Emergency Care of the Technology-Assisted Child

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Integrated community living has become an achievable goal for many medically complex children. For many children with special health care needs (CSHCN), community living means an increased dependence on emergency medical services and emergency departments (EDs) for assistance with acute medical crises. Because transport times to the tertiary care ED can be prohibitively long, the community ED is more likely to become the site of first contact for the management of acute illnesses, exacerbations of chronic illnesses, and equipment-related problems experienced by CSHCN. Therefore, it is of paramount importance that emergency providers working in community as well as tertiary settings be well equipped to handle emergencies experienced by CSHCN. This article reviews common devices and their complications.

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Advances in neonatology, critical care, emergency medicine, and other subspecialties within pediatric medicine have enabled children to survive the complications of premature birth, congenital anomalies, and severe, life-threatening illnesses. As a result, there is an increasing population of children with special health care needs (CSHCN). In the United States, there are more than 12 million CSHCN [1], who, as defined by the Maternal and Child Health Bureau and the American Academy of Pediatrics, are “children who have or are at an increased risk for a chronic physical, developmental, behavioral, or emotional condition and who also require health and related services of a type or amount beyond that required by children generally.” Technology-assisted children comprise a subset of CSHCN and are children dependent on devices for survival [2].

There is a national move toward an integrated, community-based, family-centered medical system for CSHCN [2,3]. Moreover, there is a trend for medically complicated children to reside at home with their families rather than in tertiary hospitals or chronic care facilities. For many CSHCN, community living means an increased dependence on local emergency medical services and community hospital emergency departments (EDs) for assistance with acute medical crises. Because transport times to the tertiary care ED can be prohibitively long, the community ED is more likely to become the site of first contact for the management of acute illnesses, exacerbations of chronic illnesses, and equipment-related problems experienced by CSHCN. Therefore, it is of paramount importance that emergency providers working in community as well as tertiary settings be well equipped to handle emergencies experienced by CSHCN. An emergency physician will more adeptly provide care for such patients when equipped with a working knowledge of what to expect when a special needs child arrives to the ED. Understanding how CSHCN differ from other children, knowing what complications are likely to be encountered by CSHCN, and knowing how to troubleshoot equipment failures provide the foundation for the optimal delivery of care. This article familiarizes...
the emergency provider with the clinical manifestations and the management of common problems experienced by special needs children. In particular, this article reviews the common devices and their complications including tracheostomy tubes (TTs), cerebrospinal fluid (CSF) shunts, enteral feeding tubes, and indwelling venous catheters.

**General Management Principles**

When a technology-assisted child arrives in the ED, the child’s caregiver, home nurse, and/or primary care physician may be valuable sources of information. In many situations, caregivers are able to provide important details about the child’s baseline status and offer suggestions about management of the underlying condition. Families of CSHCN should be viewed as partners in the medical decision-making process [3,4]. Many parents of special needs children are experts in their child’s medical illness and in their equipment. Many view coming to the ED as a “last resort” after attempting temporizing measures at home. Also, many families carry with them medical summaries [5] and emergency boards or “go-bags” containing all of the equipment necessary to undergo a device replacement (eg, TT change) [6]. Therefore, including family in the care of the child not only serves to improve communication and enhance the psychosocial environment, but it also potentially optimizes care by contributing a better understanding of the child’s acute and chronic issues.

It is always important to remember that medically complex children get common pediatric illnesses too. For example, a child with a ventriculoperitoneal shunt and vomiting may have gastroenteritis as opposed to a shunt malfunction. However, the presence of indwelling devices predisposes a patient to infection and other complications so it is important to take into account all diagnostic considerations. At the same time, some indwelling devices can become colonized with organisms and therefore the usual indicators for bacterial growth (eg, cell counts, Gram stain, culture) may not necessarily indicate an acute infection. Furthermore, families of special needs children often have highly specialized equipment and trained personnel to assist in caring for their children at home. Many have the capability to manage an acute infection. Therefore, including family in the care of the child not only serves to improve communication and enhance the psychosocial environment, but it also potentially optimizes care by contributing a better understanding of the child’s acute and chronic issues.

**Assessing the Special Needs Child**

Health care providers should avoid the natural tendency to become distracted by the equipment and remember to focus first on the patient, not on the machinery. The assessment and treatment of special needs children should begin as with any child: prompt attention to the patient’s airway, breathing, and circulation [4,7,8]. Children with special health care needs may be quite different than similarly aged children. They may differ developmentally; however, it should also be noted that many children with physical handicaps are cognitively normal. Examining the patient may be difficult, especially if contractures, scoliosis, and/or the devices impede the physical examination. Baseline vital signs and weights may be different than what is typical for a similarly aged child [8]. For example, a child with chronic lung disease may routinely breathe at a rate higher than normally expected and may demonstrate some baseline accessory muscle use.

Key information to elicit from the child’s caregivers includes the following:

- What are the child’s current symptoms and how do these differ from his baseline?
- What are the child’s baseline developmental level, neurological status, vital signs, and weight?
- Has the caregiver administered any medications or performed any procedures before arrival?
- What is the history of any devices that are being used? (see Table 1)

**Tracheostomy**

**Background**

Children may require tracheostomy with or without mechanical ventilation for long-term support of respiratory function. The need for tracheostomy can originate from a primary respiratory disorder (eg, chronic lung disease of prematurity, congenital airway anomalies), a neuromuscular disorder (eg, muscular dystrophy or spinal muscular atrophy), or a disorder of the central nervous system (eg, central hypoventilation syndrome, traumatic brain or spinal cord injury) [9-11]. As home health care continues to be recognized as a viable alternative to long-term hospitalization or placement in a chronic care facility, the number of children with tracheostomy and mechanical ventilation living at home will continue to increase dramatically. By necessity, most children with tracheostomies living at home will utilize the local ED when acute complications arise. The ED physician will be able to approach these patients with knowledge of the normal physiology and the expected and unexpected complications of tracheostomy patients. The ED physician will be able to approach these patients with knowledge of the normal physiology and the expected and unexpected complications of tracheostomy patients.

