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Ösophagusatresie: Versorgung zwischen Klinik und Praxis

Herausgegeben von
KEKS e. V.
Erika-Reinhardt-Stiftung
Prof. Dr. med. Franz-Josef Kretz

KEKS
Hilft bei angeborenen Erkrankungen der Luft- und Speiseröhre

ERIKA REINHARDT
STIFTUNG
Für Kinder mit Speiseröhrenfehlbildung

Thieme

Prof. Dr. med.
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Foreword

During my many years as an anesthesiologist and intensive care physician, I have had the privilege of caring for many children with esophageal atresia. This was done in close collaboration with pediatric surgeons and in consultation with the families themselves. These experiences have shown me very clearly how challenging the treatment of this rare malformation is and how important it is to provide reliable support beyond the surgery itself.

Early on, this led to close contact with KEKS e. V. Initially, this took the form of joint discussions with parents, and later also through support for the association’s work. Over the years, this has developed into a close relationship between KEKS and the Erika Reinhardt Foundation. Ms. Reinhardt herself, the founder of the foundation, was an important companion throughout her life. Today, we continue her work in her spirit by supporting the KEKS medical team, which provides expert advice to affected families.

Pediatricians in private practice play a key role in the long-term care of children with esophageal atresia. They are the first point of contact for families, help identify potential complications, and provide reassurance in everyday life. With this supplement, we would like to contribute to broadening knowledge about the specifics of this disease and provide reliable support to the doctors who care for these children.

I would like to thank everyone who contributed to the creation of this supplement, especially Julia Seifried and Annika Bürkle from KEKS and the publisher who made its publication possible; special thanks go to Dr. Judith Lucas and Melina Singer.

Prof. Dr. med. Franz-Josef Kretz
Chairman of the Erika Reinhardt Foundation

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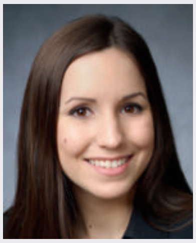

Anne Dimarakis

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Foreword to the Supplement

A diagnosis of esophageal atresia is challenging not only for the children affected, but also for their families – and for everyone involved in their medical care. After what is often an intensive hospital stay, a new phase begins when the child is discharged to their home environment: outpatient care takes over, and with it, the pediatrician assumes a key role.

Together with leading experts, KEKS has decided to make a practical contribution to pediatric practice with this supplement. The aim is to systematically highlight the often complex and heterogeneous care needs of children with esophageal atresia and to provide everyday recommendations for outpatient care.

A key motivation for the creation of this supplement is the increasing fragmentation of care. Today's hospital landscape is highly fragmented, and transitions between inpatient and outpatient treatment are sometimes inadequately structured. As a result, parents often have to take on a coordinating role, which places an emotional and organizational burden on them. Scientific literature – as well as our own survey of affected families – shows how important a continuous, cross-sectoral approach to care is.

KEKS and EAT are in close contact with a broad network of specialists, many of whom are active members of the scientific advisory boards of KEKS and EAT and their member organizations. These specialists work with their multidisciplinary teams in nationally and internationally certified clinics, together with patient organizations, to engage in a continuous quality dialogue aimed at improving care throughout the lifelong "patient journey."

The Esophageal Atresia Supplement brings together findings from clinical practice, current studies, and empirical knowledge from over 2,000 documented case histories in Germany, as well as many more from our EAT member organizations. This has allowed even rarer courses of disease and potential complications to be included in the analysis. Pediatricians working in primary care play a central role, especially in outpatient follow-up care. Families are often confronted with complications whose significance they find difficult to assess when they first occur. Parents repeatedly report that their observations have not been taken seriously in acute situations, such as in emergency rooms. A common reason for this is the lack of experience of the medical staff involved with the rare malformation. This makes it all the more important to have a reliable assessment by experienced pediatricians who can help parents weigh their concerns: whether by recommending that they return to the follow-up clinic, seek a specialized second opinion, or provide objective reassurance in cases of unremarkable or age-appropriate symptoms.

The pediatrician is therefore the most important point of contact for many families after discharge – medically, in terms of advice, but also emotionally. This role deserves professional support. We hope that this supplement will help to increase confidence in dealing with this rare disease and enable affected children to grow up in a stable environment with good support. It also aims to highlight the importance of lifelong follow-up care and a structured transition to adult medicine. KEKS and EAT are happy to support pediatricians as professionally networked, patient-oriented partners at both the national and international level.

Special thanks go to the Erika Reinhardt Foundation, whose support made this supplement possible.

Anne Dimarakis, President of KEKS
Stefanie Lorenz, Vice President of KEKS and pediatrician
Anke Widenmann, Board Member of KEKS and President of EAT

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Esophageal Atresia – The First Years

Ösophagusatresie – die ersten Jahre

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SUMMARY

The treatment of children born with an interrupted esophagus is demanding and complex. Because the esophagus and trachea develop from a shared embryonic structure, the trachea is also affected by malformation in most cases. This entails additional special features and requirements. To ensure safe care, all involved specialists must have extensive experience and practice and work together effectively. Survival and quality of life depend substantially on high-quality care in the first years of life. Clear interdisciplinary treatment pathways must be established and implemented regularly, which requires a certain annual case volume and, with it, routine.

ZUSAMMENFASSUNG

Die Behandlung von Kindern, die mit einer unterbrochenen Speiseröhre geboren werden, ist anspruchsvoll und aufwendig. Weil sie sich während der Embryonalentwicklung aus einer gemeinsamen Struktur entwickelt, ist in den meisten Fällen auch die Luftröhre von der Fehlbildung betroffen. Daraus ergeben sich zusätzliche Besonderheiten und Anforderungen. Damit diese Kinder sicher versorgt werden können, müssen alle beteiligten Fachleute viel Erfahrung und Übung haben und gut zusammenarbeiten. Die Überlebensrate und die Lebensqualität hängen ganz erheblich von einer guten Versorgung in den ersten Lebensjahren ab. Dazu müssen klare interdisziplinäre Behandlungsabläufe werden. Dies erfordert gewisse Fallzahlen, um Routine zu entwickeln.

Introduction

Because trachea and esophagus develop from a common embryonic structure, esophageal malformations are accompanied by tracheal involvement in most cases. This is readily apparent from the fact that most affected children have a tracheoesophageal fistula (TEF) which remains functionally relevant even after corrective surgery because tracheomalacia is common.

Caring for children with congenital interruption of the esophagus is challenging and requires high-level expertise and routine across all involved disciplines to ensure safety. These include, from prenatal diagnostics through obstetrics, neonatology, pediatric radiology, pediatric anesthesia, and pediatric surgery, as well as postoperative intensive care, nursing, and physiotherapy. Such a team can only operate reliably when structures and workflows are clearly defined for every step of care and the team is able to gain and maintain experience. Because this cannot be achieved with only a few cases per year, centralization is one of the most urgent requirements to ensure quality care for children with esophageal atresia.

A dataset from 1980–2015 including 6,466 children with esophageal atresia across 24 international registries from diverse countries showed an almost 90% one-month survival and an 83% five-year survival [1]. Care has undoubtedly improved over the last decades, but risks exist throughout the entire course of treatment and increase with associated anomalies and prematurity. An analysis of all German DRG discharge data from 2016–2022 showed a one-month survival of 92% [2], with providers of widely varying specialization represented. In a recent single-center series from Boston Children's Hospital, survival was only limited to 90% among neonates with a birth weight below 1,500 g or a significant cardiac malformation; all others survived [3]. It is reasonable to assume that further centralization in Germany could improve complication and survival rates [2].

Survival and quality of life depend to a very large extent on high-quality care during the first years of life. This chapter summarizes the most important aspects.

Prenatal Diagnostics

The typical prenatal ultrasound constellation for esophageal atresia is polyhydramnios combined with a small stomach. However, excessive amniotic fluid is present in only about 40–50% of cases [4]. This occurs because the fetus cannot swallow amniotic fluid, which consists of about 80% fetal urine. Because esophageal atresia can be associated with other malformations, these must be carefully evaluated when suspicion is raised. The most common association is VACTERL, an acronym standing for vertebral, anorectal, cardiac, tracheoesophageal, renal, and limb anomalies. Cardiac and renal status in particular can be important for prognosis.

If a syndrome is suspected, a maternal blood test, chorionic villus sampling, or amniocentesis can be helpful, as chromosomal anomalies such as Down syndrome (trisomy 21) and trisomies 13 and 18 are associated with esophageal atresia.

The prenatal diagnostic approach described above identifies only a portion of cases. In a meta-analysis including nearly 74,000 fetuses, the sensitivity of prenatal ultrasound was only 42% [5], which mirrors the frequency of associated polyhydramnios. Esophageal atresia is therefore among the most frequently missed congenital anomalies during pregnancy [6].

Prenatal fetal MRI is indicated only in complex, unclear cases. It has somewhat higher diagnostic accuracy than ultrasound but involves more effort. Numerous studies are currently underway to improve prenatal MRI diagnostics, and it remains to be seen whether better predictability with minimal burden for pregnant individuals and their fetuses will be possible in the future [7].

If esophageal atresia is suspected, delivery should occur in a center with sufficient experience in caring for these children. A cesarean section is not required solely due to suspected esophageal atresia. Families should be connected prenatally with a multidisciplinary team covering all aspects of care (obstetrics, neonatology, pediatric surgery, pediatric anesthesia).

Perinatal Care

Postnatal management should, when possible, promptly achieve adequate spontaneous breathing. A continuous low-pressure suction catheter should be placed to drain saliva and secretions from the mouth, and the upper body should be elevated to facilitate breathing. Lateral positioning is often helpful to allow secretions to drain. With invasive and even with noninvasive ventilation, progressive gastric insufflation can occur if a distal TEF is present, as is the case most of the time. In extreme cases, when the stomach is tense with air, ventilation may become impossible. The stomach must then be urgently decompressed to enable survival. In an emergency, this can be done with transabdominal gastric puncture; otherwise, operative gastric decompression is required. Apart from these specifics, standard neonatal care recommendations apply [8].

Postnatal/preoperative Diagnostics and Preparation

Confirming the diagnosis

Infants with esophageal atresia cannot swallow their saliva. Immediately after birth, this leads to coughing, visible oral secretions, and progressive respiratory distress. The first steps are suctioning secretions and attempting to advance a gastric tube through the nose or mouth. Failure to advance the tube into the stomach is often the first reliable indication of esophageal atresia and, despite the diagnostic caveats below, is accepted by current guidelines as diagnostic confirmation [9]. A chest and abdominal radiograph ("babygram") should be obtained to document the coiled tube; it must include the entire thorax and abdomen. Contrast instillation through the tube should be avoided due to aspiration risk; at most, a small amount of air (10 ml) may be injected during the image to better outline the proximal pouch. If the babygram shows air in the gastric bubble and the infant does have esophageal atresia, this proves a distal TEF. The babygram also shows cardiac configuration as a potential sign of congenital heart disease and the entire spine to assess vertebral anomalies. In addition, a postnatal echocardiogram and abdominal ultrasound, particularly to evaluate renal anomalies, are mandatory.

This diagnostic approach occasionally yields false positives, in which a tube cannot be placed for other reasons. Rare causes include esophageal foreign body impaction [10], pharyngeal pouching after traumatic delivery [11], pharyngeal or esophageal perforations due to traumatic endotracheal intubation attempts [12, 13], and misplaced gastric tubes [14]. False negatives also occur, where a tube appears appropriately positioned on radiograph despite malposition. Reported scenarios include perforation of the proximal esophageal pouch with passage through the mediastinum into the abdomen [15], or passage of the tube through the larynx or a proximal TEF into the trachea, then through a distal TEF into the stomach [16–18].

To avoid recognizing a misdiagnosis only intraoperatively, preoperative bronchoscopy should be required, although in practice it is often not performed [19]. A European consensus statement also recommends bronchoscopy to identify additional pathologies [9], locate a TEF, and detect cases with a second, proximal TEF [20]. The tracheal opening into a distal TEF toward the lower esophagus typically appears “fish-mouth” like (fistula tracks downward), whereas a proximal TEF appears like a volcanic crater (fistula tracks upward). There are no recommendations to routinely perform esophagoscopy beforehand. This likely reflects that most esophageal atresias have a distal TEF and demonstrating this by bronchoscopy confirms the diagnosis. However, if no TEF is found, we consider esophagoscopy essential [13], since there may be esophageal continuity or esophageal atresia without TEF (Vogt type 2). As a practical tip, esophagoscopy can be efficiently performed using a bronchoscope while applying continuous positive airway pressure (PEEP). Endoscopy can be performed immediately before surgery to avoid multiple anesthetics.

Preoperative preparation

Repairing an esophageal atresia is rarely an emergency case. Rapid action is required only when positive-pressure ventilation distends the stomach and impedes ventilation. Otherwise, careful preoperative preparation is advisable to maximize safety and success. Preparation should minimize the risk of complications and create optimal operative conditions. If there is air in the stomach and, therefore, a high likelihood of a distal TEF (recognizing the diagnostic pitfalls above), spontaneous breathing is preferable to minimize gastric overdistension.

A comprehensive evaluation to detect or exclude associated anomalies is essential, particularly echocardiography to assess cardiac function, identify anomalies, and exclude a right-sided aortic arch. Correction of acid–base balance and electrolytes is important to ensure metabolic stability. Ideally, the operation should occur within the first few days after birth, as the risk of pulmonary deterioration increases markedly over time. An irradiated blood unit should be prepared in advance in case transfusion is required.

Anesthesia for the Primary Operation

Informed consent and risk discussion

Parents or legal guardians should be thoroughly informed about the planned procedures and potential risks. Clinicians should address anesthesia-related concerns empathetically. When anesthesia is delivered by experienced pediatric anesthesiologists, it is demonstrably safer and less harmful for infants [23, 24]. As noted, tracheoscopy/bronchoscopy and, if necessary, gastroscopy are required. General anesthesia with endotracheal intubation is used. Invasive access may be necessary to optimize monitoring and management, especially in very small newborns or those with cardiac defects.

Blood transfusions may be required; indications and risks should be discussed. Postoperatively, care and monitoring will take place in the intensive care unit. The consent discussion must also address the risks of serious complications (including death or disability; see Introduction). Before corrective surgery, parents should be given ample time for decision-making between the consultation and the surgery date, whenever, as is usually the case, the procedure is elective.

Anesthesia preparation

Beyond guideline-based standard preparations for neonatal and infant surgery, additional, esophagus-specific measures are required.

Masks, laryngeal masks, and endotracheal tubes in appropriate sizes should be at hand. An endoscopic system capable of tracheoscopy must be available; if no TEF is identified, esophagoscopy must also be feasible. In addition, a large “stoch” or “pusher” tube (placed orally to splint and expose the proximal esophageal pouch) and a thin nasally placed gastric tube (to advance across the newly created anastomosis) should be prepared. Selection and sizes should be coordinated closely with the surgeons.

Standard monitoring should include pulse oximetry, ECG, noninvasive and invasive blood pressure (NIBP, IBP), temperature probe, near-infrared spectroscopy (NIRS), and neuromuscular monitoring (NMT). A urinary catheter, central venous access, and an arterial line should be placed. The arterial line enables frequent blood gas analyses and glucose checks.

Endoscopy should be performed carefully, examining the posterior tracheal wall for additional fistulas. This also helps exclude misdiagnoses (e.g., a birth-trauma associated pharyngeal hematoma). Ideally, findings are shown to the surgeon beforehand, and the subsequent plan (tube position, possible TEF cannulation) is agreed upon.

Conduct of anesthesia

Induction proceeds as usual, adapted to the clinical situation. After endoscopic assessment, particular care must be taken to avoid intubation into a TEF. Correct tube placement can be verified bronchoscopically. Induction should occur with the surgeons present so they can perform an emergent gastrotomy if severe gastric distension occurs. A pharyngeal “stoch” tube is placed and kept accessible to facilitate intraoperative exposure of the proximal esophageal pouch.

The child is positioned in left lateral decubitus for open surgery; for thoracoscopy, slightly more prone. Ventilation should be particularly gentle until the fistula is closed. Target peripheral oxygen saturation is 97–98 % (87–92 % in preterm infants). Maintain normovolemia. Catecholamines should be immediately available and, ideally, started at very low doses from the outset to ensure prompt effect if needed.

After the anastomosis, a nasally placed gastric tube may be inserted and secured, as agreed with the pediatric surgeons.

Intraoperative challenges

Owing to numerous and complex intraoperative challenges, the presence of an experienced pediatric anesthesiologist is essential.

Thick bronchial secretions may obstruct the tube and airways, impairing oxygenation. Surgical manipulation can compress the lung or trachea, causing inadequate ventilation with compromised oxygenation and CO2 clearance. Compression of the atria or venae cavae by the surgeons may impair venous return, reduce cardiac output, and further worsen oxygenation. Differentiating among these complications requires an experienced anesthesiologist capable of identifying the causes and implementing targeted measures to enable full recovery.

Some compromise must be negotiated with the surgical team; ideal values may not be achievable during the "critical" phases. Given cerebral metabolic suppression under anesthesia, this is tolerable. NIRS offers additional information on cerebral oxygenation and should be part of standard monitoring.

Operative Management

The goals of surgical repair in esophageal atresia are to ligate any tracheoesophageal fistulas and restore esophageal continuity. Surgery can be performed open, via thoracotomy, or thoracoscopically. Thoracoscopy uses three or four small ports only a few millimeters in diameter. Advantages include smaller scars, less postoperative pain, faster recovery, and potentially a lower long-term risk of musculoskeletal issues such as scoliosis.

The approach depends on the type of esophageal atresia. Most esophageal atresias with a distal fistula (Gross type C or Vogt type 3b) can be repaired in a single stage.

After bronchoscopy to assess for fistulas, the child is positioned–typically left lateral for open surgery and somewhat ventrally for thoracoscopy. The lower esophagus is identified and dissected cranially to the usually present TEF. The fistula is closed at the trachea as close as possible, either with sutures or a clip. The lower esophagus is then divided as near as possible to the trachea to preserve maximal esophageal length and avoid a deep tracheal pouch at the former fistula site.

Next, the proximal esophageal pouch is located, mobilized as needed, and drawn downward. It is opened at the tip and anastomosed to the lower esophagus. This anastomosis is technically demanding, especially thoracoscopically. The connection must be sufficiently wide and watertight. Some surgeons prefer to place a transanastomotic tube from the nose into the stomach to allow early postoperative gastric feeding.

