



Perioperative management and outcomes in long-gap esophageal atresia: A retrospective analysis from the Eastern Pediatric Surgery Network ☆☆☆★



Christine Finck^{a,*}, Benjamin Zendejas^b, J. Leslie Knod^a, Peter J. Tramontozzi^c,
Dana Jungbauer^d, Megan Anderson^d, William Middlesworth^e, Stefan Scholz^f,
Shaun M. Kunisaki^g

^a Division of Pediatric Surgery, Connecticut Children's, 282 Washington Street, Hartford, CT 06106, United States

^b Department of Surgery, Boston Children's Hospital, 300 Longwood Ave, Boston, MA 02115, United States

^c MD Program, University of Connecticut School of Medicine, UConn John Dempsey Hospital, 100 Hospital Drive, Farmington, CT 06030, United States

^d Division of Research Operations and Development, Connecticut Children's Medical Center, 282 Washington Street, Hartford, CT 06106, United States

^e Division of Pediatric Surgery, Columbia Irving Medical Center, 622 W 168th St, New York, NY 10032, United States

^f Division of Pediatric General and Thoracic Surgery, UPMC Children's Hospital of Pittsburgh, One Children's Hospital Drive, 4401 Penn Ave, Pittsburgh, PA 15224, United States

^g Department of Surgery, Johns Hopkins University School of Medicine, 1800 Orleans Street, Suite 7353, Baltimore, MD, United States

ARTICLE INFO

Keywords:

Esophageal atresia
Eastern Pediatric Surgery Network
Long gap esophageal atresia

ABSTRACT

Background: Less than 10% of infants born with esophageal atresia (EA) have a long gap that precludes primary anastomosis at birth. The purpose of this study was to evaluate the diagnosis, management, and outcomes of infants with long-gap esophageal atresia (LGEA) within a regional consortium of children's hospitals.

Methods: After IRB approval, a multicenter, retrospective cohort study was conducted of LGEA patients managed by 13 member institutions between 2009 and 2018. LGEA was defined as Type A or B esophageal atresia with the inability to perform a primary esophageal anastomosis at birth based on preoperative imaging or operating findings. Study outcomes, including operative repair, postoperative outcomes, and complications, were collected to detect significant associations between variables ($p < 0.05$).

Results: There were 62 LGEA patients identified, including 50 (81%) with Type A and twelve (19%) with Type B esophageal atresia. Most (77%) were diagnosed prenatally with 98% undergoing a gastrostomy before attempted EA repair. The mean gap length at repair was 3.24 ± 1.59 cm ($n = 21$). Most (95%) were managed with delayed repair (median age at repair of 96 days (IQR: 67.5-131)), and 22 (35%) underwent an esophageal traction-induced lengthening process. Two (3.2%) required esophageal replacement. There was no significant difference in complications between different treatment strategies.

Conclusions: In this collaborative descriptive study of 62 infants with LGEA, delayed primary repair with or without traction was the preferred approach, with outcomes that were comparable between strategies, and with high rates of esophageal preservation. We encountered variability in gap length measurement and reporting. This study highlights a critical need for a prospective, multi-institutional registry with uniform care pathways to help aid in the development of evidence-based guidelines for these challenging patients.

Abbreviations: EA, esophageal atresia; EPSN, Eastern Pediatric Surgery Network; FOIS, functional oral intake score; INoEA, International Network of Esophageal Atresia; IRB, institutional review board; LGEA, long-gap esophageal atresia; REDCap, Research Electronic Data Capture; VACTERL, vertebral anorectal cardiac tracheoesophageal fistula renal and limb.

☆ Statement of competing interests: There are no competing interests

☆☆ Previous Communication: Parts of these data were previously presented at a poster at the 2021 American Academy of Pediatrics Annual Meeting, Philadelphia, PA (A Multi-center retrospective study of long gap esophageal atresia: Report of the EPSN Consortium).

* Financial Support Statement: None

* Corresponding author.

E-mail address: cfinck@connecticutchildrens.org (C. Finck).

<https://doi.org/10.1016/j.yjpso.2023.100029>

Received 30 May 2023; Accepted 31 May 2023

Available online 15 June 2023

2949-7116/© 2023 The Author(s). Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license

(<http://creativecommons.org/licenses/by-nc-nd/4.0/>)

1. Introduction

The most common congenital anomaly of the esophagus is esophageal atresia (EA). This occurs in 1/4000 live births, and long-gap esophageal atresia (LGEA) accounts for roughly 4-10% of all cases [1–3]. Several studies have defined EA as long gap when primary repair is not possible, whether perceived or real [4]. A more restrictive definition of LGEA can be considered Gross type A or type B [5]. In 2017, the International Network of Esophageal Atresia (INoEA) working group defined a long gap as any esophageal atresia that lacks intra-abdominal air on radiographs suggesting a Gross Type A or B configuration [6,7]. Further, they surmised that other types may not fit this definition, but if the esophageal ends cannot surgically be brought together, the patient should be referred to a center with experience in managing LGEA [6,7].

