Nonoperative Management of Complete Tracheal Rings

Michael J. Rutter, FRACS; J. Paul Willging, MD; Robin T. Cotton, MD

Background: Children with complete tracheal rings are often challenging to manage. Most children will present early with a severely compromised airway and will require tracheal reconstruction.

Objective: To show that a small number of minimally symptomatic patients with complete tracheal rings experience airway growth over time and do not require tracheoplasty.

Design: A retrospective medical chart review over a 10-year period.

Setting: A tertiary care pediatric hospital.

Patients: Children (N = 10) with a diagnosis of complete tracheal rings, confirmed on bronchoscopy, who were observed for a minimum of 1 year prior to determining the need for tracheoplasty.

Main Outcome Measures: Patient symptoms, bronchoscopic findings, airway size, and the progression of these over time. Other congenital anomalies, the reason for initial diagnosis, and the need for tracheoplasty were documented.

Results: The 10 patients in our series fell into the following 3 categories: 5 patients were minimally symptomatic or asymptomatic, showed bronchoscopic evidence of progressive airway growth, and did not require tracheoplasty; 2 patients had worsening symptoms of exercise intolerance, showed minimal airway growth, and ultimately required tracheoplasty; and 3 patients are still being clinically observed and may eventually require tracheoplasty. Periods of observation have varied from 1 year to over 12 years.

Conclusions: Not all patients with complete tracheal rings require tracheoplasty. Some have satisfactory airway growth and do not require airway reconstruction. A period of observation to monitor airway growth and clinical symptoms is safe and may spare some patients from undergoing unwarranted airway reconstruction.

Arch Otolarngol Head Neck Surg. 2004;130:450-452

METHODS

We conducted a retrospective medical chart review of patients seen from April 1993 to March 2003 with a confirmed diagnosis of complete tracheal rings made on bronchoscopy, and in whom tracheoplasty was either not required or postponed for at least 1 year after initial diagnosis. Approval for this project was granted by Cincinnati Children’s Hospital Medical Center Institutional Review Board.

Airway diameter was estimated by the size of the largest endotracheal tube, endoscope, or bronchoscope that could be easily accommodated. If a 2.0-mm (inner diameter) endotracheal tube was used, the outer diameter of the tube (2.9 mm) was the estimate of airway size. Intervals between bronchoscopic
evaluations varied from 3 months for younger patients to 2 years for older asymptomatic patients. Patients were followed up until they stopped growing.

**RESULTS**

Ten patients fulfilled our inclusion criteria (Table 1), of whom 7 were male and 3 female. The age at initial diagnosis ranged from 5 to 98 months, with a mode of 14.5 months. Most patients, however, had been symptomatic since birth. One patient (patient 5) has been completely asymptomatic, with the diagnosis being made after difficulty with intubation for an incidental surgical procedure. In 3 patients, complete tracheal rings were not diagnosed on initial bronchoscopy. Additional congenital anomalies were present in 6 patients; these were cardiovascular in 3. Follow-up ranged from 12 to 138 months.

All patients had tracheal stenosis (Table 2). Two patients (patients 9 and 10) with progressive exertional dyspnea required tracheoplasty. These patients showed little evidence of airway growth over the period of observation. The 3 youngest patients (patients 6-8) are being closely observed, and 2 of these patients currently show evidence of airway growth. The third (patient 6) is likely to require future tracheoplasty. Five patients (patients 1-5) have shown evidence of significant airway growth, and we do not anticipate the need for future tracheoplasty. Four of these patients are asymptomatic, while 1 is only minimally symptomatic. One patient has been discharged from further follow-up with a mild airway stenosis (Figure).

**COMMENT**

The incidence of undiagnosed complete tracheal rings is unknown, and although this diagnosis has been made in adults, it is likely that most patients with complete tracheal rings are symptomatic and are diagnosed during infancy. The percentage of patients who will not require intervention or can safely have intervention delayed is also unknown. Since we are a tertiary care center, our referral pattern prevents us from accurately estimating this. However, we believe that up to 10% of patients with complete tracheal rings will not require tracheoplasty.

