Costal Cartilage Tracheoplasty for Congenital Long-Segment Tracheal Stenosis

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Objectives: To evaluate and report the outcome of costal cartilage tracheoplasty for the treatment of congenital long-segment tracheal stenosis.

Design: Retrospective chart review.

Setting: Academic tertiary care children’s hospital.

Patients: Consecutive series of 10 patients who presented with congenital long-segment tracheal stenosis.

Intervention: All patients underwent costal cartilage tracheoplasty while receiving cardiopulmonary bypass.

Measurements: Age at repair, weight at repair, length of stenosis, minimal diameter of stenosis, postoperative days receiving ventilator support, postoperative days until discharge, postoperative bronchoscopies, postoperative complications, associated anomalies, survival rate, and current status.

Results: There were 8 males and 2 females with an average age at repair of 18 weeks. Average weight was 5.2 kg. Average length of stenosis was 3.2 cm, and average minimal diameter was 1.9 mm. Average postoperative days receiving ventilator support was 17 with a median of 9.5. Average postoperative days until discharge was 35.2 with a median of 17. Average postoperative bronchoscopies was 18 with a median of 4.5. There was a 40% major postoperative complication rate. Seven of the patients had associated anomalies. No patient died from an inadequate tracheal airway, though 2 patients ultimately died from other cardiopulmonary complications for a survival rate of 80%. Average time since surgery for survivors is 8.0 years. Two patients still require treatment. Seven of the original 10 patients are fully active without tracheostomy.

Conclusion: We report one of the largest series of costal cartilage tracheoplasty for congenital long-segment tracheal stenosis and one that has met with a relatively high success rate.


Congenital long-segment tracheal stenosis (CLSTS) is a rare and life-threatening disorder. By convention, the stenosis involves greater than one half of the length of the trachea and usually involves complete cartilaginous tracheal rings. The pathogenesis of CLSTS is poorly understood.

Congenital tracheal stenosis was first described by Gregor in 1899. Cantrell and Guild categorized congenital tracheal stenosis due to complete tracheal rings into 3 types. Type 1 is hypoplasia of the entire trachea with subsequent stenosis from the cricoid to the carina. Type 2 is funnel-shaped stenosis with the narrowest portion of the trachea located distally, near the carina. Type 3 is segmental tracheal stenosis where the stenotic segment gives the trachea an hourglass appearance. Any of the 3 types may result in long-segment stenosis. Type 1 is usually the most severe. There is a high association between CLSTS and various other congenital anomalies. In particular, cardiac malformations along with vascular rings and slings are common.

It is generally believed that the pediatric patient can tolerate up to 50% narrowing of the trachea before becoming symptomatic. Stenosis greater than 50% usually requires intervention. However, prior to the 1980s, management of CLSTS routinely met with dismal results to the extent that some authors recommended conservative treatment. In the last 20 years, numerous changes have occurred that have cumulatively resulted in a dramatic
improvement in the prognosis for patients with these complications. Education has led to heightened clinical awareness so that the entity of CLSTS is better understood and recognized earlier. Technological improvements, particularly with the development of fiberoptic telescope and bronchoscope systems, have allowed for more accurate diagnoses and safer airway management. Better anesthetic techniques and improved cooperation between anesthesiologist and surgeon have evolved. All of the advances have subsequently allowed for the development of more aggressive and successful surgical interventions.

The work of Grillo7 demonstrated that stenoses involving less than half of the trachea could be managed by segmental resection and end-to-end anastomosis. Treatment for longer stenoses has inspired a search for various materials to augment the deficient caliber of the tracheal lumen. The ideal graft would be safe, readily available, easy to shape, biocompatible, rigid, quickly epithelialized, and ultimately able to grow with the patient. No single material has perfectly addressed all of these requirements. To date, a number of augmentation tracheoplasties have been performed using synthetic mesh, periosteum, dura, omentum, anterior esophageal wall, pericardium, and costal cartilage.15-20

For the past 15 years, we have treated CLSTS with costal cartilage tracheoplasty (CCT) at our institution. Twice, we have reported our results in the thoracic surgery literature.21,22 This article adds several patients to our series, provides further follow-up information, and is now presented in the otolaryngology literature.