The optimal diagnosis and management strategy for many LGEA patients remains controversial. At most institutions in North America and elsewhere, the diagnosis and management of LGEA have been based on surgeon training or experience rather than on evidence-based guidelines [7]. Surgical options available for the treatment of LGEA include (1) delaying the repair while expecting spontaneous esophageal growth, (2) replacing the esophagus with stomach, colon, or jejunum, and, more recently, (3) pursuing tension-induced lengthening of the native esophageal tissue to facilitate delayed primary repair [8–15]. A newer, innovative approach, the magnamosis technique, utilizes magnets to bring the ends of the esophagus together [16]. Single institution reports have shown a wide spectrum of clinical outcomes with each of these approaches and may be biased by the approach adopted by that institution. Moreover, drawing meaningful conclusions from systematic reviews and meta-analyses has been challenging due to significant heterogeneity in the definition, diagnosis, and follow up of LGEA patients [6,10].

Given the paucity of multicenter studies with substantial numbers of LGEA patients [2,3] we sought to understand current practice patterns and clinical outcomes of LGEA patients within a regional consortium of tertiary care children's hospitals. Our group hypothesized that delayed primary repair, with or without traction procedures, would be the most common approach in the management of LGEA cases.

2. Methods

2.1. Study design and eligibility criteria

Institutional review board (IRB) approval was obtained from 13 member children's hospitals associated with the Eastern Pediatric Surgery Network (EPSN, www.easternpediatricsurgery.org). A retrospective cohort study was conducted based on an operative registry of infants with LGEA managed between December 1, 2009 and December 31, 2018. LGEA cases were identified at each center based on review of all esophageal atresia cases queried in the electronic medical record by International Disease classification codes (Q39.0-Q39.9). After consensus was established among a subgroup of EPSN surgeons, we defined LGEA as a Gross type A or B esophageal atresia with a long gap length that precluded the ability to perform a primary esophageal anastomosis at birth based on review of preoperative diagnostic studies or neonatal operative reports. Re-operative LGEA patients were excluded.

2.2. Data collection

Charts were reviewed for demographics, comorbidities, and peri-operative management in accordance with a manual of operations developed based on consensus from institutional representatives. This resulted in a database containing over 225 different data field elements organized into four modules (demographic, preoperative, operative, follow up). Operative repair was classified as follows: (1) Delayed primary repair: primary anastomosis after 30 days of age (no traction procedure

Table 1

Demographic and comorbidity information of LGEA patients.

Patient characteristics	N (%)
Sex	
- Male (%)	37 (60%)
- Female (%)	25 (40%)
LGEA type	
- Type A	50 (81%)
- Type B	12 (19%)
Race	
- White	36 (58%)
- Black/African American	8 (13%)
- Asian	2 (3%)
- More than One Race	2 (3%)
- Unknown	11 (18%)
Ethnicity	
- Hispanic or Latino	8 (13%)
- Not Hispanic or Latino	51 (82%)
- Unknown	3 (5%)
Comorbidities	
- Congenital heart defects	28 (45%)
- VACTERL	9 (16%)
- Other	13 (21%)
- Anorectal Malformations	5 (8%)
- Trisomy 21	7 (11%)
- Limb anomalies	2 (3%)
- Duodenal Atresia	3 (5%)

Table 1. Breakdown of patients with long gap esophageal atresia (LGEA). The majority of patients had Type A LGEA and were white and not Hispanic or Latino. Congenital heart defects were the most common comorbidity.

used), (2) Early traction repair: traction repair (e.g., Foker process, internal static) before 30 days of age followed by primary anastomosis, (3) Delayed traction repair: traction utilized with repair after 30 days of age by primary anastomosis, and (4) Esophageal replacement using an autologous conduit (e.g., stomach, colon, jejunum), regardless of whether traction techniques were used. All data were collected and managed using REDCap (Research Electronic Data Capture) electronic data capture tools hosted at Connecticut Children's [17,18].

The Gross classification was used to classify esophageal atresia types and this study focused on types A and B esophageal atresia [5]. Study outcomes included operative repair as well as postoperative outcomes and complications. Oral feeding was evaluated based on the functional oral intake scale [19]. Frequency of complications were evaluated at 30 day intervals up to 90 days and included: (1) esophageal leak which was defined as contrast extravasation on esophagram, (2) esophageal stricture which was defined as a symptomatic luminal narrowing of the esophageal anastomosis managed by dilation [20], and (3) vocal fold movement impairment defined as documented paralysis of the vocal cord secondary to injury to the recurrent laryngeal nerve.

2.3. Statistical analysis

Descriptive and inferential statistics were calculated using SPSS (Version 17.0, SPSS Inc). Independent sample t-tests were used to compare means of continuous variables, and Fisher's exact and Chi-square test were used for categorical variables. Data were presented as means with standard deviations unless indicated otherwise. A p-value < 0.05 was considered statistically significant.

Results

There were 62 patients who met our inclusion criteria for LGEA. The median number of cases contributed per institution was 3 (interquartile range, 2-7) over the nine-year study period. Patient demographics and comorbidity data are shown in Table 1. Fifty (81%) had type A esophageal atresia (no fistula), and 12 (19%) had type B (proximal fistula). Congenital heart defects were reported in 28 (45%) patients

Table 2
Type A/B Management strategy.

