

# **Variability in the reporting of baseline characteristics, treatment and outcomes in esophageal atresia publications: a systematic review**

## **Authors and affiliations:**

Nadine M. Teunissen<sup>1,2</sup> MD, Julia Brendel<sup>3</sup> MD, Simon Eaton<sup>1,4</sup> PhD, Nigel J. Hall<sup>5</sup> PhD, Rebecca M. Thursfield<sup>6</sup> MBChB MD(Res), L.W. Ernest van Heurn<sup>7</sup> MD PhD, Benno M. Ure<sup>3</sup> MD PhD, René M.H. Wijnen<sup>1</sup> MD PhD

<sup>1</sup> Department of Pediatric Surgery and Pediatric Intensive Care Unit, Erasmus University Medical Centre - Sophia Children's Hospital, Rotterdam, The Netherlands

<sup>2</sup> Dutch Institute for Clinical Auditing, Leiden, The Netherlands

<sup>3</sup> Department of Pediatric Surgery, Hannover Medical School, Hannover, Germany

<sup>4</sup> Developmental Biology and Cancer Research and Teaching Programme, UCL Great Ormond Street Institute of Child Health, London, United Kingdom

<sup>5</sup> University Surgery Unit, Faculty of Medicine, University of Southampton, Southampton, UK

<sup>6</sup> Department of Pediatric Respiratory Medicine, Alder Hey Children's Hospital NHS Foundation Trust, Liverpool, United Kingdom

<sup>7</sup> Department of Pediatric Surgery, Emma Children's Hospital, Amsterdam UMC, University of Amsterdam & Vrije Universiteit Amsterdam, Amsterdam Reproduction and Development Research Institute and the Amsterdam Public Health Research Institute, Amsterdam, The Netherlands

## **Corresponding author:**

Nadine Teunissen, MD

Department of Pediatric Surgery and Pediatric Intensive Care Unit

Erasmus University Medical Center – Sophia Children's Hospital

Wytemaweg 80, 3015 CN Rotterdam, The Netherlands

Telephone: +31 (0) 6 52 55 26 15

E-mail: n.teunissen@erasmusmc.nl

### **Contributions**

The study was designed by NT, RW, JB and SE. Inclusion and exclusion criteria, assessment strategy, and the utilized Excel-based data extraction framework were developed by all authors. NT developed the search strategy, and NT, JB, and SE were responsible for screening, as well as data extraction and analyses. NT and JB drafted the manuscript which was revised by SE; all authors read, provided feedback, and approved the final manuscript.

### **Funding**

The European Commission funded the EPSA|ERNICA Registry in the 3<sup>rd</sup> Health Program, HP-PJ-2019.

## INTRODUCTION

With approximately one new case in 3000 to 4000 births, esophageal atresia (EA) – with or without tracheoesophageal fistula – is a rare congenital anomaly, which requires surgical repair during the neonatal period.<sup>1</sup> Survival rates of EA patients have improved remarkably over the last decades of the 20<sup>th</sup> century and have since remained stable, with the probability of survival depending mainly on the presence of additional anomalies or chromosomal or genetic syndrome diagnoses such as VACTERL association.<sup>2,3</sup> Because of the improved survival, the focus of EA research has shifted from mortality to parameters such as long-term outcomes, and quality of life for EA patients, as well as quality of care.

However, the evaluation and comparison of (quality of) EA care between hospitals, regions or even countries through clinical audits is made difficult by the lack of standardization of measured process indicators and clinical outcomes. The current implementation of an European clinical audit for esophageal atresia care necessitates the generation of an overview of all possible outcomes described in EA research, as well as patient characteristics and treatment- and care process characteristics to - in time - be able to correct for case mix in (quality of) care comparisons.<sup>4</sup>

EA research is equally hindered by a lack of standardization of measured outcomes. As EA is rare, prospective trials with adequate patient numbers are generally even more rare. Best practice and "gold standard" in EA care is mainly based on expert consensus.<sup>5,6</sup> The significant increase in the number of publications on EA over the last

decades could potentially contribute to more evidence-based practice,<sup>7</sup> but overarching comparisons of results - such as meta-analyses - are difficult because measured outcomes and their corresponding definitions and manner of reporting widely vary. A core outcome set would enable such comparison of published research, and is under development (OCELOT).<sup>8</sup>

Hence a comprehensive overview of all possible EA outcomes, would be essential in the development of: (i) indicators (process, outcome) for use in audits and registries to allow comparisons between centers, regions, care pathways etc. with appropriate case-mix adjustment; and (ii) a core outcome set for use in EA research.

The primary aim of this explorative systematic review was to create such a comprehensive list of all reported outcomes in recently published peer-reviewed research on the main EA care process, as well as the variability in utilization, definition and reporting thereof. The secondary aim was to identify and define patient characteristics, as well as treatment and care process characteristics that could enable future interpretation of comparison of (quality of) care and outcome results. For the purpose of this paper, the term 'studied parameters' refers to all patient characteristics, treatment- and care process characteristics and outcomes. To our knowledge, this is the first effort to create such a detailed summary of studied parameters in EA research.

## **METHODS**

This review was performed according to the Preferred Reporting Items for Systematic Reviews and Meta-analysis (PRISMA) statement and guidelines.<sup>9</sup> A broad search strategy for Medline, Embase, and the Cochrane Library was developed in collaboration with an experienced medical librarian of the Erasmus University Medical Center. The search was based on the search term 'esophageal atresia', combined with the following terms: 'morbidity' or 'mortality' or 'survival' or 'outcome' or 'complication'. Complete search strategies are provided in Supplementary File 1. The search was performed in September 2021.

### **Inclusion and exclusion criteria**

The search aimed to find all papers concerning any aspect of the main esophageal atresia care process, including surgical and non-surgical management. Hence, studies only focusing on the outcome of redo surgery were excluded. Subsequently, papers published before 2015 were excluded to warrant contemporaneousness. Lastly, non-English-language publications were excluded, as well as animal research and *in vitro* studies, case series with less than ten patients, editorials, letters, meeting abstracts and reviews, guidelines, and consensus statements.

### **Selection process**

NT and JB independently screened the titles and abstracts of all search results to evaluate eligibility on the grounds of reporting care for and management of patients with esophageal atresia. Subsequently, the full texts of eligible articles were screened on

relevance for the review. Any disagreement was discussed and, if necessary, resolved by SE. Reviewing authors were not blinded for the title, authors, or journal name.

### **Data extraction, analysis, and results**

An Excel-based framework facilitated data extraction. Similar studied parameters were categorized and, following agreement between NT and JB, merged into an overarching term. The most frequently reported parameters were noted in the framework.

Uncommon parameters, those mentioned in less than 5% of papers, were noted in a separate file. If available, definitions of studied parameters were extracted and noted, as well as utilized standardized ways of assessing the parameters, such as medical scores, scales and questionnaires, if the result thereof was directly reported in the included publication. Estimates of the parameters themselves were not extracted nor interpreted, and the methodological quality of included publications was not assessed.

## **RESULTS**

### **Included articles**

The applied search identified 3577 publications. After removal of duplicates and limiting the results to those articles published after 2015, 818 articles remained, of which 209 met the inclusion criteria (Fig. 1). A summary of study characteristics of all included publications is presented in table 1. An individualized overview of included studies and corresponding study characteristics can be found in Supplementary File 2.

## **Data extraction**

Full-text analysis of the 209 included manuscripts identified 731 parameters described in at least one of the included publications, which were then categorized into three overarching categories: patient characteristics (n=128), treatment- and care process characteristics (n=338), and outcomes (n=265). As several parameters could arguably be included in more than one category (e.g. length of primary hospital stay; number of surgeries), categorization followed agreement between the first two authors. We further arranged the parameters by topic, such as comorbidities, primary treatment, specific complications and long-term outcome, to facilitate the comparison of variation in studied parameters within similar subjects. The complete lists of all identified and extracted patient characteristics, treatment- and care process characteristics, and outcomes are included in Supplementary Files 3 through 5.

## **Extracted studied parameters**

Of 731 identified studied parameters, 92 parameters were described in more than 5% of included publications. These studied parameters are listed in table 2. Patient characteristics mentioned in more than two-thirds of included publications were sex, gestational age, presence of a cardiac malformation, and birth weight. The type of esophageal atresia according to the Gross classification was mentioned in 74% of papers. The type of surgical repair was stated in 60% of included articles. All other primary (surgical) treatment characteristics were mentioned in fewer than half of included publications.

Anastomotic stricture was the most frequently described complication; i.e., in 72% of included publications, followed by anastomotic leakage (69%). Mortality rate was reported in 66% of included publications. Length of primary hospital stay and duration of follow-up were mentioned in only one-third of articles (37% and 39%, respectively). In general, definitions of studied parameters varied widely between publications, rendering the extraction of unambiguous definitions impossible.

Variability in utilized standardized instruments to measure parameters such as quality of life was equally wide. To illustrate, health-related quality of life was described in 16/209 publications (8%), using twelve different tools or instruments. Other long-term outcomes, such as (motor) development, cognitive functioning, and behavior, were mentioned less often, yet the variability of used assessment tools was comparably large. A list of standardized scales, scores or instruments that were used in included publications to assess these outcomes is displayed in table 3.

## **DISCUSSION**

To the best of our knowledge, this is the first systematic review that summarizes all parameters studied in recent clinical research on esophageal atresia. More than 730 different patient characteristics, treatment characteristics and outcomes were identified in 209 included publications, demonstrating substantial variation in research interest and reporting. However, most of the parameters were mentioned solely in a handful of papers. Of 265 identified outcomes, only 5 (2%) were mentioned in more than 50% of included publications. The proportions of patient characteristics (5/128, 4%) and



treatment- and care process characteristics (1/338, <1%) that were studied and reported in more than 50% of included publications, proves to be similarly small. Additionally, studied parameters were defined and assessed inconsistently across included publications, thereby impeding comparison and benchmarking of parameters, even if they were more frequently studied.

In addition to generating an extensive list of all studied parameters, our study reveals several noteworthy observations. First, as expected, most studies were retrospective cohort studies, which emphasizes the need for more prospective studies (audits, registries, research studies, randomized trials) to establish best practice. Second, the studied parameters mainly refer to primary surgical repair of esophageal atresia and its complications. Although some studies addressed long-term outcomes such as follow-up and transition programs or long-term complications,<sup>10–16</sup> the complete list of parameters suggests that the esophageal atresia research of recent years generally had a short-term focus. By contrast, a recent overview of publication trends and global collaborations on esophageal atresia research found particular interest in the long-term outcome, surgical techniques, and epidemiology when assessing key points of the ten most-cited EA publications since 1945.<sup>7</sup> Thus, there is a mismatch between the short-term focus of the large majority of EA research versus the long-term outcomes that professionals and advocacy groups deem most important. Obviously, the reporting of short-term outcomes is easier than the reporting of long-term outcomes, for sequelae such as motor/neurodevelopment, chest wall deformities, chronic respiratory diseases, and lung function impairment often occur later in childhood or adolescence – and thus

require longer follow-up. The third remarkable trend is that only eighteen studies described quality of life outcomes through patient-reported outcome measures (PROMs), and the applied instrument varied across the studies.<sup>10,12–14,17–27</sup> The most used PROM was the generic, child-specific PEDS-QoL, which, however, was mentioned in only 8/209 papers (4%). This scarce use of PROMs is surprising, as there is a general movement to patient-centered care and PROM research across the medical world. This scarce use could possibly be attributed to the lack of a disease-specific PROM during the study period. Meanwhile, an EA-specific PROM for children has been developed in Sweden and Germany and is currently being validated in other countries.<sup>28</sup> Additionally, an EA-specific PROM for adults is being developed and validated in a Dutch nationwide study.<sup>29</sup> Lastly, although the large variety of studied parameters reflects attention for comorbidity and quality of care, mortality remains one of the most reported ones in recently published research.

Our study has some limitations. First, to warrant contemporaneity of results, reports published before 2015 were excluded. It is possible that extension of the time period to include earlier years would have yielded further outcomes that were not included in our review; it would also have potentially allowed an analysis of trends over time. However, on balance it was felt that the 466 different patient- and treatment characteristics and 265 outcomes from 209 publications were representative of current practice. Additionally, non-English publications were excluded, which may have led to underestimation of the variation in studied parameters, considering that geographical differences and local practices could influence the selection thereof.

The extensive list of studied parameters resulting from this systematic review, will serve as the foundation of projects aimed at standardizing EA data collection. Clinical audits or registry-based research will benefit from standardization of data regarding patient characteristics and characteristics of treatment and care processes, enabling further interpretation and correction of measured variation in outcome.

Additionally, these datasets must enable comparison or benchmarking between medical centers, thus be able to capture between-hospital variation and reflect (good) care. This review was undertaken as the first stage of defining a “Core Indicator Set” for data collection within the EPSA/ERNICA EA registry, which has the aim of improving EA care across Europe.

Although also aiming to improve patient care, the intention is somewhat different from that of a Core Outcome Set, which is *“an agreed standardized set of outcomes that should be measured and reported, as a minimum, in all clinical trials in specific areas of health or health care.”* The focus of a core outcome set is on trials and therefore the outcomes by which different treatments can be compared. As an example of the distinction between core indicator sets and core outcomes sets, we can take examples from our literature review. The number of patients having intra-operative bronchoscopy may be an interesting and useful process indicator to look at variation across European centers (and may be represented in a core indicator set) but it is unlikely to become part of a Core Outcome Set. Anastomotic leak, on the other hand, might be selected for both a Core Indicator Set and a Core Outcome set, as it is relevant to both the variation

between centers (e.g. leak rate in center X vs. others) and might also be useful to compare different treatments (e.g. thoracotomy vs. thoracoscopy). The application of core outcome sets has become increasingly important to achieve consistency of outcome reporting, with consideration of patients' perception of the importance of outcomes.<sup>30</sup> Defining a core outcome set for EA is already underway (OCELOT).<sup>8</sup> Core outcome sets often suffice to interpret differences between study arms in RCTs, in which baseline-characteristics are balanced, and treatment pathways protocolized.

To ensure recognition and implementation of both core data sets by everybody involved in esophageal atresia care and research, it is of utmost importance to involve all stakeholder groups (health care providers, patients, and researchers) in both endeavors.

