Best Practices In The Emergency Department Management Of Children With Special Needs

Abstract

Children with special needs have a wide variety of physical and developmental challenges. These children often have medical devices, subtle presentations, and behavioral or psychiatric issues that demand consideration when delivering emergency care. Some of the more common issues include the evaluation and management of complications in devices such as a gastrostomy/jejunostomy feeding tubes, cerebroventricular shunts, and tracheostomy tubes, as well as impediments to the performance of common emergency procedures (eg, laceration repair, sedation for imaging) on children with developmental or behavioral disorders. This review will examine each of these circumstances and provide guidance on the best approaches to managing these patients.

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CME Objectives

Upon completion of this article, you should be able to:

1. Troubleshoot a gastrostomy tube that is not functioning.
2. Assess the best method of management of a hypoxic patient who has a tracheostomy.
3. Recognize the symptoms of ventriculoperitoneal shunt obstruction and manage it efficiently.
4. Describe strategies for managing sedation in the autist or developmentally delayed child for emergency department procedures.

Prior to beginning this activity, see “Physician CME Information” on the back page.

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Case Presentations

A 7-year-old girl with an intellectual disability arrives to the ED with her parents, who state that the girl has been lethargic all day, is not tolerating her gastrostomy tube feeds, and has some leakage and redness around the exit site of her tube. She has been vomiting nonbloody, nonbilious material with every feeding attempt. Her parents describe her breathing as “harder than usual,” with an increase in her baseline home oxygen requirement. In addition, they noticed some scant blood from her tracheostomy tube after suctioning thick whitish-yellow secretions from it. Her urine output has decreased from 4 times per day to 1 time per day. She has a low-grade fever to 38.2°C. Her past medical history includes premature birth at 25 weeks with a complicated NICU stay, including hypoxic-ischemic encephalopathy, grade IV intraventricular hemorrhage requiring placement of a ventriculoperitoneal shunt, gastrostomy tube placement, prolonged intubation with chronic lung disease, tracheostomy tube placement, a seizure disorder, and significant developmental delay (the child is nonverbal) with static encephalopathy. With her many problems, how do you evaluate the cause of this child’s vomiting, lethargy, and difficulty breathing?

A 3-year-old boy with Down syndrome is brought to the ED 1 hour after pulling out his percutaneous endoscopically placed gastrostomy tube, which was placed 2 years ago. The mother believes he broke the balloon when he yanked the tube out. His past medical history includes repair of a ventricular septal defect and failure to thrive. What are the best ways to manage the dislodged tube?

An 8-year-old boy with autism arrives to the ED with a 5-cm scalp laceration extending to the temporal area of the head that he sustained after falling from a 5-foot height. His parents report a loss of consciousness for 1 to 2 minutes as well as abnormal behavior. You decide he needs a CT scan to evaluate for intracranial injury, and the laceration will need to be repaired. How do you approach the sedation of this child so he can safely and effectively undergo the interventions he requires?

Introduction

Children with special healthcare needs are defined as children “who have or are at 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.”¹ An estimated 12% to 18% of children in the United States have special needs,²⁻⁵ and they account for 5 times the number of doctor visits as children without special needs.

Children with pervasive neurodevelopment disorders, autistic spectrum disorder (ASD), and developmental disorders, and behaviorally complex children are frequent patients in the emergency department (ED). These children often have complex medical histories and various comorbid conditions that complicate their care. In addition, these children may be limited in their capacity for cooperation and communication, making diagnosis and treatment difficult. In a Canadian study of patients with ASD who presented to the ED at a tertiary care center, 33 of 160 patients (20%) were admitted, 71% had an emergency triage score of 1 to 2 (suggesting high-acuity problems), and 79% had > 1 chronic health problem. Furthermore, the study found that patients with ASD comprised 2% of all ED visits. Parents of children with developmental disorders may also be more likely to use the ED as their medical home, especially if they do not have a well-established relationship with a primary care provider. In a survey of ED usage by children in the United States with developmental disorders, children with ≥ 3 ED visits were found to report less access to primary care sources than patients with no ED visits, who often reported well-established primary care.⁷

The term “children with special needs” covers a broad array of children, including those born prematurely, children who suffered hypoxia in the perinatal period, children with developmental and behavioral conditions, and medically complex patients, such as those with congenital cardiac and brain disorders, genetic disorders, inborn errors of metabolism, and mitochondrial diseases. This review focuses on identification and management of common problems with enteral feeding tubes, tracheostomies, and ventricular shunts, and will include a discussion on performing emergency procedures in children with autism and other cognitive disabilities.

Critical Appraisal Of The Literature

An extensive literature search was conducted on PubMed using combinations of the following search terms: special needs, children, disabled, technology dependent, epidemiology, prevalence, emergency department, gastrostomy, percutaneous endoscopic, complications, jejunostomy, migration, ventriculoperitoneal shunt, infection, cellulitis, malfunction, tracheostomy, tracheoinnominate fistula, tracheitis, mucociliary clearance, emergency procedure, bleeding, sedation, autism, developmental delay, virtual reality, child life specialist, autism, behavior, procedure, management, acute setting, imaging, MRI, agitation, and aggressive. Only articles whose subjects included children aged 0 to 18 years were identified. Abstracts were reviewed for relevance to the topic. Supporting articles were gathered from related articles and the reference lists of review articles. All relevant articles were selected, reviewed, and included in the bibliography. Overall, there is a dearth of literature in this patient population, and more studies are needed.
**Gastrostomy Tube Types And Indications**

Enteral feeding of children with special needs is necessary when a patient requires supplemental nutrition because oral feedings cannot be adequately tolerated. This is commonly due to swallowing dysfunction (eg, neurologic impairment, anatomic problem with the upper gastrointestinal tract, and pulmonary aspiration). Occasionally, nutritional needs exceed the child’s capacity to take in enough calories orally (eg, cystic fibrosis, congenital heart disease, and genetic syndromes that lead to failure to thrive). Some children are unable or unwilling to take a life-preserving medication or diet on their own (eg, metabolic diseases, ketogenic diet for refractory seizures, and medications with poor palatability in a young or uncooperative child), necessitating tube feeding.

Different types of tubes allow for enteral nutrition. Nasogastric (NG) tubes can be used for short-term (usually < 1 month) enteral nutrition. For children requiring long-term (> 1 month) enteral feeding, NG tubes are not an ideal solution, as problems of discomfort, dislodgment, and sinusitis complicate the course. For long-term enteral nutrition, gastrostomy tubes (G tubes) can be placed to create a direct connection from the stoma at the skin surface through the anterior abdominal wall, tracking directly into the child’s stomach. Percutaneous endoscopic gastrostomy (PEG) tubes are placed endoscopically under conscious sedation or general anesthesia by pediatric gastroenterologists. Open or laparoscopically placed G tubes are placed in the operating room by surgeons, and have the disadvantage of increased cost and recovery time from the procedure when compared to PEG tubes. While traditional G tubes with a length of external tubing are initially placed, they are often switched to a button-type tube that sits flatter to the skin. Button-type tubes, while more expensive than traditional G tubes, have a decreased risk of dislodgement from pulling or migration distally into the gastrointestinal tract than traditional G tubes.

Some children have severe gastroesophageal reflux or severely delayed gastric emptying leading to pulmonary aspiration, which makes placement of a G tube alone undesirable. For these patients, combining a G tube with gastric fundoplication surgery can theoretically mitigate the risks of aspiration. The fundoplication procedure involves wrapping the fundus of the stomach around the distal esophagus, thereby preventing reflux of stomach contents into the upper aerodigestive tract where contents can be aspirated. Among neurologically impaired children, fundoplication surgery has high rates of failure. In a study by Martinez et al, 71% of disabled children had recurrent symptoms (pneumonia, choking, gagging, vomiting) within 11 months of their operation. Among infants undergoing antireflux procedures concomitantly with G tube placement, there were no differences in hospitalizations for reflux-related illness 1 year after surgery. For these children, a reasonable and safe alternative to fundoplication surgery is the placement of a gastrojejunostomy (GJ) tube. GJ tubes have a proximal port entering the stomach and a more-distal port that feeds directly into the jejunum. They are usually placed under fluoroscopic guidance by a gastroenterologist or interventional radiologist. Medications (often requiring an acidic environment for ideal absorption) are usually given via the gastric port, while continuous drip feeds are given via the jejunal port. Bolus feeding into the jejunum is not feasible, as the osmotic load leads to diarrhea. The use of GJ tubes is advantageous, as potentially medically fragile patients can avoid fundoplication surgery, but if it becomes dislodged, kinked, broken, or clogged, replacement of the tube must be carried out by a specialist (usually fluoroscopic replacement by the interventional radiologist).

**Tracheostomy Tube Types And Indications**

Pediatric tracheostomies are performed in patients with congenital or acquired malformations of the airway or lungs, in premature babies who require prolonged ventilatory support, patients with acquired subglottic stenosis, children with neuromuscular disease requiring ventilatory support, and, rarely, as an emergent rescue airway. Tracheostomies are now more often being placed in children for prolonged intubation and less often for anatomic airway, obstructive, and infectious complications than in years past. Different brands of tracheostomy tubes exist, with Shiley® and Bivona® among the most common brands used in the United States. Tracheostomy tubes are described by 3 dimensions: the inner diameter, the outer diameter, and the length of the tube. Despite having the same inner diameter, different brands of tubes may have different outer diameters and different lengths. Some children have custom-made tracheostomy tubes that are unique to their anatomy. When the tube size is noted, it refers to the inner diameter in millimeters, in the same manner that endotracheal tubes are measured.

Tracheostomy tubes may be cuffed or uncuffed. Cuffs assist in decreasing air leakage in a patient who is being ventilated or who is at risk for aspiration. Before removal of a tracheostomy tube, do not forget to deflate the cuff. Some tubes are fenestrated with an aperture in the posterior aspect of the tube to facilitate movement of air through the vocal cords, allowing a patient to speak. An obturator is a stiff plastic piece that is inserted into the tube to maximize the stiffness of the tube during placement. When replacing a dislodged tube, make sure to remove the obturator prior to ventilating through the new tube.

**Ventricular Shunt Types And Indications**

Central nervous system shunts are placed to drain excess or poorly draining cerebrospinal fluid (CSF)
from within the ventricles, cyst, or subdural spaces of the brain to an area outside the cranium. Ventricular shunts are the most common type of central nervous system (CNS) shunt. Congenital reasons for shunt placement include anatomic malformations such as Arnold-Chiari malformation, spina bifida, aqueductal stenosis, arachnoid cysts, and Dandy-Walker malformation. Acquired causes of hydrocephalus requiring shunting of CSF include intraventricular hemorrhage (especially common in premature babies), meningitis (CSF uptake can be damaged in the arachnoid villi), traumatic brain injury, and brain tumors.