Patient characteristics	Type A and B (n = 62)	
Gestational age/birthweight, mean (std.)	34.7 weeks (3.18), 2170.7 (604.5)	
Weight at time of repair, mean (std.)	4944.2 (1979.5)	
Diagnosis of LGEA during prenatal care (%)	47 (77%)	
Gastrostomy Tube Placed Before Repair (%)	61 (98.4%)	
Gap length in cm, mean (std.)	N = 21, 3.24 (1.59)	
Management Type	Type A and B (n = 62)	Mean weight at repair within management group, g (std.)
Delayed without Traction (%)	37 (59.7%)	4849.8 (2099.5)
Delayed with Traction (%)	22 (35.5%)	5056.1 (2068.1)
Non-delayed with Traction (%)	1 (1.6%)	3330.0
Esophageal replacement used (%)	0 (0)	-
Delayed with Traction and Esophageal Replacement (%)	2 (3.2%)	7100.0 (4101.2)

Table 2. Gestational age, birthweight, weight at repair, and management type of patients with LGEA type A/B. The majority of patients were diagnosed with LGEA during prenatal care and had a gastrostomy tube placed before repair. The most common management types was delayed repair without traction followed by delayed repair with traction.

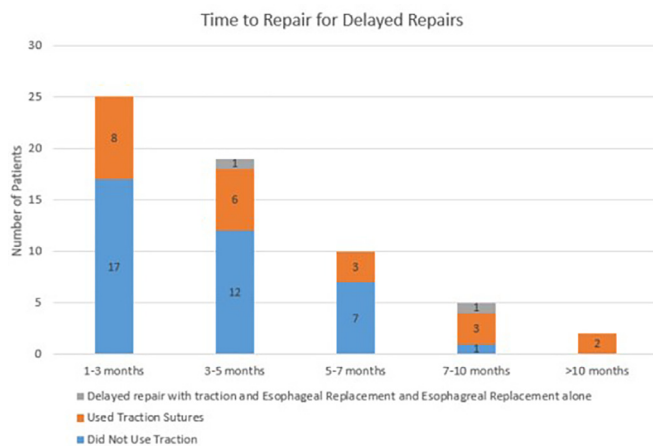


Fig. 1. Schematic of Timing of Delayed Repairs

The timing of repair in patients that had a delay in repair > 1 month. Bars in blue color had a delay in repair without the use of esophageal lengthening techniques; Bars in orange reflect delayed repair with use of esophageal lengthening techniques; Bars in grey reflect delayed repair with the use of esophageal lengthening techniques and/or an esophageal replacement. Most repairs occurred between 1 and 3 months of age.

and VACTERL association, defined by vertebral, anorectal, cardiac, tracheoesophageal fistula, renal and limb anomalies, was noted in 9 (16%) patients.

3.1. Management in type A/B LGEA

Table 2 summarizes the operative repair approaches used in the management of LGEA. Seventy-seven percent of patients were diagnosed with LGEA during prenatal care and 98.4% had a gastrostomy tube placed before repair. The mean birthweight was 2171 ± 605 g and the mean weight at time of repair was 4944.2 ± 1979.5 g. The gap length was reported in 21 (34%) patients and was 3.2 ± 1.6 cm. Thirty-seven (59.7%) patients had delayed repair without traction followed by 22 patients (35.5%) undergoing delayed repair with traction.

In the delayed repair group *without* traction, most ($n = 29$, 47%) were repaired before 5 months of age, with the majority ($n = 17$, 27%) being repaired between 1 and 3 months of age (**Fig. 1** blue bars). The median time to repair was 96 days (IQR: 67.5-131).

Twenty-five (40%) patients underwent esophageal lengthening with either internal static or external dynamic traction as part of the operative

Table 3
Management of LGEA with traction.

Management included Traction	
Average weight at repair	5180 grams (4850 in non-traction repair)
Traction applied (n = 25)	N (%)
- Open thoracotomy	18 (72%)
- Thoracoscopy	5 (20%)
- Unknown	2 (8%)
-	-
Frequency of Traction Manipulation (n = 25)	N (%)
- Daily	3 (12%)
- Every other day	6 (24%)
- Every other week	2 (8%)
- Unknown	14 (56%)
-	-
Separate procedure to readjust traction (n = 12)	N (%)
- Thoracoscopic evaluation and replacement sutures	3 (25%)
- Thoracotomy evaluation and replacement sutures	6 (50%)
- Thoracoscopic converted to open external traction	2 (17%)
- Removal and cervical esophagostomy	1 (8%)
Length of gap at readjustment (n = 4)	Median 2.75cm (IQR 2-3.5 cm)

Table 3. Patients that had dynamic or static esophageal lengthening as part of LGEA repair. Traction was applied in 25 patients with the majority being applied through an open thoracotomy.

management approach (**Table 2**). Twenty-two (35.5%) cases had traction applied in the setting of a delayed primary repair, and one (1.6%) case had early repair (non-delayed with traction). A minimally invasive traction technique was utilized to place sutures in five (20%) traction cases compared to thoracotomy in 18 (72%) cases (**Table 3**). The approach for traction was not specified in two cases. Eleven patients had the frequency of planned traction manipulation recorded. There were three patients where it was recorded as daily, six as every other day and two patients as every other week. The median weight at definitive repair in patients undergoing traction was 4835 gms (IQR: 3800-6200). Median time to repair in traction group is 126 days (IQR: 83-221)

In the one patient that underwent non-delayed repair with traction, the traction placed on day of life 9 with definitive repair on day 24. The weight at time of repair for this patient was 3330 g.

