Care Recommendations for the Respiratory Complications of Esophageal Atresia – Tracheoesophageal Fistula

Internationale Empfehlungen zur Versorgung respiratorischer Komplikationen bei Ösophagusatresie – tracheoösophagealer Fistel – NEWS2025

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ABSTRACT

This article summarizes publications, guidelines, position statements as well as consensus recommendations from the working groups of INoEA, the European Reference Networks ERNICA and ERN-LUNG as well as the European Respiratory Society (ERS) about Esophageal atresia (EA) – tracheoesophageal fistula (TOF).

EA-TOF is frequently associated with lifelong and sometimes severe respiratory impairments. Respiratory diseases and restraints include tracheobronchomalacia, aspiration risk, recurrent pulmonary infections, bronchitis, and atelectasis, which may occur both before and after surgical repair.

The article provides a structured approach to the diagnosis, management, and long-term care of these respiratory complications. The overarching aim is to sustainably improve health outcomes and quality of life in affected individuals.

ZUSAMMENFASSUNG

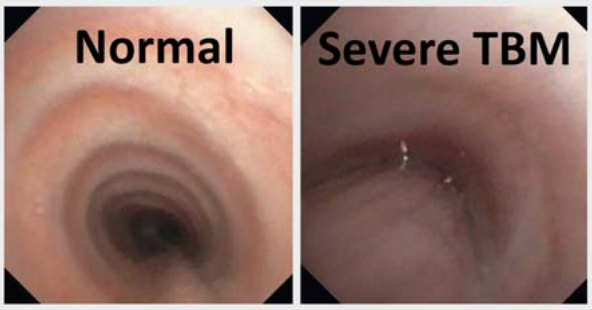
Dieser Artikel fasst aktuelle Veröffentlichungen, Leitlinien, Stellungnahmen sowie Konsensempfehlungen der Fachgruppen von INoEA, der Europäischen Referenznetzwerke ERN-Lung und ERNICA sowie der European Respiratory Society (ERS) zur Ösophagusatresie (ÖA) – Tracheoösophageale Fistel (TÖF) zusammen.

Die ÖA-TÖF ist häufig mit lebenslangen, teils erheblichen respiratorischen Einschränkungen assoziiert. Zu den typischen Komplikationen zählen Tracheobronchomalazie, Aspirationsrisiken, rezidivierende pulmonale Infektionen, Bronchitiden und Atelektasen, die sowohl vor als auch nach der chirurgischen Korrektur auftreten können. Der Artikel stellt einen strukturierten Ansatz zur Diagnostik, Therapie und Langzeitbehandlung dieser respiratorischen Folgeprobleme vor. Ziel ist die nachhaltige Verbesserung der Gesundheitssituation und Lebensqualität der Betroffenen.

Introduction

This summary document is based on a publication by Koumbourlis et al. [1] and incorporates recent statements and guidelines of the European Respiratory Society as well as consensus recommendations of INoEA, ERNICA and ERN-LUNG working groups.

Esophageal atresia (EA) and tracheoesophageal fistula (TEF) present significant lifelong respiratory challenges. Involvement of a physician with expertise in respiratory care is therefore recom-



▶ **Fig. 1** Normal trachea during bronchoscopy on the left, severe tracheomalacia (> 75% of lumen narrowing) shown on the right. The degree of obstruction may vary among patients.

mended, specifically in infancy and at the preschool age. The recommendations of the INoEA's Respiratory Complications Working Group (RCWG) [2] provide a structured approach to the diagnosis, treatment, and long-term care of these complications, aiming to improve patient outcomes. The RCWG identified the primary respiratory conditions faced by patients before and/or after EA-TEF repair, including:

Tracheobronchomalacia (TBM)

Tracheobronchomalacia is the partial or complete collapse of the main windpipe (trachea and/or main stem bronchi) due to abnormal airway wall softness or floppy cartilage. It is present in nearly all EA-TEF patients, with severity varying based on airway narrowing (▶ **Fig. 1**).

TBM may cause acute symptoms in neonates and infants but can also result in long-term complications (▶ **Fig. 2**, **Table 1**).

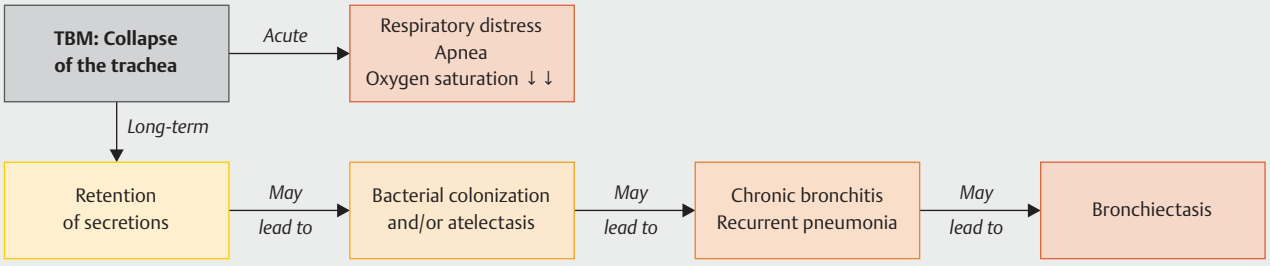
The symptoms of TBM vary according to the age of the patient. They tend to be most severe in the neonatal period and generally may improve with time (▶ **Table 1**).