While the proximal end of the shunt lies in the ventricle, cyst, or subdural space, the distal end can terminate in a number of places. Most commonly, the distal end of the shunt ends in the peritoneum (as in a ventriculoperitoneal [VP] shunt). The distal end of the shunt has a 1-way valve, causing CSF to flow only into the peritoneal space and not retrograde. Retrograde flow can pathologically occur and lead to ascending infections with gut flora.

Certain clinical scenarios (eg, history of peritonitis) require the distal end of the shunt to be placed elsewhere. Ventriculocisternal, ventriculopleural, and even ventriculocholecystal (gall bladder) shunts have been performed when VP shunts are not usable due to prior infection or anatomic reasons. Ventriculocisternal shunt use has fallen out of favor due to potential complications of shunt-related thrombi, infection, and nephritis. Some shunts are programmable, allowing the pressure of CSF flow to be adjusted using an external device.

### Children With Developmental And Behavioral Disabilities

Patients with ASD, neurodevelopmental disorders, and other behavioral disorders frequently utilize the ED, as they may have more medical issues than developmentally normal children that arise from having a lower respiratory reserve, hemodynamic instability, or changes in acid-base balances when febrile illnesses strike (eg, congenital heart disease, metabolic disease). Additionally, if the patient is nonverbal, he or she may not be able to fully express how they feel physically and may present with vague symptoms such as crying, which may lead to an ED visit. These children require a different approach to management than children with behaviorally and developmentally typical needs. Depending on the level of impairment, these children may be unable to process information about their medical complaints and treatment, which makes caring for them challenging. This is common in ASD where patients have varying levels of impairment in social communication, repetitive restrictive behaviors, interests, and activities. Simply being in the ED exposes patients with ASD to a number of potentially upsetting triggers, including overstimulation with noise and touch, exposure to large groups of people, and disruption of their normal routine. This may lead to a patient becoming aggressive and agitated, making diagnostic and therapeutic interventions difficult for the emergency clinician.

### Differential Diagnosis

#### Gastrostomy And Gastrojejunal Tubes

Placement of PEG tubes is a generally safe procedure in pediatric patients. Complication rates in the early postoperative period occur in approximately 14% of patients, and they are mostly minor issues, such as surgical site infection or unintentional dislodgement. Gastrocolic or gastrocolocutaneous fistula, occurring when the transverse colon moves in front of the anterior portion of the stomach during PEG placement, is one of the most common early major immediate postoperative complications of PEG placement, and, based on data from the early 1990s, the incidence reported is 2% to 3%.

Other major complications (such as peritonitis, subcutaneous abscess, septicemia, death, and gastrointestinal bleeding) occurred at a rate of 5%, according to a study by Gauderer, while minor complications (which the authors of the study point out some would consider to be tube maintenance problems), such as dislodgement, leakage, G tube site infection, migration, obstruction, intussusception, and other complications (such as abdominal pain and vomiting), occurred in up to 73% of patients over a 5-year period.

Though the exact incidence of peritonitis is unknown, there are case reports of peritonitis following placement of the tube. Rarely, fungal infections can cause peritonitis after G tube placement. Peritonitis is the most common of the major complications of G tube placement, and it manifests as fever, vomiting, irritability, and abdominal rigidity. The diagnosis is easily missed in the child with neurologic disabilities. It can occur from occult leakage of gastric contents into the peritoneal cavity from either a replaced tube that has entered a false tract in the peritoneum or from subtle leakage into the peritoneum directly from the stomach with a correctly placed tube.

Dislodgement of G tubes is the leading reason for presentation to the ED, accounting for approximately 60% of all ED visits related to G tubes. This occurs by intentional or unintentional pulling of the tube by the child or caregiver, catching the tube on an object, or by malfunction of the tube itself from breakage of the balloon at the distal tip of the tube. In a head-to-head study of children receiving gastrostomy tubes versus button-type tubes, traditional G tubes were dislodged >3 times as frequently as button tubes. Breakage of all types of tubes occurs, as the plastic can crack with age or with repeated manipulation.

Clogging of the feeding tube can occur, and is much more common when feeding via GJ tubes.
pared to G tubes), as the smaller caliber and extended length of GJ tubes can cause increased obstruction of the lumen. Medications and formulas can cause sludging within the lumen of G tubes and GJ tubes, and the tube should be flushed with saline after these are administered.

Migration of a G tube can occur, leading to intestinal obstruction. On average, the length of the traditional G tubing or GJ tubing that protrudes from the stoma at the skin level is between 4 and 6 cm, but may be shorter on very small children and infants. Checking the centimeter markings on the outside of the tube can provide evidence that a tube has migrated inward (eg, if the 10-cm marking at the skin level is seen when the parent reports normally seeing the 5-cm marking at the skin level, there is indication that migration has occurred). Pulling back on a G tube until the balloon is against the abdominal wall can relieve obstruction due to migration. Rarely, G tubes can cause small bowel perforation or sepsis, or act as a lead point for intussusception.

“Buried bumper” is another uncommon complication in which the internal flange (the bumper) of the G tube is buried within the abdominal wall with symptoms that include leakage around the tube, pus, discharge or bleeding from the stoma site, stiffness on feeding (eg, difficulty advancing feeds into the tube), and inability to push the flange when trying to manipulate the tube.

Aspiration pneumonia should be considered as a cause of respiratory distress in a child with a G tube who has not had antireflux surgery (eg, Nissen fundoplication), or in a patient who has had an antireflux procedure, but, as a result of a loosening of the gastric fundoplication, stomach contents are able to ascend and be aspirated (known as a slipped Nissen).

Redness around the exit site can indicate the presence of granulation tissue, localized infection, or irritation from leakage of gastric contents around the exit site. Formation of granulation tissue is common postoperatively, and this fragile, shiny pink tissue can easily bleed if there is minor trauma or rubbing against the area. In a study by Naiditch et al, 58% of children undergoing G tube placement developed granulation tissue, and 89% of those children developed it within 4 months of the surgery. In the same study, 31% of patients developed leakage around the exit site. Leakage of gastric juices from the stoma occurs due to inadequate countertraction of the tube or the presence of large granulation tissue that prevents the correct positioning of the tube flush to the anterior abdominal wall. The incidence of leakage at the exit site of the G tube reported in the literature is 1% to 2%. Leakage accounted for 17% of ED visits (the second most common reason after dislodgement).

Cellulitis of the G tube site can occur, and symptoms typically include painful redness around the exit site with or without purulent discharge and fever. Bleeding at the exit site of the tube may be due to trauma from dislodgement or trauma to friable granulation tissue. Bleeding from within the tube may arise from irritation of gastric mucosa from the tube itself or from a primary upper gastrointestinal bleed from medications, peptic ulcers, swallowed blood, or Mallory-Weiss tears from retching or vomiting.

Table 1 summarizes possible differential diagnoses based on common G tube-related complaints.

**Tracheostomy Tubes**

Dislodgement of the tracheostomy tube is a potentially life-threatening emergency, and the patient may present in respiratory distress or acute hypoxia. Occlusion occurs most commonly when mucus or secretions block the lumen of the tube. Less commonly, granulation tissue or the placement of the tube into a false tract can lead to obstruction of the tracheostomy tube.

Bleeding from a tracheostomy tube is not uncommon, and it is most often caused by inadequate humidification of air entering the tracheostomy, from aggressive or frequent suctioning of the tube, from friable tissue resulting from acute infectious tracheitis, or from disruption of granulomatous tissue in the proximal airway during routine care or suction of the tube. A rare, but life-threatening, late complication of tracheostomy placement is a tracheoinnominate vessel fistula. This condition presents as bleeding from the tracheostomy site, usually 3 to 4 weeks after surgery. Patients may present with a sentinel small bleed or massive hemorrhage. The incidence of tracheoinnominate vessel fistula is between 0.1% and 1%. Tracheal mucosal necrosis from high cuff pressures and excessive movement of a malpositioned tracheostomy tube tip or cuff

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**Table 1. Differential Diagnosis Of Gastrostomy Tube Complications**

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Possible Tube-Related Diagnoses</th>
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</thead>
<tbody>
<tr>
<td>Vomiting</td>
<td>• Tube obstruction</td>
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<tr>
<td></td>
<td>• Tube migration</td>
</tr>
<tr>
<td></td>
<td>• Intestinal obstruction (adhesions)</td>
</tr>
<tr>
<td>Redness</td>
<td>• Cellulitis</td>
</tr>
<tr>
<td></td>
<td>• Gastric content leakage</td>
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<tr>
<td></td>
<td>• Granulation tissue</td>
</tr>
<tr>
<td>Blood at site or in tube</td>
<td>• Dislodgement</td>
</tr>
<tr>
<td></td>
<td>• Trauma</td>
</tr>
<tr>
<td></td>
<td>• Granulation tissue</td>
</tr>
<tr>
<td></td>
<td>• Ulceration of gastric mucosa from the tube</td>
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<tr>
<td></td>
<td>• Blood from other GI tract site</td>
</tr>
<tr>
<td>Pain at site</td>
<td>• Cellulitis</td>
</tr>
<tr>
<td></td>
<td>• Buried bumper</td>
</tr>
<tr>
<td></td>
<td>• Irritation from gastric content leakage</td>
</tr>
<tr>
<td></td>
<td>• Occult trauma</td>
</tr>
<tr>
<td></td>
<td>• Dislodgement of tube</td>
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</tbody>
</table>

Abbreviation: GI, gastrointestinal.
are all factors that are thought to lead to erosion of the tracheal wall into the innominate artery that lies posterior to it.\textsuperscript{32}

Tracheitis usually presents with fever, increased color or change in color of tracheal secretions, and increased frequency of suctioning required to maintain a patent airway.

Skin infections around the stoma site may occur, and they are usually caused by micro-organisms from the skin. Table 2 summarizes common complaints and possible tracheostomy-related diagnoses.

