to come to the hospital setting at all. Once again, it cannot be stressed enough the importance of speaking to a hematologist or other pertinent pediatric specialist before administration of these blood products to a pediatric patient.

**SUMMARY**

In conclusion, there are many special considerations in the care and treatment of the pediatric oncology and hematology patient. However, if a systematic approach is taken, the ED care provider should feel confident in his or her ability to choose the best course of treatment for this very special type of patient. Always regard the families of these patients as partners in emergency care and as vital sources of patient- and disease-specific information. Finally, timely consultation with the child’s specialty care provider will guide medical decision making in the ED and decisions regarding disposition.

**REFERENCES**


**Additional Reading**

Emergent Complications and Management of Children With End-Stage Renal Disease

Julie Phillips, MD, Jennifer Gaskins, RN, Laurie Lawrence, MD

You receive a call from the home of a 14-year-old boy who is undergoing peritoneal dialysis. He has chronic kidney disease secondary to posterior urethral valves. He has been complaining of abdominal pain and has a fever of 103°F (39.4°C). His mother called 911 because he continues to have a fever and has become less responsive.

The main function of the renal system is to eliminate excess water and waste from the body. In renal failure, patients lose the ability to remove toxins from the blood, control blood pressure, maintain the balance of fluids and electrolytes, and produce red blood cells. In end-stage renal disease (ESRD), there is complete or near-complete loss of renal function. End-stage renal disease is a rare but devastating cause of morbidity and mortality in the pediatric population. Common etiologies in this age group include congenital renal hypoplasia/dysplasia or obstructive uropathy, such as posterior urethral valves. Renal transplant is the modality of choice for renal replacement, especially in young children.
However, organ availability and technical difficulties in this patient population limit the feasibility for transplant. Children with ESRD have difficulty achieving normal growth. Because it is difficult for these patients to consume enough calories for adequate growth, they will often have a nasogastric tube or gastrostomy tube to provide supplemental nutrition.  

**PERITONEAL DIALYSIS**

Peritoneal dialysis (PD) is the renal replacement modality of choice while awaiting transplantation. There are 2 common types of PD, continuous ambulatory PD (CAPD) and automated PD. In CAPD, dialysis fluid is infused into the peritoneal cavity where it remains for 4 to 6 hours. During this time, it absorbs the body’s waste products. The fluid is then drained from the body in a procedure called an exchange. This is repeated 4 times a day. CAPD permits children to maintain a normal lifestyle. In automated PD, the dialysate solution is changed by a machine over 8 to 10 hours, typically overnight. During this time, the machine performs a number of cycles. Typically, the machine will fill the abdomen at the end of the night with dialysate that will remain in the abdomen until the next night.

In PD, the dialysis catheter is inserted into the peritoneal cavity and, therefore, cannot be used for vascular access. Ideally, the exit tunnel for the dialysis catheter is found outside of the diaper area, on the opposite side from a gastrostomy tube, and with a down-facing opening. The tunnel is positioned in this manner in attempt to minimize catheter-related infections. The dialysate fill volume is based on a patient’s body surface area. Children treated with PD have better control of blood pressure, acidemia, fluid balance, and growth as opposed to those children undergoing hemodialysis (HD).  

**HEMODIALYSIS**

In HD, blood is taken from the body, passed through a filtering system, and returned. The amount of blood flow required for HD is too great for a peripheral vein to handle. Initially, a central line may be used. For long-term HD, an arteriovenous shunt is created surgically in the patient’s arm. This connection between an artery and vein allows enough flow for HD to occur. Dialysis access limits the use of HD in pediatric populations.

Within the dialysis filtering system, blood flows through a series of membranes. Waste products pass through the membrane into the dialysate, while the blood is returned to the body. In addition to removing waste, excess fluid can be removed via ultrafiltration. Typically, a patient undergoes a 4-hour session of dialysis approximately 3 times a week. Hemodialysis is generally performed in a specialized medical facility.

**PERITONEAL DIALYSIS CATHETER-ASSOCIATED INFECTIONS**

Infectious complications of PD range from localized infection at the catheter exit site and along the subcutaneous catheter tunnel to peritonitis. The diagnosis of exit-site infections should be made when there is marked pericatheter swelling, redness, tenderness, or the presence of purulent discharge from the catheter sinus tract. A pathogenic organism may be cultured from the exit site. *Staphylococcus aureus* causes most of the exit-site infections. Other causative organisms include enterococci, *Escherichia coli*, *Pseudomonas*, Klebsiella, and other Gram-negative species. *Staphylococcus epidermidis*, although usually not causative of infection, is frequently cultured.  

Exit-site infections can progress to peritonitis when bacteria travel along the outer catheter surface into the peritoneal cavity. Peritonitis, although rare, is more common in children younger than 2 years. Historically, Gram-positive bacteria are responsible for most of the peritonitis in children undergoing PD. Gram-negative peritonitis occurs 20% to 30% of the time. Fungal infections account for approximately 3% of peritonitis episodes in the pediatric population with the most common species being *Candida*. Bacteria can seed the peritoneal cavity from several routes. Coagulase-negative *Staphylococcus* characteristically results from transcatheter introduction of bacteria, often from skin flora contamination. Pericatheter transmission occurs as an extension of an exit-site or tunnel infection with *S aureus*, a common pathogen. Bacteria may also be introduced enterically, hema
togenously, and rarely via ascending infection through the vagina.  

**HYPERKALEMIA**

End-stage renal failure places a patient at increased risk for hyperkalemia because of impairment of potassium excretion from the body. The normal range for serum potassium in children and adults is 3.5 to 5.5 mEq/L; in young infants, the upper limit of normal is as high as 6.5 mEq/L. When a child’s laboratory sample is obtained by heel or finger stick, a false elevation of serum potassium
may be reported because of hemolysis during collection of the blood; most laboratories will note the presence of hemolysis when results are reported. It is imperative to detect hyperkalemia because it can result in lethal cardiac arrhythmias by lowering the cell membrane’s resting potential. Hyperkalemia produces classic electrocardiographic (ECG) changes that correlate with the degree of potassium elevation. Progressive ECG changes seen in hyperkalemia begin with peaking of the T wave and progresses to disappearance of the P wave, widening of the QRS complex, ventricular tachycardia resembling a sinusoidal pattern, and finally, ventricular fibrillation. Patients at risk for the development of hyperkalemia must be placed on continuous cardiac monitoring, even during transport. In most clinical settings, an ECG can be obtained more quickly than a serum potassium level; in patients at risk for significant hyperkalemia, a 12-lead ECG should be obtained as soon as possible.

**HYPERTENSIVE EMERGENCIES AND URGENCIES**

Patients with renal failure are at increased risk for severe hypertension because of the integral role that healthy kidneys play in blood pressure regulation. In the pediatric age group, hypertension is diagnosed by comparing the individual patient's blood pressure to standard values for age and sex. The National High Blood Pressure Education Working Group on High Blood Pressure in Children and Adolescents has developed tables listing the blood pressure values in the 50th, 95th, and 99th percentiles. Severe hypertension is defined by pressures generally well above the 99th percentile for age and sex.

A hypertensive emergency is defined as a severe symptomatic blood pressure elevation with evidence of acute end-organ damage. Organs generally affected by acute elevations in blood pressure include the brain, eye, heart, and kidneys. Most commonly, patients will present with encephalopathy marked by confusion, lethargy, seizures, or coma. They may complain of visual symptoms due to the development of papilledema, retinal hemorrhages, and exudates. Pulmonary edema, tachycardia, hepatomegaly, and pedal edema may be present as the result of hypertension-induced congestive heart failure.

**VIGNETTE, CONTINUED**

EMS arrives on the scene and is lead to the patient by family. He is sleeping on the couch but wakes when his mother calls his name. His mother reports that he developed a fever this morning, and he has vomited 3 times. He has been on peritoneal dialysis for the last 20 months. On physical exam, he is febrile, his heart rate is 135 and his blood pressure is 96/62. His abdomen appears distended, and it is diffusely tender to palpation with guarding and rebound pain.

**EMERGENCY MEDICAL SERVICES EVALUATION AND TREATMENT**

The evaluation by emergency medical services (EMS) providers begins with a primary and secondary assessment at the scene, applying the principles of ABC (airway, breathing, circulation).

- **Primary Assessment**
  - Airway/Breathing—Patients with renal failure are at risk for volume overload. Watch for tachypnea or increased work of breathing.
  - Circulation—Assessing the patient's volume status is important. Before dialysis, patients are at risk for volume overload. These patients may have crackles, generalized edema, jugular venous distension, or hypertension. After dialysis, patients are at risk for hypovolemia, in which case the patient's skin may be cold and clammy with poor turgor. Check for tachycardia and hypotension.

- **Secondary Assessment**
  - Obtain a focused SAMPLE history from the caregivers. Ask caregivers what dry weight is used for the patient. A dry weight is an estimate of the patient's weight at a state of normal fluid balance. Dry weights should be used when calculating drug doses and intravenous (IV) fluid rates. Obtain information about the patient's dialysis schedule and any missed sessions. Ask caregivers if the patient makes urine. Patients in renal failure may retain up to 20% of normal renal function.

Many questions may arise when caring for patients with renal failure. Liberal use of online medical control is encouraged. Often, there is concern about administering fluids for fear of fluid overload. Patients with clinical evidence of hypovolemia should receive a fluid bolus. However, IV fluid administration in euvoletic patients should be limited. Furosemide may be helpful in fluid overloaded patients who still make urine. Patients in renal failure often require large doses of diuretics; it may be helpful to seek advice regarding diuretic therapy from medical control or the patient's nephrologist. In children with renal failure, it may be difficult to obtain IV access. If a child needs
access and an IV cannot be placed, consider placement of an intraosseous (IO) needle.

**VIGNETTE, CONTINUED**

En route to the emergency department (ED), 2 attempts at vascular access were unsuccessful. An IO was placed in the proximal right tibia, and a normal saline bolus was started. Upon arrival in the ED, he is placed in an acute care room. His temperature is 38.4°C. His heart rate is 120, and his blood pressure is 104/72. A sample of cloudy dialysate is obtained from his PD catheter. A cell count of the fluid shows 6300 white blood cells (WBC) per milliliter with 83% polymorphonuclear leukocytes and 2% bands. The Gram stain shows Gram-positive cocci in clusters. He is empirically started on vancomycin and cefazidime given intraperitoneally.

**ED NURSING MANAGEMENT**

Nursing care for the child with chronic renal failure is geared toward assessing, monitoring, and treatment of acute complications related to the disease process. In a community or general hospital setting, after initial stabilization, the patient should be transferred to a tertiary care center for definitive care.

Upon the patient’s arrival, assess for airway patency and ventilation quality, both of which may be altered related to fluid volume accumulation or excessive depletion. Tachypnea may be present because of fluid overload, anemia, and acidosis. Blood pressure may be too high or too low depending on fluid balance. Hypotension in a pediatric patient is a very late and worrisome sign of shock. Monitor for arrhythmias related to electrolyte imbalance and fluid overload. Finally, the patient may have an altered mental status because of impaired oxygenation, uremic toxin build up, hypertension, increased intracranial pressure, and other metabolic factors. Keen observation with very frequent (at least hourly) neurologic checks is necessary.

Place the patient on full cardiac and apnea monitoring, obtain peripheral vascular access, obtain blood specimens, and place a Foley catheter per physician orders. Anticipate orders for venous blood gas, complete metabolic panel, complete blood count, and blood and urine cultures when fever is present.

At times, when directed by the patient’s nephrologist, it may be necessary to obtain cultures from a peritoneal catheter and central vascular access devices. These devices should only be accessed by staff with appropriate experience with these devices and under the direct supervision of a physician or specially trained nurse.

Most parents of young PD patients have been trained to access their child’s PD site and change PD bags. If the family has a used dialysate bag, it may be sent for cultures when the patient presents with complaint of fever, abdominal pain, cloudy or fibrinous dialysate. If it is necessary to draw a specimen directly from the PD catheter, everyone in the room must use thorough sterile technique, donning mask, gown, and sterile gloves. Clean the end of the catheter with betadine, unscrew the cap and place it on a sterile field. Unclamp the PD catheter and allow fluid to drain into a sterile culture collection device without allowing the device to touch the PD catheter. The cap should then be replaced, and the PD catheter should be clamped.

Hemodialysis catheters should only be accessed in rare instances, for immediate life-saving measures when IV and IO access attempts fail. Everyone in the room should mask, and the procedure should remain sterile. Follow individual institution guidelines for central line specimen collection, keeping in mind that the patient’s central line is dwelling with a large amount of heparin and gentamycin, and a generous waste should be obtained before any flushing is done.

**PHYSICIAN EVALUATION AND TREATMENT**

**Peritoneal Dialysis–Related Infections**

In cases where catheter exit-site infection is suspected, a culture should be obtained from the site. Antibiotic treatment can be initiated after culture results are available unless there are signs of severe infection. A first-generation cephalosporin or a penicillinase-resistant penicillin with or without the addition of rifampin is a recommended antibiotic regimen for exit-site infections. The child’s subspecialty care provider should be consulted on antimicrobial selection. Vancomycin should not be used for the routine treatment of these infections because of emerging bacterial resistance. In Gram-negative infections, children older than 12 years should be treated with oral ciprofloxacin. Intraperitoneal cefazidime should be used in younger children. Drainage from the subcutaneous catheter sinus tract should be cleaned with nonalcoholic disinfectants. Povidone iodine solutions and hydrogen peroxide should be avoided; they can irritate the skin and may impair local host defenses. The exit site should be kept dry with a nonocclusive sterile dressing between dressing changes. Secure the catheter with a dressing to protect from trauma.
To evaluate for peritonitis, a sample of fluid from the PD catheter should be obtained. Regardless of dwell time, the initial sample from the PD catheter is ideal for evaluation if the fluid is cloudy. To maximize the bacterial recovery, a sample of 50 to 100 mL of the PD fluid should be obtained and concentrated in the laboratory before being cultured. A sample of the first cloudy bag of dialysate can be taken if antibiotics were initiated before obtaining a sample. If a sample can be immediately delivered to the laboratory for testing, it should be transported at room temperature. If delivery is going to be delayed more than 1 hour, the sample should be refrigerated, but not frozen. The sample should be sent for Gram stain, bacterial culture, fungal culture, and cell count and differential. In 20% of pediatric peritonitis cases, the culture is negative. The diagnosis of peritonitis should be made if the peritoneal effluent is cloudy with a WBC count of more than 100/mm$^3$ and a WBC differential of at least 50% polymorphonuclear leukocytes.

Once a sample is obtained, empiric treatment with intraperitoneal antibiotics should be started if the effluent is cloudy. Before the initiation of antibiotics, several flushes of dialysis solution may be performed to reduce the severity of abdominal pain. Antibiotics should be administered intraperitoneally to ensure immediate bioavailability. Antibiotics can be given via continuous or intermittent dosing. When intermittent dosing is used, a dwell time of at least 6 hours is necessary to allow adequate systemic absorption. It is recommended that full volume exchange of approximately 1100 mL/m$^2$ BSA be used for the antibiotic dwell. Again, it is advisable to consult with the patient’s kidney disease specialist on antibiotic choices and methods for intraperitoneal administration (Table 1).

Afebrile patients presenting with mild or absent abdominal pain and no risk factors for severe infection should receive a first-generation cephalosporin and ceftazidime. If there is a history of a prior methicillin-resistant S aureus infection, recent or current exit-site or tunnel infection, fever, severe abdominal pain, or if the child is younger than 2 years, initial antibiotic therapy should include vancomycin and ceftazidime. Aminoglycosides should be avoided in children because these drugs are nephototoxic and ototoxic. If fungi are identified on Gram stain or culture, IV amphotericin B or a combination of oral or intraperitoneal fluconazole and flucytosine should be added to the antibiotic regimen. Once the results of the dialysate culture are available, antibiotic therapies should be tailored.

In patients with extremely cloudy effluent, there is risk of occlusion of the catheter by fibrin. These patients may benefit from the addition of low-dose heparin into the dialysate fluid at a dose of 500 to 1000 U/L. An inflamed peritoneum can lead to

<table>
<thead>
<tr>
<th>TABLE 1. Intraperitoneal antibiotic dosing recommendations. $^3$</th>
</tr>
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<tbody>
<tr>
<td><strong>Continuous Therapy</strong></td>
</tr>
<tr>
<td><strong>Glycopeptides</strong></td>
</tr>
<tr>
<td>Vancomycin</td>
</tr>
<tr>
<td><strong>Cephalosporins</strong></td>
</tr>
<tr>
<td>Cephazolin/cephalothin</td>
</tr>
<tr>
<td>Cefuroxime</td>
</tr>
<tr>
<td>Cefotaxime</td>
</tr>
<tr>
<td>Ceftazidime</td>
</tr>
<tr>
<td>Ceftizoxime</td>
</tr>
<tr>
<td><strong>Antifungals</strong></td>
</tr>
<tr>
<td>Amphotericin B</td>
</tr>
<tr>
<td>Fluconazole</td>
</tr>
<tr>
<td><strong>Abbreviations:</strong> IV, intravenous; IP, intraperitoneal; PO, by mouth; q, every.</td>
</tr>
</tbody>
</table>
substantial loss of γ-globulin in infants with peritonitis. If these patients appear septic or have low serum immunoglobulin G levels, they may benefit from IV immunoglobulin therapy.

Management of Hyperkalemia

The goal of treatment in renal patients with hyperkalemia is to enhance potassium elimination. Unfortunately, unless dialysis is immediately available, elimination of potassium in these patients cannot be quickly achieved. To prevent lethal cardiac arrhythmias, interventions that act to stabilize the cell membrane and shift the potassium into the cell must be used as temporizing measures until definitive treatment can be initiated. Options for the early ED management of hyperkalemia are listed in Table 2.

Management of Hypertensive Emergency

Patients should be placed on continuous cardiac and oxygen saturation monitoring. The child’s neurologic status should be evaluated frequently. The blood pressure should be checked every minute during the administration of medications, either by automatic cuff monitors or the placement of an arterial line. The goal of treatment is to gradually lower the blood pressure to normal values. Rapid drops in blood pressure may result in poor tissue and central nervous system perfusion. The mean arterial blood pressure should be reduced no lower than 25% over the first 2 to 8 hours of treatment with the goal to normalize the blood pressure within 24 to 48 hours. In most cases, this requires an IV infusion of antihypertensive medication. The choice of a particular antihypertensive agent depends on several factors including the patient’s overall medical condition, capabilities of the treating facility, and consultation with the patient’s nephrologist. Table 3 lists IV antihypertensive agents found to be effective for the management of a hypertensive emergency in the setting of renal failure.