PARTICIPANTS AND METHODS

DIAGNOSIS

The majority of the patients had been referred to the Division of Pediatric Otolaryngology, St Louis Children’s Hospital, St Louis, Mo, for evaluation of noisy breathing or respiratory distress. Others were initially evaluated by the Division of Pediatric Cardiothoracic Surgery for management of cardiovascular anomalies with secondary consultation by otolaryngology.

A pediatric otolaryngologist made the diagnosis of CLSTS with a rigid bronchoscope and with the patient under general anesthesia. The diagnosis of CLSTS was confirmed when the stenosis involved greater than one half of the length of the trachea. In addition to measuring the length of the stenosis, an attempt was made to determine the narrowest diameter of the stenosis. The minimal diameter value was obtained by sizing the stenosis with a bronchoscope or telescope of known outer diameter. The smallest telescope had a diameter of 1.9 mm and, in several patients, could not be passed through the narrowest aspect of the stenosis. In these cases, the endoscopist made an estimate of the minimal diameter of the stenosis.

SURGERY

Our technique for CCT has been described elsewhere22 and will be reviewed here.

An otolaryngologist working in conjunction with a cardiothoracic surgeon performed all repairs. All patients were administered general anesthesia per endotracheal intubation. Because of compromised respiratory status, some patients arrived in the operating room already receiving mechanical ventilation. The otolaryngology team began by making a transverse incision through the skin overlying the anterior aspect of one rib. Under continued bronchoscopic guidance, the trachea was then incised from superior to inferior so that the entire stenosis was opened (Figure 2). It is important that all complete rings are incised. The length of the stenosis cannot be ascertained by external assessment of the trachea. The vertical defect in the opened trachea was measured with a caliper and the graft fashioned accordingly. If the graft was not long enough, a second piece of cartilage was obtained through the initial harvest site. The graft had been sewn into place using nonabsorbable monofilament sutures with the otolaryngologist closing the left side of the defect and the cardiothoracic surgeon the right side. This arrangement was technically efficient and saved time. The perichondrial surface of the graft faced the lumen (Figure 3). Care was taken so that the graft did not prolapse into the lumen of the trachea and so the sutures also remained extraluminal. None of the sutures were tied until all had been placed. After they had been tied, the position of the graft was verified by another bronchoscopy. The closure was intended to be airtight and any leaks were identified by placing isotonic sodium chloride solution in the wound and applying positive pressure ventilation.
Leaks were closed with further sutures. An appropriate-sized nasotracheal tube was placed so that it did not fit snugly. Because CCT is a rigid reconstruction, the nasotracheal tube did not serve as a primary stent and did not need to pass distal to the repair site. The distal tip of the tube usually resided near the midportion of the graft. Once adequate ventilation was obtained, the patient was removed from cardiopulmonary bypass and both wound sites were closed.

Patients were sedated in the intensive care unit for 7 to 10 days. Specific sedation regimens varied according to the preferences of the managing intensivist. In general, patients received midazolam hydrochloride and fentanyl citrate. An attempt was made to avoid prolonged complete paralysis, but when paralysis was necessary, vecuronium bromide was used. Patients were returned to the operating room for another bronchoscopy to check the status of the graft and to remove any granulation tissue that may have formed. Formal extubation was performed later in the intensive care unit after the patient had fully awakened. Any further diagnostic or therapeutic bronchoscopies were performed on an as-needed basis.

**CHART REVIEW**

A retrospective chart review of all patients admitted to St Louis Children's Hospital from January 1986 to January 2000 with the diagnosis of CLSTS was undertaken. All study subjects underwent CCT as the primary repair of their tracheal disease. This procedure became the preferred technique for CLSTS at St Louis Children’s Hospital in 1986 and has been the primary repair technique used since then. This report documents our entire experience with CCT.

The following patient data were extracted: date of birth; sex; race; length of stenosis; minimal diameter of stenosis; associated anomalies; age at tracheal repair; weight at repair; major postoperative complications; number of postoperative days on ventilator; number of postoperative days until discharge from the hospital; and number of postoperative bronchoscopies. Complications, which were minor and did not affect the patient’s management or course, were not recorded.