Two patients underwent esophageal replacements with stomach after failed attempts at traction induced esophageal lengthening – both with the stomach being pulled up into the chest. The mean age at repair for these patients was 174 days with their average weight at time

Table 4
Comparison of outcomes in the type A/B LGEA.

	Delayed with Traction (n = 22)	Delayed without Traction (n = 37)	Non-delayed with Traction (n = 1)	p-value
Anastomotic Leak	4 (18%)	9 (24%)	0	0.749*
Anastomotic Stricture	15 (68%)	25 (67%)	1 (100%)	1.00
Vocal Cord Injury	1 (4.5%)	1 (3%)	0	1.00*
Anti reflux Procedure	1 (4.5%)	3 (8%)	0	1.00*

*Comparison only including delayed with traction and delayed without traction for anastomotic leak and vocal cord injury.

Table 4. Comparison of outcomes with different management strategies in Type A/B LGEA. Results are reported as aggregates for complications that occurred over 90 days post-repair, for patients with a complication occurring at least once in that timeframe.

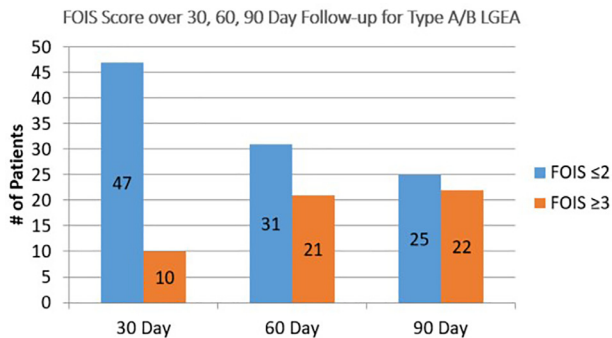


Fig. 2. Functional oral intake scale (FOIS) Scores for LGEA Patients at 30, 60, and 90 Days Follow-Up

Fig. 2. FOIS scores over time in patients with Type A/B LGEA. Blue bars indicate a FOIS of ≤ 2 indicating gastrostomy tube dependent with minimal attempt at foods or liquids by mouth at 30 days, orange bars indicate a FOIS of ≥ 3 indicating consistent oral intake.

of repair being 7100 ± 4101 g. Gap length at time of replacement was recorded in one patient at 3 cm.

3.3. Postoperative complications

A review of complications between management groups over 90 days postoperatively revealed an anastomotic leak in 4 patients (18%) in the delayed *with* traction group and 9 patients (24%) in the delayed *without* traction group (Table 4). Anastomotic stricture was reported in 15 patients (68%) of the delayed *with* traction management group and 25 patients (67%) in the delayed *without* traction group. In the non-delayed with traction patient there was an anastomotic stricture reported within the first 90 days postoperatively (100%). Vocal cord dysfunction was reported in a total of 2 patients in this series. Anti reflux procedures were reported in the first 90 days postoperatively in 4 patients (Table 4)

3.4. Functional oral intake scores

Evaluation of functional oral intake scores (FIOS) at postoperative intervals of 30, 60, and 90 days. At the 30-day interval, the majority (82%) were gastrostomy tube dependent with minimal attempt at foods or liquids by mouth (FOIS of ≤ 2). Yet this decreased to 53% at 90 days. Patients with a FOIS of ≥ 3 , defined as consistent oral intake of foods or liquids, were 18% of patients at 30 days and increased to 40% at 60 days and 47% at 90 days (Fig. 2).

4. Discussion

In rare congenital diseases, collaboration among children's hospitals is required to obtain adequate numbers of patients to understand patient outcomes and to develop evidence-based guidelines. The purpose of this

study was to characterize the diagnosis and management strategies of LGEA among a consortium of U.S. Children's Hospitals to understand current multi-institutional practices. This approach is similar to the recent study on LGEA from Nordic countries which included 71 patients with Type A/B LGEA [3].

The definition and diagnosis of LGEA is highly variable [4,6]. Sixty-two patients met the inclusion criteria for LGEA defined as a Gross Type A or B esophageal atresia with a long gap length that precluded the ability to perform a primary esophageal anastomosis at birth. Like other studies, most of these patients had blind ending esophageal pouches (Type A) [1,4,6]. Our data on prenatal diagnosis supports the literature that Type A/B LGEA is commonly diagnosed prenatally [21,22].