Hypertensive Urgency

Hypertensive urgency is defined as a marked elevation in blood pressure without severe symptoms or evidence of acute end-organ damage. The mode of treatment is determined by the duration of the blood pressure elevation. Patients who have developed a gradual rise in blood pressure over the course of days or weeks can be managed with oral medications. The treatment goal with these patients is to gain control of the blood pressure over the course of 1 to 2 days. Patients with rapid elevations of blood pressure or those unable to tolerate oral

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**TABLE 2. Treatment of hyperkalemia.**

<table>
<thead>
<tr>
<th>Shifting potassium from extracellular to intracellular fluids:</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Sodium bicarbonate</td>
<td>1-2 mEq/kg per dose over 5-10 minutes (Use even in the absence of acidosis)</td>
</tr>
<tr>
<td>Glucose and insulin</td>
<td>0.5 g/kg (2 ml/kg) dextrose 25% and water (D25W) and 0.1 unit/kg regular insulin IV (Monitor glucose hourly)</td>
</tr>
<tr>
<td>Albuterol nebulization</td>
<td>0.5% solution; 0.15 mg/kg; max dose 5 mg</td>
</tr>
</tbody>
</table>

Stabilization of cell membrane (use in setting of ECG changes):

| Calcium chloride 10% | 10-30 mg/kg/kg IV per dose (0.1-0.3 ml/kg per dose) slowly at 0.5-1 ml/min; repeat pm up to 5 ml (Caution: stop infusion if bradycardia develops) |
| Calcium gluconate 10% | 100 mg/kg/dose (1 ml/kg/dose) over 3-5 min, Repeat in 10 minutes |

Increasing elimination of potassium:

| Kayexalate | 0.5 -1 g/kg PO or PR with retention enema |
| Dialysis |  |

**TABLE 3. Intravenous antihypertensive agents used for the treatment of hypertensive emergencies in renal failure.**

<table>
<thead>
<tr>
<th>Drug</th>
<th>Action</th>
<th>Dosage</th>
<th>Onset</th>
<th>Duration</th>
</tr>
</thead>
<tbody>
<tr>
<td>Esmolol</td>
<td>β-blocker</td>
<td>Loading dose: 100 500 μg/kg over 1 min then 25 200 μg/kg/min IV infusion</td>
<td>Immediate</td>
<td>9-30 min</td>
</tr>
<tr>
<td>Nicardipine</td>
<td>Calcium-channel blocker</td>
<td>0.5-3 μg/kg/min IV</td>
<td>&lt;1 min</td>
<td>30-60 min</td>
</tr>
<tr>
<td>Labetalol</td>
<td>α- and β-blocker</td>
<td>0.2-1.5 mg/kg/h IV or 0.2-1 mg/kg IV bolus; Max 20 mg single dose</td>
<td>2-4 min</td>
<td>Up to 4 h</td>
</tr>
<tr>
<td>Fenoldopam</td>
<td>Dopamine receptor agonist</td>
<td>0.2-2 μg/kg/min IV infusion</td>
<td>5-15 min</td>
<td>8-10 min</td>
</tr>
<tr>
<td>Enalapril</td>
<td>Angiotensin-converting enzyme inhibitor</td>
<td>5-10 μg/kg IV bolus; Max 1.25 mg/dose</td>
<td>Up to 60 min</td>
<td>4-6 h</td>
</tr>
<tr>
<td>Phentolamine</td>
<td>α-adrenergic blocker</td>
<td>0.05-0.1 mg/kg IV; Max 5 mg/dose</td>
<td>Immediate</td>
<td>30-60 min</td>
</tr>
</tbody>
</table>
medications may require IV medications following the same goals as outlined in the treatment of hypertensive emergencies. Table 4 outlines the oral antihypertensive agents recommended for the treatment of hypertensive urgency.

### SUMMARY

It is important to have basic knowledge of ESRD in children and dialysis and kidney disease–related complications. Vigilance for fluid and electrolyte abnormalities and infection are key components of medical screening in the ED. Also important is familiarity with the signs of hypertensive urgencies and emergencies and their initial management. Parents and caregivers of special-needs children typically serve as excellent sources for relevant information regarding these children, including management advice. Finally, timely communication with pediatric primary or subspecialty care providers of medically complex children will also serve to inform care.

### REFERENCES


### OTHER RESOURCES


A 1-year-old girl is transported by emergency medical services to the emergency department (ED) with a 1-day history of fever, increased fussiness, and decreased intake. Her medical history is significant for gastroschisis and multiple intestinal atresias, after bowel resection and short bowel syndrome (SBS); she is currently on the bowel transplant list. She has failure to thrive and is currently on total parenteral nutrition (TPN) and oral and gastrostomy tube (G tube) feeds. The child's father states that he started her TPN on the evening before admission as per usual without problem. The following day, the father noted her to be fussy, and she had decreased oral intake. Per parents, the TPN was not running and had not been running for the last 20 hours. A finger-stick blood glucose was 146 mg/dL. The parents fixed the pump, and the TPN was resumed. On a repeat finger-stick blood glucose, her glucose had increased to 315 mg/dL and her TPN was stopped. Her gastroenterology specialist was contacted and recommended that the TPN should be continued and to have TPN laboratory tests redrawn in the morning. She developed a fever to 102°F later that night and was taken to the ED for fever, chills, and decreased activity concerns. The parents also noted that she had increased watery voluminous stool production. She had tolerated her G tube feeds well without vomiting or abdominal distention. The parents also reported decreased urine output. Although a slight dry cough started the day of presentation, she had no respiratory distress or nasal discharge.

In addition to the above, her medical history is notable for multiple line infections. She also has a history of a superior vena cava...
The patient was felt to be dehydrated, and a normal saline 20 mL/kg bolus was initiated after laboratory studies were obtained. A bedside glucose was 91 mg/dL. A complete metabolic panel was obtained, and significant values included the following: sodium, 126 mEq/L; chloride, 100 mEq/L; CO\(_2\), 9 mEq/L; blood urea nitrogen, 30 mg/dL; and creatinine, 12 mg/dL. These values were consistent, with an impression of dehydration and metabolic acidosis. Her liver enzymes were mildly elevated with an aspartate aminotransferase of 67 U/L and alanine aminotransferase of 128 U/L; this was felt to be due to the TPN. The rest of the complete metabolic panel was normal. The fever workup included blood cultures from the central line and peripheral vein with a complete blood count (CBC) and a catheter-obtained urine analysis and culture. The CBC showed a white blood cell count of 19 800/mm\(^3\), with a differential of 90% granulocytes, 7.9% lymphocytes, 2% monocytes, and 0.1% eosinophils. The hemoglobin level and hematocrit were normal, and the platelet count was decreased to 120 000/mm\(^3\). The erythrocyte sedimentation rate was normal at 9 mm/h, and the C-reactive protein was mildly elevated at 2.4 mg/L. The urine analysis was significant for 1+ ketones and a specific gravity of 1.030 or greater, with a moderate amount of bacteruria.

After the initial bolus, her heart rate improved to 130 beats/min, and after a second bolus, there was a further decrease to 120 beats/min. Vancomycin and Rocephin (Roche; USA) were given empirically, and the patient was admitted to the gastroenterology service.

The complex nature of this patient’s medical history exemplifies the need for an understanding by emergency care providers of the complications associated with pediatric gastroenterology conditions and the adjuncts used to promote patient survival.

**ENTERAL FEEDING TUBES**

Recent advances in the medical and surgical management of pediatric patients with chronic medical conditions have resulted in many children receiving nutritional support from surgically or endoscopically placed enteral feeding devices. The most common devices are the G tube, jejunostomy tube, and gastrojejunostomy tube (GJ tube). The G tube is placed through a surgically or endoscopically created stoma and provides access to the stomach from the level of the skin. It is used in children who are unable to take adequate oral intake for prolonged periods. Jejunostomy tubes are surgically created stomas that bring the jejunum to the skin. They are used when postpyloric feeding is required, such as in patients with delayed gastric emptying and recurrent aspiration pneumonia. The GJ tube is becoming more common. It has an outward appearance similar to the conventional G tube, but in addition to the gastric port, it has a long extension on the inside that travels transpyloric and into the jejunum.

Emergency care providers practicing in an emergency medical services, acute care, or ED setting must become comfortable handling the many complications that can occur with these devices. Problems with enteral feeding tubes can be divided into 2 groups, tube-related mechanical problems, and problems with the stoma.

When considering tube-related problems, dislodgement is one of the most commonly occurring. Dislodgement of the tube is typically caused by accidental trauma leading to tension on external tubing, occult balloon deflation, or rupture of the balloon. Patients who present with tube dislodgement can have either a benign stoma or active bleeding from the stoma secondary to trauma. In these cases, the goal is to keep the stoma open and...
operational with the least amount of trauma. This can be achieved by providing a temporizing measure with a Foley catheter until a permanent replacement can be obtained. For successful placement of a G tube, the interval since dislodgement is crucial. As postdislodgement time increases, the risk of the stoma constricting and closing increases. Multiple dilations with a series of Foley catheters advancing to the original size of the enteral feeding tube may be required. Another important factor to consider when replacing enteral feeding tubes is the interval since the initial placement. Tubes dislodged in the initial perioperative period (usually defined as within 1-3 months of initial placement) should be gently reinserted as soon as possible by a trained health care provider. In these early cases, a mature fistula tract has not yet developed, and the surgeon or gastroenterologist who originally placed the tube would ideally be involved in the replacement. At a minimum, surgical consultation should be obtained before resuming enteral feeds, and a radiologic contrast study should be performed to verify correct position within the stomach.

Conversely, replacement of a tube with a mature (not in the immediate perioperative period) tract does not require radiology confirmation as long as the tube is replaced into the gastrostomy site without resistance, enteral feeds flow into the stomach by gravity, and gastric contents can be aspirated to confirm correct position. Tubes dislodged for a long period often require serial dilatation with Foley catheters of 1 or 2 sizes smaller. One common technique for tube replacement is to use the wooden end of a cotton swab as a stylet in the opening of the G tube while gently applying consistent downward force until the tube is in correct position. Care must be taken not to inadvertently create a false track or cause stomach perforation. The provider must use caution when reinserting a G tube because excessive force or inattention can lead to tube insertion into the peritoneal cavity through a false tract.  

Gastrojejunostomy tubes that dislodge must be replaced under fluoroscopy. If no GJ tube is available, inserting a G tube to keep the stoma open would be an option, realizing that the usual feeding cannot be resumed until the GJ tube is replaced.

In addition to dislodgement, other minor tube-related complications can occur. These include gastrostomy leakage, excessive tension on the tube, and skin erosion. Gastrostomy leakage can occur for a variety of reasons, but it is most commonly due to improper tube fit or inadequate volume of water in the balloon. Enlargement of the stomal site due to poor wound healing may also occur, leading to gastrostomy leakage. In this instance, replacement with a Foley catheter at least 2 sizes smaller for 24 to 48 hours may allow the stoma to contract. In addition, the enteral feeding tube may be entirely removed for several hours, allowing constriction of the stomal tract.

Infants and children with poor gastric motility and small stomach sizes can experience gastric distention with high-volume bolus enteral feeds and have leakage of formula. By changing to low-volume continuous feeds, this problem will typically resolve. Leakage coming from the opening of the G tube but not around the tube indicates malfunction of the 1-way valve and warrants replacement of the device.

Finally, skin erosion due to excessive tension or leakage from the gastrostomy is also commonly seen. Zinc oxide ointment provides a good barrier to protect against constant moisture and acidity. Another barrier topical, Coloplast Critic-Aid Moisture Barrier Cream (Minneapolis, MN), composed of Calmoseptine cream, a zinc oxide cream and menthol mixture, is used in some institutions to promote skin healing.

The development of granulation tissue at the insertion site is another common problem. The etiology for this tissue has yet to be definitively identified but is most likely due to a multifactorial foreign body reaction. Chemical cautery with silver nitrate applicator sticks daily or every other day is the traditional treatment. An alternative approach is the application of triamcinolone cream (3 times daily). Prolapse of gastric mucosa at the gastrostomy site, a rare problem, can be difficult to distinguish from granulation tissue and may require surgical consultation and operative intervention.

Gastrostomy stomas rarely become infected, and antibiotic creams are not usually needed. Infection, if present, tends to occur in the immediate postoperative period and may present with anything from a small superficial infection to frank cellulitis of the site. Satellite lesions reflecting Candida infection should be treated with nystatin.

Two other infrequent but major complications of enteral feeding tubes that need to be considered are tube misplacement into the peritoneal cavity and balloon occlusion of the pylorus, duodenum, or more distal bowel. These present with signs of peritonitis and bowel obstruction, respectively.

**INTRA-ABDOMINAL WALL DEFECTS**

The term *gastrochisis* was coined in 1894 to describe conditions in which the abdomen remains open at birth. It is an intrauterine birth defect where there is herniation of the bowel outside the abdominal wall, usually to the right of the umbilicus. The intestines are thickened and matted with no
overlying membrane. Gastroschisis is typically associated with a 10% risk of other congenital anomalies. For reasons that are poorly understood, the incidence of gastroschisis has been increasing since the 1940s. Typically, affected infants are diagnosed prenatally, and plans for delivery at a tertiary care center can be arranged. However, on the rare occasion an infant with such a defect has a precipitous delivery at home or en route to the hospital, emergency care providers should be aware of this condition. Resuscitation of the infant should proceed as usual, with the additional consideration of providing optimal handling of the viscera. Ideally, this should be done by placing a transparent sterile dressing or sterile plastic bowel bag or Clingfilm over the defect, stabilizing it over the middle of the abdomen with the infant supine. The dressing should encompass the entire torso, abdomen, and lower extremities and be loosely secured around the chest just above the nipple line; this will allow direct visualization of the bowel during transport. Bowel congestion can then be promptly identified and any kinking in the mesentery alleviated by repositioning, usually with the child in a right lateral decubitus position. Ideally, an 8- or 10-Fr orogastric decompression tube should be placed, and intravenous (IV) access should be obtained with liberal IV fluid hydration. If bowel is exposed, it may take up to 200 to 300 mL/kg/day or rehydration. In addition, these children typically require coverage with broad-spectrum antibiotics. Omphalocele is another congenital abdominal wall defect. It consists of a central defect in the umbilical ring that allows herniation of the viscera into the umbilical cord, resulting in a membrane-covered defect. Omphalocele appears to have a lower incidence and is associated with advanced maternal age. It carries a higher mortality rate than gastroschisis, probably related to the higher incidence of associated birth defects, which range from 50% to 70%. As with other intra-abdominal wall defects, most of these infants are diagnosed prenatally and are delivered at a tertiary care center. In contrast to gastroschisis, the omphalocele has a membranous covering, and a dressing with non-adherent gauze to prevent injury or desiccation is usually all that is needed. If the omphalocele is ruptured, then urgent bowel coverage is needed. The clinician should be familiar with the commonly associated defects in affected infants, particularly Beckwith-Wiedemann syndrome (hypoglycemia), increased incidence of malignancy (Wilms tumor, neuroblastoma, and hepatoblastoma), cardiac defects, and chromosomal anomalies, so as to expedite evaluation and treatment of associated problems. Complications related to the surgical repair of these intra-abdominal wall defects may occur. Of primary importance is the complication of small bowel obstruction (SBO). Although an SBO can occur anytime, it appears that most cases occur in the first year or two postoperatively. A study conducted by Van Eijck et al found that 85% of SBO occurred in the first year of life. In addition, a history of sepsis and fascial dehiscence were noted to be highly predictive risk factors for the development of a SBO.

Hirschsprung disease, although not an abdominal wall defect, also deserves mention. It is a condition in which the parasympathetic ganglion cells of Auerbach plexus between the circular and the longitudinal muscle layers of the colon are absent. These children present with a long history of constipation. They usually seek urgent/emergent care with explosive diarrhea and colitis. Although a detailed discussion on the diagnosis, immediate management, and postoperative treatment is not appropriate at this time, emergency care personnel caring for these children should be aware of the risk for enterocolitis in the presence of abdominal distention and the associated morbidity and mortality, so that appropriate treatment may be initiated rapidly.

**TRANSPLANTATION EMERGENCIES**

Pediatric solid organ transplantation, which began in the 1960s, has now become a realistic option for organ failure. The introduction of calcineurin inhibitors for immunosuppression has improved the management of graft rejection. With refinements in surgical technique, medical management, and recognition and prevention of long-term complications, 80% of pediatric patients survive into adolescence and young adulthood. Transplantation essentially replaces end-stage organ disease with a more viable, chronic disease process, which has its own set of complications. These next few paragraphs will focus on mainly liver and small bowel transplantation, or combined liver/small bowel, looking at the pretransplant, peri-transplant, and post-transplant factors involved.

The pediatric patient awaiting liver-bowel transplant will frequently seek emergency care related to impending or fulminate liver failure. Liver failure can develop acutely or be chronically progressive. It can present with coagulopathy, hypoglycemia, hyperbilirubinemia, hypoprothrombinemia, and encephalopathy. Symptoms of liver failure can be vague and may include fatigue, nausea and vomiting, and diffuse abdominal pain. Patients often present with...
jaundice. Disease progression will result in worsening jaundice, tremors, lethargy, confusion, easy bruising, and ultimately, encephalopathy. Cerebral edema is associated with significant mortality and must be treated aggressively. Increasing portal hypertension may rapidly lead to bleeding esophageal and gastric varices and ascites. Emergency treatment of bleeding varices in the pediatric patient may be treated by placing a Foley in the stomach, blowing up the balloon, and applying traction, as opposed to the Sengstaken Blakemore tube used in adults. Laboratory evaluation is needed to provide definitive evidence of liver failure. These studies should include serum transaminases, CBC, coagulation profiles, bilirubin levels, albumin, and glucose. Hypoglycemia almost always accompanies liver failure and must be treated rapidly and aggressively. Metabolic acidosis, which occurs in about 30% of patients, will need to be corrected. Patients with ascites and hyponatremia require their serum sodium to be corrected slowly but rarely need anything more than a diuretic to achieve a gradual change. If respiratory or renal involvement is severe, ascites can be treated more aggressively with direct paracentesis or IV 25% albumin (1 mg/kg) followed by IV furosemide.

Children represent a disproportionate number of candidates on the Organ Procurement and Transplantation Network and Scientific Registry of Transplant Recipients’ annual report when it comes to bowel transplantation. The mortality rate is the highest in patients with combined liver and bowel transplant. Up to 70% of pediatric patients on the waiting list develop liver failure related to complications from long-term TPN and ultimately require a combined liver-intestinal transplant.

Emergency evaluation of the posttransplant pediatric patient is usually directed by an acute problem that arises from either the surgical procedure or medical complications. These can manifest as fever, graft dysfunction, pain, or drug toxicity. One of the most common problems with the combined intestine/liver transplant is graft vs host disease from the donor intestine. A recently published article by Ngô et al documented independent factors that predicted 6-month patient and graft survival after pediatric liver transplant. They concluded that the most significant factors predicting patient and graft loss are posttransplant surgical complications. Of the many medical problems that may cause a pediatric patient with transplant to seek care in an ED, medically induced immunosuppression probably plays the most significant role. A detailed discussion of the many medications being used for immunosuppression is beyond the scope of this article. The risk for infectious complications cannot be overemphasized. Both conventional and opportunistic infections are possible in the posttransplant patient. The level of immunosuppression, immunization status and the pretransplant health of the child all play a role in guiding the ED evaluation of these patients. All symptoms in the transplant patient require investigation regardless of how trivial they appear. Fever especially deserves an aggressive investigation and can result from a myriad of infections or acute graft rejection. Immunosuppressive medications such as corticosteroids have potential adverse effects in addition to the obvious effects on the immune system. Patients on calcineurin inhibitors are at risk for nephrotoxicity, which can be exacerbated by concurrent gastroenteritis and dehydration or low cardiac output. An accurate blood pressure must be recorded on all transplant patients because the combined use of steroids and calcineurin inhibitors poses a significant risk of hypertension. The posttransplant patient with a headache deserves special mention considering the increased risk of meningitis, pseudotumor cerebri, malignancy, and malignant hypertension.

Posttransplantation lymphoproliferative disorder (PTLD) is noted to be more common in pediatric patients with liver transplant as compared with their adult counterparts. Posttransplantation lymphoproliferative disorder represents a wide disease spectrum resulting from immunosuppression and is associated with Epstein-Barr virus infection. It is suspected that children are more susceptible due to their naïve immunity against Epstein-Barr virus. Posttransplantation lymphoproliferative disorder can present with signs and symptoms ranging from a simple polyclonal lymphocyte expansion resembling infectious mononucleosis to an aggressive, life-threatening lymphoma.

Although rare, patients status post liver transplant are at increased risk for small bowel obstruction (SBO), a serious complication. In the nontransplant patient, adhesions represent the cause for most SBO. However, in the posttransplant patient, a small study demonstrated that no cases of SBO were due to adhesions. Rather, right-sided diaphragmatic hernia and PTLD were the cause of the obstruction. One patient presented with intussusception.

Another entity that needs to be recognized as a complication of pediatric liver or combined liver and small bowel transplant is autoimmune hemolytic anemia (AIHA). Thought to be quite rare, a recent study by Li et al noted that AIHA may occur in patients with solid organ transplant at a much higher frequency than previously believed.
They noted that hemolysis can occur many years after transplantation and is often severe and resistant to steroid treatment. These findings were consistent with the retrospective study of Botija and colleagues, published in 2009, citing an AIHA incidence of 12.2% in 49 pediatric patients with intestinal transplant.