Diagnosis of TBM

Flexible bronchoscopy under light general anesthesia with spontaneous breathing without positive end expiratory pressure (PEEP) is the most informative method for assessing TBM presence, extent, and severity. Dynamic CT scans and advanced MRIs can also aid evaluation. The shape of the flow volume curve from spirometry (▶ **Fig. 3**) can be indicative of TBM.

Management of TBM

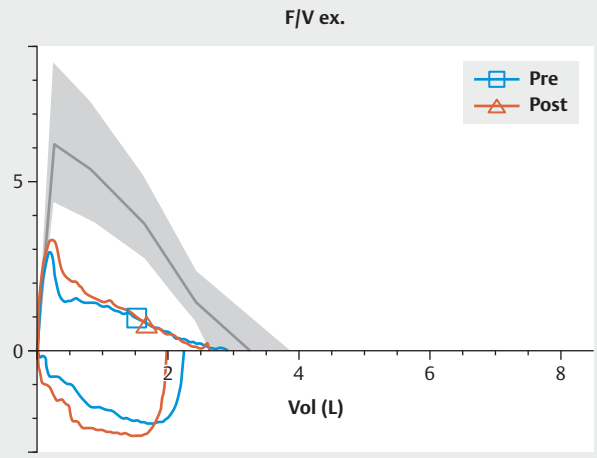
Interventions aim to reduce symptoms due to TBM and, when it is severe, reduce TBM severity by surgery. In neonates, the primary goal is to prevent respiratory distress and low oxygen levels. Treat-



▶ **Fig. 2** Acute and long-term consequences of tracheobronchomalacia.

▶ **Table 1** Symptoms of TBM according to age.

Neonates-Infants	Children-Adolescents	Adults
Respiratory distress	Recurrent wheeze	Recurrent wheeze
Apnea	"Honking"/"Brassy" and/or "wet" cough	Recurrent "wet" and/or "honky" cough
Episodes of severe hypoxemia (low oxygen saturation or "blue" or "dusky" spells)	Hoarse voice/Stridor	Hoarse voice
Recurrent harsh "wheeze"	Exercise Intolerance	Exercise intolerance
"Honking"/"Brassy" cough	Obstructive lung function, low peak expiratory flow (PEF)	Obstructive lung function, low peak expiratory flow (PEF)
Stridor/"hoarse" cry		
Recurrent airway infections	Recurrent airway infections	Recurrent airway infections



▶ **Fig. 3** Example of flow-volume curves during forced expiration before (pre) and after (post) inhalation of a short acting beta-mimetic in a patient with esophageal atresia and tracheobronchial malacia. In this patient, there was no major change with bronchodilator. Normal range is shown in the shaded grey area. In some cases, beta-mimetics have a negative effect on expiratory flow. Curves vary between patients.

ments range from supplemental oxygen use to non-invasive positive airway pressure. Severe cases may require intubation, mechanical ventilation, or surgery (tracheopexy/aortopexy) generally after the initial EA repair. Rarely, a tracheostomy is needed.

Long-term management of TBM

Long-term management of TBM focuses on clearing airway secretions and preventing airway infections. This can be achieved through age-appropriate airway clearance techniques (in adults: active cycle of breathing techniques), positive expiratory pressure devices such as PEP masks or oscillatory positive pressure devices [2, 3]. Nebulized hypertonic saline may help to improve mucociliary clearance and inhaled corticosteroids may reduce airway inflammation in selected patients. Infections, often signaled by a "wet" cough or fever, are treated aggressively with antibiotics. Bronchodilators should be used cautiously, as they can worsen airway collapse.

Aspiration Risk

Aspiration risk in EA-TEF

EA-TEF patients are highly prone to aspiration due to multiple factors:

- Esophageal dysmotility and/or strictures, causing food or fluids to enter the airway.
- Recurrent TEF
- High prevalence of gastroesophageal reflux (GER)
- Occasionally, a laryngeal cleft

Diagnosis

A systematic approach is essential to evaluate all potential aspiration causes. Swallowing function can be assessed via videofluoroscopic or modified contrast swallow studies. Esophageal narrowing and function are diagnosed by contrast swallow studies and sometimes with manometry. Recurrent TEF is diagnosed using a "pull-back" contrast study, bronchoscopy and/or upper endoscopy. GER is identified clinically or through contrast studies, pH-impedance monitoring, or endoscopy. Chronic bacterial infection due to aspiration can be detected through repeated sputum cultures and chest imaging, and if symptoms persist despite optimal treatment by bronchoscopy with bronchoalveolar lavage (BAL) to determine pathogens. Collaboration with a gastroenterologist otolaryngologist, occupational therapist, speech therapist, physiotherapist, dietician, and/or respiratory therapist, depending on local practice is recommended.