If the gap between esophageal ends is too large for primary anastomosis, this is termed long-gap esophageal atresia. Most of these patients have atresia without a fistula (Gross type A or Vogt type 2) or with a proximal TEF (Gross type B or Vogt type 3a). A proximal TEF is identifiable on preoperative bronchoscopy but is overall rare. In esophageal atresia without fistula, a Replogle tube is placed to continuously suction saliva from the proximal pouch over the following weeks. This is a double-lumen sump catheter with three holes at the tip that continuously "sips" saliva from the pouch. A gastrostomy is also placed to feed directly into the stomach, either open or laparoscopic. This is made more difficult by the often very small stomach in these infants. The combination of the sump catheter and gastrostomy allows the infant to be fed and to grow. Several strategies exist for managing long-gap atresia. The esophagus grows faster than the child overall, so waiting weeks to months may allow relative approximation of the ends for later anastomosis. Approximation can be aided by applying traction to the ends with internal sutures (internal Foker) or externalized sutures through the skin with daily incremental tension (external Foker) until the ends overlap. Alternatively, esophageal replacement can be considered, for example gastric pull-up or interposition of small or large intestine. Managing long-gap esophageal atresia is complex, prolonged, and commonly accompanied by complications to varying degrees.

Postoperative Care

General postoperative principles for neonates apply. If there were no intraoperative issues, especially after thoracoscopic repair, the infant may be extubated and transferred breathing spontaneously to recovery. Prolonged ventilation and muscle relaxation should be considered only in cases of significant anastomotic tension. If a transanastomotic tube was placed intraoperatively, it can be used for feeding after 24 hours. If the infant manages on spontaneous breathing without CPAP support, careful oral milk feeding should be added. If an anastomotic leak is suspected, a contrast study should be performed. A clinical checklist should be available covering all measures required before first discharge (for example, ultrasound of the urinary tract and spine, and caregiver CPR training).

Follow-up: Prophylaxis, Evaluations, Support

Structured follow-up is essential after esophageal atresia repair to ensure long-term success and prevent complications. About three weeks after primary anastomosis, a contrast swallow can be helpful for an initial assessment. Gastroesophageal reflux is common in infants and even more frequent in children with esophageal atresia. It can usually be managed well with proton pump inhibitors (PPIs). All children should routinely receive a PPI during the first year of life to prevent acid exposure and the resulting esophagitis. Ideally, after a trial discontinuation, biopsies should be obtained to determine whether continued therapy is necessary. If reflux causes persistent problems such as esophagitis, inadequate intake, or respiratory symptoms, antireflux surgery (fundoplication) may be indicated in selected cases, preferably after the first year of life.

Regular checkups should assess feeding and eating behavior, infection frequency, overall development, and physical growth (especially spinal development), as well as include endoscopic evaluations. Early identification of anastomotic stricture allows relatively simple treatment and avoids progression that could necessitate reoperation. Further details on musculoskeletal, airway and pulmonary care, and nutrition are covered in other articles in this special issue.

At routine visits, a careful history should address feeding, swallowing, breathing, presence of stridor or other respiratory noises, and signs of possible reflux. Physical examination should include auscultation of the chest and inspection of surgical scars. Watch for signs of scapula alata, thoracic asymmetry, or scoliosis. Weight and length are important indicators of growth. Values should be plotted on growth charts to best detect deviations from typical trajectories. Condition-specific growth and weight charts for esophageal atresia are already available for Germany [22].

A child who feeds and eats well, breathes normally, gains weight appropriately, and grows along expected centiles does not need additional diagnostics in the first year of life. Introduction of complementary foods, purees, and later solids can proceed normally, gradually increasing bite size. If problems are suspected, a chest radiograph can be helpful, or a contrast esophagram. If there are signs of anastomotic narrowing–such as impaired swallowing and a stricture visible on contrast study–the anastomosis should be gently dilated under anesthesia.

Laryngomalacia and tracheomalacia are also common in children with esophageal atresia. These involve a soft larynx or trachea that partially collapses during breathing, potentially obstructing airflow. Symptoms usually improve over time. In rare cases of significant respiratory compromise, intervention may be necessary, such as aortopexy or posterior tracheopexy. Rarely, tracheostomy may be required.

About six weeks after surgery, an initial endoscopic evaluation can be useful. Ideally, this starts with a functional assessment of the airways using a flexible scope transnasally under sedation to detect vocal cord paralysis. This should be followed by a full airway examination under general anesthesia with muscle relaxation to reliably assess tracheomalacia and visualize the former fistula site. Subsequently, the esophagus should be examined to assess anastomotic caliber and the competence of the gastroesophageal junction in both antegrade view and retroflexion. If indicated, balloon dilation of the anastomosis can be performed during the same session. The maximum achievable diameter here is defined by the diameter of the distal esophagus; typically, there is a caliber discrepancy between the proximal and distal esophagus. This is because fetuses swallow prenatally into the proximal pouch, stretching it, whereas the distal stump is not exposed to this.

If repeated dilations are necessary, intralesional depot corticosteroid injection can be considered. Leaving a transnasal balloon catheter in place to allow twice-daily home dilations by parents can also be considered. Successful therapy requires extensive experience through regular evaluations and interventions. Lifelong periodic endoscopic surveillance is recommended [21].

In the first years of life, affected children often endure numerous hospitalizations and diagnostic procedures. The care team should make these phases as comfortable as possible. Stress during this sensitive period can have long-term negative effects on development (see other article in this supplement). It is sensible to coordinate evaluations efficiently to avoid multiple anesthetics or repeated blood draws. Supportive pain-reducing measures, such as topical anesthetic patches (e.g., EMLA), can help make needle procedures more comfortable [25].

For successful follow-up, actively involving parents in the care process is crucial. This can include shared decision-making and inviting their presence during evaluations when appropriate. Parents can then participate actively, feel better informed, and make sound decisions together with the care team.

Conflict of Interest

The authors declare that they have no conflict of interest.

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

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

Author

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Keywords

oesophageal atresia/esophageal atresia, respiratory complications, tracheomalacia, tracheoesophageal fistula, atelectasis, structured long-erm care

Schlüsselwörter

Ösophagusatresie, respiratorische Komplikationen, Tracheomalazie, tracheoösophageale Fistel, Atelektasen, strukturierte Langzeitbetreuung

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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

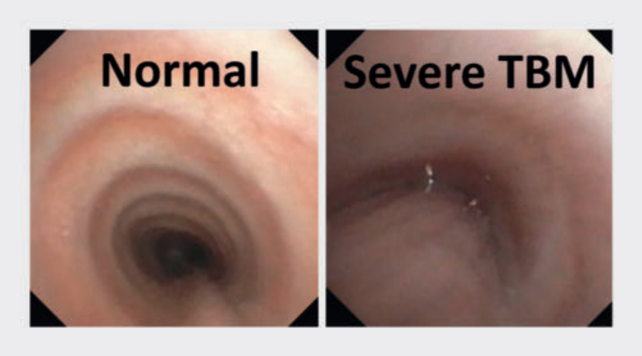
Dieser Artikel fasst aktuelle Veröffentlichungen, Leitlinien, Stellungnahmen sowie Konsensempfehlungen der Fachgruppen von INoEA, der Europäischen Referenznetzwerke ERN-Lung und ERNICA sowie der European Respiratory Society (ERS) zur Ösophagusatresie (ÖA) – Tracheoösophageale Fistel (TÖF) zusammen.

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**).

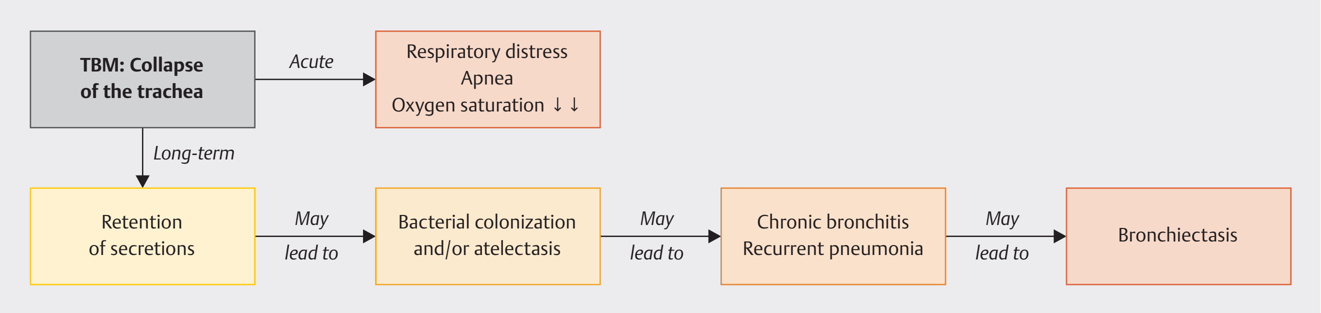
The symptoms of TBM vary according to the age of the patient. They tend to be most severe in the neonatal period and generally may improve with time (▶ **Table 1**).

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

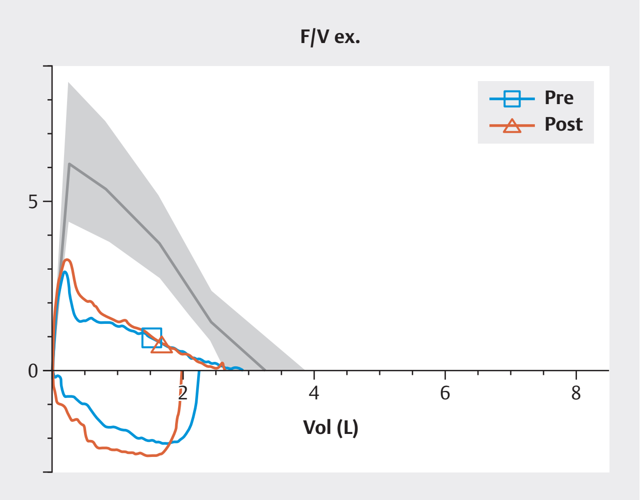
Interventions aim to reduce symptoms due to TBM and, when it is severe, reduce TBM severity by surgery. In neonates, the primary goal is to prevent respiratory distress and low oxygen levels. Treat-



▶ **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.

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Finally Home – Successful Transition from Inpatient to Outpatient Care

Endlich daheim – gelungener Übergang vom stationären in den ambulanten Sektor

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Keywords
oesophageal atresia/esophageal atresia, rare disease, patient support, discharge management, holistic follow-up

Schlüsselwörter
Ösophagusatresie, seltene Erkrankungen, Entlassmanagement, Patientenunterstützung, ganzheitliche Nachsorge

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ABSTRACT

Children with rare malformations and their families often experience gaps in care at the interface between inpatient and outpatient sectors. They require reliable and coordinated care–extending beyond the hospital. The article highlights areas of overlap with pediatric and adolescent medicine in private practice. It also illustrates possible support systems and a best practice example. For the best possible outcome, it's essential to ensure multidisciplinary care across all sectors and at all interfaces.

ZUSAMMENFASSUNG

Kinder mit seltenen Fehlbildungen und ihre Familien erleben häufig Versorgungsbrüche an der Schnittstelle zwischen dem stationären und ambulanten Sektor. Sie benötigen eine verlässliche und koordinierte Versorgung – über die Klinik hinaus. Der Artikel zeigt auf, in welchen Bereichen Überschneidungen mit der niedergelassenen Kinder- und Jugendmedizin existieren. Zudem werden mögliche Unterstützungssysteme und ein Best-Practice-Beispiel illustriert. Für einen bestmöglichen Verlauf ist es essenziell, dass eine multidisziplinäre Versorgung in allen Sektoren und an allen Schnittstellen gewährleistet ist.

Background

In particular, people with chronic illnesses are affected by the fragmentation of the German healthcare system. The transition from inpatient to outpatient therapy can lead to a discontinuous course of treatment, with therapy interruptions and loss of information [1].

The literature shows that parents of children with rare diseases experience the fragmentation of the healthcare system particularly strongly and often must overcome it themselves [2]. In addition, parents of children who require inpatient care as infants report insufficient support during and after discharge from the hospital [3, 4]. Parents of children with esophageal atresia (EA) have higher anxiety levels, increased stress symptoms, and a lower Quality of Life (QoL) index compared to the general population [6–8]. These values correlated, among other things, with the subjectively experienced lack of support during the transition from the inpatient to the outpatient sector [7, 8].

When a child is discharged from the surgical clinic to their home, inpatient care by a professional team ends and outpatient treatment begins. Especially in the first period after discharge, a lack of disease-specific expertise on the part of outpatient service providers poses a problem in adequately meeting the support needs of parents. This is a specific challenge of rare diseases and cannot be expected from outpatient pediatric care. Rather, it requires close support for parents and pediatricians from the expert clinic to be able to respond appropriately to specific questions.

In 2023, the German patient organization for EA (KEKS) interviewed 25 parents (or parent couples) about their experiences during and after the first hospital discharge. The results show that parents often feel inadequately prepared for discharge and, especially in the outpatient sector, have to organize and manage their child's care themselves [9]. It has been shown that, in contrast to the inpatient sector, parents have more control options in the outpatient sector and are more likely to make decisions together with therapeutic and pediatric staff [9]. On the other hand, therapists and pediatricians also rely more on the parents' experiential knowledge. The respondents found this responsibility to be a burden and, not least, there are often disagreements between parents and healthcare professionals on specific issues [9]. The literature shows that parents want to be involved in the care process on an individual basis, especially in the form of shared decision-making [10, 11]. However, the people concerned must receive clear information about the medical problem and the available options [12, 13]. At the same time, parents do not want to feel the main burden of responsibility for decisions [10]. In this regard, the high emotional stress can have a negative effect on parents' motivation to participate in the processes [10].

To make matters worse, parents often cannot explain or even know exactly what treatment their child needs at the beginning and therefore cannot pass it on to providers of follow-up and supportive care [14]. In general, people with rare diseases are dissatisfied with the coordination of care in the outpatient sector. This could be explained by the fact that coordinating tasks and better interprofessional networking are currently not remunerated [15].

In the KEKS survey, most parents expressed the wish to have a contact person who would accompany them from the beginning and, together with them, keep an eye on which symptoms require further clarification and which therapies and examinations are necessary [9].

Concepts of Support

To facilitate the transition between inpatient and outpatient care areas and to secure continued treatment, there are transitional structures, such as discharge and case management [16, 17].

Discharge management

Patients with complex service needs are particularly vulnerable to experiencing care disruptions during the transition from the inpatient to the outpatient sector [1]. To ensure continuity of care, professional discharge planning is of particular importance [1]. According to § 39, paragraph 1a of the SGB V, hospitals are obliged to effectively support patients during the transition to the next form of care [16]. Since 2017, hospitals have had to comply with the requirements of the framework agreement on discharge management [16]. This agreement sets out the goals and minimum requirements for discharge management [18].

The goal of discharge management is to ensure continuity of treatment after a hospital stay. For this purpose, the individual needs must be assessed and addressed in a structured way by means of a discharge plan [18]. All measures necessary to meet the identified need for services should be initiated by the hospital as early as possible [18]. The hospital must contact the continuing service providers as early as possible to ensure a secure transition. This also includes the transfer of information to the referring physician or family doctor. The continuing care structures must be informed, among other things, about the upcoming discharge date [18]. However, the scope of discharge management ends as soon as these tasks have been fulfilled and the patient has transitioned to the outpatient sector. Discharge management is no longer responsible for problems that arise afterward.

Case Management

Another form of support for people with complex illnesses is Case Management (CM). This is an approach from the social and health care sector that involves the "needs-based management of a case situation to deal with a personal problem" [17].

The goal is the case-related integration and coordination of care processes, especially at interfaces [19]. In addition to the objective assessment of needs, the consideration of individual and subjective needs plays an important role. These must be organized effectively and efficiently within the regional care structure [17].

A characteristic feature of CM is the assumption of multiple roles. People working in CM take on a gatekeeping position by arranging targeted services for the case. They also accompany the people through the care process and the service system. The CM represents the interests of the person to be cared for, initiates available services, and advocates for the appropriate quality and fulfillment of needs [20].

The prerequisite for a functioning CM is Care Management. This is understood as the organization and networking of service providers (e.g., health insurance companies), service providers (e.g., nursing services), and service recipients (e.g., the person to be cared for) in the local care landscape. The goal is to be able to offer a continuous, cross-sectoral care tailored to the individual case [20]. Examples of this are care support centers according to § 92c SGB XI, disease management programs according to § 137 f SGB V, and in the pediatric field the follow-up care model "Bunter Kreis e. V." which organizes medical, nursing, and social services for seriously and chronically ill children and their families [20, 21].

A form of CM used specifically for vulnerable groups in the medical field are patient navigators [22]. These are currently being tested in model projects. The German Society for Case and Care Management (DGCC) defines patient navigators as "care and case managers for people in complex life and care situations with one or more medical indications" [19]. However, there is currently no uniform definition of their areas of responsibility and the required skills [22].

A project at the Augsburg Central Hospital as early as 2008 showed that the structured support of parents with premature babies, starting in the clinic and continuing into the home, can lead to a reduced stress level for the mother [23].

In an international context, there are already structures in which specialized nurses employed in clinics accompany families with children with congenital malformations from their first stay to follow-up care and transition (Family Liaison Nurses). They are contact persons for the families and children, but also for outpatient specialists, daycare centers, and schools. In Germany, a pilot project has established a family liaison nurse model since November 2024 for families with children with congenital malformations. In addition, there are other liaison nurse models in Germany, such as the psychosomatic liaison nurse, who accompanies and advises oncology patients, their relatives, and on-site nursing staff [24]. An evaluation showed that nursing and medical staff perceived the liaison nurse as a relief [24].

Optimizing the Transition to Outpatient Care: Important Aspects for Successful Follow-up Care

As described in the previous chapter, there are many ways to support families in the transition of their child from inpatient to outpatient care and to make this transition as smooth and effective as possible.

The pediatrician plays a central role in this. In their function as the first point of contact after discharge from the clinic, they are often the most important authority for questions about the child's further recovery, therapy, and general care.

The following describes key elements that should help outpatient service providers to identify individual support needs and to sustainably improve the care of patients and their families:

Structured follow-up care: coordinated care and binding guidelines

Structured follow-up care is essential for continuous, high-quality care after an inpatient stay. This includes binding guidelines and standards that both families and professionals can follow–for example, through the KEKS Health Folder or specific international follow-up care recommendations. Information about the KEKS Health Folder can be found in the article “KEKS Health Folder and KEKS Portal” in this supplement [25].

The goal is holistic and long-term, cross-sectoral care. This includes, in particular, the coordinated cooperation of various specialist disciplines and support systems. The inclusion of relevant information and findings at the time of the outpatient doctor's appointment is just as important as targeted communication with the discharging clinic to avoid treatment gaps.

Complete information transfer: relevant medical documents available

All relevant medical documents – especially discharge reports, surgical reports, and findings – should be available to the treating pediatrician completely and in a timely manner. This is the only way to ensure sound medical follow-up care.

