The term congenital tracheal stenosis (CTS) describes a wide range of tracheal abnormalities. CTS may be the result of an abnormality inherent to the trachea itself or be the effect of external forces compressing the airway, such as cardiovascular malformations. However, the common characteristic in all cases of CTS is an innate narrowing of the trachea producing airway obstruction.

CTS is relatively rare. In the majority of cases, it consists of a funnel-shaped part of the trachea characterized by complete circular cartilaginous tracheal rings. CTS may involve only a short segment of the trachea, or it may affect more than 50% of the trachea, including the main bronchi. The second type of stenosis is referred to as long-segment tracheal stenosis (LSTS) and is the most severe and challenging form of CTS. Except for some cases of mild CTS, all forms of CTS are usually life threatening.

In 1964, Cantrell and Guild classified the 3 morphological types of CTS as generalized hypoplasia, funnel-shaped stenosis, and segmental stenosis. However, CTS has no standard definition. Most reports in the literature describe, by means of diagrams, several variants of CTS. Hoffer et al proposed that CTS be categorized into 3 classes. Class 1, short-segment stenosis, is associated with 8% mortality and often responds to conservative therapy. Class 2 is extensive stenosis and associated anomalies, excluding marked heart or lung disease; the mortality rate in this class is 45%. Class 3 includes any stenosis with marked heart or lung disease and has the highest mortality rate of 79%. Eliot et al proposed that the evaluation and subsequent definition of CTS be uniformly based on 4 elements: the narrowing of the trachea, the extent of tracheal involvement, the involvement of the bronchi, and the presence or absence of complete tracheal rings. Complete tracheal rings (Figure 1), in which the membranous part of the trachea is absent and the cartilage is circumferential, are common in CTS.

CTS is often associated with other congenital malformations of the pulmonary, cardiovascular, and gastrointestinal systems (Table 1). Vascular rings are among the most common associated anomaly in children with CTS. An associated pul-

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Pulmonary artery sling (Figure 2) is a common feature that occurs in 30% to 50% of patients with CTS. This association was designated the ringsling complex by Berdon. Until recently, the outlook for infants and children with CTS was unpromising because medical management was the only way to treat these patients. Most patients died of acute airway obstruction. In the past 20 years, the development and advancement of diagnostic, surgical, and intensive care medical and nursing skills have dramatically improved the outcomes of these patients. Recent reported survival rates have been as high as 78% to 92%. Currently, the challenge is to decrease the high morbidty rate associated with postoperative and therapeutic procedures rather than to increase survival.

**Embryology**

Although the anatomical features of CTS include a variety of deformities, the common denominator in all instances is congenital narrowing of the trachea. Hoffer et al describe the embryological processes of the respiratory system, the timing of developmental abnormalities leading to CTS, and associated anomalies. The trachea develops from the respiratory primordium. In the third to fourth week of life, the hepatic primordium migrates from the respiratory primordium, allowing the respiratory primordium to dilate and bifurcate ventrocaudally into lung buds. The lung buds give rise to the trachea, infraglottis, and the glottic opening. By week 8, the mesenchymal rudiments of the tracheal cartilages are present. During the next 2 weeks, the cartilages form fibroelastic tissue, and smooth muscle is incorporated into the trachea. Researchers have hypothesized that CTS is a result of abnormalities in development that occur about week 4. Hoffer et al further hypothesized that 2 possible embryological periods exist in which abnormal development may produce CTS. Abnormalities in the fourth gestational week would affect the developing respiratory and hepatic primordia and cause the more severe forms of the disease, which are associated with other anomalies, including heart and skeletal malformations. Between weeks 8 and 10, abnormalities in development would be confined to the developing cartilages and their supporting tissue. Abnormalities at this stage probably produce less severe stenosis, with fewer associated anomalies.

Backer and Mavroudis described the intricacies of the embryological processes involved in the formation of tracheal anomalies associated with vascular malformations. In embryonic vascular development, 6 pairs of aortic arches connect the 2 primitive ventral and dorsal aortas (Figure 3). The pathophysiological changes depend on the preservation or deletion of specific segments of the rudimentary aortic arch complex. These vascular malformations often lead to tracheal or tracheoesophageal compression. The most common vascular anomalies involved are double aortic arch, right aortic arch, pulmonary artery sling, innominate artery compression syndrome, and aberrant right subclavian artery.