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<td>What are the child’s usual presenting symptoms of a malfunctioning device?</td>
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calmly and systematically if he/she has a familiarity with the equipment and an understanding of the potential complications and management options for children with a tracheostomy.

**Tracheostomy Equipment**

When a patient dependent upon airway devices presents to the ED, it can be overwhelming and often challenging to elucidate the inciting problem, particularly if the child demonstrates respiratory distress. Often it is helpful for the examining physician to step back and spend a brief moment identifying the individual components of the patient's apparatus. Orienting oneself by beginning at the patient's neck, the first device that is encountered is the TT, often followed by a swivel device, and then either the ventilator tubing (if the patient is attached to the ventilator) or the heat-moisture exchanger [7].

**Tracheostomy Tube**

No longer made of metal, modern TTs are generally made of polyvinyl chloride, a synthetic substance that causes little tissue reactivity and conforms better to the shape of the trachea. Tracheostomy tubes are available from several manufacturers, packaged under sterile conditions for one-time use. Although there are manufacturer-specific differences among the tubes, in the acute setting, it is important to know only a few general characteristics of the patient's TT so that a replacement tube can be obtained and inserted if indicated. Important characteristics of a TT are its size and dimensions, whether it is cuffed or uncuffed, if it has an inner cannula, and if it is fenestrated [7,12-14] (see Figure 1).

Tracheostomy tubes are sized according to three dimensions: the inner diameter, the outer diameter, and the length. The inner diameter is always imprinted on the flange of the TT. The inner diameter ranges from 2.5 mm neonatal to 10.0 mm adult TTs. The inner diameter generally is kept constant among manufacturers and it is also the same number that is used to size endotracheal tubes. However, the outer diameter and length, which may or may not be imprinted on the flange, can vary considerably among manufacturers so it is important to keep this in mind during replacement. When a TT change is indicated and an identical replacement tube is not available, one must select a tube that is available with suitable dimensions. If a replacement tube of the same inner diameter as the patient's usual TT is not easily passed through the stoma, impediment may be due to a larger outer diameter. Therefore, it might be necessary to “size down” the tube by selecting one with a smaller outer diameter. Sizing charts have been published elsewhere [8].

Tracheostomy tubes may be cuffed or uncuffed. It is imperative to check in every patient with a TT for the presence of a cuff and deflate it before removal. The rule learned for endotracheal tube placement that cuffed tubes are to be used only for older children does not hold true for TTs. Even a young infant may have a cuffed TT in place if his underlying disease requires it (eg, tracheal anomaly).

Some patients may have a TT with an inner cannula. Tubes with an inner cannula are commonly used in patients with particularly precarious airways for whom transient removal as with routine cannula changes can be dangerous. For these patients, the inner cannula can be removed for cleaning while the outer cannula maintains the airway. Importantly, the inner cannula must be in place for proper connection of the resuscitator bag for hand ventilation. Children with a fenestrated tube can be taught how to force air out of the fenestration and then through vocal cords.

**Swivel Device**

To avoid traction on the TT when movement is attempted, a swivel device is interposed between the TT opening and the ventilator tubing. In addition, the few centimeters imparted by the swivel device serves to extend the TT opening beyond the soft tissues of the neck thereby preventing occlusion of the tube with neck flexion.

**Heat-Moisture Exchanger**

Anatomically, placement of a tracheostomy occurs at a level below the vocal cords. Therefore, air inspired into the lungs via the tracheostomy bypasses the important natural warming and humidification systems of the upper airway. For children who are maintained on a ventilator, heating and humidification systems are part of the stationary ventilator circuit. For children who do not require mechanical ventilation, or for those who will be managed off of mechanical ventilation for a period (either
during chest physiotherapy or during periods of hand ventilation), a heat-moisture exchanger is placed on the end of the swivel device or the TT. The heat-moisture exchanger is a plastic device that surrounds a honey-combed, hydrophilic substance that captures the patients own heat and humidity on exhalation so that it can be inspired on inhalation (see Figure 2).

Dislodgment or Obstruction
A child with an artificial airway with respiratory distress is considered to have a cannula obstruction or dislodgment until proven otherwise. Airway patency and the adequacy of breathing should be assessed through the usual physical examination and monitoring means. Supplemental oxygen should be administered. Suctioning of the tracheal tube to evaluate patency and to clear secretions may be helpful in alleviating symptoms. Do not be falsely reassured by a tube entering into the stoma as it may actually descend into the soft tissues of the neck rather than into the trachea, especially if a prior TT change was attempted and unknowingly resulted in a false passage.

