

# **ERNICA Consensus Conference On The Management Of Patients With Esophageal Atresia And Tracheoesophageal Fistula: Diagnostics, Preoperative, Operative And Postoperative Management**

Dingemann C<sup>1</sup>, Eaton S<sup>2</sup>, Aksnes G<sup>3</sup>, Bagolan P<sup>4</sup>, Cross KM<sup>5</sup>, De Coppi P<sup>5</sup>, Fruithof J<sup>6</sup>, Gamba P<sup>7</sup>, Husby S<sup>8</sup>, Koivusalo A<sup>9</sup>, Rasmussen L<sup>10</sup>, Sfeir R<sup>11</sup>, Slater G<sup>12</sup>, Svensson JF<sup>13</sup>, Van der Zee D<sup>14</sup>, Wessel L<sup>15</sup>, Widenmann-Grolig A<sup>16</sup>, Wijnen R<sup>17</sup>, Ure B<sup>1</sup>

<sup>1</sup>Department of Pediatric Surgery, Hannover Medical School, Hannover, Germany

<sup>2</sup>Paediatric Surgery & Metabolic Biochemistry, UCL Great Ormond Street Institute of Child Health, London, UK

<sup>3</sup>Department of Pediatric Surgery, Oslo University Hospital, Oslo, Norway

<sup>4</sup>Neonatal Surgery Unit, Bambino Gesù Children's Hospital-Research Institute Rome, Rome, Italy

<sup>5</sup>Specialist Neonatal and Paediatric Surgery, Great Ormond Street Hospital for Children, London, UK

<sup>6</sup>Esophageal Atresia and Tracheo-Esophageal Fistula Support Group VOKS, Lichtenvoorde, Netherlands

<sup>7</sup>Department of Pediatric Surgery, University of Padua, Padua, Italy

<sup>8</sup>Department of Pediatric Gastroenterology, Odense University Hospital, Odense, Denmark

<sup>9</sup>Department of Pediatric Surgery, University of Helsinki, Helsinki, Finland

<sup>10</sup>Department of Pediatric Surgery, Odense University Hospital, Odense, Denmark

<sup>11</sup>Department of Pediatric Surgery, Centre Hospitalier Régional Universitaire de Lille, Lille, France

<sup>12</sup>Esophageal Atresia and Tracheo-Esophageal Fistula Support Group TOFS, Nottingham, United Kingdom

<sup>13</sup>Department of Pediatric Surgery, Karolinska University Hospital, Stockholm, Sweden

<sup>14</sup>Department of Pediatric Surgery, Utrecht University Medical Center, Utrecht, Netherlands

<sup>15</sup>Department of Pediatric Surgery, University of Mannheim, Medical Faculty of Heidelberg, Mannheim, Germany

<sup>16</sup>Esophageal Atresia and Tracheo-Esophageal Fistula Support Group KEKS, Stuttgart, Germany

<sup>17</sup>Department of Pediatric Surgery, Erasmus MC Rotterdam, Rotterdam, Netherlands

## **Correspondence to:**

Carmen Dingemann; MD

Department of Pediatric Surgery, Hannover Medical School, Hannover, Germany

Email: [dingemann.carmen@mh-hannover.de](mailto:dingemann.carmen@mh-hannover.de)

Tel: 0049-511-532-9260

Fax: 0049-511-532-9059

## **Disclosure:**

Nothing to disclose.

ERNICA provided financial support for the conference.

SE, KMK and PDC gratefully acknowledge supported by the National Institute for Health Research Great Ormond Street Hospital Biomedical Research Centre. The views expressed are those of the authors and not necessarily those of the NHS, the NIHR or the UK Department of Health.

## **INTRODUCTION**

Esophageal atresia is a rare congenital condition with an estimated prevalence varying between 1 and 2 in 5,000 live births in Europe [1, 2]. The malformation is characterized by a discontinuity of the esophagus with or without tracheoesophageal fistula. Since the first successful primary repair of esophageal atresia in 1941, improvements in operative and perioperative care have led to better outcomes, and mortality has decreased to a single-digit range [3, 4, 5] with most deaths due to comorbidities. Evolution from concerns about mortality to concerns about morbidity and quality-of-life issues has occurred [5], and long-term morbidity remains high until adulthood [5, 6]. Morbidities include esophageal [7], gastrointestinal [8], pulmonary [9, 10] and various developmental [11] problems which may have a considerable impact on the quality of life of patients and their families [6, 12-16]. Therefore, esophageal atresia is no longer a mere neonatal surgical problem, but rather lifelong requires attention in individual patients [5, 17]. Well-designed clinical trials dealing with diagnostic and therapeutic concepts for patients with esophageal atresia are still scarce [3, 18-20] and generally accepted algorithms are lacking. As a result, there is a variety of co-existing protocols on the perioperative and surgical management of patients with esophageal atresia based on opinion rather than on evidence [21]. Diversity of concepts and the use of different operative techniques even for the same type of atresia have a significant impact on the course of the disease in single patients. The European Reference Network on Rare Inherited and Congenital Anomalies (ERNICA) has been established in response to the European Commission's call for the setup of European Reference Networks for rare diseases in 2017 [22]. ERNICA is one of 24 European Reference Networks co-funded by the European Union (Health Program), and involves teams from 20 European hospitals from 10 member states [22]. The mission is to promote optimal patient care for rare inherited and congenital digestive track-related disorders from pediatric age to adulthood providing high quality and accessible education, supporting research, improving clinical standards and services and reducing health inequalities in Europe [23-26]. The ERNICA Workstream Congenital Malformations and Diseases of the Esophagus is mainly focused on patients with esophageal atresia. During recent ERNICA meetings in Rotterdam (April 2017), Helsinki (November 2017), and Stockholm (April 2018) it became evident that protocols dealing with the management of esophageal atresia considerably differ even between ERNICA institutions. Therefore, the members of ERNICA agreed to establish consensus on all relevant aspects of the surgical management of patients with esophageal atresia as an urgent objective of ERNICA. The aim of this first ERNICA consensus conference was to establish consensus on the relevant aspects regarding the treatment of children with esophageal atresia and tracheoesophageal fistula based on expert opinions referring to the latest evidence from current literature. The conference was systematically prepared to include clinical and non-clinical members of ERNICA. In particular, representatives of several national

patient support groups, which have become full members of ERNICA [27, 28], have been invited to participate in the conference.

## **METHODS**

### ***Participants***

Members of the ERNICA Workstream Congenital Malformations and Diseases of the Esophagus were invited to take part in the preparation and implementation of the conference. In total, 19 participants originating from 15 institutions and nine countries confirmed participation. Three representatives from national patient support groups acting under the umbrella of the Federation of Esophageal Atresia and Tracheo-Esophageal Fistula Support Groups (EAT), [27] were included. Altogether 14 pediatric surgeons, one pediatric gastroenterologist, three representatives of patient support groups, and one non-surgeon pediatric surgery academic took part in all steps of the preparation and the conference itself. The conference took place in Berlin on the 25th and 26th October 2018. The preparation and implementation of the conference included three steps: (i) generation of a list of items; (ii) prioritization of the items; (iii) discussion of all items during the conference, formulation of statements; (iv) anonymous voting.

### Focus of the conference

The conference dealt exclusively with the management of patients with esophageal atresia with tracheoesophageal fistula who undergo primary anastomosis. Items dealt with the surgical, perioperative and long-term management of these patients, and the conference was focused on operative and perioperative aspects. The conference did not deal with the management of other forms of esophageal atresia such as long-gap atresia or those with secondary anastomosis. Item generation A systematic literature search was performed by CD and BU to identify relevant aspects in the management of esophageal atresia with tracheoesophageal fistula. A PubMed literature search was conducted for the years 2008 to 2018 using the keywords "esophageal/oesophageal atresia", "tracheoesophageal fistula", "diagnostics", "management", "surgery", "operation", "complications", "outcome", "experience", and "follow-up/follow up". In case of absent relevant literature, the time period was extended to include more historic literature. Reports on long-gap esophageal atresia only, non-English articles, case reports, and reviews without original patient data were excluded. After literature search, a preliminary item list was developed which was presented during an ERNICA conference in Stockholm on the 18th – 20th April 2018. Members of the Workstream Congenital Malformations and Diseases of the Esophagus discussed this list in detail. The list was modified according to this discussion by exclusion of some suggested items and inclusion of new items which were considered to be relevant. The list was distributed to all participants who were invited to revise the items and to test for plausibility. Finally, items were attributed to the following domains: (i) Diagnostics; (ii) Preoperative management after confirmation of diagnosis; (iii) Operative management; (iv) Postoperative management; (v) Follow-up; (vi)

Miscellaneous. The consensus results and discussion for the domains Diagnostics, Preoperative, Operative and Postoperative management are presented in this manuscript, the domains Follow-up and Miscellaneous will be addressed in a separate manuscript as the methodological approaches differed. Item prioritization The online REDCap electronic data capture tools [29] hosted at University College London was used for prioritization of the items. Priority was assigned to each item by all participants using a five point Likert scale (1 as the highest priority and 5 as the lowest priority). Scoring each item was compulsory. Thereafter, the final list of items to be discussed at the conference was set up by CD, BU and SE, taking the prioritization into account. The threshold for exclusion or inclusion of items was scoring as “lowest” or “highest” priority respectively by two or more participants; entirely new items were added after suggestion by two or more participants independently. Identification of relevant literature for item discussion Prior to the conference, two participants were allocated to each domain (domain leaders). Domain leaders performed a literature research and identified the most relevant publications for each item of their domain. Publications with the highest grade of evidence according to the CEBM (Centre for Evidence-Based Medicine) classification were suggested to be preferred. The highest grade of evidence was defined as Level 1 evidence derived from studies with a “high” quality of evidence (“further research is unlikely to change our confidence in the estimate of effect”). Level 1 evidence was derived from systematic reviews (with homogeneity) or metaanalyses of randomized controlled trials (CEBM Level 1a), a well-designed individual randomized controlled trial (with narrow confidence interval; CEBM Level 1b), or all or none randomized controlled trials (CEBM Level 1c) [30]. Literature was pooled by CD. After sorting, it was distributed and made available to all participants via a DropBox link prior to the conference. Conference, voting and consensus The final list of domains, items and references was accessible to all participants via an online link prior to and throughout the conference. Each domain was discussed in a separate session. Domain leaders introduced the literature and relevant aspects on single items. Subsequently, participants discussed each item and formulated a statement to facilitate voting. The wording of the statements evolved during group discussion. Participants voted on each statement via the internet-based system VoxVote [31]. An event code was provided and participants were able to vote using either an internet browser, or dedicated apps for Android or iOS. The wordings of the statements on items were updated during the discussion by the non-surgical academic (SE) who did not vote. Participants were able to vote using a 1-9 scale (9 meaning ‘fully agree’ and 1 ‘strongly disagree’). Participants were suggested not to vote on an individual item when they felt that it was not an item that they had expertise or an opinion on. Therefore, participants were allowed to vote online for ‘no relevant expertise on this statement’. As a result, the number of scoring participants varied for single statements. Consensus was defined as  $\geq 75\%$  of those voting scored 6, 7, 8 or 9, excluding those who declared no relevant expertise

on that statement. The voting was anonymous and scores were not visible to the participants during the voting process. The final scores and the consensus results were shown to all participants after all votes were obtained, but individual scores remained anonymous. The details of the discussions, in particular the controversial aspects, were documented throughout the conference by CD.