**Incidence or Occurrence**

The overall incidence of CTS in the general population is not clear.

<table>
<thead>
<tr>
<th>Common anomalies associated with congenital tracheal stenosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary artery sling</td>
</tr>
<tr>
<td>Atrioventricular septal defects or other congenital heart defects</td>
</tr>
<tr>
<td>Dextrocardia</td>
</tr>
<tr>
<td>Laryngomalacia/bronchomalacia</td>
</tr>
<tr>
<td>Bronchus suis (ie, tracheal bronchus)</td>
</tr>
<tr>
<td>Subglottic stenosis</td>
</tr>
<tr>
<td>Aplastic lung</td>
</tr>
<tr>
<td>Hydrocephalus</td>
</tr>
<tr>
<td>Trisomy 21</td>
</tr>
<tr>
<td>Imperforate anus</td>
</tr>
<tr>
<td>Single kidney</td>
</tr>
<tr>
<td>Tracheoesophageal fistula</td>
</tr>
<tr>
<td>Gastroesophageal reflux</td>
</tr>
<tr>
<td>Diaphragmatic hernia</td>
</tr>
<tr>
<td>No gall bladder</td>
</tr>
<tr>
<td>Cleft lip and palate</td>
</tr>
</tbody>
</table>

Table 1: Common anomalies associated with congenital tracheal stenosis.

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**Figure 2** Pulmonary artery sling. The trachea is compressed by the LPA.

Abbreviations: LPA, left pulmonary artery; MPA, main pulmonary artery; RPA, right pulmonary artery.
The literature on this aspect consists largely of reports of the long-term experiences with CTS at single medical centers. A small number of authors have contributed relatively few patient histories to the literature, mostly in the past 20 years. In 1994, Hoffer et al described fewer than 70 reported cases. In 1981, Benjamin et al reviewed 70,000 hospital admissions and found 21 cases of CTS. Hoffer et al later described 3 patients with CTS in 3 years, and Koopman et al reported cases in 3 patients in 2 years.

Reported survival rates in the preceding 20 years have ranged from 25% to 77%. Historically, LSTS has been associated with the highest mortality rates. However, with the current surgical options, survival rates in patients treated with different procedures have been as high as 78% to 92%; although, again, in most series, the number of patients was small. Today, mortality is often associated with other abnormalities, mainly cardiac lesions or chronic complications.

Clinical Manifestations

Depending on its severity, CTS may be manifested simply as stridor or as a catastrophic near-death episode requiring cardiopulmonary resuscitation. The worst scenario would be a child in extremis who could not breathe at all and thus would require immediate extracorporeal membrane oxygenation.

Most infants with CTS have signs of the abnormality within the first year of life, most often within the first several weeks to months. Signs may include biphasic stridor, tachypnea, retractions, nasal flaring, apnea, cyanosis, wheezing, noisy breathing, recurrent upper respiratory “cold symptoms,” persistent cough, and pneumonia. Dysphagia has also been reported. Dysphagia may be accompanied by apnea or cyanotic spells when an infant or a child with CTS attempts to swallow solid food. Failure to thrive may be a result of increased work of breathing, leading to increased energy expenditure and poor feeding. Patients may hyperextend their heads, as if to “stent” the trachea open and improve breathing.

Most often, the initial clinical manifestation is a life-threatening crisis precipitated by an acute respiratory illness. In most cases, patients have been taken to their local hospital in extremis after collapse at home. In our experience, and as similarly reported in the literature, the infants have respiratory failure and intubation is difficult. A common statement by the referring physician is that the child “keeps trying to die.”

Other causes of respiratory distress and apnea in infants and children must be ruled out, including asthma, foreign body aspiration, gastroesophageal reflux, and bacterial or viral illness. Additionally, a complete neurological workup is often needed to rule out brain tumor, head trauma, or other neurological cause that can result in apnea and subsequent respiratory arrest.

