# Management of Severe Tracheomalacia

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Tracheomalacia (TM) is defined as a generalized or localized collapse of the tracheal lumen causing luminal obstruction during respiration and is considered pathologic when obstruction exceeds more than 50% of the lumen. TM is present in 75% of patients with esophageal atresia (EA) with distal tracheoesophageal fistula (TEF), but severe TM often presents with acute life-threatening events (ALTEs) in only 15% to 33% of patients (1). TM is 1 of the major causes of morbidity and late death in patients with EA/TEF. Death from TM may be prevented by its early diagnosis, adapted assessment, and aggressive management.

Physiopathology of TM in patients with EA/TEF remains unclear but histologic and experimental studies report malformations of cartilage with fragmented cartilage, absence of lateral arms, or distortion of tracheal rings (2). Cartilaginous anomalies are associated with a concomitant increase in the length of muscle in the membranous part of the tracheal ring. Moreover, tracheal deformation is frequently associated with a bulging of the posterior wall related to dilated former esophageal pouch that can lead to a total anteroposterior collapse of the trachea. In addition, TM is occasionally associated with laryngeal or tracheobronchial malformations or vascular anomalies that can participate in airway obstruction (3,4).

Clinically, mild-to-moderate TM may be asymptomatic apart from the characteristic barking cough, stridor, or recurrent chest infections. Severe TM presents with dyspnea and recurrent ALTE, which are usually associated with feeding, crying, or coughing.

The current criterion standard of diagnosing TM is by visual assessment of tracheal collapse during bronchoscopy. Flexible bronchoscopy is superior to rigid bronchoscopy for evaluation of airway collapses (5). Flexible bronchoscopy provides the best assessment for the dynamic airway changes found in tracheobronchomalacia, and allows a complete laryngotracheal examination. Spiral computed tomography gives high-resolution and advanced volumetric assessment of tracheal lumen and reveals the spatial relation of airways and mediastinal structures (6). Dynamic acquisitions with inspiration and expiration spirals are difficult to obtain in young infants.

Severe respiratory symptoms are not only attributed to TM but also may be due to gastroesophageal reflux, recurrent TEF, associated laryngotracheal malformations, and esophageal stricture, although many patients have multiple causes (7). Moreover, the severity of symptoms is not directly correlated with the degree of tracheal collapse observed during endoscopy. Finally, the diagnosis of severe TM may be considered in cases of tracheal collapse associated with severe respiratory symptoms and without or after control of other etiologies.

When the diagnosis of severe TM is established and based on the results of the functional and morphological assessment, appropriate management must be proposed as soon as possible to avoid more ALTE. In cases of TM associated with EA/TEF, aortopexy is still the criterion standard treatment. Aortopexy was described in 1948 for the treatment of tracheal vascular compression (8) and used later for TM. We prefer anterior left thoracotomy, but right thoracotomy is also possible. When possible, the pleura is not opened and the left thymic lobe is mobilized and excised with great attention to the phrenic nerve. The pericardium is opened from the aortic root to the aortic reflection. Two to 3 rows of stitches are placed in the adventice of the ascending aorta and passed through the sternum. The surgeon's assistant applies with his or her fingers a pressure on the sternum while the surgeon ties the sutures. Sternal pressure is then released, and the effect on tracheal lumen opening is evaluated by bronchoscopy through the tracheal tube. Before tying the sutures, mobilization of the aorta under endoscopic control may show insufficiency in the tracheal opening; the sutures may be changed or more may be needed even on the innominate artery. Aortopexy had a high success rate in the resolution of ALTE (9), but some major complications occurred (Table 1). Phrenic nerve injuries must be avoided by meticulous excision of the thymic lobe; the pericardium must be left open to prevent postoperative pericardic effusion with a chest tube left in the mediastinum. Aortopexy may be repeated if signs do not improve, but some patients show amelioration only after fundoplication. Midline incision with sternal split allows a larger approach of the anterior mediastinum and may help to obtain the aorta optimal mobilization (10). A cervical approach also has been proposed, allowing tracheopexy if needed in the same operation (11). Thoracoscopic aortopexy was also reported; much experience in thoracoscopic procedures is needed but cosmetic and functional results are better (12).