**RESULTS**

Ten patients were identified as having undergone CCT for CLSTS. There were 8 males and 2 females. Seven of the patients were white and 3 were black.

Cumulated data are shown in Table 1 and Table 2. Age at repair ranged from 1 to 56 weeks with an average age of approximately 18 weeks. Weight at repair ranged from 3.1 kg to 9.9 kg with an average of 5.2 kg. Length of tracheal stenosis ranged from 2.0 cm to 5.0 cm with an average of 3.2 cm. Minimal diameter of stenosis ranged from 1.0 mm to 2.5 mm with an average of 1.9 mm. Postoperative days receiving ventilator support ranged from 7 to 81 days with an average of 17 days and a median of 9.5 days. Postoperative days until discharge ranged from 11 to 180 days with an average of 35.2 days and a median of 17 days. Postoperative bronchoscopies (both diagnostic and therapeutic) ranged from 1 to 139 with an average of approximately 18 and a median of 4.5.

Forty percent of the patients experienced major postoperative complications. Seventy percent of the patients had other congenital anomalies and 40% were cardiac in nature. Three of the children had syndromes (Down and VATER [vertebral, anal, tracheal, esophageal, radial, and renal anomalies]).

No patient died from an inadequate trachea. Patient 6 died 1.8 years after the tracheoplasty due to a cardiac event. Patient 9 died 16 days after the repair because of left lung air trapping secondary to a stenotic left mainstem bronchus along with a hypoplastic right lung. The intermediate survival rate for the group was 90% and the long-term survival rate is 80%. Seven of the 8 living patients are active and without tracheostomy. The average length of time since surgery for the survivors is 8 years.
Patient 4 did well for 11 years after his tracheoplasty until he developed stridor and dyspnea on exertion. Bronchoscopy showed a 3-cm soft stenosis of the midtrachea. This was initially treated with balloon dilation, but eventually required excision and end-to-end anastomosis of the tracheal ends. He is again fully active.

Patient 5 has experienced a prolonged and very complicated course. His data for postoperative days receiving ventilator support, postoperative days until discharge, and postoperative bronchoscopies skew the group average for these information items. Therefore, median group values are also given and probably more accurately represent the group. Patient 5 developed distal tracheal stenosis after his repair and required placement of a tracheostomy tube 1 month later. Recurring granulation tissue near the carina has required frequent carbon dioxide laser treatments working through the stoma site to maintain a functional airway. For 5 years the patient was physically active until he experienced respiratory arrest at home resulting in anoxic encephalopathy. He now exists in a vegetative state, yet continues to require laser treatments to open his distal airway.

Patient 7 underwent CCT after a tracheostomy had been placed at an outside hospital for CLSTS. One month after the repair, she developed focal stenosis at the old tracheostomy site, which required placement of a second cartilage graft. Her further course was uncomplicated.

Pediatric tracheal stenosis may be classified as acquired, extrinsic congenital, or intrinsic congenital. Acquired lesions are usually secondary to trauma, intubation, tracheostomy, ingestion, burn, or tumor. Extrinsic congenital lesions are usually due to abnormalities of the great vessels or mediastinal or esophageal disease. Intrinsic congenital lesions include tracheal aplasia, primary tracheomalacia, hamartomas, webs, and complete tracheal rings.

As described earlier, congenital tracheal stenosis due to complete rings may be subclassified as generalized hypoplastic, funnel shaped, or segmental. If any of these malformations involve greater than one half of the length of the trachea, it is considered CLSTS. Patients in whom CLSTS causes less than 50% narrowing of the trachea may
not be particularly symptomatic and can usually be managed conservatively. It is thought that the stenosis may “grow” with the child.4 Those in whom CLSTS is responsible for greater than 50% narrowing of the trachea will surely come to medical attention. To further complicate the management of these patients is the fact that the majority will also have associated congenital anomalies, particularly of the cardiovascular system.23 Pulmonary artery sling and pulmonary hypoplasia are common associated anomalies. Our series is unusual in that no vascular slings or rings were encountered. Three of our patients were syndromic.

The timing and manner of presentation of children with CLSTS are variable. The infant may have respiratory distress at birth. Others present with failure to thrive, or initially do well until their growth outstrips their ability to adequately ventilate. Some are incidentally found after a difficult intubation for an unrelated surgery or after superimposition of an upper respiratory tract infection. The majority of patients with CLSTS will present with biphasic stridor and increased work of breathing.