Diagnostic Evaluation

The diagnostic evaluation of a child with suspected tracheal stenosis should begin with plain chest radiographs. These may show unusual air distribution, deviation of the heart and mediastinum, evidence of tracheal compression, and altered tracheobronchial angles, often the result of abnormal vasculature (ie, pulmonary artery sling) impinging on the normal course of the main bronchi. Several other diagnostic imaging studies may be used, including barium esophagography, tracheobronchography, echocardiography, bronchoscopy, and cardiac catheterization. Computed tomography (CT) and magnetic resonance imaging of the chest are useful to evaluate vascular anatomy. Magnetic resonance imaging may be the best procedure for showing the vascular structures of the mediastinum without added exposure to radiation. CT has also been used to image vascular anomalies of the aortic arch and great arteries. Spiral sequencing or 3-dimensional reformattting of the CT scans and virtual bronchoscopy are

Figure 3 Embryology of the aortic vasculature. Three possible variations of the rudimentary aortic arch complex are shown.
used to reconstruct the trachea and proximal bronchi (Figure 4). However, these diagnostic procedures cannot be used to evaluate the dynamic nature of any associated bronchomalacia, in which collapse of the airway may occur only with inspiration. Additionally, other subtle anatomical findings may be missed with CT, including complete tracheal rings. Bronchoscopy coupled with bronchography is a simple procedure, is less expensive than CT, and has excellent spatial and temporal resolution.

**Initial Management**

The initial management of an infant or child with CTS must be to stabilize the airway and provide adequate ventilation and oxygenation. Pediatric Advanced Life Support guidelines should be followed for cardiorespiratory resuscitation and stabilization. Intubation may be difficult and may require a smaller endotracheal tube than recommended in both age-based (Pediatric Advanced Life Support) and length-based (Broselow tape) guidelines. Because CTS is an anatomical abnormality associated with the trachea and does not affect the lung parenchyma, most patients with CTS have no difficulty with oxygenation. Inability to provide ventilation is the most common difficulty encountered after the airway is secured. Ventilation with helium as an inert oxygen carrier can be helpful to reduce drag on gas flow and reduce air trapping.\(^3\)\(^{,12}\) Emergency tracheostomy is almost never necessary, because the stenotic area lies more distally than can be relieved with a tracheostomy. Often, neither intubation nor tracheostomy can provide relief for patients with CTS and respiratory compromise because of the severe nature and/or distal location of the obstruction. For patients with CTS who have an airway that is difficult or impossible to ventilate, extracorporeal membrane oxygenation may be the only lifesaving palliative therapy pending surgical correction.

The diagnosis of CTS and the subsequent treatment and management required inflict immense stress on patients’ families. Most likely, a child with CTS will need to be transferred to a center with the facilities to provide the child’s complex surgical and medical needs.

After complete evaluation, a thorough discussion between all members of the intensive care and surgical teams with the child’s family is imperative. All treatment options must be presented, and stress must be placed on the ambiguity of the long-term outcomes and the limited data available on which to base advice. Important financial and emotional implications must be considered when treatment is started. Members of the pediatric intensive care unit (PICU) nursing staff are paramount in providing emotional support to the families of children with CTS and in facilitating realization of the long-term consequences associated with this abnormality. Nurses should be the liaisons between patients’ families and ancillary services, including social work, clergy, and other support personnel. Nurses should also ensure that communication between patients’ family members and the medical team is ongoing.

**Surgical Intervention**

Classically, surgery is the ultimate form of treatment of CTS. The reported techniques of repair have evolved. Essentially, the length of the trachea dictates the type of reconstruction.\(^12\)

Until 1984, the reported survival rate for children with tracheal stenosis who had medical management was 25%.\(^9\) That same year, Farouk Idriss of Children’s Memorial Hospital in Chicago reported the first successful operation in which cardiopulmonary bypass was used during placement of an autologous pericardial patch in a child with tracheal stenosis due to complete tracheal rings.\(^13\) Since then, several authors have reported different surgical techniques and various results.

In 1994, Andrews et al published an analysis of the different techniques, including tracheal resec-
tion, end-to-end anastomosis, resection of stenosis involving the carina, augmentation of the tracheal lumen with costal cartilage, the use of prosthetic materials, and use of autologous pericardial grafts. Additionally, they described other innovative techniques involving various forms of tracheal resection: slide tracheoplasty for a funnel-shaped stenosis, posterior tracheal division with use of the anterior esophageal wall to create a membranous posterior tracheal wall, and balloon dilatation of a long segment of tracheal stenosis that resulted in a posterior split of the complete tracheal rings. The reported mortality rates for these surgical techniques were 40% to 77%.