Designated contact persons: clear communication channels for all involved parties

A clear contact person should be named in the discharging clinic, whom both the pediatrician and the family can contact with questions. This person does not necessarily have to be a doctor, but could be, for example, a liaison nurse or another qualified specialist.

In addition, parents are informed before discharge about how to behave in an emergency and what contact options or presentation options are available outside of regular hours. If parents have not received this information, they should be encouraged to actively request it. This can help to avoid unnecessary or emergency inpatient readmissions.

Training for families: strengthening their competence

Ideally, families receive training on emergency measures (e.g., resuscitation, dealing with bolus events, first aid for aspiration) during the initial hospital stay. The pediatrician should actively ask during follow-up care whether this training has taken place and, if necessary, suggest refresher courses.

In socio-pedagogical contexts such as settling into daycare centers, there is often a greater need for information and support – both for families and for the care facilities. Close cooperation and education about the child's illness can help to reduce fears and ensure adequate integration.

Networking with self-help and support services

Families should be actively informed about existing support services offered by self-help organizations – such as KEKS If this has not already happened in the inpatient setting, the recommendation to contact them should be explicitly made by the outpatient specialist.

Patient organizations offer valuable information, advice, and emotional relief to affected families.

Psychosocial support and parents' mental health

Pediatricians should be aware of the emotional and psychological stress on parents and use suitable, possibly standardized, tools to assess their support needs. If necessary, a referral to psychosocial or psychotherapeutic institutions is advisable. Additional information is provided in the article “EA as a family disease” in this supplement [26].

Involvement of the Social Pediatric Center (SPZ)

The Social Pediatric Centre, as a multi-professional, outpatient facility, offers a valuable addition to the care of chronically ill children. Pediatricians should find out whether there is already contact with the regional SPZ and, if not, suggest a connection.

An appointment at the SPZ can represent a valuable extension of care and promote holistic therapy, especially for children with complex or rare diseases (e.g., esophageal atresia).

Complications of esophageal atresia

Esophageal atresia is a rare, congenital malformation with a high need for medical care. Due to the complexity of the disease and the individual anatomical and functional differences in the affected children, the clinical picture is extremely heterogeneous. Therefore, differentiated, continuous observation in both the outpatient and inpatient sectors is essential.

Even minor changes in the child's general condition can be an indication of serious complications and should therefore not be trivialized but always clarified by a doctor. Early diagnosis and intervention can prevent or mitigate serious courses. Well-informed and educated parents can serve as an important and competent resource in this regard.

The most common and relevant long-term complications include:

- Recurrent fistulas: A recurrence of the fistula between the trachea and esophagus can manifest as increased coughing, choking, failure to thrive, or recurrent respiratory infections.
- Stenoses (narrowing of the anastomosis): These often lead to swallowing difficulties, gagging, or food refusal and may require repeated dilation treatment.
- Gastroesophageal Reflux (GERD): Severe reflux is common in children with EA and can lead to inflammation, pain when eating, or food refusal. If left untreated, GERD can lead to reflux esophagitis, aspirations, or Barrett's esophagus in the long term.
- Dumping Syndrome: A rarer but relevant complication after gastric surgery. Symptoms can include rapid bowel movements after eating, restlessness, paleness, hypoglycemia, or circulatory problems. Early detection and dietary adjustments play a central role here.

Best practice example – how the problem at the interfaces can be solved

High-quality and well-coordinated outpatient care is essential for the long-term course of the disease.

In order to further professionalize the care of children, the pilot project of family liaison nursing was established at the Marien Hospital in Witten in cooperation with KEKS in November 2024.

The concept is based on the liaison nursing model, which is based on “Primary Nursing” implemented by Marie Manthey as early as the 1960s [27]. Family liaison nursing at Marien Hospital Witten represents a low-threshold offer in a counselling and supportive function for people affected by rare, congenital malformations and their families. Since the liaison nurse closely accompanies the patients and their parents during their hospital stay and is part of a multidisciplinary team, they are seen as an active contact person for the parents regarding the course of treatment. After the hospital stay, consistent follow-up care for those affected is essential for the further course of treatment. To ensure continuity of treatment, especially in follow-up care, the coordination of the various specialist areas, but also support systems, is important. This task is taken over by the family liaison nurse and thus serves as an interface coordinator for resident pediatricians.

The focus is therefore on holistic, long-term care for patients beyond the clinical setting. The pilot project is designed so that it can be expanded to other expert clinics. Supported by KEKS, the concept is to be established in other clinics in the next few years. The detailed description of the concept can be found here.

Opportunities for Support for Pediatric Practices

As already mentioned in the introduction to this text, care in an expert center is of central importance. This is based on the so-called indirect evidence, as has already been shown for comparable diseases in adult medicine. Since 2021, KEKS has been certifying clinics in Germany for the care of people with esophageal atresia and their relatives. The certification is based on the ERNICA (European Reference Network for rare inherited and congenital gastrointestinal malformations and diseases) consensus recommendations [28, 29] as well as other national and international recommendations [5, 30]. Accordingly, people with rare congenital malformations should be given access to clinics with special expertise [28, 29]. The principle of a second opinion is suitable for this, as it can increase patient safety during the course of the disease. KEKS supports the search for suitable contact persons. A list of the clinics certified by KEKS so far can be viewed on the KEKS website. The primary responsibility for adequate follow-up care lies with the institution where the surgical care took place or takes place. Accordingly, it must be demanded that the corresponding professional expertise and the resources necessary for all aspects of the disease are available. This also includes the provision of structured follow-up care plans and concrete recommendations for action for frequently occurring problems in the everyday lives of the affected families. Good care for these children requires close cooperation between all participating specialist disciplines. Clinics that want to guarantee or ensure high-quality follow-up care should be open and accessible for questions or consultations at all times. A significant part of this accessibility can be achieved through a family liaison nurse.

Summary

Children with rare malformations and their families need reliable, coordinated care–beyond the clinic. Family liaison nursing shows how individual support and professional coordination can provide security and close care gaps as a supplement to existing structures such as discharge and case management.

The role of the pediatrician as the first point of contact for parents' concerns about their child's development after discharge from the clinic cannot be overestimated for the child's further recovery, therapy, and general care. KEKS is available to both pediatricians and families as a further source of support.

Conflict of Interest

The authors declare that they have no conflict of interest.

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From the Swallowing Reflex to Family Mealtimes

Recognizing Challenges and Providing Eating and Drinking Support to Children Born with Esophageal Atresia

Von Schluckreflex bis Familientisch

Herausforderungen erkennen und sicher begleiten bei Kindern mit Ösophagusatresie

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ABSTRACT

Children with EA cannot drink immediately after birth and require surgical treatment. Delayed initiation of oral feeding and (recurring) structural problems can lead to acceptance issues and challenges in eating development. In addition to evaluation of the esophagus, it is essential to examine the larynx and lower respiratory tract. Speech therapy/pediatric swallowing therapy and, if necessary, an imaging swallowing examination (e.g., FEES) are important to determine the nature of the swallowing problem, especially if aspiration is suspected. A child- and family centered, cue-based approach is central to all eating/drinking interventions. Children should learn to eat independently. For children who experience "stickies", individual trial and error–e.g., drinking after eating, using movement–helps as long as there is no acute danger. With good oral motor development, therapy in the narrower sense is often not necessary. Regarding swallowing and eating development, it is important to thoroughly evaluate eating in relation to developmental milestones and to provide continuous and needs-based counselling to families – ideally from birth.

ZUSAMMENFASSUNG

Kinder mit Ösophagusatresie (ÖA) können nach der Geburt nicht trinken und benötigen eine chirurgische Behandlung. Der verspätete Einstieg in die orale Nahrungsaufnahme sowie (wieder) auftretende strukturelle Probleme können zu Akzeptanzproblemen und Herausforderungen in der Essentwicklung führen. Neben der Diagnostik des Ösophagus müssen unbedingt auch der Larynx und die unteren Atemwege untersucht werden. Bei Schluckproblemen sind Logopädie/Kinderschlucktherapie und ggf. eine bildgebende Schluckuntersuchung (z. B. FEES) wichtig, besonders bei Verdacht auf Aspiration. Eine kindzentrierte, signalorientierte Haltung ist bei allen Interventionen zentral. Kinder sollen selbstbestimmt essen lernen. Bei Steckenbleibern hilft individuelles Ausprobieren – z. B. Nachtrinken, Bewegung – solange keine akute Gefahr besteht. Bei einer guten oralmotorischen Entwicklung ist oft keine Therapie im engeren Sinne notwendig. Wichtig ist eine gründliche Evaluation der Entwicklungsschritte und Meilensteine der Essentwicklung und eine kontinuierliche und bedarfsorientierte (logopädische) Beratung der Familien – im Idealfall von Geburt an.

Swallowing Problems in Children Born with Esophageal Atresia

Until surgical repair, children born with esophageal atresia cannot drink, i.e. breast- or bottle-feeding is not possible. In addition, many children have an abnormal connection, a fistula, between the trachea and esophagus, which carries an additional risk of aspiration, i.e. secretions, saliva, milk and stomach contents passing into the lower airway. The initiation of oral feeding is dependent upon successful surgical repair. If repair is delayed for weeks or even months after birth, so too is the initiation of breast- or bottle-feeding. A delayed introduction to drinking can lead to difficulties in acceptance. The lack of experience makes the processes of sucking and swallowing more susceptible to coordination difficulties or sensory processing difficulties which can lead to coughing or milk refusal. In addition, abnormal esophageal peristalsis and the development of narrowing/strictures can lead to slow esophageal bolus transit or bolus obstruction, often called “stickies”. The entire eating/drinking development is therefore more vulnerable than in children without EA and the caregivers are often anxious about their child’s swallowing.

What Needs to be Checked?

As part of the initial diagnosis of esophageal atresia, the larynx and trachea should be assessed using rigid endoscopy to rule out upper and lower airway abnormalities, such as tracheomalacia and laryngeal cleft. A laryngeal cleft is a congenital malformation of the posterior larynx which creates a connection between the larynx/trachea and esophagus. Undetected, it leads to chronic aspiration during swallowing. Unfortunately, even today, routine endoscopy by ENT colleagues is not standard practice in all operating centres, resulting in missed diagnosis.

As such, referral to an appropriate specialist centre should be initiated for children with frequent coughing or choking episodes associated with eating and drinking. If swallowing problems (re) occur, the first point of contact is often the pediatric surgeon providing treatment and follow-up care. Consider esophageal and oro-pharyngeal causes of eating/drinking difficulties and their underlying aetiology (e.g. GOR, dysmotility, stricture, recurrent TOF, swallow-respiratory incoordination due to tracheomalacia laryngeal cleft, VCP, oro-motor difficulties, sensory based feeding difficulties etc.). If oro-pharyngeal swallowing difficulties are suspected, instrumental swallow assessment should be undertaken, such as FEES (fibreoptic evaluation of swallowing) or VFSS (videofluoroscopy swallow study). If aspiration events are seen or an increased risk of aspiration are suspected, further assessment is necessary to determine the underlying cause of the aspiration. A rigid endoscopy under anesthesia (ideally performed jointly by pulmonology, ENT and pediatric surgery) with the question of refistula or laryngeal cleft is recommended.

How Can Children with EA Learn to Enjoy Eating and Swallow Safely?

A cue-based, child-centered approach has proven successful for the development of eating and drinking, from sucking to chewing. This means that the infant, and later the toddler’s, feeding cues are observed and taken seriously. Such cues include hunger and satiation cues and signs of distress or discomfort with feeds/mealtimes adjusted to promote positive experiences for the child and parent/caregiver. With older children, the focus is not only on basic autonomy for eating, but also on good self-awareness regarding possible problems. The more autonomy children experience while eating and drinking, the quicker they will develop appropriate coping and adaptation strategies and enjoy mealtimes.

Occasional gagging or coughing is perfectly normal when children are learning to eat independently. Food preferences are also not unusual. However, EA children have altered esophageal peristalsis, which can lead to slower movement of food into the stomach or bolus obstruction. These are the so-called “stickies”. In addition to close observation to determine whether they need specific treatment, these children also need to develop self-management skills. Unless there is an acute danger, “trial and error” applies here. This means that the children and families are encouraged to try to find out what promotes the passage of food. The most commonly used adaptations are frequent water drinking while eating, chewing food well and eating more slowly and avoiding very challenging foods (such as some types of meat and bread). The rule here is: what helps is good.

For some children, esophagus-related problems may co-occur with dysphagia arising from the oral or pharyngeal phases of swallowing. These children have difficulty preparing food in the mouth or closing the airway securely during swallowing. This manifests itself in various symptoms. Such symptoms require clinical examination, often by a speech and language therapist, and instrumental assessment of swallowing if pharyngeal difficulties are suspected (see “What needs to be checked”).

Professional Support for Eating Development

Experience shows that children with EA and their families benefit from professional support with swallowing and eating development right from the start. This involves the assessment of dysphagia and initial counselling. The form of dysphagia or the affected phase and the clinical signs also inform the need for further swallowing therapy. If oral-motor development is good, therapy in the narrower sense is often not necessary. However, support to achieve developmental eating/drinking milestones through continuous and needs-oriented counselling of the families ensures mealtimes are safe, enjoyable and nutritionally sufficient for growth. Ideally, the children are referred to speech therapy during their initial hospital stay and the parents are supported before discharge. This reduces anxiety and ensures that the need for therapy is reviewed individually. Counselling for all families should cover areas such as dietary adjustments, expanding the food repertoire, safe consistencies and meal planning or playful promotion of oral motor skills in everyday life. Where required, targeted swallowing therapy can address specific elements of dysfunction, depending on the child's age.

The expertise of swallowing therapists in the outpatient sector in relation to rare diseases is very heterogeneous. Of course, this also applies to esophageal atresia. This can lead to therapists feeling insecure. KEKS NEST, the network for eating and swallowing therapists, has therefore been in place since 2023 to support healthcare providers in the field of pediatric swallowing therapy. In this joint project between KEKS e. V. and the Department of Pediatric Surgery of the Dr. von Hauner Children's Hospital at LMU Hospital Munich, therapists meet online four times a year to strengthen the treatment of children with esophageal atresia (close to home) by sharing knowledge and experience as well as discussing cases. The project is managed by Sandra Bergmann (LMU Klinikum) and Julia Seifried (KEKS e. V.) in terms of content and organization. Interested parties can register informally or find out more at nest@keks.org

Summary

Children with EA cannot drink after birth and require surgical treatment. Often there is an abnormal connection between the trachea and esophagus (fistula), which increases the risk of aspiration. Delayed initiation of drinking can lead to acceptance problems and an increased susceptibility to swallowing disorders. Later, abnormal peristalsis or narrowing at the anastomotic site may result in food "sticking". It is important that the larynx and trachea are also examined carefully to rule out laryngeal cleft, as this can lead to chronic aspiration. Instrumental swallowing examinations such as FEES (Flexible Endoscopic Evaluation of Swallowing) are useful to detect and treat aspiration.

A child-centred, cue-based approach is key. Children should learn to eat in a self-determined way. Individual trial and error – e.g. re-swallowing, exercise – can help with stuck food as long as there is no acute bolus obstruction. Difficulties at the oral and pharyngeal swallowing phases (e.g. lack of airway protection) should also be considered with targeted assessment and therapy as necessary.

Professional support from specialists, such as speech therapists, is essential to support eating development, reduce anxiety and develop individual strategies. The aim is to strengthen children's autonomy and support their eating and swallowing skills in the best possible way.

INFO

Worth knowing from the family counseling/ parents' perspective

- Tube weaning takes time, often a similar amount of time as the children needed the tube. This is highly individual.
- It requires trust from the children – they usually have a reason for not swallowing/eating – a second opinion can be useful.
- Eating should be enjoyable – eating should primarily be a positive social interaction, not just calorie intake. Enjoyment of eating is important for overall development
- When a child has a respiratory infection, a regression in quantity and consistency of foods eaten is possible
- After dilatations, there are occasional setbacks and stagnation in (learning to) eat for a few days, especially with regard to consistency.

Conflict of Interest

The authors declare that they have no conflict of interest.

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Eating, Growing, Living – Challenges in Children and Adolescents with Esophageal Atresia

Essen, Wachsen, Leben – Herausforderungen bei Kindern und Jugendlichen mit einer Ösophagusatresie

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ABSTRACT

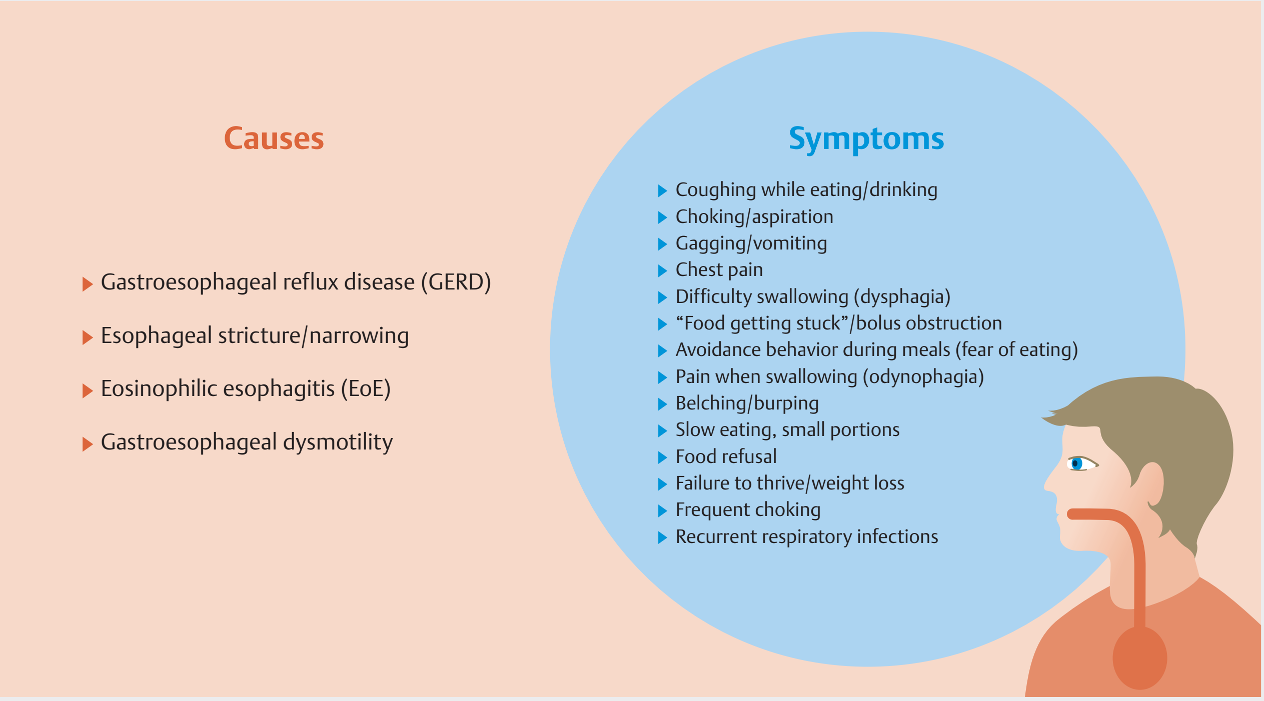
Children who have undergone surgical correction of esophageal atresia (EA) often experience significant feeding difficulties, impairing growth and overall development. Despite significant improvements in surgical techniques, long-term nutritional problems remain, which significantly affect growth, health and quality of life. Factors include anatomical alterations, motility disorders, gastroesophageal reflux disease (GERD), eosinophilic esophagitis (EoE) and sensory-motor deficits. This article provides an overview of underlying causes, diagnostic approaches, and therapeutic strategies for timely identification and management of nutritional problems. Multidisciplinary care and regular monitoring are crucial to support optimal growth and development in these children.

