Objective: To evaluate the treatment, perioperative management, and outcome of infants who underwent repair of congenital tracheal stenosis. We hypothesized that early resection and tracheoplasty with early weaning of ventilatory support results in less mucosal injury, and thus better outcome.

Design: Retrospective study from 1986 to 1996.

Setting: Tertiary care children’s hospital.

Patients: Seventeen consecutive infants with congenital tracheal stenosis, aged from birth to 16 months. Fifteen patients had complete tracheal rings, 6 of whom also had a left pulmonary artery sling. Fourteen patients underwent either tracheoplasty or resection and reanastomosis of the trachea, both facilitated by cardiopulmonary bypass.

Results: Six patients underwent resection and reanastomosis; 4 patients were extubated within 2 to 5 days without sequelae. There was 1 unrelated perioperative death. Two patients required reintubation. Eight patients required tracheoplasty due to severe tracheal stenosis and had variable postoperative courses. Seven of 14 patients required 0 to 1 postoperative bronchoscopies. Seven of 14 patients required 2 to 7 bronchoscopies for granulation tissue formation, cicatrix, graft collapse, and tracheitis. One patient required numerous procedures and revision tracheoplasty for cicatrix and stenosis.

Conclusions: Correction of short-segment (<5 rings) tracheal stenosis by resection and reanastomosis of the trachea with the aid of cardiopulmonary bypass and early weaning of ventilatory support is recommended. Tracheoplasty using either the castellation technique or slide tracheoplasty is recommended in the treatment of infants with severe (long segment) tracheal stenosis.


**DISTAL TRACHEAL** stenosis is a rare and challenging disorder with historically a poor prognosis. Tracheal stenosis is often associated with anomalies of the respiratory tract, esophagus, cardiovascular system, and skeleton. The association with H-type tracheoesophageal fistula, pulmonary hypoplasia, vascular sling, and, recently, trisomy 21, has been documented.

This malformation is most commonly secondary to complete tracheal rings. In 1964, Cantrell and Guild described 3 types of congenital tracheal stenosis: segmental, funnel-shaped, and generalized hypoplasia. The indications for surgery and the surgical management are controversial, since most institutions have few case reports in their series. Some authors propose tracheotomy with serial dilatation and ventilator support. Severe segmental lesions usually require resection and reanastomosis. Funnel-shaped lesions and generalized tracheal hypoplasia are managed in a variety of ways, usually incorporating tracheoplasty with autologous grafts of costal cartilage or pericardium. Recent case studies suggest good results with expandable stents, balloon dilatation, and facilitation of the procedure with extracorporeal membrane oxygenation. We present our experience of congenital tracheal stenosis, and the evolution of our current technique in treating infants with severe tracheal stenosis.

RESULTS

Seventeen patients were diagnosed as having congenital tracheal stenosis, aged from birth to 16 months. Their lesions are described in the tabulation below.

<table>
<thead>
<tr>
<th>Endoscopic Findings</th>
<th>No. of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Funnel-shaped stenosis</td>
<td>9</td>
</tr>
<tr>
<td>Midtracheal stenosis</td>
<td>5</td>
</tr>
<tr>
<td>Hypoplastic trachea</td>
<td>2</td>
</tr>
<tr>
<td>Complete tracheal rings</td>
<td>15</td>
</tr>
<tr>
<td>Normal bronchi</td>
<td>10</td>
</tr>
<tr>
<td>Hypoplastic bronchi</td>
<td>5</td>
</tr>
<tr>
<td>Tracheal bronchus</td>
<td>1</td>
</tr>
</tbody>
</table>

The majority of lesions had complete tracheal rings with normal mainstem bronchi. Some of the stenoses had narrowing the size of a pinhole, such that a 2-mm telescope would not pass.
PATIENTS AND METHODS

From January 1986 to June 1996, 17 infants were diagnosed as having congenital tracheal stenosis at Children's Hospital, Boston, Mass. Nine infants with hypoplastic or funnel-shaped stenoses were diagnosed in the last 18 months of the study. We have no explanation for the large number of infants diagnosed in such a short period. The study was performed retrospectively, using the operative reports, medical records, and clinical notes to collect data regarding presenting symptoms, operative technique, postoperative management, and outcome.

Fourteen patients required surgical intervention of the trachea. The patients ranged in age from birth to 16 months. Presenting symptoms and signs included stridor, wheezing, respiratory distress, cyanosis, and croup, requiring diagnostic laryngoscopy and bronchoscopy in all cases. Computed tomography, magnetic resonance imaging, echocardiography, and cardiac catheterization were performed when necessary to better evaluate the intrathoracic anatomy.

Surgical repair of the trachea consisted of either resection and reanastomosis or tracheoplasty using 1 of 4 techniques: (1) splitting of the trachea with insertion of the pericardial patch and endotracheal tube stenting, (2) castellated incision of the stenosis with sliding tracheoplasty and pericardial patch, (3) splitting of the trachea with insertion of a costal cartilage graft, and (4) resection of the most stenotic portion of the trachea with splitting of the hypoplastic portion and application of an anterior tracheal graft using the resected trachea.

There was a high association of cardiovascular anomalies (Table 1). The most common association was a left pulmonary artery (LPA) sling. One patient with midtracheal stenosis had an LPA sling that required rerouting of the artery to relieve extrinsic compression of the trachea without tracheal repair.