In 2001, Backer et al13 published a review of 50 patients with tracheal stenosis who had surgery at Children’s Memorial Hospital of Chicago with 4 different techniques during a period of 18 years. For LSTS, the techniques used were pericardial patch tracheoplasty, tracheal autografts, and slide tracheoplasty. Tracheal resection with end-to-end anastomosis was used for short-segment stenosis and became the preferred method. For LSTS, the autologous pericardial patch was the most frequently used approach and the preferred method; children who had this treatment accounted for the largest group of long-term survivors. All procedures were done through a median sternotomy, and cardiopulmonary bypass was used.

More recent reports4,14,15 have confirmed the use of various surgical techniques based on the length of tracheal involvement. All the authors of these reports4,14,15 agree that concomitant repair of any cardiovascular lesion is imperative and provides the best results with the repair of the trachea.

In 2003, Elliot et al4 presented a review of the most commonly used concepts and surgical techniques described in the literature in the preceding decade to manage patients with CTS. In patients with short-segment stenosis, the preferred method is end-to-end anastomosis or resection and primary anastomosis. Balloon dilatation combined with posterior laser treatment of the trachea is also described; however, Elliott et al indicate that studies in more patients are needed before recommendations for the use of this procedure can be made. Various stents to enlarge the stenotic part of the trachea have also been used, but indications for their use are not clear.

For patients with medium-length stenosis, the slide tracheoplasty has become a preferred method. This procedure essentially doubles the circumference of the trachea and the cross-sectional area is quadrupled.

In patients with LSTS, much controversy remains about the best method to repair the defect. Resection with slide tracheoplasty should be attempted. Acosta et al12 reported improved success with this method compared with resection and anastomosis in patients with LSTS because with slide tracheoplasty, the trachea is shortened less than it is in resection and anastomosis and thus results in less tension on the tracheal repair. Many patients, however, require some type of patch tracheoplasty.4 Several materials have been used to patch the stenotic area of the trachea, including autologous pericardium, rib cartilage, tracheal autografts, tracheal allografts, and most recently, carotid artery. Heterograft tissue and prosthetic material do not work.4

Once again, few data are available on surgical repair of CTS, and most of the reports are reviews of the results with procedures used at a single center during a period of many years. Interpretation of these data is difficult.

**Medical Management**

Postoperative and ongoing medical management of patients with tracheal stenosis is poorly described in the literature. Although several authors4,12 have mentioned “constant communication with the intensive care team,” little information is available on the optimal strategies necessary to ensure positive outcomes. The rarity of CTS and the complex challenges presented by patients with CTS argue strongly for a multidisciplinary, dedicated-team approach.

Pre operative and postoperative medical tactics are varied and are tailored to the specific conditions of each patient. First and foremost is ensuring adequate ventilation and oxygenation. This goal may be secondary only to establishment of a patent airway. In certain circumstances, extracorporeal membrane oxygenation may be used to provide cardiopulmonary support before and after surgical intervention. Cardiovascular resuscitation is unlikely during the initial manifestations of CTS unless severe acidosis occurs. In our experience, a systems approach is the best approach for medical management (Table 2).

**Central Nervous System**

Sedation and concurrent use of neuromuscular blockade are used
for each child when airway management or oxygenation and/or ventilation is difficult. Often, continuous infusions of narcotics and benzodiazepines are required. Appropriate sedation is maintained according to the Penn State Children’s Hospital Sedation Algorithm. This practice allows each patient’s nurse to determine the frequency of administration of sedatives and paralytic agents given as needed. Each patient’s level of sedation is based on the amount of ventilatory participation the patient is allowed or can tolerate. For example, a higher level of sedation is required during acute decompensation. A methadone-weaning period is often required to prevent narcotic withdrawal. In addition to pharmacological means of sedation, other methods are used to achieve the ordered level of sedation. These include music therapy, presence of the patient’s family when tolerated, television, and therapeutic touch by the nursing staff.

Respiratory System

Various methods are used to monitor the respiratory system, including cardiopulmonary waveforms, capnography, measurement of oxygen saturation, pulmonary function graphics, and measurement of central venous pressures. Each patient’s nurse is responsible for maintaining all monitoring adjuncts. The ventilatory goals for each patient are made by the attending physician on the basis of the patient’s disease process, and the bedside nurse ensures that the goals are met. For example, patients with pulmonary hypertension may require an alkaloic pH, and ventilatory strategies are adjusted accordingly.