ZUSAMMENFASSUNG

Kinder mit operierter Ösophagusatresie (ÖA) zeigen häufig erhebliche Schwierigkeiten bei der Nahrungsaufnahme, was wiederum das Gedeihen beeinträchtigen kann. Trotz deutlicher Verbesserungen der chirurgischen Techniken bleiben langfristige Ernährungsprobleme bestehen, die Wachstum, Gesundheit und Lebensqualität maßgeblich beeinflussen. Gründe für die z. T. komplexen Ernährungsschwierigkeiten sind anatomische Veränderungen, Motilitätsstörungen, gastroösophageale Refluxerkrankung (GÖRK), eosinophile Ösophagitis (EoE) sowie sensorische und motorische Einschränkungen. Der Artikel gibt einen Überblick über Ursachen, diagnostische Ansätze und Therapieoptionen, um Ernährungsprobleme frühzeitig zu erkennen und zu behandeln. Multidisziplinäre Betreuung und regelmäßiges Monitoring sind essenziell, um das Wachstum und die Entwicklung dieser Kinder bestmöglich zu fördern.

Introduction

Esophageal atresia (EA) is a congenital malformation in which the continuity of the esophagus is interrupted and requires surgical correction. Although surgical techniques have improved considerably, long-term challenges remain regarding nutrition, growth, and quality of life. These issues affect not only the child's physical development but also the psychosocial well-being of the affected children and their families.



▶ **Fig. 1** Overlapping symptoms of gastrointestinal complications in pediatric patients after surgical repair of esophageal atresia (according to data from [12]).

Causes of Feeding Difficulties

The reasons for feeding difficulties in children with repaired esophageal atresia (EA) are multifactorial and include anatomical, functional, sensory, and psychological aspects. Below are the main causes, with detailed explanations and prevalence data based on current PubMed-listed studies [1] (▶ **Fig. 1**):

Anatomical alterations

Esophageal stenosis and anastomotic strictures

One of the most common postoperative complications is the development of scar-related narrowing (strictures) at the anastomotic site. Studies report stricture rates of up to 57 % within the first year after surgery [2]. Such anastomotic strictures can severely hinder oral food intake, impair bolus transport, and cause pain, vomiting, or outright refusal to eat [3].

Esophageal motility disturbances

Impaired peristalsis

After surgery, many children present with esophageal dysmotility, resulting in swallowing difficulties. Food is not transported efficiently from the mouth to the stomach, which complicates eating and can lead to frequent choking. Up to 80 % of children with repaired EA show motility disorders, while only about 20 % exhibit normal peristaltic function [4].

Delayed clearance

Due to impaired esophageal motor function, food remains in the esophagus for longer periods, which can cause gagging, pain, and food refusal [5].

Gastroesophageal reflux disease (GERD)

Reflux

Between 22 % and 58 % of patients with EA suffer from clinically significant gastroesophageal reflux. This may cause esophagitis, which is often manifested by pain during or after meals [5]. The pathophysiology of reflux in this patient population is complex and often results from a combination of factors: anatomical changes such as a gastric pull-up or a flattened angle of His after surgical repair compromise the natural anti-reflux barrier at the gastroesophageal junction. In addition, esophageal dysmotility, present in many patients, not only delays bolus passage but also reduces clearance of refluxed gastric acid. This prolongs mucosal contact with acidic and irritant contents. In some children, pronounced eosinophilic infiltration of the esophageal mucosa (eosinophilic esophagitis, see below) also plays a role [5, 6].

Oropharyngeal dysphagia

Swallowing disorders at the oropharyngeal level

This form of dysphagia occurs in 38–85 % of cases and often includes aspiration problems caused by poor coordination between the mouth, pharynx and esophagus [4].

Eosinophilic esophagitis (EoE)

Tissue remodeling

In the esophageal mucosa of patients with EA, a marked increase in eosinophilic granulocytes is frequently found. Inflammatory mediators such as cytokines and growth factors promote tissue remodeling, leading to fibrosis of the esophageal wall, luminal narrowing, and further impairment of motility. Structural changes in the esophageal nervous system and impaired epithelial barrier function are also discussed as underlying pathophysiological mechanisms [7]. The development of EoE may be further facilitated by factors such as impaired esophageal motility, prolonged bolus transit, frequent and early-life exposure to antibiotics, and even proton pump inhibitor (PPI) therapy [8].

Bolus obstruction

As a result of inflammation-induced remodeling and preexisting motility disorders, acute bolus impaction may occur after eating–when a piece of food gets lodged in the esophagus (colloquially referred to as a “food sticking”). This is a medical emergency and usually requires prompt endoscopic removal [9].

Sensory and motor impairments

Oral aversion and sensory integration issues

Negative early oral experiences, such as surgeries and prolonged hospital stays, frequently lead to sensory integration disorders and strong aversion to oral feeding. Many children develop oral aversion due to negative associations (e.g., pain, frequent medical interventions, tube feeding), manifesting as fear, refusal, or even panic during meals [10].

Lack of oral motor skills

Particularly in toddlers, impaired oral motor patterns hinder the acquisition of normal eating skills and result in persistent difficulties with chewing and swallowing solid foods. In children with long-gap EA, oral feeding is often introduced later than normal, further delaying the development of age-appropriate eating habits [5].

Comorbidities and associated conditions

Neurological developmental delays and cardiac anomalies

Conditions such as neurological impairment or congenital heart defects increase the risk of prolonged dependence on enteral feeding tubes [11].

Vocal cord motion disorders

These issues such as vocal cord paresis or recurrent laryngeal nerve palsy are not uncommon after surgical correction of esophageal atresia and increase the risk of aspiration and cause significant swallowing difficulties [4, 12].

Psychosocial factors

Stress and anxiety in the family context

Feeding difficulties negatively affect the child’s physical and social quality of life. The need for tube feeding, feeding pumps, or high-calorie specialized formulas is associated with a significantly reduced quality of life [13].

Chronic stress due to repeated hospitalizations, invasive medical procedures, and constant fear of choking or aspiration can impact the psychological stability of the family and greatly exacerbate existing feeding difficulties [5, 14].

In practical terms, mealtimes often become stressful for both the child and the parents, leading to a vicious cycle of anxiety, refusal, and further stress, which worsens the feeding situation [10].

Overall prevalence of feeding difficulties

A large proportion of children with repaired EA show persistent abnormalities in eating behavior, often continuing into adolescence and adulthood. Up to 75 % exhibit problematic eating patterns such as slow eating, selective eating, or complete food refusal [4, 10].

Severe feeding difficulties, such as long-term dependence on feeding tubes, are particularly common in children with additional risk factors (e.g., neurological deficits or long-gap EA) [11].

Clinical Presentation

The clinical symptoms in children with repaired esophageal atresia (EA) vary greatly between individuals. Typical features include prolonged mealtimes, gagging, coughing during meals, recurrent vomiting, food refusal, and significantly impaired growth. These symptoms can occur not only in early childhood but may persist into adolescence and adulthood, often changing in nature over time.

Red flags for feeding impairment

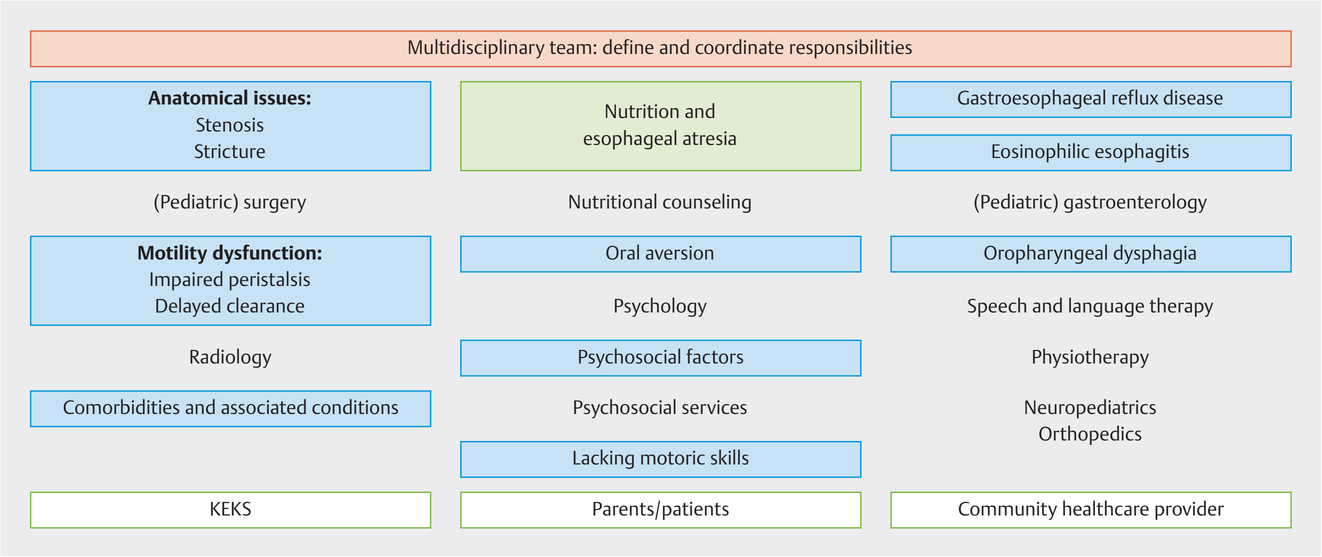
Although there is no single standardized definition of feeding problems, several so-called red flags have been described to help identify feeding disorders. These include: very long mealtimes, coughing, gagging, or vomiting during meals, pronounced aversion to food (oral aversion) or refusal of food with marked selectivity, as well as parental reports of difficult or stressful mealtimes

Puntis et al. identified the most common problems in children with EA as especially prolonged eating times, coughing or gagging during meals, and food refusal. These difficulties were notably more frequent in the first two years of life compared to healthy control children [15].

Quality of life

Children with EA who require special feeding arrangements (e.g., feeding tube, pump system, small portion sizes, prolonged mealtime duration) generally show significantly reduced quality of life–particularly in the physical, social, and school domains. In older children, a lack of independence when eating further impacts quality of life.

In a study on the quality of life of affected children and adolescents, the following aspects were described as particularly burdensome for both patients and parents: feeding through a gastrostomy tube (PEG), the use of a feeding pump, supplemental nutrition to increase caloric intake, and the need for smaller meal portions. Among the older age group (8–17 years), prolonged



▸ **Fig. 2** Multidisciplinary care.

mealtimes (over 30 min) and the necessity of parental assistance during feeding were also reported as significant stressors [13].

Diagnostic approaches

Diagnosis begins with a thorough medical history to capture individual symptoms and risk factors. Key investigations include:

- Impedance–pH monitoring to objectively assess the extent of gastroesophageal reflux, potentially even during ongoing acid-suppressive therapy to optimize dosing.
- Contrast swallow studies (barium swallow) to visualize structural narrowings.
- Endoscopy for detailed inspection and, if necessary, treatment of strictures.
- Videofluoroscopic swallow studies to evaluate oropharyngeal dysphagia and possible aspiration events.

Therapeutic strategies

A multidisciplinary approach is essential. This includes: regular nutritional counseling to ensure adequate caloric intake, speech and language therapy to improve oral motor skills and swallowing, pharmacologic measures for managing gastroesophageal reflux disease (GERD) – typically proton pump inhibitors – and for treating eosinophilic esophagitis (EoE) if present, surgical interventions such as fundoplication and regular dilatation of strictures when indicated.

Gastroesophageal reflux disease (GERD)

GERD often leads to severe complications, including esophagitis and peptic strictures, and in some cases, the development of Barrett's esophagus requiring treatment. Early and systematic management is strongly recommended, especially in the first year of life. Long-term medical therapy and surgical procedures such as fundoplication may be necessary.

Eosinophilic esophagitis (EoE)

Treatment of EoE may involve elimination diets, proton pump inhibitors, topically active steroids, or–if these measures fail–dupilumab, an interleukin-4/13 inhibitor [9]. Treatment success must be evaluated endoscopically with biopsies taken from three esophageal levels 8–12 weeks after initiation, as there are no alternative markers to confirm improvement or deterioration.

Dependence on feeding tubes

Many children require tube feeding initially or for prolonged periods. Risk factors for long-term dependence include tight anastomoses, neurodevelopmental impairments, vocal cord motion disorders, and associated cardiac anomalies. These factors should be identified early and addressed proactively.

Long-term growth and development

A considerable number of children with EA show delayed or impaired weight gain [16]. Continuous multidisciplinary interventions are crucial for achieving positive developmental outcomes and maintaining good quality of life. Regular follow-up visits and treatment adjustments are needed to detect and manage long-term complications early.

Multidisciplinary care and parental counseling

An interdisciplinary team consisting of pediatric surgeons, gastroenterologists, nutritionists, speech therapists, psychologists, and social workers is indispensable. Regular counseling and practical family support significantly improve therapy adherence and outcomes (▸ **Fig. 2**).

Practical tips for introducing food and drink

A gradual and gentle introduction of feeding is of great importance. Initially, it is advisable to feed the child small amounts in an upright position to detect coughing or reflux early. Thickened liquids should be introduced first, followed by a slow and controlled transition to smooth purees.

Soft-textured foods are the next stage and should be introduced gradually to promote acceptance and avoid overtaxing the swallowing function. Melt-away foods–items that dissolve easily in the mouth and can be swallowed safely and comfortably–can help bridge the gap toward the introduction of solid foods.

The introduction of textured foods should proceed step-by-step, always monitoring for signs of aversion or discomfort. It is important not to pressure the child, but instead to offer gentle encouragement and positive reinforcement.

Establishing a regular, calm, and consistent mealtime routine can facilitate the introduction of new foods. If difficulties arise, early speech and language therapy support is beneficial to specifically promote oral motor skills and swallowing ability.

Conclusion

Feeding difficulties in children with repaired esophageal atresia are a common and complex challenge. Integrating evidence-based recommendations (e.g., ESPGHAN–NASPGHAN guidelines [5]) with practical, experience-based strategies enables comprehensive and effective management.

An early, multidisciplinary treatment approach–combining continuous support, regular monitoring, and targeted, practical feeding measures–plays a decisive role in sustainably improving the quality of life and developmental outcomes of affected children.

The causes of feeding difficulties in this patient group are multifaceted and require an interdisciplinary perspective. Comprehensive care involves close collaboration between pediatric surgery, gastroenterology, speech therapy, nutrition counseling, and psychology to ensure adequate long-term oral feeding and to support healthy growth.

Equally important is the consistent integration of parental experiences, resources, and individual burdens into the care plan. Parents are not merely companions in the process but active partners, whose involvement is essential to fostering the child's long-term health, development, and well-being.

Conflict of Interest

The authors declare that they have no conflict of interest.

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Pulmonary Problems in Children with Esophageal Atresia

Pneumologische Probleme bei Kindern mit operierter Ösophagusatresie

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ABSTRACT

Most children with esophageal atresia (EA) have pulmonary problems. The causes include associated malformations of the central airways, tracheomalacia, consequences of intubation and mechanical ventilation, as well as prematurity. This leads to an increased frequency of pulmonary infections with bacterial involvement. Long-term consequences are bronchiectasis and other structural damage to the lungs, resulting in a restrictive ventilatory defect, reduced physical resilience, and increased mortality. To prevent these late pulmonary complications, early and consistent multidisciplinary follow-up care and treatment for children with EA are essential, with continuation into adulthood. Only a few large centers can maintain such structures and acquire the necessary expertise.

ZUSAMMENFASSUNG

Die meisten Kinder mit Ösophagusatresie (ÖA) haben pulmonale Probleme. Ursachen sind begleitende Fehlbildungen der zentralen Atemwege, Tracheomalazie, Intubations- und Beatmungsfolgen sowie Frühgeburtlichkeit. In der Folge kommt es zu gehäuften pulmonalen Infektionen mit bakterieller Beteiligung. Spätfolgen sind Bronchiektasen und andere strukturelle Schädigungen der Lungen, mit restriktiver Ventilationsstörung, verminderter Belastbarkeit und erhöhter Mortalität. Um pulmonale Spätfolgen zu verhindern, ist eine frühzeitige und konsequente multidisziplinäre Nachsorge und Behandlung bei Kindern mit ÖA zu fordern sowie die Fortsetzung auch im Erwachsenenalter. Nur wenige große Zentren können solche Strukturen vorhalten und die notwendige Expertise erwerben.

Introduction

Esophageal atresia (EA) is primarily a condition requiring pediatric surgical treatment. However, it often becomes clear very early that the airways and lungs play a critical role in a child’s subsequent development. To date, there is a lack of prospective controlled studies on airway problems in children with EA. This is certainly due to the absence of structured care and, in particular, follow-up. As long as 111 hospitals in Germany perform surgery on children with EA–29 of them only once every five years–this situation is unlikely to change. In some countries (France, Australia, Scandinavia), there are initiatives toward centralization, which has led to more available data. In the literature, reports on pulmonary sequelae after esophageal atresia are found primarily in the form of case reports or small case series. Nevertheless, an initiative by INoEA (International Network of Esophageal Atresia) successfully established a working group (Respiratory Complications Working Group), which has published recommendations for the detection and treatment of these problems [1].

Prevalence of Pulmonary Problems after EA surgery

Data from the French registry, including 1,287 children with EA, show a 12-month mortality rate of 7 %. Nearly one-third of the children were hospitalized within the first year of life due to respiratory problems. Significant risk factors include: an initial hospital stay of more than 90 days, tube feeding, inhalation therapy at the time of first discharge, recurrent fistula, aortopexy, reflux, antireflux surgery, dilatations, and insufficient weight gain [2].

Most studies do not specifically assess respiratory symptoms. In a meta-analysis, case series that did assess respiratory symptoms reported chronic respiratory problems in up to one-third of children. However, these figures are difficult to compare and interpret. The main problems are chronic cough, recurrent respiratory infections, and chronic lung disease [3].