## **RESULTS AND CONSENSUS STATEMENTS**

**Item generation and prioritization** The systematic literature search and the discussion of the members of the ERNICA Workstream Congenital Malformations and Diseases of the Esophagus during the ERNICA conference in Stockholm on the 18th – 20th April 2018 resulted in a total of 41 items. After the online prioritization phase, two items were excluded. Following the participants' suggestions, 7 items were added as new items. Consequently, the list included 46 items prior to the conference, for which literature was obtained and circulated. As a result of the presentations by the domain leaders and discussion during the conference, 5 items were excluded, and 11 items were added as new items as some items were split into several separate questions. Finally, 52 items were confirmed for voting and included six items in the domain Diagnostics, eight items in the domain Preoperative Management, 28 items in the domain Operative Management, and ten items in the domain Postoperative Management [Table 1a-d].

**Relevant literature** A total of 116 relevant manuscripts were selected by literature search conducted by all participants [see online Supplement]. Ten of these were studies had been identified for several items, and ten were quoted for more than one domain. In summary, 18 articles addressed the domain Diagnostics, 30 articles Preoperative Management, 63 Operative Management, and 17 Postoperative Management. CEBM Level 1 evidence was only available for four items (7.7%) and included four single randomized controlled trials and one pilot randomized trial which represented 4.3% of all selected articles [Table 2]. Ten Level 2 evidence studies (8.6%), 26 Level 3 evidence studies (22.4%), 43 Level 4 evidence studies (37.1%), and 11 Level 5 evidence studies (9.5%) were also included. Moreover, 15 review articles (12.9%) and 6 book chapters (5.2%) have been considered to be relevant for the discussion, despite not including original data [Figure 1].

**Consensus** Total agreement, defined as 100% consensus amongst voters, was achieved on 20 items (38%), and consensus ( $\geq 75\%$  of those voting having scored 6-9) on 37 items (71%). Thirteen items (25%) were particularly controversial in that the votes ranged from 1-9; in eight of these no consensus was reached. Detailed results are summarized in Table 1a-d.

**Abstention** Participants were supposed not to vote when they felt a lack of competence. One or more participants declared 'no relevant expertise on this statement' on the online voting system on 48 (92.3%) questions; for 3 (5.8%) questions, one participant abstained; for 2 (3.8%) questions, two participants abstained; in 21 questions (40.4%) three participants abstained; in 20 (38.5%) processes, four participants abstained; in 2 (3.8%), five participants abstained from voting. Controversial items discussed

without voting Several items were discussed that were controversial and the participants agreed verbally that more data from future research would have to be available before a meaningful question and vote could be conducted [Table 3]. The discussion included in particular the following aspects: Diagnostics The participants agreed that magnetic resonance imaging represents an important tool in identifying anomalies of the aortic arch and its branches, and might be considered as the imaging technique of choice when planning surgical management, especially in cases of associated cardiac anomalies. However, as the evidence from the literature [37] and personal experience on the routine use of magnetic resonance imaging in the preoperative work-up of esophageal atresia patients was limited, it was decided not to vote on this item. Preoperative Management Several participants postulated that preoperative measurement of the esophageal gap length may promote comparability between centers, may reduce the incidence of unsuspected difficult cases, and may contribute to a general definition of long-gap esophageal atresia [38, 39]. After extensive discussion it was decided not to vote on a statement on preoperative gap measurement as a routine procedure due to the lack of data and literature on this item. Operative Management A vote on routine posterior tracheopexy during primary repair of esophageal atresia was rejected. Most participants felt that the available data were scarce [40, 41], and the experience with this new technique remained too limited to allow a meaningful vote. Postoperative Management The management of anastomotic leakage was discussed extensively. No consensus was achieved on surgical revision of anastomotic leakage within the first 4 postoperative days. Vaghela et al. [36] presented data from a randomized controlled trial on the application of glycopyrrolate as a therapeutic option for patients with postoperative anastomotic leakage. However, despite this evidence the conference participants felt that is too early to vote on the role of glycopyrrolate. Participants supported the concept that a clinical checklist of tasks and examinations should be compiled before initial hospital discharge, including abdominal and renal ultrasound and resuscitation training for caregivers [Table 1d]. The full content of such a list remained a matter of discussion and agreement could not be achieved.

## **DISCUSSION**

According to the Council of Europe a Medical consensus is a public statement on a particular aspect of medical knowledge that is generally agreed upon as an evidence based, state-of-the-art knowledge by a representative group of experts in that area [42]. Its main objective is to counsel physicians on the best possible and acceptable way to address a particular decision-making area for diagnosis, management or treatment [43]. Most recently, multiple consensus conferences have been organized both in the field of adult [44, 45] and pediatric medicine [46, 47]. Importantly for esophageal atresia, Krishnan et al. published in 2016 the ESPGHANNASPGHAN guidelines for the evaluation and treatment of gastrointestinal and nutritional complications in children with esophageal atresia and tracheoesophageal fistula

[19]. The guidelines were set up during two consensus meetings using the nominal voting technique. Expert opinion was used where no randomized controlled trials were available to support the recommendations [19]. This consensus statement focused on the medical aspects of follow-up, and few surgical aspects were included. We hereby present for the first time the results of a consensus conference focused on surgical aspects of the management of patients with esophageal atresia with tracheoesophageal fistula. In line with the ESPGHAN-NASPGHAN guidelines, this conference was based on two keystones: (i) on expert opinion, and (ii) on evidence from literature. Modern medicine increasingly places emphasis on evidence-based medicine [48], defined by Sackett et al. as "(...) the conscientious, explicit and judicious use of current best evidence in making decisions about the care of individual patients" [49]. Nonetheless, the paucity of high level evidence in the literature on pediatric surgical procedures was highlighted in 1999, when Hardin et al. [50] reported that only 0.3% of the literature relevant for pediatric surgery consisted of CEBM Level 1 evidence studies. In 2010, Ostlie and St. Peter [51] demonstrated that prospective articles represented less than 0.05% of all pediatric surgical literature. In line with these findings, literature meeting the criteria of CEBM Level 1 evidence was only available for four out of 52 items (7.7%) that were discussed at this conference. Consequently, the votes of the participants of the conference were based on a combination of lower level evidence (e.g. retrospective reviews) combined with expert opinion. The voting resulted in 100% consensus for 20 (38%) of items. This indicates a considerable level of agreement but also a certain diversity of opinions among specialists in the field of esophageal atresia. However, general consensus (defined by  $\geq 75\%$  of votes scoring higher 6-9) was achieved for 71% of items suggesting predominantly homogeneous approaches in ERNICA institutions. A maximum range of voting from 1-9, indicating widely diverse opinions, was evident for only 25% of cases. Interestingly, consensus was achieved on most, but not on all items with Level 1 evidence in the literature. Askarpour et al. recently confirmed better outcomes of muscle-sparing versus standard posterolateral thoracotomy in newborns [32]. This statement was adopted unanimously by the participants of the conference. However, advantages of the preservation of the azygos during esophageal atresia repair were confirmed in two randomized trials [33, 34] but no consensus could be achieved on this topic. Several participants questioned the quality of both trials and as a result consensus was not achieved. Dingemann and Ure [52] reported on low CEBM levels of evidence in the field of endoscopic pediatric surgery. Dingemann et al. [21] also demonstrated that only retrospective comparative studies pursuant to CEBM Level 3 were available on pediatric video-assisted thoracoscopic surgery. With regard to the management of esophageal atresia, it has been stated that the best available evidence comparing the minimally invasive versus the open approach for esophageal atresia repair was CEBM Level 3 [53]. Since then, minimally invasive esophageal atresia repair has been subject of numerous studies [54-62]. Despite the low level

of evidence of these reports, consensus was achieved on several advantages of thoracoscopic esophageal atresia repair based on the experience of the participants. One pilot randomized controlled trial on 20 neonates dealt with potential disadvantages of the technique [35]. Patients with congenital diaphragmatic hernia but not those with esophageal atresia had severe intraoperative and prolonged hypercapnia and acidosis. However, it should be emphasized that pulmonary compromised patients had been included in this study as well as the intraoperatively applied insufflation pressures were rather high which could explain the described poor outcome. Taking this into account, the participants of the conference also felt that the numbers of patients of this pilot trial were too small to draw valid conclusions on management of infants with esophageal atresia. The management of anastomotic leakage is still the subject of ongoing discussions [63, 64]. Vaghela et al. [36] recommended the application of glycopyrrolate as a therapeutic option. Despite these results, the conference participants felt that is too early to vote on the role of glycopyrrolate. In 2014, the EUPSA international survey on the management of esophageal atresia demonstrated that approximately 60% of respondents measure the gap intra-, but not pre- operatively [18]. After a controversial discussion, it was decided not to vote on a statement on preoperative gap measurement as a routine procedure. The issue of a routinely performed posterior tracheopexy during primary repair of esophageal atresia was a matter of debate. Shieh et al. concluded from their experience with 118 patients that posterior tracheopexy should be selectively considered at the time of initial repair [40]. Tytgat et al. recently presented nine patients who underwent thoracoscopic posterior tracheopexy during primary esophageal atresia repair [41]. The authors postulated that this technique prevents potential sequelae of mild to severe tracheomalacia and a second surgical procedure. The participants felt that evidence was too low to allow voting and that potential disadvantages of the technique remain unclear. A clinical checklist may serve as an aid for clinicians not to miss essential measures before discharge. Recent literature provides an example for such similar standardized checklist [65]. Conference participants strongly supported the concept of a checklist of tasks and examinations to be used before the first discharge. However, consensus could not be achieved on which items should be included, except for abdominal and renal ultrasounds and resuscitation training for caregivers. The strength of this conference is the pool of participating specialists with extensive expertise in the field. The multidisciplinary approach allowed discussing items from various perspectives. An additional advantage of this conference was the methodology characterized by meticulous item generation and prioritization, systematic literature search and anonymous voting ensuring a high quality of results. The ability to modify wording of questions flexibly and rapidly via the online voting system was an advantage, which allowed consensus to be reached on 37 items, with a further 15 voted on without consensus. The approach that we used might be advocated for other surgical consensus meetings, in which there might be a somewhat



