

Tracheal and bronchial stenoses and other obstructive conditions

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Abstract: Although tracheal stenosis and bronchial stenosis are relatively rare in the pediatric population, they are both associated with significant morbidity and mortality. While most cases of congenital tracheal stenosis in children present as complete tracheal rings (CTRs), other congenital tracheal obstructions are also encountered in clinical practice. In addition, acquired obstructive tracheal conditions stemming from endotracheal trauma or previous surgical interventions may occur. Many affected children also have associated cardiovascular malformations, further complicating their management. Optimal management of children with tracheal or bronchial stenoses requires comprehensive diagnostic evaluation and optimization prior to surgery. Slide tracheoplasty has been the operative intervention of choice in the treatment of the majority of these children.

Keywords: Tracheal stenosis; bronchial stenosis; complete tracheal rings (CTRs); slide tracheoplasty

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Overview

Unlike subglottic stenosis, tracheal and bronchial stenoses are relatively rare in children and are generally congenital. Most pediatric tracheal stenosis manifests as complete tracheal rings (CTRs), with the absence of a membranous trachea extending for varying lengths of the airway. Other congenital malformations as well as acquired lesions are also encountered (1). Children with congenital tracheal or bronchial stenosis are frequently complex and can present with concurrent anomalies, including pulmonary artery sling, ventricular septal defects, lung hypoplasia, lung agenesis, and gastrointestinal defects (2,3).

Our article will briefly describe tracheal and bronchial stenoses and other obstructive conditions, presenting an overview of their diagnostic workup. We will subsequently present a succinct description of techniques used for surgical repair and the optimization of patients to ensure optimal surgical outcomes.

Diagnostic workup

Varying degrees of respiratory distress can be observed in patients with tracheal or bronchial stenosis. This may be accompanied with retractions and distal airway sound. Stridor, a “barking” or brassy cough, and “washing machine” airway sounds may be present. Dependent on the level of involvement, stridor may be inspiratory, expiratory or biphasic. Cyanosis and dying spells trigger urgent action (4).

Patients with severe tracheal stenosis may need urgent treatment because of life-threatening airway obstruction. On the other hand, asymptomatic patients with mild stenosis may not require surgical repair.

Diagnosis is frequently delayed because of the rarity of the lesion or because other more apparent associated malformations draw the attention of the clinician. For this reason, experience and a high level of clinical suspicion are essential in establishing an accurate diagnosis (5).

Bronchoscopy remains the most important tool to the

diagnosis of tracheal and bronchial stenosis. Both flexible and rigid instrumentation can be used to determine the type of lesion, localization, extension, and severity (5).

Imaging can provide excellent definition of an abnormal tracheobronchial tree and its mediastinal counterparts. A CT scan provides good anatomic delineation of the airway and magnetic resonance imaging (MRI) enables full assessment of the vascular structures and their relationship to the adjacent trachea. Nevertheless, definitive assessment requires endoscopic evaluation.

Congenital tracheal stenoses

CTRs

Although CTRs are rare, they are the most common cause of congenital tracheal stenosis. In children with this anomaly, the cartilaginous rings are circular and smaller than the unaffected trachea. This anomaly is characterized by the absence of a membranous trachea and the trachealis muscle.

More than 75% of patients have other associated anomalies, which can be severe; these include pulmonary artery sling, cardiac defects, and lung hypoplasia or agenesis. A large series conducted at Cincinnati Children's Hospital Medical Center [2011] revealed that 60% of patients with CTRs had cardiovascular abnormalities, most commonly a pulmonary artery sling (1).

Four specific patterns of CTRs may be recognized. These patterns include (I) complete rings that are relatively patent proximally but come down to a very small distal ring near the carina; (II) the "stovepipe" airway with a long segment of CTRs of similar diameter; (III) a short-segment stenosis, often in the mid-trachea; and (IV) CTRs associated with a high pig bronchus (6,7).

Infants typically present with increasing respiratory distress, stridor, retraction, apnea, cyanosis, and occasionally, dying spells. Some children present with a "wet" sounding airway due to secretions adhering to the area of stenosis, sometimes described as "washing machine respiration". Symptoms are exacerbated when the infant is agitated or when feeding (4).