## **CONCLUSION**

This review found substantial variability in reported patient characteristics, treatment- and care process characteristics, and outcomes in research regarding the main care process for patients with esophageal atresia. The resulting list of studied parameters could aid in the development of a standardized core outcome set, as well as a core indicator set. Standardized measurement and reporting is necessary to invoke less reporting bias, more interpretable results, and the possibility of more cross-study or cross-healthcare comparisons, which could significantly improve future outcome measurement and reporting in esophageal atresia care, as well as esophageal atresia research. The development and validation of both the standardized core indicator set

and core outcome set has started in 2021 with involvement of European expert health care providers as well as patient representatives.

## REFERENCES

1. Nassar N, Leoncini E, Amar E, et al. Prevalence of esophageal atresia among 18 international birth defects surveillance programs. *Birth Defects Res Part A Clin Mol Teratol.* 2012;94(11) doi:10.1002/bdra.23067
2. Bell JC, Baynam G, Bergman JEH, et al. Survival of infants born with esophageal atresia among 24 international birth defects surveillance programs. *Birth Defects Res.* 2021;113(12) doi:10.1002/bdr2.1891
3. Zimmer J, Eaton S, Murchison LE, De Coppi P, Ure BM, Dingemann C. State of Play: Eight Decades of Surgery for Esophageal Atresia. *Eur J Pediatr Surg.* 2019;29(1):39-48 doi:10.1055/s-0038-1668150
4. Teunissen NM. ERNICA | EPSA Registry. Published 2022. Accessed June 10, 2022. <https://ern-ernica.eu/registry/>
5. Dingemann C, Eaton S, Aksnes G, et al. ERNICA Consensus Conference on the Management of Patients with Esophageal Atresia and Tracheoesophageal Fistula: Diagnostics, Preoperative, Operative, and Postoperative Management. *Eur J Pediatr Surg.* 2020;30(4):326-336 doi:10.1055/s-0039-1693116
6. 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 doi:10.1097/MPG.0000000000001401

7. Feng X, Martynov I, Suttkus A, Lacher M, Mayer S. Publication Trends and Global Collaborations on Esophageal Atresia Research: A Bibliometric Study. *Eur J Pediatr Surg.* 2021;31(2):164-171 doi:10.1055/s-0040-1702223
8. Ocelot study. Research projects endorsed by TOFS. Published 2022. Accessed May 10, 2022. <https://tofs.org.uk/oa-tof-information/oa-tof-research/ocelot-study/>
9. Page MJ, Moher D, Bossuyt PM, et al. PRISMA 2020 explanation and elaboration: updated guidance and exemplars for reporting systematic reviews. *BMJ.* Published online March 29, 2021 doi:10.1136/bmj.n160
10. 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(01) doi:10.1055/s-0038-1672144
11. Bakal U, Ersoz F, Eker I, Sarac M, Aydin M, Kazez A. Long-Term Prognosis of Patients with Esophageal Atresia and/or Tracheoesophageal Fistula. *Indian J Pediatr.* 2016;83(5):401-404 doi:10.1007/s12098-015-1930-0
12. Gallo G, van Tuyll van Serooskerken ES, Tytgat SHAJ, et al. Quality of life after esophageal replacement in children. *J Pediatr Surg.* 2021;56(2) doi:10.1016/j.jpedsurg.2020.07.014
13. Hannon E, Eaton S, Curry JI, Kiely EM, Spitz L, De Coppi P. Outcomes in adulthood of gastric transposition for complex and long gap esophageal atresia. *J Pediatr Surg.* 2020;55(4):639-645 doi:10.1016/j.jpedsurg.2019.08.012
14. Mikkelsen A, Boye B, Diseth TH, et al. Traumatic stress, mental health and quality of life in adolescents with esophageal atresia. *J Pediatr Surg.* Published online November 7, 2020 doi:10.1016/j.jpedsurg.2020.10.029

15. Okuyama H, Tazuke Y, Ueno T, et al. Long-term morbidity in adolescents and young adults with surgically treated esophageal atresia. *Surg Today*. 2017;47(7) doi:10.1007/s00595-016-1462-x
16. Vergouwe FWT, IJsselstijn H, Biermann K, et al. High Prevalence of Barrett's Esophagus and Esophageal Squamous Cell Carcinoma After Repair of Esophageal Atresia. *Clin Gastroenterol Hepatol*. 2018;16(4) doi:10.1016/j.cgh.2017.11.008
17. Kumari V, Joshi P, Dhua AK, et al. Developmental Status of Children Operated for Esophageal Atresia with or without Tracheoesophageal Fistula Along with Maternal Stress, Their Quality of life, and Coping Abilities at AIIMS, New Delhi. *Eur J Pediatr Surg*. 2019;29(1):125-131 doi:10.1055/s-0038-1676825
18. di Natale A, Brestel J, Mauracher AA, et al. Long-Term Outcomes and Health-Related Quality of Life in a Swiss Patient Group with Esophageal Atresia. *Eur J Pediatr Surg*. Published online July 29, 2021 doi:10.1055/s-0041-1731391
19. Sreeram II, Ten Kate CA, van Rosmalen J, et al. Patient-Reported Outcome Measures and Clinical Outcomes in Children with Foregut Anomalies. *Child (Basel, Switzerland)*. 2021;8(7) doi:10.3390/children8070587
20. Tannuri ACA, Angelo SS, Takyi P, da Silva AR, Tannuri U. Esophageal substitution or esophageal elongation procedures in patients with complicated esophageal atresia? Results of a comparative study. *J Pediatr Surg*. 2021;56(5):933-937 doi:10.1016/j.jpedsurg.2020.07.028
21. van Hoorn CE, van der Cammen-van Zijp MHM, Stolker RJ, van Rosmalen J, Wijnen RMH, de Graaff JC. Associations of perioperative characteristics with motor

- function in preschool children born with esophageal atresia. *Paediatr Anaesth.* 2021;31(8):854-862 doi:10.1111/pan.14204
22. Bal HS, Sen S, Karl S, Mathai J, Thomas RJ. An assessment of quality of life of operated cases of esophageal atresia in the community. *J Indian Assoc Pediatr Surg.* 21(3):131-138 doi:10.4103/0971-9261.182588
  23. 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.* 2016;27(01) doi:10.1055/s-0036-1587334
  24. Miyano G, Seo S, Nakamura H, et al. Changes in quality of life from infancy to school age after esophagoesophagostomy for tracheoesophageal fistula: thoracotomy versus thoracoscopy. *Pediatr Surg Int.* 2017;33(10):1087-1090 doi:10.1007/s00383-017-4141-0
  25. Amin R, Knezevich M, Lingongo M, et al. Long-term Quality of Life in Neonatal Surgical Disease. *Ann Surg.* 2018;268(3):497-505 doi:10.1097/SLA.0000000000002918
  26. 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 doi:10.1016/j.jpedsurg.2017.05.033
  27. Youn JK, Park T, Kim SH, et al. Prospective evaluation of clinical outcomes and quality of life after gastric tube interposition as esophageal reconstruction in children. *Medicine (Baltimore).* 2018;97(52):e13801 doi:10.1097/MD.00000000000013801
  28. Dellenmark-Blom M, Abrahamsson K, Quitmann JH, et al. Development and pilot-



testing of a condition-specific instrument to assess the quality-of-life in children and adolescents born with esophageal atresia. *Dis Esophagus*. 2017;30(7) doi:10.1093/dote/dox017

29. Ten Kate CA, Teunissen NM, van Rosmalen J, et al. Development and Validation of a Condition-Specific Quality of Life Instrument for Adults with Esophageal Atresia [Unpublished Manuscript].; 2022
30. Williamson PR, Altman DG, Bagley H, et al. The COMET Handbook: version 1.0. *Trials*. 2017;18(S3):280 doi:10.1186/s13063-017-1978-4

## **LEGENDS OF FIGURES AND TABLES**

**Figure 1.** PRISMA Flowchart: systematic review of studied parameters in esophageal atresia research

**Table 1.** Study characteristics of included publications

**Table 2.** Identified studied parameters in EA research

**Table 3.** Identified tools/instruments utilized and reported in one or more included publications

## **ACKNOWLEDGEMENTS**

This article is supported by ERNICA. The authors would like to thank the European Pediatric Surgical Audit group (EPSA|ERNICA Registry) for scientific input.

Additionally, the authors wish to thank Sabrina T.G. Meertens-Gunput. from the Erasmus MC Medical Library for developing and updating the search strategies. And thanks to Ko Hagoort for editing the manuscript. SE acknowledges support from the National Institute of Health Research Biomedical Research Centre at Great Ormond Street Hospital.

## **CONFLICTS OF INTEREST**

The authors declare no conflict of interest.

**Table 1.** Study characteristics of included publications

		<b>n =</b>	<b>%</b>
<b>Originated in</b>	Africa	8	(4%)
	Asia	49	(23%)
	Europe	87	(42%)
	North-America	44	(21%)
	Oceania	10	(5%)
	South-America	2	(1%)
	Intercontinental	9	(4%)
<b>Study design</b>	Retrospective	148	(71%)
	Prospective	21	(10%)
	Cross-sectional	37	(18%)
	Mixed design	3	(1%)
<b>Type of study</b>	Observational	119	(57%)
	Comparative	90	(43%)
<b>Study design</b>	Cohort	198	(95%)
	Case-control	8	(4%)
	Trial	3	1%
<b>Year of publication</b>	2015	19	(9%)
	2016	23	(11%)
	2017	36	(17%)
	2018	27	(13%)
	2019	23	(11%)
	2020	35	(17%)
	2021	46	(22%)
<b>Included type of EA</b>	Only type A <sup>a</sup>	2	(1%)
	Only type C <sup>a</sup>	31	(15%)
	Only type E <sup>a</sup>	4	(2%)
	Long gap <sup>b</sup>	10	(4%)
	Multiple types <sup>a</sup>	31	(15%)
	All types <sup>a</sup>	129	(62%)
	Other <sup>c</sup>	2	(1%)

<sup>a</sup>According to Gross Classification;

<sup>b</sup>Following the definition of the included publication.

<sup>c</sup>Inclusion of patients based on other criterion: long gap and complication in primary repair (1), prenatal suspicion of EA (1).

**Table 2.** Identified studied parameters in EA research

		<b>n =</b>	<b>%</b>
<b>Baseline characteristics</b>	Sex	178	(85%)
	Gestational age or prematurity	154	(74%)
	Type of esophageal atresia	154	(74%)
	Cardiac malformation / Congenital heart disease	149	(71%)
	Birth weight	144	(69%)
	Age at surgery	98	(47%)
	Long Gap / Gap length	97	(46%)
	VACTERL <sup>a</sup> -association	88	(42%)
	Any other congenital malformation	83	(40%)
	Chromosomal / Genetic abnormalities	82	(39%)
	Renal/Genitourinary anomalies	76	(36%)
	Musculoskeletal / Limb anomalies	74	(35%)
	Anorectal malformation	67	(32%)
	Intestinal malformation	57	(27%)
	Age at time of study (survey, follow-up, intervention)	56	(27%)
	Weight at operation	41	(20%)
	Pulmonary / respiratory anomalies or conditions	36	(17%)
	Neurologic / Central nervous system anomalies	31	(15%)
	Fistula: yes/no	28	(13%)
	Previous esophageal surgery	27	(13%)
	Referred from other hospital	25	(12%)
	"Other" comorbidity (unspecified)	23	(11%)
	CHARGE <sup>b</sup> syndrome	23	(11%)
	Age at presentation / admission / diagnosis	22	(11%)
	Race / Ethnicity	18	(9%)
	Otolaryngeal anomalies / Auditory / Hearing issues	13	(6%)
	Laryngeal cleft / Laryngo-tracheo-oesophageal cleft	12	(6%)
Spitz classification	12	(6%)	
Intra-uterine growth retardation/ Small-for-	11	(5%)	
Twin / Multiple birth	11	(5%)	
<b>Treatment- and care process characteristics</b>	Type of repair (primary, secondary, interposition)	125	(60%)
	Age at surgery	98	(47%)

	Duration of follow-up	82	(39%)
	Gastrostomy at any point in time	81	(39%)
	Thoracotomy or thoracoscopy	79	(38%)
	Length of primary hospital stay	78	(37%)
	Number of dilatations	74	(35%)
	Duration of ventilation/intubation	64	(31%)
	Use of anti-acid medication (at any point in time)	61	(29%)
	Operation time	37	(18%)
	Aortopexy: yes or no	33	(16%)
	Cervical esophagostomy until surgery / at any point	33	(16%)
	Conversion thoracoscopy to thoracotomy	29	(14%)
	Time to start oral feeding (postoperatively)	29	(14%)
	Tracheostomy	27	(13%)
	Tension-free anastomosis / Concern regarding	27	(13%)
	Time on (neonatal) intensive care (days)	27	(13%)
	Prenatal diagnosis	26	(12%)
	Intra-operative chest tube	23	(11%)
	Bronchoscopy (intraoperative)	22	(11%)
	Transanastomotic tube	21	(10%)
	Polyhydramnios on antenatal ultrasound	19	(9%)
	Elongation procedure	19	(9%)
	Need for preoperative intubation/ventilation	19	(9%)
	Time to start tube feeding	14	(7%)
	Contrast study postoperative	13	(6%)
	Lung function (spirometry) in follow-up	13	(6%)
	Time between diagnosis and surgery	13	(6%)
	Time to full oral feeding	13	(6%)
	Thoracotomy: left or right approach	12	(6%)
	Use of inhalation medication	12	(6%)
	Echocardiography	11	(5%)
	Number of surgeries that patient underwent	11	(5%)
<b>Outcome</b>	Anastomotic stricture/stenosis	150	(72%)
	Anastomotic leakage	143	(69%)
	Mortality	137	(66%)

Dilatations: yes or no	128	(61%)
Gastroesophageal reflux	110	(53%)
Anti-reflux surgery: yes or no	88	(42%)
Recurrent fistula	84	(40%)
Redo surgery (esophageal)	79	(38%)
Tracheomalacia	63	(30%)
Growth / Weight / Failure to thrive	61	(29%)
Oral feeding issues	61	(29%)
Respiratory complications / symptoms / chronic	59	(28%)
Pneumonia	57	(27%)
Dysphagia / Swallowing difficulties	52	(25%)
Sepsis	43	(21%)
Recurrent respiratory infections	38	(18%)
Pneumothorax	36	(17%)
Aortopexy: yes or no	33	(16%)
Vocal cord complications / Voice changing	28	(13%)
Gastro-intestinal symptoms	23	(11%)
Blood loss during primary surgery	22	(11%)
Wound complications	20	(10%)
Chylothorax	19	(9%)
Chest wall deformity	19	(9%)
Quality of Life	16	(8%)
Readmission	16	(8%)
Graft necrosis / Graft failure / Graft loss	15	(7%)
Complications: yes or no	14	(7%)
Motor-/Neurodevelopment	13	(6%)
Complications (intra-operative)	11	(5%)
Esophageal perforation after dilatation	11	(5%)

---

<sup>a</sup>VACTERL: Vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies and limb abnormalities. <sup>b</sup>CHARGE: Coloboma, heart defects, atresia choanae, growth retardation, genital abnormalities and ear abnormalities.