higher number of (often technical) items than for medical consensus meetings in which consensus statements might be generated on, for example, a treatment approach. Abstention from voting in case of lacking expertise additionally indicates a high validity of votes. It may be speculated that representatives of the patient support groups and the pediatric gastroenterologist refrained from voting in cases of entirely surgical issues. Finally, representatives of patient support groups were able to contribute to the discussions and voting process from the patients' perspective. The items on which either no consensus was reached, or where it was felt that there was insufficient evidence to hold a meaningful vote, could be suggested to be future areas which are priorities for future research. There are several drawbacks of the conference. As the conference was focused on pediatric surgeons, only one pediatric gastroenterologist and no pediatric pulmonologist, pediatric anesthesiologist and neonatologist were involved. Numerous perioperative aspects may have been discussed differently by representatives from non-operative fields. Secondly, the results of the conference were mainly based on expert opinion. Therefore, the statements of conference participants must be considered as opinions of ERNICA representatives only. **CONCLUSION** Participants of this ERNICA conference reached significant consensus on the pre-, peri- and postoperative management of patients with esophageal atresia and tracheoesophageal fistula who undergo primary anastomosis. The consensus may facilitate standardization and development of generally accepted guidelines for patient care. The conference methodology may serve as a blueprint for further conferences on the management of other congenital malformations in pediatric surgery.

## REFERENCES

- 1 Pedersen RN, Calzolari E, Husby S, Garne E.; EUROCAT Working group. Oesophageal atresia: prevalence, prenatal diagnosis and associated anomalies in 23 European regions. *Arch Dis Child* 2012; 97 (03) 227-232.
- 2 Cassina M, Ruol M, Pertile R., et al. Prevalence, characteristics, and survival of children with esophageal atresia: a 32-year population-based study including 1,417,724 consecutive newborns. *Birth Defects Res A Clin Mol Teratol* 2016; 106 (07)542-548.
- 3 Zimmer J, Eaton S, Murchison LE, Ure BM, Dingemann C. State of Play: Eight Decades of Surgery for Esophageal Atresia *Eur J Pediatr Surg.* 2018.
- 4 Morini F, Conforti A, Bagolan P. Perioperative Complications of Esophageal Atresia.*Eur J Pediatr Surg.* 2018;28(2):133-140.
- 5 Wijnen RM, Ure B. Bridging the Gap-More than Surgery Only. *Eur J Pediatr Surg.* 2015;25(4):311.
- 6 Dingemann J, Szczepanski R, Ernst G, et al. Transition of Patients with Esophageal Atresia to Adult Care: Results of a Transition-Specific Education Program. *Eur J Pediatr Surg.* 2017;27(1):61-67. Erratum in: *Eur J Pediatr Surg.* 2017b;27(1):e1-e2.
- 7 Rayyan M, Allegaert K, Omari T, Rommel N. Dysphagia in Children with Esophageal Atresia: Current Diagnostic Options. *Eur J Pediatr Surg.* 2015;25(4):326-32. Review.
- 8 Acher CW, Ostlie DJ, Leys CM, Struckmeyer S, Parker M, Nichol PF. Long- Term Outcomes of Patients with Tracheoesophageal Fistula/Esophageal Atresia: Survey Results from Tracheoesophageal Fistula/Esophageal Atresia Online Communities. *Eur J Pediatr Surg.* 2016;26(6):476-480.
- 9 Nurminen P, Koivusalo A, Hukkinen M, Pakarinen M. Pneumonia after Repair of Esophageal Atresia-Incidence and Main Risk Factors. *Eur J Pediatr Surg.* 2018.
- 10 Snijders D, Barbato A. An Update on Diagnosis of Tracheomalacia in Children. *Eur J Pediatr Surg.* 2015;25(4):333-5. Review.
- 11 Harmsen WJ, Aarsen FJ, van der Cammen-van Zijp MHM, et al. Developmental problems in patients with oesophageal atresia: a longitudinal follow-up study. *Arch Dis Child Fetal Neonatal Ed.* 2017;102(3):F214-F219.
- 12 Witt S, Dellenmark-Blom M, Dingemann J, et al. Quality of Life in Parents of Children Born with Esophageal Atresia. *Eur J Pediatr Surg.* 2018.
- 13 Flieder S, Dellenmark-Blom M, Witt S, et al. Generic Health-Related Quality of Life after Repair of Esophageal Atresia and Its Determinants within a German- Swedish Cohort. *Eur J Pediatr Surg.* 2019;29(1):75-84.
- 14 Witt S, Dellenmark-Blom M, Flieder S, et al. Health-related quality of life experiences in children and adolescents born with esophageal atresia: A Swedish-German focus group study. *Child Care Health Dev.* 2019;45(1):79- 88.
- 15 Witt S, Dellenmark-Blom

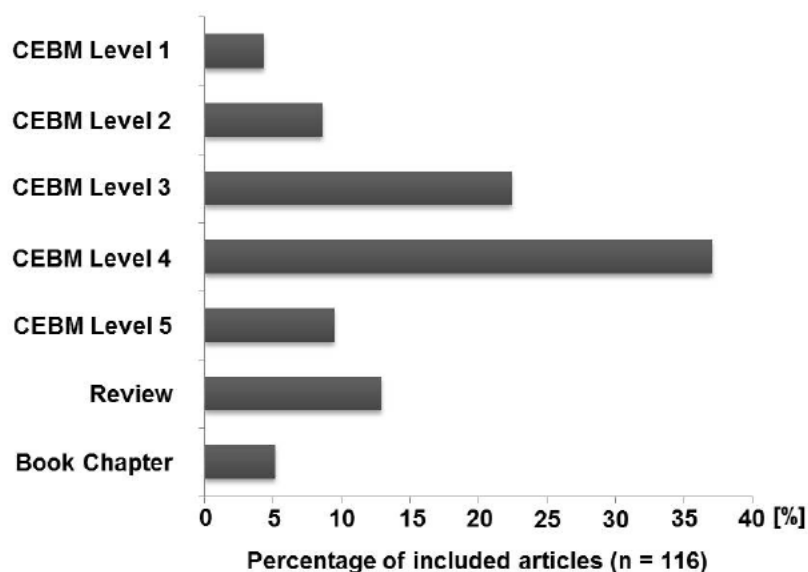
M, Dingemann J, Dingemann C, Ure BM, Gomez B, Bullinger M, Quitmann J. Quality of Life in Parents of Children Born with Esophageal Atresia. *Eur J Pediatr Surg.* 2018. 16 Rozensztrauch A, Śmigiel R, Patkowski D. Congenital Esophageal Atresia- Surgical Treatment Results in the Context of Quality of Life. *Eur J Pediatr Surg.* 2018. 17 Vergouwe FW, IJsselstijn H, Wijnen RM, Bruno MJ, Spaander MC. Screening and Surveillance in Esophageal Atresia Patients: Current Knowledge and Future Perspectives. *Eur J Pediatr Surg.* 2015;25(4):345-52. 8 Zani A, Eaton S, Hoellwarth ME, et al. International survey on the management of esophageal atresia. *Eur J Pediatr Surg.* 2014;24(1):3-8. 19 Krishnan U, Mousa H, Dall'Oglio L, et al. ESPGHAN-NASPGHAN Guidelines for the Evaluation and Treatment of Gastrointestinal and Nutritional Complications in Children With Esophageal Atresia-Tracheoesophageal Fistula. *J Pediatr Gastroenterol Nutr.* 2016 ;63(5):550-570. 20 Lal D, Miyano G, Juang D, Sharp NE, St Peter SD. Current patterns of practice and technique in the repair of esophageal atresia and tracheoesophageal fistula: an IPEG survey. *J Laparoendosc Adv Surg Tech A.* 2013;23(7):635-8. 21 Dingemann C, Ure B, Dingemann J. Thoracoscopic procedures in pediatric surgery: what is the evidence? *Eur J Pediatr Surg.* 2014;24(1):14-9. 22 Available at: <https://ern-ernica.eu/about/european-reference-networks/>. Accessed February 25, 2019. 23 Wijnen R, Anzelewicz SM, Petersen C, Czauderna P. European Reference Networks: Share, Care, and Cure-Future or Dream? *Eur J Pediatr Surg.* 2017;27(5):388-394. 24 Rolle U. Centralization of Pediatric Surgery: European Perspective. *Eur J Pediatr Surg.* 2017;27(5):387. 25 Pakarinen M, Bjørland K, Qvist N, Wester T. Centralized Pediatric Surgery in the Nordic Countries: A Role Model for Europe? *Eur J Pediatr Surg.* 2017;27(5):395-398. 26 Héon-Klin V. European Reference networks for rare diseases: what is the conceptual framework? *Orphanet J Rare Dis.* 2017;12(1):137. 27 Available at: <http://www.we-are-eat.org/>. Accessed February 25, 2019. 28 Svoboda E, Fruithof J, Widenmann-Grolig A, et al. A patient led, international study of long term outcomes of esophageal atresia: EAT 1. *J Pediatr Surg.* 2018;53(4):610-615. 29 Harris PA, Taylor R, Thielke R, Payne J, Gonzalez N, Conde JG. Research electronic data capture (REDCap)--a metadata-driven methodology and workflow process for providing translational research informatics support. *Journal of biomedical informatics* 2009; 42: 377-381 30 OCEBM Levels of Evidence Working Group. Levels of Evidence (March 2009)". Oxford Centre for Evidence-Based Medicine. Available at: <http://www.cebm.net/index.aspx?o1/41025> 31 Available at: [www.voxvote.com](http://www.voxvote.com).