An emergent TT change may be indicated if respiratory distress persists and/or the cannula clearly is dislodged. This procedure is best accomplished by two people: one to secure the patient, deflate the cuff (if present), and remove the old tube and the other to replace the new tube and assess it for proper positioning. All of the necessary equipment should be readily available including a replacement cannula with obturator, endotracheal tubes of smaller sizes, lubricant, securing tapes or ties, suction, oxygen, and monitoring devices (cardiorespiratory, pulse oximetry, end-tidal carbon dioxide). The stoma is exposed by laying the patient supine with a rolled towel under the shoulders. The tube and obturator combination is lubricated and placed through the stoma and into the trachea using gentle pressure and an arc-like motion, pushing the cannula posteriorly and downward until the flanges are flush against the neck. After confirmation of proper positioning via physical examination and non-invasive monitoring, the tube is secured with TT ties. Sufficient padding is placed under the ties to maximize comfort and dryness [7,8]. In the event that stomal constriction prevents insertion of the replacement TT, several options are available: (1) insert a TT one-half size smaller; (2) insert a smaller-sized endotracheal tube (being careful not to advance it into a mainstem bronchus) and then slowly dilate the stoma by inserting tubes of successively increasing size; and (3) cover the stoma and bag-valve-mask ventilate using traditional methods via the patient’s upper airway. Oral intubation may be exceptionally difficult, especially if the original indication for tracheostomy was to overcome an airway anomaly such as tracheal stenosis or a tracheal shelf. Therefore, neuromuscular blockade should be used with caution and anesthesiology and/or otolaryngology assistance should be considered early.

Asthma/Reactive Airway Disease
Many patients with tracheostomies have underlying chronic lung disease that may predispose them to respiratory exacerbations and/or flares of asthma. The usual asthma triggers including viral illnesses, pollens, dust, and smoke commonly precipitate respiratory exacerbations. Management parallels that of other asthmatic children and primarily consists of bronchodilator and steroid therapy. Nebulizer or metered-dose inhaler equipment can be placed in-line with the ventilator circuitry to deliver bronchodilator medications. Most respiratory therapists and even some family members or home care nurses can provide assistance with assembling this equipment. For patients with a tracheostomy and no ventilator, nebulized medications can be administered by placing the mask over the TT opening or by directing the medication toward the TT in a “blow-by” fashion.

Infection
Patients with TTs commonly are colonized with one or more organisms in the trachea [15]. This colonization usually poses no issues. However, these same organisms can become pathogenic resulting in tracheitis and/or pneumonia. It can be difficult to distinguish colonization from an infection. Here, the patient’s history is paramount. The emergency provider should elicit a history of systemic signs and symptoms such as fever and malaise. In addition, changes from baseline respiratory status including worsened tachypnea, cough, accessory muscle use, increased ventilation, or supplemental oxygen requirements and changes in the quantity and quality of the tracheal secretions may provide clinical support for the diagnosis of acute infection. Because there is no perfect, noninvasive test that can distinguish between colonization and active infection with 100% sensitivity and specificity, some recommend sampling the tracheal secretions for Gram
but bacterial culture as well as for viral respiratory detection assays [7]. Treatment is with oral or parental antibiotics depending on the severity of the illness.

Bleeding

The most common cause of minor bleeding from the TT is dryness and friability of the tracheal mucosa due to inadequate humidification. Increasing the humidification in the ventilator circuit and/or ensuring that a heat-moisture exchanger is used consistently will help to avoid bleeding from this cause. Bleeding can also result from granuloma formation. Granulomas can form at the tip of the tube, at the cuff, and/or at the stoma. For the latter, cautery with silver nitrate may be temporizing or curative. Finally, minor bleeding may also signify infection, either viral or bacterial tracheitis. Large amounts of bleeding may indicate a rare but life-threatening complication—erosion of the TT into a major vessel. This should be treated as a surgical emergency. Vascular access should be obtained and fluid resuscitation initiated while calls are made to notify surgeons and the operating room staff. Importantly, the TT should be kept in place as it may be the only way to ensure an adequate airway. Frequent suctioning may help to minimize aspiration. Overexpansion of the TT cuff may help to tamponade a bleeding vessel.

Cerebrospinal Fluid Shunts

Background

Cerebrospinal fluid shunts are placed to divert CSF from the brain to another area of the body, most commonly the peritoneal cavity. The choroid plexus produces CSF at a rate of 0.5 mL/kg per hour, or about 500 mL/d for an adult. Obstruction of flow at the third or fourth ventricle, or impaired resorption of CSF, leads to increased intracranial pressure (ICP). Cerebrospinal fluid shunts divert fluid from the intracranial vault to a caudal location. The emergency management of children with CSF shunts focuses on diagnosing shunt obstruction and shunt infection.

CSF Shunt: Anatomy of the Device

Most CSF shunts have three components: proximal shunt tubing, a reservoir system, and distal shunt tubing [16] (see Figure 3). The distendable, Silastic shunt tubing has a fenestrated tip that is located in a ventricle or a noncommunicating cyst. The reservoir, or “bubble,” is located directly over or slightly distal to the burr hole. The distal shunt tubing may be located in the peritoneum, vascular system, or pleural cavity and may contain a metal connector in case surgical replacement of the distal portion is required.

CSF Shunt Malfunction

Patients with mechanical shunt failure often complain of headache, visual disturbances, vomiting, lethargy, and irritability [17]. The most common complaint is alteration of mental status. Parental history can provide crucial insight into the possibility of shunt malfunction and may note that the child “just isn’t acting right” or is less active than usual. In many cases, parents report symptoms that are similar to a previous shunt obstruction. Other symptoms that reflect increased ICP include sunsetting or gaze preference secondary to cranial nerve neuropraxia and vital sign changes such as hypertension and bradycardia.

Many clinicians attempt to assess CSF shunt function by “pumping” the shunt reservoirs. This involves depressing the reservoir bubble and then checking for poor filling upon release. Piatt [18] found that this maneuver had a positive predictive value of 21% and a negative predictive value of 78% in patients for whom the diagnosis of shunt patency or malfunction was definite. This suggests that up to 22% of patients with shunt obstructions will have a normal pumping “test.” In addition, frequent pumping of the shunt can cause entrapment of choroid plexus in the proximal shunt tubing and lead to proximal catheter obstruction.