In a study conducted approximately ten years ago, 80 of 110 surviving adults who had undergone EA surgery were invited to participate, and 28 responded. Almost 80 % had pulmonary problems, predominantly restrictive ventilatory disorders. In most cases, the lung disease had not been diagnosed before the study [4].

One difficulty in assessing pulmonary problems in adulthood is that predominantly the healthier patients survived [5]. This leads to data bias, especially considering that the main cause of mortality in children with EA beyond the first year of life has pulmonary causes. In some countries, structured care has led to a significant improvement in this regard.

Symptoms

Dry or barking cough is a typical symptom in children with EA. This is due to tracheomalacia, which also changes the resonance characteristics of the airways, in addition to altering respiratory mechanics. Even without excessive mucus production, children with EA very frequently have chronic cough that is difficult to suppress. This occurs independently of infections and is significantly more frequent than in their peers [6].

Chronic cough with secretions is a strong indicator of a relevant functional or anatomical problem in the central airways and/or peripheral lungs. The underlying cause must be identified to prevent further damage to the airways.

Recurrent (viral) respiratory infections are normal in early childhood. However, when such infections are regularly prolonged or complicated, this is also indicative of a relevant pulmonary problem.

Hilar “central” bilateral bronchopneumonia occurs in toddlers with a frequency of around 4 % per year, usually in the context of viral infections, and are relatively harmless. In children with EA, pneumonias occur very frequently in the first years of life, almost always as complications during or after viral respiratory infections, and typically with localized infiltrates (middle lobe, basal unilateral or bilateral, among others). About half of EA children have had at least one pneumonia requiring hospitalization, and many have had three or more pneumonias in the first years of life [6–8].

Typical Complex Problems in Children with EA

Aspiration

Children aspirate with EA very frequently, usually recurrently and often in small amounts. Causes include anatomical or functional abnormalities in the larynx, esophagus, and trachea. Typical anatomical examples are unrecognized laryngeal clefts, fistulas (recurrent or unrecognized), and vocal cord paralysis.

Functional swallowing disorders without anatomical stenosis are very common and often remain undiagnosed for a long time. Symptoms can be misinterpreted by both pediatric surgeons and pulmonologists. For instance, liquids can become trapped due to interrupted or chaotic peristalsis of the esophagus. A typical case history: drinking carbonated soft drinks from a glass leads to coughing (= aspiration due to a large gulp and foaming), whereas drinking through a straw is easier (= smaller sips that pass more easily and foam less). Children with EA usually eat more slowly because peristalsis is not functioning normally or is uncoordinated and “chaotic” in the distal segment.

The accumulation of secretions (saliva), reflux, or regurgitation after food impaction can also lead to aspiration. Protective laryngospasm against aspiration does not always work well in EA and, in itself, can also be an additional problem [9]. Asthma-like symptoms can be mimicked by microaspiration, leading many EA patients to receive inappropriate and ineffective asthma therapy. This is particularly detrimental when an inhaled steroid with high local absorption (e.g., beclomethasone) is used, as this may cause fungal infection and further impair laryngeal function.

Growth and physical performance

Although physical growth and performance are not primarily pulmonary symptoms, there are interrelationships. Severe lung impairment reduces physical growth. Conversely, for example, poor growth due to insufficient nutritional intake can limit physical performance, and lung growth is also affected.

EA patients tend to be below the age-matched average in height percentiles. They exhibit lower physical activity than their peers. Across all age groups–and most pronounced in adolescents–the sports index (minutes/week) is significantly lower in children with EA than in healthy comparison groups [10].

Orthopedic symptoms

Adolescents and young adults after EA have a several-fold increased risk of scoliosis: 12 % have scoliosis > 20°, and an additional 22 % have mild scoliosis. In the cohort studies, most patients did not have additional vertebral malformations. Many exhibited reduced physical endurance. The risk of scoliosis increases with age [11].

Since these are usually relatively rigid scolioses that are not comparable to classic adolescent idiopathic scoliosis (AIS), it can be assumed that there is additional impairment of cardiopulmonary capacity.

Typical Clinical Diagnoses or Complications

Tracheomalacia

Most children with EA have tracheomalacia, regardless of what type of fistula(s) exists or existed. With an expiratory residual lumen of more than 50%, there are usually no symptoms apart from barking cough. When the expiratory residual lumen is below 10%, children often have significant episodes of dyspnea, especially during infections. These cyanotic spells can be highly dramatic. This is further exacerbated by a food bolus–when food gets stuck–and particularly if there are also cardiac malformations, especially vascular anomalies or a right-to-left shunt.

Tracheomalacia not only causes the typical barking cough but also recurrent lower respiratory tract infections. Auscultatory findings with “wheezing” often lead to inappropriate treatment with beta-mimetics. In cases of severe tracheomalacia, approximately 50% of children show aspiration during contrast examination [7].

In pronounced tracheomalacia, symptoms often persist, so many surviving adults continue to have typical respiratory problems [8].

In addition, vocal cord disorders are very common (up to 30%), often as a result of (long-term) ventilation and/or surgical interventions, or due to reflux.

Impaired airway clearance

Tracheal narrowing causes not only mechanical problems. Due to chronic inflammation and mucosal abnormalities in the fistula region, there is a loss of cilia in the central airways [1]. This results in mucus retention with secondary infection and airway inflammation causing bacterial bronchitis. The long-term consequences are bronchiectasis and destruction of lung structure. These problems are particularly significant in young children but persist into adulthood for many patients [8]. Causes include the malformation itself, acquired surgical injury, inadequate infection management in childhood, comorbidities, recurrent lower respiratory tract infections, atopy, and smoking.

Bronchiectasis

Extremely few newborns are born with bronchiectasis. Bronchiectasis is a late symptom and, in most cases, a consequence of respiratory infections. It is most common in cystic fibrosis and ciliary dysfunction disorders and is not entirely avoidable in these conditions even with structured therapy.

Recurrent pneumonias in early childhood are an important risk factor for the development of non-CF bronchiectasis, regardless of the underlying disease or malformation. Consequences include progressive loss of lung function, frequent pulmonary exacerbations, reduced quality of life, and death in early adulthood [8, 12]. Immotile cilia (primary or secondary, as in EA) promote the development of bronchiectasis.

In children with EA (6 months to 12 years), bronchiectasis is found in 30% [13], often combined with atelectasis, e.g., in the right middle lobe. Only a few larger CT studies exist in EA children. In one series, in children with a mean age of 7.4 years, bronchiectasis was found in 31%. Parallel bronchoscopy showed no signs of this [14]. In 14%, tracheal diverticula were also present–mostly in children referred from smaller centers.

Bronchiectasis is so common that there is an explicit recommendation to exclude it via CT in adults after EA with chronic cough [9].

Associated malformations

Many EA patients have other associated malformations, most frequently cardiac defects or anomalies of the central vessels. EA can occur in various syndromes and chromosomal abnormalities such as trisomy 21.

The most well-known is the VACTERL association, much more rarely the CHARGE association.

Diagnostics

Children with EA are entitled to qualified airway diagnostics. Already during the initial hospital stay, it should be determined whether there is an associated laryngeal malformation, most often a dorsal cleft. This is not uncommon and is very often missed initially [9]. Furthermore, the extent of tracheomalacia should be known. It is also important to detect atypical bronchial branching patterns (e.g., of the right upper lobe bronchus), additional narrowing caused by aberrant major vessels/cardiac malformations, and other anomalies before pulmonary complications develop. Flexible bronchoscopy under sedation with spontaneous breathing, e.g., during induction of anesthesia for surgery, is suitable for this purpose. If a fistula is suspected, combined bronchoscopy and esophagoscopy should be performed.

Even beyond the neonatal period, there are indications for–if necessary invasive–evaluation, e.g., chronic (wet) cough or repeated pneumonia episodes.

Bronchoalveolar lavage (BAL) may be useful to rule out chronic aspiration.

A low-dose chest CT or, alternatively, lung MRI (as per cystic fibrosis standards) performed during a symptom-free interval after pneumonia is suitable for detecting long-term damage. A normal chest X-ray does not rule out bronchiectasis. Annual routine chest X-rays are not recommended [1], and certainly not annual CT scans.

Another method not yet generally available is Real-Time MRI. This can be used to document the interaction of the larynx, swallowing, and breathing.

Blood count and CRP are not suitable parameters for ruling out bacterial superinfection in the context of pulmonary symptoms [1]. This may delay necessary antibiotic therapy, with corresponding consequences.

Pulmonary function testing is of limited value in diagnosing bronchomalacia (misinterpretation as asthmatic obstruction). The reduced lung function in children with EA has long been known and was previously, at least in part, justifiably attributed to prematurity, intensive care, and ventilation [15].

However, the problem persists even with improved neonatal care. In a follow-up study of adolescents with EA, 63% had abnormal lung function [16]. The length of the EA gap correlates with the degree of restriction.

Registry data from Sweden show that lung function declines with age: mean forced expiratory volume in one second (FEV1) is 82% at age 8 and 76% at age 15. Many patients exhibit “obstruction,” restriction, or a combination of both [17]. In adults, vital capacity is significantly reduced at 74% compared to 104% in control groups. It should be noted that these data include only patients with fewer complications, since they survived. Despite these limitations, regular monitoring of pulmonary function parameters is important.

Therapy

In very severely ill infants, non-invasive ventilation with PEEP may be useful in the initial phase, and a small number of children benefit from a tracheostomy, possibly with a long cannula. These are usually EA children with multiple severe associated malformations.

In cases of very severe tracheomalacia with frequent cyanotic episodes and recurrent aspirations, aortopexy (with intraoperative bronchoscopic monitoring) can be helpful [1]. A characteristic clinical sign is frequent hyperextension, which children instinctively adopt to “stretch” the trachea. Highly successful and prophylactically effective is the technique of posterior tracheopexy, which can be performed in the context of minimally invasive primary surgery. This technique is not yet widely available–another argument for centralized care in a small number of highly specialized centers.

Recommendations for anti-reflux therapy are based only on “expert opinion” [1]; good clinical studies are lacking. Bronchodilators such as salbutamol should generally be avoided, as they may worsen bronchial collapse [1]. Mucolytics have a very limited effect. Inhaled steroids are only useful in exceptional cases (see above) and can lead to infectious complications.

It is crucial to detect and treat bacterial or bacterial/viral mixed infections at an early stage. In principle, every pneumonia must be prevented to avoid the development of bronchiectasis. A low threshold for prescribing antibiotics is explicitly recommended [1]. Early and consistent treatment of infections with beginning superinfection takes priority over acute diagnostics, as neither blood count, CRP, nor current chest X-rays are reliable decision-making tools [14]. The bacterial spectrum must be taken into account: staphylococci are the second most common pathogen in non-CF bronchiectasis. Therefore, an antibiotic should be chosen that reliably covers the triad of Haemophilus, S. pneumoniae, and S. aureus. Resistance development is very rare. Resistant pathogens (particularly MRSA) are practically only seen after long-term inpatient treatment of the EA patient or close contact to people harboring MRSA.

To enable timely therapy, a practical and responsible approach is to prescribe parents two antibiotic courses of 7–10 days each, to be used as needed. Parents are usually well able to decide when therapy is appropriate. Emergency antibiotic prescriptions outside regular office hours, upon parental request with reference to EA, work neither with general practitioners or pediatrician nor in emergency departments, and when given, often involve less suitable substances. Long-term antibiotic prophylaxis–e.g., during the infection season–should be reserved for exceptional cases.

Adults with EA differ only slightly from the general population with respect to the pulmonary microbiome [5], though the available numbers are small and include only relatively uncomplicated EA cases.

Based on a meta-analysis of available studies on pulmonary outcomes, an algorithm for management has been proposed, which is roughly consistent with the approach outlined here [18].

Airway clearance techniques, including respiratory physiotherapy and, if necessary, autogenic drainage, are helpful in many children [7, 9]. This can be learned in physiotherapy practices with cystic fibrosis experience.

Summary

The available data clearly demonstrate the urgent need to break the vicious cycle of early pulmonary complications and progressive, permanent destruction of lung structure [14]. It is crucial to identify children with pulmonary problems in EA in a timely manner, something that does not occur in many surgical centers. Approximately half of children with EA experience such pulmonary problems. Therefore, all children must undergo regular evaluation. In some cases, problems arise only during adolescence.

The risk factors for a problematic course are known. Children who receive primary care in smaller hospitals with low numbers of EA surgery have a higher risk of late pulmonary complications [14]. All available data support the need for multidisciplinary follow-up care, involving at least (pediatric) surgery, (pediatric) pulmonology, (pediatric) gastroenterology, orthopedics, and other specialties as required [1, 7]. Particularly in the first three years of life, short-interval monitoring every 3–6 months is essential [14].

This leads to the following recommendations:

- Pulmonology follow-up care from the neonatal stage should be actively recommended by the treating pediatricians.
- Regular follow-up by pediatric pulmonologists with specific expertise in bronchopulmonary malformations and in recognizing and treating bronchiectasis. If necessary, parents should be advised to seek a second opinion in a certified center.
- A transition concept aimed at continued pulmonary care in adulthood. Here too, the pediatrician can raise awareness of this need at an early stage.

Only by implementing these recommendations lung damage in EA patients can be avoided–damage that diminishes quality of life and ultimately shortens life expectancy.

Conflict of Interest

The authors declare that they have no conflict of interest.

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Transition in Esophageal Atresia: Medical, Psychosocial, and Structural Aspects of Lifelong Care

Transition bei Ösophagusatresie: medizinische, psychosoziale und strukturelle Aspekte einer lebenslangen Versorgung

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ABSTRACT

Esophageal atresia (EA) is often perceived as a pediatric surgical diagnosis with treatment concluded after repair. In reality, EA represents a complex chronic condition requiring lifelong monitoring. Beyond surgical sequelae, patients are at risk for gastrointestinal, respiratory, and musculoskeletal long-term complications, frequently accompanied by associated malformations and psychosocial challenges. Transition from pediatric to adult-centered healthcare is particularly critical but remains insufficiently structured in Germany. Comprehensive, multidisciplinary teams for adults are scarce, and existing pilot projects are limited to single institutions. Consequently, pediatricians often act as key coordinators during transition, while patient organizations provide essential support through information, peer networks, and practical tools. Sustainable care structures – including dedicated transition centers, interdisciplinary networks, and standardized transfer protocols – are urgently needed to ensure continuity of care. Such frameworks would help to secure medical safety, psychosocial stability, and an independent quality of life for individuals with EA into adulthood.

ZUSAMMENFASSUNG

Die Ösophagusatresie (ÖA) wird vielfach als kinderchirurgische Diagnose verstanden, deren Behandlung mit der operativen Korrektur abgeschlossen scheint. Tatsächlich handelt es sich jedoch um eine komplexe chronische Erkrankung, die ein lebenslanges Monitoring erfordert. Neben operationsbedingten Folgen sind insbesondere gastroösophageale, muskuloskelettale und pulmonale Langzeitprobleme, zusätzliche Fehlbildungen sowie Aspekte der Lebensqualität zu berücksichtigen. Der Übergang in die Erwachsenenmedizin stellt dabei eine besondere Herausforderung dar. In Deutschland fehlen bislang flächendeckende Transitionsstrategien und spezialisierte multidisziplinäre Versorgungsteams; vorhandene Modellprojekte blieben bisher auf einzelne Standorte begrenzt. Eine kontinuierliche, interdisziplinäre Betreuung, die medizinische, psychosoziale und familiäre Dimensionen integriert, ist daher essenziell. Bis zur institutionellen Implementierung strukturierter Konzepte übernehmen Kinder- und Jugendärzt*innen eine Schlüsselrolle als Lots*innen im Transitionsprozess. Ergänzend leisten Selbsthilfeorganisationen durch Information, Peer-Support und praxisnahe Hilfen einen wichtigen Beitrag. Nachhaltige Versorgungsstrukturen sind Voraussetzung für eine erfolgreiche Transition und tragen dazu bei, jungen Menschen mit ÖA ein stabiles, gesundes und selbstbestimmtes Leben zu ermöglichen. Langfristig bedarf es politischer und institutioneller Rahmenbedingungen zur Etablierung von Transitionszentren, interdisziplinären Netzwerken und verbindlichen Übergabestrukturen, um eine adäquate Versorgung erwachsener ÖA-Betroffener sicherzustellen.

Introduction

Esophageal atresia (EA) has traditionally been understood as a pediatric surgical condition. Parents are often left with the impression that the disease is resolved following surgery and childhood. However, the reality is more complex: the patient journey does not end with surgical correction nor with the attainment of adulthood. While some families develop considerable acceptance of ongoing medical problems, this often results in relevant complications remaining undetected or untreated. The transition from pediatric to adult medical care is especially challenging. In Germany, major gaps persist, as specialized multidisciplinary care teams for adults with EA are still rarely established. This article highlights the key medical, psychosocial, and structural aspects of transition into adult care.

Transition in Medicine – Definition, Current Status, and Relevance in Esophageal Atresia

Defining Transition vs. Transfer

Transition (from the Latin transitio = passage) refers to the planned, multi-year process of moving from pediatric to adult health care. Its goal is to promote self-management, psychosocial maturity, and health-related autonomy [1, 2]. By contrast, transfer refers to the specific moment of handover, such as referral to an adult clinic [3]. Without preparatory transition, this often results in care discontinuities and poorer health outcomes [1–3]. National and international guidelines therefore emphasize structured preparation, individualized planning, and interprofessional coordination [4–6]. The German S3 guideline explicitly calls for the involvement of adolescents, parents, and adult health providers [6].

Current Situation in Germany

Germany currently lacks a nationwide, standardized transition strategy [7, 9]. Model initiatives such as TRANSLATE-NAMSE have provided valuable approaches, but remained locally limited [7, 12]. Surveys indicate that most pediatric departments have not implemented formal concepts [8, 9]. Moreover, the legal age threshold of 18 often enforces an abrupt transition, with insufficient preparation and psychosocial support.

A survey from pediatric nephrology illustrates this gap: more than 80% of clinics had no written agreements in place, yet emphasized the need for structured transition strategies [8]. Similarly, adults with EA are often inadequately connected to primary care structures [15] (see ▶ **Table 1**).

Esophageal Atresia – An Example of Complex Follow-up Care

Esophageal atresia (EA) is a congenital malformation of the esophagus and trachea that cannot be fully corrected by anastomotic surgery alone [33, 34]. Consequently, lifelong monitoring is essential. Long-term care must address both medical and psychosocial aspects, including additional anomalies associated with VACTERL as well as long-term sequelae of surgical treatment. This underscores the importance of continuous, interdisciplinary follow-up care that integrates both physical and psychological dimensions and actively includes family systems.