Accessed February 25, 2019. 32 Askarpour S, Peyvasteh M, Ashrafi A, Dehdashtian M, Malekian A, Aramesh MR. Muscle-sparing versus standard posterolateral thoracotomy in neonates with esophageal atresia. *Arq Bras Cir Dig*. 2018;31(2):e1365. 33 Upadhyaya VD, Gangopadhyaya AN, Gopal SC, et al. Is ligation of azygos vein necessary in primary repair of tracheoesophageal fistula with esophageal atresia? *Eur J Pediatr Surg*. 2007;17(4):236-40. 34 Sharma S, Sinha SK, Rawat JD, Wakhlu A, Kureel SN, Tandon R. Azygos vein preservation in primary repair of esophageal atresia with tracheoesophageal fistula. *Pediatr Surg Int*. 2007;23(12):1215-8. 35 Bishay M, Giacomello L, Retrosi G, et al. Hypercapnia and acidosis during open and thoracoscopic repair of congenital diaphragmatic hernia and esophageal atresia: results of a pilot randomized controlled trial. *Ann Surg*. 2013;258(6):895-900. 36 Vaghela MM, Mahajan JK, Sundram J, Bhardwaj N, Rao KL. Role of glycopyrrolate in healing of anastomotic dehiscence after primary repair of esophageal atresia in a low resource setting - A randomized controlled study. *J Pediatr Surg*. 2017 ;52(3):420- 423. 37 Cantinotti M, Hegde S, Bell A, Razavi R. Diagnostic role of magnetic resonance imaging in identifying aortic arch anomalies. *Congenit Heart Dis*. 2008;3(2):117-23. 38 Bagolan P, Valfrè L, Morini F, Conforti A. Long-gap esophageal atresia: traction-growth and anastomosis - before and beyond. *Dis Esophagus*. 2013;26(4):372-9. 39 Gross ER, Reichstein A, Gander JW, Stolar CJ, Coran AG, Cowles RA. The role of fiberoptic endoscopy in the evaluation and management of long gap isolated esophageal atresia. *Pediatr Surg Int*. 2010;26(12):1223-7. 40 Shieh HF, Smithers CJ, Hamilton TE, et al. Posterior Tracheopexy for Severe Tracheomalacia Associated with Esophageal Atresia (EA): Primary Treatment at the Time of Initial EA Repair versus Secondary Treatment. *Front Surg*. 2018;4:80. 41 Tytgat SHAJ, van Herwaarden-Lindeboom MYA, van Tuyll van Serooskerken ES, van der Zee DC. Thoracoscopic posterior tracheopexy during primary esophageal atresia repair: a new approach to prevent tracheomalacia complications. *J Pediatr Surg*. 2018;53(7):1420-1423. 42 Council of Europe. Developing a methodology for drawing up guidelines on best medical practice- Recommendation Rec(2001)13 and explanatory memorandum, Council of Europe Publishing, Strasbourg (2002). 43 De Boeck K, Castellani C, Elborn JS; ECFS Board. Medical consensus, guidelines, and position papers: a policy for the ECFS. *J Cyst Fibros*. 2014;13(5):495-8. 44 Cenzato M, Boccardi E, Beghi E, et al. European consensus conference on unruptured brain AVMs treatment (Supported by EANS, ESMINT, EGKS, and SINCH). *Acta Neurochir (Wien)*. 2017;159(6):1059-1064.

45 Jeyarajah DR, Berman RS, Doyle MB, et al. Consensus Conference on North American Training in Hepatopancreaticobiliary Surgery: A Review of the Conference and Presentation of Consensus Statements. *Am J Transplant*. 2016;16(4):1086-93. 46 Goldstein B, Giroir B, Randolph A; International Consensus Conference on Pediatric Sepsis. International pediatric sepsis consensus conference: definitions for sepsis and organ dysfunction in pediatrics. *Pediatr Crit Care Med*. 2005;6(1):2-8.Review. 47 Galli E, Neri I, Ricci G, et al. Consensus Conference on Clinical Management of pediatric Atopic Dermatitis. *Ital J Pediatr*. 2016;42:26. Review. 48 Zani-Ruttenstock E, Zani A, Bullman E, Lapidus-Krol E, Pierro A. Are paediatric operations evidence based? A prospective analysis of general surgery practice in a teaching paediatric hospital. *Pediatr Surg Int*. 2015;31(1):53-9. 49 Sackett DL, Rosenberg WM, Gray JA, Haynes RB, Richardson WS. Evidence based medicine: what it is and what it isn't. *BMJ*. 1996;312(7023):71-2. 50 Hardin WD Jr, Stylianos S, Lally KP. Evidence-based practice in pediatric surgery. *J Pediatr Surg*. 1999;34(5):908-12. 51 Ostlie DJ, St Peter SD. The current state of evidence-based pediatric surgery. *J Pediatr Surg*. 2010;45(10):1940-6. Review. 52 Dingemann J, Ure BM. Systematic review of level 1 evidence for laparoscopic pediatric surgery: do our procedures comply with the requirements of evidence-based medicine? *Eur J Pediatr Surg*. 2013;23(6):474-9. Review. 53 Dingemann C, Ure BM. Minimally invasive repair of esophageal atresia: an update. *Eur J Pediatr Surg*. 2013;23(3):198-203. Review. 54 Bastard F, Bonnard A, Rousseau V, et al. Thoracic skeletal anomalies following surgical treatment of esophageal atresia. Lessons from a national cohort. *J Pediatr Surg*. 2018;53(4):605-609. 55 Wei S, Saran N, Emil S. Musculoskeletal deformities following neonatal thoracotomy: long- term follow-up of an esophageal atresia cohort. *J Pediatr Surg*. 2017;52(12):1898-1903. 56 Lawal TA, Gosemann JH, Kuebler JF, Glüer S, Ure BM. Thoracoscopy versus thoracotomy improves midterm musculoskeletal status and cosmesis in infants and children. *Ann Thorac Surg*. 2009;87(1):224-8. 57 Wu Y, Kuang H, Lv T, Wu C. Comparison of clinical outcomes between open and thoracoscopic repair for esophageal atresia with tracheoesophageal fistula: a systematic review and meta-analysis. *Pediatr Surg Int*. 2017;33(11):1147-1157. 58 Yang YF, Dong R, Zheng C, et al. Outcomes of thoracoscopy versus thoracotomy for esophageal atresia with tracheoesophageal fistula repair: A PRISMA-compliant systematic review and meta-analysis. *Medicine (Baltimore)*. 2016;95(30):e4428. 59 Costerus S, Vlot J, van Rosmalen J, Wijnen R, Weber F. Effects of Neonatal Thoracoscopic Surgery on Tissue

Oxygenation: A Pilot Study on (Neuro-) Monitoring and Outcomes. *Eur J Pediatr Surg.* 2017. 60 Tytgat SH, van Herwaarden MY, Stolwijk LJ, et al. Neonatal brain oxygenation during thoracoscopic correction of esophageal atresia. *Surg Endosc.* 2016;30(7):2811-7. 61 Zani A, Lamas-Pinheiro R, Paraboschi I, et al. Intraoperative acidosis and hypercapnia during thoracoscopic repair of congenital diaphragmatic hernia and esophageal atresia/tracheoesophageal fistula. *Paediatr Anaesth.* 2017;27(8):841-848. 62 Soliman HA, Faure C, Berubé G, Mac-Thiong JM, Barchi S, Parent S. Prevalence and natural history of scoliosis and associated congenital vertebral anomalies in patients operated for esophageal atresia with or without tracheoesophageal fistula. *J Pediatr Surg.* 2018. 63 Zhao R, Li K, Shen C, Zheng S. The outcome of conservative treatment for anastomotic leakage after surgical repair of esophageal atresia. *J Pediatr Surg.* 2011; 46: 2274-8. 64 Zhu H, Shen C, Xiao X, Dong K, Zheng S. Reoperation for anastomotic complications of esophageal atresia and tracheoesophageal fistula. *J Pediatr Surg.* 2015;50(12):2012-5. 65 Soong C, Daub S, Lee J, et al. Development of a checklist of safe discharge practices for hospital patients. *J Hosp Med.* 2013;8(8):444-9.

**Figure 1: CEBM Level of Evidence of articles considered as relevant for the consensus statements** In accordance with the Oxford CEBM Levels of Evidence as published in 2009 [30]



**Table 1a: Diagnostic procedures in patients with suspected esophageal atresia**

	DIAGNOSTICS	Consensus	%	Votes	Median [range]
1	A nasogastric tube 10Fr or larger (modified for preterm infants) should be routinely inserted as a diagnostic procedure in cases with suspected EA.	+	100	15/15	9 [7-9]
2	A thoracoabdominal X-ray should be routinely performed as a preoperative diagnostic procedure.	+	100	15/15	9 [8-9]
3	An ultrasound of the abdomen (including kidney/urinary tract) should be routinely performed as a preoperative diagnostic procedure.	-	40	6/15	3 [1-9]
4	An ultrasound of the spine should be routinely performed as a preoperative diagnostic procedure.	-	6.7	1/15	1 [1-9]
5	Echocardiography should be routinely performed as a preoperative diagnostic procedure, especially to exclude a right descending aorta.	+	100	15/15	9 [7-9]
6	A contrast-study of the upper esophageal pouch should be routinely performed as a preoperative diagnostic procedure.	-	13.3	2/15	1 [1-9]

**Table 1b: Preoperative management of patients with esophageal atresia after confirmation of the diagnosis before transfer to the operation theatre**