Further evaluation with a noncontrast computed tomography scan often requires comparison with the most recent prior study, if available. A plain radiograph of the skull, chest, and abdomen (“shunt series”) assesses the integrity of the shunt connection and can identify the components of the working system. However, normal radiographic studies should not supersede a strong clinical suspicion of a shunt malfunction based on symptoms and prior history [19].

It may be both diagnostic and therapeutic to “tap” the shunt under sterile conditions by inserting a 23- or 25-gauge butterfly obliquely into the reservoir [20]. (see Figure 4). Although this procedure should be done
Erosion or pressure necrosis from the shunt apparatus is more likely due to Gram-negative infections, due to bowel perforation. Infections that occur more than 6 months after shunt placement are generally due to antibiotic resistance. The most common etiologic organisms are Staphylococcus aureus and Staphylococcus epidermidis. They tend to adhere well to Silastic tubing [23,24]. Infections that generally occur within 2 months of shunt placement [22]. Most CSF shunt infections are perioperative in nature and are less likely to be due to antibiotic-resistant organisms such as methicillin-resistant Staphylococcus aureus (MRSA) [25]. Fungi are rare pathogens seen occasionally in premature infants.

Shunt infections can also include direct invasion. Necrosis or infection of the area around the reservoir can occur as a result of the constant pressure in infants or nonambulatory patients. Skin breakdown leading to visualization of the shunt mechanism is, by definition, a shunt infection and must be treated accordingly. Primary peritoneal infection may result in pseudocysts. These may be indolent in their presentation, and the shunt tap from the reservoir may not show evidence of infection. Shunt nephritis is a rare but serious complication of ventricular-atrial shunts, whereby renal deposition of antigen-antibody complexes leads to complement activation, which damages the renal tissue.

Children with CSF shunt infections do not necessarily present with signs and symptoms of meningitis. Fever is not always present in patients with shunt infections and is uncommonly the only sign. Meningismus is not often present, but patients often complain of lethargy or irritability. Children with shunts infected by Staphylococcus epidermidis may look particularly well despite the presence of bacteria in their spinal fluid [26]. The symptoms of shunt infection overlap considerably with those of shunt malfunction, and in fact, the infection can directly cause obstruction of the fenestrations in the catheters. Infection of the distal shunt mechanism may manifest as abdominal pain or vomiting. Patients with ventriculoperitoneal shunts who complain of abdominal pain, with or without fever, may benefit from abdominal radiographs and ultrasound to search for a loculated CSF collection or pseudocyst or visceral perforation.

In the absence of overlying infection, patients with suspected shunt infection should undergo aspiration of a small amount of CSF from the shunt system, performed by a neurosurgeon whenever possible. The absolute leukocyte count is not always diagnostic because counts can range from 0 to 2600 in the face of infection, and patients without infection can have a white blood cell count of up to 500/mm³ [27]. Gram stain of the fluid that demonstrates many white blood cells with a predominant presumptive organism may be helpful in diagnosing shunt infection, but should not be used to narrow broad spectrum antibiotic coverage until the culture and sensitivities of the causative organisms are obtained. In one series, 17% of patients with shunt infection had normal CSF gram stain, cell count, and chemistries [28]. A reasonable choice of empiric therapy in this era of methicillin-resistant Staphylococcus aureus (MRSA) is vancomycin and cefotaxime.

For treatment of proximal CSF shunt infections, medical therapy alone has been found to have a relatively low success rate compared with a combined medical-surgical approach [29]. Potential surgical interventions include immediate shunt replacement or the insertion of an extraventricular drainage catheter followed by delayed shunt revision.

Figure 4 “Tapping” a CSF shunt.

CSF Shunt Infection

Most CSF shunt infections are perioperative in nature and generally occur within 2 months of shunt placement [22]. Staphylococcus epidermidis and Staphylococcus aureus are the most common etiologic organisms. They tend to adhere well to Silastic tubing [23,24]. Infections that occur more than 6 months after shunt placement are more likely due to gram-negative infections, due to bowel erosion or pressure necrosis from the shunt apparatus.
Overdrainage
Occasionally, the CSF shunts can work “too well,” resulting in low ICP. “Overshunting” is more common in infants who have had initial shunting before 6 months of age. Young infants may exhibit sunken fontanelles, microcephaly, or overriding parietal bones, and older children may exhibit intermittent symptoms of headache, nausea, vomiting, and lethargy [21]. In contrast to symptoms related to increased ICP, patients with intracranial hypotension are often worse when in the standing position or after they are awake for several hours. Computed tomography scans may be unchanged from baseline or may reveal small ventricles. Oral analgesics are usually effective in the management of these symptoms, although some cases need to be addressed surgically by altering the resistance of the valve [30,31]. If left unchecked, overdrainage can lead to shrinkage of brain tissue and concomitant subdural hematomas or effusions. Similarly, a decreased rate of head growth because of overdrainage can result in craniosynostosis in the infant [32].

Other Complications
Patients with CSF shunts can experience benign postoperative leakage of CSF around the proximal shunt tubing into the subgaleal space around the reservoir. The resulting extra-axial fluid collection resolves spontaneously, so drainage of this fluid should be avoided. In patients who are not postoperative, a new extra-axial fluid collection can suggest shunt malfunction, as the CSF takes the newest “path of least resistance.”

Patients with CSF shunts have an increased risk of seizures, caused by epileptogenic scars [33]. They are more common in patients with other abnormalities correlated with seizures, such as porencephalic cyst or intracranial hemorrhage.

Rarely, the distal portion of a CSF shunt apparatus can migrate, causing damage to viscera or acting as a fulcrum for intestinal volvulus. Ascites and abdominal cysts can form as a result of drainage of excess fluid into the peritoneum. Increased intra-abdominal pressure can precipitate the formation of an inguinal hernia through a patent processus vaginalis.