Neurologic problems after liver and combined liver-bowel transplantation appear to be seen more frequently in pediatric patients (up to 46%) as compared with adults. Neurologic problems identified in decreasing order of presentation include seizures, encephalopathy, and posterior reversible leukoencephalopathy. First described in 1996 by Hinchey et al., posterior reversible leukoencephalopathy is characterized by the radiologic finding of bilateral gray and white matter edema in the posterior regions of the cerebral hemispheres. Central nervous system infections, cerebrovascular events, peripheral neuropathies, tremors, blurry vision, auditory hallucinations, and choreoathetosis have also been reported. The recent article by Fernandez et al. is the largest pediatric study looking at neurologic problems after liver and combined liver-bowel transplantation. They found a lower mortality in these patients when compared with previously reported studies.

Despite the short- and long-term complications listed, recipients with solid organ transplant usually have good long-term psychosocial outcomes with improved quality of life in comparison with the pretransplant period and in relationship to those patients with chronic disease.

**SHORT BOWEL SYNDROME**

Short bowel syndrome is a functional definition of a significant malabsorptive state occurring after massive resection of the small intestine or congenital intestinal anomalies. Common etiologies in the infant requiring resection leading to SBS include necrotizing enterocolitis, gastrochisis, volvulus, and atresia. The extent of SBS is variable, depending on the extent and segment of resected small intestine, patient age, and the adaptive response of the small intestine to the resection. Initially after the bowel resection, the patient will require parenteral nutrition. The length of the residual small bowel, the absence of the ileocecal valve, and successful bowel adaptation influence weaning from parenteral nutrition and transitioning to enteral feeds. If adaptation to enteral feeds and independence from parenteral nutrition are not achieved, intestinal failure occurs.

Watery diarrhea is a common complication seen when the osmotic load in the lumen of the small bowel exceeds the absorptive capacity. This occurs commonly during advancement of the enteral feeds. Electrolyte imbalances can occur, especially hyponatremia. The output of diarrhea can be large and can result in dehydration. Immediate resuscitation in the ED includes rehydrating the patient with normal saline if dehydration is present and identifying and correcting electrolyte abnormalities.

Further management may include antisecretory agents such as somatostatin to slow gut transit. Cholestyramine may help to decrease the effects of secretory diarrhea related to bile acid malabsorption. Dietary changes to low-carbohydrate, high-fat enteral formula to decrease the osmotic load, and switching from bolus feeds to continuous enteral feedings may be beneficial. Gastric hypersecretion, which is commonly seen after the small bowel resection, may lead to increased fluid production and losses; therefore, acid suppressive medications are commonly used.

Patients with SBS who require prolonged parenteral nutrition are at risk for multiple complications. Central venous or peripherally inserted central catheter–related problems include catheter breakage and occlusion. Catheter repair kits should be available to be used by knowledgeable staff. Catheters that have clots and cannot be accessed may benefit from clot dissolving agents to restore patency. Careful handling, protection of the line in an active child, and flushing protocols for catheter line patency are useful preventative measures.

Catheter line infections are common and recurrent in infants with small bowel syndrome. In addition to poor venous access technique, bacterial translocation may be a factor. A recent pilot study suggests that small bowel bacterial overgrowth may increase the risk in recurrent line infections. Catheter line sepsis can be life threatening. When evaluating fever in children with small bowel syndrome and an indwelling catheter, a blood culture from the catheter and one from a peripheral vein should be obtained and broad-spectrum antibiotic coverage should be initiated. Patients should be carefully evaluated for sepsis and treated with fluid resuscitation and advanced pediatric life support as needed. Prevention of catheter line sepsis is important with a good sterile technique. In a retrospective study, Bishay et al. found a decreased incidence of sepsis with the use of chlorhexidine for central venous catheter antisepsis in infants after surgical repairs that required parenteral nutrition. Patients with SBS who require prolonged parenteral nutrition are at risk for developing
liver disease and cholelithiasis. Parenteral nutrition–associated liver disease (PNALD), also known as parenteral nutrition–associated cholestasis, occurs in 40% to 60% of infants with SBS that are TPN dependent. Risk factors for PNALD include prematurity, lack of enteral feedings, duration of TPN, and recurrent sepsis. Parenteral nutrition–associated liver disease can be progressive to liver failure and cirrhosis with continued parenteral nutrition. Cholestasis is present with a serum direct bilirubin concentration greater than 2 mg/dL. When evaluating a patient with small bowel syndrome and PNALD, ensure a direct bilirubin is obtained in addition to a complete metabolic panel, γ-glutamyltransferase and prothrombin time. Ursodeoxycholic acid may reduce gallbladder stasis and improve bile flow. The incidence of biliary sludge and gallstone formation increases with duration of parenteral nutrition. Cholecystokinin did not show any reduction in development of PNALD in a randomized double-blind controlled trial. A small amount of enteral nutrition may be helpful to stimulate the gallbladder.

Patients with SBS are at an increased risk of developing small bowel bacterial overgrowth. Contributing factors to development of bacterial overgrowth include no ileocecal valve, which allows reflux of bacteria from the colon into the small intestine, and progressive dilatation of the small intestine because abnormal intestinal peristalsis exists. The bacterial overgrowth causes an inflammatory response, which damages the absorptive surface that results in malabsorption and protein loss. The breath hydrogen test is used to diagnose bacterial overgrowth. Clinical manifestations of small bowel bacterial overgrowth include weight loss and abdominal distention, which leads to colitis. Antibiotic treatment is effective for control of small bowel bacterial overgrowth but may require cycling of dosing and antibiotic class.

Neurologic symptoms of ataxia and altered mental status with a metabolic acidosis can occur uncommonly with accumulation of D-lactic acid in patients with SBS and bacterial overgrowth. After ingestion of a large carbohydrate load, carbohydrate malabsorption and delivery of nutrients to the colon in association with the bacterial overgrowth produce D-lactic acid. D-Lactate is normally excreted in the urine. D-Lactic acid can be normally present in the serum of patients with small bowel syndrome without acidemia. Levels of D-lactate were found to be higher after meals in adults and children with small bowel syndrome. Normal or elevated L-lactic acid levels may be present. The standard serum lactate level (L-isomer) should be evaluated in a patient with small bowel syndrome and metabolic acidosis. With multiple episodes of acidosis or signs of encephalopathy, obtaining a D-lactate level should be considered. The etiology of these neurologic symptoms is unclear but is hypothesized to be due to the innate D-lactate toxicity to the brain and the unknown compounds associated with D-lactate production. Treatment modalities include recolonization of the intestine through the oral administration of a bacterial flora, low-carbohydrate diets, and rectifying acidosis with bicarbonate or through hemodialysis.

After parenteral support is decreased or discontinued, patients with SBS are at risk for macronutrient and micronutrient deficiencies. The most common deficiencies are fat-soluble vitamins, especially A, D, and E. Zinc, iron, magnesium, and selenium need to be monitored as well because of the risk for deficiencies. Oral supplementation is implemented simultaneously. However, even with multivitamin supplementation, one study found that almost 50% of patients developed at least 1 biochemical abnormality. Vitamin B12 deficiency is common with an ileal resection. Osteoporosis can occur with the vitamin D and magnesium and calcium deficiencies, and serum phosphorous and calcium levels need to be monitored.

Patients with SBS have an increased risk of renal stone formation secondary to hyperoxaluria, lower urine volumes, larger stool volumes, and hyponatremia. Treatment is variable with diet and cholestyramine. After small bowel resection, patients can develop strictures at anastomotic sites and bowel obstruction from adhesions or develop ulcers at anastomotic sites that can bleed.

**SUMMARY**

The number of pediatric patients with gastrointestinal disease who survive with improved medical management and high technical adjuncts continues to increase and will likely be encountered in the ED. Complications resulting from these disease processes and hardware are varied in scope and difficulty due to the wide variety of pathophysiology. Simple dislodgement of a gastric tube, posttransplantation fever, or electrolyte abnormalities in a patient with SBS to life-threatening septic shock may occur. A common simple complaint can be indicative of more serious problems in this high-risk population. Rapid recognition and an understanding of the medical needs of these patients at risk can help streamline their evaluation and improve treatment and care outcomes.
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Emergency Care of Children with High-Technology Neurologic Disorders

Courtney Horton, MD,
Lindsay Byrd, RN, BS,
Heather Lucht, RN, BSN,
Nicholas Higby, MD

VIGNETTE

A call comes in at 9:15 AM to assist a 4-year-old girl with a history of a brain tumor, status post–tumor resection, and ventriculoperitoneal shunt placement. She had a hemorrhagic stroke during the resection and is now neurologically devastated. Her mother called 911 because the patient has been vomiting with decreased activity all morning.

HYDROCEPHALUS

Hydrocephalus literally means “water on the brain.” Patients become symptomatic from hydrocephalus when an increase of cerebrospinal fluid (CSF) volume exerts pressure on the brain. Normally, CSF is produced and naturally reabsorbed by the body at a constant rate. There are several pathophysiologic mechanisms that can lead to a disturbance in the amount of CSF. Certain tumors actually cause an increase in the production of CSF; other tumors or brain malformations can impair or block...
CSF drainage. Infections and hemorrhage inside the skull can also block CSF drainage. Regardless of the cause of the increased CSF, the result is increased pressure on the brain that needs to be corrected, typically with a shunt.\textsuperscript{1,2}

**CSF SHUNTS—BACKGROUND**

A CSF shunt is a conduit to drain excess fluid in patients with hydrocephalus, typically to the peritoneum, pleural space, or heart (see Figure 1). A CSF shunt has 4 basic components; these include the proximal end, which is placed in the ventricle; the valve that allows for 1-way flow of CSF; the reservoir that can be accessed to sample CSF and check shunt pressure; and the tubing tunneled beneath the skin that carries the fluid to its end point. If the end point is the heart, the shunt is referred to as a ventriculoatrial shunt. If the end point is the pleural space, this is a ventriculopleural shunt. If the end point is the abdomen, the most common CSF shunt placed, it is referred to as a ventriculoperitoneal (VP) shunt. This is a terminology that the EMS provider will need to be comfortable with when you discuss the patient's shunt with his/her caregiver.\textsuperscript{1-3}

**SHUNT COMPLICATIONS**

Emergency care providers should be aware of common shunt complications because they can be life threatening. One relatively rare, although important, complication is shunt infection, which typically occurs in the first few months after shunt placement or revision, with an overall incidence of 0.3%. The most common organisms responsible for shunt infections are *Staphylococcus epidermidis* and *Staphylococcus aureus*. A shunt infection should be considered in any patient with a CSF shunt and fever, but as time progresses from the time of placement or last revision, it becomes less likely. Another major complication is a shunt malfunction or obstruction, which leads to a partial or complete blockage of CSF outflow. There are different reasons a shunt can become obstructed. Some examples include the tubing becoming blocked by debris or cells, a break or kink in the tubing, or migration of the end point of the tubing. Another complication, less common than obstruction, is when the shunt malfunctions and drains too much CSF. In this condition, the overactive shunt can produce the same signs and symptoms of a shunt obstruction (Table 1). A rare complication of an overproductive shunt is subdural hematoma, caused by tension on the bridging veins. Regardless of whether the patient has a shunt infection, malfunction, or a subdural hematoma, the primary concern is the presence of increased intracranial pressure (ICP), which needs prompt recognition and intervention.\textsuperscript{4-7}

<table>
<thead>
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<th>TABLE 1. Signs and symptoms of increased ICP.</th>
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<td>Headache</td>
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<td>Abnormal gait</td>
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<td>Altered mental status</td>
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<td>Apnea</td>
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<td>Episodic pain</td>
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<td>Bradycardia</td>
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<td>Bulging fontanel in infants</td>
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<td>Dilated or sluggish pupils</td>
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<td>Downward gaze (sun setting sign)</td>
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<td>Fever</td>
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VIGNETTE, CONTINUED

EMS responds with a crew of 2 rescuers. When they arrive on the scene, the child is laying on her bed. She is febrile and tachycardic. She responds to the EMS team but seems fussy and does not tell you her name or age. Mom tells you she is non-verbal at her baseline, but she is more sleepy than usual.

SYMPTOMS OF SHUNT COMPLICATIONS

As mentioned earlier, several different shunt complications can all lead to increased pressure on the brain. This means that patients with CSF shunt complications present with similar symptoms, regardless of the insult. Table 1 contains a list of symptoms that should be promptly recognized by EMS providers and ED staff as red flags and that should immediately brought to the attention of a physician.

EMS ASSESSMENT

A child with a CSF shunt is at risk for complications that can be fatal. It is important to look for red flags in a systematic way. Try to remember “ABCDEF” every time you are caring for a patient with a CSF shunt.

- Primary assessment: ABCDE
  Airway/Breathing: make sure the patient has an intact airway and check for irregular breathing patterns or apnea
  Circulation: document if the patient is tachycardic or bradycardic
  Disability: check for irritability, lethargy, or altered mental status
  Eyes: check for sluggish, dysconjugated, or fixed and dilated pupils

- Secondary assessment: F
  Focused history: Ask the caretaker if the patient has any fever, seizures, nausea/vomiting, ataxia, headaches, increased sleepiness, or irritability.

Apply the ABCDEF method to the patient in the vignette. As for the primary assessment, the patient is tachycardic and altered; it is important to maintain her airway and monitor her breathing. Attach a pulse oximetry probe if available, and document her oxygen saturation. It is also important to open her eyes and do a quick pupillary light reflex and document reactivity. For the secondary assessment, the patient is febrile, vomiting, and more sleepy than usual. Have suction ready to prevent the patient from aspirating. This patient has multiple red flags and is at a very high risk for seizures or herniation en route. Key questions that will enhance the assessment for the possibility of a shunt malfunction include the following: (1) When did the last shunt revision occur? (2) What signs and symptoms have been present during prior shunt malfunction events? These key questions will assist critical thinking by emergency care providers and preparation for shunt malfunction complications. If possible, make contact with the receiving hospital and inform emergency department (ED) staff of your findings in this critical patient.

VIGNETTE, CONTINUED

During transport, the patient's arms and legs begin jerking in rhythmic motions. Her heart rate increases and she is breathing spontaneously.

The patient is now experiencing a seizure; this could simply be a breakthrough seizure in a child with epilepsy or it may be a sign of increased pressure on the brain; however, it is important to be prepared for seizures during transport. Support the patient's head, and maintain a patent airway using a jaw thrust. Keep a bag-mask device and suction nearby. If the seizure lasts more than 5 minutes, deliver lorazepam 0.1 mg/kg intravenously (IV) or versed 0.15 mg/kg IV, intramuscularly, or IN.

VIGNETTE, CONTINUED

During and after a seizure, it is important to constantly reassess the patient. At this point, she is no longer fully protecting her airway and does not have adequate respiratory effort. This is a common event after termination of a seizure, and frequently, is time limited. Support her head, maintain a jaw thrust, and suction her oropharynx. She requires supplemental oxygen, and at least briefly, assisted ventilation. If she is not able to maintain her oxygen saturations and her respiratory effort does not improve, intubation should be considered. Another serious complication to be aware of is brain herniation. Suspect herniation if a patient becomes unresponsive with unequal pupils, fixed and dilated pupils, unresponsive pupils, hypertension, or bradycardia. If the patient has a rapid decline in mental
status and is unable to protect her airway and the gag reflex is absent, place an oropharyngeal airway and ventilate with a bag-mask device and 100% oxygen while preparing for intubation.

EMS report is a critical part of this patient's medical care. For this patient, an appropriate report would include the following:

**VIGNETTE, CONTINUED**

Transporting a 4 year old female with history of a VP shunt and report of fever, vomiting and lethargy. Upon arrival, the patient's vital signs were HR 115, BP 90/60, RR 12, T 101 F. Pupils were reactive to light bilaterally. She had a 2 minute seizure en route that resolved spontaneously, she is currently breathing spontaneously but requiring 10 L oxygen to maintain saturations > 95%. She does not have seizures regularly and does not take any medications for seizures at home. At baseline this patient is nonverbal and non-ambulatory. ETA is 7 minutes. Upon arrival to the emergency department, the patient is on 10 L of O₂ by facemask, is unresponsive, but withdraws from pain. She is now breathing on her own, but rapidly. You immediately elevate the head of her bed to 30 degrees and apply a non-rebreather face mask.

**ED NURSING ASSESSMENT AND INTERVENTIONS**

Similar to EMS, the primary nursing assessment should apply the ABCDEF method.

- Before the patient's arrival ED, staff should make sure suction and an ambu-bag with a range of appropriate masks are at the bedside. If possible, develop a plan with the physician to anticipate medications that may be needed and have a resources available at the bedside to calculate a weight-based dose. If the weight is not available, then use a length-based measuring tool to estimate the weight. Cardiac monitor and pulse oximetry should be available. Medications to anticipate include anticonvulsants, 3% normal saline, and rapid sequence intubation (RSI) medications. All medications should be double-checked with another registered nurse and dosed for patient's current weight measured in kilograms. Similar to EMS, the primary ED nursing assessment should apply the ABCDEF method.

Airway/Breathing: first assess the airway, and ensure that it is intact and clear of any emesis or secretions. If secretions are present, suction the airway and be prepared to turn the patient on her side if vomiting occurs to decrease the risk of aspiration. Assess breath sounds and work of breathing. If abnormalities are noted, assist the physician to provide an appropriate level of respiratory support. Bag-mask ventilation may be needed before intubation if the patient is apneic or not protecting her airway. The head should be midline with the nose in a “sniffing position” for optimal ventilation. Jaw thrust and/or chin-lift technique should be used with bag-mask ventilation. Be prepared to assist the physician in providing cricoid pressure with intubation if indicated. If the patient is intubated, an oral gastric tube should be placed to low-intermittent suction to decompress the patient's stomach.

- Circulation: assess heart rate, pulses, blood pressure, and capillary refill. Skin temperature and color are also important to document. Apply cardiac monitoring and pulse oximetry immediately upon arrival, and immediately notify the physician of any changes noted. Obtain vascular access; if unsuccessful after 2 attempts, prepare supplies for intraosseus needle placement. If the patient arrives via EMS and has an IV catheter in place, assess this for patency before use. Have isotonic fluids, such as normal saline, primed and at the bedside ready to administer. Obtain laboratory work according to physician orders. If bradycardia or hypertension is noted, immediately notify the physician.

Disability: assess patient's level of consciousness using the Glasgow coma scale or the AVPU pneumonic (Alert, responds to Verbal stimuli, responds to Painful stimuli, or Unresponsive). Alert the physician for any change in the level of consciousness. Assess pupillary response. Notify the physician if the pupils are unequal, fixed, dilated, pinpoint, or unresponsive. In patients with concern for increased ICP, some appropriate nursing interventions would include the following: decreasing stimuli in the room, elevation of the head of bed to 30°, and a minimum of hourly neurologic assessments. If the patient had or is at risk for seizures, pad the side rails of the bed and protect the patient from hitting her head on any hard objects. Place the patient on 100% oxygen via non-rebreather mask. Administer medications according to the physician's orders.

- Exposure: completely undress the patient and assess the entire body and perform a full
secondary assessment. Place warm blankets on the patient to maintain normal body temperature. If any findings such as bruising, abrasions, deformities, or edema are noted on secondary assessment, immediately notify the physician.

- Focused history/full set of vital signs: obtain a full set of vital signs at least hourly, including heart rate, respiratory rate, blood pressure, and temperature, and monitor pulse oximetry. Notify the physician of any abnormalities or significant changes. Ask the family for any medical history including drug/food allergies and current medications. Note any treatment given via EMS or at home before arrival.

**VIGNETTE, CONTINUED**

The patient now has a blood pressure of 155/95 and a heart rate of 64, with a respiratory rate of 18, and an end-tidal CO₂ of 78 mmHg. Her mother has not arrived yet and EMS does not know her weight. You use the length-based tape and find her weight estimate to be 18 kg. She remains unresponsive to verbal stimuli, but is still responsive to painful stimuli on the AVPU assessment. She still has sluggish but responsive pupils bilaterally. You place her on supplemental oxygen and obtain IV access. You then discuss the vital sign and examination findings with the physician as well as diagnostic evaluation and treatment plans.

**PHYSICIAN ASSESSMENT**

Similar to the EMS and nursing assessments, apply the ABCDEF method to a primary and secondary assessment of the patient.

Airway/Breathing: if the patient is unable to maintain an airway, prepare for RSI. Given the concern for increased ICP, appropriate medications would include etomidate and rocuronium. After securing an airway, gently hyperventilate if there is concern for increased ICP with P₃O₂ target of 30 to 35 mm Hg.