Management of LGEA varies among institutions. Single institutions have reviewed their experience and one study advocated utilizing an algorithmic personalized approach to managing LGEA patients [9] with 36% of patients having a prior attempt at repair, and 49 patients had a primary esophageal tension-induced lengthening procedure (Foker process). The authors concluded that with a customized approach based on the patient's unique anatomic configuration, esophageal preservation is possible in almost all cases [9].

Other authors have discussed replacing the esophagus when the gap is too wide. In a 2017 systematic review, colonic interposition and gastric pull up were the most favorable approaches to esophageal replacement in LGEA [23]. However, current evidence on long-term outcomes is limited by small numbers. Another meta-analysis determined that gastric pull up is associated with higher respiratory morbidity but lower gastrointestinal morbidity when compared to colonic interposition. Jejunal interposition was cited to be a valid technique in a center with experience [10]. In our cohort, only 2 patients had esophageal replacements, all with a prior attempt at traction and utilizing a stomach pull up when the ends were unable to be brought together. This study demonstrates that most patients were treated by delayed repair suggesting that utilizing native esophageal tissue is preferred by surgeons [24,25].

In the study presented here, 98% of patients underwent a delayed approach as a treatment strategy. In this delayed approach, almost 40% used traction sutures to statically or dynamically lengthen or grow the esophagus at some point in the treatment, mostly through an open thoracotomy. Two to three months of age was the most common timeframe of delay for repair. This is consistent with other studies that report delaying until 3 months of age and then performing a repair when there is the least amount of tension possible on the anastomosis [24]. However, other groups aim to begin the traction process when the child is roughly 3 to 3.5kg, regardless of age [9]. Another recent study indicated that chest radiographs can sequentially assess gap length while undergoing external esophageal traction (Foker process) [15]. Alternatively, multi-stage thoracoscopic approaches have been reported [14,26,27] with internal traction sutures including slipknots. The failure rate for esophageal lengthening techniques has recently been reported in a survey in the UK of up to 24% [28]. In our study, 12 patients (19%) had readjustment of their traction sutures, however, we did not capture whether it was planned or due to a complication.

There were no mortalities in this study, signifying advances in perinatal care. Complications such as anastomotic leak (18-24%) and stricture (49-68%) are similar to reports in the current literature on LGEA [8,29]. In this study, Type A/B esophageal atresia patients had a gap length of 3.24cm, which is in line with gap lengths reported in the literature [24]. Unfortunately, due to the limited numbers of gap measurements reported, and the variability in the method of assessment (on or off-tension, dilators, endoscopic, fluoroscopic, etc.) we are not able to draw any meaningful conclusions regarding gap length. Additionally, given the small numbers and short length of follow up, it is difficult to draw conclusions about complication rates between management strategies.

One of the long-term morbidities of EA includes poor feeding and poor growth [30]. A recent prospective case-control cohort study reported that long gap esophageal atresia patients suffer more from digestive morbidity, defined as clinical symptoms of gastroesophageal reflux disease at age 6 years when compared to a cohort of non-long gap EA [28]. Use of a standardized measure such as the FOIS is important in tracking how these children do over time [19]. Additionally, optimizing a regimen of sham feeding before repair has been shown to improve oral motor abilities in children with esophageal atresia [31]. Most children suffering from LGEA have weight for age that is below average [30–36]. Optimizing feeding therapies and tracking FOIS over the continuum will allow for improved management strategies and outcomes. In this study, we noted that most patients had an FOIS score ≤ 2 at the 30-day post-operative interval, and this improved over time. A weakness of our study is that we did not capture other nutritional variables such as growth scores (weight for age) or duration of parenteral nutrition [30]. Our future prospective database will aim to capture these variables.

Finally, in this study, an anti-reflux procedure was performed in 4 patients (6%) within 90 days of the procedure. Reflux was reported in 50-65% of patients with 55-80% of patients being placed on anti-acid medications, despite the recent suggestion that anti-acid medications may not influence stricture formation [37]. One drawback in this study is that reflux can be clinically diagnosed without concrete measurement with things like a pH probe. It is possible that most patients are placed on anti-acid medications for other reasons such as prevention of esophagitis, which is known to be prevalent in this patient population, regardless of symptoms [32].

There are several limitations of this study worth noting. This was a retrospective and largely descriptive study. There were multiple challenges in the data collection process due to variability among institutions in data reporting and lack of clearly defined definitions for diagnosing and managing LGEA. We have already initiated a prospective LGEA database where there will be a hard stop built in to ensure accurate recording as well as the review of the operative report by a central data monitoring group similar to Children's Oncology Group (COG) practice. Standardizing the approach to measure gap length along with the definition of long gap would be instrumental in enabling accurate long-term comparisons. We have suggested adopting an operative report template that includes key elements for LGEA including Gross type, gap length (ideally in both number of vertebral bodies and centimeters) and method of measurement (such as fluoroscopy, Hegar dilators, endoscope, contrast, and whether tension was applied) as well as the approach used for repair. By standardizing our reporting of LGEA, we aim to develop guidelines, a potentially a customized algorithmic approach, to help guide optimal surgical approach for repair as suggested by the Midwest consortium for the management of Type C EA [38].