Ventricular-vascular shunts can be associated with an increased risk of bacteremia and subsequent “shunt nephritis” from complement activation and renal deposition of bacteria. Bacterial endocarditis, cardiac foreign body, atrial perforation, cardiac dysrhythmias, and mural thrombus are rare but notable complications of vascular shunts.

Enteral Feeding Tubes

Background
Many CSHCN have difficulty with oral intake necessitating alternative routes for nutrition. The route selected is dependent upon the patient’s clinical situation. One must also consider the predicted duration of the nutritional support and the risk of aspiration. Nasogastric tube or gastrostomy tube (GT) feedings are preferred over nasojejunal tube or jejunostomy tube (JT) feedings. The nasogastric tube is appropriate for a patient whose duration of enteral feeding is expected to be less than 3 months. Long-term use is associated with esophagitis and nasal septum injury.

When a patient requires long-term enteral feeding, the most common mode of feeding is a GT. Indication for a GT is most often related to impaired swallowing due to a number of etiologies such as anoxic brain injury, congenital esophageal anomalies, and oropharyngeal muscle discoordination. A gastrostomy is a surgically created stoma that permits access to the stomach from the level of the skin. A jejunostomy is analogous to a gastrostomy but it brings the jejunum to the skin surface. Jejunostomy feedings are utilized preferentially when postpyloric feeding is required, such as in patients with delayed gastric emptying, recurrent aspiration pneumonia, and severe gastroesophageal reflux.

Enteral feeding via GT or JT has become more common recently. Therefore, it is recommended that ED physicians develop a level of comfort with GT and JT feeding. Equally important are the associated types of devices and the complications related to their use.

Mechanics of Gastrostomy and Jejunostomy Tubes

Both GT and JT may be inserted in one of two ways: (1) via open technique or (2) via percutaneous endoscopic gastrostomy/jejunostomy. Percutaneous endoscopic gastrostomy tubes were introduced in 1980 [34]. If the GT is inserted surgically, a left upper quadrant or midline incision is used to place the GT through the abdominal wall. The GT then passes through the anterior wall of the stomach and through the lumen of the stomach at the level of the fundus. The wall of the stomach is sewn to the abdominal wall where the tube makes its exit. Percutaneous endoscopic gastrostomy is the most popular of the nonsurgical procedures for placing a GT and it is thought to be safer and simpler than surgical gastrostomy [35]. This approach involves placing a tube into the stomach through a percutaneous opening in the anterior abdominal wall. An endoscope is used as a light source and illuminates the exact site on the anterior abdominal wall as a guide for needle puncture. Jejunostomy can also be completed via an open technique or percutaneously. In addition, jejunal feeding can be accomplished by placing a JT along with the GT. This method allows continuous jejunal feeding and enables venting of gastric air via the GT.
Equipment: Nasogastric, Gastrostomy, and Jejunal Tubes

The devices for enteral nutrition delivery are selected based on the route of feeding. There are several types of GT available. Most of these devices are made of polyurethane or silicone. The apparatus may vary in length, luminal diameter, the number of ports and lumens, and the type of catheter tip (see Figure 5). The “mushroom” types (Button by Bard Interventional Products Division, Billerica, Mass) have soft flexible tips that require a stylet to stretch the tip upon insertion. They have a single lumen. The “balloon-tip” devices (MIC-KEY, Medical Innovations Corporation, Draper, Utah) have become very popular and have replaced the mushroom-tip devices for the most part. The balloon is located at the tip of the device and is inflatable to 5 to 20 cc. The balloon-tip devices are easy to secure and do not dislodge as easily owing to the balloon anchor. The low-profile GT or button is the most recent advance in GT modalities (see Figure 6). It was developed to eliminate many of the constraints associated with the traditional GT. Buttons are made of silicone rubber and are self-retaining devices. The buttons have unidirectional antireflux valves that are fragile. A removable connector piece is attached to facilitate infusion of feeds and medications. Jejunal tubes that pass through the gastrostomy stoma are small-diameter tubes (8 F) (see Figure 7). These tubes require placement under fluoroscopy. Several types of surgical JT are available, including Malecot and MIC-KEY jejunal tubes. Gastric and jejunal tubes can be associated with a number of mechanical complications [36].

Enteral Feeding Tube Complications

Emergency department physicians should first perform a focused evaluation when a patient presents with a GT or JT problem. If the problem is isolated to the apparatus, the physician will be better able to offer efficient evaluation and therapy if he or she is familiar with potential complications. Complications related to GT and JT are divided into tube-related problems and problems associated with the stoma [37]. Further categorization is based on short-term and late-onset problems [38].

Tube Dislodgment

The most common reason a family seeks medical care for their child with a GT or JT is dislodgment. Dislodgment can be accidental or intentional. Accidental dislodgment usually occurs from a traumatic event, such as unexpected tension on the external tubing, rupture of the balloon, or unintentional balloon deflation. A GT or JT can deteriorate over time and require removal because of dysfunction. Tube obstruction that is not amenable to repair can necessitate intentional removal. When GT or JT dislodgment leads to an ED visit, one should realize that prompt tube replacement is crucial. Questioning the parents to determine the size of the replacement tube is
important but should not delay the replacement of the GT or JT [4]. If the tube size cannot be recalled, accessing the patient's medical record may provide the most recent tube measurements.