- Circulation: monitor closely for bradycardia and hypertension. Obtain 2 sites for peripheral vascular access. If IV access is not possible, place an IO. If there were unstable vital signs associated with increased ICP, the patient could benefit from either 3% normal saline (3-5 mL/kg IV) or mannitol (0.25-2 g/kg IV). In addition, a 20 mL/kg bolus of normal saline is indicated if the patient appears hypovolemic, although excessive fluid administration should be avoided in these patients because this can further contribute to the problem. Monitor for signs of shock because the patient may need vasopressors. Always start maintenance IV fluids before any transfer.

- Disability: if you need to intubate a patient who has had a recent seizure, it is important to provide a loading dose of fosphenytoin (20 mg/kg IV) in case the patient has another seizure that is not observed because the patient has been paralyzed for intubation.

- Eyes: be sure to document pupillary light reflex before intubation because it will be difficult to illicit with sedatives/paralytics on board. If the patient does not need to be intubated, avoid narcotics until a neurologic examination is completed because this will interfere with the pupillary light reflex.

- Focused history: check with the caregiver for any additional history.

**VIGNETTE, CONTINUED**

After receiving the report from EMS, obtaining vital signs from nursing and evaluating the patient, she is noted to be obtunded with agonal respiration. You decide to emergently intubate to protect and maintain her airway and to initiate hyperventilation to reduce ICP. The RSI agents selected are atropine, lidocaine, etomidate, and rocuronium. While preparing for intubation, you discuss with ED staff the next steps in treatment, which include obtaining laboratory data and a chest x-ray to check airway placement, and administration of 3% saline or mannitol, and fosphenytoin. After the airway is secured, you request elevation of the head of the bed to 30°.

**EMERGENCY DEPARTMENT EVALUATION AND TREATMENT**

Imaging is a key component of the evaluation of shunt function and shunt complications. Imaging should include a shunt series (x-rays of the shunt to rule out discontinuity in the hardware) and either computed tomography or magnetic resonance imaging (MRI) of the brain. Due to the clinical state of these patients, it is typically challenging to accurately detect sometimes subtle or even gross malfunction. The accuracy of clinical acumen alone in determining shunt function in the neurologically devastated child is fraught with error and has great potential for patient morbidity and mortality. Noncontrast computed tomography (CT) of the brain is a fast and reliable method for evaluating ventricle size and screening for
hemorrhage. Imaging will determine if there is a relative increase in ventricular size compared with the most recent study if available for comparison. CT is an appropriate choice for emergency imaging in all children, and especially, in a younger child who cannot hold still for an MRI.

A limited MRI is more sensitive and is becoming a standard of care at many tertiary children's hospitals. However, an MRI may take longer and should not be performed on a patient with a programmable shunt. A caregiver may not always know if the patient has a programmable shunt. If there is any question, an MRI should not be performed because this can be detrimental to the patient. Never let imaging delay the transfer of a patient to a higher level of care. Any patient with a CSF shunt and symptoms of a complication should prompt consultation and eventually evaluation by a neurosurgeon. If a neurosurgeon is unavailable at your hospital, one should arrange for transport as soon as possible to a facility with a neurosurgeon able to care for children. Once a neurosurgeon has evaluated the patient, it will be determined if the shunt needs to be tapped or if an extraventricular device or a new shunt needs to be placed in the operating room. Regardless, this patient's final disposition will eventually be to the pediatric intensive care unit.

Useful laboratory test results for this patient would include a complete blood count and blood culture given the fever and high risk for CSF infection. If seizures are recurring or the patient is in status epilepticus, then a complete metabolic panel is also recommended. One complication of increased ICP as a result of a malfunctioning shunt is hyponatremia, for which the patient can present with persistent seizures or status epilepticus. These seizures may be resistant to anticonvulsant therapy and only responsive to hypertonic saline infusion. If the patient has a seizure disorder, medication levels should be obtained. Coagulation studies are also important before going to the operating room. Once cultures have been drawn, if there is suspicion for a shunt infection, antibiotics should be started without attempting a lumbar puncture. An appropriate antibiotic regimen might include cefepime (50 mg/kg IV) and vancomycin (10-15 mg/kg IV) at meningitic dosing levels. Other important medications to consider are lorazepam as needed for seizures, ondansetron (0.1 mg/kg IV) for nausea, and morphine (0.1 mg/kg IV) for headaches once the neurologic examination is complete. Whenev-
increased ICP, an emergency physician can perform a shunt tap in an effort to relieve the ICP (Figure 2). This should only be done when there is no option for safe and timely transfer of the patient to a facility with a neurosurgeon available. There are considerable risks with this procedure, including infection, CSF leak, and local hematoma. A description of the procedure follows:

1. Place the patient supine with his face turned to the side allowing for the shunt reservoir to face up.
2. Shave any hair around the reservoir, prepare the site with povidone-iodine using sterile technique, then clean off with alcohol swab and allow it to dry.
3. Using sterile technique, palpate the reservoir and puncture the skin entering the reservoir with 25-gauge butterfly needle.
4. Attach a manometer; standardize the zero mark at the level of the cerebral ventricles.
5. CSF should be collected into sterile tubes.
6. If a distal obstruction is present, the CSF will flow readily. If there is a proximal obstruction or a shunt infection, using slight negative pressure with a 10-mL sterile syringe may improve CSF removal.
7. Continue to drain CSF until the measured pressure is between 10 and 20 cm H₂O.

Avoid applying suction to the syringe to reduce the chance of collapsing the ventricle and choroid plexus entering the shunt.²

SPASTICITY AND DYSTONIA ASSOCIATED WITH A BACLOFEN PUMP

VIGNETTE 2

A call comes in at 4 pm to assist a 5 year old ex-26 week premature male with a history of a mental retardation and cerebral palsy (MR/CP). The patient suffers from severe spasticity, which is controlled with a baclofen pump. His mother called 911 because the patient had two generalized tonic clonic seizures today, each lasting around 3 minutes and now he has a fever.

SPASTICITY AND DYSTONIA ASSOCIATED WITH A BACLOFEN PUMP

MUSCLE SPASTICITY AND DYSTONIA—OVERVIEW

Spasticity is a result of upper motor neuron injury, which causes certain muscles to be constantly contracted. Dystonia is a movement disorder that occurs when continuous muscle contractions lead to repetitive movements. Patients with spasticity and dystonia experience debilitating muscle spasms and frequent pain. Patients with cerebral palsy or a spinal cord injury are examples of the types of pediatric patients that may benefit from an intrathecal baclofen pump. The goal of therapy is to provide enough muscle relaxation to improve quality of life.²,⁹,¹⁰
BACLOFEN PUMP

Baclofen is a muscle relaxant used to treat spasticity and dystonia. It can be given orally in patients with mild to moderate symptoms. Patients with severe symptoms benefit from an intrathecal pump that delivers baclofen directly into the spinal canal. The benefit of the intrathecal pump is the delivery of smaller doses to achieve the desired effect rather than the larger oral doses that can lead to toxicity. The pump contains a reservoir for the drug and typically needs to be refilled every few months.2,9-11

The vignette describes symptoms of an abrupt discontinuation of the baclofen, causing withdrawal symptoms. There are 3 different sources for this presentation: a disruption in the catheter (kink, break, dislodged tubing), mechanical failure of the pump, or the pump reservoir may be empty. All can result in disruption of baclofen delivery, leading to life threatening withdrawal. Another potentially fatal complication of an intrathecal pump is when the delivered baclofen dose is too high, leading to baclofen toxicity. Lastly, the pump and tubing can become infected similar to any indwelling hardware.

EMS responds with a crew of 2 rescuers. When they arrive on the scene, the child is in his wheelchair. He is febrile and tachycardic. He is not moving much when you quickly evaluate his vitals. He is making moaning noises. Mom tells you at baseline he is non-verbal and wheelchair bound due to severe contractures, but he is more irritable and “tight” than usual. The last seizure was 20 minutes ago, it resolved spontaneously.

Focused history: Ask the caretaker if the patient has any fever, seizures, nausea, vomiting, ataxia, headaches, increased sleepiness, or irritability. Also, ask if the parent has a prescription for oral baclofen and to bring the medication to the hospital if possible.


d - Primary Assessment: ABCDE
  - Airway/Breathing: make sure the patient has an intact airway, and check for slow respirations or apnea.
  - Circulation: document if the patient is tachycardic or bradycardic.
  - Disability: check for irritability, lethargy, or altered mental status
  - Eyes: check for sluggish or fixed and dilated pupils.
- Secondary assessment: F

Apply the ABCDEF method to the patient in the vignette. As for the primary assessment, she is tachycardic and irritable; it is important to maintain the airway and monitor his breathing. Attach a pulse oximetry probe if available, and document O2 saturation. It is also important to open the eyes and do a quick pupillary light reflex and document reactivity. For the secondary assessment, he is febrile, no longer seizing, and more spastic than usual. Vomiting is common; have the suction ready to prevent the patient from aspirating. Be sure to support his head to maintain a patent airway. These patients often have severe contractures, making positioning difficult. Never try to stretch an extremity in these patients if you meet resistance because this can inflict pain and serious injury. If possible, make contact with the receiving hospital and inform them of your findings in this critical patient.

A baclofen pump complication is always a medical emergency, and it is important to be aware of the consequences.2,9-11,13 Symptoms of baclofen withdrawal and toxicity are listed in Figure 3.

EMS ASSESSMENT

A child with a baclofen pump is at risk for complications that can be fatal. It is important to look for red flags in a systematic way. Try to remember “ABCDEF” in your primary and secondary assessment:

- Primary Assessment: ABCDE
  - Airway/Breathing: make sure the patient has an intact airway, and check for slow respirations or apnea.
  - Circulation: document if the patient is tachycardic or bradycardic.
  - Disability: check for irritability, lethargy, or altered mental status
  - Eyes: check for sluggish or fixed and dilated pupils.
- Secondary assessment: F

Focused history: Ask the caretaker if the patient has any fever, seizures, nausea, vomiting, ataxia, headaches, increased sleepiness, or irritability. Also, ask if the parent has a prescription for oral baclofen and to bring the medication to the hospital if possible.

Figure 3. Signs of baclofen withdrawal and toxicity.
Communication from EMS is a critical part of this patient's medical care. For this patient, an appropriate report would include the following:

**VIGNETTE, CONTINUED**

We are transporting a 5 year old male with a history of MR/CP and baclofen pump for spasticity. At baseline this patient is nonverbal and non-ambulatory. 911 was called for fever and three seizures today. Upon arrival the patient's vital signs were HR 115, BP 90/60, RR 12, T 101 F. His pupils are reactive to light bilaterally. His last seizure was 20 minutes before arrival, lasted 3 minutes and resolved spontaneously. Currently the patient is stable on room air and has required no interventions en route. Our ETA is 7 minutes.

**NURSING ASSESSMENT/INTERVENTIONS**

Similar to the EMS assessment, apply the ABC-DEF method. Please refer to the CSF shunt malfunction scenario for details. Be sure to fully expose all skin surfaces, and check for pressure ulcers on these patients. These can be a source of infection and are often missed.

**PHYSICIAN ASSESSMENT**

This patient has a suspected baclofen pump malfunction, which is a medical emergency. When this patient arrives to the ED, similar to the EMS and ED nursing assessment, apply the ABCDEF method.

- **Airway/Breathing:** If the patient is unable to maintain an airway, set up for RSI. Given the concern for seizures, appropriate RSI medications would include versed and rocuronium. Assess breath sounds and work of breathing, and assist as indicated with bag/mask ventilation.
- **Circulation:** Obtain peripheral vascular access—preferably 2 lines. If IV access is not possible, place an IO needle. Monitor for signs of shock because this patient is reported to be febrile and could be septic. If there is a pump malfunction, tachycardia may be a sign of pain, and it may not respond to fluid boluses.
- **Disability:** If you need to intubate a patient with a recent seizure, it will be important to load with fosphenytoin in case the patient has another seizure that is not observed because the patient has been paralyzed for intubation.
- **Eyes:** Be sure to document pupillary light reflex before intubation or giving benzodiazepines for spasticity.

**ED EVALUATION AND TREATMENT**

This patient has a suspected pump malfunction, and early recognition is important. This patient also has a fever, which can be due to baclofen withdrawal or may be due to infection. Given the increased spasticity and seizure activity, baclofen withdrawal should be on the top of the differential and should be promptly evaluated. There are 3 important components in the workup of pump complications: laboratory work, medications, and pump interrogation. Laboratory studies for this patient would include a complete blood count and blood culture, given the fever and risk for infection. Coagulation studies are also important in case the pump requires surgical revision. Prolonged spasticity or seizures can cause rhabdomyolysis; a urinalysis can provide very timely data toward this diagnosis. A metabolic panel is important to monitor renal function and serum creatinine, as well as creatine phosphokinase, to monitor patient response to fluids. Once cultures have been drawn, if there is suspicion for a pump infection, antibiotics should be started without attempting a lumbar puncture. One appropriate antibiotic regimen would be cefepime and vancomycin at meningitic dosing levels. Whenever possible, consultation should be obtained with the child's specialist to determine the optimal medication regimen. Other important medications to consider are lorazepam as needed for seizures or spasticity.

Given that the symptoms may be caused by withdrawal, another medication to consider is oral baclofen. All patients with intrathecal pumps should have a prescription for oral baclofen from their specialist for emergencies such as this. Once the correct dose is verified, under physician discretion, it would be appropriate to try a dose if one is confident that the patient is not at risk for aspiration.

Fluids are important in these patients because they are at high risk for rhabdomyolysis. If present, this should be managed with bolus infusions of normal saline. Maintenance fluids will be necessary throughout ED care and transport to the tertiary center. Imaging is often pursued at the discretion of the neurosurgeon; this typically includes plain films.
Complex Neurology Patient Assessment
Airway
Breathing
Circulation
Disability
Exposure/Eyes
Focused History (allergies, medications, last meal, event)

Common Medications
Lorazepam 0.1 mg/kg/dose IV
Midazolam 0.1 mg/kg/dose IV; 0.5 mg/kg IN up to 10 mg
Diazepam 0.5 mg/kg rectal (max 5 mg if <5yo; max 10 mg if >5yo)
3% Saline 3-5 mL/kg/dose IV
Mannitol 0.25-2 g/kg/dose IV
Fosphenytoin 20 mg/kg/dose IV
Cefipime 50 mg/kg/dose IV
Vancomycin 15 mg/kg/dose IV
Ondansetron 0.1 mg/kg/dose IV

Cerebrospinal Fluid Shunts
Signs of increased intracranial pressure
- Vomiting
- Headache
- Episodic pain
- Abnormal gait
- Altered mental status
- Lethargy/somnolence
- Seizures

Management
- Elevate head of bed to 30 degrees
- If intubated keep pCO$_2$ 30-35 mm Hg
- Medication
- Imaging
- CBC, BMP, blood culture if fever, CPK if concern for rhabdomyolysis

Baclofen Pumps
Withdrawal signs and symptoms
- Irritability
- R rigidity
- Vomiting
- Seizures
- Fever/sepsis picture
- Rhabdomyolysis

Toxicity signs and symptoms
- Lethargy
- Hypotonia
- Urinary retention
- Respiratory distress or failure
- Coma

Figure 4. Pediatric high-technology neurology pocket card.
and a MRI of the spine. However, an MRI takes a significant amount of time and places the patient in a relatively unstable environment and should not be performed before transfer. Imaging should never delay transferring a patient to a higher level of care.

If this patient was having a baclofen overdose, the most important part of management would be establishing an airway. Any patient with a baclofen pump presenting with any symptoms suggestive of a possible complication needs to be evaluated by a neurosurgeon. If a neurosurgeon is unavailable, arrange for transport to an appropriate facility as soon as possible. Part of the evaluation by neurosurgery will include a pump interrogation to determine the cause of the malfunction.2,9,11-13

**SUMMARY**

It is important to have basic knowledge of devices and their complications in patients with complex neurologic diseases. Remember to always confirm airway, breathing and circulation first, and then consider possible complications specific to the patient. Simple complaints like fever, vomiting, or sleepiness can be red flags for underlying problems that can be life threatening. Every patient encounter that involves a CSF shunt or a baclofen pump with any signs or symptoms suggestive of malfunction or potential complication should be treated as an emergency until proven otherwise. Familiarity with the disease process, the hardware itself, and relevant differential diagnoses can expedite care and ultimately improve outcomes. Readily accessible references (see Figure 4) and timely contact with the patient’s specialty care provider will help to guide care. Always consider the child’s parents or home caregivers as key resources about the patient’s care needs.

**REFERENCES**

Disaster Management and Emergency Preparedness for Children and Youth With Special Health Care Needs

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VIGNETTE

A small bus carrying 5 children with special health care needs and 2 care providers is involved in a collision with a tree after sliding off a highway because of severe weather. The rural scene is located 15 miles from a community hospital emergency department and 80 miles from a major urban center with a pediatric tertiary care facility whose pediatric intensive care unit beds are full at present. A basic life support unit is on the scene within 5 minutes, with a first responder and basic emergency medical technician, and an advanced life support unit arrives shortly thereafter with an emergency medical technician paramedic and emergency medical technician intermediate IV.

Abstract:
The aim of this article is to discuss key aspects of the assessment and management of multiple children and youth with special health care needs (CYSHCN) in a mass casualty scenario, from prehospital to emergency department nursing and physician perspectives. We aim to highlight why CYSHCN are at increased risk for poor outcomes during disaster events and the importance of developing and maintaining an updated emergency information form along with early identification and thoughtful community disaster preparedness for CYSHCN, who are our most vulnerable patients when emergency resources are stressed or stretched beyond normal capacity.

Keywords: disasters; children with special health care needs
Disaster medicine faces the challenge of preparing for the limitless possibilities of critical scenarios that could play out unexpectedly in daily life. Although some disaster scenarios are considered more likely than others (earthquakes in California, for instance), the timing and scope of disasters are impossible to predict and may vary in scale from a mass casualty event (MCE) to a multiple casualty incident (MCI) as demonstrated in the case scenario above.

Children pose a wide variety of unique challenges for emergency preparedness and disaster scenarios at the national, state, and local levels. All children, especially children and youth with special health care needs (CYSHCN), are a vulnerable population during disaster events for several reasons, and major deficiencies have been described in the disaster preparedness plans of prehospital emergency medical services (EMS) agencies across the United States for the care of children. In its 2010 annual report to the President and Congress, the National Commission on Children and Disasters provided a sobering assessment of the current fragile state of disaster preparedness for children in our country and highlighted the limited availability and inaccessibility of resources related to pediatric disaster preparedness at that time.

It is well-known that a single critically ill or injured child is a major stress to prehospital providers, so MCIs or MCEs involving large numbers of injured and distressed children are bound to elicit emotional responses from emergency care providers. Furthermore, the added complexity of triage, life-sustaining equipment, and resource use for CYSHCN places these patients at even greater risk, as evidenced by the increased morbidity and mortality of CYSHCN in the wake of hurricane Katrina.

The aim of this article is to discuss key aspects of the assessment and management of multiple CYSHCN victims from prehospital, emergency department (ED) nursing, and ED physician standpoints, and to highlight the importance of thoughtful community disaster preparedness for CYSHCN, who are our most vulnerable patients when emergency resources are stressed or stretched beyond normal capacity.

**VIGNETTE, CONTINUED**

Both adult care providers appear to have minor injuries and can communicate some immediate concerns about the children:

- **Child 1** is ventilator dependent with a tracheostomy in place, now taking long pauses in between shallow breaths with a ventilator that is visibly damaged and not functioning.
- **Child 2** also has a tracheostomy (with no associated ventilator), and his tracheostomy ties and tube are lying on the ground, and he is in mild respiratory distress.
- **Child 3** has a ventriculoperitoneal shunt, hit his head, seems to be in pain, and has new swelling over his shunt site.
- **Child 4** has cerebral palsy with spastic quadraparesis and a baclofen pump. The pump is located in the right lower abdomen, and there is surrounding abdominal swelling and bruising, and the child appears uncomfortable.
- **Child 5** is awaiting a heart transplant and has a continuous central infusion of “something for his heart” via a peripherally inserted central catheter line, which appears to still be infusing. She also has a gastrostomy-jejunostomy (GJ) tube, which appears to be pulled out several centimeters.

