Fixing the "Presumed" Unfixable: The Tracheopexy Procedure Improves the Lives of Infants and Children with Tracheomalacia

**Introduction:** Tracheomalacia (TM) is defined as a deformity of the tracheal cartilage and posterior membrane, leading to tracheal collapse, especially when increased airflow is demanded. Symptoms most often are seen in infants and children during expiration due to an increase in intra-thoracic pressure causing collapse. This occurs most dramatically during forced expiration, as when the infant or child is crying or coughing. Parents may describe a brassy or barking cough. Tracheomalacia results in mucous trapping, which can lead to an increased number of respiratory infections, increased oxygen demand, and irreversible long-term lung damage such as bronchiectasis. Collapse of the trachea decreases air exchange, and affects exercise tolerance. It can also present as a Brief Resolved Unexplained Event (BRUE).

**Review:** The Esophageal and Airway Treatment Center at Boston Children’s Hospital manages patients with complex medical needs, including esophageal atresia/tracheo-esophageal fistula (EA/TEF). These patients often have TM in addition to their underlying condition. Our center has also seen a rise in the number of patients with isolated TM without EA/TEF. Dr. Russell Jennings has developed the posterior tracheopexy; a surgical procedure in which the posterior membrane of the trachea is sutured to the anterior spinal ligament thus opening the airway and decreasing the degree of collapse. Our multidisciplinary team cares for these patients and has seen improved outcomes at an even greater level than with the aortopexy procedure for TM, once thought to be the mainstay of treatment. These patients are able to avoid tracheostomy, those with tracheostomies have been successfully decannulated following tracheopexy, and patients have seen decreased respiratory infections and hospitalizations, and have been more able to live a normal and active life.

**Results:** Patients who undergo tracheopexy have had a decrease in symptoms such as persistent cough, noisy breathing, prolonged or recurrent respiratory infections, oxygen/ventilator dependence, and ALTE’s. In addition, families also report improved quality of life and better psychosocial well-being.

**Conclusion:** Our patients who have undergone the tracheopexy procedure at our institution have experienced a decrease in their preoperative symptoms of tracheomalacia. In addition, the surgery has improved children’s lives allowing for an active and healthy life in a condition that was presumed unfixable in the past.

**Future project:** Examination of long-term (5 year) follow-up of families whose infant or child has undergone tracheopexy. We will be evaluating symptoms and quality of life with a survey that will be filled out by the caregiver/patient. Objective measures will be scored such as symptom frequency, tracheostomy/ventilator dependence, pulmonary function tests, and follow up bronchoscopy.

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Background

Tracheomalacia (TM) is a deformity of the tracheal cartilage and posterior membrane that leads to tracheal collapse, especially when airflow demand is increased.
- Occurs most dramatically during forced exhalation, as when the infant or child is crying or coughing.
- Results in mucus trapping, leading to an increased number of respiratory infections, increased oxygen consumption, and irreversible long term lung damage such as bronchiectasis.
- Parents report brassy or barking cough. It may affect exercise tolerance and can be the cause of BRUE (brief resolved unexplained events, formerly ALTE).

Purpose

The posterior tracheopexy procedure is a surgical technique in which the posterior membrane of the trachea is sutured to the anterior spinal ligament, thus opening the airway and decreasing the degree of collapse. Mucus can then clear more effectively, oxygen exchange is easier and more effective.

Here we describe our approach to patients who have tracheomalacia, with or without esophageal atresia/tracheoesophageal fistula (EA/TEF), at the Boston Children's Hospital Esophageal and Airway Treatment Center.

Methods: (Interdisciplinary team approach)

- Preoperative: triage phone calls and clinic visits for initial and ongoing patient assessments in conjunction with multidisciplinary team to evaluate whether symptoms fit the tracheomalacia picture.
- Inpatient: involvement in daily collaborative clinical decision making and ongoing discharge preparation and planning.
- Outpatient: post discharge evaluation of improvements in symptoms, phone triage, clinic visits, discontinuation of medications, and monitoring improvements in quality of life.

Meet Sarah

4 year old girl with history of EA/TEF repaired at birth. Found to have tracheomalacia and subglottic stenosis requiring tracheostomy. Underwent laryngotracheal repair (LTR) in Nov 2015. Presented to BCH for eval/treatment of tracheomalacia to optimize success of decannulation Prior to tracheopexy, suffered pneumonia every month, required PT vest, mechanical ventilator support when hospitalized, and medical management.

Airway CT

Sarah's airway CT reveals absence of airflow at end of expiration.

Bronchoscopy

Sarah's bronchoscopy reveals severe TM with complete airway collapse. Medical treatment includes:
- Atrovent
- Inhaled steroid
- PT vest
- Hypertonic saline

Tracheotomy

Sarah underwent tracheotomy and tracheal diverticulum resection in April 2016.

Post Op

Sarah was decannulated May 2016. She has had no lower airway symptoms since the tracheotomy. She has been able to stop the vest, hypertonic saline and all airway medications with no recurrence of symptoms. She has no chronic cough, no exercise intolerance, and no frequent or prolonged upper respiratory infections (URI).

Outcomes

Through observation and parental report in outpatient clinic and phone communication, we have seen improvement:
- In symptoms such as persistent cough/noisy breathing, prolonged or recurrent respiratory infections, and BRUE's.
- In oxygen/ventilator dependency.
- In quality of life, better psychosocial well-being, enabling a more active and healthy life.

Future

To further investigate symptom management and quality of life by examining the long term (5 year) follow up of families whose infant or child has undergone tracheotomy. We will create a preoperative and postoperative symptom and quality of life survey to be filled out by the family.

Lessons Learned

- Not always able to diagnose TM in newborns due to their decreased intra-thoracic pressures and compliant airways.
- It takes the expertise of a multidisciplinary team with ongoing communication for successful management of these unique patients.
- Needs to be on the differential diagnosis list when evaluating all symptomatic infants and children with EA/TEF and for patients who suffer from frequent respiratory infections and cough without comorbidities.
- Not necessary to see patients back for bronchoscopy until one year post surgery with a clinic visit unless symptomatic earlier.

References


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