When a GT or JT has dislodged, one should carefully inspect the stoma for evidence of trauma characterized by bleeding or tears. The most important next step is placement of a Foley catheter if the tube size is unknown or unavailable. This will allow the stoma to be stented until the appropriate replacement tube is located. The choice of size of the replacement tube is guided by the length of time that has elapsed since the tube has been dislodged. If the duration has been long, the stoma may be constricted and require insertion of a smaller replacement tube. A series of progressively larger Foley catheters should be used to dilate the stoma if it is partially closed.

When evaluating patients with dislodged GT or JT, the time interval since initial placement of the gastrostomy or jejunostomy should be considered. Perioperative displacement (within 1 month of initial placement) is managed differently than tube dislodgment from a stable stoma. In perioperative cases, the ED physician should seek the input of the service (eg, interventional radiology, gastroenterology, pediatric surgery) that inserted the device. On the other hand, a tube that has dislodged from an older stoma should be replaced urgently with the same size and type of tube to avoid shrinkage of the stoma. When reinserting a GT, one must use caution to avoid inserting the tube into the peritoneal cavity through a false tract. A dislodged JT should be managed by the surgeon or radiologist, depending on the type of initial placement. Once the GT is replaced, it is important to check for proper location and functioning. The stomach should be aspirated with a syringe to obtain gastric fluid. Next, one should insert 10 to 15 cc of air while listening over the stomach for borborygmi. If the results are positive for both of the above, the use of gastrografin and radiography is not indicated [34].

**Tube Obstruction**

The second most common problem related to enteral tubes is obstruction of the lumen. This occurs as a result of accumulation of solidified formula or kinking of the tube. Caretakers report that they cannot easily infuse fluids. If formula or another liquid is suspected as the cause, gentle flushing should be attempted. Aspiration of the concretion may prove to be successful. Warm water is still recommended as the most effective fluid. Carbonated drinks have been used in this situation but their effectiveness is controversial. When a GT becomes obstructed, insertion of a stylet or other device is not recommended. Repositioning of the tube should be attempted if dissolving the concretion is not effective. However, if repositioning is not successful, removal and replacement are necessary. If the gastrostomy is new (placed within 1 month), the subspecialist who placed the device should be consulted before the ED physician removes the clogged tube.

**Leaking**

It is not unusual for a stoma to become widened over time. This stretching may lead to leakage of gastric or jejunal contents from the space around the tube. Once leakage is detected by caregivers and brought to medical attention, it is important to note what the fluid consists of (ie, formula, pus, gastric fluid). Of greatest concern is the drainage of pus signaling a stomal infection. If fluid appears to be leaking from the lumen of the tube itself, the practitioner should assess the tube position and check the balloon's status. If a low-profile GT or JT is leaking, one should consider problems with valve patency. When the stoma is judged to be widened, the options are to remove the tube briefly and allow constriction or to replace the existing tube with a larger-diameter tube. If the tube has dislodged traumatically, the stoma may be injured and stretched in this manner. This scenario requires surgical consultation and management.

**Gastroesophageal Reflux**

Many children with GT or JT have a preexisting history of gastroesophageal reflux disease (GERD). In some cases, placement of a GT actually worsens the GERD. This is attributable to the effect of bolus feeds directly into the stomach. The performance of a Nissen fundoplication precludes this problem. Patients with an exacerbation of GERD may present with an increase in vomiting, severe retching, and symptoms of esophageal irritation. In the event of these symptoms, initiating continuous enteral feedings may alleviate the problem.

**Gastric Irritation and Ulceration**

When the GT is too long or if the balloon becomes overinflated, friction results and gastric irritation occurs. On occasion, this can lead to ulceration of the gastric mucosa [39]. This situation is exacerbated when the stomach is empty. To avoid balloon over dilatation, one must use caution when administering medications, ascertaining that the correct port is utilized.

Gastric ulceration symptoms include those that are seen with peptic ulcer disease: abdominal pain, irritability, dark stools, coffee-ground drainage, or emesis. If a patient with a GT presents to the ED with presence of bright red blood or large amounts of coffee-ground content, saline lavage should be performed immediately. The standard approach to gastric bleeding should be followed if blood is obtained with lavage. If the fluid withdrawn is not bloody, medications such as H2-blockers, antacids, and Carafate should be administered. Upper endoscopy is a crucial adjunct. The GT should be removed and changed and the patient's symptoms and vital signs monitored carefully.
Gastric Outlet Obstruction

Rarely, the GT tip migrates into the pyloric channel resulting in partial gastric outlet obstruction [40]. This is quite rare but deserves mention so that the symptoms can be recognized. Even less common is migration of the GT into the distal esophagus, causing partial blockage of the esophagus. The child is noted to have retching or sudden onset of emesis and appears very uncomfortable. The GT needs to be radiographically located and retracted back to its proper location so that it is snug against the abdominal wall. If this procedure is not successful, the GT must be removed completely.

Stomal Complications

Irritant Dermatitis/Allergic Hypersensitivity

Irritation of the stomal skin is fairly common and results from recurrent leakage of gastric or jejunal fluid. If the stoma stretches, the leakage may increase, resulting in a more serious dermatitis. Although therapeutic when indicated, overzealous application of adhesives and stomal cleansing solutions may result in irritation or an allergic rash around the stoma.

If a patient presents with an irritated stoma, the skin should be thoroughly cleansed and dried before inspection. The area should be kept as dry as possible and the practitioner should recommend the use of barrier creams to protect the skin from further breakdown. Stomahesive Power (Convatec, Princeton, NJ) is effective when spread on to the stomal skin surface. It functions as an adsorbent and keeps the area dry.

Granuloma

Children with GT or JT may present with granulomas in the peristomal area. These lesions are usually painless but can occasionally become infected or develop bleeding. Rarely, the granulomas can cause blockage of the stoma. The best treatment is to apply silver nitrate to the granulomas.