Ventilation strategies are often a trial-and-error process. Time-cycled, volume-limited and time-cycled, pressure-limited modes are used, along with a combination of the 2 modes, otherwise known as pressure-regulated volume control. Pressure-regulated volume control is a control mode of ventilation with a decelerating inspiratory flow pattern. Pressure is constant during the entire preset inspiratory phase. The ventilator constantly analyzes the patient’s pressure-volume relationship to facilitate the delivery of the preset tidal volume at the lowest possible pressure. When changes in compliance occur, the delivery pressure is adjusted in small (3 cm H₂O) increments until the preset tidal volume is again achieved, so long as the maximum pressure setting has not been violated. If the maximum pressure setting is reached, the breath will still be delivered, but at a lower tidal volume. This mode is often used for patients who need controlled tidal volumes during pressure-controlled ventilation. The levels of positive end-expiratory pressure that we use vary widely. Most commonly, the level is of 4 to 15 cm H₂O; the higher ranges are used to help “stent” the airway open. Oscillatory ventilation may also be used.

All 3 patients treated at Penn State Children’s Hospital (see box) required tracheostomy tubes (because of severe malacia) and mechanical ventilation at home. Multiple types of tracheostomy tubes were used in the management of these patients. Standard, rigid tubes were used initially after tracheostomy to promote a good track and good healing of the stoma. Flexible tubes in standard sizes were also used but were not effective in providing support to the patients.

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Table 2  Summary of medical management of patients with congenital tracheal stenosis: a systems-based approach

<table>
<thead>
<tr>
<th>System or category</th>
<th>Intervention/management strategy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Central nervous system</td>
<td>Use of a sedation scale to optimize ventilation and oxygenation</td>
</tr>
<tr>
<td></td>
<td>Provision of nonpharmacological comfort measures by pediatric intensive care unit nurses</td>
</tr>
<tr>
<td>Respiratory system</td>
<td>Ventilation and oxygenation goals set by healthcare team and individualized on the basis of patient’s physiological needs</td>
</tr>
<tr>
<td></td>
<td>Vigilant monitoring of patient’s airway</td>
</tr>
<tr>
<td></td>
<td>Familiarization with multiple airway adjuncts and of ventilator modes and types</td>
</tr>
<tr>
<td></td>
<td>Teaching of patient’s family started early</td>
</tr>
<tr>
<td>Cardiovascular system</td>
<td>Hemodynamic monitoring and support started early in hospital course</td>
</tr>
<tr>
<td></td>
<td>Additional therapies used according to presence of associated cardiac anomalies</td>
</tr>
<tr>
<td>Fluids, electrolytes, and nutrition</td>
<td>Early enteral feedings used whenever possible</td>
</tr>
<tr>
<td></td>
<td>Use of long-term nutritional support adjuncts, including placement of a gastrostomy tube, as needed</td>
</tr>
<tr>
<td>Hematology</td>
<td>Blood products used when optimization of oxygenation needed</td>
</tr>
<tr>
<td></td>
<td>Coagulopathies corrected when applicable</td>
</tr>
<tr>
<td>Infectious disease</td>
<td>Antibiotics administered only when clinically indicated</td>
</tr>
<tr>
<td>Development</td>
<td>Therapies started early to address physical, occupational, and speech/swallowing needs</td>
</tr>
</tbody>
</table>
Pediatric Care

Case Series at Penn State Children's Hospital

**Patient 1**
Patient 1 was born full term with multiple congenital anomalies, including cardiac defects, right pulmonary agenesis, and long-segment tracheal stenosis. Tracheal reconstruction with a bovine pericardial patch was performed early in the neonatal course. A tracheostomy was performed, and the patient ultimately required long-term mechanical ventilation at home. Additionally, a Nissen fundoplication and a gastrostomy tube were required to ensure adequate nutrition. At 2 years of age, the patient was weaned from the ventilator, and the tracheostomy tube was removed. Unfortunately after many months of uncontrolled pulmonary hypertension, the patient died at 27 months of age.