Gastroesophageal Long-term Problems

Recent reviews highlight increasing evidence for the lifelong gastrointestinal vulnerability of patients with EA [10]. While surgical reconstruction restores anatomical continuity, it only incompletely replaces the complex neuromuscular control of the esophagus. This frequently results in persistent motility-associated complaints. A personalized follow-up approach, guided by functional symptoms as well as endoscopic and manometric findings, is increasingly advocated [10].

Motility Disorders and Neural Control

Many patients suffer from disturbances of the oral phase, bolus transport, and lower sphincter control. Damage to the vagus nerve and its branches–particularly the recurrent laryngeal nerve–is thought to play a role [28, 29]. As these pathways are essential for motility, swallowing, and vocal cord function, dysfunction can cause not only dysphagia but also hoarseness, vocal cord paresis, and an increased risk of aspiration. These problems often emerge only in later childhood or adolescence, requiring interdisciplinary follow-up (e.g., gastroenterology, speech therapy, otolaryngology).

Gastroesophageal Reflux Disease (GERD)

GERD is one of the most common long-term complications, affecting about half of adolescents and nearly all adults with EA [18, 19]. Symptoms are often nonspecific, and even asymptomatic patients may develop clinically significant pathology. Possible complications include strictures, eosinophilic esophagitis, dumping syndrome, Barrett's metaplasia, and even carcinoma [18]. Regular surveillance endoscopies (EGD), ideally every 5–10 years, are therefore mandatory [18, 19].

▶ **Table 1** Gaps in Care in Germany – Current vs. Desired State.

Area	Current State	Target State
Transition Programs	Only pilot projects, locally restricted	Implemented nationwide
Age Limit	Abrupt at 18 years	Flexible, individually adapted
Adult Care Structures	Few specialized centers	Multidisciplinary centers nationwide
Follow-up Planning	Rarely documented	Standardized plans with handover meeting
Pediatrics–Adult Communication	Fragmented	Clear interfaces, designated contacts
Psychosocial Support	Insufficient	Integral part of transition concept

Nutrition and Growth

Children with EA often develop aversions to certain foods or fear of choking, [16] which may lead to eating disorders and growth impairment. In adulthood, distinct differences from the general population become apparent: 21% of patients are underweight, while only 2% are obese [15]. Speech and nutritional therapy remain important beyond childhood and adolescence to prevent malnutrition and related complications [15, 17]. This is particularly relevant in cases where primary care was surgical only and long-term structured follow-up was lacking.

Respiratory Long-term Complications

Respiratory problems are among the central sequelae of EA [11, 13, 14]. Tracheomalacia, occurring in up to one-third of patients, [11] is especially common. Tracheal instability may lead to chronic cough, recurrent infections, atelectasis, and eventually bronchiectasis. Symptoms such as stridor, wheezing, or dyspnea are often misinterpreted in childhood as asthma or croup. Microaspiration-related bronchitis is also frequent.

Diagnostics

Assessment is interdisciplinary and includes pulmonary function tests, flexible and rigid bronchoscopy, and imaging studies. Low-dose CT or real-time MRI with dynamic breathing maneuvers allow visualization of tracheal and bronchial collapse phenomena [13, 14, 35].

Therapy and Management

Therapeutic options range from inhalations (saline, N-acetylcysteine) and structured respiratory physiotherapy (e.g., autogenic drainage, PEP systems) to surgical interventions such as tracheopexy in severe cases [11, 13, 14]. Antibiotic strategies modeled after cystic fibrosis care are also established [14]. Education of parents–and later patients themselves–on disease management, including keeping standby antibiotics for acute situations, is crucial, even in adulthood.

Structured Long-term Care

Ongoing respiratory physiotherapy should be prescribed as a standard part of follow-up care [13]. In cases of recurrent infections or signs of chronic lung damage, transition to specialized bronchiectasis clinics (pediatric or pulmonary) is advisable to ensure regular monitoring and early intervention [14]. For further details, see “Pulmonary Problems in Children with Esophageal Atresia” in this issue.

Musculoskeletal Sequelae and Scoliosis

After open thoracotomy, many EA patients develop musculoskeletal sequelae such as rib fusions, thoracic asymmetries, and scoliosis [17, 32]. Minimally invasive thoracoscopic techniques significantly reduce this risk [20].

A recent prospective MRI study from Leipzig provided the first morphological and functional evidence in real time: children after open EA repair showed rib fusions and adhesions (78%) as well as scoliosis (15%) significantly more often than thoracoscopically operated children or healthy controls [32]. Thoracic development was also impaired, with reduced right-sided lung volumes and asymmetric thoracic movement. The number of thoracotomies emerged as a key risk factor for reduced thoracic volumes.

These findings highlight that musculoskeletal and pulmonary sequelae after thoracotomy are frequently underestimated and require targeted follow-up. In addition to clinical and imaging surveillance, physiotherapy, breathing exercises, and sports activities should be actively encouraged. Appropriate sports not only support physical development but also strengthen psychosocial stability (See “Supporting Physical Literacy in Patients with Esophageal Atresia”).

Psychosocial Aspects and Quality of Life

Despite substantial medical progress, many patients continue to report impairments in everyday life. These include reduced physical capacity, limitations in school, education, or work, and challenges in relationships and quality of life [15, 21–23]. Dysphagia-related problems in social situations (e.g., eating together) and school absenteeism due to recurrent respiratory infections are particularly distressing, contributing to emotional stress and reduced quality of life [21, 22, 31].

Moreover, studies show an increased prevalence of psychiatric disorders such as depression, ADHD, and autism in EA patients [24]. Therefore, psychological health should be routinely assessed during follow-up. In addition to medical care, involvement of patient support groups (e.g., KEKS, SoMA) and empathetic, open physician–patient communication are critical factors for sustaining long-term quality of life (see ▸ **Table 1**).

Recommendations for Outpatient Care (See ▸ **Table 2**)

Transition into adult medicine is particularly critical, as specialized multidisciplinary teams remain rare in Germany. Against this background, the following recommendations apply for outpatient care:

- Gastrointestinal follow-up: Regular endoscopies (EGD) with biopsies, even if asymptomatic, to detect complications such as strictures, GERD, Barrett’s metaplasia, or eosinophilic esophagitis early.
- Respiratory diagnostics: Pulmonary function testing every 1–2 years; bronchoscopy for recurrent infections or chronic cough; respiratory physiotherapy and encouragement of age-appropriate physical activity.
- Nutritional status: Regular assessment of weight, BMI, and eating behavior; early referral to nutrition specialists and speech therapists if needed.
- Musculoskeletal screening: Clinical exam for postural issues and scoliosis; imaging if indicated.
- Psychosocial support: Routine assessment of quality of life (e.g., EA-QoL questionnaire); referral to psychotherapy or social services as appropriate.
- Prevention: Verification and updating of vaccinations, particularly pneumococcus, influenza, pertussis, and Hib, optional: RSV (Respiratory Syncytial Virus) – new vaccination options:

▶ **Table 2** Recommendations for Outpatient Follow-Up in Esophageal Atresia.

Area	Recommended Measure	Frequency/Comment
Gastrointestinal	EGD with biopsies (strictures, GERD, Barrett's, EoE)	Every 5–10 years or symptom-based
Respiratory	Pulmonary function, if needed bronchoscopy, respiratory therapy	Every 1–2 years, immediately if infections occur
Nutrition	Weight, BMI, eating behavior	Annually
Musculoskeletal	Inspection for posture, scoliosis	Annually
Psychosocial	EA-QoL, screening for depression/ADHD/autism	Regularly
Vaccinations	Pneumococcus, influenza, pertussis, Hib, optional: RSV (Respiratory Syncytial Virus) and COVID-19	Check vaccination status regularly
Networking	KEKS/NEKS health folder, participation in self-help groups	Continuously

e. g., nirsevimab (passive immunization during the first winter of life), especially for infants with risk factors. Consider also: COVID-19 (according to STIKO recommendations for children from 6 months of age with underlying conditions).

- Self-help and networking: Recommendation to engage with patient organizations such as KEKS or SoMA; use of the KEKS health folder as a structured documentation and communication tool.

These recommendations emphasize that follow-up care for EA patients must be understood not as a completed surgical intervention but as a lifelong interdisciplinary task.

Stakeholders in Comprehensive Follow-up Care

Pediatricians as Key Actors

Pediatricians, who often accompany chronically ill children for many years, play a central role in the transition process. With their knowledge of disease course, family dynamics, and psychosocial factors, they can initiate transition early, integrate age-appropriate education, and develop individualized plans.

Targeted assessments enable evaluation of self-management, disease processing, and psychosocial maturity. On this basis, measures such as transition clinics, internships in adult outpatient care, or structured handover discussions can be planned. In practice, however, these approaches often fail due to lack of resources and insufficient networking with adult medicine. Adult physicians are frequently unprepared for the complex needs of EA patients, underscoring the importance of binding communication structures and interdisciplinary collaboration.

Patient Organizations as Bridge Builders

Self-help organizations such as KEKS or SoMA complement medical care with information, peer support, and transition programs. Their offerings range from workshops and transition booklets to mentoring by older patients. These initiatives promote health literacy, adherence, and quality of life [25, 26].

In addition, patient organizations act as mediators between families and care structures: they address fears, clarify expectations, and build trust in new medical teams. Their continuous involvement helps mitigate disruptions in the transition process [30].

Designing a "Transfer Arrival Station"

A central goal for future care structures should be the development of a Transfer Arrival Station–a real or virtual space where handovers from pediatric to adult medicine are coordinated and supported [7, 27]. Essential components could include:

- Joint transition clinics with pediatric and adult providers
- Structured transfer plans with relevant disease and treatment history
- Transition coordinators or "navigators"
- Psychosocial support from social work and psychology
- Participatory follow-up discussions to evaluate the transition

Such a "station" would be more than just a handover point: it would provide empowerment, validation, and collaboration, closing the "black hole" between care systems and offering adolescents and families both safety and orientation.

Conclusion

The transition of children and adolescents with chronic diseases–particularly rare and complex conditions such as esophageal atresia–remains a critical yet insufficiently structured phase of care. While surgical repair resolves the acute phase, numerous gastrointestinal, pulmonary, musculoskeletal, and psychosocial sequelae necessitate continuous interdisciplinary follow-up.

Currently, Germany faces major deficits: lack of nationwide transition programs, rigid age limits, insufficient networking with adult medicine, and very few established multidisciplinary structures. These care gaps pose not only medical risks but also impair quality of life and psychosocial stability.

Until structured transition concepts are institutionally anchored, pediatricians remain key actors: as continuous points of contact, they can actively shape transition through early education, motivation, and referral to supportive networks. Patient organizations such as KEKS and SoMA provide essential additional resources by offering information, peer support, and practical assistance.

In the long term, however, binding political, institutional, and financial frameworks are required: transition centers, interdisciplinary networks, structured handover processes, and defined “arrival stations” that coordinate and accompany the move into adult care. Only through such sustainable structures can the transition truly succeed–ensuring not only the prevention of complications but also a stable, healthy, and self-determined adult life for individuals with esophageal atresia and comparable conditions.

Conflict of Interest

The authors declare that they have no conflict of interest.

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Esophageal Atresia as a Family Disease – Effects of EA on the Infant, on the Family System and on the Mental Health of Parents

Ösophagusatresie als Familiendiagnose – Auswirkungen der ÖA auf den Säugling, auf das Familiensystem und auf die psychische Gesundheit der Eltern

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ABSTRACT

The diagnosis of esophageal atresia (EA) in a newborn presents a significant medical and psychosocial burden for the entire family system. This article addresses the reactions of affected infants, the psychological impact on parents–with a special focus on the risk of post-traumatic stress disorders–as well as potential consequences for siblings and parental relationships. Additionally, practical recommendations are provided for pediatricians regarding the early recognition of family distress and suitable intervention options. The article emphasizes persistent neonatal stress–resulting from repeated medical interventions, separation from parents and EA-typical symptoms like shortness of breath, swallowing difficulties, reflux problems, and oral aversion–can be associated with impaired central nervous stress processing. Such early stressors can manifest in elevated cortisol levels, sleep disturbances and regulatory problems and can negatively influence the child’s social and neurological development in the long term. The findings underline the need for holistic care that includes psychosocial aspects of the family in addition to medical care.

ZUSAMMENFASSUNG

Die Diagnose einer Ösophagusatresie (ÖA) beim Neugeborenen stellt eine erhebliche medizinische und psychosoziale Belastung für das gesamte Familiensystem dar. Der vorliegende Beitrag widmet sich den Reaktionen des betroffenen Säuglings, den psychischen Auswirkungen auf Eltern – mit besonderem Augenmerk auf das Risiko posttraumatischer Belastungsstörungen – sowie möglichen Folgen für Geschwisterkinder und elterliche Paarbeziehung. Zudem werden praxisnahe Empfehlungen für Kinderärztinnen und -ärzte zur frühzeitigen Erkennung familiärer Belastungen sowie geeigneter Interventionsmöglichkeiten aufgezeigt. Der Artikel betont, dass anhaltender neonataler Stress – infolge wiederholter medizinischer Eingriffe, Trennung von den Eltern und ÖA-typischen Symptomen wie Atemnot, Schluckstörungen, Refluxproblematik und oraler Aversion – mit einer beeinträchtigten zentralnervösen Stressverarbeitung assoziiert sein kann. Solche frühen Belastungen können sich in erhöhtem Kortisolspiegel, Schlafstörungen und Regulationsproblemen äußern und langfristig die soziale und neurologische Entwicklung des Kindes negativ beeinflussen. Die Ergebnisse unterstreichen die Notwendigkeit einer ganzheitlichen Betreuung, die neben der medizinischen Versorgung auch psychosoziale Aspekte der Familie einbezieht.

Neurological and Developmental Psychological Implications

Maturation of the central nervous system

Chronic pain, interruptions of sleep cycles, and stress caused by medical interventions can impair neurological maturation [1–3]. This particularly affects:

- Regulation of alertness and affect
- Sensory integration
- Development of socio-emotional responses

Risk for regulatory disorders

Infants with EA more frequently present with:

- Persistent crying
- Sleep disturbances
- Impaired self-regulation

These are described as "regulatory disorders" in infancy and are closely associated with early medical stress.

Attachment and relationship aspects

The illness affects not only the infant's physiological development but also their social environment:

- Limited interaction with parents due to separation after birth and the presence of medical devices.
- Negative oral experiences (e.g., from suctioning, (feeding) tubes,) lead to aversions towards oral stimulation.
- Delayed relationship-building can have long-term consequences on emotional development.

A disrupted development of the "body-self" and the initial self-environment boundaries has been described in neonates receiving intensive medical care [4].

Immediate medical intervention after birth often results in physical separation between mother and child. The absence of direct skin-to-skin contact and delayed first breastfeeding make it difficult to establish an emotional bond [5]. Early attachment is characterized by non-verbal communication, physical contact, and intuitive parental responses to infant signals. For infants with EA, these interactions are often complicated by medical equipment, hospitalization, and the infant's physical condition. Parents often experience uncertainty in caring for their child, particularly during feeding or care. This can lead to feelings of alienation [6].

Supportive interventions through parent-centered care

- An interdisciplinary team (nurses, psychologists, social workers) can provide early support to parents and address their emotional needs. Involving parents in caregiving and medical decisions promotes a sense of control and strengthens attachment.
- Early psychological support for families.
- Psychoeducational programs and low-threshold psychological support reduce the risk of developing depression and foster parental resilience.
- Promoting closeness despite the hospital routines.
- Measures such as "kangaroo care" (skin-to-skin contact in the intensive care unit), rooming-in, and targeted support of parent-child interaction can help strengthen the bond despite technical barriers [7].

Long-term consequences and course

Even after a successful operation, many infants face long-term issues that can affect their development:

- Dysphagia (swallowing disorders), reflux, feeding problems
- Growth delays
- Frequent infections (especially of the respiratory tract)
- Sensory integration problems

These factors, in turn, affect play, learning, and exploratory behavior in infancy and may contribute to developmental delays.

Psychological Impact on Parents

The diagnosis of EA is usually made immediately after birth and leads to acute psychological distress for parents. Feelings of shock, fear, helplessness, and grief over "lost normality" are common.

Research has found that 20–30% of parents of critically ill newborns develop symptoms of PTSD [6, 8]. Risk factors include fear for the child's life, loss of control in the medical setting, separations immediately after birth, and insufficient social support. Mothers are particularly often affected. The diagnosis of a rare, chronic illness like EA, along with the necessity of prolonged medical treatment, represent a significant emotional burden for caregivers.

The diagnosis of a congenital malformation in the child is perceived as traumatic by 88% of mothers and 83% of fathers [9]. Compared to parents of healthy children, parents of children with rare diseases report reduced quality of life, increased stress symptoms, loneliness, and post-traumatic stress disorders [10–12]. A French study found that 59% of parents of children with EA experience PTSD symptoms, regardless of the medical severity of the child's illness and their long-term health prognosis [13].

In order to allow for a more nuanced consideration of the diagnosis of post-traumatic stress disorder and to refined the definition of the term "traumatization", which in our view is often used in an inflationary way, the diagnostic criteria are summarized in this chart.

To enable a nuanced consideration of the diagnosis of post-traumatic stress disorder and to sharpen the definition of the term "traumatization", which in our view is often used in an inflationary way, the diagnostic criteria are summarized in this chart below.

Psychological Impact on the Affected Child

From the very beginning, the development of a child with EA is influenced by pain, hospital stays, and attachment insecurities. More than half of mothers of children with EA reported that their infants had experienced a medically induced trauma and that they observed PTSD symptoms in their infants, including distress when reminded of the trauma, difficulty falling asleep, and the emergence of new fears [15]. Additionally, it has been described that children with EA employ various illness-specific coping strategies [16]. It became evident that children with EA use a wide range of positive coping strategies to overcome health-related challenges.

The preschool and school years may be associated with an increased need for care and a continued higher incidence of infections, placing the affected child in a special role. Young children born with an EA experience a significant symptom burden. In particular, respiratory problems (e.g., frequent infections, coughing, and breathing difficulties/dyspnea) can lead to severe stress symptoms even in children aged 0,5 to 7 years [17]. School and preschool absenteeism can be seen as indicators of the burden experienced by both the EA child and the family. Due to absenteeism and limitations in performance, EA children often experience more stress, which can restrict their participation in social activities.

THE CONNECTION BETWEEN THE DIAGNOSIS OF ESOPHAGEAL ATRESIA IN INFANTS AND POST-TRAUMATIC STRESS DISORDER (PTSD) IN PARENTS

The combination of shock, fear, medical uncertainty, and physical separation between parents and child can trigger traumatic stress reactions in parents. In some cases, these reactions develop into clinically significant symptoms of post-traumatic stress disorder (PTSD). PTSD is a mental health condition that can occur as a result of an extremely distressing or traumatic event. Diagnosis is based on the criteria of the DSM-5 or ICD-11 and includes, among others, intrusive memories, avoidance, hyperarousal, and negative alterations in thoughts and emotions.