### Ventricular Shunts

Shunt failure rates within the first year of placement are 35% to 40%.\textsuperscript{33} Over the course of 15 years, 85% of patients with shunts will need a revision.\textsuperscript{34} Shunt obstruction occurs when a distal or proximal segment fails to drain CSF, and pressure increases in the cranium. Proximal shunt obstruction is more common than distal obstruction. Symptoms are the same as those seen in other causes of increased intracranial pressure (ICP), including headache, vomiting, and changes in behavior. Advanced symptoms may include signs of herniation, such as cranial nerve palsies (especially cranial nerve VI) and Cushing triad (hypertension, irregular respirations, and bradycardia). Although vomiting and headache are important symptoms that may indicate elevated ICP, bradycardia (without hypertension) may be a more sensitive and specific sign. One small study demonstrated that 45% of children with shunt malfunction had bradycardia (heart rate < 2nd percentile for age) compared to only 11% of controls. The positive predictive value (PPV) of bradycardia was 90%. Only reduced level of consciousness had a higher PPV for shunt malfunction (92%), while headache and vomiting each had a PPV of only 65%.\textsuperscript{35} Though epilepsy is a frequent comorbidity of patients with indwelling cerebral shunts, it is unclear whether isolated seizure activity is a symptom that should require evaluation of shunt failure. Rates of shunt revision requirements in patients presenting with seizures range from 2.9% to 28%.\textsuperscript{36-38} Among patients with VP shunt malfunctions who had underlying Chiari II malformations, neck pain was the presenting symptom in 41% of cases.\textsuperscript{39} Severe constipation can cause enough intra-abdominal pressure to lead to a clinical picture consistent with distal shunt obstruction. Relief of the constipation can abate the symptoms without the need for shunt revision surgery.\textsuperscript{40}

A broken or disconnected shunt is most likely to have occurred when there is direct trauma, the patient describes a popping sensation with sudden movement, or if there is localized swelling around the shunt site in the scalp or tubing. Once the disconnection occurs, and if the CSF fails to drain, clinical symptoms of shunt failure may follow. After obstruction, this is the second most common cause of shunt malfunction.\textsuperscript{41}

Infections of cerebral shunts are usually caused by \textit{Staphylococcus aureus} and coagulase-negative \textit{Staphylococcus} species. Gram-negative rods, \textit{Streptococcus}, \textit{Enterococcus}, and anaerobes may be implicated, although this is rare. Most shunt infections come from colonization at the time of placement of the shunt.\textsuperscript{42} Most (> 90%) of shunt infections occur within 6 months of shunt placement or revision.\textsuperscript{43} Symptoms of shunt infection include fever, headache, vomiting, irritability, redness over the shunt site, abdominal pain, and having a CSF fluid leak, although the presence of headache and vomiting alone are not independent risks for shunt infection.\textsuperscript{44} Historical risk factors for shunt infection include history of recurrent shunt infections,\textsuperscript{45} shunts placed for hydrocephalus due to hemorrhage, shunt placement, or revision within 90 days.\textsuperscript{43}

Abdominal pseudocysts are an infrequent complication of VP shunt placement, leading to vague complaints such as abdominal pain and vomiting, sometimes in the absence of headache or altered behavior. A collection of fluid around the distal shunt tip accumulates and is surrounded by tissue causing the presenting symptoms. In a substantial percentage of cases of abdominal pseudocysts, concurrent shunt infection is present (> 30% of the time in most reports).\textsuperscript{46,47} Shunt migration can lead to shunt malfunction. There are case reports of distal shunt tubing migrating through the bowel, bladder, into the scrotal sac, anus, chest, heart, and even spontaneous knot formation leading to obstructive symptoms.\textsuperscript{48-54} Table 3 (see page 7) summarizes common presenting symptoms related to VP shunts and their possible diagnoses.

### Table 2. Differential Diagnosis Of Tracheostomy-Related Complications

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Possible Diagnosis</th>
</tr>
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<tbody>
<tr>
<td>Hypoxia</td>
<td>• Tracheitis</td>
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<tr>
<td></td>
<td>• Pneumonia</td>
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<tr>
<td></td>
<td>• Dislodgement</td>
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<tr>
<td></td>
<td>• Obstruction by granulation tissue</td>
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<tr>
<td></td>
<td>• False tract</td>
</tr>
<tr>
<td>Fever</td>
<td>• Tracheitis</td>
</tr>
<tr>
<td></td>
<td>• Pneumonia</td>
</tr>
<tr>
<td></td>
<td>• Other infectious source</td>
</tr>
<tr>
<td>Bleeding</td>
<td>• Over-suctioning</td>
</tr>
<tr>
<td></td>
<td>• Tracheoinnominate vessel fistula</td>
</tr>
<tr>
<td></td>
<td>• Tracheitis</td>
</tr>
<tr>
<td></td>
<td>• Inadequate humidification</td>
</tr>
<tr>
<td></td>
<td>• Friable granulomatous tissue</td>
</tr>
</tbody>
</table>

### Children With Developmental And Behavioral Disabilities

Children with developmental delay or developmental disorders, ASD, and behavioral disorders may
have presenting symptoms that are different than developmentally typical children. There is a paucity of studies that focus on the presentation of acute illnesses, trauma, or mental status changes in these children in an acute care setting. Caregivers who routinely care for these children can be an invaluable resource to help delineate normal from abnormal behaviors. Presentation of common illnesses may have atypical features in this group of children. For example, a nonverbal child with ASD presenting with gastrointestinal symptoms may show signs of agitation, behavioral changes, wincing, grimacing, or other behaviors. These behaviors, coupled with decreased oral intake, constipation, vomiting, or diarrhea may be suggestive of gastrointestinal pathology. Children with complex chronic conditions have higher rates of severe sepsis and increased rates of mortality from sepsis compared to children without these conditions, so abnormal vital signs suggestive of sepsis should not be ignored.

Children with developmental and behavioral disabilities are often on several medications with myriad side effects. Changes in their behavior may be due to medication side effects, acute illness, intracranial processes (infectious, traumatic, anatomic), or occult accidental or nonaccidental trauma.

### Prehospital Care

Emergency medical services (EMS) providers may be contacted to transport patients with G tubes, tracheostomies, or cerebral shunts to the ED. They should be prepared to suction tracheostomies, and they should be able to provide respiratory support should a tracheostomy tube become dislodged. G tube issues usually do not require emergent care from a prehospital provider. Cerebral shunt malfunction may require the prehospital provider to administer basic and advanced life support, with an emphasis on managing increased intracranial pressure. Basic maneuvers, such as raising the head of the bed and providing rescue breaths may help in these situations. In a stable child with special needs, consider transporting the child to the nearest hospital capable of caring for children with complex needs. If the child is unstable, the nearest hospital should be the transfer destination.

### Emergency Department Evaluation

#### General Approach To The Initial Evaluation

Once airway, breathing, and circulation are assessed and stabilized, a more thorough evaluation can take place. When evaluating the child with special needs in the ED, conduct a thorough review of past medical conditions, ideally by reviewing a printed or computerized medical record. Noting baseline vital signs from recent prior well visits, oxygen requirements and saturations at baseline, and baseline mental status can be invaluable information. Listen to the caregiver’s assessment of how the child is doing, as they know the child’s normal behavioral baseline best. If they are concerned that the patient is not acting normally (eg, too agitated, too lethargic, temperature of 99°F is high for them), take these concerns seriously.

Occasionally, children with very specific special needs (eg, inborn errors of metabolism) may require very specific studies or interventions upon arrival to the ED. Ask the caregiver about this to see if they have a letter that gives clear specialized care instructions. Obtain a current medication list, and ask about recent changes in medications (eg, discontinuations, changes in doses, or additions of new medications).

The physical examination should take place with the child in a gown, on a gurney, in order to fully examine the patient. When it comes to the technology-dependent child, a deeper understanding of the equipment is paramount to successful care. In the more critical patient, inquiring about end-of-life care preferences, such as Do Not Resuscitate (DNR) or Allow Natural Death orders may be appropriate.

#### History And Physical Examination For Gastrostomy And Gastrojejunal Tubes

When a feeding tube is dislodged, not functioning properly, or there is concern about the site itself (bleeding, leaking, redness), important questions to ask the caregiver include: timing of initial placement of the tube, size of the tube (usually measured in French), type of tube (eg, button, PEG, CORPAK, mushroom tip, GJ), and how long it has been dislodged (if that is the complaint).

The integrity of the tract between stoma and stomach is usually well formed by 6 weeks postoperatively. Inspect the exit site on the skin for any bleeding, erythema, warmth, pus, or granulation

<table>
<thead>
<tr>
<th>Table 3. Differential Diagnosis Of Ventricular Shunt Complications</th>
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<tbody>
<tr>
<td><strong>Symptom</strong></td>
</tr>
</tbody>
</table>
| Vomiting | - Obstruction  
- Shunt migration  
- Disconnection of shunt tubing  
- Abdominal pseudocyst |
| Fever | - Shunt infection (if revised or placed within the previous 6 months) |
| Abdominal pain | - Pseudocyst  
- Bowel perforation  
- Shunt migration |
| Headache | - Obstruction  
- Shunt infection  
- Overdrained ventricles |
| Seizure | - Shunt infection  
- Obstruction (uncommon) |
tissue. Note the length of the tube at the level of the skin, as this might be an indicator of inward or outward movement of the tube. Ask about vomiting, and examine the patient’s abdomen for distension, rigidity, or tenderness. Vital signs should be noted, and the presence of fever and/or tachycardia should alert the emergency clinician to the possibility of infection at the exit site or due to peritonitis.

History And Physical Examination For Tracheostomy Tubes
Note the size (length and diameter) and type of tracheostomy tube, and have an extra tube ready at the bedside when the patient arrives, if time allows. If the tube is dislodged and the patient is in respiratory distress, consider plans for replacement of the tracheostomy tube or obtaining an alternative airway. Often these children have complicated airways, so having a backup airway plan (such as video laryngoscopy or anesthesia involvement) is advisable.

It takes 5 to 7 days after placement of a tracheostomy tube for the tract to mature. If a tube becomes dislodged after this period of time, attempt to replace it with the same size tube or a smaller tube. If this fails, traditional orotracheal intubation can be used emergently. In a more stable patient, the subspecialist can revise the tracheostomy surgically. Have suction and saline available for clearing mucus secretions in cases of occlusion of the tracheostomy tube. When replacing a tracheostomy tube, use water-soluble lubrication (rather than petroleum-based), in case the lubricant unintentionally enters the lungs.

If the patient has excessive bleeding from the tracheostomy site, an experienced otolaryngologist should evaluate the source of bleeding with a bronchoscope. If a tracheoinnominate vessel fistula is suspected, overinflate the balloon that surrounds the tracheostomy tube in order to occlude the bleeding vessel, or alternatively, place a curved clamp (or use a finger) into the tracheostomy hole and apply pressure anteriorly to occlude the vessel. Rarely, excessive granulation tissue will obstruct the tracheostomy tube. Replacing the tube is necessary in these cases. Be aware that tubes can be inserted into false tracts, so if a patient with a freshly replaced tube cannot be ventilated, you may need to reinsert the tube into the correct tract.

In cases of suspected tracheitis, send tracheal aspirate cultures, obtain a chest x-ray to rule out concomitant pneumonia, and review old cultures to assess which pathogens the patient has grown in the past to guide antibiotic choice. Beware that many of these tracheostomy tubes are colonized with bacteria. Symptoms and concurrent white blood cells on a Gram stain typically differentiate an acute infection from chronic colonization. Mucus plugging during an episode of tracheitis is a common cause of tracheostomy tube obstruction. Suctioning can be performed using an 8F suction catheter, which should fit inside most tracheostomy tubes.

History And Physical Examination For Ventricular Shunts
Important historical information includes the following: why the patient had a shunt placed, what type of shunt the patient has, when it was initially inserted, when it was last revised, the child’s typical symptoms during shunt malfunction (each child’s presentation may vary), and any history of shunt infection. Most cases of shunt infection (> 90%) will occur within 6 months of placement, revision, or tapping of a shunt. However, risks of shunt infection from tapping the shunt are, realistically, very low. A recent study of 542 shunt taps showed no infections related to a prior tap (among 14 overall VP shunt infections). Therefore, shunt infection should be placed farther down on the differential diagnosis of fever if > 6 months have elapsed since the shunt was surgically manipulated.