**CHILDREN AND YOUTH WITH SPECIAL HEALTH CARE NEEDS IN A DISASTER: PREHOSPITAL EMS PREPARATION AND ASSESSMENT**

Regardless of event classification as MCI or MCE, it is easy to see that any such situation involving multiple ill or injured CYSHCN would likely feel like a disaster for anyone involved and would quickly stress local EMSs resources beyond their normal capacity. A high level of family and local community preparedness for the emergency care of these children involves preidentification of CYSHCN and their emergency medical information and advance consideration of necessary emergency care resources. This may make the difference between a needlessly chaotic response and one that successfully triages and stabilizes multiple medical needs.

The role of EMS providers in an MCI or MCE may include

(a) instituting an incident command structure, if necessary;
(b) providing primary triage and implementing a disaster triage system, if necessary;
(c) early notification of base hospitals;
(d) requesting additional transport resources; and
(e) direct communication with online medical direction.

In a scenario involving only a handful of CYSHCN, EMS providers would likely be called on to do all of the above. As an EMS provider in your community,
you should ask yourself how you would initiate the 5 steps above at the onset of a disaster.

Just like any other child, CYSHCN should have airway, breathing, and circulation that should be assessed and addressed. There are several established mass casualty triage systems available, with JumpSTART being a pediatric-specific algorithm designed to parallel the START triage algorithm. The assessment of CYSHCN in mass triage systems has not been well described, although accurate triage of this population could become an arm of defined pediatric secondary and tertiary triage assessments with further work. It should be noted that primary pediatric decision points for JumpSTART includes responses to simple airway repositioning and rescue breaths and, when evaluating CYSHCN with a tracheostomy, could include response to assisted ventilation via the tracheostomy as rescue breaths. This assumes that backup equipment is immediately available (ie, a transport “go” bag that should accompany the child at all times) and, thus, underscores the importance for family preparedness.

To illustrate this point in the case scenario, both Children 1 and 2 need immediate airway assessment and assistance. Child 1 would need to be removed from the useless ventilator circuit and have manually assisted breaths via the tracheostomy tube. An end-tidal carbon dioxide capnometer with color change, chest rise, and auscultation of breath sounds should help distinguish adequate tube position from the need to change out an obstructed or dislodged tube. Child 2 is in respiratory distress and will obviously need an immediate tracheostomy change, which should be performed in conjunction with the caregiver (if able) by the EMS provider with the highest available level of training using backup equipment (eg, replacement tracheostomy) in the child’s “go bag.” Children 3, 4, and 5 will also require careful assessment of their vital signs and need to be transported without delay for evaluation of other potentially life-threatening injuries.

With increasing frequency, EMS personnel will be called on to perform triage and assessment of critically ill or injured CYSHCN whether in daily emergency or disaster scenarios. As demonstrated above, this may involve procedures immediately necessary but rarely performed (ie, tracheostomy change, suctioning, and/or assisted ventilation) with unfamiliar equipment. Being aware ahead of time of the CYSHCN in your area and the medical devices that your team may encounter is of the utmost importance. If available, enlisting the help of the child's family member or care provider is essential, as they are likely “experts” regarding their child and may be highly knowledgeable about their devices.

What happens if a family member or care provider is not available or is seriously injured? Prehospital personnel should be trained to look for MedicAlert jewelry and other medical information on hand, such as an emergency information form (EIF). The creation of an EIF is absolutely essential and has been strongly advocated for over a decade by the American Academy of Pediatrics and the American College of Emergency Physicians. The EIF was proposed as a means to provide rapid access to a concise health summary for children with special health care needs in a 1999 joint American Academy of Pediatrics and American College of Emergency Physicians policy statement (reaffirmed in 2002 and updated in 2010). The EIF was introduced as a resource to aid emergency care providers in caring for medically complex children. Projects are underway nationally to create electronic EIFs, and repositories of such information and templates are available online. Both electronic and waterproof hard copies have been advocated to have on hand in case of an emergency with power failure.

It should be noted that any EIF is only as good as the information put into it. Although the onus should be on families and their medical home provider to keep such a document up to date and useable, these preventative emergency preparedness services may be viewed as onerous and as a nonreimbursable activity. In addition, it is absolutely imperative that local EMS agencies pursue awareness of the CYSHCN in their area by requesting and reviewing EIF data with these families. Furthermore, it is essential for EMS systems to arrange ongoing training for their EMS providers regarding specific areas of knowledge and actions they may be called on to perform and the contingency plans to deploy if that child and community are involved in a disaster. Comprehensive templates already exist and are available to assist in prehospital education and protocol development for CYSHCN.

**CHILDREN AND YOUTH WITH SPECIAL HEALTH CARE NEEDS IN A DISASTER: NURSING ASSESSMENT AND CRITICAL THINKING**

Moving from the prehospital setting into the hospital, ED nursing staff will become the next “front line” in continuing triage, reassessment, and
care for an unusual patient load in a disaster. This section will highlight key aspects of situational and patient assessment and critical thinking from both the charge nurse and bedside nurse vantage points when CYSHCN are involved in a disaster scenario.

**Charge Nurse**

Effective disaster response is a significant challenge for even the most experienced ED charge nurse. Determining who is the “sickest of the sick,” managing crowd control, and the appropriate allocation of personnel and resources are just a few of the immediate challenges that present themselves in a disaster situation. The added complexity of organizing care for pediatric patients who have special care needs and related equipment can make a difficult situation even more challenging and requires advance preparation and practice for these children to ensure successful outcomes.

First and foremost, when anticipating a sudden surge in critically injured or ill patients, clear and concise communication with all involved personnel is a must. Knowing the overall strengths and weaknesses of your team, your available personnel, space, and equipment are all essential components of maintaining order in this potentially stressful and chaotic time.

Upon receiving notice of an impending influx of patients, be prepared to activate your institution’s emergency or disaster response plan. It is imperative to familiarize yourself with this plan ahead of time and ensure that the plan includes memoranda of understanding (MOUs) and/or transfer agreements with other regional institutions (including centers with pediatric tertiary care capabilities) and respiratory care suppliers to identify resources that can be used during patient volume surges. In addition, be aware that in MCEs, many patients will self-transport rather than arrive via EMS. As a charge nurse, you will be a leader in initiating mass patient triage and decontamination procedures as well.

It is imperative to conduct a brief patient assessment to determine “sick vs not sick.” Once that is established, it is advantageous to rank the presenting patients and to begin treating the “most sick” accordingly. Having 1 dedicated person or team to conduct initial triage is very beneficial to facilitating overall patient flow. As mentioned previously, the JumpSTART triage system is a pediatric-specific algorithm that could be initiated within activation of your institution’s emergency or disaster response plan.

In an ED that is already at full capacity, it is essential to have each bedside nurse identify at least 1 patient who could be moved from their room to an alternate care area expeditiously and have a plan for who will care for these patients. It is also vital to identify and form more than 1 trauma team per shift as well as to determine what equipment would be necessary to convert a standard examination room into a trauma room quickly.

**Bedside Nurses**

When demand is great and available resources are not, mass triage systems are designed to quickly identify the sickest patients and direct how limited resources are initially used. It is here that CYSHCN may face an increased risk of being overlooked; this underscores the importance of advance preparation. Nursing staff caring for CYSHCN should determine the presence and availability of a “go” bag accompanying the patient because these may offer timely access to backup equipment and supplies. The need for a backup power supply for children supported by home ventilators and other battery-powered devices should be assessed immediately.

It is vital to remember that a CYSHCN’s home caregiver and an EIF are generally the best sources of information regarding the child’s specific condition(s), medications and equipment, required care interventions, and any prearranged transfer agreements/MOUs. Do not hesitate to ask the caregiver specific questions and involve them in care decisions, as they are usually much more familiar with what is needed to offer the most effective level of care for the patient.

**VIGNETTE, CONTINUED**

All 5 children have been transported to the local community hospital. Emergency department staff are working to assess each victim and are directly communicating with the ED charge nurse and attending physician.

Child 1: The first priority is to stabilize the child’s airway, if not already done by EMS. Assess whether bilateral breath sounds are present, and if not done already, discontinue the use of the damaged ventilator and designate someone to initiate the use of an ambu bag for ventilation. Suction the tracheostomy tube as necessary. Confirm appropriate tube position with end-tidal carbon dioxide monitoring and chest rise. Anticipate the need for chest x-ray and other supportive care to maintain airway, breathing, and circulation. Evaluate for presence of backup equipment, caregiver, and/or an EIF. Assess need for ongoing respiratory support.

Child 2: Confirm that EMS correctly replaced the tracheostomy tube, and stabilize the tube as needed.
Check for chest rise and the presence of equal breath sounds bilaterally, and evaluate work of breathing. Assess for any neck crepitus, stoma site bleeding, integrity of tracheostomy tube ties, and presence of an inflated cuff. Anticipate the need for chest x-ray. Evaluate available equipment: was the child's backup trach tube used? Evaluate for presence of backup equipment, caregiver, and/or an EIF.

Child 3: Assess for signs of increased intracranial pressure and shunt malfunction. This child should be kept with the head midline and the head of the bed elevated to 30° at all times. This patient has the capability to decompensate quickly, and neurologic status should be monitored very closely with frequent neurological checks and for vital signs indicative of deterioration (eg, Cushing's triad: hypertension, irregular respirations, and bradycardia) and for which increased intracranial pressure interventions may need to be administered emergently. Because CYSHCN can be difficult to assess, it is important to understand the patient's baseline level of function. This may be facilitated by involving the caregiver and/or evaluating the EIF. This patient will eventually need neurosurgical evaluation with imaging for shunt integrity. Evaluate for presence of a caregiver and/or an EIF.

Child 4: Assess for signs of baclofen withdrawal or toxicity. This patient has the capability to decompensate quickly and should be monitored very closely with frequent neurological checks (including muscle tone) and vital signs for symptoms of baclofen withdrawal or overdose. The patient may have sustained serious blunt injury to the abdomen or pelvis and should receive a trauma assessment. This

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Risk</th>
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<tbody>
<tr>
<td><strong>All Children</strong></td>
<td>• Dependent on others for food, clothing, shelter</td>
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<td><strong>General</strong></td>
<td>• Limited communication/verbal skills</td>
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<td></td>
<td>• Decreased cognitive ability to perceive danger, evacuate, and seek help</td>
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<td></td>
<td>• Targets of terrorism: child victims elicit emotional response</td>
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<td></td>
<td>• Separated from family at school for large portions of time</td>
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<tr>
<td><strong>Anatomic/Physiologic</strong></td>
<td>• More susceptible to injury from traumatic forces</td>
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<td>Smaller body mass, fat</td>
<td>• Vulnerable to fluid loss, hypothermia</td>
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<tr>
<td>Smaller circulating blood volume</td>
<td>• More exposure to accumulated heavier gases (sarin, chlorine)</td>
</tr>
<tr>
<td>Height lower to ground</td>
<td>• Greater risk for hypothermia, greater exposure to toxins absorbed through the skin</td>
</tr>
<tr>
<td>Higher BSA: mass ratio</td>
<td>• Increased permeability to toxins, prone to hypothermia</td>
</tr>
<tr>
<td>Thinner, less keratinized skin</td>
<td>• Susceptible to infectious diseases &amp; complications</td>
</tr>
<tr>
<td>Immature immune system, less herd immunity</td>
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<tr>
<td><strong>CYSHCN</strong></td>
<td>• May rely on life-sustaining equipment that requires a power source, is easily broken, and/or difficult to decontaminate, replace, or fix</td>
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<tr>
<td>Technology/hardware-assisted</td>
<td>• May have multiple medications or diets that are difficult to find or replace, have complex schedules, and/or rely on vulnerable equipment to administer</td>
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<tr>
<td>Medication/diet-dependent</td>
<td>• May limit communication with emergency personnel if a familiar adult provider is not available</td>
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<tr>
<td>Developmental delay</td>
<td>• If complicated, emergency personnel may avoid in MCE</td>
</tr>
<tr>
<td>Multiple medical problems</td>
<td></td>
</tr>
</tbody>
</table>

*CYSHCN = Children and Youth with Special Health Care Needs

Figure 1. Population at risk: why children and CYSHCN are vulnerable during a disaster. Note: This is not an exhaustive list nor is it intended to characterize CYSHCN as severely handicapped. It is intended for robust emergency preparedness and planning purposes only.
patient will also need neurosurgical evaluation with imaging. Assess for any beeping coming from the implanted baclofen pump. Evaluate for presence of a caregiver and/or EIF, paying particular attention to any information regarding the baclofen pump dose and infusion rate.

Child 3: Assess the integrity of the patient’s central line infusion, and assess/secure the dislodged feeding tube. This patient appears to be the most stable of the group but also has the potential to decompensate very rapidly. It is imperative to monitor this patient closely for signs and symptoms of heart failure and to identify the medication being infused. Be prepared to establish additional vascular access if there is any indication that the infusion is not working and that the child is deteriorating in addition to performing a chest x-ray and an electrocardiogram. Given the GJ tube dislodgement, assess the abdomen for signs of trauma. Anticipate the need for abdominal imaging to assess tube positioning and for evidence of free air. If the GJ tube was used solely for feeds, it may be appropriate to secure it and leave it alone until a higher level of care can be obtained. Evaluate for presence of a caregiver and/or EIF.

**CYSHCN IN A DISASTER: ED PHYSICIAN ASSESSMENT AND CRITICAL THINKING**

As shown in Figure 1, several factors place CYSHCN at heightened risk during disaster situations. In addition to the anatomical, physiological, and developmental susceptibilities of all children during a disaster, CYSHCN are even more susceptible to poor outcomes based on their potential to present for care with high-tech medical equipment, special medication or dietary needs, developmental difficulties, and/or multiple and complex medical problems.

Other articles in this issue of *Clinical Pediatric Emergency Medicine* discuss specific management issues for the technology-assisted CYSHCN and will not be repeated here. Instead, we take a final look at the case scenario that raises questions that, as an ED physician, you should ask yourself about each patient and the scenario as a whole.

**VIGNETTE, CONTINUED**

**Child 1:** The patient is ventilator-dependent with apnea, and no working ventilator is present.

1. Can you continue to support the patient’s airway until a ventilator is found?
2. Do you have appropriate ventilator circuits at your facility?
3. Does the family or referring facility have backup equipment/power sources?
4. Are there home supply companies available?
5. If no home supplies are available, does the patient need to be transferred?

**Child 2:** The patient is breathing on his own but required a tracheostomy change.

1. Can your staff perform this procedure, if needed, again?
2. Does your facility have the appropriate equipment and sizes?
3. Does the family have backup equipment available?
4. What are your alternatives if there is no additional equipment? Can you modify available equipment to suit the patient’s needs?
5. Do you have airway support at your facility, such as otolaryngology or anesthesiology?
6. If the patient has an uneventful tracheostomy change, can he be discharged?
7. What are your options for transport to a higher level of care if the patient decompensates?

**Child 3:** The patient has possible damage to his ventriculoperitoneal shunt.

1. What is the appropriate workup for the tubing and shunt function?
2. Can you perform the necessary imaging at your facility?
3. If there is a malfunction, who can fix it?
4. What is the closest facility that can take care of this patient?
5. What temporizing measures could you perform in case of a delayed transfer?

**Child 4:** The patient has cerebral palsy with developmental delay and blunt abdominal injury with possible trauma at his baclofen pump site.

1. Can you interrogate the pump or evaluate it for damage?
2. If the patient was nonverbal and alone, how would you obtain his history?
3. What are the signs of baclofen withdrawal or toxicity?
4. How would you treat signs of baclofen withdrawal or toxicity?
5. Does your institution have baclofen? What are your alternatives?
6. Do the parents have extra medications?
7. How would you pursue evaluation of intra-abdominal trauma in a child? Can you perform the necessary imaging in your facility? Can your local surgeon and anesthesiologist manage a surgical emergency?
8. What is the closest facility that can take care of this patient?

Child 5: The patient has a complex heart disease on a continuous life-sustaining infusion awaiting transplant.

1. What are the patient’s baseline vitals?
2. What other medications is the patient taking?
3. What procedures has the patient undergone?
4. How much medication does the patient have or need to get through the disaster?
5. Is the pump functioning and have back up power?
6. Does the family have a backup plan for medication, equipment, and power sources?
7. Does your facility carry the necessary medication or equipment?
8. What is the closest facility that does, and how can they get there?

Finally, have you carefully reviewed your hospital’s emergency/disaster response plan?

The answers to these questions are not always easy in day-to-day practice. Add to this the stress of caring for multiple patients and limited resources, it becomes extremely difficult to find the answers that could make the difference for a critically ill or injured child in the midst of a MCI. Understanding what special needs can be reasonably well-supported locally, and those that cannot, can inform decisions regarding which patients should be prioritized for transfer for definitive care.

Accessible information about CYSHCN’s history and treatment plans is invaluable in a disaster situation. However, as noted earlier, parents or caregivers might not always be available, so alternative sources of information should be pursued, such as primary and specialty care providers. As mentioned earlier, use of an EIF with input from the patient’s family, primary caregiver, subspecialist, emergency services, and hospital providers along with having backup supplies on hand can help mitigate risk to special needs children during a disaster.

The EIF should include emergency plans for backup equipment, medications, power sources, dietary needs, and emergency contacts. Regional transport and transfer agreements and MOUs with tertiary care centers should be established ahead of time and may obviate the need for unnecessary local ED use during periods of surge capacity. Advance planning for access to emergency power sources during electrical outages may also serve to reduce excess ED use during a crisis. Building community resilience for emergency and disaster management of CYSHCN involves parents, primary and specialty care providers, EMS agencies, regional hospitals, schools, public safety, public health, and emergency management agencies. Whenever possible, pediatricians should be involved in all levels of disaster planning. Identifying the needs of CYSHCN in your area ahead of time, conducting a local hazard vulnerability analysis, and planning for a disaster response that also addresses the needs of children take motivation and effort on the front end.

PLANNING RESOURCES

In addition to the EIF templates mentioned above, many other pediatric disaster preparedness resources have been consolidated online. The federal Emergency Medical Services for Children program has recently developed an online pediatric disaster clearinghouse of resources for health care providers, emergency and community planners, and families to help communities achieve an optimal level of emergency readiness for children who are involved in a disaster. The American Academy of Pediatrics also offers a comprehensive website resource on children and disasters for health care providers and families, with links to both federal and private sector information sources.

SUMMARY

Children and youth with special health care needs are at increased risk for poor outcomes during disasters for many reasons. The importance of developing and maintaining an updated EIF along with early identification and thoughtful community disaster planning for CYSHCN cannot be understated. Children and youth with special health care needs are our most vulnerable patients when emergency resources are stressed or stretched beyond normal capacity.

REFERENCES

Nursing and Equipment Troubleshooting for Special Needs Children in the Emergency Department

Catherine Porter Moore, MD, PhD, Jennifer Frizzell, MSN, RN, CPN, Anna Richmond, MSN, FNP-C, Kate Copeland, RN, BSN, CPN

Emergency medical services (EMS) is called to the home of a 6-year-old boy in respiratory distress. On arrival to the scene, EMS finds a smaller-than-stated-age boy with a small head with sun-setting eyes, who is coughing and sputtering sputum from a tracheostomy, which is dislodged from the home ventilator. His arms are flailing, and he is in obvious distress. He then begins to jerk rhythmically, his eyes roll back in his head, and he begins to turn quite pale.

MEDICALLY COMPLEX AND FRAGILE CHILDREN

Medically complex and fragile children living at home often require technological support such as supplemental oxygen, respiratory or cardiac monitors, mechanical ventilation, vascular access devices, and/or feeding tubes. These children are primarily
cared for by parents/guardians in the home, frequently with the assistance of home health nursing. The complex medical problems of these patients put them at increased risk for frequent hospitalizations, and often, their point of entry is by EMS transport to the closest hospital’s emergency department (ED).

Vignette Continued

The patient received a dose of rectal diazepam from the home health nurse. The motor activity slows down, but the patient continues to have copious thick yellow secretions from his tracheostomy. His mother describes him as her “miracle baby, born 4 months early and nobody thought he'd make it.” He is tracheostomy, ventilator, and gastrostomy tube dependent and does not walk or talk. He has a shunt in his head to “help with the fluid and seizures,” and although he does not walk or talk, he sometimes nods and mother knows when he is unwell.