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Age at Diagnosis, mo</th>
<th>Initial Symptoms</th>
<th>Tracheal Segment Involved</th>
<th>Other Congenital Anomalies</th>
<th>Most Recent Age (or Age at Tracheoplasty), mo</th>
<th>Current Symptoms (or at Time of Tracheoplasty)</th>
<th>Other Observations</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>18</td>
<td>Stridor</td>
<td>Middle third</td>
<td>...</td>
<td>156</td>
<td>...</td>
<td>...</td>
</tr>
<tr>
<td>2</td>
<td>7</td>
<td>Stridor</td>
<td>Middle third</td>
<td>...</td>
<td>144</td>
<td>...</td>
<td>...</td>
</tr>
<tr>
<td>3</td>
<td>71</td>
<td>Stridor, recurrent cough</td>
<td>Middle half</td>
<td>...</td>
<td>120</td>
<td>...</td>
<td>...</td>
</tr>
<tr>
<td>4</td>
<td>18</td>
<td>Stridor with URI</td>
<td>Lower two thirds</td>
<td>Down syndrome, AV canal defect</td>
<td>60</td>
<td>Mild stridor with URI</td>
<td>Cardiac repair as an infant incidental finding during intubation</td>
</tr>
<tr>
<td>5</td>
<td>17</td>
<td>...</td>
<td>Middle third</td>
<td>Lacrimal stenosis</td>
<td>54</td>
<td>...</td>
<td>...</td>
</tr>
<tr>
<td>6</td>
<td>12</td>
<td>Stridor, SOBOE</td>
<td>Lower half</td>
<td>Bronchomalacia</td>
<td>42</td>
<td>Stridor SOBOE</td>
<td>Will need tracheoplasty required tissue expander right chest</td>
</tr>
<tr>
<td>7</td>
<td>5</td>
<td>Stridor</td>
<td>1-2 Distal rings</td>
<td>Right pulmonary agenesis, vascular compression</td>
<td>30</td>
<td>...</td>
<td>...</td>
</tr>
<tr>
<td>8</td>
<td>6</td>
<td>Stridor</td>
<td>Lower two thirds</td>
<td>...</td>
<td>18</td>
<td>Intermittent stridor</td>
<td>Cardiac repair as an infant incidental finding during intubation</td>
</tr>
<tr>
<td>9</td>
<td>98</td>
<td>Stridor, SOBOE</td>
<td>Middle two thirds</td>
<td>Pig bronchus</td>
<td>168</td>
<td>Stridor SOBOE</td>
<td>Slide tracheoplasty</td>
</tr>
<tr>
<td>10</td>
<td>5</td>
<td>Stridor, SOBOE</td>
<td>Lower third</td>
<td>Pig bronchus, anomalous SVC</td>
<td>54</td>
<td>Worsening SOBOE</td>
<td>Cardiac repair at 2 years, slide tracheoplasty</td>
</tr>
</tbody>
</table>

Abbreviations: AV, atrioventricular; SOBOE, shortness of breath on exertion; SVC, superior vena cava; URI, upper respiratory tract infection; ellipses, none.
Five patients in our series (patients 1-3, 5, and 7) have shown proven growth of the airway diameter in the region of the complete rings. Four of these patients have become asymptomatic over time, and 1 (patient 5) has never been symptomatic. After a period of observation, 2 patients (patients 9 and 10) required tracheoplasty at ages 4 and 14 years, respectively, owing to worsening symptoms. The 3 remaining patients are young, and we are not yet certain of the final outcome. It does, however, appear likely that 1 patient will require tracheoplasty in the future.

Three patients (patients 4, 7, and 10) required cardiovascular intervention. Patient 4 required an atrioventricular canal repair as an infant, with the diagnosis of complete tracheal rings not being made until 18 months of age. Patient 7 had a right pulmonary agenesis, with the heart lying in the right chest cavity, and the aorta stretched across the distal trachea causing marked tracheal compression, similar to that occasionally seen in postpneumonectomy syndrome. Because the aorta did not permit access for tracheoplasty, a skin expander balloon was placed extrapleurally in the right chest cavity and slowly inflated to move the heart back to the midline and lift the aorta off the trachea. Later tracheoplasty was planned but has not been required because of symptom resolution and continued airway growth. In patient 10, an increasing oxygen requirement disproportionate to the degree of stenosis prompted a Glenn procedure to correct the anomalous superior vena cava. Two years later, increasing shortness of breath on exertion with no further increase in tracheal diameter prompted tracheoplasty. No patients in this series had a pulmonary artery sling. Whether this represents a higher likelihood that patients with the ring/sling complex will require tracheoplasty or is simply a statistical anomaly due to the small number of patients in this series cannot be ascertained.

Most patients with complete tracheal rings require tracheoplasty. Surgical intervention is indicated if there is respiratory failure or a worsening of respiratory status. The younger the age at initial presentation, the more likely the need for tracheoplasty. However, for patients who present with few symptoms, a period of observation may be appropriate. This study demonstrates that in a proportion of these patients complete tracheal rings can grow, thus allowing nonoperative management. In patients with persistent or worsening symptoms and minimal increase in the tracheal diameter, tracheoplasty is eventually required. This procedure is not without risk, and intervention is better justified in a patient with significant and worsening symptoms. Moreover, tracheoplasty is technically easier to perform in a larger child.

Submitted for publication August 12, 2003; accepted September 9, 2003.

This study was presented at a meeting of the American Society of Pediatric Otolaryngology; May 5, 2003; Memphis, Tenn.

Dr Rutter had full access to all the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis.

Corresponding author and reprints: Michael J. Rutter, FRACS, Division of Pediatric Otolaryngology/Head & Neck Surgery, Cincinnati Children’s Hospital Medical Center, 3333 Burnet Ave, Cincinnati, OH 45229-3039 (e-mail: mike.rutter@cchmc.org).

REFERENCES