The initial assessment includes determining whether the patient is showing signs of impending herniation. Vital signs may demonstrate hypertension, irregular respirations, and bradycardia (Cushing triad) in cases of impending herniation. Note the patient’s level of consciousness, pupil size and reactivity, presence of posturing, and Glasgow Coma Scale score. In infants, a bulging fontanelle has a 44.6 likelihood ratio of being related to shunt failure. Rapidly increasing head circumference in this age group may also be a helpful data point indicating worsening hydrocephalus and shunt failure. Palpate for any disconnection of the shunt tubing running down the patient’s neck or subcutaneous fluid collection along the shunt route. This area is a common site for shunt disconnection.

Diagnostic Studies
Gastrostomy And Gastrojejunal Tubes
Checking the pH of the secretions suctioned from a replaced G tube can help confirm correct placement of the tube. Often, this is all that is necessary to confirm correct placement, if the replacement procedure was uncomplicated and the G tube tract is mature (> 6 weeks old). X-rays of the abdomen with contrast in the feeding tube can confirm placement of the replaced tube, if replacement was difficult, the tract is immature (< 6 weeks old), or there is difficulty obtaining gastric secretions from the newly replaced tube. Dye studies can also evaluate for obstructive processes. Approximately 5 to 10 mL of radiocontrast material is placed into the tube, and plain films are performed by ordering a G tube contrast dye study. If the outline of the stomach (or jejunum if there is a jejunal tube) is clearly seen, then the tube is appropriately positioned.

Tracheostomy Tubes
Tracheal aspirate cultures should be obtained in suspected cases of tracheitis. Chest radiographs are often
obtained to rule out pneumonia or pleural effusion when symptoms of tracheitis are present. Respiratory viral panels may aid in determining the etiology of an infectious tracheitis. If the patient is in distress, blood gases can be used serially to monitor changes in respiratory status. In cases of bleeding from a tracheostomy site, specialist evaluation with a bronchoscope is necessary to determine the source of the bleeding.

In the event of massive bleeding related to tracheoinnominate vessel fistula, diagnosis is traditionally made by bronchoscopy and angiography.

**Ventricular Shunts**

**Computed Tomography And Magnetic Resonance Imaging**

Head computed tomography (CT) or limited (so-called “fast”) MRI scans can be utilized to evaluate for ventricular dilation in association with shunt malfunction. It is necessary to compare old images when evaluating for changes in ventricular size, as changes in size are more important than the actual size. The radiation dose of CT scans can be minimized by up to 90% by decreasing the number of slices obtained (utilizing “thick cuts”) without sacrificing the ability to evaluate ventricle size. Alternatively, rapid-sequence brain MRI imaging provides information about ventricle size and VP shunt catheter tip placement without risk of radiation. Most protocols only require the patient to be still for 8 to 40 seconds, avoiding the need for sedation in most patients.

Some disadvantages of MRI include the cost of MRI (approximately $650 more than CT scans) and the potential need for resetting of programmable shunts after MRI is performed. Programmable shunts allow for the neurosurgeon to adjust the CSF pressure at which ventricular drainage will occur using an external electronic device.

**Plain Radiography**

Plain films examining the entire length of the shunt (a shunt series) are sometimes used to evaluate the integrity of the shunt tubing and look for breakages in the system, especially in the setting of external trauma to the shunt or local findings around the tubing (localized swelling near the tubing). Occasionally, tube migration is detected using plain films. There is evidence that a plain film shunt series is a low-yield study, showing the anatomic reason for malfunction in 4% to 5% of cases. In contrast, CT scan demonstrated abnormalities in almost all of those cases, making the shunt series uniquely responsible for picking up a shunt malfunction in only approximately 1% of cases.

**Tapping The Shunt**

Patients who have had past shunt infections are at risk for developing shunt malfunction without ventricular dilation, which occurs in 9% to 11% of pediatric patients. Slit ventricle syndrome is a group of disorders featuring severe headaches in patients with ventricular shunts that have been placed for the treatment of hydrocephalus and normal or smaller than normal ventricles seen on CT. Pathophysiologically, different processes may be responsible for the symptoms, including intracranial hypotension (analogous to spinal headaches from CSF overdrainage), intermittent obstruction of the proximal ventricular catheter, intracranial hypertension with small ventricles and a failed shunt (stiff ventricles that do not dilate), intracranial hypertension with a working shunt (analogous to pseudotumor cerebri), and shunt-related migraine.

Tapping the shunt may be necessary to evaluate for shunt infection and to determine flow and pressure within a shunt system. CSF obtained from these taps can be sent for culture, cell counts, protein, and glucose levels, as one would do in a routine lumbar puncture. Shunt tapping should be performed in a sterile manner. Prepare the scalp in standard sterile fashion, and access the plastic reservoir (usually in the right parietal area of the scalp) using a 23-gauge butterfly needle attached to 30 cm of tubing. Attach a 3-mL syringe to apply back pressure upon entering the reservoir if no CSF spontaneously returns. If CSF returns spontaneously or with < 1 mL of back pressure, that is considered good flow. Poor flow is demonstrated by being unable to obtain any CSF or if CSF can only be obtained with 2 to 3 mL of back pressure.

Determine opening pressure by measuring a column of CSF in the butterfly tubing or in a manometer that is held up vertically at a level above the child’s ear while the patient is in a supine position. CSF pressure is deemed to be elevated if it rises above 20 to 25 cm H₂O. Shunt malfunction is present if there is poor flow or elevated CSF pressure. Proximal shunt obstruction is usually the culprit in poor flow situations. Elevated pressure with good flow indicates distal obstruction or valve issues. In a small prospective study, among subjects with good flow but low pressure and concerning clinical presentation, 64% had a proximal shunt obstruction, so shunt tap results should not be taken in isolation without consideration of clinical context. Whenever possible, it is preferable for the neurosurgeon to perform this procedure, but in cases of impending herniation, it is important that the emergency clinician is capable of performing the procedure to acutely decrease ICP.

To perform this procedure, cleanse the scalp with povidone-iodine solution at the site under which you will puncture. Shunt tapping should be performed in a sterile manner. Prepare the scalp in standard sterile manner. Place the reservoir if no CSF spontaneously returns. If CSF returns spontaneously or with < 1 mL of back pressure, that is considered good flow. Poor flow is demonstrated by being unable to obtain any CSF or if CSF can only be obtained with 2 to 3 mL of back pressure.

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To perform this procedure, cleanse the scalp with povidone-iodine solution at the site under which you can palpate the shunt reservoir. Puncture the skin and reservoir with a 25-gauge butterfly needle, and attach a 5- or 10-mL syringe to the tubing. With slight negative pressure (not needed in cases of distal obstruction, where the CSF will forcefully emerge), remove CSF into sterile butterfly tubing (localized swelling near the tubing). Occasionally, tube migration is detected using plain films. To perform this procedure, but in cases of impending herniation, it is important that the emergency clinician is capable of performing the procedure to acutely decrease ICP.

To perform this procedure, cleanse the scalp with povidone-iodine solution at the site under which you can palpate the shunt reservoir. Puncture the skin and reservoir with a 25-gauge butterfly needle, and attach a 5- or 10-mL syringe to the tubing. With slight negative pressure (not needed in cases of distal obstruction, where the CSF will forcefully emerge), remove CSF into sterile butterfly tubing (localized swelling near the tubing). Occasionally, tube migration is detected using plain films. To perform this procedure, but in cases of impending herniation, it is important that the emergency clinician is capable of performing the procedure to acutely decrease ICP.
Shunt Flow Studies
In cases where CT scan and tapping the shunt has not yielded definitive results and patients have equivocal symptoms of elevated ICP, radionuclide studies (shunt flow studies or “shuntograms”) are performed to determine whether to perform a shunt revision. A small amount of radionuclide is injected into the shunt reservoir. A normal shuntogram must demonstrate filling of the ventricle, the shunt system must be entirely visualized, and the isotope must diffuse uniformly in the peritoneal cavity.73

Other Diagnostic Studies
Abdominal pseudocysts can be diagnosed by ultrasound or, less commonly, by CT scan showing a large fluid collection around the distal tip of the shunt.46 In properly functioning shunts, there should be scant or absent peritoneal fluid detectable on CT or ultrasound.74

Treatment

Gastrostomy And Gastrojejunal Tubes

Gastrostomy Tube Replacement
Tubes placed more recently than 6 weeks can be considered immature, in that the tract between the abdominal wall stoma and the stomach wall may not fully be formed. There is institutional variance as to when a tract is deemed mature, and this should be discussed with the service that placed the tube if there is doubt as to tract maturity. Dislodged G tubes with immature tracts should, ideally, be replaced by the specialist (surgeon or gastrointestinal physician) who placed the tube, but if he or she is unavailable, the emergency clinician can replace the tube, and confirmatory studies should be carried out to ensure the tube is within the stomach and that a false tract has not been created. Rates of complications of G tube replacement are low, with a pediatric study by Showalter et al showing complication rates of 1.2% (3 patients), none of which involved disruption of the tract as seen on dye study.75

Replacing the G tube should be done expeditiously, as delays will allow for stoma closure. Though not studied, some emergency clinicians apply a topical anesthetic (viscous lidocaine 1%) to the exit site area to, theoretically, decrease the pain of replacement. Replace the tube with one the same size as the one that was dislodged, but have a smaller one available in case the stoma has closed. If you cannot fit the original size tube into a narrowed stoma, start with a smaller-sized Foley or replacement tube and dilate it using progressively larger tubes or a set of Hegar dilators until the original tube size can be used. Replacement tubes can be a simple Foley catheter, traditional G tube, or button G tube, depending on availability. Do not force the replacement tubes in, to avoid creating a false tract. Once replaced, clinical confirmation of correct placement can be checked by suctioning gastric contents and checking the pH with litmus paper. Bedside ultrasound confirmation of tube placement has been described as well.76

There is a dearth of literature and no current standard of care for the exact indications for obtaining confirmatory studies. Confirmatory studies may include pH testing of gastric contents suctioned from the tube, and dye studies where a small amount of radiocontrast material (5-10 mL) is placed into the tube and plain films are taken. Use of dye studies to confirm placement is variable among institutions, but this routine practice may not be necessary if one can clinically prove the tube is properly placed. Evidence is limited, but the retrospective study by Showalter et al of 237 children undergoing replacement of G tubes demonstrated only 3 misplaced tubes. All 3 had no clinical confirmation documented on the chart and had symptoms of intestinal obstruction from an overfilled balloon (1 patient) and migration of the tube past the pylorus (2 patients).75

Gastrojejunal Tube Replacement
In cases of GJ tube dislodgement, contact an interventional radiologist to replace the tube under fluoroscopy or transfer the patient to an appropriate center for tube placement. If you cannot replace a tube definitively, at bare minimum, place a small catheter in the stoma to maintain patency until a more definitive feeding tube can be placed.