Diagnosis begins with a high degree of suspicion. Plain films and airway fluoroscopy may aid in the diagnosis, but can underestimate the extent of disease. Contrast tracheography and bronchography are dangerous procedures and no longer have a role. Computed tomography and magnetic resonance imaging can be useful in the diagnosis of CLSTS and may further define potential vascular anomalies. However, extreme caution should be used if sedation is necessary to perform these studies. Angiography and echocardiography are warranted to diagnose cardiovascular anomalies.

The gold standard for the diagnosis and evaluation of CLSTS is rigid bronchoscopy under general anesthesia. The endoscopist must make an effort to determine the width, length, and site of the stenosis. Filming the procedure on videotape to review later with a cardiothoracic surgeon is recommended. An attempt should also be made to document the status of vocal cord mobility.

Congenital tracheal stenosis involving less than 50% of the length of the trachea may be treated by segmental resection. Longer stenoses will require a different intervention and various options are discussed below. Also, a variety of intraoperative airway management techniques are available. These include endotracheal intubation, endotracheal jet ventilation, bronchoscopic ventilation, extracorporeal membrane oxygenation, and distal (bronchial) ventilation. We used the cardiopulmonary bypass technique in our series and found this optimal. By removing the need for upper airway ventilation, the surgical field is less encumbered and the surgery can proceed in an unhurried and steady fashion.

The optimal repair technique for CLSTS is controversial. Costal cartilage tracheoplasty was first described by Kimura et al in 1982.15 The advantages of CCT include a graft material that is plentiful and autologous. The cartilage provides a rigid reconstruction, which secondarily allows for a relatively short period of postoperative intubation. Subsequently, the formation of granulation tissue, though always a concern, may be less than that seen with a technique such as pericardial patch. Because of graft rigidity, the endotracheal tube need not be positioned near the carina, a site prone to granulation tissue formation. Another advantage is that the cartilage graft may be fashioned to extend over stenosis of a mainstem bronchus. Experimentally, costal cartilage has been shown to epithelialize well25 and this has been confirmed clinically with postoperative bronchoscopy. Disadvantages of CCT include the existence of 2 wound sites (though the graft may be harvested through the midline sternotomy) and that the initial graft may be too short to encompass the entire stenosis. This latter issue may be addressed by harvesting 2 pieces of cartilage or by resecting a segment of trachea. Long-term growth potential of the cartilage graft is also a concern. Oue et al26 report histological findings of costal cartilage grafts from 6 patients who died 1 month to 2 years after CCT. They found that the graft gradually diminished in size and was ultimately replaced completely by mature scar tissue. Nonetheless, the diameter of the reconstructed trachea was not reduced. Other authors have also noted that the grafts are replaced with scar, though with good functional results.15,19

The fact that patients 1, 2, and 3 are now more than 10 years out from their procedures, without significant airway compromise, would seem to support CCT as a procedure with the potential for long-term success. What truly constitutes “long-term success” is somewhat arbitrary and it will be interesting to follow these patients over the ensuing years. We are unsure as to the etiology of the restenosis seen in patient 4, though are alert to the possibility that it is secondary to complete degeneration of the graft. The poor outcome in patient 5 is related to tracheomalacia and bronchomalacia, which were present in addition to CLSTS and it is doubtful whether any other technique would have been more successful. The death of patient 9 speaks to the worsened prognosis for patients with CLSTS and associated pulmonary anomalies (left mainstem bronchus stenosis, right lung hypoplasia, and hyaline membrane disease). Our survival rate compares favorably with that of other authors.10,14,19