	PREOPERATIVE MANAGEMENT	Consensus	%	Votes	Median [range]
7	A repleg tube should be routinely placed into the upper esophageal pouch to allow continuous low pressure suction.	+	100	15/15	9 [6-9]
8	Preoperative antibiotic prophylaxis should be routinely administered as soon as the diagnosis is established.	-	13.3	2/15	2 [1-9]
9	Spontaneous breathing should routinely be favoured.	+	100	15/15	9 [9-9]
10	If assisted ventilation is required, preference should be given to intubation rather than to non-invasive ventilation.	+	100	15/15	9 [8-9]
11	Tracheobronchoscopy under spontaneous breathing should be performed preoperatively to evaluate tracheomalacia.	-	53.3	8/15	6 [2-9]
12	A central venous line should be routinely placed preoperatively.	-	14.3	2/14	2 [1-7]
13	An arterial line should be routinely placed preoperatively.	-	7.1	1/14	1 [1-8]
14	During preoperative counselling parents should be routinely informed about different surgical options such as open and thoracoscopic repair.	+	94.4	17/18	9 [2-9]

**Table 1c:**

## Operative management of patients with esophageal atresia

	OPERATIVE MANAGEMENT	Consensus	%	Votes	Median [range]
15	A stable neonate with EA should preferably be operated during working hours during the week.	+	94.4	17/18	9 [3-9]
16	Antibiotics should be routinely administered perioperatively.	+	100	14/14	9 [8-9]
17	A central venous line should be placed before the operation.	+	93.3	14/15	9 [1-9]
18	An arterial line should be placed before the operation.	+	78.8	11/14	8 [1-9]
19	Tracheoscopy should be routinely performed before the operation to evaluate the fistula(s) and other tracheolaryngeal pathology.	+	94.1	16/17	9 [2-9]
20	Horizontal or vertical or U-shaped (Bianchi) approaches (skin incision) are viable approaches for conventional thoracotomy.	+	100	15/15	9 [7-9]
21	Muscle-sparing approach is the recommended approach for conventional thoracotomy.	+	100	15/15	9 [8-9]
22	Entry through the 4 <sup>th</sup> intercostal space is the recommended approach for conventional thoracotomy.	+	100	14/14	9 [7-9]
23	The extrapleural approach is the preferred approach for thoracotomy.	+	92.9	13/14	9 [5-9]
24	In cases with suspected right descending aorta, a right-sided thoracic approach is the first option.	+	76.9	10/13	8 [1-9]
25	The azygos vein should be preserved whenever possible.	-	71.4	10/14	6.5 [2-9]
26	The tracheoesophageal fistula should preferably be closed by transfixing suture.	+	100	14/14	9 [6-9]
27	The esophageal anastomosis should be preferably performed with absorbable sutures.	+	85.7	12/14	8 [1-9]
28	The esophageal anastomosis should be preferably performed with interrupted sutures.	+	100	14/14	9 [6-9]
29	A transanastomotic tube should be routinely inserted.	+	80	12/15	8 [1-9]
30	A chest drain should be routinely placed.	-	21.4	3/14	1 [1-9]
31	The thoracoscopic approach is a viable option.	+	87.5	14/16	9 [5-9]
32	The thoracoscopic approach should be only performed if suitable expertise is available.	+	100	17/17	9 [6-9]
33	The thoracoscopic approach offers the advantage of magnification compared to the conventional approach.	+	92.9	13/14	9 [5-9]
34	The thoracoscopic approach offers the advantage of faster recovery compared to the conventional approach.	-	53.3	8/15	6 [1-9]
35	The thoracoscopic approach offers the advantage of better cosmesis compared to the conventional approach.	+	94.1	16/17	9 [5-9]
36	The thoracoscopic approach offers the advantage of less musculoskeletal sequelae compared to the conventional approach.	+	86.7	13/15	8 [5-9]
37	The maximum insufflation pressure of CO <sub>2</sub> during thoracoscopy should not exceed 5 mmHg.	+	100	14/14	9 [6-9]
38	Maximum duration of thoracoscopic operation should be 3 hours.	+	92.9	13/14	8 [4-9]
39	The thoracoscopic approach has the disadvantage of longer operative time compared to the conventional approach.	+	92.9	13/14	7.5 [2-9]
40	The thoracoscopic approach has a negative pathophysiological impact (acidosis, cerebral oxygenation impairment) compared to the conventional approach.	-	30.8	4/13	5 [1-7]
41	The thoracoscopic approach has the disadvantage of a higher complication rate compared to the conventional approach.	-	35.7	5/14	3.5 [1-8]
42	There is no place for routine fundoplication in patients with EA during the initial operation.	+	100	18/18	9 [8-9]

**Table 1d:**  
**Postoperative management of patients with esophageal atresia**

	POSTOPERATIVE MANAGEMENT	Consensus	%	Votes	Median [range]
43	Postoperative ventilation and relaxation should not be routine and should be reserved for selected patients such as those with tension anastomosis.	+	100	14/14	9 [6-9]
44	Routine postoperative antibiotic treatment for longer than 24 hours should be recommended.	-	13.3	2/15	2 [1-9]
45	A postoperative contrast study of the esophagus should be routinely performed before the initiation of oral feeding.	-	20	3/15	1 [1-7]
46	Feeding via the transanastomotic tube may be routinely initiated at 24 hours postoperatively.	+	100	15/15	9 [7-9]
47	Oral feeding may be routinely initiated after 24 hours postoperatively.	+	100	15/15	9 [6-9]
48	An anastomotic leakage should be routinely managed with a chest drain.	+	92.9	13/14	8 [2-9]
49	An anastomotic leakage within the first 4 postoperative days may be considered for surgical revision.	-	71.4	10/14	8 [1-9]
50	A contrast study, tracheoscopy and esophagoscopy are necessary to exclude a re-fistula, or missed upper pouch fistula, if suspected.	+	93.8	15/16	9 [3-9]



51	A re-fistula may be initially managed by either endoscopic or surgical approach.	+	100	14/14	9 [6-9]
52	A clinical checklist should be made available including items which should be performed before first discharge (i.e. abdominal and renal ultrasound, re-suscitation training for parents/caregivers).	+	100	18/18	9 [9-9]

**Table 2: Literature meeting the criteria of CEBM Level 1 Evidence<sup>§</sup>**

Statement	Domain	Reference	Study Type
The muscle-sparing approach is the recommended approach for conventional thoracotomy.	Operative Management	Askarpour S et al., Arq Bras Cir Dig 2018 [32]	RCT
The azygos vein should be preserved whenever possible.	Operative Management	Upadhyaya VD et al., Eur J Pediatr Surg. 2007 [33]	RCT
		Sharma S et al., Pediatr Surg Int 2007 [34]	RCT
The thoracoscopic approach has the disadvantage of a negative pathophysiological impact (acidosis, cerebral oxygenation impairment) compared to the conventional open approach.	Operative Management	Bishay M et al., Ann Surg 2013 [35]	Pilot RCT
An anastomotic leakage within the first 4 postoperative days may be considered for surgical revision.	Postoperative Management	Vaghela MM et al., J Pediatr Surg 2017 [36]	RCT

<sup>§</sup> In accordance with the Oxford CEBM Levels of Evidence as published in 2009 [30]  
RCT, randomized controlled trial

**Table 3**  
**Controversial discussion items leading to exclusion from voting due to inability to formulate a meaningful voting question**

<b>Domain</b>	<b>Discussed item</b>	<b>Result</b>
<b>Diagnostics</b>	Role of routine preoperative magnetic resonance imaging (MRI)	<b>No voting</b>
<b>Preoperative Management</b>	Measurement of the gap length before operation	<b>No voting</b>
<b>Operative Management</b>	Routine posterior tracheopexy during primary repair of esophageal atresia	<b>No voting</b>
<b>Postoperative Management</b>	Application of glycopyrrolate as a therapeutic option for anastomotic leakage.	<b>No voting</b>
	Content of a checklist for first discharge	<b>No voting</b>

## Supplement - Relevant Literature

### Domain *Diagnostics* (in alphabetical order)

Reference	Level of Evidence <sup>§</sup>
Aguilera-Pujabet M, Gahete JAM, Guillén G, et al. Management of neonates with right-sided aortic arch and esophageal atresia: International survey on IPEG AND ESPES members' experience. <i>J Pediatr Surg.</i> 2018 Oct;53(10):1923-1927.	5
Bradshaw CJ, Thakkar H, Knutzen L, et al. Accuracy of prenatal detection of tracheoesophageal fistula and oesophageal atresia. <i>J Pediatr Surg.</i> 2016;51(8):1268-72.	3
Broemling N, Campbell F. Anesthetic management of congenital tracheoesophageal fistula. <i>Paediatr Anaesth.</i> 2011;21(11):1092-9.	REVIEW
Cunningham BK, Khromykh A, Martinez AF, Carney T, Hadley DW, Solomon BD. Analysis of renal anomalies in VACTERL association. <i>Birth Defects Res A Clin Mol Teratol.</i> 2014;100(10):801-5.	4
Eghbalian F, Monsef A, Mousavi-Bahar SH. Urinary tract and other associated anomalies in newborns with esophageal atresia. <i>Urol J.</i> 2009 Spring;6(2):123-6.	4
Encinas JL, Luis AL, Avila LF, et al. Impact of preoperative diagnosis of congenital heart disease on the treatment of esophageal atresia. <i>Pediatr Surg Int.</i> 2006;22(2):150-3.	3
Fallon SC, Ethun CG, Olutoye OO, et al. Comparing characteristics and outcomes in infants with prenatal and postnatal diagnosis of esophageal atresia. <i>J Surg Res.</i> 2014;190(1):242-5.	3
Garabedian C, Sfeir R, Langlois C et al; French Network on Esophageal Atresia. Does prenatal diagnosis modify neonatal treatment and early outcome of children with esophageal atresia? <i>Am J Obstet Gynecol.</i> 2015;212(3):340.e1-7.	2
Gedicke MM, Gopal M, Spicer R. A gasless abdomen does not exclude distal tracheoesophageal fistula: the value of a repeat x-ray. <i>J Pediatr Surg.</i> 2007;42(3):576-7.	4
Kimble RM, Harding J, Kolbe A. Additional congenital anomalies in babies with gut atresia or stenosis: when to investigate, and which investigation. <i>Pediatr Surg Int.</i> 1997;12(8):565-70.	4
Kunisaki SM, Bruch SW, Hirschl RB, Mychaliska GB, Treadwell MC, Coran AG. The diagnosis of fetal esophageal atresia and its implications on perinatal outcome. <i>Pediatr Surg Int.</i> 2014;30(10):971-7.	3
Lautz TB, Mandelia A, Radhakrishnan J. VACTERL associations in children undergoing surgery for esophageal atresia and anorectal malformations: Implications for pediatric surgeons. <i>J Pediatr Surg.</i> 2015;50(8):1245-50.	4
McDuffie LA, Wakeman D, Warner BW. Diagnosis of esophageal atresia with tracheoesophageal fistula: is there a need for gastrointestinal contrast? <i>J Pediatr.</i> 2010;156(5):852.	5
Parolini F, Bulotta AL, Battaglia S, Alberti D. Preoperative management of children with esophageal atresia: current perspectives. <i>Pediatric Health Med Ther.</i> 2017;8:1-7.	REVIEW
Pedersen RN, Calzolari E, Husby S, Garne E; EUROCAT Working group. Esophageal atresia: prevalence, prenatal diagnosis and associated anomalies in 23 European regions. <i>Arch Dis Child.</i> 2012;97(3):227-32.	2
Soccorso G, England RJ, Godbole PP, Fisher RM, Marven SS. Mind the gap:delayed diagnosis of oesophageal atresia and tracheo-oesophageal fistula due to passage of a nasogastric tube. <i>Arch Dis Child Fetal Neonatal Ed.</i> 2012;97(6):F463-4.	5
Solomon BD, Baker LA, Bear KA, et al. An approach to the identification of anomalies and etiologies in neonates with identified or suspected VACTERL (vertebral defects, anal atresia, tracheo-esophageal fistula with esophageal atresia, cardiac anomalies, renal anomalies, and limb anomalies) association. <i>J Pediatr.</i> 2014;164(3):451-7.e1.	3
Spitz, Lewis; National Library of Medicine. Esophageal atresia. Lessons I have learned in a 40-year experience. <i>Journal of pediatric surgery Vol. 41, Iss.10,2006: 1635-1640.</i>	5