Removal Of Gastrojejunal Tube Obstruction
If a tube becomes clogged (which is especially common with the long narrow lumen of the GJ tube), a common practice (but unsubstantiated by compelling research) is to dwell room-temperature carbonated liquid for 15 minutes followed by a saline flush.77 Alternatively, pancreatic enzymes can be used to achieve the same relief of tube obstruction. If this fails, consultation with an interventional radiologist or surgeon may be necessary to replace the GJ tube.

Correcting Leakage At The Stoma Site
In cases where there is leakage at the stoma site, replacing the tube with a larger one may result in a progressively expanding stoma, and it is not recommended. In these patients, correct probe position and balloon inflation should be verified by pulling the tube until the balloon is snug against the internal stomach wall.78 Skin-protecting medications (such as liquid sucralfate) can be placed on the skin surrounding the exit site.79 Topical agents such as magnesium hydroxide or menthol/zinc oxide can be used as adjuncts as well. When the leakage persists for several days, the G tube can be removed (in consultation with the specialist who placed the tube) to allow partial stoma closure, and a new PEG tube can then be positioned through the same site.80 Granulation tissue may be treated using silver nitrate sticks (to stanch
minor bleeding) or topical steroids.

Irritation from gastric content leakage can sometimes be mitigated by ensuring that the tube is flush against the anterior stomach wall. This can be done by first ensuring that the balloon is inflated, and then gently pulling the G tube portion outward until resistance is met, followed by pushing the external retention disc down toward the skin. This helps to seal any defects in the space between the stoma and the part of the G tube within the stomach lumen, and it prevents further leakage of stomach contents. Place a gauze pad between the disc and the abdominal wall skin. Additionally, topical sucralfate, magnesium hydroxide, or menthol/zinc oxide may be used to relieve the irritation of acidic stomach contents on the skin.

**Infection**

Oral antibiotics covering skin flora are usually adequate to treat simple peristomal cellulitis. If the child is ill-appearing, there is a large area or rapidly progressing cellulitis, or the child is vomiting (and unable to take oral antibiotics), admission for intravenous antibiotics is indicated.

**Tracheostomy Tubes**

Replace tubes that have broken cuff balloons. Tracheostomy tube ties should be applied to prevent accidental decannulation of the airway once a tracheostomy tube is replaced and location is confirmed by x-ray. Suction the tube if there is evidence of secretions or occlusion from mucus. The use of albuterol may help to break up mucus plugging, as beta-2-adrenoceptor agonists increase beat frequency of cilia and may, therefore, facilitate mucociliary clearance. Humidification can help thin secretions that are thick.

Treatment of a tracheoinnominate fistula includes surgical diversion of blood flow and endovascular interventions, such as stent grafts, until more definitive surgical therapy can be instituted. Temporizing measures may include inflation of the cuff of the tracheostomy tube to occlude the innominate vessel.

Antibiotics, ideally based on prior respiratory cultures, can be given enterally or intravenously (for more severe cases) to treat tracheitis and bacterial pneumonia. Oral or even topical antibiotics covering skin flora (for mild infections) are usually adequate to treat peristomal cellulitis. Ill-appearing children or children who are unable to maintain hydration or take medications may require admission for intravenous antibiotics.

**Ventricular Shunts**

**Shunt Obstruction**

Shunt obstruction is treated with revision of the shunt, either the distal segment for distal or valve obstructions or a total shunt revision for proximal shunt obstruction. In cases of possible shunt obstruction in which imaging is normal or flow studies are normal, but symptoms persist or worsen, it is reasonable to admit the patient for observation and undergo delayed revision. Emergent removal of CSF from the shunt reservoir (ie, tapping the shunt) can acutely reduce ICP. Hypertonic saline or mannitol can theoretically be life-saving in cases where herniation is imminent, though there are no studies examining the efficacy of these medications in this particular clinical scenario. While awaiting definitive surgical treatment, keep the head of the patient’s bed elevated to 30° and maintain systemic blood pressure within a normal range to avoid a decrease in cerebral perfusion pressure.

**Shunt Infection**

Shunt infections are treated by removing the infected shunt, placing an external ventricular drain, and administering intravenous antibiotics. The use of intravenous antibiotics in conjunction with removal of the infected shunt has been shown to be superior to the use of intravenous antibiotics alone without shunt removal. Some centers also give intraventricular antibiotis to provide antibiotics directly into the CSF, but this is not without controversy. There are reports of antibiotic-related toxicity and elevated endotoxin levels in infants with meningitis and ventriculitis who received intraventricular antibiotics when compared with patients who received intravenous antibiotics alone. However, other studies have demonstrated higher long-term cure rates using intraventricular antibiotics without increased morbidity.

Antibiotics should initially include coverage for gram-positive and gram-negative organisms until culture results are known. Review past cultures, if available, and use these to guide therapy. Due to the increasing incidence of methicillin-resistant *S aureus*, vancomycin should be considered, often as the first-line therapy. If an abdominal pseudocyst is present, consider empiric addition of an antibiotic that covers anaerobes and coliforms. Although antibiotic-impregnated shunt systems are initially more expensive than standard shunts, they reduce shunt-related infections without breeding antibiotic-resistant organisms, and they may save money spent on revisions and hospitalizations for shunt infections.

**Abdominal Pseudocyst**

Abdominal pseudocyst treatment involves seeking and treating the underlying infection of the shunt, as well as replacing either the distal part of the shunt (when no infection is present), or completely replacing the shunt (when infection is present and after antibiotic therapy is completed). If infection cannot be eradicated or the peritoneum is no longer suitable for the distal shunt, alternative sites can be used (eg, pleural space, right atrium, gall bladder) for the distal segment of the new shunt.
Children With Developmental And Behavioral Disabilities

Behaviorally complex children can be challenging to manage, due to their lack of ability to communicate and their varying levels of cooperation. Patients may be nonverbal and/or unable to understand the need for diagnostic imaging or a simple procedure. Because of this, patients with ASD or developmental disorders may require sedation for diagnostic procedures and therapeutic interventions that developmentally normal children would tolerate without such measures.

Nonpharmacologic Measures

Patients with ASD may be amenable to nonpharmacologic methods of management. There are few studies on communicating and managing behaviorally complex children with neurodevelopment disorders. Seasoned caregivers can offer valuable insight into what works in a given situation. Recommendations from experts in the management of these children suggest several different techniques. Communicate in a manner consistent with the patient’s primary method of communication. A reward-based system, such as giving toys or treats to the patient after cooperation, may be an effective way to promote desired behaviors. Address the need for routine and sameness for the child. While it is difficult to recreate the child’s normal environment in the ED, there are some steps (as outlined in Table 4) that can be taken to increase cooperation in the ED. If available in your ED, a child life specialist can be a valuable resource in communicating with these patients exactly how a procedure will be performed. Setting expectations and laying out the plan of care with the patient and caregiver is a way to familiarize the patient with the procedure and minimize unwelcome surprises. Use of a child life specialist may help decrease stress in children undergoing painful procedures such as venipuncture and laceration repair, but no large controlled trials have been performed for children with developmental disorders or ASD.99,100

Sedative/Analgesia Considerations

When nonpharmacologic interventions are not possible, or when they fail, then sedation may become necessary to safely and effectively care for these children. In selecting the level of sedation required, it is important to consider the level of cooperation by the patient to perform a needed procedure. Simple procedures such as intravenous line placement or splinting may only require minimal sedation (anxiolyis) with oral agents. Minimal sedation allows patients to continue to respond to commands and maintain cognitive function, and it requires minimal intermittent observation rather than continuous monitoring. In general, oral agents are effective for minimal sedation. Moderate sedation is necessary when a depressed level of consciousness is needed to obtain patient compliance. Moderate sedation maintains a level of consciousness and patient cooperation to verbal commands and tactile stimulation. Drugs used should be such that there is low risk of the patient slipping further into losing consciousness and into a level of deep sedation.103 Deep sedation is a state of depressed consciousness where the patient is difficult to arouse and responds only to repeated verbal commands or vigorous stimulation. Deep sedation requires a higher level of monitoring with personnel trained in the management and rescue of sedated children.104

Determine the level of sedation needed for a given procedure using the lowest dosage of medication necessary in order to reduce adverse events. In behaviorally complex children with developmental disorders or ASD, sedation with oral agents can be effective, allowing for minimal monitoring of the patient. Titration of oral agents can be challenging, as absorption of these agents can vary among patients. If a higher level of sedation than desired occurs, the emergency clinician should be prepared to switch to an increased level of monitoring as required for moderate sedation levels.105 If a patient requires more than minimal sedation, the physician should be prepared for appropriate cardiac and respiratory monitoring, including continuous electrocardiography, oxygen saturation monitoring, airway interventions, and, if available, capnography. When selecting an agent for sedation, the following should be considered: the level of sedation desired, analgesic effect, amnestic effect, pharmacokinetic profile, and side-effect profile.

There are few studies related to the use of specific medications for procedural sedation in behaviorally complex children with neurodevelopmental disorders. In a study assessing sedation for MRI among developmentally normal children versus children with developmental disorders, of 486 patients aged 1 to 18 years, 53.5% of patients were classified as having developmental disabilities. Patients were sedated with pentobarbital and fentanyl. Results revealed no difference in the doses required for sedation between the 2 groups. Children with developmental disorders had a threefold increase in hypoxic events (defined as oxygen saturation [SaO2]...
<93%) compared to normal children. However, in a follow-up case-control study by the same authors, 140 children aged 3 to 10 years with developmental disorders had no increase in medication requirement with pentobarbital and fentanyl or pentobarbital and midazolam, when compared with developmentally normal children. This study also showed no increase in hypoxic events (defined in this study as $\text{SaO}_2$ of < 90% for > 30 seconds requiring airway intervention), compared to the control group. The differences in hypoxic events in these 2 studies are likely related to differences in definitions, with the latter study having a lower threshold for the definition of hypoxia and reporting of hypoxic events within the studies. The latter study suggests that there is no increase in hypoxic events among children with developmental disorders, and that they may be as sedated as developmentally typical children in the ED setting. Furthermore, Ross et al reported that medication requirements for sedation of autistic children were no different than developmentally typical children, showing that there was no increase in medication requirement when sedated with pentobarbital. In fact, the ASD group had a decreased requirement for fentanyl when compared to controls.

Sedation for children with developmental disorders may have some key differences compared to sedation in the general pediatric population. Physically, airway diameters in children with developmental disorders under pentobarbital sedation for MRI have been shown to be smaller than those with normal development, by 40% at the level of the palate. Theoretically, this may be related to abnormal muscle tone.