---

**Table 3.** Identified tools/instruments utilized and reported in one or more included publications

		<b>n =</b>	<b>%</b>
<b>Behavior</b>	(Modified) Vineland Social Maturity Scale	2	(<1%)
	Behavior Assessment System for Children (BASC II)	1	(<1%)
	Behavior Rating Inventory of Executive Function Preschool	1	(<1%)
	Child Behavior Checklist	1	(<1%)
<b>Birth</b>	Score for Neonatal Acute Physiology with Perinatal Extension (SNAPPE-II)	1	(<1%)
<b>Cognition</b>	Wechsler Intelligence Scale for Children (WISC)	3	(1%)
	Wechsler Preschool & Primary Scale of Intelligence (WPPSI)	2	(<1%)
	Ankara Developmental Screening Inventory	1	(<1%)
	Children's Memory Scale	1	(<1%)
	Developmental Neuropsychological Assessment (NEPSY)	1	(<1%)
	Revised Amsterdam Intelligence Test (RAKIT)	1	(<1%)
	Test of Everyday Attention for Children	1	(<1%)
	Wide Range Achievement Test (WRAT)	1	(<1%)
<b>Coping, transition and commitment to care</b>	Coping Strategy Checklist	1	(<1%)
	Fragebogen zur Messung der Patientenzufriedenheit (ZUF-8)	1	(<1%)
	Patient Activation Measure (PAM)	1	(<1%)
<b>Development</b>	Bayley Scales of Infant and Toddler Development (BSID)	4	(2%)
	Movement Assessment Battery for Children (M-ABC)	3	(1%)
	Ages and Stages Questionnaire	1	(<1%)
	Developmental Assessment Scale for Indian Infants	1	(<1%)
	Gross Motor Function Classification System (GMFCS)	1	(<1%)
	Kinderturntest plus / Deutscher Motorik Test	1	(<1%)
	Trivandrum Development Screening Chart (TDSC)	1	(<1%)
<b>Feeding</b>	Functional Oral Intake Score (FOIS)	3	(1%)
	Montreal Children's Hospital Feeding Scale (MCH-FS)	2	(<1%)
	International Dysphagia Diet Standardization Initiative (IDDSI)	1	(<1%)
	Karaduman Chewing Performance Scale (KCPS)	1	(<1%)
	Penetration and aspiration scale	1	(<1%)
	Turkish Feeding / Swallowing Impact Survey	1	(<1%)



<b>(Mental) health status and symptomatology</b>	German Health Survey for Children and Adolescents (KIGGS)	2	(<1%)
	Gastrointestinal Symptom Rating Scale (GSRS)	1	(<1%)
	Strength and Difficulties Questionnaire – 20 (SDQ-20)	1	(<1%)
	Risk Adjustment for Congenital Heart Surgery Score (Rachs)	1	(<1%)
<b>Quality of life</b>	Pediatric Quality of Life Inventory (PedsQL)	8	(4%)
	Gastrointestinal Quality of Life Index (GIQLI)	4	(2%)
	Self-developed questionnaire	2	(<1%)
	World Health Organization Quality of Life Questionnaire (WHOQOL-BREF)	2	(<1%)
	Child Health Questionnaire (CHF87-BREF)	1	(<1%)
	DISABKIDS Chronic Generic Measure - 47	1	(<1%)
	KIDSCREEN-27	1	(<1%)
	Short Form Survey (SF-36)	1	(<1%)
	TNO AZL Children's Quality of Life (TACQOL)	1	(<1%)
	TNO AZL Adult's Quality of Life (TAAQOL)	1	(<1%)
	WHO-5 / WHO-5 parental	1	(<1%)
<b>Trauma and stress</b>	Impact of Events Scale (IES-13)	1	(<1%)
	Parental stress scale	1	(<1%)

---

Multiple instruments or tools may have been used in one publication.

## Identification of studies

Identification

Records identified from\*:  
Embase (n = 2002)  
Medline (Ovid) (n = 1517)  
Cochrane Central (n = 58)

Records removed *before screening*:  
Duplicate records removed (n = 1274)  
Records removed before 2015 (n = 1485)

Screening

Records screened on title and abstract (n = 818)

Records excluded (n = 569)

Reports sought for retrieval (n = 249)

Reports not retrieved (n = 0)

Reports assessed for eligibility (n = 249)

Reports excluded:  
Not concerning main EA care process (n = 34)  
Case series n<10 (n = 4)  
Non-English language (n = 1)  
Conference abstract (n = 1)

Included

Studies included in review (n = 209)  
Reports of included studies (n = 209)

## **Supplementary material of the manuscript:**

### **Variability in the reporting of baseline characteristics, treatment and outcomes in esophageal atresia publications: a systematic review**

#### **Authors and affiliations:**

Nadine M. Teunissen<sup>1,2</sup> MD, Julia Brendel<sup>3</sup> MD, Simon Eaton<sup>1,4</sup> PhD, Nigel J. Hall<sup>5</sup> PhD, Rebecca M. Thursfield<sup>6</sup> MBChB MD(Res), L.W. Ernest van Heurn<sup>7</sup> MD PhD, Benno M. Ure<sup>3</sup> MD PhD, René Wijnen<sup>1</sup> MD PhD

<sup>1</sup> Department of Pediatric Surgery and Pediatric Intensive Care Unit, Erasmus University Medical Centre - Sophia Children's Hospital, Rotterdam, The Netherlands

<sup>2</sup> Dutch Institute for Clinical Auditing, Leiden, The Netherlands

<sup>3</sup> Department of Pediatric Surgery, Hannover Medical School, Hannover, Germany

<sup>4</sup> Developmental Biology and Cancer Research and Teaching Programme, UCL Great Ormond Street Institute of Child Health, London, United Kingdom

<sup>5</sup> University Surgery Unit, Faculty of Medicine, University of Southampton, Southampton, UK

<sup>6</sup> Department of Paediatric Respiratory Medicine, Alder Hey Children's Hospital NHS Foundation Trust, Liverpool, United Kingdom

<sup>7</sup> Department of Pediatric Surgery, Emma Children's Hospital, Amsterdam UMC, University of Amsterdam & Vrije Universiteit Amsterdam, Amsterdam Reproduction and Development Research Institute and the Amsterdam Public Health Research Institute, Amsterdam, The Netherlands

**Corresponding author:**

Nadine Teunissen, MD

**Contents:**

S1. Search Strategy

S2. Included publications

S3. Overview of all identified baseline characteristics

S4. Overview of all identified treatment- and care-process characteristics

S5. Overview of all identified outcomes

## **SUPPLEMENTARY 1: search strategy**

### **Embase.com**

('esophagus atresia'/de OR (EA-TEF OR TOF-OA OR ((esophag\* OR oesophag\* OR tracheoesophag\*) NEAR/3 (atresia\* OR atretic\*))) :ab,ti,kw) AND ('morbidity'/exp OR 'mortality'/exp OR 'survival'/exp OR 'treatment outcome'/exp OR 'complication'/exp OR (morbidity\* OR mortality\* OR survival\* OR outcome\* OR complication\*) :ab,ti,kw) NOT ([conference abstract]/lim OR [editorial]/lim OR [letter]/lim) AND [English]/lim NOT ('review'/exp OR (review):ti) NOT ((animal/exp OR animal\*:de OR nonhuman/de) NOT ('human'/exp))

### **Medline (Ovid)**

(Esophageal Atresia/ OR (EA-TEF OR TOF-OA OR ((esophag\* OR oesophag\* OR tracheoesophag\*) ADJ3 (atresia\* OR atretic\*))) .ab,ti,kf.) AND (Morbidity/ OR exp Mortality/ OR mortality.fx. OR Survival/ OR exp Treatment Outcome/ OR (morbidity\* OR mortality\* OR survival\* OR outcome\* OR complication\*) .ab,ti,kf.) NOT (letter\* OR news OR comment\* OR editorial\* OR congress\* OR abstract\* OR book\* OR chapter\* OR dissertation abstract\*) .pt. AND english.lg. NOT (exp Review/ OR (review).ti.) NOT (exp animals/ NOT humans/)

### **Cochrane Central**

((EA NEXT TEF OR TOF NEXT OA OR ((esophag\* OR oesophag\* OR tracheoesophag\*) NEAR/3 (atresia\* OR atretic\*))) :ab,ti,kw) AND ((morbidity\* OR mortality\* OR survival\* OR outcome\* OR complication\*) :ab,ti,kw)

## SUPPLEMENTARY 2: included publications

Author	Title	Year	Continent	n =	Study design		
Askarpour S, et al.	Evaluation of risk factors affecting anastomotic leakage after repair of esophageal atresia	2015	Asia	61	Comparative	Retrospective	Cohort
Baird R, et al.	A pilot investigation of feeding problems in children with esophageal atresia	2015	North-America	30	Observational	Cross-sectional	Cohort
Bairdain S, et al.	Foker process for the correction of long gap esophageal atresia: Primary treatment versus secondary treatment after prior esophageal surgery	2015	North-America	52	Comparative	Retrospective	Cohort
Bevilacqua F, et al.	Factors affecting short-term neurodevelopmental outcome in children operated on for major congenital anomalies	2015	Europe	41	Observational	Prospective / Cross-Sectional	Cohort
Gallo G, et al.	A two-center comparative study of gastric pull-up and jejunal interposition for long gap esophageal atresia	2015	Europe	24	Comparative	Retrospective	Cohort
Huynh-Trudeau V, et al.	Dysphagia among adult patients who underwent surgery for esophageal atresia at birth	2015	North-America	41	Observational	Cross-sectional	Cohort
Kay-Rivest E, et al.	Evaluation of aortopexy in the management of severe tracheomalacia after esophageal atresia repair	2015	North-America	132	Observational	Retrospective	Cohort
Koivusalo AI, et al.	Revisional surgery for recurrent tracheoesophageal fistula and anastomotic complications after repair of esophageal atresia in 258 infants	2015	Europe	258	Observational	Retrospective	Cohort
Koziarkiewicz M, et al.	Long-term complications of congenital esophageal atresia—single institution experience	2015	Europe	77	Observational	Retrospective	Cohort
Mochizuki K, et al.	Impact of an external lengthening procedure on the outcome of long-gap esophageal atresia at our hospitals	2015	Asia	16	Comparative	Retrospective	Cohort
Okuyama H, et al.	Current practice and outcomes of thoracoscopic esophageal atresia and tracheoesophageal fistula repair: A multi-institutional analysis in Japan	2015	Asia	58	Observational	Retrospective	Cohort
Pini Prato A, et al.	A cross-sectional nationwide survey on esophageal atresia and tracheoesophageal fistula	2015	Europe	146	Observational	Retrospective	Cohort
Shah R, et al.	Predictive factors for complications in children with esophageal atresia and tracheoesophageal fistula	2015	Oceania	100	Observational	Retrospective	Cohort
Vukadin M, et al.	Analysis of Prognostic Factors and Mortality in Children with Esophageal Atresia	2015	Europe	60	Observational	Retrospective	Cohort
Woo S, et al.	Thoracoscopic versus open repair of tracheoesophageal fistulas and rates of vocal cord paresis	2015	North-America	31	Comparative	Retrospective	Cohort
Yalcin S, et al.	The evaluation of deglutition with videofluoroscopy after repair of esophageal atresia and/or tracheoesophageal fistula	2015	Eurasia	32	Observational	Cross-sectional	Cohort
Murase N, et al.	Prophylactic effect of H2 blocker for anastomotic stricture after esophageal atresia repair	2015	Asia	27	Comparative	Retrospective	Case-control
Yeung A, and Butterworth SA	A comparison of surgical outcomes between in-hours and after-hours tracheoesophageal fistula repairs	2015	North-America	28	Comparative	Retrospective	Cohort
Zhu H, et al.	Reoperation for anastomotic complications of esophageal atresia and tracheoesophageal fistula	2015	Asia	21	Observational	Retrospective	Cohort
Abouzeid AA, et al.	Posterior cologastric anastomosis: An effective antireflux mechanism in colonic replacement of the esophagus	2016	Africa	16	Observational	Retrospective	Cohort
Acher CW, et al.	Long-term outcomes of patients with tracheoesophageal fistula/esophageal atresia: Survey results from tracheoesophageal fistula/esophageal atresia online communities	2016	North-America	445	Observational	Cross-sectional	Cohort