5. Conclusions

Better evidence to guide initial and long-term management of long gap esophageal atresia is needed. Variability in diagnostic techniques, management approaches and small numbers of patients per center confounds the ability to make logical management guidelines as highlighted in this multi-institutional consortium retrospective review. The results

reported here set the stage for the creation of a prospective multi-institutional registry with uniform care pathways, which can then aid development of evidence-based guidelines for LGEA management. In addition, the wide range in number of patients treated by different institutions suggests that there might be benefit in early referral to centers with multi-disciplinary teams well-versed in various strategies of initial surgical management as well as post-operative and longitudinal EA management (33-36).

Author contributions

Study conception and design: Christine Finck, Benjamin Zendejas, J. Leslie Knod, Peter Tramontozzi, Shaun Kunisaki, William Middlesworth, Stefan Scholz

Data acquisition: Christine Finck, Benjamin Zendejas, J. Leslie Knod, Peter Tramontozzi, Shaun Kunisaki, William Middlesworth, Stefan Scholz

Analysis and data interpretation: Christine Finck, Peter Tramontozzi, Dana Jungbauer, Megan Anderson

Drafting of the manuscript: Christine Finck, Benjamin Zendejas, J. Leslie Knod, Peter Tramontozzi, Dana Jungbauer, Megan Anderson, Shaun Kunisaki, William Middlesworth, Stefan Scholz

Critical revision: Christine Finck, Benjamin Zendejas, J. Leslie Knod, Peter Tramontozzi, Dana Jungbauer, Megan Anderson, Shaun Kunisaki, William Middlesworth, Stefan Scholz

(*): Modified from the authorship requirements of the J Am Coll Surg

Declaration of Competing Interest

There are no conflicts of interest.

Acknowledgments

ESPN is a clinical research collaboration with the following members who also made contributions as authors to this work by instituting and overseeing local site data collection and contributing to the drafting and/or revision of the manuscript: Myron Allukian III, MD; Amina Bhatia, MD; Chase Binion, MD-Candidate; Robert A. Cowles, MD; Rosa Hwang, BS; Saleem Islam, MD, MHP; Harold Leraas, MD; Sean McLean, MD; Jay Meisner, MD; Aaron Lipskar, MD; Valerie Erica Polcz, MD, MS; Oswaldo G. Quevado, MD; Robert T. Russell, MD, MPH; Andres Gonzalez Salazar, MD; Elisabeth Tracy, MD; Lucy Zhang, MD.

The following 13 hospitals contributed data for this study: Boston Children's Hospital, Children's Healthcare of Atlanta, Children's of Alabama, Children's Hospital of Philadelphia, Cohen Children's Medical Center, Connecticut Children's Hospital, Duke Children's Hospital & Health Center, Johns Hopkins Children's Center, Morgan Stanley Children's Hospital, University of North Carolina Children's Hospital, University of Florida Shands Children's Hospital, UPMC Children's Hospital of Pittsburgh, and Yale New Haven Children's Hospital.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at [doi:10.1016/j.yjps.2023.100029](https://doi.org/10.1016/j.yjps.2023.100029).

References

- [1] H.F. Shieh, R.W. Jennings, Long-gap esophageal atresia, *Semin. Pediatr. Surg.* 26 (2017) 72–77 Apr/Epub 2017 Feb 3. PMID: 28550874, doi:10.1053/j.sempedsurg.2017.02.009.
- [2] D.R. Lal, S.K. Gadepalli, C.D. Downard, et al., Perioperative management and outcomes of esophageal atresia and tracheoesophageal fistula, *J. Pediatr. Surg.* 52 (2017) 1245–1251, doi:10.1016/j.jpedsurg.2016.11.046.
- [3] T. Stadil, A. Koivusalo, M. Pakarinen, et al., Surgical repair of long-gap esophageal atresia: a retrospective study comparing the management of long-gap esophageal atresia in the Nordic countries, *J. Pediatr. Surg.* 54 (2019) 423–428, doi:10.1016/j.jpedsurg.2018.07.023.