There are some anatomic differences in specific syndromes that should be considered prior to moving forward with sedation. Patients with craniofacial abnormalities (e.g., Crouzon, Apert, Pierre-Robin sequence, and others) may have difficult airways should the sedation become complicated. Children with Down syndrome tend to have smaller chins and larger tongues, and they can become a difficult airway. Airway anatomy (laryngeal mask airways, video-assisted laryngoscopy, emergency tracheostomy kit, or an anesthesiologist) should be arranged before sedating these children. Alternatively, one may consider performing the procedure with an anesthesiologist in the operating suite, if the risk of a difficult airway is deemed to be too great for the ED setting. The less likely that the patient will be able to control his behavior, the deeper the level of sedation required. Children aged < 6 years and children with neurodevelopmental disorders are more likely to require a higher level of sedation compared to older children with developmental disorders and developmentally typical children. However, despite these differences, as shown earlier, children with developmental disorders can be safely sedated when required, if a standardized, ordered approach to minimize adverse events is taken.

**Painful Procedures**

**Topical Anesthetics**

The use of topical anesthetics for painful procedures should be maximized if a painful procedure is planned. Lidocaine, epinephrine, tetracaine (LET) gel is useful for open wounds, allowing for effective analgesia with minimal stress to the patient. Apply it directly to an open wound and allow it to seep in for at least 30 minutes prior to cleansing and repairing the wound. Transdermal lidocaine can also be administered through a needle-free system for placement of intravenous lines or for venipuncture, although the system makes a popping sound that the child may find disagreeable. Topical lidocaine creams can be used on intact skin for venipuncture or lumbar puncture, if transdermal lidocaine is deemed to be unnecessary.

**Opioids And Benzodiazepines**

When selecting a sedating agent for a painful procedure, using an agent or a combination of agents that provides analgesia is preferable. Opioids are a good choice when performing painful procedures, as they not only offer pain control, but also act as a hypnotic. While usually well tolerated, opioids have potential side effects. Respiratory depression, bradycardia, and hypotension may occur. They should be used carefully in any hemodynamically unstable patient or in patients at risk for hypoventilation. They may also cause constipation, urinary retention, pruritus, rash, nausea, and vomiting. Opioids do not have amnestic properties, and if amnesia is desired, then coadministration with another agent, usually a benzodiazepine such as midazolam, is a good choice. Opioids and benzodiazepines may be dosed intramuscularly, orally, or intravenously, making them helpful in large autistic or aggressive patients in whom initial intravenous line placement may not be possible.

**Ketamine**

Another agent useful for sedation for painful procedures is ketamine. Ketamine is an N-methyl-D-aspartate (NMDA) antagonist, phencyclidine derivative with both amnestic and analgesic properties. Ketamine is a sympathomimetic agent with effects on systemic blood pressure and potentially cerebral perfusion pressures. Other effects of ketamine include bronchodilation, as well as increased respiratory secretions. Care should be taken in neurologically impaired patients who are unable to control their secretions.

Ketamine can be given intravenously, intramuscularly, orally, or intranasally, making it a very flexible agent. It offers good analgesia, making it an effective choice for fracture or dislocation reductions, as well as for prolonged or complicated laceration repairs. Given orally, it is an effective choice for nonpainful procedures, such as...
MRI. One case study showed it to be a good option when used in conjunction with midazolam for an aggressive autistic patient.102

**Nonpainful Procedures**

For nonpainful procedures in the ED setting (such as obtaining a CT scan), there are several options. Inhaled nitrous oxide, barbiturates, benzodiazepines, propofol, dexmedetomidine, and etomidate, either as single agents or in combination, are options for sedation, despite there being few studies that focus on sedation in patients with developmental disorders. Nitrous oxide should be used with caution in patients with seizure disorders.103

**Benzodiazepines**

Benzodiazepines have several benefits for sedating behaviorally complex children and children with developmental disorders. Benefits include sedation, anterograde amnesia, anxiolysis, and muscle relaxation. This makes them useful in many different situations, including in agitated autistic patients, anxiolysis for MRI, or in conjunction with opioids for painful procedures. Benzodiazepines work on gamma-aminobutyric acid (GABA) receptors in the brain, decreasing the rate of depolarization of neurons. This makes them useful agents in epileptic patients as well. Common adverse effects are hypotension and hypoventilation.

Midazolam is a short-acting agent that is beneficial for sedation in children with developmental disorders. It may be administered intravenously, intramuscularly, intranasally, orally, or buccally, and it has a rapid onset and rapid metabolism by the liver. Midazolam can cause idiosyncratic paradoxical excitatory reactions, such as hallucinations, disorientation, uncontrollable crying or verbalization, agitation, restlessness, involuntary movement, self-injury, and aggressive or violent behavior. These reactions usually occur within 5 minutes of administration and are more likely in younger patients.104 These reactions do not usually resolve with time and will require another agent to treat them. Several agents have been shown to be effective in treating paradoxical reactions, including ketamine, flumazenil, propofol, increased doses of midazolam, and haloperidol. One study showed that ketamine was effective in the treatment of paradoxical midazolam reactions when compared to control groups of placebo or an increased dose of midazolam.105

**Barbiturates**

Barbiturates (such as pentobarbital, methohexital, and thiopental) may be used in children with developmental disorders for procedural sedation or sedation for nonpainful procedures. They have a neuroprotective effect, making them ideal agents in the setting of seizures or head trauma with elevated ICP.106 Methohexital and thiopental each have an onset of effect within 1 minute, but the duration of effect for methohexital is 5 to 7 minutes, whereas the duration of thiopental is 10 to 30 minutes. Pentobarbital may last 15 to 20 minutes when given intravenously, and it has been shown to effectively sedate children for diagnostic imaging with a success rate of up to 98%.107 Despite its success rate, however, pentobarbital was shown to have a higher incidence of paradoxical reactions (such as agitation) after administration, compared to chloral hydrate.108 In a study comparing sedative medications for functional MRI, pentobarbital was found to have better sedative effect than propofol; however, it had a higher rate of adverse events.108 Adverse events of pentobarbital include myocardial depression, hypotension, and respiratory depression. Thiopental can be used in patients who require rapid procedures, such as CT or rapid sequence intubation. It may be given intravenously or rectally, and the side effect profile remains the same as with pentobarbital.109

**Propofol**

Propofol is an excellent medication for procedural sedation for children in the ED setting or for diagnostic imaging. It has a rapid onset and offset, and it is well tolerated. It may be easily titrated for moderate or deep sedation. Its rapid metabolism and rapid patient recovery allows the emergency clinician to continue neurologic examinations post sedation. Propofol has been shown to be efficacious for MRI in pediatric patients. It offers a titratable level of sedation with minimal movement by the patient, allowing for shorter MRI times and better image quality compared to dexmedetomidine.110 Propofol has some possible side effects, including hypotension, bradycardia, and respiratory depression. It does not have any analgesic effects, but it is often used in combination with opioids for painful procedures. One uncommon side effect is propofol infusion syndrome, which results in a lactic acidosis, rhabdomyolysis, cardiac dysrhythmias, hyperlipidemia, and multisystem organ failure in patients who are continuously infused with propofol for > 24 hours. While rare, this complication is potentially fatal. Therefore, caution should be used in pediatric patients requiring prolonged sedation.111,112 Though not reported in the literature, it seems prudent to avoid propofol in children with special needs where there is the potential for lactic acidosis (such as certain inborn errors of metabolism or mitochondrial diseases), or in patients with underlying cardiac dysrhythmias.

**Dexmedetomidine**

Dexmedetomidine may be a good choice for sedation of children with ASD and developmental disorders. It is an alpha-2 agonist that works in the central nervous system. It can be given orally for short procedural sedation or intravenously for prolonged sedation in intubated patients. Its efficacy in patients
Chloral Hydrate

Chloral hydrate has been a commonly used agent for sedation for diagnostic imaging. However, due to its side-effect profile, variable bioavailability, and variable sedative effects when compared to other agents, its use has decreased in the ED setting. Despite decreased use, it still remains an option for emergency clinicians who are comfortable with its administration, which can be either oral or rectal. Oral administration has a more consistent absorption profile than rectal. It is believed to work in the CNS by increasing GABA type-A receptor activation. The onset of action is usually within 30 to 60 minutes, and the duration of action is 4 to 8 hours. Repeat dosing at half of the initial dose is often required to achieve sedation.

Common adverse effects include vomiting and diarrhea. It has also been associated with paradoxical reactions of hyperactivity and anxiety. Rarely, it may cause respiratory depression or airway obstruction. Chloral hydrate has a 95% success rate for moderate sedation, with a rate of adverse events of 1.8% in a retrospective review by Delgado et al. Overall, it is considered a safe option for sedation for children, but its sporadic absorption and sometimes prolonged duration of action makes this agent a less useful choice in an ED setting.

Nitrous Oxide

Nitrous oxide is an option for procedures where a noninvasive means of sedation is desired. It is an inorganic inhalant that is generally well tolerated. It has a rapid onset, and it can be titrated rapidly. It can achieve amnesia while maintaining hemodynamic stability. As a single agent, nitrous oxide easily achieves minimal sedation and does not require continuous monitoring at minimal sedation levels. In a prospective observational study of 7802 procedures on children aged 33 days to 18 years, children were administered 70% nitrous oxide for various procedures. The overall adverse event rate was 4.3%, with the majority of adverse events being nausea, vomiting, and diaphoresis. The authors of the study also reported 9 potentially serious adverse events; however, these all resolved without complication.

There is evidence that nitrous oxide can be administered without fasting or postoperative monitoring. A retrospective review looked at its efficacy and safety in 1058 children who received nitrous oxide as a single agent when undergoing elective procedures. Procedures included incision and drainage of an abscess, cyst, or mole removal, foreign body removal, and central line removal. The patients received a topical anesthetic at the procedure site and were asked to eat within 2 hours of the procedure. Their findings showed that 98.2% of patients had no adverse events. The most common adverse event was vomiting, occurring in 0.7% of the patients.

There are case reports in the literature of nitrous oxide being temporally related to seizures. However, these events are rare, and no causality has been established. Nitrous oxide has been routinely used in dental offices for patients with seizure disorders, without reported ill effects, but caution is urged in using nitrous oxide in patients with known seizure disorders.

Given the ease of administration and overall safe profile of nitrous oxide, it is another reasonable option for sedation in pediatric patients, including children with developmental disorders, ASD, and behaviorally complex children, or when intravenous access is not established.

Alternate Routes Of Sedation/Analgesia

Although not formally studied in children with developmental disorders or ASD, intranasal fentanyl, ketamine, and midazolam are promising options for effective administration of analgesia and anxiolysis without having to resort to painful injection or intravenous starts. Doses of 1.7 mcg/kg of fentanyl have been demonstrated to be equivalent to 0.1 mg/kg of morphine. One study investigated the efficacy of intranasal midazolam, ketamine, or a combination of both in young, uncooperative dental patients (aged 2-6 years). Doses of intranasal medications used were midazolam 0.3 mg/kg, ketamine 6 mg/kg, and a mixture of both midazolam and ketamine at 0.2 mg/kg and 4 mg/kg, respectively. The overall success rate
Clinical Pathway For Management Of Pediatric Patients With Suspected Cerebroventricular Shunt Malfunction (Headache, Vomiting, Altered Behavior)

Does the patient have a fever?