**Patient 2**
Patient 2 was a full-term baby girl who had undiagnosed respiratory problems from birth thought to be the result of laryngomalacia. Before the patient’s first birthday, an upper respiratory infection caused intractable respiratory distress and subsequent cardiopulmonary arrest. Extracorporeal membrane oxygenation was a necessary bridge to surgical correction. Subsequently, a pulmonary artery sling and associated complete tracheal rings extending down both main bronchi were diagnosed. Tracheal reconstruction with a bovine pericardial patch and division of the vascular ring was performed. Tracheostomy, Nissen fundoplication, and gastrostomy were ultimately performed. Management of the patient’s airway continued to be difficult. After several episodes of cardiopulmonary arrest, the patient had hypoxic brain injury. Long-term mechanical ventilation at home was required. The patient was referred for a tracheal allograft transplant and stenting of her airway at an out-of-state hospital. After 4 years of tenuous medical and surgical care, the tracheostomy tube was removed.

**Patient 3**
Patient 3 was an otherwise healthy full-term baby boy who had severe respiratory distress when he was 9 weeks old. Computed tomography of the chest revealed a true vascular ring: an aberrant right subclavian artery and long-segment tracheal stenosis with complete tracheal rings. A bovine pericardial patch repair with removal of the aberrant subclavian artery was performed. After several unsuccessful extubations, a tracheostomy was performed. The patient ultimately required long-term mechanical ventilation at home. Throughout his second year of life, he required frequent hospitalizations for management of granulation tissue. Unfortunately, he had a cardiac arrest at home and could not be resuscitated. The cause of death remains unknown.

Cardiovascular System

Historically, infants and children with tracheal stenosis initially have respiratory distress without cardiovascular compromise. Most of the intensive care is associated with maintaining a patent airway and extended past the area of malacia, giving support to the floppy part of the airway. An adjustable-length tracheostomy tube was used in 2 of the 3 patients. The length could be manipulated via a wraparound locking system, allowing the barrel of the tube to be adjusted to achieve the desired depth in the airway. The other tube that was used was also adjustable in length but was bifurcated at the distal end to provide support for a part of both main bronchi (Figure 6). This particular tube was put in place via bronchoscopy to guide the bifurcated ends into the left and right main bronchi.

Levels of mechanical ventilation at home vary for each patient. The Chronic Ventilation Service is consulted early and assists with the management of patients with CTS even while the patients are in the PICU. Once a patient’s condition is stable, he or she is transferred to the rehabilitation unit, where intensive education of the patient’s family begins. Members of the Chronic Ventilation Service staff closely monitor patients’ respiratory mechanics, ventilation, oxygenation, growth, and activity tolerance. Members of the nursing staff provide extensive in-hospital education to each patient’s family before the family can take the patient home. This training begins early, before the patient leaves the PICU. Education includes suctioning, airway management, use of oxygen, tracheostomy tube changes, and ventilator mechanics, including troubleshooting. This education along with comprehensive social support continues through discharge and into the home until the tracheostomy tube is removed.
adequate gas exchange. However, uncompensated respiratory acidosis can quickly progress to cardiovascular collapse, and full cardiorespiratory support may be required. Central venous access is almost always mandated, and placement of an arterial catheter may also be warranted. A pulmonary artery catheter and echocardiographic evaluation are justified if pulmonary hypertension or congenital heart disease is suspected. Nurses must be able to anticipate and detect subtle changes in the child’s cardiopulmonary status. For example, tachycardia, pulsus paradoxus, and changes in oxygen saturations are often detected before a patient decompensates. Proficiency in hemodynamic monitoring is paramount for PICU nursing staff.

**Fluids, Electrolytes, and Nutrition**

Early enteral feeding is used whenever possible. For each patient, the bedside nurse in the PICU places a nasojejunal feeding tube within the first 48 hours postoperatively to ensure adequate energy intake. In our PICU, a nasojejunal placement team inserts the feeding tubes. Patients with CTS often require more permanent feeding adjuncts, including placement of a gastrostomy tube with or without Nissen fundoplication, in which the fundus of the stomach (top part of the stomach) is wrapped around the back of the esophagus until the fundus is once again in front. The part of the fundus that is now on the right side of the esophagus is sutured to the part on the left side to keep the wrap in place. This procedure has the effect of creating a 1-way valve in the esophagus that allows food to pass into the stomach but prevents stomach acid from flowing into the esophagus and thus prevents reflux of food or medications.