Author	Title	Year	Continent	n =	Study design		
Askarpour S, et al.	End-to-end versus end-to-side anastomosis in the treatment of esophageal atresia or tracheoesophageal fistula	2016	Asia	72	Comparative	Retrospective	Cohort
Bakal U, et al.	Long-Term Prognosis of Patients with Esophageal Atresia and/or Tracheoesophageal Fistula	2016	Asia	57	Observational	Cross-sectional	Cohort
Bal HS, et al.	An assessment of quality of life of operated cases of esophageal atresia in the community	2016	Asia	79	Observational	Cross-sectional	Cohort
Bradshaw CJ, et al.	Accuracy of prenatal detection of tracheoesophageal fistula and oesophageal atresia	2016	Europe	58	Comparative	Retrospective	Cohort
Cartabuke RH, et al.	Long-term esophageal and respiratory outcomes in children with esophageal atresia and tracheoesophageal fistula	2016	North-America	43	Observational	Retrospective	Cohort
Choudhury SR, et al.	Pediatric esophageal substitution by gastric pull-up and gastric tube	2016	Asia	22	Observational	Retrospective	Cohort
Conforti A, et al.	Cervical repair of congenital tracheoesophageal fistula: Complications lurking!	2016	Europe	18	Comparative	Retrospective	Cohort
Deboer EM, et al.	Multidisciplinary care of children with repaired esophageal atresia and tracheoesophageal fistula	2016	North-America	29	Observational	Cross-sectional	Cohort
Dingemann C, et al.	Early complications after esophageal atresia repair: analysis of a German health insurance database covering a population of 8 million	2016	Europe	75	Observational	Retrospective	Cohort
Donoso F, et al.	Outcome and management in infants with esophageal atresia – A single centre observational study	2016	Europe	129	Comparative	Retrospective	Cohort
Hiradfar M, et al.	Thoracoscopic Esophageal Atresia with Tracheoesophageal Fistula Repair: The First Iranian Group Report, Passing the Learning Curve	2016	Asia	24	Observational	Retrospective	Cohort
Jönsson L, et al.	Treatment and Follow-Up of Patients with Long-Gap Esophageal Atresia: 15 Years' of Experience from the Western Region of Sweden	2016	Europe	16	Observational	Retrospective	Cohort
Malakounides G, et al.	Esophageal Atresia: Improved Outcome in High-Risk Groups Revisited	2016	Europe	200	Comparative	Retrospective	Cohort
Milickovic M, et al.	Gastric tube esophageal reconstruction in children with esophageal atresia and caustic stricture Study of clinical value based on 25 single-center. Centre experience	2016	Europe	22	Observational	Retrospective	Cohort
Okata Y, et al.	Evaluation of the intraoperative risk factors for esophageal anastomotic complications after primary repair of esophageal atresia with tracheoesophageal fistula	2016	Asia	28	Comparative	Retrospective	Cohort
Raitio A, et al.	Fluoroscopic balloon dilatation for anastomotic strictures in patients with esophageal atresia: A fifteen-year single centre UK experience	2016	Europe	137	Observational	Retrospective	Cohort
Rassiwala M, et al.	Determinants of gap length in esophageal atresia with tracheoesophageal fistula and the impact of gap length on outcome	2016	Asia	69	Comparative	Prospective	Cohort
Sayari AJ, et al.	Weekday vs. weekend repair of esophageal atresia and tracheoesophageal fistula	2016	North-America	861	Comparative	Retrospective	Cohort
Shah PS, et al.	Does continuous positive airway pressure for extubation in congenital tracheoesophageal fistula increase the risk of anastomotic leak? A retrospective cohort study	2016	Oceania	51	Comparative	Retrospective	Cohort
Walker K, et al.	Developmental outcomes at three years of age of infants with esophageal atresia	2016	Oceania	24	Comparative	Prospective	Case-control
Zani A, et al.	Preservation of native esophagus in infants with pure esophageal atresia has good long-term outcomes despite significant postoperative morbidity	2016	North-America	12	Observational	Retrospective	Cohort
Dingemann J, et al.	Transition of Patients with Esophageal Atresia to Adult Care: Results of a Transition-Specific Education Program	2017	Europe	29	Comparative	Cross-sectional	Cohort

Author	Title	Year	Continent	n =	Study design		
Dittrich R, et al.	Pulmonary outcome of esophageal atresia patients and its potential causes in early childhood	2017	Europe	27	Observational	Cross-sectional	Cohort
Donoso F, and Lilja HE	Risk Factors for Anastomotic Strictures after Esophageal Atresia Repair: Prophylactic Proton Pump Inhibitors Do Not Reduce the Incidence of Strictures	2017	Europe	128	Comparative	Retrospective	Cohort
Elfiky MMA, et al.	Gastric tube esophagoplasty for pediatric esophageal replacement	2017	Africa	27	Observational	Retrospective	Cohort
Friedmacher F, et al.	Postoperative Complications and Functional Outcome after Esophageal Atresia Repair: Results from Longitudinal Single-Center Follow-Up	2017	Europe	109	Comparative	Retrospective	Cohort
Gallo G, et al.	Respiratory function after esophageal replacement in children	2017	Europe	15	Comparative	Cross-sectional	Cohort
Gibreel W, et al.	Swallowing Dysfunction and Quality of Life in Adults with Surgically Corrected Esophageal Atresia/Tracheoesophageal Fistula as Infants	2017	North-America	46	Observational	Cross-sectional	Cohort
Harmsen WJ, et al.	Developmental problems in patients with oesophageal atresia: A longitudinal follow-up study	2017	Europe	58	Observational	Cross-sectional	Cohort
Hölscher AC, et al.	Quality of Life after Surgical Treatment for Esophageal Atresia: Long-Term Outcome of 154 Patients	2017	Europe	154	Observational	Cross-sectional	Cohort
Koivusalo AI, et al.	Long-term outcomes of oesophageal atresia without or with proximal tracheoesophageal fistula – Gross types A and B	2017	Europe	68	Observational	Retrospective	Cohort
Lal DR, et al.	Perioperative management and outcomes of esophageal atresia and tracheoesophageal fistula	2017	North-America	396	Comparative	Retrospective	Cohort
Li XW, et al.	A scoring system to predict mortality in infants with esophageal atresia	2017	Asia	198	Comparative	Retrospective	Cohort
Long AM, et al.	Oesophageal atresia with no distal tracheoesophageal fistula: Management and outcomes from a population-based cohort	2017	Europe	21	Observational	Prospective	Cohort
Menzies J, et al.	Prevalence of malnutrition and feeding difficulties in children with esophageal atresia	2017	Australia	75	Observational	Cross-sectional	Cohort
Miyano G, et al.	Changes in quality of life from infancy to school age after esophagoesophagostomy for tracheoesophageal fistula: thoracotomy versus thoracoscopy	2017	Asia	37	Comparative	Retrospective	Cohort
Narayanan SK, et al.	Is routine use of transanastomotic tube justified in the repair of esophageal atresia?	2017	Asia	33	Comparative	Retrospective	Cohort
Nomura A, et al.	Evaluation of developmental prognosis for esophageal atresia with tracheoesophageal fistula	2017	Asia	47	Comparative	Retrospective	Cohort
Okuyama H, et al.	Long-term morbidity in adolescents and young adults with surgically treated esophageal atresia	2017	Asia	69	Comparative	Retrospective	Cohort
Pedersen RN, et al.	Long-term pulmonary function in esophageal atresia—A case-control study	2017	Europe	59	Comparative	Cross-sectional	Case-control
Peters RT, et al.	Mortality and morbidity in oesophageal atresia	2017	Europe	248	Observational	Retrospective	Cohort
Porcaro F, et al.	Respiratory problems in children with esophageal atresia and tracheoesophageal fistula	2017	Europe	105	Observational	Retrospective	Cohort
Rattan KN, et al.	Clinical profile and short-term outcome of neonates with esophageal atresia and tracheoesophageal fistula at tertiary care center in a developing country: A 25-year experience	2017	Asia	693	Comparative	Retrospective	Cohort
Saiad MO	The Modified Posterior Thoracotomy for Esophageal Atresia	2017	Africa	56	Observational	Retrospective	Cohort
Schmidt A, et al.	Outcome of primary repair in extremely and very low-birth-weight infants with esophageal atresia/distal tracheoesophageal fistula	2017	Europe	35	Observational	Retrospective	Cohort



Author	Title	Year	Continent	n =	Study design		
Shieh HF, et al.	Posterior Tracheopexy for Severe Tracheomalacia Associated with Esophageal Atresia (EA): Primary Treatment at the Time of Initial EA Repair versus Secondary Treatment	2017	North-America	118	Comparative	Retrospective	Cohort
Smithers CJ, et al.	Categorization and repair of recurrent and acquired tracheoesophageal fistulae occurring after esophageal atresia repair	2017	North-America	66	Observational	Retrospective	Cohort
Stenström P, et al.	Prolonged Use of Proton Pump Inhibitors as Stricture Prophylaxis in Infants with Reconstructed Esophageal Atresia	2017	Europe	63	Comparative	Prospective / Retrospective	Cohort
Stenström P, et al.	Dilations of anastomotic strictures over time after repair of esophageal atresia	2017	Europe	131	Comparative	Retrospective	Cohort
Tanaka Y, et al.	Comparison of outcomes of thoracoscopic primary repair of gross type C esophageal atresia performed by qualified and non-qualified surgeons	2017	Asia	17	Comparative	Retrospective	Cohort
Toussaint-Duyster LCC, et al.	Determinants of exercise capacity in school-aged esophageal atresia patients	2017	Europe	63	Observational	Cross-sectional	Cohort
Tröbs RB, et al.	Isolated tracheoesophageal fistula versus esophageal atresia – Early morbidity and short-term outcome. A single institution series	2017	Europe	24	Comparative	Retrospective	Cohort
Vaghela MM, et al.	Role of glycopyrrolate in healing of anastomotic dehiscence after primary repair of esophageal atresia in a low resource setting—A randomized controlled study	2017	Asia	42	Comparative	Prospective	Trial
Vergouwe FWT, et al.	Longitudinal evaluation of growth in oesophageal atresia patients up to 12 years	2017	Europe	126	Observational	Cross-sectional	Cohort
Wei S, et al.	Musculoskeletal deformities following neonatal thoracotomy: long-term follow-up of an esophageal atresia cohort	2017	North-America	52	Comparative	Retrospective	Cohort
Zani A, et al.	Long-term outcomes following H-type tracheoesophageal fistula repair in infants	2017	North-America	16	Observational	Retrospective	Cohort
Zeng Z, et al.	Outcomes of primary gastric transposition for long-gap esophageal atresia in neonates	2017	Asia	14	Observational	Retrospective	Cohort
Amin R, et al.	Long-term Quality of Life in Neonatal Surgical Disease	2018	North-America	62	Observational	Prospective	Cohort
Askarpour S, et al.	Muscle-sparing versus standard posterolateral thoracotomy in neonates with esophageal atresia	2018	Asia	40	Comparative	Prospective	Trial
Bastard F, et al.	Thoracic skeletal anomalies following surgical treatment of esophageal atresia. Lessons from a national cohort	2018	Europe	322	Observational	Cross-sectional	Cohort
Baxter KJ, et al.	Structural airway abnormalities contribute to dysphagia in children with esophageal atresia and tracheoesophageal fistula	2018	North-America	145	Comparative	Retrospective	Cohort
Bradshaw CJ, et al.	Outcomes of Esophageal Replacement: Gastric Pull-Up and Colonic Interposition Procedures	2018	Europe/Africa	32	Comparative	Retrospective	Cohort
Dai J, et al.	Experience of diagnosis and treatment of 31 H-type tracheoesophageal fistula in a single clinical center	2018	Asia	31	Observational	Retrospective	Cohort
Dylkowski D, et al.	Repair of congenital esophageal atresia with tracheoesophageal fistula repair in Ontario over the last 20 years: Volume and outcomes	2018	North-America	465	Observational	Retrospective	Cohort
Foster JD, et al.	Esophageal replacement by gastric transposition: A single surgeon's experience from a tertiary pediatric surgical center	2018	Europe	17	Observational	Retrospective	Cohort
Garabedian C, et al.	Management and outcome of neonates with a prenatal diagnosis of esophageal atresia type A: A population-based study	2018	Europe	88	Comparative	Retrospective	Cohort
Goodarzi M, et al.	Esophageal atresia: Recent five years' mortality and morbidity	2018	Asia	43	Comparative	Retrospective	Cohort

Author	Title	Year	Continent	n =	Study design		
Kamran A, et al.	Slide Esophagoplasty vs End-to-End Anastomosis for Recalcitrant Esophageal Stricture after Esophageal Atresia Repair	2018	North-America	50	Comparative	Retrospective	Cohort
Koivusalo A, et al.	Location of TEF at the carina as an indicator of long-gap C-Type esophageal atresia	2018	Europe	247	Comparative	Retrospective	Cohort
Konig TT, and Muensterer OJ	Physical Fitness and Locomotor Skills in Children With Esophageal Atresia-A Case Control Pilot Study	2018	Europe	12	Observational	Cross-sectional	Cohort
Kovesi T, et al.	Vocal cord paralysis appears to be an acquired lesion in children with repaired esophageal atresia/tracheoesophageal fistula	2018	Europe	64	Observational	Retrospective	Cohort
Lal DR, et al.	Challenging surgical dogma in the management of proximal esophageal atresia with distal tracheoesophageal fistula: Outcomes from the Midwest Pediatric Surgery Consortium	2018	North-America	292	Comparative	Retrospective	Cohort
Leibovitch L, et al.	Infants born with esophageal atresia with or without tracheo-esophageal fistula: Short-and long-term outcomes	2018	Asia	46	Comparative	Cross-sectional	Cohort
Lin CH, et al.	Thoracoscopic repair of esophageal atresia: Comparison with open approach	2018	Asia	21	Comparative	Retrospective	Cohort
Macchini F, et al.	Classification of Esophageal Strictures following Esophageal Atresia Repair	2018	Europe	40	Observational	Retrospective	Cohort
Masuya R, et al.	Predictive factors affecting the prognosis and late complications of 73 consecutive cases of esophageal atresia at 2 centers	2018	Asia	73	Comparative	Retrospective	Cohort
Mawlana W, et al.	Neurodevelopmental outcomes of infants with esophageal atresia and tracheoesophageal fistula	2018	Africa	253	Observational	Retrospective	Cohort
Serel Arslan S, et al.	Chewing Function in Children with Repaired Esophageal Atresia-Tracheoesophageal Fistula	2018	Eurasia	30	Observational	Cross-sectional	Cohort
Stenström P, et al.	Congenital heart disease and its impact on the development of anastomotic strictures after reconstruction of esophageal atresia	2018	Europe	96	Comparative	Retrospective	Cohort
Svoboda E, et al.	A patient led, international study of long term outcomes of esophageal atresia: EAT 1	2018	World	928	Observational	Cross-sectional	Cohort
Vergouwe FWT, et al.	High Prevalence of Barrett's Esophagus and Esophageal Squamous Cell Carcinoma After Repair of Esophageal Atresia	2018	Europe	148	Observational	Prospective	Cohort
Yamoto M, et al.	New prognostic classification and managements in infants with esophageal atresia <sup>101</sup>	2018	Asia	65	Observational	Retrospective	Cohort
Youn JK, et al.	Prospective evaluation of clinical outcomes and quality of life after gastric tube interposition as esophageal reconstruction in children	2018	Asia	25	Observational	Retrospective / Cross-sectional	Case-control
Zhu H, et al.	Diagnosis and management of post-operative complications in esophageal atresia patients in China: A retrospective analysis from a single institution	2018	Asia	172	Observational	Retrospective	Cohort
Ammar S, et al.	Management of esophageal atresia and early predictive factors of mortality and morbidity in a developing country	2019	Africa	42	Observational	Retrospective	Cohort
Boumas N, et al.	Surgical treatment of esophageal atresia: our experience after 24 years	2019	Europe	76	Observational	Retrospective	Cohort
Chiarenza SF, et al.	The Use of Endoclips in Thoracoscopic Correction of Esophageal Atresia: Advantages or Complications?	2019	Europe	32	Observational	Retrospective	Cohort
Ferrand A, et al.	Postoperative noninvasive ventilation and complications in esophageal atresia–tracheoesophageal fistula	2019	North-America	91	Observational	Retrospective	Cohort
Firriolo JM, et al.	Supercharged Jejunal Interposition: A Reliable Esophageal Replacement in Pediatric Patients	2019	North-America	17	Observational	Retrospective	Cohort