NO

< 6 months since placement or revision?

NO

Consider tapping the shunt or consulting a neurosurgeon to tap the shunt to rule out infected shunt
• Send culture and routine CSF studies
• Administer antibiotics for pleocytosis or positive Gram stain (Class II)

YES

YES

Abdominal pain? Distension? Vomiting?

Consider shunt flow studies (Class III)

NEGATIVE STUDY

GOOD FLOW

POOR/ABSENT FLOW

Consider tapping the shunt or consulting a neurosurgeon to tap the shunt to evaluate shunt function and CSF flow (Class II)

NEGATIVE STUDY

POSITIVE STUDY

Tap the shunt or have the consulting neurosurgeon tap the shunt to evaluate shunt function and CSF flow (Class II)

Call neurosurgery for definitive management
• Make the patient NPO
• Manage increased intracranial pressure

Send culture and routine CSF studies
• Administer antibiotics for pleocytosis or positive Gram stain (Class II)

Consider abdominal ultrasound or CT scan to evaluate for abdominal pseudocyst (Class II)

Consider discharge home if well, or admit for observation if still symptomatic

Abdominal pain? Distension? Vomiting?

NO

Consider shunt flow studies (Class III)

NEGATIVE STUDY

POSITIVE STUDY

Consider abdominal ultrasound or CT scan to evaluate for abdominal pseudocyst (Class II)

YES

NO

< 6 months since placement or revision?

YES

This clinical pathway is intended to supplement, rather than substitute for, professional judgment and may be changed depending upon a patient’s individual needs. Failure to comply with this pathway does not represent a breach of the standard of care.

Class Of Evidence Definitions

Each action in the clinical pathways section of Pediatric Emergency Medicine Practice receives a score based on the following definitions.

Class I
• Always acceptable, safe
• Definitely useful
• Proven in both efficacy and effectiveness

Level of Evidence:
• One or more large prospective studies are present (with rare exceptions)
• High-quality meta-analyses
• Study results consistently positive and compelling

Class II
• Safe, acceptable
• Probably useful

Level of Evidence:
• Generally higher levels of evidence
• Nonrandomized or retrospective studies: historic, cohort, or case control studies
• Less robust randomized controlled trials
• Results consistently positive

Class III
• May be acceptable
• Possibly useful
• Considered optional or alternative treatments

Level of Evidence:
• Generally lower or intermediate levels of evidence
• Case series, animal studies, consensus panels
• Occasionally positive results

Indeterminate
• Continuing area of research
• No recommendations until further research

Level of Evidence:
• Evidence not available
• Higher studies in progress
• Results inconsistent, contradictory
• Results not compelling

Abbreviations: CSF, cerebrospinal fluid; CT, computed tomography; MRI, magnetic resonance imagining; NPO, nothing by mouth.
Clinical Pathway For Management Of Pediatric Patients With Tracheostomy Malfunction (Hypoxia, Respiratory Distress, Cyanosis)\(^a\)

- Suction the tracheostomy lumen
- Provide supplemental oxygen
- Reposition the patient
- Establish the size and brand of the tracheostomy tube (Class I)

**YES**

- Able to oxygenate and ventilate?

**NO**

- Increased tracheal secretions or change in sputum, cough, or fever?

**NO**

- Send tracheal aspirate to the laboratory for culture
- Consider chest x-ray
- Obtain respiratory viral panel (rule out tracheitis, pneumonia) (Class II)

**YES**

- Replace tube in correct position
- Have the patient under observation in the ED prior to discharge

**DISPLACED**

**NOT DISPLACED**

- Address other causes of hypoxia
- Utilize alternate method of oxygenation/ventilation, including bag-valve mask ventilation, otorrheal intubation, or endotracheal tube into tracheal stoma
- Call for back-up airway assistance (ear, nose, and throat or anesthesia specialist)

**Able to oxygenate and ventilate?**

**YES**

- Change tracheostomy tube and attempt to oxygenate and ventilate (Class I)
- Consider specialist involvement

**NO**

- Admit if patient is in respiratory distress or if the patient requires frequent suctioning, are ill, or are hypoxic with concomitant pneumonia requiring a significant increase in home oxygen requirements.

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\(^a\)Mild cases of tracheitis can be treated with enteral antibiotics as long as close follow-up is ensured and the patient does not appear ill.

Abbreviation: ED, emergency department.

For Class of Evidence definitions, see page 16.
Clinical Pathway For Management Of Pediatric Patients With Developmental Disorders Who Require A Procedure

Involve a child life specialist, if available, to assist in patient cooperation and understanding of the procedure

Will distraction techniques work?

- Distract the patient and complete the procedure

Is the procedure painful?

- Use topical/local anesthetic, oral/intramuscular morphine, intranasal fentanyl, or oral/intramuscular ketamine, based on clinical scenario (Class II)
- Add oral/intramuscular midazolam for anxiolysis as desired (Class III)

What level of analgesia does the patient need?

- Minimal
  - Use topical/local anesthetic, oral/intramuscular/morphine, intranasal/intravenous fentanyl, oral/intramuscular/intravenous ketamine, or nitrous oxide (Class II)
  - Add oral/intramuscular midazolam for anxiolysis as desired (Class III)

- Moderate
  - Obtain intravenous access after using oral or intramuscular agents, or use nitrous oxide, if needed
  - Use sedative agent of choice based on clinical scenario, plus opioid (Class II)
  - Sedate with ketamine alone or in combination with opioid. (Class II)

- High
  - Restrain, distract, or sedate as appropriate (Indeterminate)
  - Sedate with ketamine alone or in combination with opioid. (Class II)

Length of imaging or other diagnostic test or procedure?

- SHORT DURATION
  - Use topical/local anesthetic, oral/intramuscular/intravenous morphine, intranasal/intravenous fentanyl, oral/intramuscular/intravenous ketamine, or nitrous oxide (Class II)
  - If needed, sedate with agent of choice plus an opioid, nitrous oxide, or intravenous ketamine alone (Class III)

- LONG DURATION
  - Administer propofol, etomidate, thiopental, or chloral hydrate based on clinical scenario (Class II)

Abbreviation: MRI, magnetic resonance imaging.
For Class of Evidence definitions, see page 16.
was 89% with ketamine alone, 84% with the combination of drugs, and 69% with midazolam alone. Intranasal midazolam has been successfully used to abort seizures in epileptic children.

### Special Circumstances

Fractures, patterned or excessive bruising, and vomiting in the child with special needs should raise the possibility of nonaccidental trauma. Children with special needs are sometimes nonverbal and can be challenging and frustrating to caregivers. Children with special needs are 2 to 3.6 times more likely than nondisabled peers to be physically abused. Children who were born prematurely are also at higher risk for abuse. Specifically at risk are children with behavior and mental health problems. In a 2008 study, young (aged < 6 years) developmentally delayed patients were abused at rates similar to developmentally normal children without special needs, and children with behavior and mental health issues were almost twice as likely to be abused than control peers.

Care should be taken to inspect the entire child with clothing removed to examine for bruises, patterned injuries, or other signs of nonaccidental trauma. Munchausen by proxy, also called pediatric condition falsification, may also be a factor in children with special needs. Disturbingly, children and adolescents with developmental disabilities are at 2 to 3 times greater risk for sexual abuse than their developmentally typical peers. Urine or serum pregnancy tests should be performed on all females of child-bearing age when they present with vomiting, abdominal pain, abdominal mass/distension, genitourinary complaint, or secondary amenorrhea.

### Controversies And Cutting Edge

#### Ventricular Shunt Flow

A noninvasive thermal technique (Shunt Check™) to evaluate for flow in ventricular shunts by application of an ice cube for 1 minute at the shunt reservoir site to detect changes in temperature, seems promising, as it accurately preoperatively detected flow (or the absence of flow) in patients in the operating room for shunt revision (80% sensitive, 100% specific). Unfortunately, half of the patients seen in routine office visits did not have detectable flow, although none required shunt revision on clinical symptoms. Furthermore, among a group of patients admitted to the hospital for possible shunt obstruction, ShuntCheck™ was unable to accurately predict which patients progressed to requiring revision due to ongoing symptoms. Several subjects with flow documented by Shunt Check™ had to undergo surgery for revision, while several with no documented flow remained clinically stable and did not go to the operating room even within 7 days.

Further studies with ShuntCheck™ may be warranted to establish its role in the diagnosis of shunt malfunction.

### Disposition

#### Gastrostomy And Gastrojejunal Tubes

Patients with dislodged G tubes that have been replaced with Foley catheters, G tubes, or buttons can be discharged home to follow up with the service that placed the tube (the gastrointestinal specialist or surgeon). Dislodged GJ tubes in patients requiring continuous feeds who cannot have replacement of the tube by an interventional radiologist should be admitted to the hospital for intravenous hydration and careful monitoring of electrolytes until the GJ tube is replaced under fluoroscopy. A Foley catheter or feeding tube may be placed to keep the stoma patent, but it should not be used until the definitive tube is placed. Most patients with G-tube cellulitis may be discharged home on oral antibiotics; however, if oral antibiotics have failed and they have worsening cellulitis or appear ill, they should be admitted for intravenous antibiotics.

#### Tracheostomy Tubes

Patients with dislodged tracheostomy tubes that are successfully replaced can be discharged home after a period of observation. Patients with tracheitis should be admitted if they require frequent suctioning, are ill, or are hypoxic with concomitant pneumonia requiring a significant increase in home oxygen requirements. Mild cases of tracheitis can be treated with enteral antibiotics as long as close follow-up is ensured and the patient does not appear ill. Patients with mild bleeding from a tracheostomy may be discharged home after consultation with otolaryngological services. Hemodynamically unstable or persistently bleeding tracheostomy sites need to be investigated either at the bedside or in the operating room with bronchoscopy to locate and contain the source of bleeding.

### Ventricular Shunts

Patients with shunt obstructions must be admitted for revision or transferred to a center with specialists capable of performing the revision. Infected shunts require intravenous antibiotics and, in some cases, shunt externalization as determined by the neurosurgical service. Migrated shunt tubing and disconnected or broken shunts must be managed operatively by neurosurgery. Patients with abdominal pseudocysts should also be admitted for surgical intervention and antibiotics. Patients with a suspected shunt malfunction may be discharged if all studies and clinical appearance are normal, after discussion with the subspecialist to ensure close follow-up.
1. “The G tube is leaking around the exit site. I’ll just upsize it to close the hole.”
Upsizing a leaking G tube runs the risk of expanding the size of the stoma, which may lead to further expansion of the hole. Instead, pull the tube toward you until the tube is flush against the anterior abdominal wall. Make sure the balloon is inflated and functioning. If these maneuvers fail, consult with the service (usually surgery) that placed the tube.