Cellulitis

Occasionally, irritant dermatitis of the stoma can progress to stomal cellulitis. In general, the cellulitis is superficial and appears first as a skin irritation. However, if untreated, it can evolve into a deeper infection heralded by erythema, induration, tenderness, and swelling. As the peristomal infection progresses to a more diffuse picture, local symptoms and signs are accompanied by systemic symptoms. Manipulation of the tube and stoma becomes challenging because of the pain associated with the cellulitis. Patients with stomal and peristomal cellulitis require systemic antibiotics. The most common etiologic organisms are staphylococci and streptococci, which usually respond to a first-generation cephalosporin. Recurrent moisture and drainage can also lead to fungal skin infections. Topical clotrimazole is efficacious in this scenario. Rarely, peristomal cellulitis results in a peristomal abscess, indicated by a localized area of fluctuance. Incision and drainage are required before antibiotic administration. Methicillin-resistant Staphylococcus aureus must be considered when choosing antibiotic therapy.

Central Venous Access

Overview

Central venous catheters provide long-term venous access for infusion of medications and parenteral nutrition as well as for frequent sampling of blood in patients that require these interventions. Oncologic patients receiving chemotherapy, sickle cell patients requiring chronic transfusions, and patients with short-gut syndrome with the need for total parenteral nutrition are all examples of patients in whom the placement of a long-term indwelling central venous catheter may be advantageous [4,41,42].

Equipment

In general, the distal tip of a central venous catheter is located in the right atrium or at its junction with the superior vena cava [41]. The proximal insertion site of the catheter varies depending on the type of device used, but the most common sites utilized are the internal or external jugular, subclavian, or cephalic veins [42]. A portion of the catheter is then tunneled through the subcutaneous tissues to a site distant from the venous insertion point, which serves as the external access point. Most modern catheters are made of silicone rubber (silicone elastomer or Silastic), which offers some resistance to thrombus formation and infection [43] and causes less trauma on insertion [41].

There are two main types of central venous catheters: externalized (see Figure 8) and internalized (see Figure 9).

![Figure 8 Externalized or partially implantable central venous catheter. A, Female Luer lock. B, Clamp. C, Dacron cuff.](image-url)
The externalized (or partially implantable) central venous catheters include the Broviac, Hickman, and Groshong catheters. These are externalized after being tunneled through the subcutaneous tissue such that a portion of the catheter remains outside the skin. A female Luer lock is present at the most proximal end of the catheter and this may in turn be connected to most syringes and infusion devices. These catheters have a Dacron cuff present at the site of insertion into the skin, which stimulates the development of a fibrous sheath and thus aids in anchoring the device [8,42]. It may also serve as a mechanical barrier to infection [43]. The Groshong catheter deserves special mention because, unlike other forms of externalized devices, it has a valve on the distal tip that prevents back-bleeding. As such, external clamping and heparin flushes are not required (saline flushes are used instead) [8,42].

For the internalized (or implantable) devices, the proximal portion of the catheter is connected to a reservoir that is implanted in the subcutaneous tissue after tunneling. This reservoir consists of a self-resealing silicone membrane, which may be accessed through the skin, and a hard posterior surface. Common brand names for internalized devices include Port-A-Cath, Infuse-A-Port, and Mediport [8,42].

Accessing the Central Line

The principles of access are similar for both the internalized and externalized types of catheters. Aseptic technique is absolutely imperative to minimize the risk of infection. In general, no infusion should be started until patency is clearly established to avoid extravasation of caustic medications and solutions. Three to 5 mL of normal saline should be injected into the system, followed by withdrawal of 3 to 5 mL of blood to confirm patency. In general, 5 mL of blood should be withdrawn unused before withdrawing blood samples for laboratory tests. At no point should fluid be forcefully injected into the system. If resistance to the infusion is encountered or free-flowing blood is not obtained on aspiration, infusion should be stopped immediately to avoid rupture of the catheter or dislodgment of a thrombus. Syringes 3 mL or smaller should never be used because of the high pressures they can generate. To prevent air emboli, clamps must be kept closed at all times when any portion of the circuit is open. In addition, the entire IV circuit to be used should be flushed before connection to the system. The catheter should be flushed with 10 mL of saline between medications and with heparin after being accessed [4,8,42]. For externalized catheters, tincture of iodine should never be used as this may dry out Silastic catheters, leading to cracking and other damage. Avoiding the use of clamps and hemostats with teeth will also help to prevent damage to the externalized portion of the catheter [4,8,42]. These catheters also require heparin flushes daily in addition to flushes with each use. The exception would be Groshong catheters that only require saline flushes as mentioned above (see Figure 10).

For the internalized catheter device specifically, non-coring Huber needles should be placed directly into the reservoir to provide access. Because the skin must be penetrated with every access, a topical anesthetic such as EMLA (eutetic mixture of local anesthetics) should be applied at least 60 minutes before the procedure whenever possible (see Figure 11).

Catheter Occlusion

Occlusion may occur for many reasons. The catheter tip could be malpositioned, abutting a vessel wall. Thrombus formation within or outside the catheter may also be a cause. Certain infusions can cause precipitation of deposits within the lumen of the catheter. Waxy deposits, for example, have been noted to occur with the infusion of parenteral nutrition solutions containing mixtures of carbohydrates, protein, and fat. Specific medications such as phenytoin and diazepam can precipitate within the lumen of the catheter. Poorly soluble substances such as calcium and phosphorus may precipitate out of solutions [8,42].
When poor blood return is noted, certain maneuvers can be attempted that increase venous pressure and may facilitate phlebotomy. Examples include raising the patient's arms above the head, placing the patient in the reverse Trendelenburg position, or asking the patient to cough or perform a Valsalva maneuver.