Author	Title	Year	Continent	n =	Study design		
Flieder S, et al.	Generic Health-Related Quality of Life after Repair of Esophageal Atresia and Its Determinants within a German-Swedish Cohort	2019	Europe	192	Comparative	Cross-sectional	Cohort
François B, et al.	Predictors of the Performance of Early Antireflux Surgery in Esophageal Atresia	2019	Europe	682	Comparative	Prospective	Cohort
Fung SW, et al.	Vocal cord dysfunction following esophageal atresia and tracheoesophageal fistula (EA/TEF) repair	2019	North-America	197	Comparative	Retrospective	Cohort
Grunder FR, et al.	Should Proton Pump Inhibitors be Systematically Prescribed in Patients with Esophageal Atresia after Surgical Repair?	2019	North-America	73	Comparative	Prospective	Cohort
Ishimaru T, et al.	Impact of congenital heart disease on outcomes after primary repair of esophageal atresia: a retrospective observational study using a nationwide database in Japan	2019	Asia	431	Comparative	Retrospective	Cohort
Jönsson L, et al.	Long-Term Effectiveness of Antireflux Surgery in Esophageal Atresia Patients	2019	Europe	99	Comparative	Retrospective	Cohort
Kumari V, et al.	Developmental Status of Children Operated for Esophageal Atresia with or without Tracheoesophageal Fistula Along with Maternal Stress, Their Quality of life, and Coping Abilities at AIIMS, New Delhi	2019	Asia	51	Observational	Cross-sectional	Cohort
Lal DR, et al.	Infants with esophageal atresia and right aortic arch: Characteristics and outcomes from the Midwest Pediatric Surgery Consortium	2019	North-America	396	Comparative	Retrospective	Cohort
Lawrence, A E et al	Relationships between hospital and surgeon operative volumes and outcomes of esophageal atresia/tracheoesophageal fistula repair	2019	North-America	3085	Comparative	Retrospective	Cohort
Nurminen P, et al.	Pneumonia after Repair of Esophageal Atresia-Incidence and Main Risk Factors	2019	Europe	104	Observational	Retrospective	Cohort
Petit LM, et al.	Prevalence and Predictive Factors of Histopathological Complications in Children with Esophageal Atresia	2019	North-America	85	Observational	Prospective	Cohort
Rayyan M, et al.	Neonatal factors predictive for respiratory and gastro-intestinal morbidity after esophageal atresia repair	2019	Europe	93	Observational	Retrospective	Cohort
Shirota C, et al.	Therapeutic strategy for thoracoscopic repair of esophageal atresia and its outcome	2019	Asia	39	Observational	Retrospective	Cohort
Slater BJ, et al.	Use of magnets as a minimally invasive approach for anastomosis in esophageal atresia: Long-term outcomes	2019	North-America	13	Observational	Retrospective	Cohort
Sømme S, et al.	Neonatal surgery in low- vs. high-volume institutions: a KID inpatient database outcomes and cost study after repair of congenital diaphragmatic hernia, esophageal atresia, and gastroschisis	2019	North-America	1280	Comparative	Retrospective	Cohort
Stadil T, et al.	Surgical repair of long-gap esophageal atresia: A retrospective study comparing the management of long-gap esophageal atresia in the Nordic countries	2019	Europe	71	Comparative	Retrospective	Cohort
Thompson A, et al.	Not all neonates with oesophageal atresia and tracheoesophageal fistula are a surgical emergency	2019	Europe	64	Observational	Retrospective	Cohort
Vergouwe FWT, et al.	Risk factors for refractory anastomotic strictures after oesophageal atresia repair: A multicentre study	2019	Europe	454	Observational	Retrospective	Cohort
Askarpour S, et al.	INCIDENCE OF MALNUTRITION, ESOPHAGEAL STENOSIS AND RESPIRATORY COMPLICATIONS AMONG CHILDREN WITH REPAIRED ESOPHAGEAL ATRESIA	2020	Asia	43	Observational	Retrospective	Cohort
Badran EF, et al.	Esophageal atresia: Associated anomalies, mortality, and morbidity in Jordan	2020	Asia	55	Observational	Retrospective	Cohort
Bevilacqua F, et al.	Fixed the gap, solved the problem? Eating skills in esophageal atresia patients at 3 years	2020	Europe	54	Observational	Cross-sectional	Cohort
Blanco AJ, et al.	Comorbidities and course of lung function in patients with congenital esophageal atresia	2020	Europe	97	Observational	Retrospective	Cohort

Author	Title	Year	Continent	n =	Study design		
Bludevich BM, et al.	30-Day Outcomes Following Esophageal Replacement in Children: A National Surgical Quality Improvement Project Pediatric Analysis	2020	North-America	78	Comparative	Retrospective	Cohort
Campos J, et al.	The burden of esophageal dilatations following repair of esophageal atresia	2020	Oceania	258	Observational	Retrospective	Cohort
Chou CS, et al.	Fiberoptic bronchoesophagoscopy-assisted evaluation and prognostic factor analysis in children with congenital esophageal atresia and tracheoesophageal fistula	2020	Asia	33	Observational	Retrospective	Cohort
Cui X, et al.	Clinical Analysis of Azygos Vein Preservation Under Thoracoscope in the Operation of Type III Esophageal Atresia	2020	Asia	70	Comparative	Retrospective	Cohort
Cui X, et al.	The value of thoracic lavage in the treatment of anastomotic leakage after surgery for type III esophageal atresia	2020	Asia	92	Comparative	Retrospective	Cohort
Dey S, et al.	First-Year follow-up of Newborns Operated for Esophageal Atresia in a Developing Country: Just Operating is not Enough!	2020	Asia	70	Observational	Retrospective	Cohort
Dingemann C, et al.	Low gestational age is associated with less anastomotic complications after open primary repair of esophageal atresia with tracheoesophageal fistula	2020	Europe	75	Observational	Retrospective	Cohort
Donoso F, et al.	Pulmonary function in children and adolescents after esophageal atresia repair	2020	Europe	46	Observational	Retrospective	Cohort
Elbarbary MM, et al.	Outcome of thoracoscopic repair of type-C esophageal Atresia: A single-center experience from North Africa	2020	Africa	46	Observational	Prospective	Cohort
Elhattab A, et al.	Thoracoscopy Versus Thoracotomy in the Repair of Esophageal Atresia with Distal Tracheoesophageal Fistula	2020	Europe	187	Comparative	Retrospective	Cohort
Fernandes E, et al.	Is thoracoscopic esophageal atresia repair safe in the presence of cardiac anomalies?	2020	Oceania	49	Comparative	Retrospective	Cohort
Garcia HJ, et al.	Experience of Minimally Invasive Surgery in Neonates with Congenital Malformations in a Tertiary Care Pediatric Hospital	2020	South America	22	Observational	Prospective	Cohort
Hannon E, et al.	Outcomes in adulthood of gastric transposition for complex and long gap esophageal atresia	2020	Europe	64	Comparative	Cross-sectional	Cohort
Jones CE, et al.	Association Between Administration of Antacid Medication and Anastomotic Stricture Formation After Repair of Esophageal Atresia	2020	Europe	114	Comparative	Retrospective	Cohort
Jones CE, et al.	Repair of oesophageal atresia by consultants and supervised trainees results in similar outcomes	2020	Europe	120	Comparative	Retrospective	Cohort
Kim W, et al.	The learning curve for thoracoscopic repair of esophageal atresia with distal tracheoesophageal fistula: A cumulative sum analysis	2020	Asia	50	Observational	Retrospective	Cohort
Koivusalo A, et al.	Indications, Surgical Complications, and Long-Term Outcomes in Pediatric Esophageal Reconstructions with Pedicled Jejunal Interposition Graft	2020	Europe	14	Observational	Retrospective	Cohort
Kulshrestha S, et al.	Conservative Management of Major Anastomotic Leaks Occurring after Primary Repair in Esophageal Atresia with Fistula: Role of EYestrapleural Approach	2020	Asia	203	Observational	Retrospective	Cohort
Lieber J, et al.	Functional outcome after laparoscopic assisted gastric transposition including pyloric dilatation in long-gap esophageal atresia	2020	Europe	14	Observational	Retrospective	Cohort
Mikkelsen A, et al.	Traumatic stress, mental health and quality of life in adolescents with esophageal atresia	2020	Europe	68	Comparative	Cross-sectional	Case-control
Nguyen MVL, et al.	The value of prophylactic chest tubes in tracheoesophageal fistula repair	2020	North-America	109	Comparative	Retrospective	Case-control
Pelizzo G, et al.	Esophageal Atresia: Nutritional Status and Energy Metabolism to MaYesimize Growth Outcome	2020	Europe	21	Observational	Cross-sectional	Cohort

Author	Title	Year	Continent	n =	Study design		
Pruitt LCC, et al.	Impact of consolidation of cases on post-operative outcomes for indexes pediatric surgery cases	2020	North-America	673	Comparative	Retrospective	Cohort
Puliński M, et al.	Congenital esophageal atresia treated with thoracoscopic approach – Results of surgical treatment	2020	Europe	28	Observational	Retrospective	Cohort
Quiroz HJ, et al.	Nationwide analysis of mortality and hospital readmissions in esophageal atresia	2020	North-America	3157	Observational	Retrospective	Cohort
Ritz LA, et al.	Outcome of Patients With Esophageal Atresia and Very Low Birth Weight ( $\leq 1,500$ g)	2020	Europe	48	Comparative	Cross-sectional	Cohort
Rozeik AE, et al.	Thoracoscopic versus conventional open repair of tracheoesophageal fistula in neonates: A short-term comparative study	2020	Africa	30	Comparative	Prospective	Trial
Serel Arslan S, et al.	Assessment of the Concerns of Caregivers of Children with Repaired Esophageal Atresia-Tracheoesophageal Fistula Related to Feeding-Swallowing Difficulties	2020	Eurasia	64	Observational	Cross-sectional	Cohort
Soyer T, et al.	The effect of azygos vein preservation on postoperative complications after esophageal atresia repair: Results from the Turkish Esophageal Atresia Registry	2020	Eurasia	315	Comparative	Retrospective	Cohort
Tan Tanny SP, et al.	Predictors of Mortality after Primary Discharge from Hospital in Patients with Esophageal Atresia	2020	Oceania	650	Observational	Prospective	Cohort
Zhang J, et al.	Clinical analysis of surgery for type III esophageal atresia via thoracoscopy: a study of a Chinese single-center experience	2020	Asia	92	Comparative	Retrospective	Cohort
Acharya SK, et al.	Gastric pull-up by the retrosternal route for esophageal replacement: Feasibility in a limited-resource scenario	2021	Asia	18	Observational	Retrospective	Cohort
Aworanti OM, et al.	Extubation strategies after esophageal atresia repair	2021	Europe	46	Comparative	Retrospective	Cohort
Ax SÓ, et al.	Parent-reported feeding difficulties among children born with esophageal atresia: Prevalence and early risk factors	2021	Europe	114	Observational	Cross-sectional	Cohort
Baghdadi O, et al.	Initial Esophageal Anastomosis Diameter Predicts Treatment Outcomes in Esophageal Atresia Patients With a High Risk for Stricture Development	2021	North-America	121	Observational	Retrospective	Cohort
Bence CM, et al.	Clinical outcomes following implementation of a management bundle for esophageal atresia with distal tracheoesophageal fistula	2021	North-america	170	Comparative	Retrospective	Cohort
Besendörfer M, et al.	Association of clinical factors with postoperative complications of esophageal atresia	2021	Europe	43	Observational	Retrospective	Cohort
Burnett AC, et al.	Cognitive, academic, and behavioral functioning in school-aged children born with esophageal atresia	2021	Oceania	98	Observational	Cross-sectional	Cohort
Chiang CM, et al.	Risk factors and management for anastomotic stricture after surgical reconstruction of esophageal atresia	2021	Asia	40	Observational	Retrospective	Cohort
Di Natale A, et al.	Long-Term Outcomes and Health-Related Quality of Life in a Swiss Patient Group with Esophageal Atresia	2021	Europe	30	Observational	Cross-sectional	Cohort
Etchill EW, et al.	Association of operative approach with outcomes in neonates with esophageal atresia and tracheoesophageal fistula	2021	North-America	855	Comparative	Retrospective	Cohort
Flatrès C, et al.	Prevalence of acid gastroesophageal reflux disease in infants with esophageal atresia/tracheoesophageal fistula	2021	Europe	70	Observational	Prospective	Cohort
Folaranmi SE, et al.	Influence of birth weight on primary surgical management of newborns with esophageal atresia	2021	Europe	198	Comparative	Retrospective	Cohort
Francesca B, et al.	Neurodevelopmental outcome in infants with esophageal atresia: risk factors in the first year of life	2021	Europe	82	Observational	Prospective	Cohort