Management

Aspiration risks are addressed through careful (e.g., small bites, well-chewed, sipping water between bites of dense foods) or postural feeding, dilation for symptomatic esophageal strictures, GER treatment, and surgical repair of recurrent TEF or laryngeal cleft.

Recurrent Infections and Bronchitis

Complications

Patients with EA-TEF are prone to recurrent lower respiratory infections (pneumonias) and chronic bronchitis due to impaired airway clearance caused by tracheomalacia and recurrent aspiration.

Diagnosis

The diagnosis of bronchitis is based primarily on symptoms. A diagnosis of pneumonia or atelectasis should be confirmed with chest X-ray and/or ultrasound. When recurrent pneumonia is documented, a CT scan is recommended to identify bronchiectasis and exclude alternative diagnoses. Sputum or bronchoalveolar lavage (BAL) can help detect airway colonization by bacteria.

Treatment

Start age-appropriate airway clearance techniques to aid in the removal of mucus (in adolescents/adults: active cycle of breathing techniques) such as positive expiratory pressure devices like PEP masks or oscillatory positive pressure devices [2, 3]. Oral antibiotics – usually for 10–14 days – are recommended when infection is strongly suspected or confirmed, and azithromycin may be used preventatively due to its anti-inflammatory and antibiotic properties.

Atelectasis

Pathogenesis

Endobronchial secretions and/or aspiration may lead to atelectasis (areas of lung collapse). This is a particular problem with tracheobronchomalacia, which impairs mucociliary clearance. Atelectasis can also occur after surgery.

Diagnosis

In cases of new or worsening or recurrent respiratory symptoms, atelectasis should be suspected and imaging (lung ultrasound, chest X-ray, thoracic MRI and/or CT scan) should be performed to exclude or confirm the diagnosis.

Treatment

Conservative therapy may include airway clearance techniques, positive airway pressure devices and/or high-flow or CPAP support, use of hypertonic saline nebulization, mucolytic agents, and eventually systemic steroids, and systemic antibiotics. In persistent atelectasis, bronchoscopy – preferably within 4–6 weeks – may be required to reopen the lung and prevent permanent scarring and bronchiectasis. Follow-up imaging is required to confirm that atelectasis has resolved.

Bronchiectasis Monitoring

Definition

Bronchiectasis, a serious and potentially permanent complication, involves the floppiness and dilation of the airways caused by recurrent and/or persistent bacterial infection and aspiration, leading to chronic bacterial infection.

Diagnosis

When bronchiectasis is suspected, it should be confirmed with a CT scan or MRI of the chest. Depending on the clinical course, annual chest X-rays can be considered to monitor for bronchiectasis, likewise, regular lung function testing.

Management

Early detection, regular treatment with airway clearance techniques, nebulization of hypertonic saline, long-term use of azithromycin, and prompt use of antibiotics during exacerbations can help slow the progression of bronchiectasis.

Chronic Respiratory Symptoms (Wheezing, Coughing)

Diagnosis

Symptoms such as wheezing are, after TEF, often caused by airway collapse (TBM) rather than asthma. Misdiagnosis can lead to unnecessary treatments. Nevertheless, a diagnosis of asthma needs to be excluded.

Treatment

Bronchodilators are generally not recommended in case of TBM as they may worsen airway collapse, but they can be trialed for cases with airway hyperreactivity. In infants and preschool children, risk factors (e.g. family history of atopic diseases, eczema) and allergy testing may identify children with co-existent asthma. Pulmonary function testing, including bronchodilator response, could additionally be used in individuals 5–6 years and older.

Routine Health Maintenance

Regular follow-up

Patients should be monitored by a multidisciplinary team (which usually include a pediatric pulmonologist, gastroenterologist, surgeon and/or ear, nose and throat surgeon), with at least annual evaluations.

Vaccinations

In addition to routine vaccinations, annual influenza vaccination is strongly recommended. RSV vaccination should be performed in infants, in accordance with local guidelines.

Lifestyle and care

Parents and patients should be educated on airway clearance, recognizing early signs of infection, identifying changes in their respiratory status, monitoring for signs of aspiration, and the importance of long-term follow-up care.

Notice: This document is endorsed by the Esophageal ATresia global support groups (EAT), the European Reference Network for rare Inherited and Congenital Anomalies (ERNICA), the European Reference Network for rare respiratory diseases (ERN-LUNG), and the International Network of Esophageal Atresia (INoEA).

Conflict of Interest

The authors declare that they have no conflict of interest.

References

[1] Koumbourlis AC, Belessis Y, Cataletto M et al. Care recommendations for the respiratory complications of esophageal atresia-tracheoesophageal fistula. Pediatr Pulmonol 2020; 55: 2713–2729

[2] Chang AB, Fortescue R, Grimwood K et al. European Respiratory Society guidelines for the management of children and adolescents with bronchiectasis. Eur Respir J 2021; 58: 2002990

[3] Herrero-Cortina B, Lee AL, Oliveira A et al. European Respiratory Society statement on airway clearance techniques in adults with bronchiectasis. Eur Respir J 2023; 62: 2202053