- [4] D.C. van der Zee, P. Bagolan, C. Faure, et al., Position Paper of INoEA Working Group on Long-Gap Esophageal Atresia: For Better Care, *Front. Pediatr.* 5 (2017) 63, doi:10.3389/fped.2017.00063.
- [5] S. Al-Shanafey, J. Harvey, Long gap esophageal atresia: an Australian experience, *J. Pediatr. Surg.* 43 (2008) 597–601 Apr; PMID: 18405702, doi:10.1016/j.jpedsurg.2007.12.001.
- [6] S. Chittmitrappap, L. Spitz, E.M. Kiely, et al., Oesophageal atresia and associated anomalies, *Arch. Dis. Child.* 64 (1989) 3648.
- [7] C. Dingemann, S. Eaton, G. Aksnes, et al., ERNICA consensus conference on the management of patients with long-gap esophageal atresia: perioperative, surgical, and long-term management, *Eur. J. Pediatr. Surg.* 31 (2021) 214–225 Jun; Epub 2020 Jul 15. PMID: 32668485, doi:10.1055/s-0040-1713932.
- [8] R. Baird, D.R. Lal, R.L. Ricca, et al., Management of long gap esophageal atresia: a systematic review and evidence-based guidelines from the APSA Outcomes and Evidence Based Practice Committee, *J. Pediatr. Surg.* 54 (2019) 675–687 Apr; Epub 2019 Feb 7. PMID: 30853248, doi:10.1016/j.jpedsurg.2018.12.019.
- [9] W.J. Svetanoff, B. Zendejas, K. Hernandez, et al., Contemporary outcomes of the Foker process and evolution of treatment algorithms for long-gap esophageal atresia, *J. Pediatr. Surg.* 56 (2021) 2180–2191 Dec; Epub 2021 Feb 26. PMID: 33766420, doi:10.1016/j.jpedsurg.2021.02.054.
- [10] G. Gallo, S. Zwaveling, H. Groen, et al., Long-gap esophageal atresia: a meta-analysis of jejunal interposition, colon interposition, and gastric pull-up, *Eur. J. Pediatr. Surg.* 22 (2012) 420–425 Dec; Epub 2012 Dec 4. PMID: 23212741, doi:10.1055/s-0032-1331459.
- [11] K. Thompson, B. Zendejas, W.J. Svetanoff, et al., Evolution, lessons learned, and contemporary outcomes of esophageal replacement with jejunum for children, *Surgery* 170 (2021) 114–125 Jul; Epub 2021 Apr 1. PMID: 33812755, doi:10.1016/j.surg.2021.01.036.
- [12] J.M. Firriolo, L.C. Nuzzi, I.M. Ganske, et al., Supercharged jejunal interposition: a reliable esophageal replacement in pediatric patients, *Plast. Reconstr. Surg.* 143 (2019) 1266e–1276e Jun; PMID: 31136495, doi:10.1097/PRS.0000000000005649.
- [13] E. Platt, J. McNally, E. Cusick, Pedicled jejunal interposition for long gap esophageal atresia, *J. Pediatr. Surg.* 54 (2019) 1557–1562 Aug; Epub 2019 Jan 16. PMID: 30717983, doi:10.1016/j.jpedsurg.2018.10.108.
- [14] B. Bogusz, D. Patkowski, S. Gerus, et al., Staged thoracoscopic repair of long-gap esophageal atresia without temporary gastrostomy, *J. Laparoendosc. Adv. Surg. Tech. A* 28 (2018) 1510–1512 Dec; Epub 2018 Jul 17. PMID: 30016196, doi:10.1089/lap.2018.0188.
- [15] A.M. Foust, B. Zendejas, S. Mohammed, et al., Radiographic assessment of traction-induced esophageal growth and traction-related complications of the Foker process for treatment of long-gap esophageal atresia, *Pediatr. Radiol.* 52 (2022) 468–476 Mar; Epub 2021 Nov 30. PMID: 34845501. (clip distance on radiographs), doi:10.1007/s00247-021-05228-z.
- [16] A. Conforti, C. Pellegrino, L. Valfré, C. Iacusso, P.M.S. Schingo, I. Capolupo, S. Sgro', L. Rasmussen, P. Bagolan, Magnamosis for long gap esophageal atresia: Minimally invasive "fatal attraction", *J. Pediatr. Surg.* 58 (3) (2023) 405–411 Mar; Epub 2022 Aug 28. PMID: 36150933, doi:10.1016/j.jpedsurg.2022.08.018.
- [17] P.A. Harris, R. Taylor, R. Thielke, et al., Research electronic data capture (REDCap) – A metadata-driven methodology and workflow process for providing translational research informatics support, *J. Biomed. Inform.* 42 (2009) 377–381 Apr.
- [18] P.A. Harris, R. Taylor, B.L. Minor, et al., REDCap Consortium, The REDCap consortium: Building an international community of software partners, *J. Biomed. Inform.* 9 (2019) May; doi:10.1016/j.jbi.2019.103208.
- [19] Y.G. Yi, H.I. Shin, Psychometrics of the functional oral intake scale for infants, *Front. Pediatr.* 7 (2019) 156 Apr 18.
- [20] R. Tambucci, G. Angelino, P. De Angelis, et al., Anastomotic strictures after esophageal atresia repair: incidence, investigations, and management, including treatment of refractory and recurrent strictures, *Front. Pediatr.* 5 (2017) 120 May 29; PMID: 28611969; PMCID: PMC5447026, doi:10.3389/fped.2017.00120.