<sup>§</sup> In accordance with the Oxford CEBM Levels of Evidence as published in 2009 [9]

### Domain *Preoperative Management after Confirmation of Diagnosis*

(in alphabetical order)

Reference	Level of Evidence <sup>s</sup>
Alberti D: Esophageal atresia: pre and post-operative management. <i>J Matern Fetal Neonatal Med.</i> 2011;24 Suppl 1:4-6.	<b>REVIEW</b>
Ares G, Hunter CJ. Central venous access in children: indications, devices, and risks. <i>Curr Opin Pediatr.</i> 2017;29(3):340-346.	<b>REVIEW</b>
Aziz D. Can 'long-gap' esophageal atresia be safely managed at home while awaiting anastomosis? <i>J Pediatr Surg.</i> 2003;38(5):705-8.	<b>4</b>
Bagolan P, Valfrè L, Morini F, Conforti A. Long-gap esophageal atresia: traction- growth and anastomosis—before and beyond. <i>Dis Esophagus.</i> 2013;26 (4):372–379).	<b>4</b>
Bairdain S, Kelly DP, Tan C, et al. High incidence of catheter-associated venous thromboembolic events in patients with long gap esophageal atresia treated with the Foker process. <i>J Pediatr Surg.</i> 2014;49(2):370-3.	<b>4</b>
Borruto FA, Impellizzeri P, Montalto AS, et al. Thoracoscopy versus thoracotomy for esophageal atresia and tracheoesophageal fistula repair: review of the literature and meta-analysis. <i>Eur J Pediatr Surg.</i> 2012;22(6):415-9.	<b>3</b>
Burge DM, Shah K, Spark P, et al; British Association of Paediatric Surgeons Congenital Anomalies Surveillance System (BAPS-CASS). Contemporary management and outcomes for infants born with oesophageal atresia. <i>Br J Surg.</i> 2013;100(4):515-21.	<b>2</b>
Conforti A, Morini F, Pietro Bagolan. Difficult esophageal atresia. Trick and treat. <i>Semin Pediatr Surg</i> 2014;23:261–9.	<b>REVIEW</b>
Continent G, Costi D, Stephens P, Beringer R, Davidson A. An audit of anesthetic management and complications of tracheo-esophageal fistula and esophageal atresia repair. <i>Paediatr Anaesth</i> 2012; 22 (03) 268-274.	<b>4</b>
Fitzgerald DA. Pro -con debate: "That antibiotic prophylaxis should be used in children with repaired oesophageal atresia and tracheo-oesophageal fistula". <i>Paediatr Respir Rev.</i> 2016;18:58-9.	<b>REVIEW</b>
Friedmacher F, Puri P. Delayed primary anastomosis for management of long- gap esophageal atresia: a meta-analysis of complications and long-term out- come. <i>Pediatr Surg Int.</i> 2012;28(9):899–906).	<b>4</b>
Golonka NR, Hayashi AH. Early "sham" feeding of neonates promotes oral feeding after delayed primary repair of major congenital esophageal anomalies. <i>Am J Surg.</i> 2008;195(5):659-62; discussion 662.	<b>4</b>
Kelly DP, Bairdain S, Zurakowski D, et al. Quality improvement program reduces venous thromboembolism in infants and children with long-gap esophageal atresia (LGEA). <i>Pediatr Surg Int.</i> 2016;32(7):691-6.	<b>3</b>
Knottenbelt G, Skinner A, Seefelder C. Tracheo-oesophageal fistula (TOF) and oesophageal atresia (OA). <i>Best Pract Res Clin Anaesthesiol</i> 2010; 24 (03) 387-401	<b>REVIEW</b>
Lal DR, Gadepalli SK, Downard CD, et al.;Midwest Pediatric Surgery Consortium. Challenging surgical dogma in the management of proximal esophageal atresia with distal tracheoesophageal fistula:Outcomes from the Midwest Pediatric Surgery Consortium. <i>J Pediatr Surg.</i> 2018;53(7):1267-1272.	<b>3</b>
Lal D, Miyano G. Current patterns of practice and technique in the repair of esophageal atresia and tracheoesophageal fistua: an IPEG survey. <i>J Laparoendosc Adv Surg Tech A.</i> 2013;23(7):635-8.	<b>5</b>
Leung TS, Bayston R, Spitz L. Bacterial colonisation of the upper pouch in neonates with oesophageal atresia. <i>Z Kinderchir.</i> 1986;41(2):78-80	<b>4</b>
Merkus PJ. That antibiotic prophylaxis should be used in children with repaired oesophageal atresia and or trachea-oesophageal fistula - The case in favour. <i>Paediatr Respir Rev.</i> 2016;18:60-1.	<b>REVIEW</b>
Replogle RL. Plastic sump catheter for drainage of the proximal pouch. <i>Surgery</i> 54; 296 (1963).	<b>5</b>
Rinkel R, Bakx R. Bronchoscopy and Fogarty Balloon Insertion of Distal Tracheo-Oesophageal Fistula for Oesophageal Atresia Repair With Video Illustration. <i>Ann Otol Rhinol Laryngol.</i> 2017;126(1):6-8.	<b>4</b>
Sayari AJ, Tashiro J, Wang B, Perez EA, Lasko DS, Sola JE. Weekday vs. weekend repair of esophageal atresia and tracheoesophageal fistula. <i>J Pediatr Surg.</i> 2016;51(5):739-42.	<b>3</b>
Schindler E, Kowald B, Suess H, et al. Catheterization of the radial or brachial artery in neonates and infants. <i>Paediatr Anaesth.</i> 2005;15(8):677-82)	<b>4</b>
Till H. Preoperative Evaluation 163-6, in "Esophageal and Gastric Disorders in Infancy and Childhood". Editors Holger Till, Mike Thomson, John E. Foker, George W. Holcomb III, Khalid M. Khan. 2017 Springer.	<b>BOOK CHAPTER</b>

Wang B, Tashiro J, Allan BJ, et al. A nationwide analysis of clinical outcomes among newborns with esophageal atresia and tracheoesophageal fistulas in the United States. <i>J Surg Res.</i> 2014;190(2):604-12.	<b>2</b>
Warris A. Prophylactic antibiotics should be used in children with repaired oesophageal atresia and tracheo-oesophageal fistula: The case against. <i>Paediatr Respir Rev.</i> 2016;18:62-3.	<b>REVIEW</b>
Wu Y, Kuang H, Lv T, Wu C. Comparison of clinical outcomes between open and thoracoscopic repair for esophageal atresia with tracheoesophageal fistula: a systematic review and meta-analysis. <i>Pediatr Surg Int.</i> 2017;33(11):1147-1157	<b>3</b>
Yang YF, Dong R, Zheng C, et al. Outcomes of thoracoscopy versus thoracotomy for esophageal atresia with tracheoesophageal fistula repair: A PRISMA- compliant systematic review and meta-analysis. <i>Medicine (Baltimore).</i> 2016;95(30):e4428.	<b>3</b>
Yeung A, Butterworth SA. A comparison of surgical outcomes between in-hours and after- hours tracheoesophageal fistula repairs. <i>J Pediatr Surg.</i> 2015;50(5):805-8.	<b>4</b>
Zani A, Eaton S, Hoellwarth M, et al. International Survey on the Management of Esophageal Atresia. <i>Eur J Pediatr Surg</i> 2014;24:003–8).	<b>5</b>
Zani A, Lamas-Pinheiro R, Paraboschi I et al. Intraoperative acidosis and hypercapnia during thoracoscopic repair of congenital diaphragmatic hernia and esophageal atresia/tracheoesophageal fistula. <i>Paediatr Anaesth.</i> 2017;27(8):841-848).	<b>4</b>

§ In accordance with the Oxford CEBM Levels of Evidence as published in 2009 [9]