EMERGENCY MEDICAL SERVICES ASSESSMENT

Stabilization and transport of the medically fragile child by EMS should follow the standardized assessment of airway, breathing, and circulation (ABCs).

Airway—For a child with a tracheostomy who requires ventilator support, airway assessment and assuring that the airway is patent are key to effective ventilator support.1 Appropriate supplies and suction technique and effective bag-mask ventilation will be needed for EMS transport of the tracheostomy patient. First, assess the tracheostomy tube for proper placement and patency with assisted ventilation via a self-inflating ambu bag and end-tidal CO$_2$ (ETCO$_2$) monitoring. Also note the presence of any bleeding and the amount and type of secretions from the tracheostomy.

Breathing—An evaluation of ventilatory function should be pursued with bag-mask or bag-tracheostomy ventilation, assessing the effectiveness of chest rise and the quality of breath sounds.1 If there are any concerns regarding airway patency or the functionality of the home ventilator, children presenting with respiratory symptoms should be ventilated via bag-mask in sync with their respiratory effort and removed from their home ventilator during transport.1

Circulation—A full set of vital signs should be obtained, including a blood pressure, and the patient should be placed on a cardiac monitor. Adequacy of circulation should be determined through an assessment of central and peripheral pulses and skin perfusion. Pulse oximetry and ETCO$_2$ monitoring should be applied and watched closely during transport.

The EMS provider should attempt to obtain as thorough a medical history as reasonably possible including baseline functional ability or mental capacity and home medications. It is important to determine the patient's baseline neurologic and respiratory status from the caregivers before transporting.1-4 This information is paramount to determining acute changes in the child’s condition, and this information will be vital for the receiving ED staff.

Vignette Continued

EMS providers assess the ABCs and immediately note that the airway requires clearing with suctioning of thick yellow mucus. After suctioning, the patient begins to breathe in a much less labored fashion. His breath sounds are coarse but equal, and his heart rate is in the 110s with easily palpable pulses in all extremities. Before preparing the patient for transport, the EMS team obtains a medical history from the mother and home health nurse and notifies the receiving hospital of their expected time of arrival and provides a report that includes their patient assessment, estimated weight, and medical history. Based on this report, what would you do to be prepared if you are a member of the receiving team in the ED?

EMERGENCY DEPARTMENT ASSESSMENT AND MANAGEMENT

The ED team should approach the anticipated arrival of every EMS transport in a standardized manner to avoid oversight error. The ready availability of necessary equipment and supplies for medically complex children is key to maintaining a calm and productive environment during initial ED stabilization. To ensure that all supplies are available, the nurse must be able to anticipate what special needs each patient may have. A thorough report from EMS or a transferring facility is very useful in anticipating patient needs. This advance notice is not always possible, which is why it is so important that all EDs be stocked with the necessary equipment (see Table 1).1,3

Guidelines for ED preparedness for children, which include detailed recommendations for necessary equipment, supplies, and medications for pediatric care, have been published by the American
An equipment checklist can be used by the primary nurse at the beginning of each shift to ensure availability in the examination room or ED. Upon notification by EMS and before patient arrival, the primary nurse should complete a survey of the examination or resuscitation room. This process should take approximately 2 minutes. This survey includes assuring that anticipated required equipment type and sizes are available and functional. The monitor should be checked, and a length-based medication-dosing tool (eg, Broselow tape) and an age-appropriate thermometer should be available. Having the appropriate airway equipment, suction setup, and the monitor ready to be attached to the patient upon arrival will save valuable time and reduce distraction during initial assessment and stabilization.

The assessment of special needs patients with complex medical needs should begin with a system-based approach, always beginning with the ABCs, and should include concurrent troubleshooting of the patient's equipment supporting these same systems.\textsuperscript{1,2}

**AIRWAY**

The evaluation of the special needs child begins, as does the evaluation of any child, with the airway. The most common airway device used in special needs children is the tracheostomy tube (trach). Important questions for the patient's caregiver, when present, include the following: What size/model is the trach? When was it last changed? Any difficulty replacing and managing the trach at home? Has the trach required more suctioning than normal or has there been a change in secretion color, amount, or consistency? Does the family have a replacement tube with them?

**Tracheostomy Tube Obstruction**

Tracheostomy occlusion is common, occurring in 14% of all children with trachs and in up to 72% in infant and neonatal age groups.\textsuperscript{3} If clearance techniques (suction) are ineffective, it may be necessary to replace the trach. Table 2 details the evaluation and management of suspected tracheostomy tube obstruction. This procedure requires the appropriate type of equipment (eg, trach tubes and suction catheters) and size range. Parents or home caregivers may arrive with a replacement tube and corresponding suction catheter. Table 3 offers comparative tube size data from different manufacturers and recommendations for appropriate size suction catheters; a resource such as this may be useful in guiding selection of an appropriate replacement tube in an acute setting. When replacing a tracheostomy tube, it is prudent to prepare for the worst-case scenario, which means having equipment, skilled personnel, and medications ready to perform rapid sequence intubation in a child who may have a difficult airway.

The patient should first be placed supine with the neck fully extended. Adequate restraint should be applied if the patient is unable to cooperate. After cutting or untying the trach ties and deflation of the balloon cuff (if present), the trach is removed with minimal force applied in a downward arc, with the tube pulled out and toward the chest. Tube replacement is then accomplished by inserting the

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**TABLE 1. Baseline ED equipment to address the airway, breathing and circulation needs of medically complex children.\textsuperscript{1}**

<table>
<thead>
<tr>
<th>Airway</th>
<th>O\textsubscript{2} source</th>
<th>Flow meter</th>
<th>O\textsubscript{2} nut nipple adapter (Christmas tree)</th>
<th>Pulse oximetry monitor and age appropriate probe</th>
<th>End-tidal CO\textsubscript{2} detector (EasyCap or capnography with waveform)</th>
<th>Nasal cannula</th>
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<td>Nonrebreather face masks</td>
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<td>Self-inflating ambu bag</td>
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<td></td>
<td></td>
<td>Anesthesia breathing circuit (Jackson Reese) with appropriate sized masks</td>
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<tr>
<td>Breathing</td>
<td>Suction canister</td>
<td></td>
<td>Yankauer suction tip</td>
<td>Deep nasopharyngeal suction kits</td>
<td>Endotracheal tube suction (Sims) catheters</td>
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<tr>
<td></td>
<td>Blood pressure cuffs in all sizes</td>
<td></td>
<td>Cardiac monitor</td>
<td>Electrocardiogram leads</td>
<td>Intravenous catheter start kit</td>
<td></td>
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<tr>
<td>Circulation</td>
<td></td>
<td></td>
<td>Endotracheal tube catheters</td>
<td>Deep nasopharyngeal suction kits</td>
<td>Endotracheal tube suction (Sims) catheters</td>
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</tbody>
</table>

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Academy of Pediatrics, Emergency Nurses Association, and the American College of Emergency Physicians.\textsuperscript{3} An equipment checklist can be used by the primary nurse at the beginning of each shift to ensure availability in the examination room or ED.
4. Another option for replacing tracheostomy tubes is by using a nasogastric tube as in Figure 1.

If tube replacement efforts are not successful, an endotracheal tube should be placed, and the trach stoma bandaged with sterile vaseline gauze, providing an occlusive dressing. If initial tube replacement efforts are not successful, another option is using an ETT exchanger with the capacity for delivering high flow oxygen thru an appropriate size endotracheal tube (to a set point pass the tracheostomy tube’s end) thru the new tracheostomy tube with attachment to an ETCO2 monitoring devices and ventilatory system (BVM or Anesthesia Bag) to provided ventilation during this procedure. Slide the ETT over the ETT exchanger utilizing the ETCO2 waveform for assessing proper tube placement then bandaged the ostomy site with sterile vaseline gauze, providing an occlusive dressing (Figure 2). Utilizing this set up will give the healthcare provider the most optimal assessment of the this new tracheostomy/ventilatory system and proper placement of the new tracheostomy tube. Once the replaced tracheostomy tube has adequate oxygenation and ETCO2 readings, the ETT tube can be removed and then utilizing the ETCO2 monitor and Pulse Oximetry adjustment to ventilation and securing the new tracheostomy tube can be done. One should remember that a false tract into subcutaneous tissues may be created by traumatic reinsertion efforts. If this or any other tracheostomy complication is present (eg, unintended decannulation, stomal granulomata, suprastomal collapse, tracheomalacia, bleeding, cellulitis, tracheitis), the patient will need to be evaluated by a pediatric

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**TABLE 2.** Management of suspected tracheostomy tube obstruction.

<table>
<thead>
<tr>
<th>YES</th>
<th>NO</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Monitor patient</td>
<td>• Attempt tracheostomy change</td>
</tr>
<tr>
<td>• Determine source of increased mucus production</td>
<td></td>
</tr>
</tbody>
</table>


---

**TABLE 3.** Approximate size of tracheostomy cannulas, endotracheal tubes, and suction catheters.

<table>
<thead>
<tr>
<th>Bivona</th>
<th>Shiley</th>
<th>ETT</th>
<th>Suction catheter</th>
</tr>
</thead>
<tbody>
<tr>
<td>Size</td>
<td>ID (mm)</td>
<td>OD (mm)</td>
<td>Length (mm)</td>
</tr>
<tr>
<td>2.5 neo</td>
<td>2.5</td>
<td>4.0</td>
<td>30</td>
</tr>
<tr>
<td>3.0 neo</td>
<td>3.0</td>
<td>4.7</td>
<td>32</td>
</tr>
<tr>
<td>3.5 neo</td>
<td>3.5</td>
<td>5.3</td>
<td>34</td>
</tr>
<tr>
<td>4.0 neo</td>
<td>4.0</td>
<td>6.0</td>
<td>36</td>
</tr>
<tr>
<td>2.5 ped</td>
<td>2.5</td>
<td>4.0</td>
<td>38</td>
</tr>
<tr>
<td>3.0 ped</td>
<td>3.0</td>
<td>5.3</td>
<td>40</td>
</tr>
<tr>
<td>3.5 ped</td>
<td>3.5</td>
<td>6.0</td>
<td>41</td>
</tr>
<tr>
<td>4.0 ped</td>
<td>4.0</td>
<td>6.7</td>
<td>42</td>
</tr>
<tr>
<td>4.5 ped</td>
<td>4.5</td>
<td>7.3</td>
<td>44</td>
</tr>
<tr>
<td>5.0 ped</td>
<td>5.5</td>
<td>8.0</td>
<td>46</td>
</tr>
</tbody>
</table>

Abbreviations: ETT, endotracheal tube; ID, internal diameter; OD, external diameter; neo, neonatal; ped, pediatric.
otolaryngologist. Brisk bleeding from the tracheostomy may represent an immediate life-threatening complication, the formation of a fistula with an adjacent vascular structure; this requires immediate referral to a center capable of pediatric surgical and critical care.

**Breathing**

Ventilator dependence necessitates a well-functioning ventilator. Important questions for the ED staff to consider include the following: What do I have in place to check the ventilator? Do we know if it is delivering the prescribed pressure/volume? The easiest way to quickly assess whether a ventilator is functioning properly is to take the patient off the home device and place them on a hospital ventilator with the same settings. Some hospitals have an in-house service (eg, biomechanics) that can perform mechanical or electronic checks on these devices. Respiratory therapy is another potential resource for the evaluation of ventilator performance. When available, these resources should be used. If the ventilator is not effective in ventilating the child, the patient can be maintained on a hospital ventilator, or in a worse-case scenario, by manual bagging.

Other ventilation-related technologies that are used by special needs children include continuous positive airway pressure (CPAP) for conditions such as obstructive sleep apnea, biphasic positive airway pressure (BiPAP) for patients with advanced muscular dystrophy, spinal muscle atrophy and related disorders, and mechanical insufflator-exsufflators (coughalator) for chest percussion for cystic fibrosis and other pulmonary diseases. If the patient uses these devices at their baseline and present to the hospital with a respiratory illness, chances are that they may need additional interventions beyond what they are getting at home. Typically, home health companies provide technical support and service these machines to ensure that they are working appropriately. The hospital should have access to these same devices to maximize the care these patients receive. The assessment and management of respiratory care issues in special needs children are discussed in much greater detail in the article by Bassham et al in this issue of *Clinical Pediatric Emergency Medicine*.

**Circulation**

Another key consideration for ED staff caring for medically complex children is whether the patient has any cardiac history. Cardiac and circulation support devices to be aware of include indwelling pacemakers/defibrillators and central venous access devices (CVADs). Chronically ill children will often have indwelling skin-tunneled vascular catheters, implanted injection ports, or peripherally inserted central catheters for long-term intravenous fluid therapy or medication administration. ED staff should ask caregivers about cardiac medications and the presence of central lines/ports in these children. The assessment and management of congenital heart disorders in special needs children are discussed in much greater detail in the article by Szlam et al in this issue of *Clinical Pediatric Emergency Medicine*.

**Management of Complications of CVADs**

Occlusion and infection are the most common complications involving CVADs. Before accessing a
CVAD, one should possess sufficient skill and knowledge about the requirements for routine access, flushing, and maintenance for that specific device. Parents and home health providers may prove to be expert in these aspects of CVAD management. If no CVAD expertise is present in the ED, experienced staff from other areas of the hospital may prove to be helpful.

The most common source of catheter-related septicemia is catheter hub colonization. When accessing a nonimplanted CVAD, it is imperative that injection hubs are adequately cleaned before medication delivery or infusion of fluids. Chlorhexidine gluconate 0.5% is recommended for cleansing of injection hubs. Hubs should be cleaned for a minimum of 30 seconds and allowed to dry before being used. Table 4 offers guidelines for CVAD maintenance.

If the CVAD is not cooperating with efforts to withdraw blood or flush, it may be clamped, it may be occluded by a clot, or on occasion, it may be broken. Maintenance and repair kits are commercially available for most non–fully implanted CVADs. At most pediatric tertiary referral centers, there are staff who specialize in central-line care and repair. When faced with a broken CVAD, one should be aware of data demonstrating an increased risk of infection in patients who have had central-line repair. This emphasizes the need for skilled providers in managing this problem. If local expertise in CVAD repair does not exist, the line should be clamped, and the patient referred to a center with that expertise.

Intraluminal occlusions may be due to precipitate formation from infused fluids and medications but are more commonly a consequence of clotted blood. In the scenario with a CVAD occluded by a presumed clot, tissue plasminogen activator (TPA) can be tried in an effort to clear the line. A typical protocol for this procedure is to dilute 2 mg of TPA in 1 mL of sterile water and to infuse this into the line after the proximal contents have been aspirated. The TPA should then remain in the line for 30 minutes before being aspirated. Access can then be reattempted. If this does succeed, the TPA dose can be repeated, this time left in the line for 2 hours. In the interim, it will be necessary to obtain peripheral vascular access for laboratory test results and medication administration.

**Neurological Assessment/Devices**

As described within the EMS section earlier, the initial assessment of medically complex children in the ED should include an evaluation of their mental status and neurologic function. Because many of these children do not have a normal level of baseline function, this evaluation should reflect alterations from that baseline status. Family members and home care providers are generally an excellent source of information regarding the child's baseline neurocognitive function.

The most common indwelling device likely to be encountered in special needs children are cerebrospinal fluid shunts used for the relief of hydrocephalus. The most common version is the ventriculoperitoneal shunt. Some patients, in whom a ventriculoperitoneal shunt has failed, may require a ventriculoatrial or ventriculopleural device. The assessment and management of these shunts is discussed in much greater detail in the article by Horton et al in this issue of Clinical Pediatric Emergency Medicine. For brevity sake, it is imperative to evaluate these patients for shunt malfunction because as this can be a lethal complication.
The baclofen pump, which delivers a muscle relaxant for the management of spasticity, from a reservoir placed in the abdomen via a catheter to the spinal canal, is another device that will be encountered in special needs children. This is also discussed in greater detail in the article by Horton et al. On examination, it may be palpated as a hard mass in the abdomen. In most specialty care centers, these pumps are serviced by either the physical medicine and rehabilitation or neurosurgery services. It is appropriate to ask the family which specialist cares for the pump and to contact the appropriate provider if there is a concern for malfunction. This too can be a life-threatening complication.

Gastrointestinal Assessment/Devices

Patients unable to effectively protect their airway during feeding or those with difficulty eating or swallowing are often managed with a gastrostomy or gastrojejunostomy tube. These devices allow delivery of nutrition and fluids directly to the stomach or jejunum. Tube-related complications and tube malfunction are a very frequent cause for ED visits. Therefore, it is important for ED care providers to be familiar with their general maintenance and resolution of common problems. Table 5 lists common gastrostomy complications and their management. The assessment and management of these devices are described in greater detail in the article by Bogie et al in this issue of Clinical Pediatric Emergency Medicine.

Fluids and nutrition are commonly delivered by a pump or by gravity/bolus in medically complex children. Important questions for the child’s caregiver include the following: Have there been any recent problems with feeds or the gastrostomy site? When was the gastrostomy tube inserted or last changed? On physical examination, the child should be assessed for any erythema, induration, tenderness, or warmth adjacent to the stoma. In the scenario with a child with accidental gastrostomy tube dislodgement and no effort (or unsuccessful efforts) to reinsert or replace the tube, the stoma should be

---

**TABLE 5. Management of common gastrostomy tube complications.**

<table>
<thead>
<tr>
<th>Complication</th>
<th>Possible Causes</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Leaking around tube</td>
<td>Migration of the tube Positioning of the child</td>
<td>Make sure gastrostomy tube is firmly in place.</td>
</tr>
<tr>
<td></td>
<td>Increased size of the stoma</td>
<td>Make sure that the balloon is properly inflated.</td>
</tr>
<tr>
<td></td>
<td>Child should always be in the upright position for</td>
<td>Stabilize tube with gauze pads. Split gauze may be placed under the</td>
</tr>
<tr>
<td></td>
<td>feeding.</td>
<td>gastrostomy button.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>If the stoma is too large, a new larger tube may need to be inserted.</td>
</tr>
<tr>
<td>Dislodgement</td>
<td>Child may have pulled tube out</td>
<td>If there is leaking from the lumen of the gastrostomy tube then the tube</td>
</tr>
<tr>
<td></td>
<td>Stoma may be too large for the tube</td>
<td>may need to be replaced.</td>
</tr>
<tr>
<td>Clogged gastrostomy tube</td>
<td>Residue</td>
<td>If the gastrostomy stoma is mature (&gt;2-3 mo old) then a dislodged tube</td>
</tr>
<tr>
<td></td>
<td>Improper flushing between feeds and medications</td>
<td>may be safely replaced with the same tube type and size if available.</td>
</tr>
<tr>
<td></td>
<td>Medication</td>
<td>If the gastrostomy is not mature, then a Foley catheter should be placed</td>
</tr>
<tr>
<td></td>
<td></td>
<td>in the stoma as soon as possible to prevent closure.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Always use correct formula.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Do not mix formula and medications.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Flush the gastrostomy tube before and after bolus feeds and medications.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Flush every 3-4 h with continuous feeds.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Dissolve 1/4 tsp meat tenderizer in 10 mL water and flush to dissolve clot.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Refer to local policy on acceptable use of medications or other</td>
</tr>
<tr>
<td></td>
<td></td>
<td>components before administering in gastrostomy tube.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>If you are unable to dissolve the clot, then a new tube will have to be</td>
</tr>
<tr>
<td></td>
<td></td>
<td>placed.</td>
</tr>
</tbody>
</table>

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stabilized with the largest Foley catheter that will fit until a replacement tube can be reinserted. ED staff should be wary of any effort to reinsert a tube for patients with a new stoma (less than 3 months since surgery) due to the risk for separation and creation of a false tract during insertion.

Often, before a gastrostomy tube is placed, patients (especially babies) are fed with a nasogastric tube. The same assessment questions apply to these patients: Any recent trouble with feedings or with the tube? Other gastrointestinal devices that may be encountered include colostomy and ileostomy stomas and ostomy bags, cutaneous portal drains, and Chait buttons/appendix stoma. Chait stoma are used for the delivery of antegrade continence enemas for the prevention of severe constipation. For these devices, asking the caregivers about their function and inspection for signs of infection is fundamental in their assessment.