In the first few weeks of life, the infant may grow at a far greater rate than the stenotic airway. This occurrence may result in critical obstruction. Reports of a very difficult intubation with the need to "screw in" the endotracheal tube are unfortunately quite common. This can further exacerbate airway edema and may cause a compromised

airway to become a critical airway (4).

A small percentage of children with CTRs do not present until later in life. Moderate obstruction can accommodate growth without causing respiratory distress (8). As the child grows, so do the CTRs, though at a non-linear rate. Symptoms in these children develop when the growth of the child significantly exceeds the growth of the airway. Presentation typically manifests with exercise intolerance and is insidious in nature. Important to note, a child may have comparatively few symptoms despite a significantly narrowed airway.

Assessment is made with rigid bronchoscopy, which should be performed with extreme care. The smallest possible telescopes should be used, as rough instrumentation in the area of stenosis may cause enough swelling to convert a narrow airway to a critical airway, necessitating abrupt emergent intervention. An estimate of the size of the airway is valuable. Because 50% of the children with CTRs have a tracheal inner diameter of approximately 2.0 mm at the time of diagnosis, the smallest endotracheal tube (2.0 mm) and the smallest tracheotomy tube (2.5 mm) cannot pass through the stenotic segment without severe damage to mucosa or tracheal rupture. Intubation proximal to the complete rings is sometimes advisable. If intubation is still not able to maintain adequate ventilation, typically seen with severe distal tracheal involvement, or when there are coexistent cardiovascular anomalies requiring repair, extracorporeal membrane oxygenation (ECMO) may be necessary to stabilize the child.

In view of the high proportion of patients with other congenital anomalies, a computed tomography (CT) scan and an echocardiogram should be performed. CT scans should be performed with contrast and 3D reconstructions to best appreciate the anatomy of both the airway and surrounding vasculature.

Not all pediatric patients with CTRs require tracheal reconstruction. In some asymptomatic or mildly symptomatic patients, a period of observation helps in determining whether a tracheoplasty will ultimately be required (5,8,9). A small number of patients show evidence of significant airway growth and do not require surgery. In other cases, the growth of the child will be faster than the growth of the CTRs, with the child becoming more symptomatic over time. In this clinical scenario, intervention will ultimately be required. In general, the younger the age at initial presentation, the more likely the need for tracheoplasty (8).

When surgical repair is necessary, slide tracheoplasty is

currently the operation of choice (1).

Tracheal cartilaginous sleeve

In this condition, the trachea is not composed of 15 to 20 separate rings, but of a single sheet of cartilage that may extend proximally into the cricoid and progress distally into the bronchi. This is universally associated with craniosynostosis (Pfeiffer, Crouzon, or Apert syndromes) (10). While in most cases the sleeve trachea is not stenotic, in rare cases the posterior cartilaginous trachea may overlap, requiring operative reconstruction. In this setting, slide tracheoplasty is an effective reconstructive option; however, it is technically more challenging to perform (4).

Absent tracheal rings

Absent tracheal rings are an extremely rare intrinsic tracheal defect manifesting as tracheal stenosis or collapse. This condition typically presents in an otherwise normal child with a short-segment of the trachea (2 to 3 rings) missing cartilage. This segment is usually just proximal to the carina. Presentation is similar to CTRs; however, bronchoscopically, the stenotic segment lacks cartilage and is therefore distensible (11). This anomaly is generally managed with either a tracheal resection or a slide tracheoplasty (1,11).

Chondrodysplasia punctata

Chondrodysplasia punctata comprises a group of rare congenital disorders characterized by punctate calcification of cartilage referred to as “stippling”; this results in bone and skin lesions as well as ophthalmologic and cardiac malformations (12).

Laryngeal and tracheal calcifications in affected children are rarely reported. There is no universal management approach for these exceedingly rare anomalies. Balloon dilation of the trachea can be attempted. In refractory cases, tracheostomy should be considered (12).