### Domain *Operative Management* (in alphabetical order)

<b>Reference</b>	<b>Level of Evidence<sup>§</sup></b>
Aguilera-Pujabet M, Gahete JAM, Guillén G, et al. Management of neonates with right-sided aortic arch and esophageal atresia: International survey on IPEG AND ESPES members´ experience. <i>J Pediatr Surg.</i> 2018;53:1923-1927.	<b>5</b>
Ainsworth SB, McGuire W. Peripherally Inserted Central Catheters vs Peripheral Cannulas for Delivering Parenteral Nutrition in Neonates. <i>JAMA.</i> 2016 21;315(23):2612-3.	<b>5</b>
Alabbad SI, Ryckman J, Puligandla PS, Shaw K, Nguyen LT, Laberge JM. Use of transanastomotic feeding tubes during esophageal atresia repair. <i>J Pediatr Surg.</i> 2009 ;44(5):902-5.	<b>4</b>
Askarpour S, Peyvasteh M, Ashrafi A, Dehdashtian M, Malekian A, Aramesh MR. Muscel sparing versus posterolateral thoracotomy in neonates with esophageal atresia. <i>Arq Bras Cir Dig.</i> 2018;31(2):e1365.	<b>1</b>
Bastard F, Bonnard A, Rousseau V, et al. Thoracic skeletal anomalies following surgical treatment of esophageal atresia. Lessons from a national cohort. <i>J Pediatr Surg.</i> 2018;53(4):605- 609.	<b>2</b>
Bath S, Lines J, Loeffler AM, Malhotra A, Turner RB. Impact of standardization of antimicrobial prophylaxis duration in pediatric cardiac surgery. <i>J Thorac Cardiovasc Surg.</i> 2016 ;152:1115-20.	<b>3</b>
Bicakci U, Tander B, Ariturk E, Rizalar R, Ayyildiz SH, Bernay F. The right-sided aortic arch in children with esophageal atresia and tracheo-esophageal fistula: a repair through the right thoracotomy. <i>Pediatr Surg Int.</i> 2009 ;25:423-5.	<b>4</b>
Bishay M, Giacomello L, Retrosi G, et al. Hypercapnia and Acidosis During Open and Thoracoscopic Repair of Congenital Diaphragmatic Hernia and Esophageal Atresia: Results of a Pilot Randomized Controlled Trial. <i>Ann Surg.</i> 2013; 258; 895–900.	<b>1</b>
Bishay M, Giacomello L, Retrosi G, et al. Decreased cerebral oxygen saturation during thoracoscopic repair of congenital diaphragmatic hernia and esophageal atresia in infants. <i>J Pediatr Surg.</i> 2011;46(1):47-51.	<b>3</b>
Broemling N, Campbell F. Anesthetic management of congenital tracheoesophageal fistula. <i>Pediatr Anesth</i> 2011; 21:1092-9	<b>REVIEW</b>
Costerus S, Vlot J, van Rosmalen J, Wijnen R, Weber F. Effects of Neonatal Thoracoscopic Surgery on Tissue Oxygenation: A Pilot Study on (Neuro-) Monitoring and Outcomes. <i>Eur J Pediatr Surg.</i> 2017.	<b>2</b>
Cross K, Smith J, Walker IA. Thoracoabdominal and general surgery. Ch 10 in <i>Neonatal Anesthesia</i> ed J Lerman, Springer 2015	<b>BOOK CHAPTER</b>
Davenport M, Rothenberg SS, Crabbe DC, Wulkan ML. The great debate: open or thoracoscopic repair for oesophageal atresia or diaphragmatic hernia. <i>J Pediatr Surg.</i> 2015;50(2):240-6.	<b>REVIEW</b>
Davies RR. All the small things: The impact of central venous catheters in neonates undergoing cardiac surgery. <i>J Thorac Cardiovasc Surg.</i> 2018 ;155:1159-1160.	<b>REVIEW</b>

Dingemann C, Ure B, Dingemann J. Thoracoscopic procedures in pediatric surgery: what is the evidence? <i>Eur J Pediatr Surg.</i> 2014;24(1):14-9.	3
Dingemann C, Ure BM. Minimally invasive repair of esophageal atresia: an update. <i>Eur J Pediatr Surg.</i> 2013;23(3):198-203.	3
Evans WN, Kogut K, Acherman RJ. Preserving the azygos vein when repairing esophageal atresia and tracheoesophageal fistula accompanied by interrupted inferior vena cava. <i>Pediatr Surg Int.</i> 2014;30(3):345-7.	5
Fathi M, Joudi M, Morteza A. Evaluating Necessity of Azygos Vein Ligation in Primary Repair of Esophageal Atresia. <i>Indian J Surg.</i> 2015 ;77 (Suppl 2):543-5.	3
Fayoux P, Morisse M, Sfeir R, Michaud L, Daniel S. Laryngotracheal anomalies associated with esophageal atresia: importance of early diagnosis. <i>Eur Arch Otorhinolaryngol.</i> 2018;275(2):477-481.	4
Filston HC, Chitwood WR Jr, Schkolne B, Blackmon LR. The Fogarty balloon catheter as an aid to management of the infant with esophageal atresia and tracheoesophageal fistula complicated by severe RDS or pneumonia. <i>J Pediatr Surg.</i> 1982;17:149-51.	5
Fusco JC, Calisto JL, Gaines BA, Malek MM. A large single-institution review of tracheoesophageal fistulae with evaluation of the use of transanastomotic feeding tubes. <i>J Pediatr Surg.</i> 2017	4
Gruszka A, Sachweh JS, Schnoering H, et al. Aortopexy offers surgical options for a variety of pathological tracheal conditions in paediatric patients. <i>Interact Cardiovasc Thorac Surg.</i> 2017	4
Guo W, Li Y, Jiao A, Peng Y, Hou D, Chen Y. Tracheoesophageal fistula after primary repair of type C esophageal atresia in the neonatal period: recurrent or missed second congenital fistula. <i>J Pediatr Surg.</i> 2010;45(12):2351-5.	4
Harmon CM and Coran AG. Congenital Anomalies of the esophagus. In <i>Pediatric Surgery</i> (Grosfeld, O'Neill, Fonkalsrud, Coran eds) 7th ed Chapter 69, Sanders London 2012.	<b>BOOK CHAPTER</b>
Holcomb GW 3rd, Rothenberg SS, Bax KM, et al. Thoracoscopic repair of esophageal atresia and tracheoesophageal fistula: a multi-institutional analysis. <i>Ann Surg.</i> 2005;242(3):422-8; discussion 428-30.	4
Johnson C, Sims C. Neonatal anaesthesia chap 13 in <i>Your guide to Paediatric Anaesthesia</i> , ed by C Sims & C Johnson, McGraw Hill, Sidney 2011	<b>BOOK CHAPTER</b>
Kamran A, Smithers CJ, Manfredi MA, et al. Slide Esophagoplasty vs End-to-End Anastomosis for Recalcitrant Esophageal Stricture after Esophageal Atresia Repair. <i>J Am Coll Surg.</i> 2018 ;226(6):1045-1050.	3
Knottenbelt J, Skinner A, Seefelder C. Tracheo-oesophageal fistula and oesophageal atresia in Best Practice & Research Clinical Anaesthesiology: Neonatal anaesthesia. 2010; 24: 387- 401	<b>BOOK CHAPTER</b>
Koivusalo A, Suominen J, Rintala R, Pakarinen M. Location of TEF at the carina as an indicator of long-gap C-type esophageal atresia. <i>Dis Esophagus.</i> 2018.	4
Krosnar S, Baxter A. Repair of oesophageal atresia with tracheo-oesophageal fistula: anaesthetic and intensive care management of a series of 8 patients. <i>Pediatr Anesth</i> 2005;15 : 541-6	4
Lal DR, Gadepalli SK, Downard CD et al. Midwest Pediatric Surgery Consortium. Challenging surgical dogma in the management of proximal esophageal atresia with distal tracheoesophageal fistula: Outcomes from the Midwest Pediatric Surgery Consortium. <i>J Pediatr Surg.</i> 2018 53(7):1267-1272.	3
Lal DR, Gadepalli SK, Downard CD, et al. Infants with esophageal atresia and right aortic arch: Characteristics and outcomes from the Midwest Pediatric Surgery Consortium. <i>J Pediatr Surg.</i> 2018	3
Lal DR, Gadepalli SK, Downard CD, et al. Perioperative management and outcomes of esophageal atresia and tracheoesophageal fistula. <i>J Pediatr Surg.</i> 2017;52:1245-1251.	4
Lal D, Miyano G, Juang D, Sharp NE, St Peter SD. Current Patterns of Practice and Technique in the Repair of Esophageal Atresia and Tracheoesophageal Fistula: An IPEG Survey <i>JOURNAL OF LAPAROENDOSCOPIC &amp; ADVANCED SURGICAL TECHNIQUES</i> Volume 23, Number 7, 2013.	5
Lawal TA, Gosemann JH, Kuebler JF, Glüer S, Ure BM. Thoracoscopy versus thoracotomy improves midterm musculoskeletal status and cosmesis in infants and children. <i>Ann Thorac Surg.</i> 2009;87(1):224-8.	2
Losty P. In Losty P, Flake A, Rintala R, Hutson JM and Iwai N (editors) <i>Rickham's Neonatal Surgery</i> Springer-Verlag, London 2018 Volume 1 Chapter 23 pp 541 – 61.	<b>BOOK CHAPTER</b>
Ma L, Liu YZ, Ma YQ, Zhang SS, Pan NL. Comparison of neonatal tolerance to thoracoscopic and open repair of esophageal atresia with tracheoesophageal fistula. <i>Chin Med J (Engl).</i> 2012;125(19):3492-5.	3
Narayanan S, Vazhiyodan A, Somnath P, Mohanan A. Is routine use of transanastomotic tube justified in the repair of esophageal atresia? <i>World J Pediatr</i> 2017;13(6):584-587	3