Renal & Genitourinary Assessment/Devices

The following assessment questions should be asked: Is the patient on dialysis? If so, peritoneal vs arteriovenous? What is the patient's current regimen? Where is the dialysis catheter located? The dialysis site should be visually inspected for any redness, swelling, or drainage. Other devices that may be encountered include nephrostomy tubes and appendicovesicostomy (Mitrofanoff) tubes. These serve as alternate pathways for urine elimination. These should also be assessed in the same manner as a dialysis catheter.

The care of pediatric dialysis patients requires strict sterile technique throughout the procedure especially during connections, dressing changes, and cleaning. Most children with end-stage kidney disease are managed with peritoneal dialysis. These patients are at an increased risk for peritonitis. ED care providers should be capable of recognizing the signs and symptoms of peritonitis, which include cloudy dialysis fluid, fever, abdominal pain, and tenderness. Signs and symptoms of peritonitis signify a medical emergency and interventions should not be delayed.

Musculoskeletal Assessment/Devices

Equipment encountered in this category include braces and ambulatory assistive devices as well as splints, casts, and external fixation devices. Special needs patients, especially those with minimal activity, may develop osteopenia and fracture easily. It is vital that ED staff take special care when handling or moving these patients. Parents and caregivers will offer relevant experience on how to perform simple functions, such as wheelchair or bed transfers, toileting, and clothing changes. Furthermore, these patients should be assessed for skin breakdown from assistive devices or from infrequent position changes.

FAMILY CONSIDERATIONS FOR NURSING AND HEALTH CARE PROVIDERS

Emergency department staff must partner with parents, families, guardians, and home health providers who provide for the care needs for medically complex children. Taking the time for a thorough assessment and developing a rapport with family and home health providers will better inform ED staff about the patient and will ultimately improve patient care quality and safety. Here is a brief list of questions that should be considered for all ED patient assessments.

- How does the child communicate?
- How do you recognize when the patient is in pain?
- What are the child's normal stooling/urination patterns?
- What is the child's normal temperature
- What is the child's normal feeding regime?
- Has there been any rash or skin breakdown?
- What assistive devices are used?
- What are the contact numbers for home health and/or the durable medical equipment company?
- What are the family's/caregiver's specific concerns?

It is particularly important to embrace the concepts central to patient- and family-centered care, caring for the patient and not his condition; understanding the family's culture, values, and goals, and focusing patient in that context. Families of children with special health care needs are under very significant stress. This may be further magnified during an acute illness or ED visit. Health care providers should be sensitive to this dynamic and be proactive in partnering with families, so as to develop appropriate interventions and treatment plans for these special children.

SUMMARY

To provide optimal care for children with special health care needs, it is paramount to understand the patient's baseline function and current deviation from that, as well as the medical equipment/devices that are used maintain them. For this, nurses and physicians must establish a rapport with the family and have a basic understanding of the child's disabilities as well as the medical interventions required to best care for the patient. In addition to
leveraging the expertise of family members and home caregivers, timely consultation with the child's medical home provider and/or specialty care provider(s) will serve to inform and guide care.

REFERENCES

A 17-year-old previously healthy African American boy was roller-skating when he quickly twisted his trunk and felt a sharp pain in his chest. He felt lightheaded and sat down. He then developed lower back pain along with numbness and weakness in both legs. He also reported urinary incontinence. He denied any trauma or any previous episodes of numbness or weakness. He denied any recent illness, past medical problems, or surgeries.

He was taken to an outside emergency department via ambulance. Upon arrival, he was alert and oriented but unable to ambulate. He could not move his right lower extremity and had minimal movement of his left lower extremity. He had numbness over both lower extremities and decreased patellar deep tendon reflexes bilaterally.

Laboratory studies were obtained. The complete blood count showed the following: white blood cell (WBC) count 11,700/µL with 79.3% neutrophils, 15% lymphocytes, 4.9% monocytes, 0.5% eosinophils, and 0.3% basophils; hemoglobin 15.4 g/dL; hematocrit 44.8%; and a platelet count of 194,000/µL. A complete metabolic profile showed the following: sodium 139 mEq/L, potassium 3.7 mEq/L, chloride 102 mEq/L, CO₂ 22 mEq/L, serum urea nitrogen 13 mg/dL, creatinine 1.2 mg/dL, glucose 125 mg/dL, calcium 9.6 mg/dL, total protein 7.3 g/dL, albumin 4.5 g/dL, aspartate aminotransferase 60 IU/L, alanine aminotransferase 27 IU/L, total bilirubin 0.3 mg/dL, and alkaline phosphatase 89 IU/L. Urinalysis showed a specific gravity of 1.025, pH 6.0, and 2+ blood but was otherwise negative. Microscopic urinalysis showed 0 to 1 WBCs and 0 to 1 RBCs per high-power field, trace bacteria, and trace mucus. Urine drug screen was negative, and serum ethanol level was 0 g/dL.

Imaging studies were performed at the outside facility and dictated reports were provided. Computed tomography (CT) of the head without contrast showed no acute abnormalities. CT of the cervical spine without contrast showed a deformity of the spinous process of C2 on the left that may be related to a congenital anomaly or remote trauma. CT of the thoracic spine without contrast showed mild scoliosis of the mid and lower thoracic spine.

Abstract:
Aortic dissection is an uncommon but potentially catastrophic disorder. It typically occurs in older patients but can be seen in younger patients (usually with predisposing conditions). Aortic dissections can be classified into 2 types: type A dissections that involve the ascending aorta and type B dissections that do not involve the ascending aorta. We report a case of a teenager with a type B aortic dissection after a seemingly trivial inciting event. This article reviews important features of aortic dissection including diagnosis and management.

Keywords:
aortic dissection; Stanford classification; spinal cord ischemia; CT angiography; transesophageal echocardiography

Michael D. Baldovsky, DO, MBA

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with convexity to the right. CT of the lumbar spine without contrast showed no acute abnormalities. The patient was given the following intravenous medications: ketorolac 30 mg, dexamethasone 8 mg, ondansetron 4 mg, and morphine sulfate 4 mg. A Foley catheter was placed. He was then transferred to our hospital for further management.

When the patient arrived at our hospital, he continued to complain of numbness and weakness in both legs and was complaining of lower back pain. He denied any current chest pain or shortness of breath. His vital signs revealed a temperature of 37.3°C, heart rate of 77 beats per minute, respiratory rate of 18 breaths per minute, blood pressure of 174/98 mm Hg, and oxygen saturation of 100% on room air. On examination, he was awake and alert but was in mild distress secondary to lower back pain. His cardiac examination showed he had a regular rate and rhythm with no murmurs, rubs, or gallops. He had no chest tenderness to palpation. His lungs were clear to auscultation bilaterally with no respiratory distress. His abdomen was soft, nontender, and nondistended with normal bowel sounds. There was no hepatosplenomegaly. He was tender over his lower back and complained of pain with any movement of his lower back.

On neurologic examination, cranial nerves II-XII were intact. Muscle strength was 5/5 and sensation was intact in both upper extremities. Deep tendon reflexes were symmetric in both upper extremities. Muscle strength was 0/5 in both lower extremities. He had no sensation below the L2 dermatome on the right and below the L3 dermatome on the left. He had absent patellar and Achilles deep tendon reflexes bilaterally. There was no clonus, and the Babinski was absent. He had good rectal tone and no saddle anesthesia. Radial and femoral pulses were 2+ bilaterally. Pedal pulses were 1+ bilaterally, but capillary refill time was brisk in both feet. He had a Foley catheter in place with dark, reddish urine seen in the bag.

Laboratory studies were also obtained at our institution. A complete blood count was almost identical to the previous study. Prothrombin time was 14 seconds, international normalized ratio was 1.06, and partial thromboplastin time was 31.1 seconds. Troponin I was less than 0.05 ng/mL, and creatine kinase-MB was more than 300 ng/mL (normal range, 0.6-6.3 ng/mL). Urine drug screen was positive for opiates, but the patient received morphine sulfate at the outside hospital. An electrocardiogram showed a normal sinus rhythm with a normal axis and intervals and no ectopy. A compact disc containing radiologic images accompanied the patient in transfer, but it was blank when opened. Neurosurgery was consulted and saw the patient on arrival. Then, a definitive diagnostic study was ordered.

**DIFFERENTIAL DIAGNOSIS**

The patient's symptoms of lower back pain, acute lower extremity paralysis, and incontinence are concerning for a spinal lesion. The location of the patient's neurologic deficits suggests a lesion at the level of the lumbosacral spinal cord or cauda equina. The lumbosacral spinal cord innervates the muscles and dermatomes of the lower extremities and buttocks. Parasympathetic innervation of pelvic and abdominal organs is provided by sacral nerve roots.1 Spinal cord lesions can be grouped into several categories (Table 1).

Vascular disorders with resultant spinal cord infarction are an important cause of acute paralysis. Aortic dissections, traumatic aortic rupture, or aortic aneurysms that rupture or thrombose can cause ischemia of the spinal cord and lower extremities as well as ischemia of other major organs. Symptoms from these vascular lesions usually occur very abruptly and vary depending on the involved vascular territory. These lesions usually cause sharp pain in the chest, back, or abdomen depending on their location and can cause hypotension and death. If the patient experiences sharp chest or

### TABLE 1. Differential diagnosis of acute onset of lower back pain and lower extremity paralysis.

<table>
<thead>
<tr>
<th>Vascular/Spinal Cord Infarction</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aortic dissection</td>
</tr>
<tr>
<td>Aortic aneurysm with rupture/thrombosis</td>
</tr>
<tr>
<td>Aortic rupture</td>
</tr>
<tr>
<td>Vascular malformation with hemorrhage</td>
</tr>
<tr>
<td>Spinal epidural hematoma</td>
</tr>
<tr>
<td>Coagulopathy with thrombosis</td>
</tr>
<tr>
<td>Cardiogenic embolism</td>
</tr>
<tr>
<td>Trauma</td>
</tr>
<tr>
<td>Aortic rupture</td>
</tr>
<tr>
<td>Recent or prior aortic surgery</td>
</tr>
<tr>
<td>Vertebral fractures</td>
</tr>
<tr>
<td>Vertebral dislocation</td>
</tr>
<tr>
<td>Inflammatory</td>
</tr>
<tr>
<td>Transverse myelitis</td>
</tr>
<tr>
<td>Vasculitis</td>
</tr>
<tr>
<td>Malignancy</td>
</tr>
<tr>
<td>Spinal cord tumor with hemorrhage</td>
</tr>
<tr>
<td>Epidural metastases</td>
</tr>
<tr>
<td>Infectious</td>
</tr>
<tr>
<td>Viral myelitis</td>
</tr>
<tr>
<td>Epidural abscess</td>
</tr>
</tbody>
</table>
abdominal pain or hemodynamic instability, it can help to differentiate a vascular lesion from an isolated spinal lesion. Vascular malformations of the spinal cord can hemorrhage into the cord and cause compression or infarction. Embolisms from an underlying coagulopathy or cardiac origin can also cause infarction of the spinal cord. Symptoms depend on the involved vascular territory. Computed tomography angiography and magnetic resonance imaging (MRI)/magnetic resonance angiography (MRA) are effective in identifying these lesions. MRI can also identify spinal cord ischemia, although it may be normal in the first 24 hours. Spinal angiography may be necessary to further delineate vascular malformations.

Acute compression of the spinal cord can also lead to acute paralysis. Compression can occur secondary to a spinal epidural abscess or hematoma. Spinal epidural abscesses can result from skin and soft tissue infections, but many have no identifiable source. Spinal epidural hematomas can occur after spinal dural puncture or trauma. Rarely, they can occur spontaneously. In addition, hemorrhage of a spinal cord tumor can cause acute spinal cord compression. Epidural metastases from a primary tumor can cause vascular compression leading to spinal cord infarction. Patients commonly experience back pain followed by progressive sensory and motor deficits. MRI is optimal to delineate these spinal cord lesions. MRA is helpful in identifying a source of bleeding.

Inflammatory lesions of the spinal cord include transverse myelitis and vasculitis. Transverse myelitis causes acute inflammation of 1 or 2 segments of the spinal cord, commonly in the thoracic spinal cord. Most cases are idiopathic, but up to 60% may have a preceding infection. The inflammation usually involves the full thickness of the cord causing weakness as well as sensory disturbances below the lesion. The symptoms usually progress rapidly over several hours but may progress more slowly. Patients commonly present with back pain and a tight banding sensation around their abdomen. Acute viral myelitis occurs as part of a viral illness, commonly enterovirus or West Nile virus. The virus attacks the anterior horn cells causing asymmetric flaccid weakness and reduced or absent reflexes but minimal sensory symptoms. Patients also usually present with fever and headaches. Vasculitis resulting from infection or autoimmune disorders can cause spinal cord infarction with resultant paralysis. Symptoms vary depending on the involved vascular territory.

Trauma is another important cause of acute paralysis. Penetrating trauma can lead to direct spinal cord injury or spinal cord injury from bony fragments. Severe blunt trauma can lead to burst fractures, vertebral dislocations, or fracture dislocations, which can injure the spinal cord. Burst fractures cause fractures of the vertebral end plate with possible displacement of bony fragments into the spinal canal. Vertebral dislocations and severe fracture dislocations commonly cause complete paraplegia. In addition, trauma can cause cord compression due to a spinal epidural hematoma or disc herniation. Radiographs may identify fractures and dislocations, but computed tomography can add sensitivity and can further identify spinal cord impingement.

**CASE PROGRESSION AND DIAGNOSIS**

We attempted to order a CT angiogram of the chest, abdomen, and pelvis. However, because of concerns that the patient may have already received intravenous contrast at the outside hospital, an MRI was performed. The patient had an MRI of the cervical, thoracic, and lumbar spine with and without contrast, which showed a normal cervical, thoracic, and lumbar spine. However, an aortic dissection was seen from the origin of the left subclavian artery to the aortic bifurcation (Figure 1). CT angiography of the chest, abdomen, and pelvis was then performed. It showed aortic dissection from the origin of the left subclavian artery through the aortic bifurcation and involving both common iliac arteries (Figure 2). The intimal flap extended
superiorly to involve the left subclavian artery. In addition, the flap extended into the superior mesenteric artery, nearly occluding it just distal to its origin. Below the level of the renal arteries, the true aortic lumen was progressively compressed to the point where the distal aorta appeared completely occluded. The absence of flow continued into the common iliac arteries. There was reconstitution of flow at the level of the iliac bifurcations bilaterally. The thoracic aorta was diffusely ectatic, with the arch measuring 3.5 cm in width. The heart size was enlarged.

Bilateral lower extremity arterial ultrasound was performed and showed significantly diminished arterial flow in both lower extremity arterial circulations, suggestive of a more proximal high-grade stenosis and/or occlusion. Bilateral lower extremity venous ultrasound showed no deep venous thrombosis. Pending laboratory results from our hospital became available after the patient was transferred. Urinalysis showed a specific gravity of 1.026, pH of 6.0, large blood, moderate glucose, 4+ albumin, positive nitrates, and positive leukocyte esterase. Microscopic urinalysis showed 5 to 10 WBCs and 10 to 25 RBCs per high-power field, 2+ bacteria, no epithelial cells, and positive myoglobin. Urine myoglobin level was 2,700,000 μg/L. Serum creatine kinase was 29,638 IU/L (normal range, 33-145 IU/L).

The key to this patient’s diagnosis was his preceding severe chest pain and near syncope, which are not typical of isolated spinal cord lesions. In addition, he had decreased pedal pulses, indicating decreased perfusion to both of his lower extremities. Isolated spinal cord lesions can produce lower extremity weakness and paraplegia, but pulses would be preserved. The patient had an extensive type B aortic dissection with resultant spinal artery involvement and ischemia of the spinal cord and lower extremities. He developed rhabdomyolysis secondary to the extensive ischemia.

Cardiology and cardiothoracic surgery were consulted. He was given labetalol to control his blood pressure until an esmolol drip could be started. He was transferred to an adult hospital for further management because of the availability of vascular surgery. At the adult facility, he immediately had a spinal drain placed in an attempt to salvage some neurologic function, but he remained paralyzed. The following day, he was taken to the operating room for revascularization of his lower extremities using femoral artery grafts. He had declining renal function and was started on hemodialysis. One week later, he developed abdominal pain and was taken to the operating room for an exploratory laparotomy. He was found to have an occlusion of the superior mesenteric artery with bowel ischemia and underwent a bowel resection. He went back to the operating room later that night for a superior mesenteric artery bypass. Postoperatively, he remained intubated. He developed fevers and possible sepsis. His sedation was weaned, but he never regained consciousness and could not be extubated. He also developed seizure-like activity. It was thought that he had suffered a hypoxic ischemic injury at some point. His mental status never improved. After discussion with the family, care was withdrawn on hospital day 16. The family declined an autopsy.

**DISCUSSION—AORTIC DISSECTION**

Pathogenesis and Epidemiology

Aortic dissection is a relatively uncommon but potentially catastrophic condition that results from
a tear in the intima of the aorta. Blood flows into the tear and separates the intima from the media creating a false lumen. The dissection can extend proximally or distally from the initial tear. Aortic dissection presents more commonly in older patients, especially those with long-standing hypertension and atherosclerosis. The incidence of aortic dissection is approximately 2.9 in 100,000.\textsuperscript{11}

In a review of patients from the International Registry of Acute Aortic Dissection (IRAD), the mean age of patients with aortic dissection was 63.1 years with 65% of patients being male. A history of hypertension was present in 72.1% of patients.\textsuperscript{12} Aortic dissection can also develop in younger patients but is usually due to a predisposing condition, a traumatic event, or a transient elevation in blood pressure\textsuperscript{13} (Table 2).

There are several classifications of aortic dissection, but the Stanford classification is the most commonly used. The Stanford classification characterizes dissections into type A dissections, which involve the ascending aorta, and type B dissections, which do not involve the ascending aorta. In the review of patients from the IRAD, type A dissections occurred in 62.3% of patients with the remainder of patients presenting with type B dissections.\textsuperscript{12}

### Clinical Presentation

Patients with aortic dissection typically present with severe, sharp, “ripping,” or “tearing” chest or back pain, although painless dissection does rarely occur. In the review of patients from IRAD, 72.7% of patients reported chest pain, with anterior chest pain occurring more often in patients with type A dissections. Patients with type B dissections often presented with chest pain also but also commonly presented with back and abdominal pain. Regardless of the type of pain, the onset of pain was abrupt in 84.8% of patients and severe in 90.6% of patients.\textsuperscript{12}

Depending on the extent of the dissection, patients can experience multiple associated symptoms. Patients with dissections involving the ascending aorta can develop aortic regurgitation if there is extension near the aortic valve. Patients can develop myocardial ischemia if the coronary arteries are involved or neurologic deficits if there is carotid artery involvement. If the dissection causes rupture, patients can develop cardiac tamponade or hemothorax.\textsuperscript{14} Patients with dissections involving the descending aorta can develop ischemia of the bowel, kidneys, spinal cord, or lower extremities depending on the extent of the dissection. Syncope during dissection is associated with an increased risk of in-hospital mortality because of its relation to complications such as cardiac tamponade, stroke, spinal cord ischemia, and coma. Syncope occurs more often in proximal dissections.\textsuperscript{15}

### Diagnostic Tests

In adults, 3 predictors can stratify patients into low, intermediate, and high-probability groups:

1. Abrupt onset of thoracic or abdominal pain with a sharp, tearing, and/or ripping character.
2. Mediastinal and/or aortic widening on chest radiograph.
3. A pulse differential (absence of a proximal extremity pulse or carotid pulse on one side) and/or blood pressure differential (> 20 mm Hg difference between the right and left arm).