Other congenital tracheal obstructions

Tracheal agenesis

Tracheal agenesis is a rare embryologic anomaly that almost always results in death. Patients who are prenatally diagnosed and in whom the atresia involves only the

proximal trachea occasionally survive through the use of an *ex-vivo* intrapartum treatment (EXIT) procedure, low tracheotomy, and eventual tracheal reconstruction. In another form of tracheal agenesis, there is either a very short remnant of distal trachea, or the entire trachea is absent and the bronchi come directly off the esophagus. Neonates present at birth with severe respiratory distress. Temporary ventilation is often possible with esophageal intubation, and ventilation through a tracheoesophageal fistula; however, this is typically unsustainable. If there is no communication between the airway and the esophagus, congenital high airway obstructive syndrome (CHAOS) will result (13).

Type 4 laryngotracheoesophageal cleft

The triad of husky cry, aspiration pneumonia, and feeding problems in newborns with congenital anomalies should prompt an early workup for complete laryngotracheoesophageal cleft. Early surgical intervention is necessary to prevent aspiration and offers the only hope for survival (14).

Tracheomalacia

It is critical not to confuse malacia with stenosis, as the management of this disease is fundamentally different from the management of tracheal stenosis.

Overall, tracheomalacia is the most common congenital abnormality of the trachea. Diffuse tracheomalacia can be caused by intrinsic problems of the trachea, as in the mucopolysaccharidoses (Hunter and Hurler syndromes), but more commonly is localized. Children treated with prolonged mechanical ventilation for respiratory distress syndrome are particularly at risk (15,16).

In patients ultimately requiring tracheotomy, four levels of focal tracheal injury may induce cartilage damage and collapse: the suprastomal tracheal ring, the stoma, the overinflated cuff site, and the site of a malposition tip.

Most patients with tracheomalacia are asymptomatic. When symptomatic, however, children typically present with medium-pitched expiratory stridor. A “brassy” or “honking” cough may also be present. Despite the cough, the patient may have difficulty expectorating secretions. The most serious symptoms of tracheomalacia include dying spells and cyanosis.

In addition to the symptoms and signs attributed to tracheomalacia, the patient is often symptomatic from

a variety of associated anomalies. Commonly associated abnormalities include laryngeal clefts, tracheoesophageal fistulae, and bronchomalacia (15). Fifty percent to 75% of patients with tracheomalacia have concurrent gastroesophageal reflux disease (GERD) (17,18).

As with other tracheal anomalies, the gold standard in the diagnosis of tracheomalacia is endoscopy with the patient spontaneously breathing. This can be accomplished during flexible bronchoscopy or microlaryngoscopy and bronchoscopy using a rigid telescope. However, large-caliber rigid instruments may artificially stent open the tracheal airway, not appreciating the degree of anterior-posterior collapse. Therefore, many endoscopists prefer the use of flexible bronchoscopy to diagnose malacic conditions. If the plan of anesthesia is too deep and the patient is not spontaneously breathing, assistive ventilation will obscure disease as well (4).

In all circumstances, intervention should be based on the symptoms of the child rather than the appearance of the airway. Nonoperative management includes pneumatic stenting of the airway with continuous positive airway pressure (CPAP), positioning, and treatment of GERD, which has been shown to exacerbate laryngotracheal disease (19).

Operative management obtains the best results for symptomatic localized lesions. Clinical circumstances that mandate operative intervention include persistent symptoms that fail nonsurgical therapies, acute life-threatening events, recurrent pneumonia, and other manifestations of intermittent respiratory obstruction. Currently, aortopexy is preferred for mid-to-distal tracheomalacia (20-22). Stents can be applied endoscopically to maintain tracheal patency across focally collapsed segments. Although they do not require open neck incisions, their propensity to migrate, to cause mucosal erosions, and to form granulation tissue makes them less attractive as a permanent solution (23). Alternatively, stents may be most effectively used as a temporizing measure while the collapsed segment gains intrinsic strength or while a more extensive repair is being planned.

Tracheotomy with or without positive pressure ventilation is frequently performed for intractable tracheomalacia that does not meet criteria for, or has failed, lesion-specific surgical therapy. The other common indication for tracheotomy is tracheobronchomalacia, as this is rarely amenable to surgical intervention. Severe tracheobronchomalacia may require long-term ventilation through the tracheotomy tube.

Vascular compression

Although vascular compression is not uncommon, the majority of affected children are either asymptomatic or mildly symptomatic. When symptomatic, children present with biphasic stridor, retractions, a honking cough, and dying spells.

