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The role of conservative management in congenital tracheal stenosis: an evidence-based long-term follow-up study

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Abstract

Background/Purpose: Surgery has been the management of choice for severe congenital tracheal stenosis (CTS). The role of conservative management of CTS however is not clear. The aim of this study is to characterize the natural history of CTS, review the radiologic evidence of tracheal growth, and evaluate the clinical outcome and selection criteria of conservative management of CTS.

Methods: A retrospective study was carried out on 22 consecutive children with symptomatic CTS admitted into a single institution between 1982 and 2001. The patients were categorized into operation (n = 11) and observation (n = 11) groups. Six patients of the observation group were followed up with serial computed tomography scan. Their tracheal growth was compared with that of healthy children of the same age.

Results: The mortality rates of observation and operation groups were 9% and 27%, respectively, although the latter group consisted of more severely affected patients. The pathologic categorization of the CTS influenced the survival rates ($P = .046$, χ^2), with the long segment type having the worst prognosis (67%). Serial computed tomography scans of 6 conservatively managed patients revealed that all stenotic tracheas continued to grow ($P = .039$, 2-tailed paired Student's t test). Of the 6 stenotic tracheas, 5 grew at a faster-than-normal rate, and the stenotic tracheal diameters approached those of normal diameters by the age of 9 years.

Conclusions: The management of patients with symptomatic CTS should be individualized. A selected group of patients with CTS can be safely managed nonoperatively.

Congenital tracheal stenosis (CTS) is a rare but potentially life-threatening condition because of various degrees of airway obstruction. It is disquieting for some clinicians that the airway obstruction would progressively deteriorate as the child grows, and the child's lung would outgrow the capacity of tracheal air conductance resulting in respiratory failure or death [1]. Surgical intervention is considered to offer the best opportunity of survival for those children with CTS who have severe symptoms [2]. The surgery includes resection of the stenotic segment and primary anastomosis, slide tracheo-

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plasty, patch tracheoplasty with cartilage, pericardium, or anterior wall of esophagus, as well as tracheal dilatation with or without stent insertion [3]. These procedures require great deal of surgical expertise and carry certain degrees of morbidity and mortality [4,5].

With the advent of ventilating fiberoptic bronchoscopy and other imaging studies, a form of CTS with less severe symptoms is being increasingly recognized [6]. Whether this subgroup of patients would develop airway obstruction and eventually require surgical intervention remains unclear. Because of the scarcity of the condition, understanding the natural history of CTS remains a challenge. Patient survival after conservative management has been reported previously [7]. We speculate that conservative management may be more widely practiced than reported. Yet the rationale behind nonoperative management begets evidence. We herewith report successful outcome of a subgroup of conservatively managed patients with CTS. Follow-up with serial computed tomography (CT) measurement of the tracheal diameters in conservatively managed patients showed a faster-than-normal (catch-up) tracheal growth.

1. Materials and methods

With the internal review board approval, a retrospective study of 22 consecutive cases of symptomatic CTS managed in our institution between 1982 and 2001 was carried out. Subglottic stenosis of trachea secondary to intubation and tracheomalacia were excluded from the series. The diagnosis of CTS was based on bronchoscopic and radiologic evidences in addition to clinical findings. The patients were either operated on or observed, based on the severity, CT and/or bronchoscopy findings, surgeons' preference, and parental consents. Nonoperative management consisted of treating the respiratory tract infection, chest physiotherapy, and short-team oxygen therapy. The data were statistically analyzed with SPSS (v12.0; SPSS, Chicago, Ill), and the trend charts were generated with Excel (Microsoft Office XP; Microsoft, Redmond, Wash).

1.1. Tracheal diameters and tracheal growth rates

The lumen diameters of the most stenotic part of the trachea of each patient were measured on CT scan images (in millimeters) according to the method delineated by Griscom and Wohl [8]. Admittedly, the observers were not blinded. Normal tracheal lumen diameter (in millimeters) of a child of the same age was gauged from the published normograms and formula [7,9,10]. Normal full-term neonatal tracheal diameter average was estimated to be 3.75 mm [11]. The rate of tracheal diameter growth in the first year of life was assumed to be linear. The stenotic trachea diameters were then expressed as percentages of the tracheal diameters of the normal age-matched children. The averages of anterior-posterior and transverse diameters were used in the analysis.

Catch-up growth of the trachea was defined as tracheal growth at a rate faster than that of normal growth rate. The tracheal catch-up growth rate was estimated by the formula:

$$\frac{\text{Tracheal lumen diameter in percentage}^{\text{2nd CT}} - \text{Tracheal lumen diameter in percentage}^{\text{1st CT}}}{\text{interval period (months)}}$$

The unit of the catch-up growth rate was expressed as percentages gained per month.

Presenting tracheal diameter as a percentage of normal was purposely designed so that stenotic tracheal growth could be appreciated in relation to normal tracheal growth. If tracheal diameters were expressed as millimeters and tracheal growth rate as millimeters per month, one would find it difficult to appreciate (1) whether a given growth rate is faster or slower than that of normal tracheal growth rate, and (2) a child may be experiencing increasing tracheal stenosis in relation to his/her body size, although the actual tracheal lumen diameter is increasing. By presenting the tracheal growth rate as percentages gained per month, one would be able to evaluate the growth of stenotic trachea in relation to that of normal trachea. A positive number signifies a tracheal growth rate that is faster than that of normal growth rate (catch-up growth). Zero signifies a growth rate equal to that of normal. A negative number signifies a slower than normal growth rate or no growth.

2. Results

2.1. Patient demographics

Congenital tracheal stenosis affected predominantly male infants (M/F = 20:2) in this series, and congenital cardiac anomaly was the most common associated anomaly (36%, 8/22). Of 22 patients, 7 (32%) had anomalous left pulmonary artery sling. The most common pathology was funnel-shaped stenosis (50%), followed by segmental stenosis (36%). The long segment stenosis where more than 50% of the tracheal length was stenotic was least common (14%). The patients presented with various symptoms of airway obstruction including dyspnea (n = 6), stridor (n = 5), repeated respiratory tract infection (n = 4), cough (n = 4), wheeze (n = 4), and exercise intolerance (n = 3). The whole group (n = 22) was followed up for 10.6 years. Eleven children were observed, whereas 11 were operated on. Tracheoplasty was carried out using costal cartilages (n = 6), anterior esophageal wall (n = 3), periosteum (n = 1) and pericardium (n = 1), and tracheal dilatations (n = 2) with or without stent.

2.2. Clinical outcome

The operation group of patients presented earlier than the observation group (0.4 vs 1.5 years). The symptoms appeared to be more severe, although the recorded