Definition and symptomatology of PTSD

PTSD in parents: prevalence and risk factors

Studies show that 20–30% of parents of critically ill newborns develop clinically significant PTSD symptoms [6, 8]. Risk factors include:

- Emergency or high-risk delivery
- Premature birth or congenital malformations
- Separation of child and parents
- Perceived helplessness and loss of control
- Lack of psycho-social support

Diagnostic and early detection of PTSD in parents

Diagnosing parental PTSD requires specific screening tools tailored to the unique circumstances of parents in medical crisis situations.

- Impact of Event Scale-Revised (IES-R)
- Parental Stressor Scale: NICU
- PTSD Checklist for DSM-5 (PCL-5)

A early screening – already during the hospital stay – can help to identify at-risk families and provide targeted support.

Prevention and intervention

- Psycho-education and communication
- A transparent flow of information, empathic communication and parental involvement in care decisions are essential
- Psychological support

An interdisciplinary team with integrated psychosocial care should be a standard component of perinatal center services. Short-term therapies (e.g., trauma-focused cognitive behavioral therapy, EMDR) have proven effective in early intervention.

Connection between Esophageal Atresia and PTSD Risk

Parents of infants with EA are exposed to a variety of potentially traumatic stressors:

- Acute life-threatening condition of the child: The threat to the child's life is one of the strongest triggers for traumatic reactions.
- Immediate surgeries and complications: The need for urgent medical decisions and repeated interventions reinforces the sense of loss of control.
- Long-term burden from hospitalization: Weeks-long stays in the neonatal unit with repeated stressors (e.g., reflux, feeding tubes, failure to thrive) lead to chronic stress.
- Parental guilt: Many parents unconsciously blame themselves or each other.

The burden is intensified by uncertainty regarding the course of the disease and possible long-term complications. Studies show that mothers are particularly affected, although fathers also have an elevated risk [14].

Impact on the Rest of the Family

Once the sick child returns home, the complex influence of the disease on the entire family becomes clear. Especially in the initial stages, feelings of being overwhelmed and fearful can lead to withdrawal from the social network and a reduction in social participation. Many affected families report isolation and a withdrawal from social life, which they attribute to the limitations associated with the illness and the lack of resources to maintain and build social connections [18, 19]. The tendency to withdraw is further reinforced by societal prejudices and the burden of illness, which makes it seem easier for many to limit their lives to the domestic sphere [19].

Social participation is also further reduced by the child's limited and more complicated eating situation, along with associated emotions such as fear and frustration.

Due to the special needs of the EA child, witnessed emergency situations, the increased care and nursing time for the EA child, and the withdrawal from the social network, potential siblings are also burdened. Siblings may feel neglected and react with separation anxiety, general anxiety, or feelings of guilt.

The couple's relationship is often significantly strained due to different coping strategies in dealing with the situation, exhaustion, and the lack of time for the partnership. Studies have shown that mothers and fathers often use different forms of stress management: mothers tend to engage in intense emotional processing, whereas fathers act in a more functional-pragmatic way [20]. This discrepancy can lead to misunderstandings, communication difficulties, and emotional distance within the couple's relationship.

Grandparents and other close caregivers are also affected. Financial and organizational challenges can further increase the family's burden.

Caring for a child with EA can consume substantial time and financial resources:

- Reduction or loss of working hours for one parent
- Travel burdens to specialized centers
- Costs for accommodation, meals, and medical supplies
- Bureaucratic hurdles in applying for benefits (e.g., nursing care level, assistive devices, rehabilitation)

Coping with the demands of daily life for a child who requires special healthcare support can be associated with an increased risk of social isolation and restricted participation in community life [18, 19].

Family Dynamics in the Context of an Acute Childhood Illness

From the perspective of systemic family psychology, every family is a dynamic system in which changes to one element affect the entire system. The serious illness of an infant, such as EA, causes an acute restructuring of family roles, division of tasks, and emotional bonds [21].

This can lead to shifts in responsibilities–for example, siblings may develop feelings of guilt regarding the family's "exceptional situation" and, as a result, develop their own symptoms or react by becoming "parentified", taking on parental functions and feeling responsible for caring for the parents or younger siblings.

In the best-case scenario, the family initially responds "dynamically", meaning it activates resources in response to the illness of one member. In this way, the system does not experience itself as deficient and disadvantaged, but also develops special survival strategies that strengthen cohesion and a sense of self-efficacy. This is only reliably possible if there are sufficient external contacts and networks that were already effective and helpful for the family before the child's illness.

The classical systemic paradigm always refers to the circularity of systems (meaning that "things" do not exist in isolation but are in relation to one another). A does not act unilaterally on B; rather, B also acts on A (reciprocal interaction). Therefore, despite all the strain on the family, we assume that the system is fundamentally capable of "reinventing" itself and finding an adequate response to the challenging situation. In our work with families, we see that it is consistently possible to create space for the special needs of sick children and to establish a supportive, emotionally nurturing family atmosphere. To put it in the words of the individual psychologist Alfred Adler: it is not the burdens or traumas experienced that matter, but the response we find to them: "It's not about what one is born with, but about what one makes of it" [22].

Nevertheless, the diagnosis of EA can repeatedly push families to the limits of their resilience. As mentioned earlier, the family's network plays a major role in coping adequately with these stress factors. Since pediatricians usually play an important role in supporting families in connection with illness, we would like to conclude with a focus on how pediatricians can recognize relevant stress factors and which interventions are recommended.

How Can Pediatricians Recognize Family Distress?

Observable warning signs in the child

- Failure to thrive, which can manifest as insufficient weight gain despite adequate medical care, may indicate stressful feeding situations or psychosocial stress.
- Swallowing refusal, oral aversions, and excessively frequent vomiting or panic during feeding can be an expression of strained parent-child interactions.
- Delayed development, especially in social or emotional domains, can indicate a lack of interaction or stimulation.
- Regulatory disorders such as excessive crying, sleep disturbances, or hyperactivity are signs of insufficient fulfillment of the infant's basic needs for attachment, orientation, and control [23].

Notable parental behaviors

- Overly anxious or avoidant behavior toward the child
- Ambivalence: strong fluctuations between overprotectiveness and withdrawal
- Marked exhaustion or depressive symptoms (e.g., crying, lack of drive, irritability)
- Inappropriate feelings of guilt ("I did something wrong")
- Marital conflict or inconsistent statements by mother and father regarding the home situation
- Missed medical appointments/therapy discontinuations, can indicate being overwhelmed

Family risk factors identified in conversation

- Low social support (e.g., no extended family network)
- Financial strain due to work loss, hospital stays
- Stressed siblings (school problems, withdrawal, jealousy)
- Lack of knowledge or feeling overwhelmed with home care (e.g., feeding tubes, reflux management)

What Can Pediatricians Advise and Offer to Families?

Establish a trustful communication

- Active listening and validation: Take parents' concerns seriously ("That sounds very stressful for you.")
- Psychoeducation: Provide age-appropriate explanations about illness, prognosis, and care to reduce uncertainty
- Normalize emotions: Explain that feelings such as fear, exhaustion, and being overwhelmed are common

Arrange for early psychosocial support

- Referral to psychosocial counseling (e.g., hospital social services, SPZ, parenting support services)
- Early intervention and developmental support for the child when risks are present
- Parent–child interaction counseling for attachment difficulties or problematic behaviors
- Psychological support for post-traumatic stress, depression, or relationship conflicts
- Self-help groups (e.g., KEKS e. V. – Self-Help Organization for Children and Adults with Esophageal Atresia)
- Recommendation to seek additional support from the local youth welfare office
- Referral for specific psychological/psychiatric assessment
- Recommendation/referral for occupational therapy and psychotherapy

Strengthen Family Self-Efficacy

- Actively involve parents in decisions and care processes
- Strengthen “parental competence” by acknowledging small successes
- Establish routines and positive rituals in daily life
- Develop relief strategies (e.g., home care assistance, household help, childcare)

Don’t Forget Siblings

- Provide information about counseling services for siblings
- Recommend age-appropriate inclusion (“The baby is sick, but you are just as important.”)

Conclusion

When caring for children with EA, pediatricians should also pay close attention to the family’s overall situation. Notable changes in parent–child interaction, emotional exhaustion of parents, or developmental problems in the child are serious warning signs. Early networking with psychosocial resources, clear communication, and strengthening parental competence are key measures for supporting these families.

Conflict of Interest

The authors declare that they have no conflict of interest.

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Supporting Physical Literacy in Patients with Esophageal Atresia

Verbesserung der Physical Literacy bei Kindern und Jugendlichen mit Ösophagusatresie

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ABSTRACT

Compared to their peers, children and adolescents with esophageal atresia have reduced physical activity, motor skills and impaired lung function. Respiratory complaints caused by tracheomalacia, bronchial obstruction and chronic inflammation limit physical performance. More rarely, failure to thrive, gastroesophageal reflux and swallowing disorders can also cause problems during prolonged exertion. In addition, parental anxiety and overprotectiveness may have a negative impact on physical activity behaviour. Targeted promotion of physical literacy – motivation, self-confidence, ability and knowledge – can help to ensure physical activity in the long term. In addition to the targeted treatment of exercise-related symptoms, medical care should include the teaching of suitable everyday strategies and the promotion of motivation to actively participate in sport.

ZUSAMMENFASSUNG

Im Vergleich zu Gleichaltrigen weisen Kinder und Jugendliche mit Ösophagusatresie eine verminderte körperliche Aktivität, motorische Fähigkeiten und eine eingeschränkte Lungenfunktion auf. Respiratorische Beschwerden bedingt durch Tracheomalazie, bronchiale Obstruktion und chronische Inflammation schränken die körperliche Leistungsfähigkeit ein. Bei längeren Belastungen können seltener auch eine Gedeihstörung sowie gastroösophagealer Reflux und Schluckstörungen Probleme bereiten. Zusätzlich beeinflussen elterliche Ängste und Überfürsorglichkeit das Bewegungsverhalten möglicherweise negativ. Eine gezielte Förderung der Physical Literacy – Motivation, Selbstvertrauen, Fähigkeit und Wissen – kann dazu beitragen, die körperliche Aktivität langfristig zu sichern. Die ärztliche Betreuung sollte, neben der gezielten Behandlung belastungsabhängiger Symptome, die Vermittlung geeigneter Alltagsstrategien sowie die Förderung der Motivation zur aktiven Teilnahme am Sport beinhalten.

Physical Activity and Promoting Exercise

The first 1,000 days of life–from conception to the end of the second year of life–are crucial for shaping health factors such as nutrition and exercise habits [1]. Deficiencies during this critical developmental period are often discussed in the context of global inequality [1], but in children with chronic conditions such as esophageal atresia, who spend a large part of their first two years of life undergoing medical treatment [2], general health promotion is neglected in the context of constant need for specific therapy. Yet it is precisely in children who already live with health limitations that it is crucial to promote healthy behaviours as much as possible. Family activities play an outstanding role in early childhood development and the establishment of an active lifestyle

[1, 3], and physically active children and adolescents become active adults [4]. This makes promoting physical activity at all ages a family project. Furthermore, the foundations for lifelong mobility into old age are laid in childhood and adolescence. Physical activity in youth greatly influences long-term muscle [5] and bone mass [6] as well as lung function [7] in older ages. It’s a paediatrician’s job to plan far ahead for our patient’s long-term outcome of many years to come. Publicly accessible and free-of-charge information material of promoting physical activity, such as the new “prescription for exercise” for children and adolescents and detailed target group-specific information materials, are provided by the German Sports Youth of the German Olympic Sports Confederation (www.dsj.de) or the Federal Institute for Public Health (shop.bioeg.de).

Children and adolescents with “special medical care needs,” the term used in epidemiological research to refer to all chronic diseases, are among the recognized risk groups for reduced physical activity [8]. In 2020, the World Health Organization (WHO) published new recommendations for physical activity that, for the first time, explicitly included children and adolescents with chronic diseases [9]. According to these recommendations, children and adolescents aged six years and older should exercise for an average of 60 minutes a day, so that they sweat at least a little and are out of breath (moderate-to-vigorous physical activity). Three times a week, the activities should also include strength training [9]. The Federal Centre for Health Education (BZgA) in Germany recommends 90 minutes of exercise per day for children and adolescents, including low-intensity everyday activities such as walking [10]. For toddlers, the WHO and the BZgA even recommend 180 minutes of daily exercise [3, 10]. Both institutions emphasize limiting screen time as the key to promoting physical activity.

The positive effects of physical activity on many chronic physical and mental illnesses in children and adolescents are now well documented [11]. However, children with chronic diseases, such as congenital heart disease are exempt from physical education more often than medically strictly necessary [12]. For esophageal atresia patients, there are no specific intervention studies that prove an improvement in health through physical activity to date, but there are reports on positive experiences from clinical practice and patient organizations. At the very least, generally accepted assumptions about the positive effects of exercise and sports on health apply [11].

Since physical activity depends on more than just physical fitness, the holistic concept of “physical literacy” was introduced in 2001, which describes essential pillars for an active lifestyle in addition to pure physical competence: motivation, confidence, and knowledge and understanding [13] (▶ **Fig. 1**). Concepts for improving physical literacy are usually applied systemically, for example in schools. In the case of rare diseases, however, disease-specific support, especially the transfer of knowledge, is crucial. In addition to medical staff, patient organizations also play a key role here.

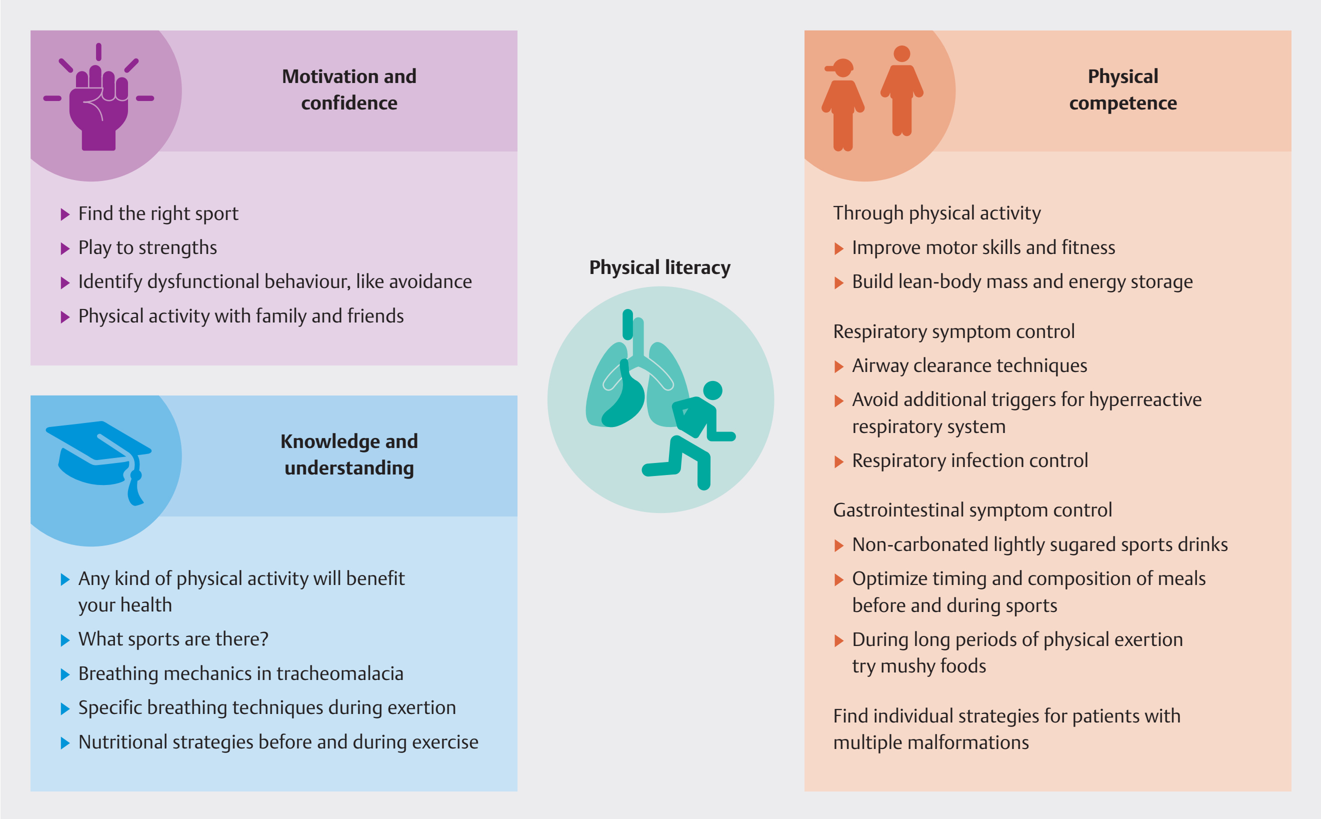
Physical Activity and Fitness

Children and adolescents with esophageal atresia generally participate in school and club sports to a normal extent, but they still spend significantly less time being physically active each week than their healthy peers [14]. While there is no relevant difference in physical activity at primary school age, the average difference increases with age, especially in girls [14]. In children and adolescents, impaired motor skills have also been demonstrated in small cohorts [15–17]. However, there are very large individual differences in physical activity [14] and performance [16, 18], which is influenced by factors related to esophageal atresia and general environmental factors, such as socioeconomic status. With regards towards physical activity, different surgical strategies, such as minimally-invasive versus open repair or additional fundoplication had no influence [14]. Symptoms during exertion and lower body weight, however, were associated with reduced physical activity [14]. Further studies with larger samples are needed to identify key factors for physical activity and fitness in these patients.

In addition to physical factors, the mindset of esophageal atresia patients and their families is crucial. In many cases, mental health and self-care of parents of chronically ill children are also significantly reduced [19]. In the context of esophageal atresia, not only physical activity of affected children and adolescents, but also of their parents was significantly impaired [20]. More than two-thirds of parents of children with esophageal atresia complained of psychological problems such as increased anxiety and depressive symptoms [21], which can be transferred to the children or lead to an overprotective and highly controlling parenting style with avoidance [19]. Avoidant behaviour, both on the part of parents and the affected subjects, is a maladaptive disengaging coping strategy that should not be legitimized by medical prohibitions whenever possible [19]. Membership of a sports club by family members, on the other hand, was associated with increased physical activity among patients. In clinical practice, avoiding behaviour with regards towards physical activity should be recognized and addressed [14].