Forms of congenital vascular compression affecting the trachea include innominate artery compression, double aortic arch, and pulmonary artery sling (4).

The diagnosis of airway compression is best established with rigid bronchoscopy. Thoracic imaging assists in determining the relevant vascular anatomy.

CPAP frequently offers a degree of temporary improvement, as segmental tracheomalacia may be present in the region of the vascular compression. Intubation or tracheostomy may be required to stabilize the airway prior to definitive treatment, but both should be utilized with caution because of the risk of forming an arterial fistula from erosion in the area of compression.

The surgical management of symptomatic vascular compression must be individually tailored to address specific pathology. Although alleviating vascular compression improves the airway, it takes time for the cartilage in the affected segment of the airway to completely normalize. Until the airway normalizes, children may require a tracheostomy (4).

Acquired tracheal stenoses

Acquired tracheal stenosis is generally traumatic in origin, either the result of direct injury (e.g., clothesline injury) or more commonly the result of trauma from the cuff or tip of an endotracheal tube. It is therefore usually a problem affecting the cervical trachea and amenable to a cervical approach if reconstruction is required (4).

Tracheal stenosis may also occur at the site of a tracheotomy, with either an A-frame deformity or suprastomal collapse due to cartilage disruption, erosion, and deformity at this site.

Patients with acquired tracheal stenosis have a tendency to present insidiously and have moderate complaints of exercise intolerance or sleep-disordered breathing. Their endoscopic exam often reveals a much more severe stenosis than their symptoms portend. These patients may present months to years following prolonged intubation, laryngotracheoplasty, or direct injury to the airway (4).

Congenital bronchial stenosis and other rare bronchial obstructions

Congenital bronchial stenosis

Isolated congenital bronchial stenosis is an unusual lesion, caused by compressive vascular anomaly, cardiac anomaly, or a congenital pulmonary cyst. The stenosis is usually seen in the left main bronchus, which is compressed between the left pulmonary artery and the descendent aorta. Symptoms and treatment vary depending on both the severity and anatomic location of the lesion. The diagnostic workup, including bronchoscopy, CT scan, and echocardiography, is similar to that used for suspected tracheal stenosis (24).

Although extremely rare, complete bronchial rings may also occur. They are usually an extension of CTRs. Butler *et al.* reported that 23.7% of their patients with CTRs have extension of the stenosis into one or both bronchi (2).

Surgical management of bronchial stenosis includes resection and reconstruction of the bronchus and slide bronchoplasty, and this can be extremely challenging in babies because of the small size of the infant airway (7,24). Postoperative complications are common after bronchial surgery. Atelectasis is the most frequent complication, followed by stenosis, anastomotic leakage, malacia, pneumonia, and empyema (24).

Other rare bronchial obstructions

Congenital bronchial lobar agenesis is characterized by either complete termination of the bronchus or significantly stenosed segmental or lobar bronchus. Bronchial obstruction results in enlargement of the peripheral bronchus, mucus accumulation, and emphysematous changes in the peripheral lung. Prenatal diagnosis remains difficult, but unlike tracheal agenesis, this disease is compatible with life (25). Most patients can be managed nonoperatively.

Absent bronchial rings (Williams-Campbell syndrome) are a rare disorder characterized by deficiency of subsegmental bronchial cartilage and the development of airway collapse and bronchiectasis that may subsequently progress to respiratory failure and death (26). Some published reports suggest a familial association (27). The bronchoscopic findings are characteristic, and reconstruction requires transection of the bronchial insertion at the carina, splitting the distal posterior wall of the bronchus and the proximal anterior wall of the trachea

and sliding the bronchus further up the trachea (4).

Acquired bronchial stenosis

Because of selective intubation of the right main bronchus, acquired stenosis in neonates is likely more frequent than congenital stenosis of the major bronchi; nevertheless, it is rare. Most cases can be managed with endoscopic balloon dilation or laser resection.

Acquired bronchial obstruction may also occur following vascular intrathoracic procedures, mainly because of a cardiovascular stent compressing the bronchus.