Formulas high in protein are used in older infants and children; fortified breast milk or infant formulas are used for younger infants. We have also advocated supplementation with zinc and vitamins C and E to support the immune system. Normalization of electrolyte levels is accomplished; many patients require sodium and potassium supplementation because of the use of diuretics. We often use gentle diuresis during the postoperative period or when ventilation is difficult. Continued use of diuretics may continue into the chronic phases of home care to optimize respiratory mechanics. Consultation with a pediatric nutritionist occurs early to provide optimal management.

**Hematology**

Blood and blood products have been used to promote oxygen delivery or correct coagulopathy during the acute period of care. We minimize phlebotomy once each patient is beyond the acute postoperative phase.

**Infectious Disease**

Colonization of the airway with bacteria is common in children and adults with artificial airways. Because of the increasing problem of resistance to antimicrobial agents, we limit the use of antibiotics. Treatment is always based on the results of cultures and tests of susceptibilities of the cultured organisms and the patient’s clinical status. The nursing and medical staff follow strict isolation for any patient with antibiotic-resistant bacteria. Universal precautions are used for all PICU patients. Members of the nursing staff are responsible for educating
patients’ families about the importance of infection control in the hospital and in the home.

**Development**

Prevention of contractures and pressure ulcers is paramount if a patient is maintained at a high level of sedation. Members of the nursing staff regularly institute range-of-motion exercises and use of skin-supporting devices in patients with decreased mobility. Our nurses use the Braden Q Scale on every patient admitted to the PICU to determine risk factors for alteration in skin integrity. Early introduction of physical, occupational, and speech therapy is a necessity. Assessing each patient’s readiness to swallow is essential for successful oral progression and speech development. These therapies are started in the hospital and are continued in the home setting if required.

**Chronic Issues: Morbidity and Mortality**

Table 3 lists the most common complications associated with surgical correction and subsequent management in patients with CTS. Granulation tissue readily develops at the surgical site or where the patch interfaces with the native trachea and carina. Granulation tissue is also a problem where the end of the tracheostomy or endotracheal tube is sitting. The development of granulation tissue resulting in airway obstruction is often the reason for an emergent trip to the operating room. Vascular endothelial growth factor is an experimental protein that is being studied in animal models to decrease the formation of granulation tissue in recipients of tracheal autografts.

Malacia, both of the patch and of the native airway, has been a difficult complication to treat on a long-term basis. This complication may require creative ventilation strategies with positive end-expiratory pressure or continuous positive airway pressure and customized tracheostomy tubes to stent the floppy area open. Various stents have been used preoperatively and postoperatively to overcome airway obstruction. These stents are also associated with risks, including development of granulation tissue and infection. Infectious complications are rare, but mediastinitis can occur and must be taken seriously.

Despite the advances in surgical repair, the high quality of medical care, and the ability to provide long-term mechanical ventilation in the home setting, morbidity and mortality remain high in CTS. Backer et al published the outcomes of 28 patients who underwent pericardial patch tracheoplasty from 1982 to 2000. They reported 2 early deaths and 5 late deaths; in addition, 6 patients required tracheotomy.

Reoperations were required in several patients, including management of air leak, patch revision, rib cartilage grafts, and placement of a balloon expandable Palmaz stent. In the review by Elliott et al of 163 patients from 1987 to 2002, mortality rates varied from 0% to 47%, depending on the surgical technique.

**Discussion**

Caring for an infant or child with CTS requires a multidisciplinary approach to ensure the best possible outcome. Each case should be assessed individually to determine which surgical technique will be most successful for the specific anomaly. Ongoing monitoring of the airway is essential along with a ability to return rapidly to the operating room if there is any hint of airway problems. Aggressive and innovative airway and ventilation management by the PICU team in conjunction with a surgical specialist is needed. Patients’ successful transition home requires a dedicated program of mechanical ventilation at home and a pediatric rehabilitation facility to ensure the caregivers’ proficiency in providing the treatment. Once home, each patient should be comanaged by his or her primary pediatrician to address general care issues. Bedside nurses can help patients’ families during this stressful time by coordinating care among the various teams and by providing psychosocial support. Nurses can also assist families in the transition to long-term home nursing care. Dedicated teaching by nurses is required to ensure a safe and successful transition home for these medically fragile children.
Acknowledgments
We thank the staff of the pediatric intensive care unit for their never-ending dedication and diligent care. We are especially grateful to Dr. Michael Dettorre for his compassionate care to the patients of the Chronic Ventilation Service and his assistance with the preparation of this manuscript.

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