Internal or external tracheal stenting has been proposed for the treatment of not only primary but also secondary TM. Experience in pediatric population with EA/TEF with TM is more limited than in isolated tracheal diseases (congenital tracheal stenosis, postheart–lung or lung transplantation, malignant mediastinal mass, and postprolonged intubation stenosis).

The severe TM related to EA/TEF occurs in early infancy in small babies, and endoscopic maneuvers require a skilled team and continuous availability. The procedure itself may be difficult, and complications of stents are numerous and may include tissue granulation, stent dislodgments, need for stents to be expanded again, difficulties in stent removal (13), and stent epithelialization (Table 2). To prevent these complications, different types of stents may be proposed. The Palmaz stent is an expandable stent in stainless steel with incrustation granulation and hemorrhagic complications. The Wallstent is an expandable cobalt-alloy tubular mesh stent, the Nitinol/Ultraflex stent is a shape memory stent covered or not with an alloy of titanium-nickel, and the silicone stents (Dumon) and the metallic stent. In addition, the size of the

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TABLE 1. Main results and complications of aortopexy

| Author | Results | Major complications |
|---|---|---|
| Vasquez-Jimenez et al (10) | 28/29 resolved, 1 resolved after fundoplication | 5 scoliosis |
| Morabito et al (11) | 15/16 resolved, ALTE abolished in all | 1 vocal cord palsy |
| Weber et al (14) | ALTE abolished in all | None |
| Abdel-Rahman et al (15) | 13/16 resolved, ALTE abolished in all | None |
| Valerie et al (13) | 11 patients 8 AE/TEF | 2 pericardial effusions drained and 2 not drained, 1 postoperative reintubation |
| Dave and Currie (9) | ALTE abolished in 28/28, 15 AE/TEF | 3 lung consolidation, 1 phrenic nerve palsy, 1 thymic engorgement |

AE/TEF = esophageal atresia with or without tracheoesophageal fistula; ALTE = acute life-threatening event.

TABLE 2. Major complications of tracheal stenting

| Author | Results | Type of stent | Longest follow-up and complications |
|---|---|---|---|
| Nicolai et al (16) | 7 patients, 19 procedures, no AE/TEF patient<br>Rapid improvement of respiratory obstruction in all | Nitinol/Titan | 50 mo with bronchial stent<br>4 deaths not related to stent placement |
| Kumar et al (17) | 17 patients, no AE/TEF patient<br>6/8 extubated after airway stenting | Wallstent | 54 mo<br>9 deaths not related to stent placement |
| Geller et al (18) | 9 patients with severe tracheomalacia | Palmaz | 86 mo with tracheal stent, 30 mo with bronchial stent<br>4 deaths, with 3 from tracheal hemorrhage and 1 not related to stent placement |
| Valerie et al (13) | 14 patients, 9 with AE/TEF, 8/14 extubated shortly after the stent<br>1 death, 2 cardiac arrest after removal with 1 death | Palmaz | Up to 2 y |

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stent needed in neonates is also a major problem, and today it seems there is no single ideal stent (19).

In our experience, 10 babies with severe TM had aortopexy with ALTE abolishment, or weaning from mechanical ventilation in all. All of them were operated before age 8 months. Their mean follow-up is 3 years; all of them still have respiratory signs.

In conclusion, severe TM in EA/TEF should be managed promptly and with a tailored plan for each patient. Aortopexy can abolish the ALTE events and have acceptable complications and no operative mortality. Long-term follow-up to detect persistent obstructive disease in these patients and a long-term respiratory follow-up are therefore mandatory.

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