Two infants were managed conservatively: 1 had Pfeiffer syndrome type III and a type I laryngeal cleft. This patient also had a severe funnel-shaped tracheal lesion and was managed with tracheotomy (Figure 1); a second patient with trisomy 21, atrioventricular canal, and a funnel-shaped tracheal lesion with normal bronchi was managed with repair of the cardiac lesion and observation for the airway. Figure 2 is an endoscopic view of a patient with complete tracheal rings following tracheoplasty.

Fourteen patients underwent repair of tracheal stenosis: 6 underwent resection and reanastomosis and 8 underwent tracheoplasty (Table 2). All procedures were facilitated by the use of cardiopulmonary bypass. The ages at diagnosis were as follows: neonates (0-4 weeks old), 6 patients; infants (1-12 months old), 10 patients; and toddlers (12-36 months old), 1 patient. Thirteen of 14 repairs were performed on infants (n = 6) or neonates (n = 7), and the remaining repair was performed on a toddler. Surgical techniques using pericardium and 6-0 Prolene or 5-0 PDS sutures were performed in the majority of patients. Management of an LPA sling currently involves resection of the affected segment of trachea and rerouting the pulmonary artery through the gap in the divided trachea. The pulmonary artery should be sutured anteriorly prior to reanastomosis of the trachea.

Exubation of patients was attempted as early as the first postoperative day in patients with uncomplicated procedures. The mean length of time of intubation was 5 days, with a range of 1 to 14 days. Seven patients required reintubation, ranging from 2 to 21 days postoperatively. One patient was reintubated on postoperative day 63 for respiratory syncytial virus infection. The causes for reintubation were granulation tissue (n = 4), cicatrix formation (n = 3), anterior graft collapse (n = 2), diffuse tracheitis (n = 1), and extrinsic compression (n = 1). Reintubation occurred almost exclusively in the patients with severe stenoses requiring tracheoplasty, and not those who underwent resection and reanastomosis.
Tracheal stenosis has historically had a poor prognosis with mortality rates near 50% in series as recent as 1997. The patients in our study had a survival rate of 93%, with 2 of 14 children currently being managed with long-term tracheostomy and the remaining doing well. Nine patients with hypoplastic or funnel-shaped stenoses were diagnosed in the last 18 months of the study. Two of these patients were managed conservatively, 1 with tracheostomy. The other 7 patients required tracheoplasty for management.

Congenital tracheal stenosis was associated with LPA sling in 7 (41%) of our patients. The majority of these patients had complete tracheal rings associated with their vascular sling. We also observed an association of tracheal stenosis with trisomy 21, pulmonary hypoplasia, VACTERL syndrome, Pfeiffer syndrome type III, and congenital heart disease.

Cardiopulmonary bypass was used in all patients who underwent repair of the trachea and LPA division and rerouting. Technically, the procedure is easier to perform when an endotracheal tube does not have to be present in the surgical field. Repair of the tracheal lesion was performed in conjunction with cardiac repair if the cardiothoracic surgeon believed this was indicated. There were no complications related to the use of cardiopulmonary bypass in this group of patients.

Patients with segmental stenosis did well with tracheal resection and reanastomosis. Rerouting of the LPA sling, if present, was performed at the time of the tracheal division to avoid division of the pulmonary artery. These patients were usually successfully extubated shortly after the repair, and had fewer postoperative bronchoscopies for stridor than did the patients who required tracheoplasty. In general, these patients had less severe stenoses that were more amenable to resection. There was only 1 patient with recurrence of stenosis at the suture line requiring dilatation and ultimately tracheotomy.

The majority of patients who required tracheoplasty for severe stenoses were treated in the last 18 months of this study. An initial poor outcome related to costal cartilage tracheoplasty caused us to reexamine our technique. We noted excessive granulation tissue forming at the cartilage graft site. We now recommend tracheoplasty with castellation or the slide technique under cardiopulmonary bypass in infants with long-
segment funnel-shaped or hypoplastic tracheas and bronchi. Resection of a portion of the tracheal stenosis with application of an anterior tracheal graft is reserved for the most severe lesions. Children with complicated repairs are kept intubated for 1 to 2 weeks to allow for stenting. Repeated bronchoscopy, sometimes urgent, is required to monitor granulation tissue formation. When patients developed significant granulation tissue, they were reintubated for 7 days, treated with antibiotics and steroids, and then extubated, usually with success.

CONCLUSIONS

Surgical and perioperative advances in the management of tracheal stenosis have made a great impact in the morbidity and mortality of a once nearly uniformly fatal disease. Preoperative evaluation of the tracheobronchial tree with direct laryngoscopy and rigid bronchoscopy is essential to ascertain the severity of the lesion. Segmental lesions are best treated with resection and reanastomosis, with early weaning of ventilatory support to reduce mucosal and anastomosis injury. Funnel-shaped stenosis and generalized hypoplasia of the trachea often require a more creative repair, combining partial resection of stenotic areas with tracheoplasty. The use of cardiopulmonary bypass facilitates precise tracheal anastomosis and is safe even in neonates. Follow-up bronchoscopy is crucial in maintaining airway patency in children with severe tracheal stenosis.

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REFERENCES