Preoperative optimization

Optimal management of children with tracheal stenosis requires comprehensive evaluation prior to repair. The temptation is to proceed straight to definitive repair should the child deteriorate. However, if the airway permits intubation with a 2.0 endotracheal tube, this should be performed via nasotracheal route in order to temporarily stabilize the child. When this approach is not possible, an endotracheal tube sized to accommodate the cricoid cartilage, but placed shallow, and proximal to the complete rings, can still permit positive pressure ventilation. It is rare that the first two tracheal rings are affected in children with CTR, and therefore most children can be intubated proximal to the complete rings. If ventilation remains difficult, ECMO is advisable but should not be taken lightly. Clearly, tracheotomy is rarely helpful as the smallest CTRs tend to be more distal, and the smallest available commercial tracheotomy tube is 3.6 mm in outer diameter. More importantly, tracheotomy may further compromise the options of subsequent operative repair (4).

All patients should undergo methicillin-resistant *Staphylococcus aureus* (MRSA) screening and treatment before the surgery. MRSA infection in open airway procedures can be a devastating complication, resulting in dehiscence, graft loss, and weakening of the cartilaginous structure of the laryngotracheal complex (28).

Although GERD plays an important role in the pathogenesis and prognosis of laryngeal stenosis, its impact on children with tracheal or bronchial stenosis is insignificant.

Surgical management

Surgical correction may not always be necessary in infants with tracheal or bronchial stenosis. These patients

may, however, present later in life with dyspnea; in this clinical scenario, surgical correction becomes essential. Concomitant cardiac or great vessel anomalies that require repair, most often a pulmonary artery sling, require a combined procedure through an anterior sternotomy with the patient under cardiopulmonary bypass to repair the trachea. A cervical approach to the trachea, without sternotomy or bypass, may be performed in patients with mid- to high tracheal lesions.

Endoscopic techniques

Although balloon dilation and laser resection can be performed in some cases of acquired tracheal stenosis and in some children with postoperative complications (29), its role in the treatment of congenital tracheal stenosis is controversial. Most authors agree that endoscopic techniques should not be employed in patients with CTRs (5).

Open surgical procedures

Tracheal resection

Tracheal resection with primary reanastomosis is required much less frequently in children than in adults. Historically, this procedure in children was considered to be less successful than in adults because of the smaller airway diameter in children and less tolerance of anastomotic tension (7). This technique is a useful tool for addressing short-segment CTRs (involving less than one-third of the trachea) and acquired stenosis of the trachea, and careful surgical technique and selection of patients have been shown to improve outcomes (30). However, it is not uncommon to discover a longer segment of CTRs following dissection of the trachea during an open procedure. Therefore, in our experience, optimal results have been achieved with a short segment slide tracheoplasty (4).

When performing a tracheal resection, one must avoid anastomosis close to the carina because any anastomotic problems may ultimately require placement of a tracheotomy tube; the tip of the tracheotomy tube must lie between the anastomosis and the carina and this may be problematic if there are only 2 to 3 mm between the anastomosis and the carina (4).

Augmentation techniques

Costal cartilage graft tracheoplasty

This procedure is reserved for upper tracheal stenosis,

since the strap muscles are not present in the lower trachea. Overlying muscle provides the vascular supply to cartilage grafts, and an intrathoracic graft may require a pedicled muscle flap to remain viable. Because cartilage grafts treating intrathoracic tracheal stenosis have an increased risk of granulation tissue formation at the site of the graft and the risk of prolapse of the graft into the airway (4), we no longer perform this procedure.

Anterior pericardial patch

For many years, pericardial patch tracheoplasty was the standard procedure for the management of CTRs. It is effective in children with mild to moderate stenosis due to CTRs. However, children with more severe forms of tracheal stenosis tend to have a difficult postoperative course following this procedure; this is usually due to granulation tissue in an airway that remains small despite the tracheoplasty or anterior malacia of the patch (4). This operation has fallen out of favor given the established success of the slide tracheoplasty for most cases of CTRs.

Tracheal autograft patch

This technique involves the resection of the middle third of a segment of CTRs, anastomosis of the upper and lower ring segments, and splitting of the remaining rings anteriorly. The resected segment of rings is then laid open and used as an anterior graft through the remaining rings (31). It is superior to the pericardial patch; however, it is associated with a significant incidence of restenosis.