If these strategies do not resolve the problem, a thrombus or precipitate should be more strongly suspected. An attempt could be made to irrigate and aspirate the thrombus with a small amount of saline (about 3 mL) infused gently in a back-and-forth motion. If care is not exercised, the clot could be dislodged and, if large enough, could lead to thromboembolic complications such as pulmonary embolism. Excessive force may also lead to catheter rupture. Should the problem still persist, recombinant tissue plasminogen activator and urokinase are two medications that can be used to dissolve fibrin clots within the lumen of the catheter [41].

In the case of other precipitates, specific agents have been used with some efficacy. For waxy precipitates from TPN, 70% ethanol is recommended, whereas for other particulate deposits, 0.1 normal hydrochloric acid has been suggested [8,42].

Infection

Tunneled catheters carry a lower risk of infection than those that are not tunneled [43]. Internalized devices (ie, the ports) in turn have a lower risk of infection than the externalized devices [43]. In general, the presence of an indwelling catheter places the patient at higher risk for infection; therefore, fever in the patient with such a device must be thoroughly evaluated, although in most cases, the likely cause may be an intercurrent viral or bacterial infection [8].

Catheter-related infections can be classified into three types that are not mutually exclusive: exit site infection, tunnel infection, and catheter-related sepsis/bacteremia [41,44]. The exit site infections are limited to the area of insertion of the catheter through the skin, are frequently caused by S epidermidis, and may not require removal of the device [41]. Infections of the tunnel or pocket are more likely to require removal [41,44]. Catheter-related bacteremia or sepsis may occur without any external signs of infection. Gram-positive bacteria such as S aureus, S epidermidis, and Streptococcus viridans are the most common culprits. Gram-negative infections are noted with increased frequency in patients receiving parenteral nutrition [8,43]. Immunocompromised patients are at risk for polymicrobial fungal and gram-negative infections [8,43].

In a patient presenting with suspected central line infection, the skin site should be carefully examined as part of the overall physical examination. Any tenderness, erythema, or purulent drainage at the site should increase the suspicion of line infection [44]. Blood cultures should be obtained from the catheter and, in many cases, from a peripheral vein as well [8,44]. A complete blood count with differential may be helpful. It should be noted, however, that a normal result does not rule out infection. Consider sending fungal cultures in immunocompromised patients [8].

Treatment consists of intravenous antibiotics (infused through the suspected catheter if it is patent) tailored to the spectrum of suspected organisms and their antibiotic susceptibilities. Gram-negative and gram-positive coverage is warranted. Vancomycin in combination with gentamicin is used at many centers. Ceftazidime may be added for patients who are neutropenic [8,44].

Catheter Displacement or Migration

Catheter displacement should be suspected if any of the following is present: Dacron cuff visualized outside the skin; mobility of the catheter inside the tunnel; fresh blood at the skin insertion site; and inability to aspirate free-flowing blood. Displacement is more common in externalized devices and tends to occur most frequently within the first 2 weeks after placement, that is, before the formation of the fibrin sheath at the Dacron cuff. Internalized devices may become displaced in rare cases of significant thoracic trauma. If the catheter becomes displaced from the reservoir, accumulation of blood in the subcutaneous space may result in an expanding painful lesion in the proximity of the reservoir. This requires urgent surgical consultation [8,42]. Catheter-tip migration occurs more infrequently and may result in cardiac dysrhythmias, pneumothorax, superior vena cava syndrome, or cardiac tamponade. Radiographic evaluation for line position is required (ie, plain films and contrast studies as necessary) in addition to surgical consultation and supportive management.

Catheter Disruption or Breakage

Catheter disruption may occur inadvertently during dressing changes or access procedures (eg, puncture of the line with a needle or accidental disruption with
scissors). It may also occur as a result of normal wear and tear \[8\]. Patients are unlikely to exsanguinate due to the natural tendency toward venous hemostasis. Management involves immediately clamping an externalized catheter and applying pressure at the site of venous insertion (not at the site of insertion into the skin) if active bleeding is occurring \[4,8,42\]. The external portion of the catheter should be cleaned with a povidone-iodine solution and covered with a clean dressing until repair can take place. Repair kits are available that include a new external catheter segment that can be fitted into the cleanly cut tip of the external portion of the catheter currently in place. Surgical consultation may be necessary.

**Air Embolus**

Signs and symptoms of air embolism include the sudden onset of tachypnea, hypotension, tachycardia, or loss of consciousness. Treatment involves providing supplemental oxygen, placing the patient in a left-sided Trendelenburg position, and immediately clamping the catheter \[8,42\]. To prevent this complication, the catheter circuit must be kept closed at all times when active infusion of treatments or withdrawal of blood is not taking place.

**Summary**

As more special needs children reside in community settings, emergency physicians will continue to play an important role in the overall care of these children. To this end, emergency medical services providers and ED physicians should have a familiarity in recognizing and managing acute complications of chronic disease states and in troubleshooting equipment problems. In addition to assisting with acute crises, the emergency provider can be instrumental in helping families with CSHCN integrate successfully into the community by reminding families of important measures that they should take to optimize their child’s medical care. In particular, families should be encouraged to have medical summary information and go-bags wherever they travel and to develop written emergency care plans in concert with their primary care provider. The Emergency Information Form co-developed by the American Academy of Pediatrics and the American College of Emergency Physicians is an excellent resource for families with CSHCN \[45\]. In addition, it is important for families of chronically ill and technology-assisted children to notify community emergency medical services departments and local utility companies of their residence \[5\]. Together, the medical community and families can partner to ensure optimal medical care and community integration of special needs children.

**References**