Possible Symptoms During Physical Exertion

Esophageal atresia is a combined malformation of the esophagus and respiratory tract that can be associated with a variety of respiratory complications in addition to gastrointestinal complaints [22]. Schoolchildren showed significantly reduced cardiopulmonary function, which was attributed to recurrent pulmonary infections and reduced physical activity [23]. In addition, both increased obstructive and restrictive ventilation disorders were detected. These were accompanied by a reduction in total lung capacity, which in turn is associated with reduced cardiopulmonary performance [23]. However, physical activity generally led to a catch-up in lung function parameters between the ages of four and ten in children with other lung disease during infancy [24]. In esophageal atresia patients, symptoms during exercise occurred in about a quarter of patients [14]: respiratory complications associated with tracheomalacia and bronchial obstruction are the most common limiting factors for physical activity in affected individuals [14]. Impaired secretion clearance in the context of tracheomalacia can also play a role during exercise [22]. In order to improve participation and enjoyment in sports, symptom control and avoidance of additional pulmonary stressors in a hyperreactive bronchial system are therefore crucial, which in turn has a positive effect on lung function through training.



Motivation and confidence

- Find the right sport
- Play to strengths
- Identify dysfunctional behaviour, like avoidance
- Physical activity with family and friends

Knowledge and understanding

- Any kind of physical activity will benefit your health
- What sports are there?
- Breathing mechanics in tracheomalacia
- Specific breathing techniques during exertion
- Nutritional strategies before and during exercise

Physical literacy

Physical competence

Through physical activity

- Improve motor skills and fitness
- Build lean-body mass and energy storage

Respiratory symptom control

- Airway clearance techniques
- Avoid additional triggers for hyperreactive respiratory system
- Respiratory infection control

Gastrointestinal symptom control

- Non-carbonated lightly sugared sports drinks
- Optimize timing and composition of meals before and during sports
- During long periods of physical exertion try mushy foods

Find individual strategies for patients with multiple malformations

▶ **Fig. 1** Condition-specific promotion of physical literacy in children and adolescents born with esophageal atresia.

Underweight is common in esophageal atresia patients, especially in early childhood, and is often associated with a complicated course, persistent nutritional difficulties, and growth disorders [25]. A reduction in fatty and fat-free body mass is associated not only with reduced strength [16], but also with a lower capacity to store substrates of both aerobic and anaerobic energy metabolism. In rare cases, the daily calorie intake of patients with esophageal atresia exceeds their basal metabolic rate [26]. However, energy expenditure during moderate physical activity as recommended daily by the WHO is defined as three-fold basal metabolic rate [8]. Parents also described slim physique and, in some cases, short stature, as well as a “lack of energy” as limiting factors in sports [14]. A lower body weight and height was associated with a reduced amount of moderate-to-vigorous physical activity [14]. In esophageal atresia, prolonged exertion with short meal breaks is particularly problematic, as patients need more time to eat and a longer digestive break between meals and renewed exertion. In these cases, individual solutions must be found. In general, foods that are low in fat, acid, and fiber and not very spicy are recommended to control reflux symptoms [27, 28]. For those affected by dysphagia, in addition to lightly sweetened non-carbonated beverages, a regimen including mushy carbohydrate-containing foods such as fruit puree or glucose gels should be tested. In general, nutritional and swallowing problems, as well as gastroesophageal reflux, which dominate the everyday lives of many families [29], have subjectively no major impact on physical activity according to patients [14]. One possible explanation is that many patients have already found effective individual strategies for timing and composition of food before planned exertion.

Esophageal atresia is associated with other congenital malformations and syndromes. A combination of several associated malformations is another significant negative factor influencing physical activity and is also the main reason for exemption from school sports [14]. The combinations of malformations are highly individual and require appropriate solutions for affected patients and their families. Special exercise programs for children and adolescents with congenital malformations without mental disabilities have not been successfully established in Germany, so that, whenever possible, integration into regular organized sports is warranted. Contrary to expectations, no statistical correlation with inactivity could be demonstrated for associated congenital heart disease in the context of esophageal atresia [14]. A specific analysis of children with isolated esophageal atresia compared to isolated congenital heart defects showed a comparable reduction in physical activity compared to healthy peers [30]. It was particularly noticeable that the active time of both patient groups decreased significantly with increasing age [30]. Adolescence is a vulnerable phase in which young people develop their own physical self-concept and become more aware of their chronic illness and physical differences [31]. Even in the normal population, there is a regular decrease in physical activity during this phase [31], but this was significantly more pronounced in adolescents

▶ **Fig. 2** Graham Slater, one of the oldest survivors with corrected esophageal atresia.

with congenital heart defects and esophageal atresia [30]. Physical activity should therefore be a particular focus of follow-up care in this vulnerable group.

Conclusion

Just as unfavourable environmental factors can negatively influence child health, the health of children and adolescents with chronic diseases can be optimized through targeted support and avoidance of dysfunctional compensation mechanisms. Currently, follow-up care for esophageal atresia focuses primarily on physical, especially surgical, aspects. However, other pillars of physical literacy should also be addressed in a disease-specific manner as part of promoting physical activity. The outcome, physical activity, and physical performance vary greatly among individuals with esophageal atresia. Whenever possible, integration into organized sports should be pursued. In cases of more severe physical limitations, a suitable program must be found that initially promotes the child's strengths rather than their deficits in order to boost motivation and self-confidence. Fun and positive experiences also increase the likelihood that patients will remain active in the long term [9]. For patients with esophageal atresia, for example, sports can be chosen in which lower body weight is irrelevant or even advantageous, sports in weight classes or with a lesser focus on endurance. In order to enable a carefree time during sports, optimal management of respiratory complaints and a robust nutritional concept for prolonged exertion are essential. Effective symptom control can in turn boost lung function parameters through improved fitness. In addition, patient education on how to deal with possible physical limitations during exercise is crucial, for patients to take responsibility for their own bodies. Further studies are needed to identify relevant factors influencing cardiopulmonary fitness and motor skills in esophageal atresia.

Graham Slater (▶ **Fig. 2**), chair of EAT (2011–2021), the global association of patient organisations for esophageal atresia, has been physically active throughout his life: "Running was for me a good way to maintain my overall fitness, maintain good mobility and provide opportunities to be outside in fresh air. It was also important in improving my lung function and this exercise helped combat the respiratory difficulties I experienced as a TOF/OA survivor."

Conflict of Interest

The authors declare that they have no conflict of interest.

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The KEKS Health Folder and the KEKS Portal

Der KEKS-Gesundheitsordner und das KEKS-Portal

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Keywords
PROM, Patient-Reported Outcome Measure, follow-up care, oesophageal atresia/esophageal atresia

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PROM, Patient-Reported Outcome Measure, Nachsorge, Ösophagusatresie

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ABSTRACT

The importance of lifelong, structured follow-up for corrected esophageal atresia led to the development of a follow-up book. This was updated and digitized in 2024. The KEKS-Portal allows families to complete the forms themselves, reflects the patient perspective, and trains parents in symptom assessment. The new digital forms serve as preparation for clinic appointments and as a “Patient-Reported Outcome Measure” (PROM). KEKS uses the data for consultations and research. Additionally, families receive a free health folder with information on esophageal atresia. These offerings are an important support for families and clinics to structure and improve follow-up care.

ZUSAMMENFASSUNG

Die Wichtigkeit einer lebenslangen, strukturierten Nachsorge einer korrigierten Ösophagusatresie führte zur Entwicklung eines Nachuntersuchungsbuches. Dieses wurde 2024 aktualisiert und digitalisiert. Das KEKS-Portal ermöglicht es Familien, die Bogen selbst auszufüllen, bildet die Betroffenenperspektive ab und schult Eltern in der Symptomeinschätzung. Die neuen digitalen Bogen dienen als Vorbereitung für Kliniktermine und als „Patient-Reported Outcome Measure“ (PROM). KEKS nutzt die Daten für Beratungen und Forschung. Ergänzend erhalten Familien einen kostenlosen Gesundheitsordner mit Informationen zur Ösophagusatresie. Diese Angebote sind eine wichtige Unterstützung für Familien und Kliniken, um die Nachsorge zu strukturieren und zu verbessern.

The Beginnings of Structured Follow-up Care – The KEKS Follow-up Examination Book

The preceding articles show the importance of lifelong follow-up care for a corrected esophageal atresia across different medical specialties. To provide clinics and families with a guide for successful follow-up care, the German patient organization for EA (KEKS) developed the KEKS Follow-up Examination Book in 2010 in collaboration with its scientific advisory board. This binder contained 15 questionnaires for follow-up examinations and a medical history that detailed the child’s initial situation before their first discharge. The follow-up clinics would fill out the forms during the examinations and send the results to the registry-managing institute (initially IMBI, Heidelberg). Since 2021, IMBEI in Mainz has been our partner institute, and the forms are first sent to KEKS.

Digitization of the Follow-up Examination Forms – The New KEKS Portal

From 2022 to 2024, KEKS fundamentally revised the follow-up examination book.

The main goal was to digitize the binder and to incorporate the patient’s perspective. For this purpose, KEKS edited the questionnaires so that they could be filled out by families themselves, independent of the follow-up examinations at the clinic.

The new forms are designed to represent the family’s perspective and to train parents to assess potential symptoms and their child’s current situation. Furthermore, the forms serve as a basis for preparation for the follow-up examinations in the clinic. This relieves the burden on doctors, and families can come to the appointment with prepared questions.

The questions focus on the child's current condition, considering everyday issues in the categories of general health, breathing, swallowing/thriving, sleeping/resting, treatments, therapies, support measures, results of early detection screenings (U-examinations), new diagnoses, and other topics (psychological support, overall situation). A total of 9 questionnaires are now filled out, starting with the medical history. This is followed by 8 follow-up examinations, which are timed to coincide with the U- and J-examinations. They should be carried out at the ages of 3, 6, 9, and 12 months, 2–3 years, 4–6 years, 9–10 years, and 16–17 years, respectively. These are routine follow-up examinations. If symptoms or problems arise, further consultations should take place at the follow-up clinic regardless of the recommended schedule. The last questionnaire (NU8) is completed by both the parents and the adolescents themselves, to prepare them to take responsibility for their own follow-up care.

This change also supports the patient-centered perspective that is becoming increasingly important in research. The new forms therefore serve as a Patient-Reported Outcome Measure (PROM). PROMs map the subjective health status of patients and make it measurable and comparable. They can help to improve quality of life and patient orientation in care [1]. KEKS also uses the data from the questionnaires as a basis for medical consultations and, at a later stage, for scientific studies.

Although the questionnaires were designed so that medical laypersons can fill them out easily, we advise that the medical history is best completed in the clinic before the first discharge, together with the treating professionals and the family. This way, any unclear terms can be clarified immediately. However, many families only receive the binder and access to the KEKS portal after being discharged from the clinic. To enable families to fill out the questionnaires completely, it is helpful to have a clear and complete doctor's or discharge letter. It cannot be ruled out that families will turn to the pediatrician's office with unclear medical terms.

The KEKS Health Folder

In addition to access to the portal and thus the follow-up examination forms, families receive a Health Folder. This contains information about esophageal atresia with materials for the family as well as for health and other professionals. It also provides space for the child's health records.

The folder includes recommendations for follow-up examinations, information for parents and affected individuals, e.g., information sheets on VACTERL, infection management, reflux management, degree of disability and care level, and starting external care. In addition, the folder contains information sheets for medical and educational professionals, e.g., for pediatric practices and family doctors, regarding the importance of lifelong follow-up care, information on infection management, and information sheets for daycare centres or schools. The appendix lists relevant literature on the topic of esophageal atresia.

We ask pediatric practices to talk to families about the Health Folder and the follow-up examinations and to point out their importance.

Families receive the folder and access to the KEKS portal free of charge and regardless of membership via KEKS The content of the folder is available for free download via the following QR code:

Importance of the KEKS Follow-Up Examination Book and the KEKS Health Folder

One important reason for creating a KEKS Follow-Up Book and the new KEKS Health Folder is that many families are confronted with scientifically unfounded statements about esophageal atresia and its consequences. The most common statements from healthcare professionals that families report to KEKS's medical advisory service are as follows:

- Esophageal atresia is cured with surgery.
- If the esophagus is sealed and wide, eating can't be a problem.
- A viral infection doesn't require antibiotics.
- Tracheomalacia will outgrow itself.
- Failure to thrive can be treated with more calories.
- Eating eliminates or prevents strictures.
- Coughing and low performance are normal.

The preceding articles clearly show that none of these "legends about esophageal atresia" are scientifically sound.

A Word from a Professional

"The follow-up examination book–in its proven and new versions–supports us doctors and the affected families in structuring follow-up care and including all aspects of the disease in every contact." – Prof. Dr. Michael Boettcher, Chief Physician of Pediatric Surgery, University Hospital Mannheim

Conflict of Interest

The authors declare that they have no conflict of interest.

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Thieme

Rehabilitation Treatment for Children and Adolescents with Corrected Esophageal Atresia

Rehabilitationsbehandlung für Kinder und Jugendliche mit korrigierter Ösophagusatresie

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Keywords

esophageal atresia/oesophageal atresia, rare disease, specialized rehabilitation programs, respiratory problems, group-oriented rehabilitation programs, holistic care

Schlüsselwörter

Ösophagusatresie, seltene Erkrankung, Lungenerkrankung, spezialisierte Rehabilitationsprogramme, gruppenorientierte Rehabilitation, ganzheitliche Betreuung

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ABSTRACT

Background Children with corrected esophageal atresia (EA) often suffer from pulmonary, nutritional, and psychosocial problems. Structured rehabilitation programs for this group are lacking.

Aim The aim was to develop a group-oriented concept to improve participation, resilience, and quality of life.

Methods In cooperation with the patient organization KEKS, Fachklinik Wangen established a four-week interdisciplinary program. Indications include recurrent infections, chronic cough, nutritional failure, reduced endurance, and high family stress. Interventions comprise medical optimization, physiotherapy, sports therapy, nutritional counseling, psychological support, educational measures, and structured caregiver training.

Results The first implementations showed high acceptance, improved physical resilience and psychosocial stability, and strong benefits from peer exchange.

Conclusion Specialized rehabilitation for EA represents an important and necessary addition to pediatric care.

ZUSAMMENFASSUNG

Hintergrund Kinder mit korrigierter Ösophagusatresie (ÖA) leiden häufig an pulmonalen, gastroenterologischen und psychosozialen Problemen. Bisher fehlen strukturierte Rehabilitationsprogramme.

Ziel Entwicklung eines gruppenorientierten Konzepts zur Verbesserung von Teilhabe, Belastbarkeit und Lebensqualität.

Methoden In Kooperation mit der Patientenorganisation KEKS wurde in der Fachklinik Wangen ein 4-wöchiges interdisziplinäres Programm etabliert. Indikationen umfassen rezidivierende Infekte, chronischen Husten, Gedeihstörung, Belastungsintoleranz sowie hohe familiäre Belastung. Therapieelemente sind medizinische Optimierung, Physio- und Sporttherapie, Ernährungstherapie, psychologische Unterstützung, Elternschulung und pädagogische Begleitung.

Ergebnisse Erste Durchführungen zeigten hohe Akzeptanz, Verbesserungen in körperlicher Belastbarkeit und psychosozialer Stabilität sowie großen Nutzen durch Peer-Austausch.

Schlussfolgerung Spezialisierte Rehabilitation für Ösophagusatresie ist sinnvoll und notwendig.

Introduction

Children living with corrected esophageal atresia (EA) and their families face significant burdens–both physical and emotional. The beginning of life for these children is often marked by intensive medical interventions, surgery, and prolonged hospital stays. This is followed by various therapeutic, sometimes invasive, procedures and ongoing medical appointments, both planned and emergency visits. These experiences leave their mark on the children themselves as well as on their parents and siblings.

The complexity of the malformation which directly and indirectly affects multiple organ systems, results in a high demand for therapy. It is therefore all the more remarkable that, to date, there are hardly any specialized rehabilitation programs for this patient group. While children with EA are indeed already being treated in rehabilitation facilities, this usually take places on an individual basis–for example, due to pulmonary or orthopedic issues.

Fachklinik Wangen has set itself the goal of developing structured, group-oriented rehabilitation programs for children and adolescents with EA and their families. The initiative originated from the patient organization KEKS.

Specific Indications for Admission to a Rehabilitation Clinic May Include

- Frequent and/or increasing pulmonary infections
- Chronic cough and airway obstruction
- Reduced physical resilience in daily life
- Nutritional problems and failure to thrive
- Increasing absences from school
- Impaired physical and psychological development
- High family stress levels

The therapeutic approach is based on close collaboration between physicians, psychologists, physiotherapists, sports therapists, educators, and nutritionists. Accompanying caregivers are trained as co-therapists through structured parent education. Rehabilitation measures are organized in a group format for a duration of four weeks, ideally with age-homogeneous participants.

Core Components of the Rehabilitation Concept

- Therapy plan, optimization of medical therapy (e.g. for reflux or bronchial hyperreactivity)
- Physiotherapy and respiratory therapy to improve lung function, mobilize secretions, and enhance breathing awareness
- Sports therapy to increase physical endurance and promote enjoyment of movement
- Psychological support
- Nutritional therapy (addressing swallowing problems, food impaction, selection of suitable foods, etc.)
- Training in relaxation techniques
- Development of strategies for everyday life
- Educational support
- Emergency management training
- Patient and caregiver education (e.g., how to manage reflux or food impaction, prevent infections, mobilize mucus, nutritional advice)
- Moderated group discussions to exchange best practices, separately for children and caregivers
- For caregivers additionally: sports and relaxation activities, psychological support

Fachkliniken Wangen is ideally suited for the rehabilitation of patients with EA. Pulmonology, nutritional counseling, sports therapy are already well established and work together routinely for other patient groups (e.g., cystic fibrosis and rare lung diseases). Training on the specific needs of patients with EA has already been implemented.

The interest in rehabilitation for EA patients is high, and in addition to the therapeutic program, the group setting offers valuable opportunities for peer exchange. The two first group program was successfully conducted, and the positive feedback underscores the need for specialized rehabilitation.

Key Goals of Rehabilitation

- Improving participation in everyday life, school, and social environments
- Promoting self-regulation and self-management
- Improving and/or restoring reduced quality of life
- Strengthen psychological resilience
- Adapt therapy to the current stage of development
- Foster healthy lifestyle habits in the family's daily life

Children with corrected EA need more than just acute medical care–they need targeted developmental support and psychosocial stability. Rehabilitation offers these children and their families precisely these opportunities.

Conflict of Interest

The authors declare that they have no conflict of interest.

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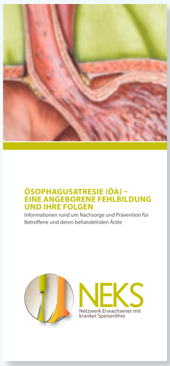
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