Slide tracheoplasty

Slide tracheoplasty was originally described by Tsang *et al.* (32) and popularized by both Grillo *et al.* (7) and by our team at Cincinnati Children's Hospital (6). This operation overlaps stenotic segments of the trachea, shortening it but doubling the diameter of the stenotic area. Slide tracheoplasty is currently the operation of choice for tracheal stenosis attributed to CTRs. This technique has a high rate of success and minimal morbidity (1).

This technique has a number of advantages relative to other methods. These advantages include immediate tracheal reconstruction with rigid, vascularized tissue with a normal mucosa; ability to extubate patients early in many cases; less postoperative granulation tissue formation; and growth potential of the reconstructed trachea (7,33). In addition, it is a versatile technique: one can perform a short-segment slide, an oblique slide, or even an inverse slide if circumstances dictate. The slide can also extend into the membranous

trachea or into the carina if required. The whole length of the trachea may be slid, even past the carina.

Briefly, patients are initially placed on cardiopulmonary bypass, with cannulation of the ascending aorta and the right atrial appendage. If a pulmonary artery sling is present, this is repaired prior to the slide tracheoplasty. The trachea is exposed by dissecting between the ascending aorta and the superior vena cava. In the process, removal of the right paratracheal lymph nodes facilitates tracheal exposure. The carina is identified deep to the right pulmonary artery and the anterior trachea is exposed from the carina to the upper aspect of the CTRs (1,4).

Intraoperative bronchoscopy is then performed to define the upper and lower limits of the complete tracheal ring segment. A 30-gauge needle is placed through the anterior tracheal wall as it is visualized by the bronchoscope to define the proximal and distal CTRs. At this point, with the patient stabilized on cardiopulmonary bypass, a more comprehensive evaluation of the distal airway can also be performed (1,4).

The length of the stenosis is then measured and the trachea is transected at the midpoint of the segment of complete rings. Each end of the transected trachea is then mobilized. The lateral vascular attachments to the trachea are preserved in this process. The anterior wall of the proximal tracheal segment is incised vertically. The posterior wall of the distal segment is cut vertically toward the carina. Cartilage is then trimmed from the corners of the proximal and distal segments, and the segments then slid over each other. Depending upon the length of the stenotic segment, this requires additional tracheal mobilization from both superior and inferior attachments. The carina is displaced superiorly as stay sutures applied to the distal trachea are attached to the head drape (1,4).

The anastomosis is performed with a running 5.0 or 6.0 double-armed polydioxanone suture (PDS), with a single knot securing the repair proximally. Prior to final closure of the anastomosis, the patient is re-intubated with an age-appropriate endotracheal tube, and the tip of the tube positioned under direct visualization. The anastomosis is then leak tested, and further sealed with fibrin glue. The proximal and distal extent of the anastomosis can be marked with small hemoclips to help identify the extent of the anastomosis on post-operative radiographs. The patient is then removed from bypass, the chest closed, and the patient is transferred to the intensive care unit. Even with near full-length tracheal reconstruction, it is unusual to need a suprahyoid release or chin to chest sutures.

Extension of the slide into a bronchus or cricoid cartilage has been performed successfully at our institution and may assist with repairing these concomitant stenoses. In children with an associated pig bronchus, a modified slide can also be performed, with the rings being split oblique to the midline, so as not to compromise the orifice to the bronchus (1,4).

At Cincinnati Children's, we have followed this surgical approach since 2001. Our experience has demonstrated that the slide tracheoplasty with cardiopulmonary bypass support can be performed with very low mortality despite the complexity of this patient population. Also, the slide technique requires airway reintervention less frequently than other techniques (1).

In our last cohort study (n=80), 29% of our population required airway reintervention within 12 months of the initial procedure. However, this rarely involved more than endoscopic management, such as balloon dilation, endoscopic resection of granulation tissue, or temporary stent placement. Four deaths (5%) were reported (1). This mortality rate was much lower than the previously reported mortality rate of up to 24% in some series (34,35).

A series of 101 children who underwent a slide procedure at Great Ormond Street Hospital was published in 2014. Seventy-two of their patients (71.3%) had associated cardiovascular anomalies. Thirty-three children (33%) had residual stenosis at 3 months and 8 (8%) had residual stenosis at 9 months after the surgery. Stenting was required in 21.8%, mainly in patients with preoperative bronchomalacia. The mortality rate was 11.8%, and bronchomalacia and the need of preoperative ECMO were associated with this outcome (2).