Author	Title	Year	Continent	n =	Study design		
Gallo G, et al.	Quality of life after esophageal replacement in children	2021	Europe	24	Comparative	Cross-sectional	Cohort
Harrington AW, et al.	Nutrition delivery and growth outcomes in infants with long-gap esophageal atresia who undergo the Foker process	2021	North-America	45	Observational	Retrospective	Cohort
Hew NLC, et al.	Predictors of poor outcomes in children with tracheoesophageal fistula/oesophageal atresia: An Australian experience	2021	Oceania	103	Observational	Retrospective	Cohort
Huang JX, et al.	Risk factors for anastomotic complications after one-stage anastomosis for oesophageal atresia	2021	Asia	107	Observational	Retrospective	Cohort
Jo Svetanoff W, et al.	The left-sided repair: An alternative approach for difficult esophageal atresia repair	2021	North-America	47	Comparative	Retrospective	Cohort
Lejeune S, et al.	Esophageal atresia and respiratory morbidity	2021	Europe	1287	Observational	Prospective	Cohort
Lu YH, et al.	Risk factors for digestive morbidities after esophageal atresia repair	2021	Asia	72	Observational	Retrospective	Cohort
Maan M, et al.	Growth and Development Assessment of Children (1-5 Years) Operated for Tracheoesophageal Fistula/Esophageal Atresia: A Case Control study	2021	Asia	40	Comparative	Retrospective	Case-control
O'Connor E, and Jaffray B	Surgeon-Level Variation in Outcome following Esophageal Atresia Repair Is Not Explained by Volume	2021	Europe	190	Observational	Retrospective	Cohort
Oliver DH, et al.	Favorable Outcome of Electively Delayed Elongation Procedure in Long-Gap Esophageal Atresia	2021	Europe	13	Observational	Retrospective	Cohort
Ozsin-Ozler C, et al.	Oral Health Status among Children with Repaired Esophageal Atresia	2021	Eurasia	35	Observational	Cross-sectional	Cohort
Oztan MO, et al.	Outcome of Very Low and Low Birth Weight Infants with Esophageal Atresia: Results of the Turkish Esophageal Atresia Registry	2021	Eurasia	389	Comparative	Prospective	Cohort
Patterson K, et al.	Quantifying Upper Aerodigestive Sequelae in Esophageal Atresia/Tracheoesophageal Fistula Neonates	2021	North-America	2509	Observational	Retrospective	Cohort
Pham A, et al.	Feeding disorders in children with oesophageal atresia: A cross-sectional study	2021	Europe	145	Observational	Prospective / Cross-Sectional	Cohort
Saiad MO	A wave-like anastomosis, a new technique of anastomosis to prevent stricture after oesophageal atresia repair	2021	Africa	49	Comparative	Retrospective	Cohort
Samraj P, et al.	Primary anastomosis in difficult cases of type "C" esophageal atresia: The atraumatic microvascular clamp technique of minimal tension with good outcome	2021	Asia	32	Observational	Prospective	Cohort
Schmedding A, et al.	Outcome of esophageal atresia in Germany	2021	Europe	287	Observational	Retrospective	Cohort
Sfeir R, et al.	Risk Factors of Early Mortality and Morbidity in Esophageal Atresia with Distal Tracheoesophageal Fistula: A Population-Based Cohort Study	2021	Europe	1008	Observational	Prospective	Cohort
Sinopidis X, et al.	Oesophageal atresia without major cardiovascular anomalies: Is management justified at a district paediatric surgical institution?	2021	Europe	33	Comparative	Retrospective	Cohort
Son J, et al.	Thoracoscopic repair of esophageal atresia with distal tracheoesophageal fistula: is it a safe procedure in infants weighing less than 2000 g?	2021	Asia	41	Comparative	Retrospective	Cohort
Sreeram, II, et al.	Patient-Reported Outcome Measures and Clinical Outcomes in Children with Foregut Anomalies	2021	Europe	93	Observational	Cross-sectional	Cohort
Svetanoff WJ, et al.	Contemporary outcomes of the Foker process and evolution of treatment algorithms for long-gap Esophageal Atresia	2021	North-America	102	Comparative	Retrospective	Cohort

Author	Title	Year	Continent	n =	Study design		
Taghavi K, et al.	H-type congenital tracheoesophageal fistula: Insights from 70 years of The Royal Children's Hospital eYesperience	2021	Oceania	56	Observational	Prospective	Cohort
Tannuri ACA, et al.	Esophageal substitution or esophageal elongation procedures in patients with complicated esophageal atresia? Results of a comparative study	2021	South America	276	Observational	Cross-sectional	Cohort
Thakkar H, et al.	Thoracoscopic oesophageal atresia/tracheo-oesophageal fistula (OA/TOF) repair is associated with a higher stricture rate: a single institution's experience	2021	Europe	95	Comparative	Retrospective	Cohort
Thompson K, et al.	Evolution, lessons learned, and contemporary outcomes of esophageal replacement with jejunum for children	2021	North-America	48	Comparative	Retrospective	Cohort
Toczewski K, et al.	Thoracoscopic repair of congenital isolated H-type tracheoesophageal fistula	2021	Europe	12	Observational	Retrospective	Cohort
Tuğcu GD, et al.	Evaluation of pulmonary complications and affecting factors in children for repaired esophageal atresia and tracheoesophageal fistula	2021	Eurasia	71	Observational	Retrospective / Cross-sectional	Cohort
van Hoorn CE, et al.	Primary repair of esophageal atresia is followed by multiple diagnostic and surgical procedures	2021	Europe	102	Comparative	Retrospective	Cohort
van Hoorn CE, et al.	Associations of perioperative characteristics with motor function in preschool children born with esophageal atresia	2021	Europe	53	Observational	Retrospective / Cross-sectional	Cohort
van Tuyll van Serooskerken ES, et al.	Childhood outcome after correction of long-gap esophageal atresia by thoracoscopic external traction technique	2021	Europe	11	Observational	Retrospective	Cohort
van Tuyll van Serooskerken ES, et al.	Thoracoscopic Repair of Esophageal Atresia	2021	Europe	11	Comparative	Retrospective	Cohort
Yasuda JL, et al.	Prophylactic negative vacuum therapy of high-risk esophageal anastomoses in pediatric patients	2021	North-America	14	Comparative	Retrospective	Case-control

### SUPPLEMENTARY 3: Extracted baseline characteristics

\*As several parameters could arguably be included in more than one category, categorization followed agreement between the first two authors. We further arranged the parameters by topic, such as comorbidities, primary treatment, specific complications and long-term outcome, to facilitate the comparison of variation in studied parameters within similar subjects. Underlined items were studied in more than 5% of included publications.

#### Patient characteristics

	n =	%		n =	%
<u>Age at time of filling in questionnaire / intervention / time of follow up</u>	<u>56</u>	27%	Family history of horse shoe kidney	1	<1%
<u>Age at presentation / admission / diagnosis</u>	<u>22</u>	11%	Aortic arch: side	5	2%
Age at referral from other hospital	1	<1%	Alcohol consumption	1	<1%
Age at study completion	1	<1%	Tobacco smoking	1	<1%
<u>Mortality prediction (Spitz classification)</u>	<u>12</u>	6%	<u>Weight at operation</u>	<u>41</u>	20%
Mortality prediction (Waterston classification)	4	2%	Weight for gestational age	4	2%
Mortality prediction (Montreal classification)	1	<1%	Weight-for-age at admission	2	1%
<u>Weight at birth</u>	<u>144</u>	69%	Head circumference	1	<1%
Birth height / length	6	3%	SD from weight at gestational age	1	<1%
Mode of delivery: C-section versus Vaginal	6	3%	Weight-for-age at operation	1	<1%
Birth order    First pregnancy	3	1%	<u>Gender</u>	<u>178</u>	85%
Inborn versus Outborn	2	1%	<u>Gestational age or prematurity</u>	<u>154</u>	74%
Mode of conception: IVF or spontaneous	2	1%	<u>Previous esophageal repair/surgery</u>	<u>27</u>	13%
Incidence conception (when)	1	<1%	<u>Referred from other hospital</u>	<u>25</u>	12%
Season of birth	1	<1%	<u>Race/Ethnicity</u>	<u>18</u>	9%
Family history of EA	3	1%	<u>Intra-uterine growth / Small for gestational age</u>	<u>11</u>	5%
Family history of congenital anomalies	1	<1%	<u>Twin / Multiple birth</u>	<u>11</u>	5%
			Education	3	1%
			Occupation	1	<1%
			Originating country	1	<1%
			Year of admission	1	<1%

#### Disease characteristics

	n =	%		n =	%
Presenting symptoms	10	5%	<u>Long Gap / Gap length</u>	<u>97</u>	46%
Site of fistula / TEF	5	2%	<u>Fistula: yes/no</u>	<u>28</u>	13%
TEF diameter (fistula)	1	<1%	Clinical EA-severity	2	1%
<u>Type of EA</u>	<u>154</u>	74%			



## Comorbidities

	n =	%		n =	%
Abdominal wall	2	1%	Number of associated anomalies	9	4%
Inguinal Hernia	1	<1%	Major associated anomaly	4	2%
Omphalocele	1	<1%	Minor associated anomaly	2	1%
<u>Pulmonary/respiratory conditions</u>	<u>36</u>	17%	No comorbidities	2	1%
IRDS / Neonatal Respiratory Distress Syndrome	8	4%	Associated malformation affecting growth	1	<1%
Anatomical variations on fiberbronchoscopy	1	<1%	Multiple anomalies	1	<1%
Lung hypoplasia (preoperative)	1	<1%	<u>Cardiac malformation / Congenital heart disease</u>	<u>149</u>	71%
Lung maturation	1	<1%	Great vessel anomalies / Vascular	9	4%
Structural Airway Anomaly	1	<1%	Cardiac severity score (based on diagnosis, clinical)	1	<1%
Three main bronchi	1	<1%	Severity of associated cardiac anomalies (Rachs score)	1	<1%
Tracheal anomaly	1	<1%	Hematologic disorder	2	1%
<u>ARM</u>	<u>67</u>	32%	<u>Laryngeal cleft / Laryngo-tracheo-oesophageal cleft</u>	<u>12</u>	6%
<u>Chromosomal/Genetic abnormalities</u>	<u>82</u>	39%	Laryngeal atresia	1	<1%
Apgar Score at birth	4	2%	Laryngeal stenosis	1	<1%
Asphyxia at birth	2	1%	<u>Musculoskeletal or limb anomalies</u>	<u>74</u>	35%
Recussitation at birth	1	<1%	Polydactyly	1	<1%
Score for Neonatal Acute Physiology with Perinatal Extension-II (SNAPPE II)	1	<1%	<u>Neurologic or central or CNS anomalies</u>	<u>31</u>	15%
Anomalies of the head or neck	10	5%	Atopy	1	<1%
Anomalies of the palatum	6	3%	Spina bifida	1	<1%
Choanal atresia	6	3%	Tethered cord	1	<1%
Cleft lip	1	<1%	<u>Renal/Genitourinary anomalies</u>	<u>76</u>	36%
Craniosynostosis	1	<1%	<u>Otolaryngeal anomalies / Auditory / Hearing issues</u>	<u>13</u>	6%
Oropharyngeal abnormalities	1	<1%	<u>VACTERL</u>	<u>88</u>	42%
CDH	4	2%	<u>CHARGE</u>	<u>23</u>	11%
Right-sided diaphragmatic relaxation	1	<1%	Poland syndrome	1	<1%
Eye anomaly / coloboma	2	1%	VATER	1	<1%
Blind	1	<1%	Hypothyroidism	1	<1%
<u>Intestinal malformation</u>	<u>57</u>	27%	Subglottic stenosis (preoperative)	4	2%
Congenital esophageal stenosis	6	3%	ASA class	3	1%
Necrotizing enterocolitis	6	3%			
<u>At least one associated anomaly</u>	<u>83</u>	4<1%			
"Other" comorbidity (unspecified)	<u>23</u>	11%			

## Paternal characteristics

	n =	%		n =	%
Maternal age	8	4%	Maternal problems	1	<1%
Insurance status	6	3%	Maternal profession	1	<1%
Socioeconomic status	6	3%	Migration background	1	<1%
Income	3	1%	Number of family members maternal side	1	<1%
Paternal age	2	1%	Parental age	1	<1%
Residential area	2	1%	Parental employment status	1	<1%
Sociodemographic factors	2	1%	Parental gender	1	<1%
(Number of) siblings	1	<1%	Parental health status	1	<1%
Age of caregivers (at time of filling in questionnaire / intervention)	1	<1%	Parental partnership	1	<1%
Consanguinity	1	<1%	Primary language spoken at home	1	<1%
Maternal education / Parental education	1	<1%	Primary payer (government, private, other)	1	<1%

## SUPPLEMENTARY 4: Extracted treatment- and care-process characteristics

\*As several parameters could arguably be included in more than one category, categorization followed agreement between the first two authors. We further arranged the parameters by topic, such as comorbidities, primary treatment, specific complications and long-term outcome, to facilitate the comparison of variation in studied parameters within similar subjects. Underlined items were studied in more than 5% of included publications.

### Hospital structure

	n =	%		n =	%
Born in tertiary center	1	<1%	Treating specialties last year	1	<1%
Hospital of delivery: pediatrician available?	1	<1%	Hospital type	3	1%
Transfer after birth	1	<1%	Hospital location / teaching	1	<1%
Multidiscipline specialist team for follow-up	4	2%	Hospital volume / Annual institutional volume (number of procedures per year) / Number of procedures per surgeon	9	4%
Lost to follow-up	1	<1%	Hospital size	5	2%
Number of visits in the outpatient otolaryngology clinic	1	<1%			

### Diagnostics

	n =	%		n =	%
Antenatal: amniocentesis / chorionic villous sampling	2	1%	Number of bronchoscopies	1	<1%
Antenatal: EA suspected during prenatal MRI	2	1%	<u>Contrast study postoperative</u>	<u>13</u>	6%
<u>Polyhydramnios on antenatal ultrasound</u>	<u>19</u>	9%	Contrast study (preop) / Prone tube esophagogram (preop, type E)	5	2%
Antenatal ultrasound: yes or no	9	4%	Contrast medium swallowing (barium) / Fluoroscopic swallow study	4	2%
Antenatal ultrasound: absent stomach	8	4%	Contrast study, post-op: time between repair and contrast study	4	2%
Antenatal ultrasound: upper pouch sign	3	1%	Contrast study, post-op: routine	2	1%
Antenatal: EA suspected during prenatal sonography	3	1%	Contrast study: regular follow-up	1	<1%
<u>Bronchoscopy (intraoperative)</u>	<u>22</u>	11%	Iodized Oil Radiography (preoperative)	1	<1%
Bronchoscopy (postoperative)	3	1%	Postoperative iodine oil contrast exam: number of days postoperative	1	<1%
Fiberoptic bronchoesophagoscopy-assisted diagnostics / interventions (a wide range)	3	1%	Postoperative iodine oil contrast exam: yes or no	1	<1%
Tracheoscopy at some point in time	3	1%	Chest CT at some point in time (yes or no)	4	2%
BAL	2	1%	Chest CT (preop)	2	1%
Age at fiberoptic bronchoesophagoscopy	1	<1%	<u>Echocardiography</u>	<u>11</u>	5%
Age at first bronchoscopy postoperative	1	<1%	Endoscopy: (first) surveillance / GI series	6	3%

	n =	%		n =	%
Endoscopy at some point in time (yes or no)	3	1%	Neutrophil count	1	<1%
Endoscopy including pH measurement (m1,m3,m6 and yearly)	2	1%	<u>Lung function (spirometry) in follow-up</u>	<u>13</u>	6%
Endoscopy: median age at first endoscopy	2	1%	Body plethysmography	2	1%
Esophagoscopy / Upper endoscopy (preoperative)	2	1%	Spirometry: age at	1	<1%
Endoscopy peroperative (type E)	1	<1%	Spirometry: diffusion capacity	1	<1%
Endoscopy: conscious sedation	1	<1%	Mode of diagnosis: H-type fistula	1	<1%
Endoscopy: surveillance, time between endoscopies	1	<1%	MRI performed	1	<1%
Time (duration) endoscopic follow-up / Surveillance	1	<1%	Manometry	2	1%
Site of fistula consistent with preoperative examination	1	<1%	Physical examination	2	1%
Gap measurement (X-ray, bronchoscopy and bronchoscopy and a rigid instrument)	3	1%	Age at first pH-metry	1	<1%
Difference between radiographic and intraoperatively measured gap	1	<1%	Determination size / volume stomach	1	<1%
Albumin after surgery	2	1%	Oral-pharyngeal motility study: any aspiration	1	<1%
Lab: pCO2	2	1%	Oral-pharyngeal motility study	1	<1%
Lab: pH	2	1%	Total screening for VACTERL	1	<1%
Albumin / globulin	1	<1%	pH impedance measures / Ri at / 24h pH monitoring	9	4%
Hemoglobin after surgery	1	<1%	<u>Prenatal diagnosis</u>	<u>26</u>	12%
Lab: Hb (preop)	1	<1%	Antenatal: counseling with antenatal specialist	1	<1%
Lab: hematocrit	1	<1%	Antenatal: gestational age at prenatal diagnosis	1	<1%
Lab: standard bicarbonate	1	<1%	Antenatal: in utero transfer	1	<1%
Lab: WBC (preop)	1	<1%	Ultrasound: renal	4	2%
Lymphocyte count	1	<1%	Ultrasound: abdominal	1	<1%
Neutrophil / Lymphocyte Ratio	1	<1%	Number of video fluoroscopic feeding studies	1	<1%
			Plain X-ray: chest (with or without nasogastric tube inserted)	6	3%
			Plain X-ray: abdominal	1	<1%