- [21] C. Garabedian, R. Sfeir, C. Langlois, et al., French Network on Esophageal Atresia. Does prenatal diagnosis modify neonatal treatment and early outcome of children with esophageal atresia? *Am. J. Obstet. Gynecol.* 212 (2015) 340 Mare1-7; Epub 2014 Sep 28. PMID: 25265404, doi:10.1016/j.ajog.2014.09.030.
- [22] S.M. Kunisaki, S.W. Bruch, R.B. Hirschl, et al., The diagnosis of fetal esophageal atresia and its implications on perinatal outcome, *Pediatr. Surg. Int.* 30 (2014) 971–977 Oct; Epub 2014 Jul 24. PMID: 25056797, doi:10.1007/s00383-014-3562-2.
- [23] J. Liu, Y. Yang, C. Zheng, et al., Surgical outcomes of different approaches to esophageal replacement in long-gap esophageal atresia: a systematic review, *Medicine* 96 (2017) e6942 (Baltimore). May; PMID: 28538385.
- [24] D.H. Oliver, S. Martin, D.I. Belkis, et al., Favorable outcome of electively delayed elongation procedure in long-gap esophageal atresia, *Front. Surg.* 8 (2021) 701609 Jul 6; PMID: 34295918.
- [25] P. Bagolan, L. Valfre, F. Morini, et al., Long-gap esophageal atresia: traction-growth and anastomosis-before and beyond, *Dis. Esophagus* 26 (2013) 372–379 May-Jun; PMID: 23679026.
- [26] D.C. Van der Zee, G. Gallo, S.H. Tytgat, Thoracoscopic traction technique in long gap esophageal atresia: entering a new era, *Surg. Endosc.* 29 (2015) 3324–3330 Nov; PMID: 25669641.
- [27] T. Tainaka, H. Uchida, A. Tanano, et al., Two-stage thoracoscopic repair of long-gap esophageal atresia using internal traction is safe and feasible, *J. Laparoendosc. Adv. Surg. Tech. A* (2017) 71–75 Jan; 27; PMID: 27792528.
- [28] K. Brennan, P. Cullis, I. Yardley, Children's Upper Gastrointestinal Surgery (ChUGS) Network. Oesophageal lengthening by traction in oesophageal atresia: The UK experience, *J. Pediatr. Surg.* 57 (2022) 187–191, doi:10.1016/j.jpedsurg.2021.10.039.
- [29] A. Bourg, F. Gottrand, B. Parmentier, Outcome of long gap esophageal atresia at 6 years: a prospective case control cohort study, *J. Pediatr. Surg.* (2022), doi:10.1016/j.jpedsurg.2022.07.023.
- [30] A.W. Harrington, J. Riebold, K. Hernandez, et al., Nutrition delivery and growth outcomes in infants with long-gap esophageal atresia who undergo the Foker process, *J. Pediatr. Surg.* 56 (2021) 2139 Dec; Epub 2021 Jul 24. PMID: 34366132, doi:10.1016/j.jpedsurg.2021.07.014.
- [31] T. Soyer, S.S. Arslan, Ö. Boybeyi, et al., The Role of Oral Feeding Time and Sham Feeding on Oropharyngeal Swallowing Functions in Children with Esophageal Atresia, *Dysphagia* 38 (1) (2023) 247–252, doi:10.1007/s00455-022-10461-1.
- [32] U. Krishnan, H. Mousa, L.D. Oglio, et al., ESPGHAN-NASPGHAN guidelines for the evaluation and treatment of gastrointestinal and nutritional complications in children with esophageal atresia-tracheoesophageal fistula, *JPGN* 63 (2016) 550–570.
- [33] S. Sømme, N. Shahi, L. McLeod, 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, *Pediatr. Surg. Int.* 35 (2019) 1293–1300, doi:10.1007/s00383-019-04525-x.
- [34] D. Dylkowski, S. Dave, J. Andrew McClure, et al., Repair of congenital esophageal atresia with tracheoesophageal fistula repair in Ontario over the last 20 years: volume and outcomes, *J. Pediatr. Surg.* 53 (2018) 925–928, doi:10.1016/j.jpedsurg.2018.02.019.
- [35] A.E. Lawrence, P.C. Minnici, K.J. Deans, et al., Relationships between hospital and surgeon operative volumes and outcomes of esophageal atresia/tracheoesophageal fistula repair, *J. Pediatr. Surg.* 54 (2019) 44–49, doi:10.1016/j.jpedsurg.2018.10.037.
- [36] S. Bairdain, T.E. Hamilton, C.J. Smithers, et al., Foker process for the correction of long gap esophageal atresia: primary treatment versus secondary treatment after prior esophageal surgery, *J. Pediatr. Surg.* 50 (2015) 933–937, doi:10.1016/j.jpedsurg.2015.03.010.
- [37] L. Dr, S.K. Gadepalli, C.D. Downard, et al., Midwest Pediatric Surgery Consortium. Challenging surgical dogma in the management of proximal esophageal atresia with distal tracheoesophageal fistula: Outcomes from the Midwest Pediatric Surgery Consortium, *J. Pediatr. Surg.* 53 (2018) 1267–1272 Jul; PMID: 28599967.
- [38] C.M. Bence, B. Rymeski, S. Gadepalli, et al., Clinical outcomes following implementation of a management bundle for esophageal atresia with distal tracheoesophageal fistula, *J. Pediatr. Surg.* 56 (2021) 47–54, doi:10.1016/j.jpedsurg.2020.09.049.