Regarding the anesthetic technique, although current methods, including jet ventilation, may allow for repair of distal and long-segment tracheal stenosis, these can often be obtrusive and cumbersome for the surgeon. Cardiopulmonary bypass is a safe alternative that allows partial deflation of the heart and lungs so that exposure of the complete trachea is optimized. Conversion of ECMO to cardiopulmonary bypass is also recommended for the procedure for this same reason. Successful surgical management thus depends upon close collaboration of the airway surgeon and the cardiovascular surgeon.

Cervical slide tracheoplasty without cardiopulmonary bypass has also been performed by airway surgeons at Cincinnati Children's since 2003. It is considered to be a valuable technique that should be added to the surgical armamentarium for patients requiring open airway

reconstruction. This procedure is an adaptation of the standard slide procedure and can be used for tracheal long-segment stenosis, tracheal “A-frame” deformities, and multilevel laryngotracheal stenosis. In our cohort published in 2012, we described 29 patients who underwent this procedure. Operation-specific success rate was 79% (23 of 29 patients), including all 10 patients with long-segment acquired tracheal stenosis. Lower operative success occurred in patients with concomitant subglottic stenosis, posterior glottic stenosis, and multilevel airway lesions. Four patients (14%) experienced complications: one patient had a minor wound infection; one had a dehiscence that was managed with a revision tracheoplasty; one had an innominate artery injury that was successfully treated intraoperatively without sequelae and one had a symptomatic “figure-8” deformity that required revision therapy (36).

A retrospective series by Matúte *et al.* (n=10) compared the slide procedure with anterior costal cartilage graft tracheoplasty (ACCGT) for the treatment of CTRs, reporting that all six patients treated by ACCGT experienced restenosis while the four patients treated with slide tracheoplasty were asymptomatic and doing well (33).

Complications that occur after slide tracheoplasty most commonly reflect the underlying health status of the child. These infants may be critically ill prior to tracheal repair, and often remain critically ill following tracheal repair. The tracheal repair may also cause problems, including granulation tissue along suture lines and restenosis or collapse at the anastomosis site. Temporary or permanent injury to the recurrently laryngeal nerve is also possible. Failure of extubation usually results from one of these issues. A worrisome complication is dehiscence of the anastomosis. However, this is extremely uncommon. Granulation tissue is usually amenable to serial bronchoscopic management. Stenosis or collapse at the anastomosis site may require either periodic dilation of the trachea or placement of a tracheotomy tube, with the tip of the tracheotomy tube bypassing the area of concern (4). However the primary predictors for poor outcomes were revision surgery, unilateral pulmonary agenesis, and bronchial stenosis.

Postoperative care

Following open tracheal repair, the aim is extubate the child at the conclusion of the procedure, or within 24–48 hours. While the patient is intubated in the pediatric intensive care unit, the child’s head is maintained in forward flexion

on a pillow, and it is advisable to maintain peak ventilation pressures below 30 cm of water pressure so as not to damage the anastomosis. Ideally, chest drains are left in place until after the extubation. In the setting of an extremely unstable preoperative ventilated child, postoperative ECMO may be required. The aim is to establish endotracheal ventilation and remove the child from ECMO as soon as possible (1).

Follow-up endoscopy to examine the repair is routinely performed 1 and 2 weeks after the operation. Gentle balloon dilation is sometimes useful during the recovery phase if the figure-8 tracheal deformity at the repair is significant. This intervention helps prevent left and right lateral suture lines from coming into contact and adhering (1). Children without cardiopulmonary anomalies are typically discharged from the hospital 2 to 3 weeks postoperatively.

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None.

Footnote

Conflicts of Interest: Dr. MJ Rutter has a suprastomal stent named after him, marketed by Boston Medical Products, for which he has declined royalties; he is the patent holder for the Aeris airway balloon marketed by Bryan Medical, and is a consultant for Bryan Medical; he has received no compensation or royalties for this product, although that may change in the future. The other authors have no conflicts of interest to declare.

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