### Primary treatment

	n =	%		n =	%
<u>Age at operation</u>	<u>98</u>	47%	Number of hospital admissions / hospitalizations / hospitalization rate	8	4%
<u>Time between diagnosis and surgery (delay)</u>	<u>13</u>	6%	Age at diagnosis	3	1%
			Time (duration) of paralysis	5	2%

	n =	%		n =	%
Number of episodes of general anaesthesia	4	2%	Parenteral feeding	6	3%
Time (duration) of anaesthesia	4	2%	Time (duration) of tube feeding	6	3%
Cumulative time (duration) of anaesthesia (during first two years)	2	1%	Time to full enteral feeding	5	2%
Mean End-tidal CO <sub>2</sub> (EtCO <sub>2</sub> ) at end of surgery	2	1%	Preoperative nutrition	4	2%
Mean End-tidal CO <sub>2</sub> (EtCO <sub>2</sub> ) at start of surgery	2	1%	Time (duration) parenteral feeding	4	2%
Mean maximum EtCO <sub>2</sub>	2	1%	Time to extubation transanastomotic tube	4	2%
Blood gas: arterial or capillary	1	<1%	Bottle use	1	<1%
Difficulty weaning from methadone and lorazepam	1	<1%	Breast milk taking methods	1	<1%
Highest heartrate during surgery	1	<1%	Breast milk taking period	1	<1%
Highest MAP during surgery	1	<1%	Breast milk: yes or no	1	<1%
Lowest MAP during surgery	1	<1%	Nasogastric tube at discharge	1	<1%
PaO <sub>2</sub> during surgery	1	<1%	Nasogastric tube postoperatively	1	<1%
ECMO (yes vs. no)	3	1%	Pacifier use	1	<1%
<u>Elongation procedure</u>	<u>19</u>	9%	Tube feeding at time of discharge from initial NICU	1	<1%
Time between beginning Foker / elongation to anastomosis	6	3%	Fistula: undiagnosed TEF	5	2%
Elongation: Loss of traction sutures	3	1%	<u>Gastrostomy at any point in time</u>	<u>81</u>	39%
Elongation: age at start Foker / elongation	2	1%	Gastrostomy: age at	2	1%
Elongation: weight at start Foker	2	1%	Gastrostomy: time to closure	2	1%
Elongation, preoperative (techniques)	1	<1%	Gastrostomy button placement	1	<1%
Elongation: number of steps	1	<1%	Gastrostomy type (image-guided percutaneous vs Stamm surgical)	1	<1%
Foker: primary or rescue	1	<1%	Gastrostomy: open versus laparoscopically	1	<1%
Gastropexy before elongation	1	<1%	<u>Number of surgeries</u>	<u>11</u>	5%
Time to anastomosis with magnets	1	<1%	Distal jejunal conduit anastomosis: roux-en-y or straight to stomach	1	<1%
<u>Cervical esophagostomy until surgery / at any point in time</u>	<u>33</u>	16%	Length of jejunal conduit	1	<1%
<u>Time to start oral feeding</u>	<u>29</u>	14%	<u>Time on intensive care (days)</u>	<u>27</u>	13%
<u>Time to start tube feeding</u>	<u>14</u>	7%	<u>Time (duration) primary hospital stay</u>	<u>78</u>	37%
<u>Time to full oral feeding</u>	<u>13</u>	6%	Age at discharge	1	<1%
Method of feeding (TPN, gastric / jejunal via transanas tube / gastrostomy tube)	8	4%	<u>Use of anti-acid medication (at any point in time)</u>	<u>61</u>	29%
			Medication: anti-acid, duration	3	1%
			Physiotherapy / Physical therapy	3	1%
			Speech pathologists	2	1%
			Cannulation of the TEF tract (preoperative)	3	1%

	n =	%		n =	%
Medication: (perioperative) steroiduse	2	1%	<u>Conversion -scopy to -tomy</u>	29	14%
Fasting time	1	<1%	<u>Tension-free anastomosis / Concern regarding tension</u>	27	13%
Incidence of TAT dislodgment (re-intubation)	1	<1%	<u>Intra-operative chest tube</u>	23	11%
Medication: antibiotic profylaxis	4	2%	<u>Transanastomotic tube</u>	21	1<1 %
Medication: vasopressors, intraoperative	2	1%	<u>Thoracotomy: left or right approach</u>	12	6%
Medication: vasopressors, preoperative	2	1%	Thoracotomy: surgical approach (axillary, lateral, pleural / extrapleural)	7	3%
Mode of blood pressure measurement	1	<1%	Experience of surgeon	5	2%
Medication: continuous muscle relaxation postoperatively / Neuromuscular blockade	3	1%	Azygos vein: division (yes or no)	4	2%
Medication: extension of postoperative antibiotics	2	1%	Interposition material between esophageal and tracheal suture lines	4	2%
Medication: inotrope support / Vasopressor	2	1%	Number of surgeons	4	2%
Medication: vasopressors, postoperative	2	1%	Anastomosis: type of sutures	3	1%
Medication: catecholamine use	1	<1%	Conversion: causes for conversion	3	1%
Medication: coagulation factor XIII postoperative	1	<1%	Emergency procedure	3	1%
Medication: duration of analgesia	1	<1%	Interposition material (autologous versus synthetic)	3	1%
Medication: type of analgesia	1	<1%	Number of surgeries until anastomosis	3	1%
Preoperative nasoesophageal replegle	2	1%	Number of thoracotomies	3	1%
Duration hospital stay preoperative	1	<1%	TEF: cervical approach (fistula)	3	1%
Home management before definitive repair with portable suction	1	<1%	Anastomosis: number of sutures	2	1%
Jejunostomy	6	3%	Esophageal replacement after first surgery	2	1%
Cervical esophagostomy: right or left sided	1	<1%	Fistula: suture ligated OR transfix and divided (type E)	2	1%
Colostomy	1	<1%	Jejunal interposition with supercharge of a and v mamma interna	2	1%
Duodenoduodenostomy	1	<1%	Livaditis myotomy	2	1%
Time to closure jejunostomy	1	<1%	TEF ligation: type of closure (sutures, clips); fistula	2	1%
<u>Type of repair (primary, secondary, anastomosis/interposition)</u>	125	6<1 %	Thoracotomy: location of skin incision	2	1%
<u>Thoracotomy or thoracoscopy</u>	79	38%	Thoracotomy: muscle sparing	2	1%
<u>Operation time/duration</u>	37	18%	Time to fistula closure	2	1%
			Timing esophageal replacement	2	1%

	n =	%		n =	%
Atraumatic microvascular clamp: application time	1	<1%	Postoperative mechanical ventilation / intubation(yes or no)	4	2%
Chest tube: extrapleural versus intrapleural	1	<1%	Time (days) on ventilator, initial	4	2%
Conversion extrapleural to transpleural	1	<1%	CPAP duration after intubation	2	1%
Conversion left thoracotomy to right thoracotomy	1	<1%	Ventilation: extubated from the OR	2	1%
Date of primary surgery	1	<1%	CPAP after intubation	1	<1%
Difficult anastomosis	1	<1%	CPAP time from repair to when CPAP was commenced	1	<1%
End-to-end / side-to-end	1	<1%	Indication for re-intubation	1	<1%
Fistula: suture ligated OR transected and divided (type E)	1	<1%	Intubation: repeated oro-tracheal (number of)	1	<1%
Incision length	1	<1%	Need for intensive respiratory support	1	<1%
Lower pouch mobilisation	1	<1%	Number of intubated days after reintubation before final extubation	1	<1%
Operation on weekday or weekendday	1	<1%	Number of times (periods) on ventilation	1	<1%
Serratus anterior status (division vs. sparing)	1	<1%	Oxygen support: duration	1	<1%
Sternotomy	1	<1%	Post-operative paralysis, ventilation and neck flexion: yes or no	1	<1%
Surgery: technique	1	<1%	Reintubation: unplanned	1	<1%
Sutures: type of intercostal	1	<1%	Route of intubation	1	<1%
TEF: age at closure	1	<1%	Single-lung ventilation	1	<1%
TEF: age at diagnosis	1	<1%	Time (duration) of ventilation prior to fistula ligation	1	<1%
Thoracotomy: intercostal incision (periostal vs muscle section)	1	<1%	Total reintubation episodes	1	<1%
Time to reaching pleural cavity by the surgeon	1	<1%	Ventilation: postoperative mode	1	<1%
Upper pouch mobilisation	1	<1%	Ventilation: preoperative mode	1	<1%
Use of endoscopic clips	1	<1%	Chest tube: required second / post-operative placement / emergency	8	4%
Stabilization time before surgery	1	<1%	Time (duration) IOCT / Chest tube	8	4%
<u>Ventilation period / Duration of ventilation</u>	<u>64</u>	<u>31%</u>	Parents receiving sufficient information during inpatient stay	1	<1%
<u>Need for preoperative intubation/ventilation</u>	<u>19</u>	<u>9%</u>	Parents referred to additional information sources	1	<1%
Need for postoperative ventilation	9	4%			
Re-intubation	6	3%			
Oxygen support	4	2%			

## Treatment of comorbidities

	n =	%		n =	%
Surgeries for associated anomalies	9	4%	Number of abdominal surgeries	1	<1%
Cardiac surgical correction	6	3%	Number of limb surgeries	1	<1%
Pylorotomy / pyloroplasty	5	2%	Number of urological surgeries	1	<1%
Surgical intervention bij ENT specialist	2	1%	Surgery for ARM	1	<1%
Musculoskeletal deformities, requiring active treatment	1	<1%	Surgery for scoliosis / chest wall deformity / winged scapula	1	<1%

## Treatment of complications

	n =	%		n =	%
Number of eVAC changes	1	<1%	Stricture, recalcitrant: electrocautery incisional therapy	1	<1%
Total eVAC duration	1	<1%	Stricture, recalcitrant: resection technique	1	<1%
<u>Number of dilatations</u>	<u>74</u>	<u>35%</u>	Time between subsequent dilatations	1	<1%
Stricture: placement of stent	7	3%	Time between surgery and anastomotic stricture	1	<1%
Stricture: resection	7	3%	Graft removal	2	1%
Dilatation: technique	6	3%	Dilatation for congenital esophageal stenosis	1	<1%
Dilatations: age at first	6	3%	Gastric transposition: pyloric dilatations	1	<1%
Time to first dilatation	5	2%	rTEF closure: endoluminal methods	4	2%
Dilatations: number of at first year	3	1%	rTEF: attempted procedures prior to referral	1	<1%
Dilatations: period of repeated dilatations (mean)	3	1%	Fundoplication: age at operation	8	4%
Stricture, recalcitrant: steroid injections intralesional	3	1%	Fundoplication: redo	3	1%
Dilatation: time during which dilatations were required	2	1%	Age at redo fundoplication	1	<1%
Number of dilatations during follow-up	2	1%	Antireflux surgery: number of surgeries required	1	<1%
Stricture, recalcitrant: timing of resection	2	1%	<u>Tracheostomy</u>	<u>27</u>	<u>13%</u>
Admission for complicated dilatation: length of hospital stay	1	<1%	Aortopexy: age at aortopexy	2	1%
Age at which there was a complication of dilatation	1	<1%	Tracheomalacia: tracheal stent	2	1%
Dilatation: diameter of dilation procedure	1	<1%	Aortopexy: operative approach	1	<1%
Number of intralesional steroid injections performed	1	<1%	Aortopexy: redo	1	<1%
Planned dilatation before discharge	1	<1%	Tests to diagnose tracheomalacia	1	<1%
Stenosis, esophageal: treatment for (Yes vs. No)	1	<1%	Tracheomalacia: time between first symptoms and investigation	1	<1%
Stent postoperative	1	<1%	Tracheostomy: age at procedure	1	<1%



## Treatment of long-term morbidity

	n =	%		n =	%
Eosinophilic esophagitis: treatment	2	1%	Elimination diet (because of esophagitis)	1	<1%
Age at laryngotracheoplasty	1	<1%	Age at asthma diagnosis	1	<1%
Laryngotracheoplasty	1	<1%	Airway clearance techniques as part of daily routine	1	<1%
<u>Use of inhalation medication</u>	<u>12</u>	6%	Age at which severe psychological stress was reported	1	<1%
Medication: prokinetics / promotility	4	2%	<u>Duration of follow-up</u>	<u>82</u>	39%
Medication: antibiotics on regular basis	3	1%	Visit to allergy consultant	1	<1%
Medication: regular medicine use	2	1%	Visit to ENT-doctor	1	<1%
Medication: antibiotics (chronic treatment, macrolide)	1	<1%	Currently under care	1	<1%
Occupational therapy	1	<1%	Vaccination status	1	<1%

## Readmission and reoperation

Time spent in hospital during first year / first five years	4	2%	Reoperation: duration IOCT (fistula)	1	<1%
Readmission: cause	2	1%	Reoperation: duration of hospital stay (fistula)	1	<1%
Readmission: number of	2	1%	Reoperation: duration postop ventilation (fistula)	1	<1%
Re-admissions (total duration)	2	1%	Reoperation: operating time (fistula)	1	<1%
Readmission: intensive care unit	1	<1%	Reoperation: unplanned	2	1%
Readmission: unplanned	1	<1%	Reoperation: age at	1	<1%
Reoperation: surgical method (fistula)	2	1%	Reoperation: elective	1	<1%

## SUPPLEMENTARY 5: Extracted outcomes

\*As several parameters could arguably be included in more than one category, categorization followed agreement between the first two authors. We further arranged the parameters by topic, such as comorbidities, primary treatment, specific complications and long-term outcome, to facilitate the comparison of variation in studied parameters within similar subjects. Underlined items were studied in more than 5% of included publications.