2. “The blood from the tracheostomy was flowing pretty heavily when he got here, but now it’s stopped, his hemoglobin is normal, and the dad told me he was suctioning him every hour before it started. I’m going to send him home. The ear, nose, and throat specialist doesn’t need to be called in.”
Tracheoinnominate fistula is a life-threatening condition that can’t be missed. Often there will be a sentinel bleed prior to a clinically important larger bleed. An ear, nose, and throat specialist should evaluate the tracheostomy with bronchoscopy if the amount of blood is impressive, even if it has since stopped. Tracheoinnominate fistula, bleeding granulomatous tissue, and tracheitis are all possible diagnoses, or excessive bleeding could be caused by aggressive suctioning.

3. “The patient’s mom told me that she put the dislodged G tube back in, and she’s been feeding him through it, so I don’t need to check its placement.”
At the very least, pull back stomach secretions from the tube, and check the pH to ensure that the tube is, in fact, in the stomach. Make sure that the tube is amenable to a parent’s replacing it, such as a G tube with balloon, and not a GJ tube or one without a balloon.

4. “He just started vomiting, so he’ll probably develop diarrhea soon. It’s probably just a virus.”
Whether this patient has a shunt or feeding tube or a history of abdominal surgery, maintain a lower threshold to work up the child with special needs who is vomiting. This could represent shunt malfunction, tube obstruction, intestinal obstruction, medication side effects, nonaccidental head trauma, or decompensation of a metabolic problem.

5. “The head CT scan shows small ventricles, so the symptoms must not be related to the VP shunt.”
A subset of patients with VP shunts has no change in ventricle size on CT, despite having a malfunctioning shunt. If the patient demonstrates symptoms of shunt obstruction, consult with neurosurgery to evaluate the patient, regardless of results of brain CT.

6. “He has tachycardia and fever, but he is an older child with special needs, so we don’t have to worry about his having a serious bacterial infection.”
Sepsis is more common in children with special needs than in the general pediatric population. Although sepsis is not a common diagnosis among children with special needs, abnormal vital signs should not be ignored in this population.

7. “This child with autism just needs a couple stitches in his lip. I don’t want to sedate him, because it’s too risky, so I’ll just hold him down.”
Children with autism respond to procedural sedation as well as children who do not have autism using similar amounts of sedation medication. Follow usual protocols, avoid polypharmacy, and be ready with backup airway maneuvers.

8. “The CT is normal, and the VP shunt flow was normal when the neurosurgeon tapped it, so it must not be the shunt causing the headaches, lethargy, and vomiting.”
In a small prospective study, 64% of the children had symptoms of shunt malfunction with a normal CT and normal shunt tap. Do not ignore signs of elevated ICP.

9. “I can’t find a source of fever in this patient who has a VP shunt, but we don’t need to tap the shunt, since he had his last revision 1 month ago.”
Ninety percent of VP shunt infections will occur within the first 6 months after placement or revision. Children who have fever within the first month are at highest risk for VP shunt infection, so the shunt should be tapped to investigate for infection if no other obvious source is present.

10. “I don’t want the family to have to watch this fragile child with special needs go through this code. This family has been through enough.”
Children with special needs are no different from other children, and parental presence should be offered in these situations. Importantly, you should determine early in the resuscitation efforts what the end-of-life care preferences are, including Allow Natural Death, DNR, or variations of these plans.
Children With Developmental And Behavioral Disabilities

Be aware that children with developmental disorders may have smaller airways than developmentally normal children and may have an increased incidence of hypoxia during sedation. Keep their comorbidities in mind when deciding whether it is safe to sedate them in the ED for procedures. When patients are deemed unsafe to sedate in the ED, admitting them for procedures to be performed in the operating room under general anesthesia is an acceptable option.

Summary

Children with special needs are a growing and diverse population presenting issues requiring a special skill set among emergency clinicians. Parents are a reliable source of knowledge of their child’s baseline behavior. If the parent feels that something is wrong with the child, this should be pursued rather than dismissed. Understanding the mechanics and complications of the various instruments that a child with special needs may have will improve the emergency clinician’s ability to provide quality care for this patient subset. Procedures can be safely and effectively performed on behaviorally complex patients, but studies specifically looking at management of these children in the emergency setting are lacking.

Case Conclusions

After examining the 7-year-old girl, you determined that she had tracheitis as well as pneumonia. After providing mild sedation, you had the girl undergo a CT scan of the head, which was normal. The blood from the tracheostomy site was from suctioning and tracheitis, and the ear, nose, and throat specialist performed a bedside bronchoscope to confirm this. The evaluation for sepsis was normal. The patient improved after admission, and she was given antibiotics based on prior cultures, pulmonary toilet, and oxygen. You determined that the G tube site blood was from local irritation. You inflated the balloon and secured it against the abdominal wall, relieving leakage of stomach contents. With application of liquid sucralfate, the skin improved during admission.

The 3-year-old boy with Down syndrome required replacement of his G tube in the ED, and this was completed by an emergency medicine resident under your supervision. She confirmed placement of the new tube by suctioning back gastric contents and demonstrating a pH of 2 on litmus paper. No other confirmatory testing was done, and the patient was discharged home in good condition.

The 8-year-old patient was sedated with intravenous pentobarbital, and his CT scan was normal. You repaired his laceration using a combination of topical local anesthetic medication, child life support, and intranasal fentanyl for pain control and anxiolysis. He was discharged home when he returned to his baseline.

References

Evidence-based medicine requires a critical appraisal of the literature based upon study methodology and number of subjects. Not all references are equally robust. The findings of a large, prospective, randomized, and blinded trial should carry more weight than a case report.

To help the reader judge the strength of each reference, pertinent information about the study, such as the type of study and the number of patients in the study will be included in bold type following the references cited in this paper, as determined by the author, will be noted by an asterisk (*) next to the number of the reference.

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6. A clogged GJ tube can be unclogged by using:
   a. Tissue plasminogen activator
   b. Pancreatic enzymes
   c. Corrosive alkali
   d. Valsalva maneuver

7. Which medication is recommended for treatment of irritation of the skin at the exit site of a gastrostomy tube that is leaking gastric contents?
   a. Oral ranitidine
   b. Topical bacitracin ointment
   c. Topical liquid nystatin
   d. Topical liquid sucralfate

8. Compared to developmentally normal counterparts, developmentally delayed children have a higher incidence of which of the following risk factors for developing hypoxia while under sedation?
   a. Lower tidal volume
   b. Higher minute ventilation
   c. Shorter neck
   d. Smaller airway diameter

9. Which strategy may help reduce anxiety in the autistic child needing to undergo a procedure in the ED?
   a. Minimize the number of staff interacting with the child.
   b. Have the child away from his usual caregiver.
   c. Remove all familiar items brought from the child’s home.
   d. Papoose the child whenever possible.

10. Compared to developmentally normal counterparts, dosage requirements of sedation medications in autistic children requiring ED procedures are:
    a. Twice as much
    b. Half as much
    c. 5 times as much
    d. The same
An Updated Approach To The Use Of Video Laryngoscopy And Intubation In Pediatric Patients

Endotracheal intubation of pediatric patients is an infrequently needed, yet high-risk, procedure in emergency medicine. Emergency clinicians should be aware of available approaches to assist with successful intubation in pediatric patients. Video laryngoscopy involves the utilization of optical and video technology to facilitate indirect visualization of laryngeal structures during intubation. This technology can be advantageous when intubating patients with both normal and difficult airways. A number of pediatric devices are now available, each with unique benefits as well as limitations and nuances in technique that guide their use. This evidence-based review provides a summary of the existing data on video laryngoscopy use in routine as well as difficult airways and provides practical instruction on the use of 3 specific devices that have been approved for use in pediatric patients.

Risk Management Pitfalls Addressed In This Review

1. “The patient has bronchiolitis and copious oral secretions. Is that a contraindication to using video laryngoscopy?”
   Secretions, vomitus, or blood can cover the camera lens and compromise visualization. Although this is not an absolute contraindication to the use of video laryngoscopy, preparations such as suction, gauze to wipe the camera, and back-up blades may be helpful. In addition, availability of a device that allows for direct visualization (either a C-MAC® or a traditional laryngoscope) may help obviate the risk of reliance on an indirect (camera) view.

2. “Video laryngoscopes are too large to be used in neonates. I would only use direct laryngoscopy in this age group.”
   Although there are no large comparative trials regarding the use of video laryngoscopy in neonates, case reports and series have shown success in this age group. Becoming comfortable with the size of the blades and the technique for use in this population requires experience. However, many devices include the C-MAC®, GlideScope®, and Airtraq all have blade sizes designed for use in children of all ages, including neonates. For emergency clinicians with training, these devices can offer the same advantages as with older pediatric patients and adult. Emergency clinicians who are comfortable with airway management in this age group and with video laryngoscopy may safely use video laryngoscopes in neonates.

Myocarditis And Pericarditis In Pediatric Patients: Recognition And Management

Myocarditis and pericarditis are inflammatory conditions of the heart commonly caused by viral and autoimmune etiologies, although many cases are idiopathic. Emergency clinicians need to maintain a high index of suspicion for these conditions given the rarity and often nonspecific presentation in the pediatric population. Children with myocarditis may present with a variety of symptoms ranging from mild flu-like symptoms to overt heart failure and shock, whereas children with pericarditis typically present with chest pain and fever. The cornerstone of therapy for myocarditis includes aggressive supportive management of heart failure, as well as inotropes and antidysrhythmic medications as indicated. The acute management of pericarditis includes recognition of tamponade and, if identified, the use of pericardiocentesis. Medical therapies may include nonsteroidal anti-inflammatory drugs and colchicine, with steroids reserved for specific populations. This review focuses on the evaluation and treatment of children with myocarditis and pericarditis, with an emphasis on currently available medical evidence.

Risk Management Pitfalls Addressed In This Review

1. “The patient’s troponin level is negative, so he can’t have myocarditis.”
   Troponin levels may have sufficient sensitivity to rule out myocarditis, but the test performance depends on the cut-off level defining a positive test. Current evidence suggests that troponin I and T tests lack adequate specificity in cases of pediatric myocarditis. While a negative troponin is reassuring, emergency clinicians should interpret this result in the context of the cut-off value used at his or her facility.

2. “I gave 60 mL/kg of intravenous fluid to a child with myocarditis, and now she is getting worse.”
   Children with myocarditis often present in shock, which prompts aggressive intravenous fluid administration. Failure to respond to an initial fluid bolus should raise concern for a cardiogenic cause such as myocarditis. In cardiogenic shock, poor cardiac contractility leads to the development of pulmonary edema. Clinically, patients will develop labored breathing as well as crackles and rales on examination. Treatment of such patients by emergency clinicians should include intravenous diuretics, such as furosemide.
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