If there is an absence of all 3 variables, the patient has a low probability (7%) of dissection. If the patient has isolated aortic pain or isolated mediastinal widening on chest radiograph, there is an intermediate probability (31% and 39%, respectively) of dissection. If the patient has an isolated pulse/blood pressure differential or any combination of variables, there is a high probability (83%) of dissection.\textsuperscript{16}

CTA is usually the diagnostic test of choice because of its widespread availability and short duration of the scan. Chest radiographs may not show mediastinal widening in 45% to 55% of cases. Other abnormalities seen on chest radiograph include abnormal aortic or cardiac contour, pleural effusion, and displacement/calciﬁcation of the aorta. In the review of patients from IRAD, the chest radiograph was completely normal in 12.4% of dissections.\textsuperscript{12} If CT angiography is equivocal or further detail of the dissection is needed, MRI or transesophageal echocardiography is recommended. MRI is limited by its availability, prolonged

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<tr>
<th>TABLE 2. Causes of aortic dissection in younger patients.</th>
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<tr>
<td>Preexisting aortic aneurysm</td>
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<tr>
<td>Inflammatory vasculitis</td>
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<td>Collagen disorders (eg, Marfan syndrome, Ehlers-Danlos syndrome)</td>
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<td>Bicuspid aortic valve</td>
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<td>Turner syndrome</td>
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<td>Trauma</td>
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<td>Rapid acceleration/deceleration</td>
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<td>Transient elevations in blood pressure (eg, weight lifting, cocaine use)</td>
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<tr>
<td>Cardiac surgery/catheterization</td>
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<tr>
<td>Aortic valve replacement</td>
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duration of scan, and inability to use in patients with metallic implants or pacemakers. Transesophageal echocardiography is limited by its availability, the need for patient sedation, and operator dependence. If the patient is unstable, transesophageal echocardiography may be the preferable test. Electrocardiogram most commonly shows non-specific ST-segment or T-wave changes, and 30% may show no abnormalities. A smaller percentage may show ischemia or infarction, which can confuse the clinical picture of a patient presenting with chest pain. Blood tests are not very helpful in the diagnosis of aortic dissection.

Treatment and Outcome

Type A aortic dissections are considered to be surgical emergencies, whereas type B dissections can usually be managed medically unless the patient has progressive dissection or occlusion of a major vessel. Some dissections are repaired using endovascular stent grafts. Blood pressure and heart rate control are essential to minimize stress on the aortic wall. Morphine should be given for pain control. Intravenous β-blockers are the first line for blood pressure and heart rate control (propranolol, labetalol, or esmolol drip). Verapamil and diltiazem are alternatives for patients who cannot tolerate β-blockers. Systolic blood pressure should be reduced to 100 to 120 mm Hg. Intravenous nitropresside can be added. The patient can be switched to oral therapy once blood pressure and heart rate targets are achieved.

Patients who survive the initial dissection require lifelong blood pressure medications, usually with a β-blocker. Patients must also avoid strenuous physical activity. MRIs are usually performed at 3, 6, and 12 months after dissection and then every 1 to 2 years. Long-term survival rates differ in different studies but appear to be similar whether the patient has a type A or type B dissection. This is likely because both types of patients can develop adverse effects such as recurrent dissection and aneurysm formation.

SUMMARY

Aortic dissection is an uncommon but potentially devastating condition. It is usually seen in adults but must be considered in pediatric patients, especially in patients with certain underlying conditions. This case outlines a patient who presented with what initially may have seemed like an isolated spinal lesion. However, vascular lesions with resulting ischemia must always be considered in these patients. This is especially important because of the life-threatening nature of vascular lesions such as aortic dissection.

REFERENCES

Abstract:
This is a case of an 8-year-old girl who presents to the emergency department unable to walk due to right hip and left ankle pain for 3 days. Her examination is notable for polyarthralgia with significant tenderness of the left ankle and a heart murmur. Arthrocentesis of the ankle was performed, and initial results were inconclusive. Magnetic resonance imaging of the left ankle demonstrated findings consistent with an infection of the joint space. Subsequent laboratory results were also diagnostic for acute rheumatic fever. This patient was treated for both septic arthritis and acute rheumatic fever. This case report reviews the differential diagnosis for arthritis as well as the diagnostic criteria for acute rheumatic fever. The case highlights the need to search for a second diagnosis if a unifying diagnosis fails to explain all of a patient’s signs and symptoms.

Keywords:
arthritis; limp; septic arthritis; acute rheumatic fever

EMERGI-QUIZ CASE PRESENTATION

A Complicated Limp: When Occam's Razor Fails

Julia Lloyd, MD

A 8-year-old girl presents to the emergency department (ED) with right hip and left ankle pain. She reports falling and landing on her back 3 days before presentation while running at school. At the end of that day, her parents noted that she was walking with a limp, although she denied any pain. Two days before presentation, she developed left ankle pain, and right hip pain began on the night before presentation. On the morning she presents to the ED, she was unable to bear weight on either lower extremity due to the pain, and she noted some swelling of the left ankle. She has not had any fever, chills, respiratory, or gastrointestinal symptoms. She reports having a mouth sore over the preceding 3 days, which resolved by the time of presentation. She had a sore throat 2 weeks before presentation, which self-resolved. Medical, surgical, and family history were noncontributory, and review of systems was as noted above.

On physical examination, she was well appearing and afebrile, with normal vital signs for age. Examination of her head, eyes, ears, nose, and throat were normal without any oral ulcers or pharyngeal erythema. Cardiac examination revealed a II/VI systolic murmur heard best at the left upper sternal border, with regular rate and rhythm. Lung examination was clear to auscultation with good aeration. Her abdomen was obese with no tenderness or organomegaly. Her genitourinary examination showed normal female genitalia, Tanner stage 1. Her extremity examination was significant for decreased active and passive range of motion (ROM) to 60° of flexion at the right hip due to pain as well as pain with any internal rotation of the hip. She had no tenderness to palpation of right hip, knee, or ankle. There was
pain with passive ROM of the right knee. The left knee was swollen with tenderness of the lateral and medial joint lines. The left ankle had no obvious swelling, but there was pain with any passive or active ROM, worse with flexion. There was no erythema or warmth of any joint. Upper extremities were normal bilaterally. There were no rashes.

In the ED, a complete blood count was obtained with a white blood cell (WBC) count of $12.3 \times 10^3$/mm$^3$; hemoglobin level of 9.8 g/dL; and platelets of $342 \times 10^3$/mm$^3$ with 69% neutrophils, 20% lymphocytes, 11% monocytes, and a mean corpuscular volume of 74. The erythrocyte sedimentation rate (ESR) was 122 mm/h (normal, 0-20 mm/h), and C-reactive protein (CRP) was 21.8 mg/dL (normal, 0.0-0.8 mg/dL). Radiographs of the right hip and left ankle were normal. Ultrasound of the right hip demonstrated no effusion. Aspiration of the left ankle effusion was performed in the ED; the fluid obtained was grossly bloody, with 2.4 million red blood cells and 222 WBCs (50% neutrophils and 45% lymphocytes). The patient was admitted to the hospital, and the diagnosis was made on the second hospital day.

**DIFFERENTIAL DIAGNOSIS**

Limp and joint pain are common complaints in the pediatric ED, and the differential diagnosis is extensive (Table 1). Both common causes and life-threatening etiologies must be considered. A careful history and physical examination can help to narrow this differential. The history should include information regarding specific joints involved, timing and duration of symptoms, associated infectious symptoms, and history of trauma. Physical examination should include assessment of the affected joints for redness, warmth, swelling, ROM, and focal tenderness. In addition, specific attention should be paid to carefully evaluate for any rashes or skin changes; heart murmurs; or abdominal pain, tenderness, or masses. Judicious use of laboratory evaluation and imaging studies should be used to confirm or rule out diagnoses.

The most common cause of limp is trauma. Plain radiographs in the acute setting may not show all subtle fractures; follow-up radiographs 1 to 2 weeks after injury may be needed. Minor trauma is often readily recalled by caregivers, but it is important to note that if the trauma did not restrict the child from their activity at the time, it was unlikely to be the cause of the current complaint. Trauma may result in fractures, contusions, hemarthroses (possibly as an initial presentation of a bleeding disorder), sprains or strains, or inflammation of a tendon or bursa. Traumatic or overuse injuries such as osteochondritis dessicans, chondromalacia patellae, Osgood-Schlatter disease, slipped capital femoral epiphysis, or Legg-Calve-Perthes disease should also be considered. The time course of pain may often help to distinguish an overuse injury from an acute traumatic injury.

The most concerning diagnoses are those which are life or limb threatening. Acute rheumatic fever, Kawasaki disease, malignancies (including leukemia, neuroblastoma, or bony tumors), and infectious causes such as septic arthritis or osteomyelitis should be considered. These disease entities have particular physical findings that a careful physical examination may reveal to aid in diagnosis.

**TABLE 1. Differential diagnosis of joint pain.**

| Trauma/overuse | Fracture | Contusion | Hemarthrosis | Sprain/strain | Tendonitis | Bursitis | Osteochondritis dessicans | Legg-Calve-Perthes disease | Osgood-Schlatter disease | Slipped capital femoral epiphysis | Infection | Septic arthritis | Osteomyelitis | Gonococcal arthritis | Other infectious arthritis | Postinfectious (transient synovitis) | Acute rheumatic fever | Lyme disease | Bacterial endocarditis | Immune-mediated/vasculitis | Juvenile idiopathic arthritis | Serum sickness—like reaction | Kawasaki disease | Inflammatory bowel disease | Henoch-Schonlein purpura | Systemic lupus erythematosus | Hematologic/oncologic | Leukemia | Neuroblastoma | Bone tumor | Hemophilia |
|----------------|---------|-----------|-------------|--------------|------------|---------|---------------------------|--------------------------|--------------------------|----------------------------|-----------|----------------|--------------|------------------------|----------------------|----------------------------|------------------------|-------------|----------------------|------------------------|------------------------|----------------------|--------------|-------------|-------------|-------------|
Initial evaluation for acute rheumatic fever should include a careful cardiac examination, looking for a new murmur as well as other physical examination findings consistent with Jones criteria for acute rheumatic fever: arthritis, carditis, skin changes (erythema marginatum or subcutaneous nodules), or Sydenham chorea (Table 2). The diagnostic criteria of Kawasaki disease include fever; unilateral cervical lymphadenopathy greater than 0.5 cm; oropharyngeal mucous membrane changes; rash; conjunctival injection (usually sparing the limbic region); and changes of the extremities, including erythema, edema, or periungual desquamation. Laboratory test results and radiographs may be helpful in diagnosing bony tumors such as osteosarcoma or Ewing sarcoma and other malignancies such as leukemia, lymphoma, or metastatic disease. Bone pain rather than joint pain is a more classic presentation for leukemia or other infiltrative malignancies.

Although infectious causes for limp, such as septic arthritis or osteomyelitis, are often accompanied by fever, the absence of fever does not rule out these possibilities. Septic arthritis classically presents with a single swollen, red, and painful joint. Pain frequently worsens with attempts to flex or extend the joint. Joint effusions, seen on plain radiographs or ultrasound, may be present. Joint aspiration is useful for definitive diagnosis. Osteomyelitis, in contrast, is characterized by focal tenderness with or without warmth and edema and typically occurs over the metaphyses of long bones. Diagnosis can be difficult and often requires a combination of laboratory test results and imaging studies. In osteomyelitis, inflammatory markers such as ESR and CRP will typically be elevated, whereas the WBC count may be normal in up to two-thirds of cases. Blood cultures should be obtained before initiation of antibiotics to aid in identification of the infecting organism. Although plain radiographs are the initial study obtained, lack of findings is not helpful, as they may not show changes until several days into the infection. If suspicion is high but plain radiographs are inconclusive, magnetic resonance imaging (MRI) or bone scan should be obtained.

Less common causes of polyarthritis that should be considered include diagnoses such as gonococcal arthritis, Lyme disease, serum sickness–like reaction, bacterial endocarditis, systemic lupus erythematosus, Henoch-Schönlein purpura, inflammatory bowel disease, juvenile idiopathic arthritis, and postinfectious arthritis. Each of these diagnoses has individual clinical characteristics that may help to distinguish them. Gonococcal arthritis should be considered in the adolescent who is ill appearing with symptoms of tenosynovitis, especially of the wrist or ankle, accompanied with a vesiculopustular rash. Lyme disease has varied clinical pictures depending on the stage of disease: erythema migrans with migratory polyarthritis early in the disease or an intermittent monoarthritis in later disease. Serum sickness–like reaction often presents with diffuse urticaria and angioedema, usually within 1 to 2 weeks of initiation of a precipitating medication, typically an antibiotic. Acute bacterial endocarditis should be considered in the patient with known congenital heart disease with fever or a patient with a new murmur and splinter hemorrhages or other evidence of embolic phenomena. Systemic lupus erythematosus has a diverse presentation but typically presents as arthritis involving the smaller joints, along with involvement of other organ systems (cardiac, pulmonary, renal, or dermatologic). Henoch-Schönlein purpura is typically diagnosed based on the classic palpable petechial and purpuric rash in dependent areas. The rash associated with Henoch-Schönlein purpura may present after the arthritis, and abdominal pain have already manifested. Inflammatory bowel disease should be considered in the child with accompanying abdominal pain, stool changes, weight loss, or anemia. Juvenile idiopathic arthritis should be suspected in

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<th>TABLE 2. Jones criteria for diagnosis of acute rheumatic fever.</th>
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<td>Major Criteria</td>
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<td>Carditis</td>
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<td>Arthritis</td>
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<td>Subcutaneous nodules</td>
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<td>Erythema marginatum</td>
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<td>Chorea</td>
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<td>Minor Criteria</td>
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<tr>
<td>Clinical findings</td>
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<tr>
<td>Arthralgia</td>
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<td>Fever</td>
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<td>Laboratory findings</td>
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<tr>
<td>Elevated acute phase reactants</td>
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<tr>
<td>Erythrocyte sedimentation rate</td>
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<tr>
<td>C-reactive protein</td>
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<tr>
<td>Prolonged PR interval</td>
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<tr>
<td>Supporting evidence of antecedent group A streptococcal infection</td>
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<tr>
<td>Positive throat culture of rapid streptococcal antibody titer</td>
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<td>Elevated or rising streptococcal antibody titer</td>
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patients with daily temperature spikes and chronic polyarthritis. Postinfectious arthritis (transient synovitis) is common, usually does not have an associated fever, and occurs 1 to 2 weeks after an acute viral infection. Transient synovitis classically involves the large joints of the lower extremities.

**CASE PROGRESSION AND DIAGNOSIS**

On the first hospital day, an MRI of the left ankle was obtained to evaluate for osteomyelitis. The MRI demonstrates a small talotibular ankle joint effusion with mild synovial thickening and enhancement of the joint space. Increased T2 signal intensity was seen around the deltoid ligament and the anterior talofibular ligament, without evidence of frank disruption. An oblique linear region of high T2 signal was seen through the medial malleolus as well as within the bone marrow (Figure 1). These findings were concerning for both a septic left ankle and a nondisplaced fracture of the medial malleolus of the left ankle. A repeat joint aspiration of the left ankle was performed, and the fluid had a WBC count of 94,000 with 92% neutrophils. The presence of such a high number of WBCs in the joint aspirate with a neutrophilic predominance is suggestive of a bacterial infection in the joint space. Evidence of fracture and bony abnormalities suggests osteomyelitis as a possible source of the joint infection. She was taken to the operating room for joint washout with drain placement. In addition, a splint was placed for the nondisplaced fracture, and she was treated with intravenous clindamycin. No organism was identified from her blood or synovial fluid cultures.

On the second hospital day, the antistreptolysin O titer returned at 870 IU/mL (normal, 0-220 IU/mL), and the anti-DNase B titer was 480 (normal, <241). On the third hospital day, a throat culture grew group A beta-hemolytic streptococci. An electrocardiogram was obtained, which showed normal sinus rhythm without abnormalities. An echocardiogram showed signs of mild mitral regurgitation, mild aortic stenosis, and mild aortic regurgitation as well as a trivial pericardial effusion. Involvement of multiple joints with a history of beta-hemolytic Streptococcus infection, an elevated antistreptolysin O titer, and signs of carditis suggests a diagnosis of acute rheumatic fever.

Conservative therapy recommendations include treatment for both septic arthritis and acute rheumatic fever. In a patient diagnosed with acute rheumatic fever, published guidelines suggest that with arthralgia and minor carditis alone, aspirin therapy is not indicated. The consensus of experts was that this patient had 2 active processes, both septic arthritis of the left ankle as well as acute rheumatic fever. The other involved joints did not show signs of arthritis, only arthralgia. Treatment of both disease entities included intravenous clindamycin for 6 weeks, followed by penicillin prophylaxis for a minimum of 10 years. She is followed by pediatric cardiology for monitoring of her valvular disease.

**DISCUSSION**

Septic arthritis is a bacterial infection within a joint space. Although various organisms may be responsible, in children from 3 months of age to adolescence, Staphylococcus aureus is the most common pathogen. The infection typically results from hematogenous spread into the joint or bony metaphysis; however, in this case, the fracture and other bony abnormalities identified by MRI suggest that the infection began as an osteomyelitis that spread into the joint space. Most septic arthritis is a monoarticular arthritis, and it is rare to have multiple joints involved. Plain radiographs obtained in the acute setting often do not show any pathologic bony changes, although an early finding may be edema of the adjacent soft tissues.

The criterion standard for diagnosis of septic arthritis is an arthrocentesis yielding purulent fluid within the joint space, as seen in our patient. Published guidelines identify 5 criteria for diagnosis: (1) a non–weight-bearing patient, (2) fever above
38.5°C, (3) CRP greater than 2 mg/dL, (4) ESR greater than 40 mm/h, and (5) a peripheral WBC count of greater than 12000. If none of these factors are present, the patient has a 17% chance of a diagnosis of septic arthritis. Sensitivity increases as the number of criteria present increase, for example, if 2 factors are present, the predicted probability of septic arthritis is 62%. If 4 factors are present, the predicted probability of septic arthritis is 93%. If all 5 factors are present, the predicted probability of septic arthritis is 97.5%.

Septic arthritis demands rapid management. Infection of the hip is the most rapidly destructive, as increased pressure within the joint space can compromise the vascular supply. In animal models, it has been shown that antimicrobial treatment without drainage does not prevent joint infection unless started in the first 48 hours of inoculation. Initial treatment relieves the pressure within the joint as well as treating the infection. All children with septic arthritis require admission for intravenous antibiotics. Antibiotic choice may vary based on patient age and Gram stain and culture of the joint fluid.

Acute rheumatic fever is an inflammatory disease that develops after a streptococcal infection and may involve the heart, joints, skin, and brain. There are well-established guidelines for the diagnosis of acute rheumatic fever. The Jones criteria include both major and minor categories; 2 major criteria or 1 major and 2 minor criteria are diagnostic. The major criteria include carditis, arthritis, subcutaneous nodules, erythema marginatum, and Sydenham chorea. The minor criteria include clinical and laboratory findings such as arthralgia, fever, elevated acute phase reactants, a prolonged PR interval, and supportive evidence of an antecedent group A streptococcal infection. Inclusion of the evidence of recent streptococcal infection as one of the minor criteria is a more recent modification to the Jones criteria. Most of the minor criteria are nonspecific indices of inflammation and may lead to overdiagnosis of rheumatic fever. In our patient with echocardiography and carditis in conjunction with the previously mentioned minor criteria of arthralgia of multiple joints and evidence of a recent streptococcal infection, the diagnosis may still be made even with an active joint infection that could result in elevated inflammatory markers.

Management of acute rheumatic fever involves treatment of the acute streptococcal infection, typically with a course of penicillin. Our patient was already receiving clindamycin for her septic arthritis, which would treat acute rheumatic fever as well. Rest is advocated during evidence of active inflammation. Anti-inflammatory agents such as aspirin are typically indicated when arthritis and not just arthralgia is present. Chorea may be treated with benzodiazepines or haloperidol if necessary.

With regard to prevention of further recurrent episodes, either monthly intramuscular bicillin or twice daily oral penicillin is recommended. In patients without evidence of carditis, prophylactic antibiotics should be given for a minimum of 5 years or until 21 years of age, whichever is longer. If carditis is identified, prophylaxis should be continued until at least the age of 40 years and may be lifelong.

**SUMMARY**

Joint pain is a common presenting complaint for children in the ED. It is important to consider all potential etiologies of joint pain, ensuring that the life- and limb-threatening conditions are first excluded. A high index of suspicion must be maintained for conditions such as septic arthritis and rheumatic fever even when faced with inconclusive laboratory and diagnostic test results. This case offers a striking argument for the need to search for either a unifying diagnosis or a second diagnosis to explain the constellation of symptoms demonstrated. Early, definitive therapy for septic arthritis is vital to preventing long-term morbidity and loss of function, and recognition of acute rheumatic fever with appropriate treatment is required to prevent long-term cardiovascular morbidity.

**REFERENCES**