### Complications

	n =	%		n =	%
<u>Anastomotic leakage</u>	<u>143</u>	68%	Stricture index, endoscopically measured	1	<1%
Anastomotic dehiscence	8	4%	Stricture index, radiologically measured	1	<1%
Anastomotic leak: time to settlement conservative	3	1%	Stricture: ability to pass an endoscope	1	<1%
Anastomotic leak requiring reintervention	2	1%	Dilatation for anastomotic stricture	1	<1%
Anastomotic leakage: duration until onset	2	1%	<u>Wound issues</u>	<u>20</u>	1<1%
Esophageal continuity at discharge	2	1%	<u>Esophageal perforation after dilatation</u>	<u>11</u>	5%
Anastomotic leakage: contents of leak	1	<1%	Esophagitis	9	4%
Anastomotic leakage: stopped, decreasing or persistent	1	<1%	Barrett's oesophagus	7	3%
Anastomotic leakage: volume of leak per day	1	<1%	Esophageal diverticulum	4	2%
Long term anastomotic failure	1	<1%	Perforation, esophageal	4	2%
Technical failure of the eVAC	1	<1%	Achalasia	2	1%
Thoracic drainage volume	1	<1%	Esophageal varices	2	1%
<u>Anastomotic stricture/stenosis</u>	<u>150</u>	72%	Columnar-lined esophagus	1	<1%
<u>Dilatations: yes or no</u>	<u>128</u>	61%	Dilated proximal esophagus	1	<1%
Refractory stricture	3	1%	Ulcer / Gastric bleeding	9	4%
Stricture, dilatation-resistant and indication for revision	3	1%	Adhesional small bowel obstruction / Ileus	4	2%
Stricture: anastomotic stricture index	3	1%	Hiatal hernia	3	1%
Stricture: before first year	3	1%	Perforation, duodenal	2	1%
Bougienage required	2	1%	Perforation, gastric	2	1%
Stricture: recurrent anastomotic stricture	2	1%	Abdominal evisceration	1	<1%
Dilatation, Complication: clinically-significant aspiration episodes	1	<1%	Pneumatosis of ascending colon	1	<1%
Dilatation: tight stricture (yes or no)	1	<1%	Pyloric stenosis	1	<1%
Esophageal stent erosion	1	<1%	Short bowel syndrome	1	<1%
Peptic stricture	1	<1%	Time between surgery and adhesive bowel obstruction / ileus	1	<1%
Result of dilatation	1	<1%	Gastrostomy-related complication	3	1%
			<u>Complication: absent/present</u>	<u>14</u>	7%
			Number of complications	5	2%
			Other complication (unspecified)	3	1%

	n =	%		n =	%
Histopathologically proven complication	2	1%	Aortic injury	2	1%
Incidence of 30-day Complication	2	1%	Fractures	2	1%
Complication of elongation	1	<1%	Hypotension requiring fluid resuscitation	2	1%
Complication: clinically serious	1	<1%	Left recurrent laryngeal nerve injury	2	1%
Number of complications > Clavien Dindo 3b	1	<1%	Acute otitis media (recurrent)	1	<1%
Short-term complications	1	<1%	Anatomic ridge / shelft	1	<1%
Time to first complication	1	<1%	Diaphragmatic paresis: transient	1	<1%
<u>Graft necrosis / Graft failure / Graft loss</u>	<u>15</u>	7%	Functional outcome shoulder	1	<1%
Perforation jejunal graft	3	1%	Hypertension	1	<1%
Degree of conduit tortuosity	1	<1%	Phrenic nerve injury	1	<1%
DVT / thrombosis / trombosis / VTE	5	2%	Sliding hernia	1	<1%
Cardiac arrest	1	<1%	<u>Recurrent fistula</u>	<u>84</u>	4<1%
Cardiac complication	1	<1%	Anastomotic fistula	2	1%
Pericardial effusion	1	<1%	rTEF: number recurrences of the TEF	1	<1%
Anemia	2	1%	<u>Gastrooesophageal reflux</u>	<u>110</u>	53%
Hemodynamic instability, postoperative	1	<1%	<u>Anti-reflux surgery: yes or no</u>	<u>88</u>	42%
<u>Sepsis</u>	<u>43</u>	21%	Dysplasia, esophageal or gastric	5	2%
Shock (also septic shock)	4	2%	Esophagitis, peptic	4	2%
Infections	3	1%	Esophageal carcinoma / cancer	2	1%
Urinary tract infection	3	1%	Fundoplication: time between EA repair and fundoplication	2	1%
<u>Complication (intra-operative)</u>	<u>11</u>	5%	Longest reflux time (min)	1	<1%
Mediastinitis	6	3%	Reflux index	1	<1%
Elongation: mediastinitis	1	<1%	Reflux with normal biopsy	1	<1%
Mediastinal abcess	1	<1%	Reflux: number of nonacid reflux per day	1	<1%
Mediastinal emphysema	1	<1%	Reflux: number of reflux >5min per hour	1	<1%
Mediastinitis after esophageal elongation	1	<1%	Reflux: number of reflux per hour	1	<1%
Intracranial hemorrhage (also preoperative)	8	4%	Renal complication	1	<1%
Bilirubin encephalopathy	1	<1%	<u>Respiratory complication / symptoms / chronic disease</u>	<u>60</u>	29%
Complication: neurologic	1	<1%	<u>Pneumothorax</u>	<u>36</u>	17%
<u>Blood loss</u>	<u>22</u>	11%	<u>Chylothorax</u>	<u>19</u>	9%
Multi-system organ failure	5	2%	Respiratory failure	6	3%
Horners syndrome (transient)	3	1%	Aspiration syndrome / Aspiration	4	2%
Aberrant subclavian artery fistula	2	1%	Pleural effusion	3	1%

	n =	%
Diverticulum of the trachea	2	1%
Empyema	2	1%
Aspiration: recurrent foreign body	1	<1%
Lung collapse (postoperative)	1	<1%
Pulmonary hemorrhage	1	<1%
Respiratory arrest	1	<1%
Jejunostomy problems	2	1%
Stoma Complication	1	<1%
<u>Tracheomalacia</u>	<u>63</u>	3<1%
<u>Vocal cord issues / Voice changing</u>	<u>28</u>	13%
Severity of tracheomalacia	1	<1%

	n =	%
Tracheal narrowing	1	<1%
Tracheomalacia: presenting signs and symptoms	1	<1%
<u>Aortopexy: yes or no</u>	<u>33</u>	16%
Aortopexy: additional procedures to open airway	2	1%
Tracheomalacia: time between EA repair and first symptoms	1	<1%
Tracheopexy (performed, yes or no)	1	<1%
Complication related to re-intubation	1	<1%
Ventilation: intraoperative ventilatory Complication	1	<1%

### Readmission and reoperation

	n =	%
<u>Readmission</u>	<u>16</u>	8%
Anastomotic leakage: reoperation	1	<1%
Foker: reoperation due to leak while on traction	1	<1%

	n =	%
<u>Reoperation (esophageal)</u>	<u>80</u>	38%
Time between primary repair and reoperation	1	<1%

### Growth

	n =	%
Undernourished / Undernutrition / Malnourished / Malnutrition	4	2%
Nutritional status at 1y	2	1%
<u>Growth / Weight / Failure to thrive</u>	<u>61</u>	29%
Weight at time of filling in questionnaire/intervention/time of follow-up	8	4%

	n =	%
Height at time of filling in questionnaire / intervention / time of follow-up	3	1%
Perioperative weight change	1	<1%

### Morbidity

	n =	%
Diameter of anastomotic opening (cm)	4	2%
Stricture: acute food bolus impaction	1	<1%
Behavior	3	1%
Parent-reported attention deficit hyperactivity disorder	1	<1%
Parent-reported autism spectrum disorder	1	<1%

	n =	%
Intelligence (Wechsler Intelligence Scale for children (WISC-III-NL))	2	1%
Start school education at time of questionnaire/intervention/follow-up	2	1%
Academic skills (WRAT-4)	1	<1%
Attention/language (WPPSI   NEPSY-II / TEA-Ch)	1	<1%
Cognitive problems	1	<1%
Daily executive functioning (proxy-reported)	1	<1%

	n =	%		n =	%
Executive functioning (NEPSY-II)	1	<1%	<u>Gastro-intestinal symptoms</u>	23	11%
Intellectual functioning (WPPSI-III/WISC)	1	<1%	Endoscopy: gastric and esophageal metaplasia	8	4%
Intellectual levels (ADSI scale)	1	<1%	Delayed emptying of the stomach	6	3%
Memory (CMS)	1	<1%	Dumping	6	3%
Revised Amsterdam Intelligence Test (RAKIT)	1	<1%	Gastroesophageal dysmotility	3	1%
<u>Motor/neuro- development</u>	13	6%	Choking	2	1%
<u>Dysphagia / Swallowing difficulties</u>	52	25%	Gastric Outlet Obstruction	2	1%
Eosinophilic esophagitis	10	5%	Gastric retention	1	<1%
Bolus impactions requiring endoscopic intervention	5	2%	Gastrointestinal symptoms (Gastrointestinal Symptom Rating Scale, GSRS)	1	<1%
Choking: number of episodes a week	1	<1%	Gastrostomy tube at 1 year	1	<1%
Delayed emptying of the esophagus	1	<1%	Gastrostomy tube at discharge	1	<1%
Endoscopy: active inflammation	1	<1%	Delayed transit of jejunal conduit	1	<1%
Esophagitis, erosive / ulcerative	1	<1%	Health status (German Health Survey for Children and Adolescents (KiGGS))	2	1%
Tolerated level of food texture (International Dysphagia Diet Standardization Initiative (IDDSI))	1	<1%	Mental and psychosocial health (SDQ-20)	1	<1%
<u>Oral feeding issues</u>	61	29%	Laryngomalacia	3	1%
Functional intake	7	3%	Mannitol challenge test: positive result	1	<1%
All oral intake at 1 year	2	1%	Metacholine challenge test: bronchial hyperresponsiveness	1	<1%
Age when eating seemed to be enjoyed	1	<1%	Spirometry: reduced respiratory capacity	1	<1%
Age when eating solid food was properly managed	1	<1%	Coping (Coping Strategy Checklist)	1	<1%
Chewing function (Karaduman Chewing Performance Scale (KCPS))	1	<1%	Traumatic stress (IES-13)	1	<1%
Consuming blenderized and mashed food	1	<1%	<u>Chest wall deformity</u>	19	9%
Diet habits	1	<1%	Dental problems	1	<1%
Eating outcomes at 3 years of age	1	<1%	Erosive teeth wear	1	<1%
Mealtime behaviors	1	<1%	Halitosis	1	<1%
Nutrient delivery	1	<1%	Oral hygiene	1	<1%
Parental anxiety during mealtime	1	<1%	Elevated / Winged scapula	4	2%
Penetration and aspiration (Penetration and Aspiration Scale)	1	<1%	Subglottic stenosis (postoperative)	4	2%
Reliant on tube feeding to some degree at one year	1	<1%	BRUEs	2	1%
			Sleep disorders	2	1%
			Depression	1	<1%
			Horner syndrome	1	<1%

	n =	%		n =	%
Exercise capacity / intolerance (Bruce protocol)	4	2%	Tracheal vascular compression	2	1%
Participation in sports	1	<1%	Aspiration risk	1	<1%
<u>Quality of Life</u>	<u>16</u>	8%	Blind ending fistula stump on fiberbronchoscopy	1	<1%
<u>Pneumonia</u>	<u>57</u>	27%	Bronchitis	1	<1%
<u>Recurrent respiratory infections</u>	<u>38</u>	18%	Mosaic perfusion on chest CT	1	<1%
ALTE / Cyanotic spells / Cyanosis	7	3%	Pneumonia: number of episodes	1	<1%
Spirometry: obstructive ventilatory impairment	5	2%	Pneumonitis	1	<1%
Bronchiectasis	4	2%	Prolonged respiratory infection	1	<1%
Lung atelectasis	4	2%	Purulent secretion in main bronchi on fiberbronchoscopy	1	<1%
Spirometry: restrictive ventilatory defect / impairment	4	2%	Respiratory tract infections: number of hospitalizations	1	<1%
Bronchomalacia	3	1%	Social maturity ((Modified) Vineland Social Maturity Scale)	2	1%
Hospital admission for respiratory tract infections	3	1%	Gastrojejunostomytube at 1 year	1	<1%
Spirometry: reversible airways obstruction	3	1%	Amount of psychological stress for patients and their families	1	<1%
Asthma	2	1%	Maternal stress (Parental stress scale)	1	<1%
Chest infections	2	1%			
Pulmonary function impairment (obstructive and / or restrictive and / or impaired DLCO)	2	1%			

## Mortality

	n =	%		n =	%
<u>Mortality</u>	<u>137</u>	66%	Adolescents knowledge of care and transition	1	<1%
Survival	6	3%	Parents' transition-specific knowledge	1	<1%
Age at death	3	1%			
In-hospital death	1	<1%			
Time between surgery and death	1	<1%			

## Other

	n =	%		n =	%
Cosmetic Result	6	3%	Returning the Moro reflex (1m, 3m)	1	<1%
Hospital expenses / Cost	4	2%	Number of medical appliances needed after discharge (feeding, respiratory, stoma)	1	<1%
Feeding at discharge (oral versus NJT or combination or parenteral)	8	4%	Adolescents commitment to own care (Patient Activation Measure-13D )	1	<1%
Inability to be fed orally by end of the first month	1	<1%	Patient satisfaction with transition programme (ZUF-8)	1	<1%
Time to shoulder function recovery (days)	1	<1%			