In addition to CCT, the other preferred technique for augmentation tracheoplasty is the pericardial patch. This was first described by Idriss et al10 in 1984. With the patient on cardiopulmonary bypass, a piece of pericardium is harvested through the midline sternotomy. It is then sewn to the anterior tracheal defect and also tacked anteriorly to the innominate artery. The repair is left stented by the endotracheal tube for 1 to 2 weeks. Advantages of this technique include a graft material that is autologous, plentiful, and harvested from the same operative field as the site of tracheoplasty. Disadvantages include the fact that this is not a rigid reconstruction and, therefore, requires longer endotracheal tube stenting. This may result in greater granulation tissue formation necessitating more therapeutic bronchoscopies, particularly when the repair site extends to the carina. Brown et al27 have shown that the pericardium becomes epithelialized and later replaced with normal mucosal and submucosal tissue. Spiral computed tomography scans in their long-term survivors document continued growth of the trachea as the patients aged.
Slide tracheoplasty is another surgical technique for the treatment of CLSTS. It was first described by Tsang et al26 in 1989 and later modified by Grillo.27 Several other authors have documented its success.28-30 With this technique, the trachea is divided transversely through the middle of the stenosis. The tracheal ends are then spatulated by longitudinal incisions on the anterior surface of one end and the posterior surface of the other end. The tracheal ends are then advanced over each other and anastomosed. The resulting repair reduces the length of the stenosis by one half and increases the lumen of the trachea by a factor of 4. The advantages of this technique include avoidance of a graft material and a rigid reconstruction. Postoperative endotracheal stenting time is reduced or unnecessary. Less granulation tissue is formed and fewer bronchoscopies are required. Disadvantages of this technique include its potentially limited applicability for extremely long stenoses and those which involve the carina or mainstem bronchi.31 It may also be difficult to perform on very young patients. In fact, the cumulative experience in young infants has been small.

Adult tracheal reconstruction with a cadaveric tracheal homograft was first described by Herberhold et al31 in 1980. The technique was first described in children in 1996 by Jacobs et al.32 Further reports have attested to its feasibility.33,34 With this technique, fixed cadaveric trachea is sewn to an opened stenotic trachea. A temporary silicone stent is placed in the lumen and endoscopically removed 10 to 12 weeks later. The advantage of this technique is its ability to reconstruct very large tracheal defects including stenoses of the mainstem bronchi. Disadvantages include the availability of cadaveric trachea, granulation tissue formation secondary to the stent, and growth concerns of the graft. The authors recommend this technique only for those patients in whom other reconstruction approaches have failed.

Backer et al35 recently reported a novel technique for the treatment of CLSTS using a free tracheal autograft. With this technique, the anterior trachea was vertically incised through the area of stenosis and the midportion of the stenotic trachea was excised. The posterior ends of the remaining trachea were anastomosed in end-to-end fashion. The excised tracheal segment was then used as a free autograft to repair the anterior tracheal defect. When the graft was not large enough to cover the entire defect, a pericardial patch was added to the repair.

In the future, repair of CLSTS may involve the use of living allogeneic tracheal tissue grown in a laboratory. Macchiarini et al36 have reported growing embryonic human tracheas in the abdominal cavity of nude mice. These tracheal segments were then used to reconstruct surgically induced tracheal stenosis in piglets. The repairs were successful with and without immunosuppression.

Less invasive techniques for the management of CLSTS include the use of balloon dilation37,38 and the placement of expandable metallic airway stents.30 The former technique usually requires multiple applications and may better serve as a bridging maneuver until definitive repair can be performed. The placement of metal airway stents is considered a salvage technique in patients with complications in whom earlier tracheoplasty has failed or who cannot tolerate it.

As the above discussion indicates, there is no consensus on the optimal repair technique for CLSTS. Costal cartilage tracheoplasty, pericardial patch, and slide tracheoplasty seem to be preferred techniques.

CONCLUSIONS

We report our experience with CCT for all patients who presented with CLSTS to St Louis Children's Hospital from 1986 to 2000. All of the repairs were performed with an otolaryngologist working in conjunction with a cardiothoracic surgeon and while the patient was on cardiopulmonary bypass. Most of the patients were small infants, many with associated anomalies or syndromes. No patient died of an inadequate tracheal airway, although one patient is tracheostomy dependent. The survival rate for the group is 80% and 7 of the 10 patients are now fully active without tracheostomy. We believe that CCT should be considered a preferred technique for the management of CLSTS.

Accepted for publication April 8, 2002.

This study was presented at the 16th Annual Meeting of the American Society of Pediatric Otolaryngology, Scottsdale, Ariz, May 11, 2001.

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REFERENCES