Parolini F, Boroni G, Stefini S, Agapiti C, Bazzana T, Alberti D. Role of preoperative tracheobronchoscopy in newborns with esophageal atresia: A review. <i>World J Gastrointest Endosc.</i> 2014;16 :6:482-7.	<b>3</b>
Pepper VK, Boomer LA, Thung AK, Grischkan JM, Diefenbach KA. Routine Bronchoscopy and Fogarty Catheter Occlusion of Tracheoesophageal Fistulas. <i>J Laparoendosc Adv Surg Tech A.</i> 2017;27(1):97-100.	<b>3</b>
Pierro A. Hypercapnia and acidosis during the thoracoscopic repair of oesophageal atresia and congenital diaphragmatic hernia. <i>J Pediatr Surg.</i> 2015;50(2):247-9.	<b>REVIEW</b>
Pinheiro PFM, Simões e Silva AC, Pereira RM. Current knowledge on esophageal atresia. <i>World Journal of Gastroenterology : WJG.</i> 2012;18 :3662-3672.	<b>REVIEW</b>
Rashid KA, Maletha M, Khan TR, Wakhlu A, Rawat J, Kureel SN. Esophageal anastomosis medial to preserved azygos vein in esophageal atresia with tracheoesophageal fistula: restoration of normal mediastinal anatomy. <i>J Neonatal Surg.</i> 2012;1(4):50.	<b>3</b>
Rinkel R, Van Poll, Sibarani-Ponsen R, Sleeboom C, Bakx R. Bronchoscopy and Fogarty Balloon Insertion of Distal Tracheo-Oesophageal Fistula for Oesophageal Atresia Repair With Video Illustration. <i>Ann Otol Rhinol Laryngol.</i> 2017;126:6-8.	<b>4</b>
Szavay PO, Zundel S, Blumenstock G, et al. Perioperative outcome of patients with esophageal atresia and tracheo-esophageal fistula undergoing open versus thoracoscopic surgery. <i>J Laparoendosc Adv Surg Tech A.</i> 2011;21(5):439-43.	<b>3</b>
Sharma N, Srinivas M. Laryngotracheobronchoscopy prior to esophageal atresia and tracheoesophageal fistula repair-its use and importance. <i>J Pediatr Surg.</i> 2014;49(2):367- 9.	<b>4</b>
Sharma S, Sinha SK, Rawat JD, Wakhlu A, Kureel SN, Tandon R. Azygos vein preservation in primary repair of esophageal atresia with tracheoesophageal fistula. <i>Pediatr Surg Int.</i> 2007;23(12):1215-8.	<b>1</b>
St. Peter SD, Valusek PA, Snyder CL, Holcomb GW III, Ostlie DJ. Impact of Suture Choice on Stricture Formation Following Repair of Esophageal Atresia. <i>Annals of Pediatric Surgery, Vol 3, No 2, 2007 PP 75-79.</i>	<b>4</b>
Shieh HF, Smithers CJ, Hamilton TE, et al. Posterior Tracheopexy for Severe Tracheomalacia Associated with Esophageal Atresia (EA): Primary Treatment at the Time of Initial EA Repair versus Secondary Treatment. <i>Front Surg.</i> 2018;4:80.	<b>4</b>
Stolwijk LJ, van der Zee DC, Tytgat S, et al. Brain Oxygenation During Thoracoscopic Repair of Long Gap Esophageal Atresia. <i>World J Surg.</i> 2017;41(5):1384-1392.	<b>4</b>
Taguchi T, Nagata K, Kinoshita Y, et al. The utility of muscle sparing axillar skin crease incision for pediatric thoracic surgery. <i>Pediatr Surg Int.</i> 2012;28(3):239-44.	<b>4</b>
Tandale SR, Dave N, Garasia M, Patil S, Parelkar S.A Study of Morbidity and Cost of Peripheral Venous Cannulation in Neonates Admitted to Paediatric Surgical Intensive Care Unit. <i>J Clin Diagn Res.</i> 2017;11(3):UC08-UC10.	<b>4</b>
Thakkar H, Upadhyaya M, Yardley IE. Bronchoscopy as a screening tool for symptomatic tracheomalacia in oesophageal atresia. <i>J Pediatr Surg.</i> 2018;53:227-229.	<b>2</b>
Turner NM. Intraoperative hypotension in neonates: when and how should we intervene? <i>Curr Opin Anaesthesiol.</i> 2015;28(3):308-13.	<b>REVIEW</b>
Tytgat SHAJ, van Herwaarden-Lindeboom MYA, van Tuyl van Serooskerken ES, van der Zee DC. Thoracoscopic posterior tracheopexy during primary esophageal atresia repair: a new approach to prevent tracheomalacia complications. <i>J Pediatr Surg.</i> 2018 53(7):1420-1423.	<b>4</b>
Tytgat SH, van Herwaarden MY, Stolwijk LJ, et al. Neonatal brain oxygenation during thoracoscopic correction of esophageal atresia. <i>Surg Endosc.</i> 2016;30(7):2811-7.	<b>4</b>
Upadhyaya VD, Gangopadhyaya AN, Gopal SC, et al. Is ligation of azygos vein necessary in primary repair of tracheoesophageal fistula with esophageal atresia? <i>Eur J Pediatr Surg.</i> 2007;17(4):236-40.	<b>1</b>
Walker S, Datta A, Massoumi RL, Gross ER, Uhing M, Arca MJ. Antibiotic stewardship in the newborn surgical patient: A quality improvement project in the neonatal intensive care unit. <i>Surgery.</i> 2017;162:1295-1303.	<b>3</b>
Wei S, Saran N, Emil S. Musculoskeletal deformities following neonatal thoracotomy: long- term follow-up of an esophageal atresia cohort. <i>J Pediatr Surg.</i> 2017;52(12):1898-1903.	<b>2</b>
Wood JA, Carachi R. The right-sided aortic arch in children with oesophageal atresia and tracheo-oesophageal fistula. <i>Eur J Pediatr Surg.</i> 2012	<b>4</b>
Wu Y, Kuang H, Lv T, Wu C. Comparison of clinical outcomes between open and thoracoscopic repair for esophageal atresia with tracheoesophageal fistula: a systematic review and meta-analysis. <i>Pediatr Surg Int.</i> 2017;33(11):1147-1157.	<b>3</b>
Yang YF, Dong R, Zheng C, et al. Outcomes of thoracoscopy versus thoracotomy for esophageal atresia with tracheoesophageal fistula repair: A PRISMA- compliant systematic review and meta-analysis. <i>Medicine (Baltimore).</i> 2016;95(30):e4428.	<b>3</b>

Zani A, Lamas-Pinheiro R, Paraboschi I, et al. Intraoperative acidosis and hypercapnia during thoracoscopic repair of congenital diaphragmatic hernia and esophageal atresia/tracheoesophageal fistula. <i>Paediatr Anaesth.</i> 2017;27(8):841-848.	<b>4</b>
--	----------

§ In accordance with the Oxford CEBM Levels of Evidence as published in 2009 [9]

### Domain *Postoperative Management* (in alphabetical order)

Reference	Level of Evidence <sup>§</sup>
Alberti D: Esophageal atresia: pre and post-operative management. <i>J Matern Fetal Neonatal Med.</i> 2011;24 Suppl 1:4-6.	<b>REVIEW</b>
Aworanti O, Awadalla S. Management of recurrent tracheoesophageal fistulas a systematic review. <i>Eur J Pediatr Surg</i> 2014; 24: 365-75	<b>4</b>
Beasley SW: Does postoperative ventilation have an effect on the integrity of the anastomosis in repaired oesophageal atresia? <i>J Pediatr Child Health</i> 1999; 35: 120-2	<b>REVIEW</b>
Golden J, Demeter NE, C Lim J, Ford HR, Upperman JS, Gayer CP. Routine post-operative esophagram is not necessary after repair of esophageal atresia. <i>Am J Surg</i> 2017; 213: 640-4	<b>4</b>
Gregory S, Chun RH, Parakininkas D, et al: Endoscopic esophageal and tracheal cauterization for closure of recurrent tracheoesophageal fistula a case report and review of the literature. <i>JPORL</i> 2017; 98: 158-61	<b>5</b>
Koivusalo A. Revisional surgery for recurrent tracheoesophageal fistula and anastomotic complications after repair of esophageal atresia in 258 infants. <i>J Pediatr Surg.</i> 2015; 50:250-4	<b>4</b>
Lal DR, Gadepalli SK, Downard CD, et al. Midwest Pediatric Surgery Consortium. Challenging surgical dogma in the management of proximal esophageal atresia with distal tracheoesophageal fistula: Outcomes from the Midwest Pediatric Surgery Consortium. <i>J Pediatr Surg.</i> 2018 53(7):1267-1272.	<b>3</b>
Lal D1, Miyano G, Juang D, Sharp NE, St Peter SD. Current patterns of practice and technique in the repair of esophageal atresia and tracheoesophageal fistula: an IPEG survey. <i>JLAST</i> 2013; 23: 635-8.	<b>5</b>
Nambirajan L1, Rintala RJ, Losty PD, Carty H, Lloyd DA. The value of early postoperative oesophagography following repair of oesophageal atresia. <i>Pediatr Surg Int</i> 1998; 13: 76-8.	<b>4</b>
Pinheiro PFM, Simoes e Silva AC, Pereira RM. Current knowledge on esophageal atresia. <i>World J Gastroenterol</i> 2012; 18: 3662-72	<b>REVIEW</b>
Smithers CJ, Hamilton TE, Manfredi MA, et al: Categorization and repair of recurrent and acquired tracheoesophageal fistulae occurring after esophageal repair. <i>J Pediatr Surg</i> 2016; 52: 424-30	<b>4</b>
Uchida K, Inoue M, Otake K, et al. Efficacy of postoperative elective ventilator support for leakage protection in primary anastomosis of congenital esophageal atresia. <i>Pediatr Surg Int</i> 2006; 22: 696-9	<b>3</b>
Vaghela MM, Mahajan JK, Sundram J, Bhardwaj N, Rao KL. Role of glycopyrrolate in healing of anastomotic dehiscence after primary repair of esophageal atresia in a low resource setting-A randomized controlled study. <i>J Pediatr Surg</i> 2017; 52: 420-3	<b>1</b>
Wang J, Zhang M, Pan W, Wu W, Yan W, Cai W. Management of recurrent tracheoesophageal fistula after esophageal atresia and follow up. <i>Diseases of the Esophagus</i> 2017; 30: 1-8	<b>2</b>
Yanchar NL, Gordon R, Cooper M, Dunlap H, Soucy P. Significance of the clinical course and early upper gastrointestinal studies in predicting complications associated with repair of esophageal atresia. <i>J Pediatr Surg.</i> 2001; 36: 815-22	<b>4</b>
Zani A, Eaton S, Hoellwarth ME, et al. International survey on the management of esophageal atresia. <i>Eur J Pediatr Surg</i> 2014; 24: 3-8	<b>5</b>
Zhao R, Li K, Shen C, Zheng S. The outcome of conservative treatment for anastomotic leakage after surgical repair of esophageal atresia. <i>J Pediatr Surg.</i> 2011; 46: 2274-8	<b>4</b>

§ In accordance with the Oxford CEBM Levels of Evidence as published in 2009 [9]