Tracheomalacia after Esophageal Replacement: Role of Aortopexy

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ABSTRACT
Diffuse malacia of the airway is a rare condition; no definite incidence rates are available. It can be associated with a variety of congenital anomalies, but internal compression by an endotracheal or tracheostomy tube also may be the culprit. We describe a one-year-old boy with tracheomalacia that developed after a gastric pull-up procedure for esophageal replacement due to esophageal atresia. After being under mechanical ventilation for 28 days and impossible to extubate, he improved dramatically after performing aortopexy. (Tanaffos 2004; 3(10): 63-67)

Key Words: Tracheomalacia, Esophageal replacement, Aortopexy

INTRODUCTION
Tracheomalacia is a structural abnormality of the tracheal cartilage allowing collapse of its walls and airway obstruction. A deficiency and/or malformation of the supporting cartilage exists, with a decrease in the cartilage-to-muscle ratio. The disease can be categorized into 3 groups based on histological, endoscopic, and clinical presentation:

Type I: Congenital or intrinsic abnormalities that can be associated with a tracheoesophageal fistula.

Type II: Extrinsic defects or anomalies, such as a vascular ring causing undue pressure on the trachea.

Type III: Acquired tracheomalacia that occurs with prolonged intubation or chronic tracheal infections.

Immaturity of the tracheobronchial cartilage is thought to be the cause in type I, whereas degeneration of previously healthy cartilage is thought to produce other types.

Inflammatory processes, extrinsic compression from vascular anomalies, or neoplasms may produce degeneration.

The definitive diagnosis depends on obtaining an accurate history combined with proper endoscopic evaluation. The airway is directly visualized during spontaneous respiration using ventilating laryngoscope and telescoping bronchoscopy.

Tracheomalacia generally is benign; most infants outgrow the symptoms by age 18-24 months. Surgical therapy is only recommended for severe symptoms and failure of conservative treatment.

Here we describe a one-year-old boy who was admitted at our hospital for esophageal replacement due to a long-gap esophageal atresia. After a gastric pull-up procedure, he became ventilator dependent...
and could not be extubated. Finally he was diagnosed as having tracheomalacia that was successfully treated by means of aortopexy. To the best of our knowledge, this is the first report of aortopexy performed in Iran.

**CASE REPORT**

A male second-child patient, from non-consanguineous parents was born with esophageal atresia and was referred to our hospital for treatment. At operation, an esophageal atresia common type was found, but both segments were so widely separated that a cervical esphagostomy and a gastrostomy were performed. After an uneventful post-op, the patient was discharged and maintained with feedings through gastrostomy without complications. At the age of one year and a body weight of 12 kg, he was readmitted for esophageal replacement. A contrast radiography through the gastrostomy showed a short esophageal distal end that was not apt for primary anastomosis. At operation through a midline laparotomy the distal esophageal end was removed, and the gastroesophageal junction oversewn. The left gastric and short vessels were divided preserving the right gastric and gastroepiploic vessels. The duodenum was mobilized by the Kocher maneuver and the stomach was passed through the esophageal hiatus. A right thoracotomy was performed in order to help accommodate the stomach at the neck for a tension-free anastomosis in one layer of absorbable material between the gastric fundus and the proximal esophagus. A pyloroplasty and a jejunostomy completed the surgical procedure. After 72 hours of mechanical ventilation and with hemodynamic and homeostatic stability, attempts of weaning failed systematically with dramatic drops in the oxygen saturation and quick development of fatigue. Besides, at the 12th post-operative day, a lateral salivary fistula appeared. The condition of the respiratory system persisted for 28 days; moreover, after this period, a bronchoscopy (flexible) depicted a proximal and distal tracheomalacia. 24 hours after the diagnosis, the patient was treated with tracheostomy as an open procedure with a cannula #4.5. Unfortunately, this operation did not improve the patient’s condition, and he could not tolerate disconnection from the respirator even for very short periods of 10 minutes. At that moment the decision of performing an aortopexy was made. The patient underwent aortopexy 32 days after the gastric pull-up through a left anterior thoracotomy. Left lobe thymectomy improved the exposure, and a single row of interrupted monofilament sutures was placed from the ascendant aorta to the undersurface of the sternum. The operation proved to be successful as the patient could be disconnected from the respirator just 24 hours after the procedure without signs of muscular fatigue or drops in the oxygen saturation.

He resumed oral feedings 48 hours later and was discharged from the ICU, 4 days after the operation.

**DISCUSSION**

Tracheomalacia is a process characterized by flaccidity of the supporting tracheal cartilage, widening of the posterior membranous wall, and reduced anterior-posterior airway caliber (1). These factors cause tracheal collapse especially during times of increased airflow such as coughing, crying or feeding. It seems evident that our patient had a type III tracheomalacia with prolonged intubation. Cases of acquired tracheomalacia occur with increasing frequency both in children and adults, and the tracheomalacia often is not recognized clearly (1,2). These lesions usually cause focal tracheomalacia and may result from indwelling tracheostomy and endobronchial tube, chest trauma, chronic tracheobronchitis, and inflammation (relapsing polychondritis). They may be secondary to pulmonary resection and tracheal malignancy.
(cylindroma), and they may be idiopathic. Tracheomalacia most commonly affects the distal third of the trachea (3,4).

By virtue of its intrinsic flexibility, or compliance, the trachea changes caliber during the respiratory cycle. Tracheal dilatation and lengthening occurs during inspiration; narrowing and shortening occurs during expiration. Accentuation of this cyclic process may cause excessive narrowing of tracheal lumen, thus, deforming the entire length or a localized segment. The functional interference with ventilation may cause expiratory flow obstruction and interfere with clearance of secretion. Functional impairment is proportional to the length of the involved segment and the degree of stenosis (5,6).

Bronchoscope provides the diagnosis. The findings consist of the following classic triad:
- Loss of normal semicircular shape tracheal lumen.
- Forward ballooning of the posterior membranous wall
- Anteroposterior narrowing of the tracheal lumen

Tracheostomy helps to maintain an airway while the child is growing and the trachea regains structural integrity, but the problem with this procedure is that the tracheostomy tube may not support the distal trachea. Aortopexy can provide relief of tracheal compression and relieves the external pressure on flaccid trachea. It is not a perfect operation because of a small but significant failure rate and potential for complications (7,8).

Surgical therapy is indicated when conservative measures fail. The indications for tracheostomy are severe symptoms, failure of conservative therapy, and proximal or diffuse tracheomalacia. The indications for aortopexy are dying spells or reflex apnea, recurrent pneumonia, intermittent obstruction, and inability to extubate airway in an infant who is intubated. For distal tracheomalacia, aortopexy is the procedure of choice.

Aortopexy has proven to be a safe, expedient way to relieve the problem of tracheomalacia in most patients. The success of aortopexy has been reported at about 75% in several small studies. Aortopexy has less long-term morbidity than tracheostomy. While not altering the structural characteristics of the tracheal wall, it widens the anterior-posterior tracheal dimension to maintain a patent lumen.

As experience accumulates, a direct surgical approach for treating tracheomalacia may replace tracheostomy in the management of proximal and diffuse tracheomalacia. These procedures include prosthetic stenting, tracheoplasty, and tracheal resection with end-to-end anastomosis (9, 10,11).

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